Perfusion for Congenital Heart Surgery

Notes on Cardiopulmonary Bypass for a Complex Patient Population

Gregory S. Matte

WILEY Blackwell

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To Bridget, Nicholas, and Justin for their love, support, and regular reminders that we are all lifelong students.

Contents

Foreword

The art and science of providing perfusion for patients undergoing surgical correction of congenital heart lesions has advanced rapidly in the past decade. The complex equipment; the unique acid–base management strategies; the specialized perfusion and ultrafiltration techniques; the wide variation in patient's age, size, and vulnerability to physiologic trespass; and the wide variety of surgical procedures performed set perfusion for congenital heart surgery apart from that provided for correction of acquired heart disease in adults. Consequently, provision of cardiopulmonary support for repair of congenital heart lesions has become a specialty unto itself.

There is no doubt that perfusionists caring for patients with congenital heart disease will find this manual invaluable. More importantly, from the perspective of a pediatric anesthesiologist and intensivist, this manual will be an essential resource for cardiac anesthesiologists and intensivists. The practical hands-on information contained herein is not currently readily available in any other publication. This manual will and should become part of the teaching curriculum for anesthesia and intensive care residents and fellows involved in the care of these complex patients.

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Preface

There are numerous excellent textbooks available today on congenital cardiac disease. Most congenital cardiac perfusion services have at their disposal updated texts regarding the disciplines of cardiology, cardiac surgery, anesthesia, nursing, and perfusion. The perfusion texts, of which I am most interested, can be found to have both vague and contradictory statements regarding how to actually "run the pump" for congenital cardiac cases. It is my hope that this book provides some direction. The aim of this book is not to provide a comprehensive textbook for congenital cardiac surgery or cardiopulmonary bypass. Nor is it to simply publish clinical practice protocols. Rather, the aim is to provide easily referenced information and reminders to the pediatric perfusionist and non-perfusionist alike, which can influence a bypass plan and perhaps become part of one's practice. The pediatric perfusionist, with at least a general understanding of the other disciplines involved with cardiac surgery, should be able to reference this book with its provided notes, as I prefer to call them, to confidently devise a plan for a pump run.

The idea of creating this book stems from over 17 years of practicing perfusion for congenital heart surgery and nearly 25 years of working with critically ill children. The academic institutions where I have trained and worked regularly host visitors from around the country and world. They include fellows of anesthesia, cardiac surgery, and the intensive care unit, perfusion and nursing students, as well as current practitioners in those fields. The most common visitor question a department receives is, "Can I get a copy of your protocols?" My standard answer is always a qualified "yes." It is quite simple to pass along perfusion protocols and customized tubing pack specifications. Those items are essential and useful. Though, in isolation, they fail to capture much of the thought and consideration put into every cardiopulmonary bypass plan for congenital cardiac cases. It is my hope that this book is a step toward filling this gap in currently available offerings.

Finally, we are all imperfect clinicians, and it has been said that no one can harm (or even kill) a patient faster than a perfusionist (or surgeon). With that being said, may your clinical errors be minor and preferably off cardiopulmonary bypass!

Gregory S. Matte, CCP, LP, FPP

Disclaimer

Perfusionists, by law, provide cardiopulmonary bypass under the supervision of a physician. This book contains information and recommendations that should be sanctioned by the supervising physician. Errors and omissions with the information provided are possible. Clinical practice is constantly evolving. Use of the information within this book is at your own discretion and risk.

Acknowledgments

A special thanks to Willis Gieser, former Chief of Perfusion at Boston Children's Hospital, who accepted my unsolicited phone call, gave me an interview, and hired me many years ago.

I am also deeply indebted to the following individuals who provided feedback regarding the content of this book and/or reviewed chapters: James DiNardo, Frank Pigula, Kirsten Odegard, Chris Baird, Kevin Connor, Robert Howe, Mark Wesley, Robert Groom, Robert Wise, and Gerard Myers.

CHAPTER 1

Equipment for bypass

The cardiopulmonary bypass plan starts with basics of patient height, weight, allergy history, original diagnosis, previous surgeries, and current indications for surgery. The perfusionist must select and assemble an array of equipment matched to the patient's size, expected pump flow rates, and other factors related to diagnosis. The following is an overview of the major components of a bypass circuit. Please refer to *Chapter 6* for equipment considerations in addition to patient size. Figure 1.1 is provided as a reference to basic equipment arranged for cardiopulmonary bypass.

Oxygenators

The contemporary "oxygenator" is actually several integrated items that in addition to the oxygenating membrane may include the arterial line filter (ALF), venous reservoir and filter, cardiotomy filter, and heat exchanger. Figure 1.2 depicts the components of the Terumo CAPIOX FX series of "oxygenators." Figure 1.3 depicts typical components of an oxygenator system.

The oxygenator membrane

- Membrane oxygenators allow for diffusion of gas, oxygen and carbon dioxide most importantly, across a material separating the blood path from the gas flow path (also called the blood and gas phases).
- True membrane oxygenators allow for diffusion of gases through a membrane separating the blood and gas phases (see Figure 1.4). The type and thickness of the membrane, as well as blood and gas flow characteristics on opposing sides, determines overall diffusion rates.
- Microporous membrane oxygenators allow for diffusion of gases through microscopic holes in the membrane material (see Figure 1.4). The gas transfers directly through these micropores and is therefore less impacted by the membrane material. However, blood and gas flow characteristics on opposing sides still impact diffusion capacity.
- The vast majority of oxygenators for cardiopulmonary bypass are microporous membrane oxygenators. True membrane oxygenators have limited applications today including the use for ECMO at some institutions.
- The membrane oxygenator size chosen for a particular patient should be the smallest which will allow for safe perfusion with some degree of functional reserve in case of decreasing efficiency during extended bypass runs or to account for markedly increased pump flows due to aorto-pulmonary connections (MAPCAs, surgical or other central shunts) or significant aortic regurgitation. Increased pump flow rates in these situations may be required to maintain adequate effective systemic perfusion.
- Using an oxygenator above its manufacturer recommended maximum flow rate may increase arterial line GME transmission and is not recommended.
- It is recommended to define the patient BSA and select an oxygenator based on the maximum expected pump flows. It is important to note that for neonates and infants in particular, their relatively higher metabolic requirement may require markedly increased pump flows during normothermic bypass (i.e., rewarming). Flows of 3.0–3.5 L/min/m^2 are not uncommon and should be considered for equipment selection.
- Primary consideration is given to the manufacturer-recommended maximum flow rate that is based on gas

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

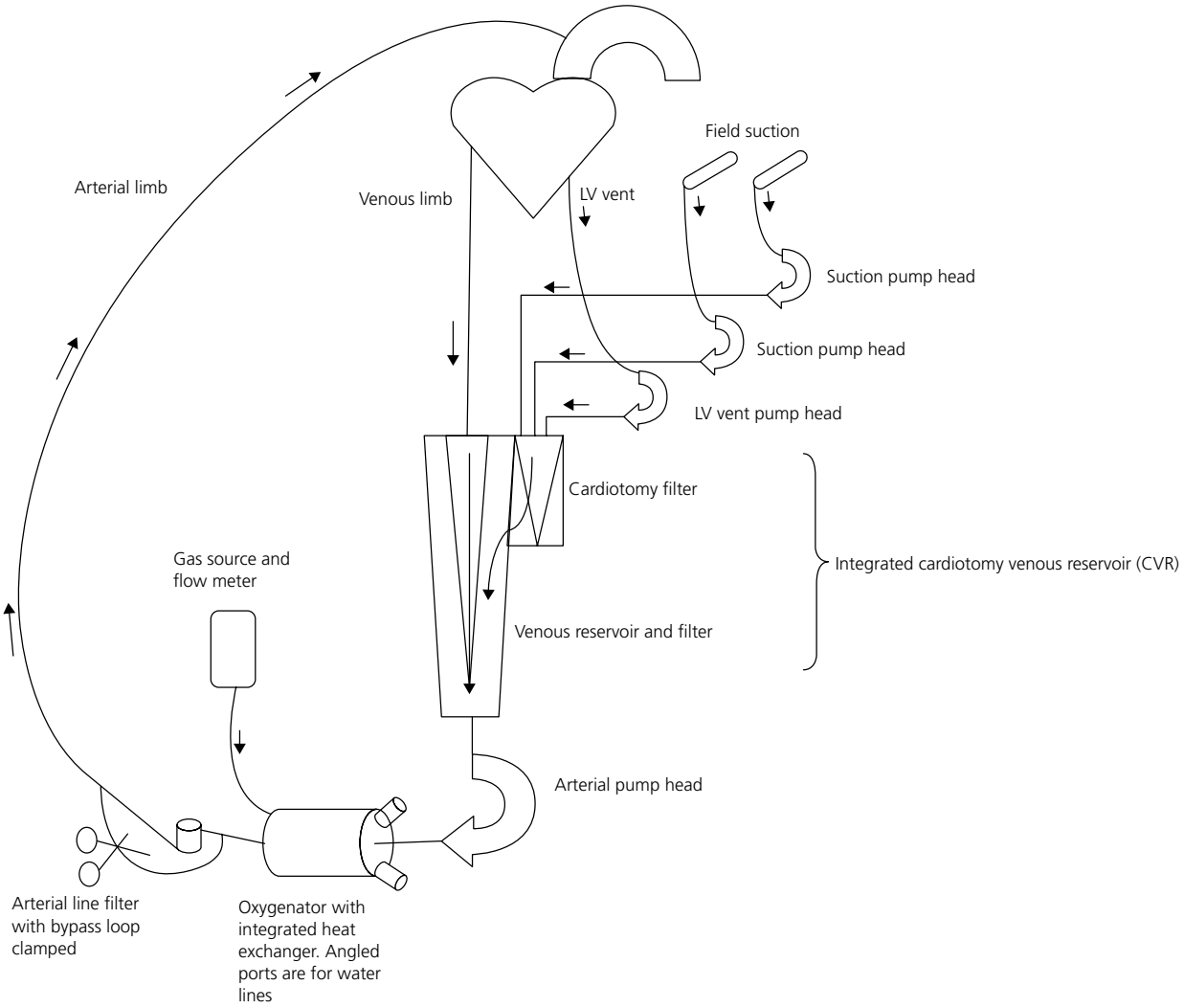


Figure 1.1 Simplified schematic for basic cardiopulmonary bypass equipment (excludes cardioplegia system).

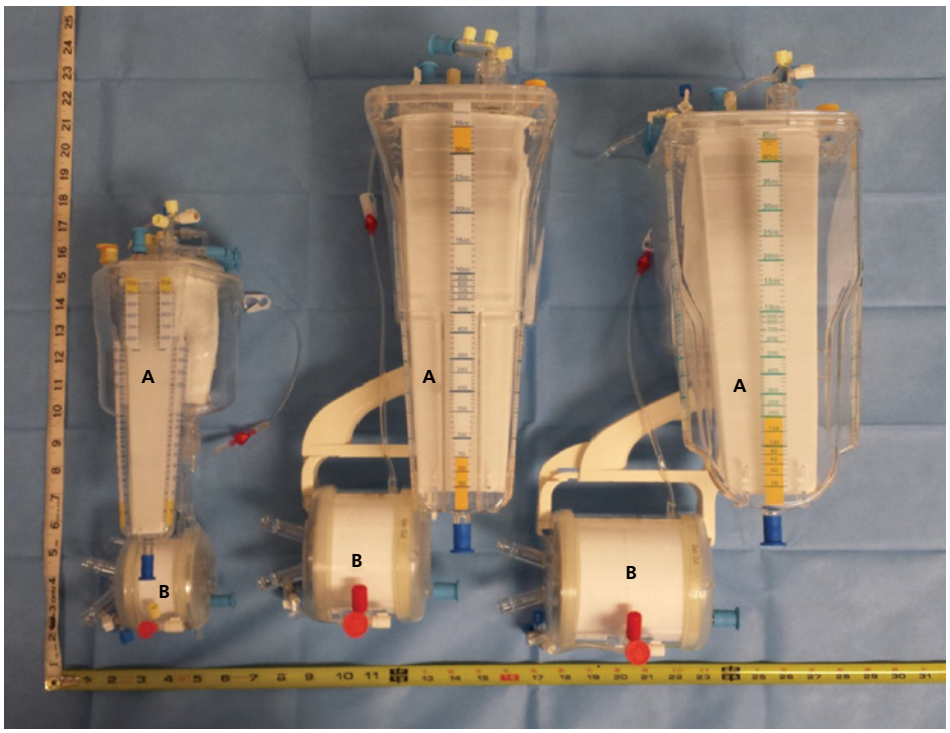


Figure 1.2 Terumo CAPIOX FX series of oxygenators. *Left to right: Terumo CAPIOX FX05, Terumo CAPIOX FX15-30, Terumo CAPIOX FX25. A—cardiotomy venous reservoir and B—oxygenator membrane with integrated arterial line filter and heat exchanger.* (*See insert for color representation of the figure.*)

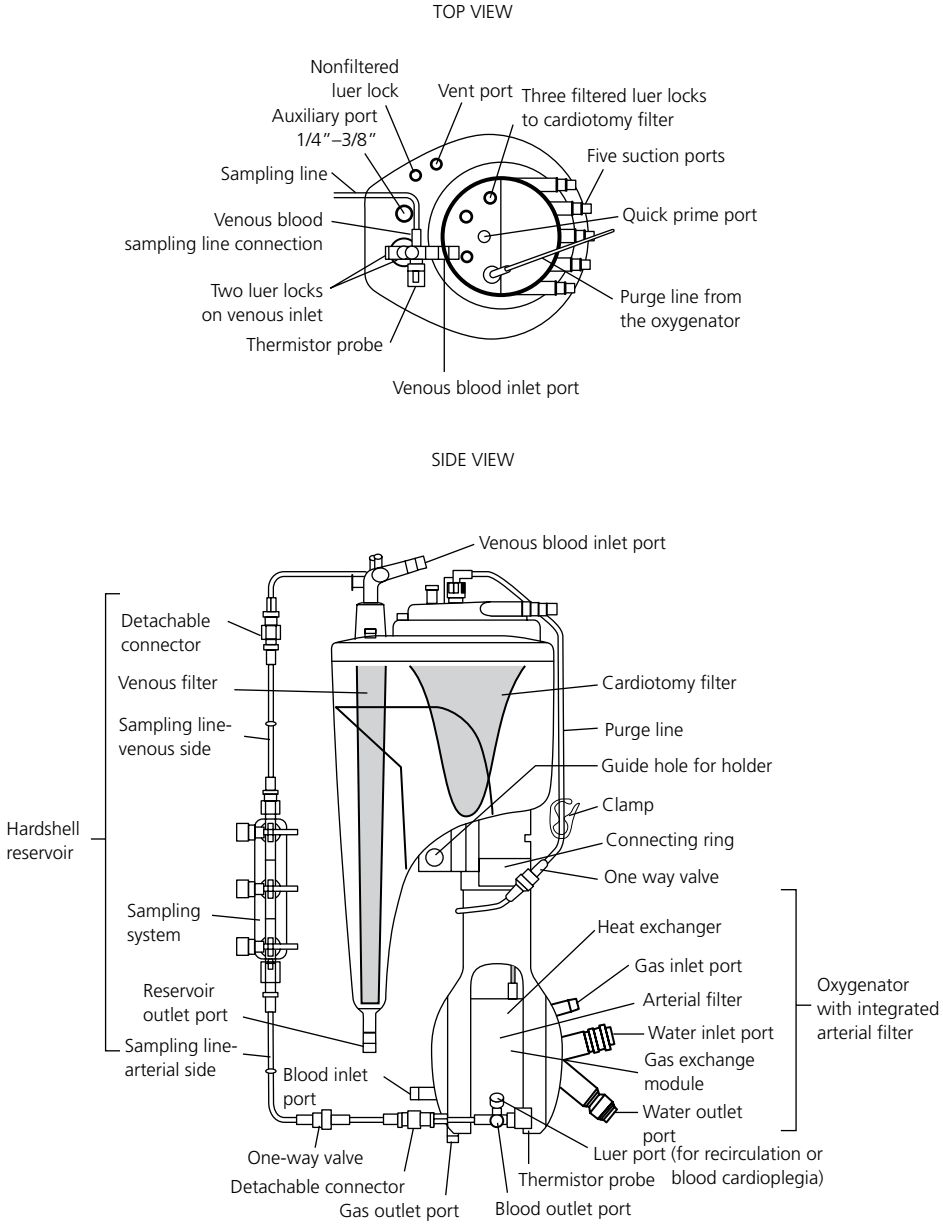
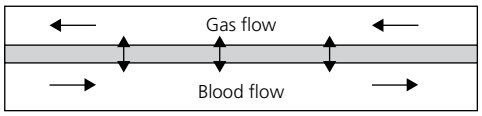
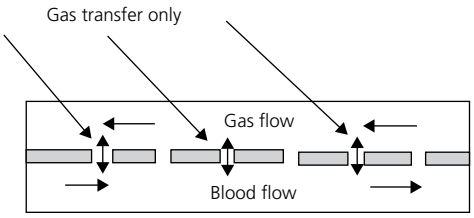


Figure 1.3 Typical components of an oxygenator system. Reproduced with permission from Terumo Cardiovascular Group, Ann Arbor, MI. All rights reserved.

True membrane oxygenators

Gas diffuses through a solid membrane (shown in gray). There are no direct communications between the gas and blood compartments.





Microporous membrane oxygenators

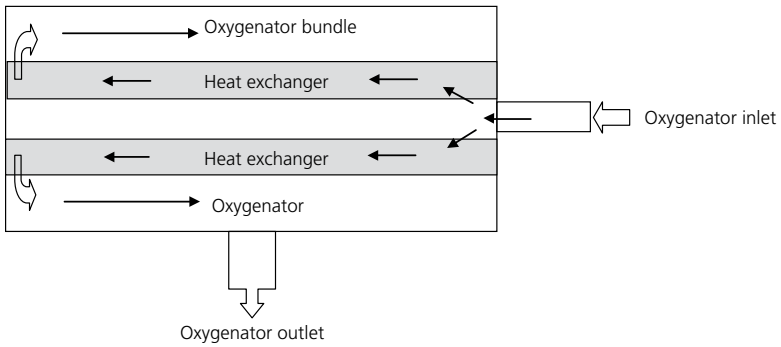
Gases diffuse through tiny holes in the membrane material (shown in gray). There are direct communications between the gas and blood compartments. Microair in the blood compartment may pass into the gas compartment to facilitate removal of gaseous microemboli in the blood. Blood cannot pass.

Figure 1.4 Primary types of oxygenators currently in use.

exchange and other aspects of oxygenator, heat exchanger, and reservoir performance. The American Association of Medical Instrumentation (AAMI) standard reference values are usually not relied solely upon since they may not take into account additional factors that the manufacturer evaluates for overall performance.

- Increased oxygenator bundle size does not linearly relate to performance since characteristics of the blood and gas flow paths vary among devices.
- Oxygenators have either radial or axial blood flow paths that affect performance in competing ways in regards to oxygenating efficiency, pressure drop, microemboli removal, and heat exchanger performance (see Figure 1.5).
- Microporous oxygenator bundles are important in the removal of air from the blood path. Some centers, particularly outside of the United States, deem the microporous oxygenator effective enough at air removal that they do not utilize a standalone ALF.
- One oxygenator on the market, the Medtronic Affinity Fusion, has taken advantage of the air handling capabilities of microporous oxygenators, and unique bundle wrapping technology, to be FDA approved for use as an ALF as well as an oxygenator.
- The perfusionist must be familiar with the *manufacturer* recommendations for treating an oxygenator suspected of "wetting out." These values are listed in Tables 1.1 to 1.4.
- Oxygenators are usually qualified for use by the manufacturer for up to 6 h. Use beyond this limit does occur and most often there is not a significant decrease in performance. However, safe use beyond this limit is not guaranteed. Consideration should be given to changing out an oxygenator after a long case in which an additional bypass run is a serious possibility. Elective change out while off bypass can be accomplished in a controlled manner and can eliminate several concerns of emergently resuming bypass with a product at the end of its rated performance limit.
- Blood proteins coat a membrane oxygenator's surface area, including the micropores through which gas exchange occurs.
- Microporous membranes may experience increased protein coating and subsequent decrease in oxygen transfer during extended bypass runs. It is important to note that this protein coating may also decrease the air handling capabilities of the membrane.
- The pressure drop across an oxygenator membrane is frequently listed as a specification. It may be measured in real time during bypass. A change in this value over time is important to consider during bypass as it can be an indicator of change in function.
- Pressure drop is frequently equated with shear stress where a lower pressure drop is considered beneficial with lower shear stress. That is not always the case since

Axial blood flow path
Blood travels along the axis of the heat exchanger first and then along the axis of the oxygenator bundle before exiting a low point in the device.



Radial blood flow path
Blood enters and radiates through the heat exchanger and oxygenator bundle before exiting a low point in the device. Some oxygenators preferentially have blood exit the heat exchanger top before flowing through the oxygenator bundle.

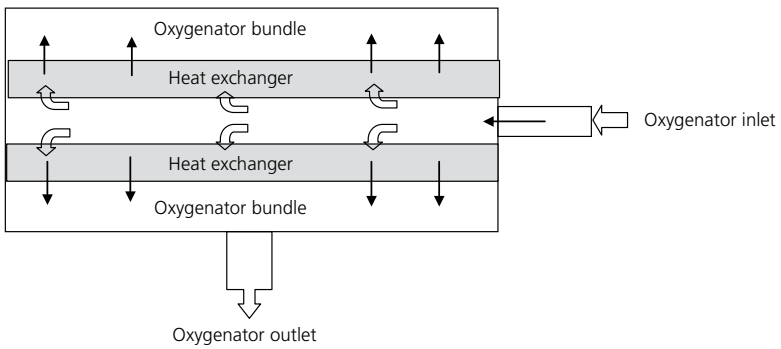


Figure 1.5 Simplified schematic of blood flow paths through an oxygenator.

Table 1.1 Oxygenators rated up to ~2 LPM.

Manufacturer	Oxygenator (microporous polypropylene except as noted)	Oxygenator bundle (m^2)	Oxygenator prime volume	Manufacturer recommended blood flow range (LPM)	Heat exchanger size (cm^2)	Heat exchanger performance factor at manufacturer maximum recommended blood flow rate	Reservoir capacity (mL)	Compatible with vacuum assisted drainage	Integrated arterial filter/pore size/surface area	Minimum operating volume	Gas flow range (LPM)	Maximum temporary gas flow for suspected wetting out of oxygenator
Sorin	D100	0.22	31	Up to 0.7	300	0.65	500	Yes	No	10	Up to 1.4 LPM with a max V/Q of 2:1	Up to 2.8 LPM with a max V/Q of 4:1
Sorin	Liiliput D901	0.34	60	0.8	200	0.72	675	Yes	No	15	Up to 1.6 LPM with a max V/Q of 2:1	None specified
Maquet	Quadrox-i Neonatal	0.38	38	0.2–1.5	700	0.62	800	Yes	No	15	0.1–3.0	None specified
Maquet	Quadrox-i Neonatal with integrated ALF	0.38	40	0.2–1.5	700	0.62	800	Yes	Yes/33 μm/20 cm^2	15	0.1–3.0	None specified
Terumo	Capiox RX05	0.5	43	0.1–1.5	350	0.65	1000	Yes	No	15	0.05–5, minimum 0.2 V/Q	5 LPM for 10s, do not repeat
Terumo	Capiox FX05	0.5	43	0.1–1.5	350	0.65	1000	Yes	Yes/32 μm/130 cm^2	15	0.05–5, minimum 0.2 V/Q	5 LPM for 10s, do not repeat
Sorin	D101	0.61	87	Up to 2.5	600	0.6	1500	Yes	No	30	Up to 5 LPM with a max V/Q of 2:1	Up to 10 LPM with a max V/Q of 4:1
Medtronic	Pixie	0.67	48	0.1–2.0	Not specified	0.65	1200	Yes	No	20	Up to 4.0 LPM with a max V/Q of 2:1	None specified

Table 1.2 Oxygenators rated ~2 to ~5 LPM.

Manufacturer	Oxygenator (microporous polypropylene except as noted)	Oxygenator bundle (m^2)	Oxygenator prime volume	Manufacturer recommended blood flow range (LPM)	Heat exchanger size (cm^2)	Heat exchanger performance factor at manufacturer maximum recommended blood flow rate	Reservoir capacity (mL)	Compatible with vacuum assisted drainage	Integrated arterial filter/pore size/ surface area	Minimum operating volume	Gas flow range	Maximum temporary gas flow for suspected wetting out of oxygenator
Sorin	Lilliput D902	0.64	105	Up to 2.3	200	0.48	1800	Yes	No	200	Up to 4.6 LPM with a max V/Q of 2:1	None specified
Maquet	Quadrox-i Pediatric	0.8	81	0.2–2.8	1500	0.62	1700	Yes	No	30	0.1–5.6	None specified
Maquet	Quadrox-i Pediatric with integrated ALF	0.8	99	0.2–2.8	1500	0.62	1700	Yes	Yes/33 μm/55 cm^2	30	0.1–5.6	None specified
Maquet	Quadrox-iD Pediatric (polymethylpentene)	0.8	81	0.2–2.8	1500	0.62	1700	Yes	No	30	0.1–5.6	None specified
Medtronic	Minimax	0.8	149	0.5–2.3	Not specified	0.45	2000	No	No	150	None specified	None specified
Maquet	Quadrox-i Small Adult	1.3	175	0.5–5.0	3000	0.62	4200	Yes, with VHK 2001 reservoir	No	300	Up to 15	None specified
Maquet	Quadrox-i Small Adult with integrated ALF	1.3	295	0.5–5.0	3000	0.62	4200	Yes, with VHK 2001 reservoir	Yes/40 μm/430 cm^2	300	Up to 15	None specified
Sorin	Inspire 6 (optional integrated arterial filter)	1.4	184/284 with ALF	Up to 6.0	4300	0.6	4500	Yes	Optional/38 μm/68 cm^2	150	Up to 12 LPM, not to exceed 2:1 V:Q	Up to 12 LPM, not to exceed 2:1 V:Q
Terumo	Capiox RX15, 30	1.5	135	0.5–4, 5 LPM with assisted venous drainage	1400	0.6	3000	Yes	No	70	0.5–15	15 LPM for 10s, do not repeat
Terumo	Capiox RX15, 40	1.5	135	0.5–5.0	1400	0.6	4000	Yes	No	200	0.5–15	15 LPM for 10s, do not repeat
Terumo	Capiox FX15, 30	1.5	144	0.5–4, 5 LPM with assisted venous drainage	1400	0.6	3000	Yes	Yes/32 μm/360 cm^2	70	0.5–15	15 LPM for 10s, do not repeat
Terumo	Capiox FX15, 40	1.5	144	0.5–5.0	1400	0.53	4000	Yes	Yes/32 μm/360 cm^2	200	0.5–15	15 LPM for 10s, do not repeat

Table 1.3 Oxygenators rated up to 8 LPM (Part I of II).

Manufacturer	Oxygenator (microporous polypropylene except as noted)	Oxygenator bundle (m^2)	Oxygenator prime volume	Manufacturer recommended blood flow range (LPM)	Heat exchanger size (cm^2)	Heat exchanger performance factor at manufacturer maximum recommended blood flow rate	Reservoir capacity (mL)	Compatible with vacuum assisted drainage	Integrated arterial filter/ pore size/surface area	Minimum operating volume	Gas flow range	Maximum temporary gas flow for suspected wetting out of oxygenator
Sorin	Inspire 8 (optional integrated arterial filter)	1.75	219/351 with ALF	Up to 8.0	4300	0.53	4500	Yes	Optional/38 μm/ 97 cm^2	150	Up to 16 LPM, not to exceed 2:1 V:Q	Up to 16 LPM, not to exceed 2:1 V:Q
Maquet	Quadrox-i Adult	1.8	215	0.5–7.0	4000	0.6	4200	Yes, with VHK 2001 reservoir	No	300	Up to 15	None specified
Maquet	Quadrox-i Adult with integrated ALF	1.8	335	0.5–7.0	6000	0.62	4200	Yes, with VHK 2001 reservoir	Yes/40 μm/430 cm^2	300	Up to 15	None specified
Maquet	Quadrox-iD Adult (polymethylpentene)	1.8	215	0.5–7.0	4000	0.6	4200	Yes, with VHK 2001 reservoir	No	300	Up to 15	None specified
Terumo	SX18	1.8	270	0.5–7.0	2200	0.5	4000	Yes	No	200	0.5–20	20 LPM for 10 s, do not repeat
Sorin	PrimO2X	1.87	250	Up to 8	1400	0.44	4300	Yes	No	250	Up to 16 LPM, not to exceed 2:1 V:Q	Up to 16 LPM, not to exceed 2:1 V:Q
Sorin	Apex HP	1.87	250	Up to 8	1400	0.48	4000	Yes	No	200	Up to 16 LPM, not to exceed 2:1 V:Q	Up to 16 LPM, not to exceed 2:1 V:Q
Sorin	Synthesis	2	500 with integrated ALF	Up to 8	1400	0.36	4300	Yes	Yes/40 μm/400 cm^2	300	Up to 16 LPM, not to exceed 2:1 V:Q	Up to 16 LPM, not to exceed 2:1 V:Q

Table 1.4 Oxygenators rated up to 8 LPM (Part II of II).

Manufacturer	Oxygenator (microporous polypropylene except as noted)	Oxygenator bundle (m^2)	Oxygenator prime volume	Manufacturer recommended blood flow range (LPM)	Heat exchanger size (cm^2)	Heat exchanger performance factor at manufacturer maximum recommended blood flow rate	Reservoir capacity (mL)	Compatible with vacuum assisted drainage	Integrated arterial filter/pore size/surface area	Minimum operating volume	Gas flow range (LPM)	Maximum temporary gas flow for suspected wetting out of oxygenator
Medtronic	Affinity NT	2.5	270	1.0–7.0	Not specified	0.45	4000	Yes	No	200	1.0–7.0	None specified
Medtronic	Affinity Fusion	2.5	260	1.0–7.0	4000	0.55	4500	Yes	Yes/25 μm via oxygenator bundle	200	None specified	None specified
Terumo	Capiox RX25	2.5	250	0.5–7.0	2000	0.53	4000	Yes	No	200	0.5–20	20 LPM for 10s, do not repeat
Terumo	Capiox FX25	2.5	260	0.5–7.0	2000	0.53	4000	Yes	Yes/32 μm/600 cm^2	200	0.5–20	20 LPM for 10s, do not repeat
Terumo	SX25	2.5	340	0.5–7.0	2200	0.5	4000	Yes	No	200	0.5–20	20 LPM for 10s, do not repeat

shear is not only related to pressure. Additionally, pressure drop plays an important role in microembolic air removal in microporous systems [1]. Today's FDA-approved oxygenators have shear values well within acceptable limits, and pressure drop across the device should not simply be minimized. Since pressure drop values may be misleading for initial consideration of a device, they are not listed in Tables 1.1 to 1.4.

The integral ALF (select models only)

- An ALF is a screen filter with a pore size generally in the 25–40 μm range.
- The ALF generally serves as the last safeguard in a cardiopulmonary bypass circuit to trap and/or remove particulate and air emboli from the blood before return to the patient.
- An integrated ALF, if used, must meet or exceed the maximum oxygenator flow rate. One should not "push" an oxygenator with an integrated arterial filter beyond its recommended flow, even if gas exchange is acceptable, without manufacturer confirmation that the filter can safely handle a higher flow.
- For additional information, *see section "Arterial Line Filters."*

The venous reservoir

- Most pediatric centers use open hard-shell reservoirs. The term "open" refers to the reservoir being open to atmosphere for use in a system using gravity siphon drainage. The reservoir must be properly vented to prevent pressurization and the risk of air embolization to the patient. Pressure in the reservoir can result from sucker, vent, and venous inflow if air is not allowed to escape through a vent port or vacuum system. (*See section "Massive air embolism" in Chapter 8 and section "Standard and augmented venous return" in Chapter 3.*)
- "Closed" bypass systems commonly incorporate a bag design for the venous reservoir, which significantly limits the blood:air interface. Closed systems have been shown to have a decreased inflammatory response and fewer hematologic disruptions. However, closed systems have less precise visual monitoring of venous return, require additional systems for purging venous bag air, and are not readily converted to vacuum-assisted venous drainage (which comes with its own set of additional concerns for safe use).
- The venous reservoir contains the venous filter.
- The venous reservoir generally collects venous blood and cardiotomy blood. Both venous blood and cardiotomy blood get filtered separately via different flow paths in the reservoir. The reservoir may therefore be referred to as the cardiotomy venous reservoir (CVR) (see Figure 1.6).
- The reservoir capacity needs to handle the patient blood volume in cases of planned or unplanned low-flow or circulatory arrest.
- The venous reservoir is an extremely important air removal device. The vast majority of air in the cardiotomy and venous blood flow paths is removed in their respective filtration systems (see Figure 1.7).
- Venous reservoirs have inflows either near the top (top feeders) or bottom (bottom feeders). Both incorporate an extension tube (venous straw) which runs low in the reservoir to help maintain a continuous column of fluid for gravity siphon drainage.
- Venous (and cardiotomy) filters are normally coated, at least in part, with a chemical compound to help prevent the formation of foam and to eliminate foam that has been introduced to the filter. Antifoam products containing silicone, simethicone, and methylcellulose are currently used for this application.

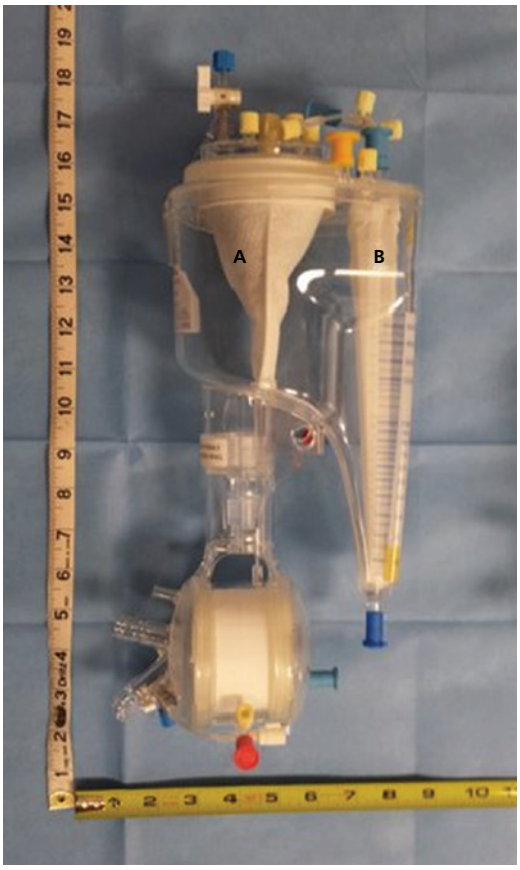


Figure 1.6 Filters in the Terumo CAPIOX FX05 oxygenator. *A—cardiotomy filter in the CVR. B—venous filter in the CVR.* (*See insert for color representation of the figure.*)

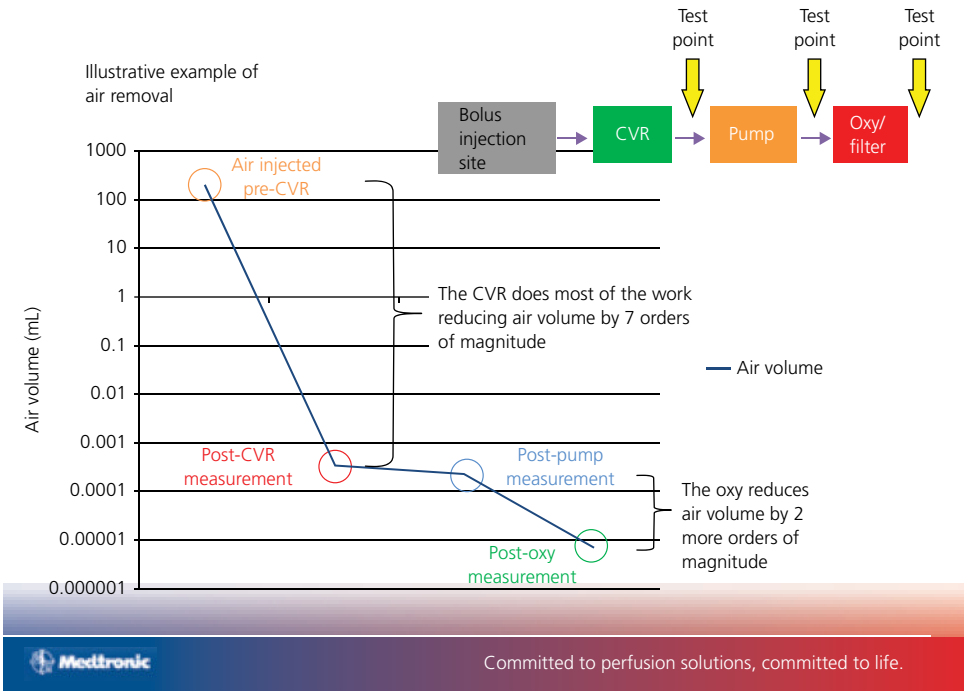


Figure 1.7 Air removal in the CVR and oxygenator. Reproduced with permission from Medtronic, Inc., Minneapolis, MN. All rights reserved. (*See insert for color representation of the figure.*)

- It is common for defoaming agents to coat only the upper levels of a filter system. This results in a system whereby it is not mandatory for the blood to pass through the defoamer. The blood/foam will only come in contact with the defoamer when it is more likely to be needed (i.e., foam rising above a certain reservoir level during periods of high sucker flow) and during periods when the reservoir level is high. If foam is seen in a CVR during bypass, the perfusionist should maintain a higher reservoir level to aid in the defoaming process. This aids in the removal of gaseous microemboli by allowing for an increased reservoir transit time.

The cardiotomy filter

- The cardiotomy filter has its own flow rating that is generally less than the maximum oxygenator flow but well within the needs for field suction and left ventricular vent flow.
- The cardiotomy filter generally provides more filtering capacity than the venous filter. Venous blood tends to be much "cleaner" with less air and particulate emboli, while cardiotomy blood generally has more.
- An integrated cardiotomy filter is normally located higher than and behind the venous reservoir. This arrangement allows cardiotomy blood to passively flow into the venous reservoir after passing through the cardiotomy filter.
- Consideration should be given to a secondary stand-alone cardiotomy reservoir if there is concern that the integrated CVR would overflow in cases of pump low-flow or circulatory arrest. The secondary reservoir can be used to temporarily store blood volume.
- A secondary cardiotomy reservoir with filter should also be considered for cases with an expected high sucker flow (reoperations, patients with significant MAPCAs, very large patients with high pump flow, surgery in or around the liver). The additional filtration, or prefiltering, of shed blood provided by a secondary cardiotomy filter may increase the useful life of the primary CVR. When utilized, a secondary cardiotomy is usually set up to process vent and sucker return with drainage to the primary cardiotomy reservoir.
- Cardiotomy (and venous) filters are normally coated, at least in part, with a chemical compound to help prevent the formation of foam and to eliminate foam that has been introduced to the filter. Antifoam products containing silicone, simethicone, and methylcellulose are currently used for this application.
- It is common for defoaming agents to coat only the upper levels of a filter system. This results in a system whereby it is not mandatory for the blood to pass through the defoamer. The blood/foam will only come into contact with the defoamer when it is more likely to be needed (i.e., foam rising above a certain reservoir level during

periods of high sucker flow) and during periods when the reservoir level is high. If foam is seen in a CVR during bypass, the perfusionist should maintain a higher reservoir level to aid in the defoaming process. Of course, this would also aid in the removal of gaseous microemboli by allowing for an increased reservoir transit time.

The heat exchanger

- An oxygenator's integrated heat exchanger must be water tested prior to the addition of crystalloid solutions for priming. Running water through the heat exchange system at a flow and pressure comparable to the operating room values and inspecting for leaks to the blood compartment are important steps in the process of setting up a heart–lung machine for bypass.
- The water pathway through an oxygenator helps dissipate static electricity charges that may develop in the roller head pumps and be transmitted through the blood pathway. This feature is especially important in preventing static electricity discharge through the heart–lung machine circuitry that can cause damage.
- Heat exchanger specifications are qualified by a "performance factor" defined as the difference of inlet (venous) and outlet (arterial) blood temps divided by the difference of inlet (venous) blood and inlet water temperatures.

$$\text{Performance factor} = \frac{\text{(arterial blood temperature} - \text{venous blood temperature)}}{\text{(water inlet temperature} - \text{venous blood temperature)}}$$

- The performance factor is frequently in the range of 0.4–0.7 at a device's maximum rated flow.
- The gradient between the venous blood and the water inlet temperature is usually limited to 10 °C (though some manufacturers allow for a gradient of up to 15°C). This is to prevent excessive gradients where there is potential for gas to come out of solution if warmed too rapidly (due to the decrease in gas solubility at higher temperatures). Limiting the temperature gradient also helps to evenly cool and warm a patient. A more homogenous warming may result from limiting the gradient during rewarm which can help prevent after-drop (whereby core patient temperature falls in the early post bypass period).
- Some manufacturers may explicitly state that the gradient should be less than 10 °C (an 8°C maximum for example), which would be an important factor in evaluating a product for use in a congenital cardiac program that uses moderate-to-profound hypothermia. Lower gradients can increase the length of a bypass case since rewarming times will be longer. (*See section "Temperature management" in Chapter 3.*)
- It is commonly accepted that limiting temperature gradients is important in preventing gas from coming out of solution. However, an animal study by Nollert *et al.* found no correlation between temperature gradient and emboli count [2]. Manufacturer-recommended gradient limits should not be exceeded.
- The minimum acceptable heat exchanger performance factor is not defined by the manufacturer or AAMI. This leaves the practitioner to decide if heat exchange performance is acceptable at the intended bypass flow and/or up to the maximum recommended oxygenator flow. This assessment comes with clinical experience. To note, an oxygenator may provide acceptable gas exchange but unacceptable heat exchange for a planned deep hypothermic circulatory arrest case. This situation, for example, would encourage the perfusionist to upsize the oxygenator and therefore the integral heat exchanger.
- It is important to note that there have been oxygenators manufactured with different oxygenator bundle sizes wrapped around the same-sized heat exchanger. Therefore, the perfusionist must be familiar with product line specifications.
- The heat exchanger performance factor must be evaluated carefully. The value stated by the manufacturer is not standardized to a heat exchanger water flow rate. Furthermore, the value must be obtained from a chart that looks at the spectrum of performance factors over the entire rated flow range. Therefore, performance factors are ideally evaluated at the expected pump flow rate for a given case. To note, the values listed in Tables 1.1 through 1.4 are performance factors at the maximum rated blood flow as stated by the manufacturer but are not always standardized to 10 LPM water flow since that information is not always available.
- Heat exchanger performance will be least efficient at the upper flow rating for a device.
- Increased heat exchanger surface area does not always equate to increased performance. The blood flow path through the oxygenator (axial vs. radial), the heat exchanger material, and characteristics of the water system will affect performance.
- It is important to consider that the rates of cooling and warming on cardiopulmonary bypass are impacted by systemic vascular resistance management and pump flow.

Tables 1.1, 1.2, 1.3, and 1.4 list devices currently on the market. Adult-sized oxygenators are included since

congenital cardiac perfusion programs must be able to accommodate patients of all sizes. Users are encouraged to trial oxygenators and evaluate all aspects of performance in their own operating room environment. Then, along with this thorough understanding of the oxygenators to be used, custom tubing packs can be created. Ultimately, a perfusion group should develop a chart identifying the components to be used for different expected pump flow ranges. (*See section "Comprehensive experience-based equipment chart" in Chapter 12.*)

Arterial line filters

The cardiopulmonary bypass circuit contains filters in several systems: the cardiotomy reservoir, venous reservoir, oxygenator bundle, bypass arterial line, prebypass, gas line, crystalloid line, blood transfusion line, cardioplegia system, and others. The ALF tends to receive the most attention since it serves as the last system in a cardiopulmonary bypass circuit to trap and/or remove particulate and air emboli from the blood before return to the patient. It is important to note that one of the most important aspects of a bypass circuit is its ability to remove (and not create) particulate and air emboli. Several authors have conclusively shown that microair emboli are delivered to patients on bypass [3–8]. These emboli can result in microvasculature blockages with the downstream effects of hyoperfusion or ischemia. Air in particular is disruptive to the endothelial glycocalyx which has important implications for vascular permeability and the potential for edema throughout the body with cardiopulmonary bypass [9–11]. The cardiotomy reservoir, venous reservoir, and oxygenator bundle are all important air handling and removal devices in a cardiopulmonary bypass circuit. The ALF is the last removal device before blood reenters the body. Figure 1.8 depicts four commonly used external ALFs. The ALF must be properly deaired with a crystalloid priming solution before bypass. If a blood prime is used, blood should only be added after crystalloid priming and thorough ALF deairing.

- ALFs are most commonly standalone add-ons to a perfusion circuit.
- ALFs are screen filters with a pore size ranging from 25 to 40 μm.
- Filter housings are preferably clear to visually facilitate deairing during crystalloid priming.
- External ALFs most commonly have folded or wrapped filter medium that is not fully visible. The filter should be carbon dioxide flushed prior to crystalloid priming. Then, with fluid flowing through at the manufacturer's recommended rate for priming, the filter should be adequately tapped to ensure no air is trapped (more precisely, any microbubbles present *should be* carbon dioxide).
- Air emboli during bypass are generally purged from the filter by its flow characteristics and venting system with the flow velocity decreasing in the external ALF encouraging any bubbles to rise and exit via the top-mounted purge line.
- The external ALF most often has a bleed line continuously returning a low flow back to the cardiotomy reservoir. Less commonly, hydrophobic material located at

Figure 1.8 External arterial line filters. **(a)** *Sorin Group D736.* **(b)** *Sorin Group D733.* **(a)** and **(b)** Reproduced with permission from Sorin Group USA Inc., Arvada, CO. All rights reserved. **(c)** *Terumo Capiox AF02.* Reproduced with permission from Terumo Cardiovascular Group, Ann Arbor, MI. All rights reserved. **(d)** *Medtronic Affinity Pixie.* Reproduced with permission from Medtronic, Inc., Minneapolis, MN. All rights reserved. (*See insert for color representation of the figure.*)

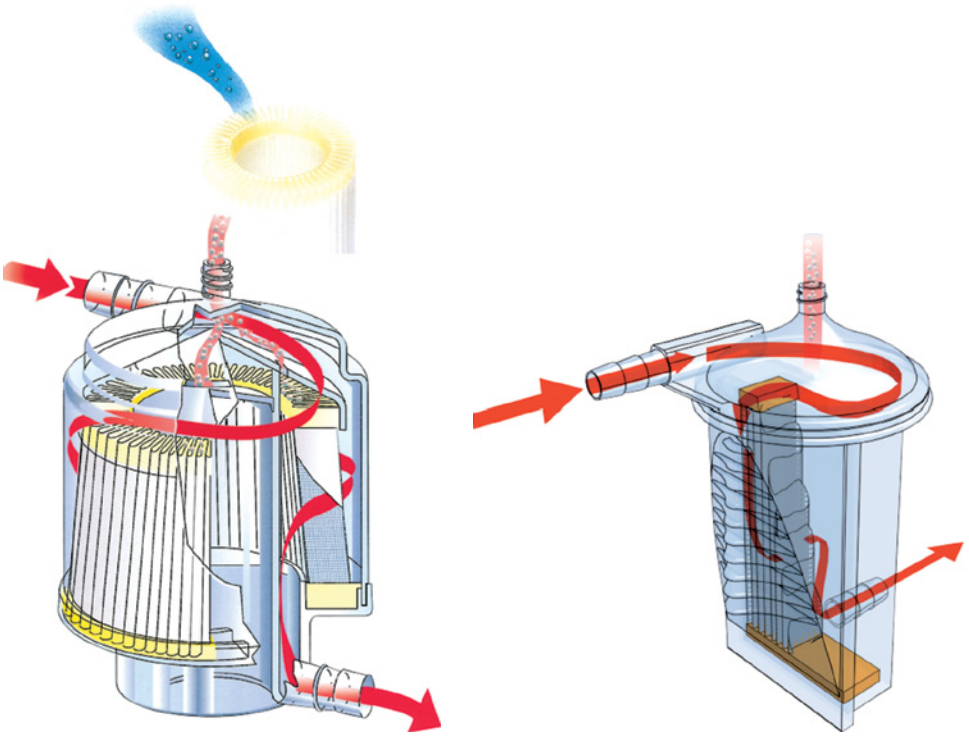


Figure 1.9 Typical flow path through external arterial line filters. The top luer connector purges continuously via a line connected to the CVR. Reproduced with permission from Sorin Group USA Inc., Arvada, CO. All rights reserved. (*See insert for color representation of the figure.*)

the top of the filter allows air to be directly vented to the atmosphere. Figure 1.9 shows the typical blood flow path through two external ALFs with top-mounted purge lines.

- Some oxygenators now have integrated screen filters wrapped around the oxygenator bundle. These integrated, or internal, ALFs additionally promote the removal of air in the blood by purging air directly through the microporous oxygenator fibers to be vented out the gas phase of the oxygenator. This is facilitated by the back pressure the internal filter provides and the proximity of the potential air to the oxygenator bundle's micropores.
- Oxygenator-integrated ALFs decrease circuit priming volume since the additional filtering medium and space usually adds less volume than a standalone external ALF to the overall prime.
- Future oxygenators may employ microporous membrane wrapping technology to serve dual function as oxygenator and arterial filter. However, the use of oxygenator membranes serving the additional role of filter medium has not been adequately validated in the literature. The adult-sized Medtronic Affinity Fusion is currently the only oxygenator on the market FDA approved for this dual role.
- The pressure drop across an ALF is an important factor to evaluate. However, filters today tend to have a generous surface area with rated flows well within acceptable values for sheer stress and turbulence.

Table 1.5 lists commonly used external ALFs. The charts in Tables 1.1 to 1.4 can be used to compare the integrated ALFs some oxygenator models offer against the external model characteristics listed in Table 1.5.

Tubing packs

Custom tubing packs for cardiopulmonary bypass circuits are commonly created by the perfusion team to aid in quick and efficient setup of the heart–lung machine (HLM). A congenital cardiac program may have 3–5 different oxygenators as well as 3–5 tubing packs with overlap between oxygenators and tubing packs. This creates numerous options that come with several considerations when choosing equipment for bypass. Table 1.6 is an example of tubing pack specifications based on anticipated maximum pump flow rates. In addition to these items, an institutional comprehensive experience-based equipment chart is helpful in defining the options for nearly all common components

Table 1.5 External arterial line filters.

Manufacturer	Model	Maximum flow (LPM)	Prime volume (mL)	Pore size (μm)
Sorin	KiDS D130	0.7	16	40
Sorin	KiDS D131	2.5	28	40
Terumo	Capiox AF02	2.5	40	32
Sorin	D736	2.5	47	40
Sorin	D735	2.5	47	27
Pall	AL3	3	28	40
Medtronic	Affinity Pixie	3.2	39	30
Sorin	D731	6	100	27
Sorin	D733	6	100	40
Terumo	Capiox AF125X	7	125	37
Maquet	QUART	7	180	40
Terumo	Capiox AF200X	7	200	37
Medtronic	Affinity	7	212	20
Medtronic	Affinity	7	212	38
Pall	AL6	8	100	40
Pall	AL8	8	170	40
Sorin	D732	8	195	27
Sorin	D734	8	195	40
Pall	AV6SV	8	220	40

Table 1.6 Example of equipment selection based on anticipated maximum pump flow rate.

	Total circuit prime (mL)	Oxygenator with integrated 32 μm ALF	Primary tubing pack components	Milliliters of prime for oxygenator (O), reservoir (R), arterial limb (A), boot (B), venous limb (V), centrifugal venous head and tubing (CH)
Up to 1.2	215	Terumo Capiox FX05 rated to 1.5 LPM	3/16 arterial 3/16 boot 1/4 venous	(O) 43, (R) 75, (A) 30, (B) 15, (V) 52
1.2–1.5	230		3/16 arterial 1/4 boot 1/4 venous	(O) 43, (R) 75, (A) 30, (B) 30, (V) 52
1.5–1.8	460	Terumo Capiox FX15-30 rated to 5 LPM (with assisted drainage)	1/4 arterial 1/4 boot 1/4 venous	(O) 144, (R) 150, (A) 75, (B) 30, (V) 61
1.8–2.1	535		1/4 arterial 1/4 boot 3/8 venous	(O) 144, (R) 150, (A) 75, (B) 30, (V) 136
2.1–3.15	570		1/4 arterial 3/8 boot 3/8 venous	(O) 144, (R) 150, (A) 75, (B) 65, (V) 136
3.15–3.6	675		3/8 arterial 3/8 boot 3/8 venous	(O) 144, (R) 150, (A) 175, (B) 65, (V) 141
3.6–4.4	750		3/8 arterial 3/8 boot 3/8 venous, Centrifugal venous assist head	(O) 144, (R) 150, (A) 175, (B) 65, (V) 141, (CH) 75
4.4–5	775		3/8 arterial 1/2 boot step up 3/8 venous, Centrifugal venous assist head	(O) 144, (R) 150, (A) 175, (B) 90, (V) 141, (CH) 75
5.0–7.0	980	Terumo Capiox FX25 rated to 7 LPM	3/8 arterial 1/2 boot step up 3/8 venous, Centrifugal venous assist head	(O) 250, (R.) 250, (A) 175, (B) 90, (V) 140, (CH) 75

for bypass. (*See section "Comprehensive experience-based equipment chart" in Chapter 12.*)

- A cardiovascular team must define their maximum acceptable flows through the various sizes of available tubing: 3/16″, 1/4″, 3/8″, and 1/2″ (other sizes are available but are less commonly used).
- Institutions vary in regards to standard bypass tubing length, table height, venous reservoir height, and typical venous cannulae style and size which all impact achievable flow rates.
- Tubing used for venous return may be evaluated with gravity versus augmented drainage.
- Boot (arterial pump raceway) tubing should be evaluated for flow based on head RPMs since spallation and the potential for failure are a function of the number of tubing compressions over time. Maximum RPMs used are commonly in the 160–170 RPM range even though roller heads may be capable of 250 RPMs.
- Smaller raceway lengths (i.e., mini-heads) increase the compression rate for a given pump flow rate.
- Table 1.7 outlines an *example* of the maximum flows used for various tubing sizes.
- Each tubing size has a prime volume that can be calculated with a known length (Table 1.8).
- The objective in defining custom tubing pack line sizes is to achieve the smallest prime volume while ensuring adequate and safe flow rates and pressures.
- Consideration must be given to the range of operative table movement since rolling the bed away in an up position may require several inches of pump tubing to safely accommodate.
- The venous line must have some slack in it to allow for "walking" air out if an air lock develops on bypass with gravity siphon drainage.
- The total system prime volume will include the tubing pack, oxygenator, starting reservoir level, dynamic volume holdup (if significant), and hemoconcentrator (if primed before bypass).
- The starting reservoir volume may be higher for surgeons who tend to give more pump volume during the cannulation process.
- Tubing lengths will vary for each case based on how much of the arterial-venous loop is discarded before connection to the bypass cannulae. Excess tubing in the pack is important to accommodate varied pump positions, particularly variances for femoral bypass.
- Some pediatric centers will claim a much lower prime volume with the use of retrograde arterial priming and venous antegrade priming. It is debatable whether these autologous priming strategies have a net positive effect since young congenital heart disease patients frequently do not have much volume to "donate" to the bypass circuit in the immediate prebypass period. In fact, these patients are frequently more sensitive to overall volume loss during cannulation. It is also debatable whether it is appropriate to use alpha agents such as phenylephrine immediately before bypass to aid in autologous priming techniques.
- The prime volume of a custom tubing pack is simply the various line lengths multiplied by the volume contained. The volume of tubing cut off at the field may be subtracted only if it is accounted for in the starting reservoir level since blood-primed circuits will have this volume returned via the pump suckers before bypass.
- Future efforts toward circuit prime reduction will focus primarily on tubing size and length since the bulk of the prime is now either there or in the venous reservoir. The use of vacuum-assisted venous drainage has allowed some centers to move the oxygenator apparatus closer to the patient since the kinetic potential requirement between the patient relative to the reservoir is eliminated. Vacuum-assisted venous drainage though is not without risk and must be carefully implemented into a congenital cardiac perfusion practice. (*See section "Standard and augmented venous return" in Chapter 3.*)

Table 1.6 is offered as an *example* of an overview for equipment selection. The maximum flow rate anticipated on

Table 1.7 Typical tubing sizes with flow maximums.

Tubing size	Max flow boot line in standard raceway (mL/min)	Max flow arterial line (mL/min)	Max flow venous line with gravity siphon drainage (mL/min)	Max flow venous line with assisted drainage (mL/min)
3/16″	1200	1500	600	800
1/4″	2100	3150	1800	2500
3/8″	4400	>3150	3750	>3750
1/2″	>4400	NA	>3750	>6000

Table 1.8 Tubing prime volume.

Internal tubing diameter	1/8″	3/16″	1/4″	5/16″	3/8″	1/2″
Milliliters per foot	2.4	5.0	9.7	15.5	21.7	38.6

bypass is identified on the left with prime volumes and circuit components required listed to the right. An individual bypass plan may require a maximum flow of 2.5–4.0 L/min/m^2, which usually corresponds with an absolute flow of 0.4–7.0 LPM. There is overlap between the ranges listed. When the anticipated maximum flow straddles two oxygenator brackets, it is important to consider whether the functional reserve built into this estimate is worth the additional prime volume, and possibly the additional minimum operating volume, which importantly affects how a bypass case is run. This chart does not list ALFs since the oxygenators in this example have integrated 32 µm screen filters wrapped around the oxygenator bundle. This chart also includes the option of kinetic-assisted venous drainage for pump flows greater than 3.6 LPM since 1/2″ venous tubing is not utilized. Perfusion programs providing the option of vacuum-assisted venous drainage for all patients may be able to further decrease priming volumes by moving the oxygenator assembly closer to the patient. Gravity drainage requires the assembly to be closer to the floor for sufficient kinetic potential.

Cardioplegia systems

There are dozens of congenital cardiac surgery cardioplegia formulas, delivery methods, and protocols that are heavily based on institutional experience and surgeon preference. In fact, a recent North American survey of the Congenital Heart Surgeon's Society showed that while 86% of respondents used blood-based cardioplegia, the crystalloid component of those solutions varied; 38% del Nido, 34% custom, 16% St Thomas/Plegisol/Baxter, 7% Custodial, and 5% microplegia [12]. Additionally, there were wide variations noted regarding dosage, delivery method, temperature, and time interval between doses. It is clear that while excellent clinical outcomes have been achieved at numerous centers, consensus regarding optimal myocardial protection for congenital cardiac surgery patients has not been reached.

Cardioplegia *systems* can broadly be categorized as recirculating, nonrecirculating, and continuous. Each system will require either a custom tubing pack or a commercially available option. It is beyond the scope of this chapter to describe the dozens of cardioplegia formulas and delivery methods in use. The following are types of cardioplegia *systems.* Articles regarding *types* of cardioplegia are listed in the reference section at the end of this chapter [12–25].

Recirculating cardioplegia system

- The recirculating circuit has an outlet, ideally near the aortic root needle, to which cardioplegia is directed during delivery (the return limb is clamped and the outlet line unclamped during cardioplegia delivery). Otherwise, the flow is recirculated (the outlet line is clamped and the recirculation limb is unclamped) (see Figure 1.10).
- Contains cardioplegia (crystalloid or blood-based) in a system in which the final product for delivery is kept flowing, well mixed, and at the desired temperature (see Figure 1.11).

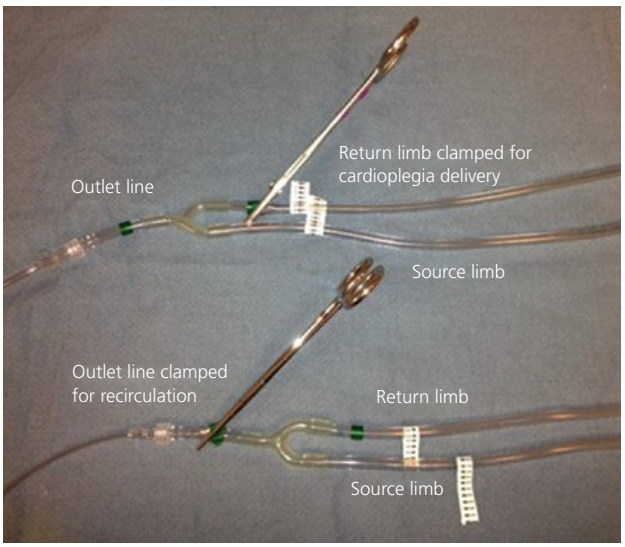


Figure 1.10 Recirculating cardioplegia system table lines. Top: Recirculating delivery system with table line return limb clamped for cardioplegia delivery. Bottom: Recirculating delivery system with table line outlet limb clamped for recirculation. (*See insert for color representation of the figure.*)

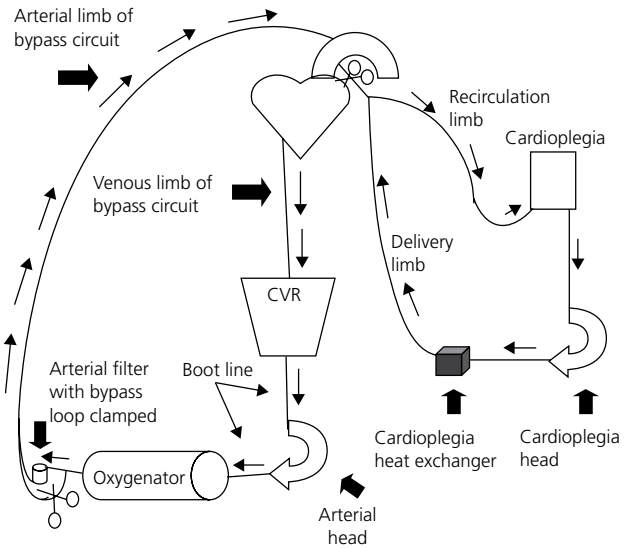


Figure 1.11 Recirculating cardioplegia schematic. Note the table line outlet limb is clamped for recirculation.

- An active cooling coil or ice bath coil is used to adjust the cardioplegia delivery temperature.
- An ideal system has minimal dead space volume. The dead space volume in a recirculating system is that which is not part of the active flow but integral to the delivery pathway. Generally, this would be the volume of the aortic root needle proximally back to the return limb of the flow path.
- The dead space volume must be kept to a minimum since it can warm to room temperature and contain poorly mixed cardioplegia solution.
- Cardioplegia in the dead space can be flushed through a vented aortic root before delivery if that volume is deemed significant.
- Recirculating systems may be used for single-dose and multidose cardioplegia strategies.
- A recirculating system is by its nature a closed system and is therefore not vented to the atmosphere. A recirculating system should not be flushed with carbon dioxide before priming unless there is a mechanism to remove the excess carbon dioxide which will be dissolved in the cardioplegia solution with priming.
- 1/8″ to 3/16″ tubing is commonly used to minimize the cardioplegia circuit prime volume.

Nonrecirculating cardioplegia system

- Generally pulls arterialized blood from a post-oxygenator source that is mixed with a crystalloid component (see Figure 1.12).

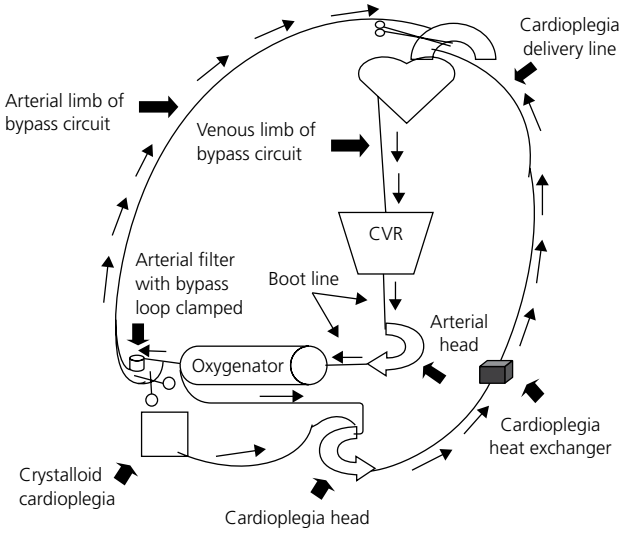


Figure 1.12 Nonrecirculating cardioplegia schematic. The cardioplegia head draws a crystalloid component together with a blood component to be delivered through a cardioplegia heat exchanger to the patient.

- May also be referred to as a single-pass system.
- The mixed cardioplegia solution flows through an active coiling coil or ice bath coil and on to the aortic root needle.
- This system inherently has more dead space volume. This volume extends from the aortic root needle back to the blood and crystalloid component sources. The cardioplegia in the dead space portion of the circuit may be at room temperature during initial and/or subsequent doses if it is not flushed from the system in some fashion.
- The dead space volume may equate to a significant portion of the intended dose volume in neonates and infants.
- Can be employed for single-dose and multidose cardioplegia strategies.
- 1/8″ to 3/16″ tubing is commonly used. The crystalloid and blood components may have different tubing sizes, which run through the same roller head to effect the desired ratio between the two components. Alternately, the crystalloid and blood components may run through different roller heads to allow for a variable and adjustable ratio of components during a bypass run.

Continuous cardioplegia system

- Continuous cardioplegia, also known as microplegia, is primarily arterialized blood drawn from a post-oxygenator source that is mixed with an arresting agent and other additives.
- It is given via continuous or cyclic low flow.
- The concentration of additives may be adjusted for effect in real time.
- The delivery temperature may be cold or warm and may vary throughout the arrest period; warm or cold induction, warm or cold maintenance, and warm terminal reperfusion dose (aka hot shot).
- Continuous cardioplegia systems are not commonly used for congenital cardiac surgery, and there are few published reports for its use in neonates and infants.

The heart–lung machine

The HLM needs several basic items for safe cardiopulmonary bypass. Most importantly, periodic preventative maintenance as recommended by the manufacturer is essential. Such checks go a long way toward

preventing the need for emergency procedures (hand cranking, arterial head change-out, etc.). Today's HLMs minimally have arterial head servoregulation for system pressure, reservoir-level sensing, internal checks for over/under-speed of the head, a calibrated system for calculating pump flow based on tubing size and raceway length, bubble detection, and master/follower head assignments (i.e., cardioplegia head should not flow more than the arterial head in systems drawing cardioplegia blood from a post-oxygenator source). It is strongly encouraged to have HLMs that can quickly and easily be adapted to any size patient. Then, knowledge of patient specifics allows one to select and set up any available HLM for the bypass run, particularly in an emergency. Figure 1.13 depicts a customized HLM configuration.

The HLM, in my opinion, has seen five significant advances over the past decade:

1 The layout of the hardware has become more customizable.
   - HLMs formerly existed as 4–6 standard pump heads on a mobile base. Today's HLM allows for regular-sized heads and mini-heads to be pole mounted, rotated toward the field, and even placed in a somewhat stacked fashion to decrease the overall console footprint and minimize the circuit prime volume. The roller pump heads may also rotate in their housings which can decrease line lengths and prevent hard angles as the tubing makes its way through the system. However, it is important to note that pump consoles still follow the law of conservation of mass. When a HLM decreases its width side-to-side, it tends to increase its front-to-back dimension. This is an important consideration for programs that occasionally use their HLMs in areas of the hospital outside of the cardiac operating rooms. Not all doorways in an institution may allow for passage of the newer style HLM as easily as the cardiac operating room doorways.
2 Belt-driven systems have been replaced with electric field motor drives.
   - This technology requires less maintenance with increased reliability since there are fewer parts (belts, brushes, bushings, etc.) to wear out over time.
3 Basic LCD displays have been replaced with touch screen color systems.
   - This advance has decreased the number of display pages that need to be scrolled through, and therefore has increased efficiency. Color systems also aid with quick detection of specific alarms and alerts.

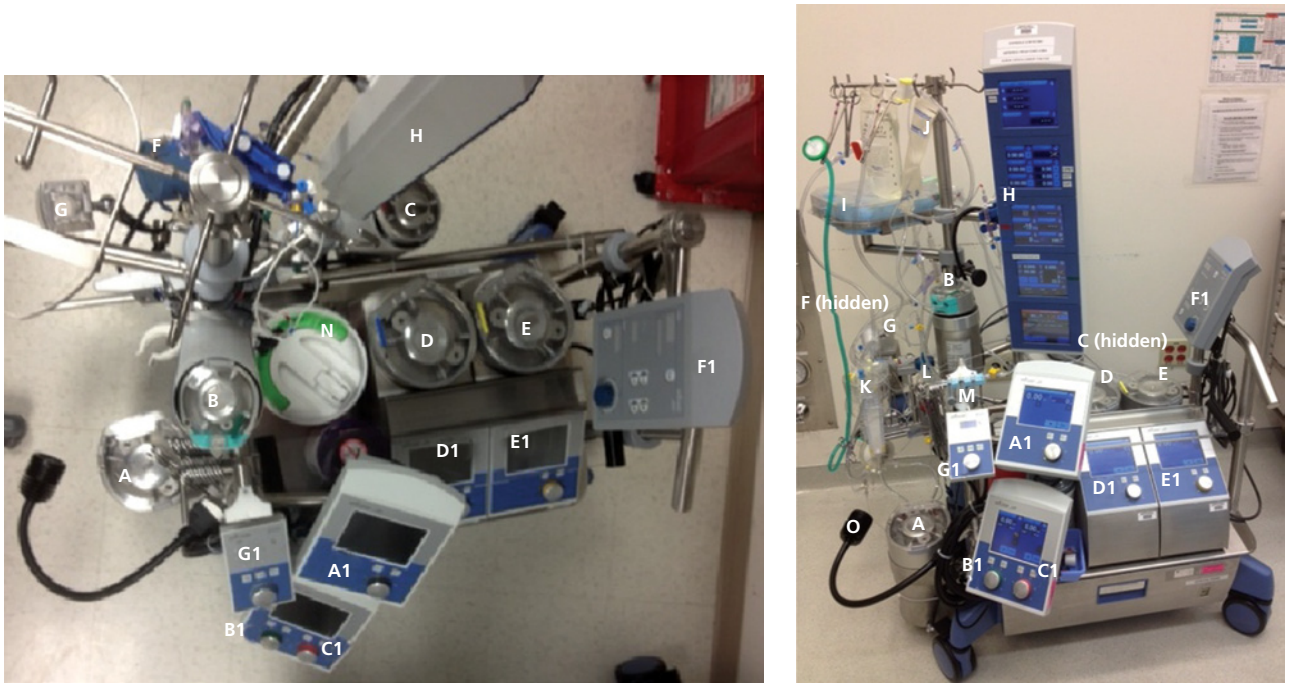


Figure 1.13 Top and side views of a customized Stockert S5 heart–lung machine. *A—Arterial head and its controller A1, B—cardioplegia head and its controller B1, C—vent head and its controller C1, D—field sucker and its controller D1, E—field sucker and its controller E1, F—centrifugal head motor (for kinetic venous-assisted drainage) and its controller F1, G—electronic venous occluder and its controller G1, H—master display tower, I—sterile custom tubing pack, J—custom cardioplegia tubing set, K—oxygenator, L—hemoconcentrator, M—blood gas sampling manifold, and N—ice bucket for cardioplegia cooling coil.* (*See insert for color representation of the figure.*)

4 Display systems and head assignments are more customizable.
   - The control and alarm displays can be assigned to different locations on the display tower to meet the needs of a perfusion program. These computerized systems have also eliminated the requirement of older systems where servoregulation was controlled by individual component power plug placement. Current systems allow for user assignment of servoregulation and alarm systems to individual pumps. This is an important safety improvement since older systems risked running without servoregulation if a replacement head could not be plugged into the one designated outlet on the console for that feature (i.e., the arterial head was servoregulated for pressure and level solely based on its power supply location in the base).

5 Internal HLM batteries are nearly universal and more powerful.
   - These upgraded batteries can power the console for an extended period of time. This is an essential feature even in hospitals with backup power systems because those systems can experience brief temporary loss of output. Loss of output in computerized systems without battery backup can result in extended boot-up processes complicating the reinstitution of bypass. Computerized systems generally do not allow for hand cranking during the startup systems-check, which adds unnecessary risk to the patient. An integrated battery backup system alleviates any concerns regarding temporary loss of hospital power and allows for transport in the unlikely event of an operating room environment emergency requiring evacuation.

The arterial pump head

HLMs for congenital heart surgery most often use roller heads for the arterial pump. Roller head pumps allow for precise control over flow and are not afterload sensitive. Centrifugal heads are less commonly used for congenital cardiac surgery, especially in younger patients. Centrifugal heads may have theoretical (and oft-cited) advantages in regards to decreasing blood trauma and preventing massive air infusion, but studies have shown that centrifugal heads can produce more hemolysis than roller pumps and they transmit gaseous microemboli just as roller heads do [26–28]. However, the inability to finely control blood flow for extended periods of time, especially when weaning support, and the inability to provide exceptionally low blood flow rates for neonates or during regional cerebral perfusion, essentially limits their utility in congenital cardiac surgery. Additionally, it is less than ideal to have a perfusion group switching between roller heads and centrifugal heads depending on the day's case mix and having more custom pack options on the shelf to accommodate such. Therefore, most congenital cardiac surgery programs utilize roller head pumps in the arterial position on their HLMs.

The heater-cooler system

The heater-cooler system is an essential cardiopulmonary bypass component. It is the piece of hardware that interfaces with the heat exchanger that is integrated in today's oxygenator housing. It may also have the capability to provide separate water flow to a cardioplegia system or surface cooling/warming blanket. The heater-cooler system may also be referred to as the thermocirculator.

A heater-cooler system comes in two varieties. The most common system is a standalone mobile unit that is capable of heating and cooling up to three water baths (Figure 1.4). One water bath is connected to the oxygenator heat exchanger with water lines capable of providing 10–18 LPM of water flow and temperatures of 15–41 °C. The other water bath(s) are generally used to provide temperature management to the cardioplegia system. These baths are sometimes separate to allow for rapid temperature change between the following cardioplegia strategies: cold (2–15 °C), tepid (28–32 °C), and warm (32–37 °C). Alternately, the other water bath(s) may be used to provide water flow through a surface cooling/warming blanket under the patient.

The standalone heater-cooler unit is a rather simple device. It provides water flow based on pump power and system resistance. Different oxygenators and different water connection lines, lengths, and hardware will result in varying water flow rates. A water flow rate of 10–18 LPM is commonly used by manufacturers to test oxygenator heat exchangers. The unit should provide flow in this range. If a flow quantifier is not included with the unit, an aftermarket product should be added. It is important to have the ability to monitor water flow since it is an important factor in cooling and warming a patient. The ability to readily quantify water flow is important when ruling out issues related to the temperature management of a patient. Suboptimal water flow, due to a kinked line for example,

can significantly decrease the ability to properly manage a patient's temperature on bypass.

A less common type of heater-cooler system is termed "wall water." This system utilizes the traditional hot and cold water supplies, or a designated system for such, specifically for the cardiac operating rooms. These systems usually have the advantage of high water flow and the ability to rapidly change temperature. Recall that standalone systems usually only have one water bath for oxygenator heat exchanger management that requires active warming and cooling when changes are required. The wall water system, on the other hand, allows for immediate temperature change since there are no recirculating baths. Wall water systems use fresh water at the desired temperature, which is then sent to a drain system. For this reason, wall water systems tend to have higher operating costs. Reliance on wall water systems also requires standalone units as backup in case of system failure, increasing operative costs.

Most perfusion programs use standalone heater-cooler systems. These systems have proven to be reliable, cost-effective, easy to maintain, and mobile. Their evolution over the past decade has decreased their size and made them quieter, both important in the operating room environment. Additionally, remote-operating control systems have allowed the units to be located away from the HLM console. This may be an advantage to the operating room layout, but it should be noted that significant water line length decreases system efficiency since more water must be heated or cooled. This is an important consideration in congenital cardiac programs regularly utilizing moderate-to-deep hypothermia.

Figure 1.14 Stockert heater-cooler system 3T (three-tank system). *A—Cold cardioplegia circuit. The first tank instantly provides 2–10°C water to the cardioplegia system. B—Warm cardioplegia circuit. The second tank instantly provides 15–41°C water to the cardioplegia system. It can also be fitted for a water-based patient surface cooling/heating blanket. C—Oxygenator circuit. The third tank provides 15–41°C water to the oxygenator heat exchanger. It has a second pump and circuit to provide water flow from the same tank to a water-based patient blanket if desired.* (*See insert for color representation of the figure.*)

Cannulae

Patients with congenital heart disease can be cannulated for bypass in a variety of ways. The ascending aorta is most commonly cannulated for arterial inflow. Some congenital cardiac defects require arterial inflow via the main pulmonary artery with flow through the ductus arteriosus to provide systemic perfusion (with branch pulmonary arteries controlled). Venous cannulation is most commonly via the superior and inferior vena cavae when intracardiac work is needed. A single right atrial venous cannula is commonly used when intracardiac work is not needed. Additionally for cannulation, a left ventricular vent may be placed via the right superior pulmonary vein and advanced across the mitral valve. Figure 1.15 depicts a structurally normal heart to illustrate cannulation sites relative to the general cardiac anatomy. Figures 1.16 and 1.17 illustrate normal left aortic arch and central venous anatomy for reference.

Regarding cannulae, is it cannulae or cannulas? These terms are used interchangeably for the plural of cannula. The term "cannulas" is more common in conversation, whereas the term "cannulae" seems to be more common in written form. Both are used in this text.

There are literally hundreds of different cannulae available for cardiopulmonary bypass. The range of cannulae that need to be readily available for a congenital cardiac program is quite extensive to cover patients of all sizes from 1 to 200 kg! One must have on hand aortic root needles of various sizes, retrograde coronary sinus and coronary perfusers, left ventricular vents, and of course connectors and tubing to incorporate these items into the

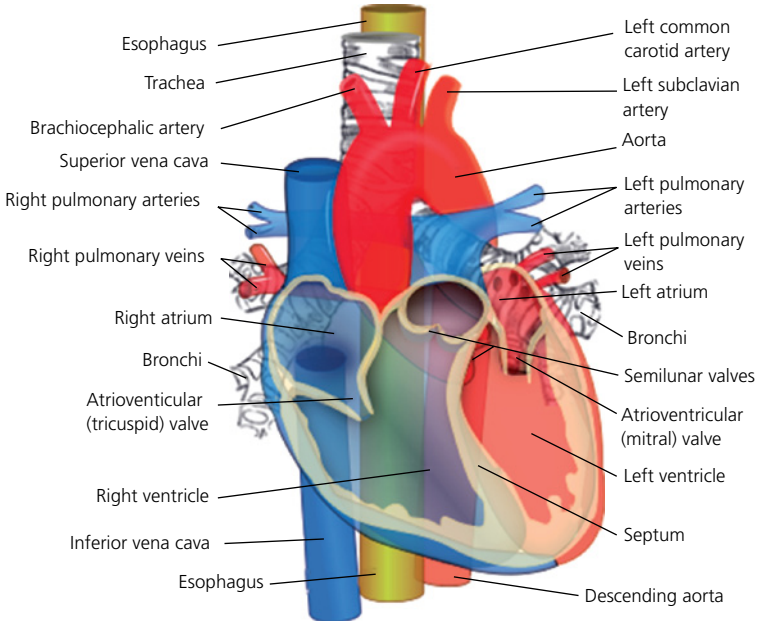


Figure 1.15 General cardiac anatomy. http://en.wikipedia.org/wiki/File:Relations_of_the_aorta,_trachea,_esophagus_and_other_heart_structures.png via Wikipedia. Reproduced with permission from under the Creative Commons Attribution-Share Alike 3.0. Unported license (accessed January 7, 2014). (*See insert for color representation of the figure.*)

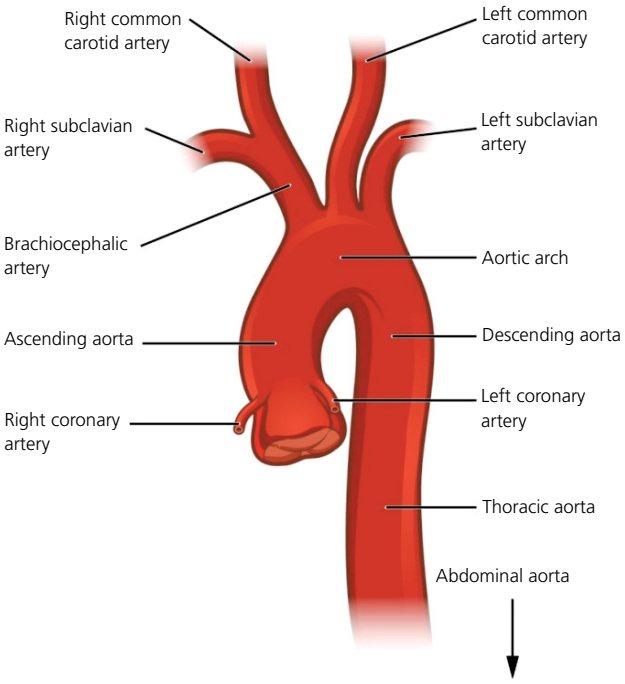


Figure 1.16 Aortic arch anatomy. http://cnx.org/content/m46646/latest/2121_Aorta.jpg. Reproduced with permission from OpenStax College on Wikimedia Commons under the Creative Commons Attribution-Share Alike 3.0. Unported license (accessed January 7, 2014). (*See insert for color representation of the figure.*)

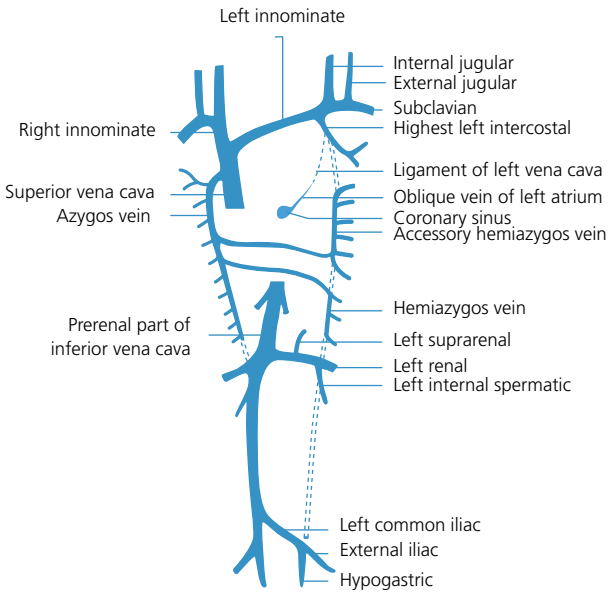


Figure 1.17 Central venous anatomy. Reproduced with permission from Gray's Anatomy of the Human Body, 1918 edition (http://en.wikipedia.org/wiki/File:Gray480.png) via Wikipedia. (*See insert for color representation of the figure.*)

bypass circuit. Arterial and venous cannulae are the most important to consider since they are required to place a patient on traditional cardiopulmonary bypass.

It is an underappreciated fact that cannulae do not come from the manufacturer with specific rated flows, as do oxygenators and ALFs. Rather, cannulae come with charts depicting flow versus pressure drop (also called pressure loss). These charts are based on manufacturer-based measurements using water flow at room temperature and may

therefore be referred to as "water charts." These charts do not factor in tubing length connecting the cannula to the bypass equipment, temperature changes during a case, hematocrit, or several other factors that may impact the actual flow capability of a specific cannula. It is therefore imperative that a perfusion program develop their own charts rating flow capabilities for cannulae within their cardiopulmonary bypass system. These experience-based charts are essential in a congenital cardiac surgery program where dozens of options exist due to considerable variation in patient and vessel size. An overview of factors determining cannulae size is provided with examples of sizing charts to follow.

Venous cannulae

Venous cannulae are graded based on their French size (Fr). Most manufacturers use the outer diameter (OD) to determine French size. It is generally accepted that with gravity drainage through a venous cannula, the pressure drop across the cannula should be limited to 35–40 mmHg. A venous cannula with a larger internal lumen will obviously have a lower pressure drop and increased flow capacity. However, there are other important considerations. Venous cannulae are made with different materials and have variable tip designs usually consisting of additional side port holes. Different cannula designs, tip materials (metal, polycarbonate, plastic, silicone), and construction methods result in different ratios between the inner and outer diameter of the cannula. All of these differences may be important to the cannulating surgeon. Metal tips tend to have the benefit of maximized inner to outer (I/O) ratios but are rigid. Silicone tips have less favorable (I/O) ratios as compared to metal tips but are less rigid and tend to have more conical tips that may aid insertion into the cavae. It is important to note that while venous cannulae are graded based on OD, flow capabilities will vary due to these factors. The choice of venous cannulae, as with all cannulae, usually comes down to surgeon judgment. Examples of pediatric venous cannulae are shown in Figure 1.18. The pediatric perfusionist must be familiar with available options for outlier cases such as difficult cannulations or LSVC, hepatic, axillary, or femoral cannulation.

In adult coronary artery surgery, the right atrium is most commonly cannulated with a single dual-stage venous cannula. This right atrial cannula has a low likelihood of occluding its side port holes since the right atrium is considerably larger than the cannula itself. In congenital cardiac surgery, patients are more likely to require bicaval cannulation since intracardiac work is more common. It is important to note that a larger venous cannula inserted into a cava does not always result in a higher expected flow rate. If the side port holes are occluded, venous return can be limited to a single hole at the tip, which may decrease overall performance. Furthermore, the IVC cannula must not occlude the hepatic venous drainage. The Eustachian valve is also of consideration for IVC cannulation since it can hinder proper cannula placement.

Table 1.9 lists recommended cannulae sizing for Medtronic DLP right-angle metal-tip cannulae and is based on one institution's experience using gravity drainage. The

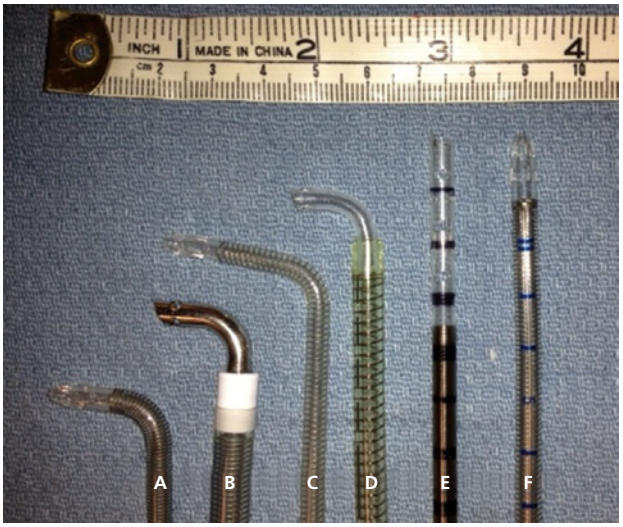


Figure 1.18 Select pediatric venous cannulae. *A—14 Fr. Terumo Tenderflow right angle PVC tip. B—14 Fr. Medtronic DLP right angle metal tip. C—12 Fr. Medtronic DLP right angle PVC tip. D—12 Fr. Edwards Lifesciences Thin-Flex right angle plastic tip. E—14 Fr. Medtronic Bio-Medicus straight with multiple side port holes. F—14 Fr. Medtronic DLP malleable straight PVC tip.* (*See insert for color representation of the figure.*)

Table 1.9 Venous cannulae for bicaval cannulation.

Medtronic DLP angled metal tip venous cannulae		
Weight (kg)	**SVC (Fr.)**	**IVC (Fr.)**
<3	12	12
3–6	12	14
6–8	12	16
8–12	14	16
12–16	14	18
16–22	16	18
22–30	16	20
30–34	18	20
34–46	18	20 or 22
46–58	20	22
58–75	20	24
75–100	22	24
>100	22 or 24	24 or 28

Table 1.10 Water chart flow rates for select venous cannulae.

Venous Cannula Size (Fr.)	Manufacturer	Water chart flow (LPM)
12	Medtronic DLP angled metal	0.8
	Medtronic DLP angled PVC	0.6
	Medtronic DLP straight PVC	0.6
	Terumo angled tenderflow	0.7
14	Medtronic DLP angled metal	1.6
	Medtronic DLP angled PVC	0.8
	Medtronic DLP straight PVC	0.8
	Terumo angled tenderflow	1.1
16	Medtronic DLP angled metal	1.9
	Medtronic DLP angled PVC	1.4
	Medtronic DLP straight PVC	1.4
	Terumo angled tenderflow	1.5
18	Medtronic DLP angled metal	2.6
	Medtronic DLP angled PVC	1.8
	Medtronic DLP straight PVC	1.8
	Terumo angled tenderflow	2.0
20	Medtronic DLP angled metal	3.0
	Medtronic DLP angled PVC	2.4
	Medtronic DLP straight PVC	2.4
	Terumo angled tenderflow	2.4
22	Medtronic DLP angled metal	3.7
	Medtronic DLP angled PVC	3.1
	Medtronic DLP straight PVC	3.1
	Terumo angled tenderflow	2.7
24	Medtronic DLP angled metal	4.2
	Medtronic DLP angled PVC	3.5
	Medtronic DLP straight PVC	3.5
	Terumo angled tenderflow	4.1
28	Medtronic DLP angled metal	5.3
	Medtronic DLP angled PVC	5.2
	Medtronic DLP straight PVC	5.2
	Terumo angled tenderflow	Cannula size not available

Table 1.11 Water chart flow rates for select femoral venous cannulae.

Weight (kg)	Size (Fr.)	Brand	Water chart flow (LPM)
<4.5	10	Edwards Lifesciences Fem-Flex	0.8
	10	Biomedicus, 7.5″	0.7
4.5–10	12	Edwards Lifesciences Fem-Flex	1.25
	12	Biomedicus, 7.5″	1.25
10–14	14	Edwards Lifesciences Fem-Flex	1.80
	14	Biomedicus, 7.5″	1.75
14–28	15	Biomedicus, 30″	1.2
28–40	17	Biomedicus, 30″	1.7
40–55	19	Biomedicus, 30″	2.5
55–75	21	Biomedicus, 30″	3.0
75–90	23	Biomedicus, 30″	3.8
>90	25	Biomedicus, 30″	4.7
	27	Biomedicus, 30″	5.5
	29	Biomedicus, 30″	>6.0

- Kinetic-assisted venous drainage or VAVD increases achievable flows.
- Significant differences in femoral vessel size can be seen in similarly sized patients. Cannula used will primarily be determined by vessel size.
- Water chart flows may overestimate achievable flows with femoral cannulation in particular since the venous line may end up being longer to accommodate connection to the bypass circuit.
- For quick reference in emergent situations, femoral venous cannula size is frequently 2 Fr. greater than the femoral arterial size (15 Fr. femoral arterial cannula will result in a 17 Fr. femoral venous cannula).
- The achievable flows with a single femoral venous cannula using gravity drainage are rarely able to support full flows on bypass.
- Femoral cannulation may simply support circulation until full cardiac exposure is achieved and central cannulation is possible. Alternately, an SVC or right atrial cannula may be added to the femoral venous circuit to allow for full flow on bypass.

use of vacuum or kinetic assist would allow for the use of smaller venous cannulae (and possibly a smaller venous line). As discussed, cannulae are rated by pressure drop and the achievable flows within an accepted pressure range. Table 1.9 is based on weight and there is overlap between the ranges. Experience-based sizing based on weight has been shown to be more useful than water flow charts, especially during emergent cannulations. The overlap encourages the clinician to consider other patient variables when contemplating stepping up or down the size of a cannula.

Tables 1.10 and 1.11 exemplify some of the options and flow differences between manufacturers and venous cannulae designs. Referencing more comprehensive charts such as these is less than ideal in emergent situations. And, these flow charts are based on water flows and so may not accurately reflect achievable flows in a bypass system. A weight based chart works well in most instances, while water flow charts may be helpful in unique situations. The water charts are useful when determining cannula size for single venous and alternate cannulation strategies. These charts are offered only as an example since different perfusion programs will have different bypass line lengths and equipment, which may affect cannulae performance. Additionally, cannula position importantly affects achievable flows. Experience-based charts should be an excellent guide resulting in proper sizing with rare exception. Ultimately, the patient's anatomy, anticipated flows on bypass, surgical needs, and surgeon judgment determine venous cannulae size.

Arterial cannulae

Arterial cannulae are graded based on their French size. Table 1.12 lists an example of weight-based arterial cannulae sizing for central aortic cannulation. Generally speaking, as with venous cannulae, arterial cannulae French size refers to the outer diameter (OD). It is generally accepted that the pressure drop across an arterial cannula should be limited to 100 mmHg to prevent excessive jetting, shear forces and damage to the formed elements of the blood. An arterial

Table 1.12 Arterial cannulae for central aortic cannulation.

Weight (kg)	Size (Fr.)	Brand	Water chart flow (LPM)
<2.5	6	Medtronic DLP	0.4
2.5–4.5	8	Biomedicus	0.7
4.5–10	10	Biomedicus	1.3
10–14	12	Biomedicus	2.1
14–28	14	Biomedicus	2.8
25–35	16	Medtronic One-Piece	3.8
28–35	18	Medtronic EOPA CAP	2.6
35–50	18	Medtronic EOPA	4.8
50–75	20	Medtronic EOPA CAP	4.1
75–100	20	Medtronic EOPA	5.7
75–90	22	Medtronic EOPA CAP	5.4
>90	22	Medtronic EOPA	>6
>90	24	Medtronic EOPA	>6

cannula with a larger internal lumen will have a higher flow capacity at the acceptable pressure drop. However, the cannula cannot be so large that it significantly blocks flow around it which is particularly important for ascending aortic cannulation. Native cardiac output ideally is not significantly impacted with arterial cannulation, particularly during the initiation, rewarming, and weaning phases of bypass or when the patient is cannulated but not on bypass. Additionally, the length of the cannula with smallest diameter, and whether the cannula is compressible (likelihood to kink or be restricted by the purse strings), will impact performance. Figure 1.19 depicts some commonly

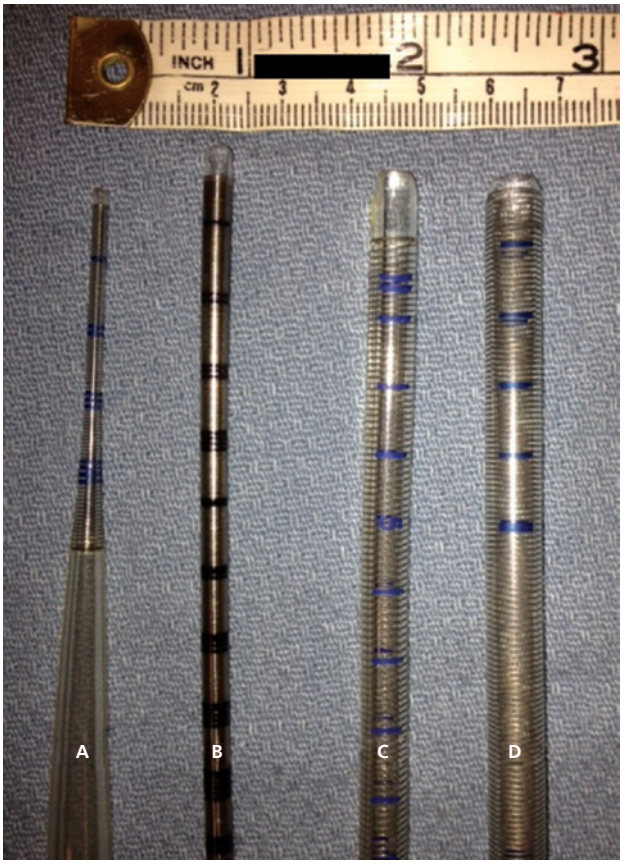


Figure 1.19 Select arterial cannulae for central cannulation. *A-6 Fr. Medtronic DLP One Piece. B-12 Fr. Medtronic Bio-Medicus. C-20 Fr. Medtronic EOPA. D-22 Fr. Medtronic EOPA CAP.* (*See insert for color representation of the figure.*)

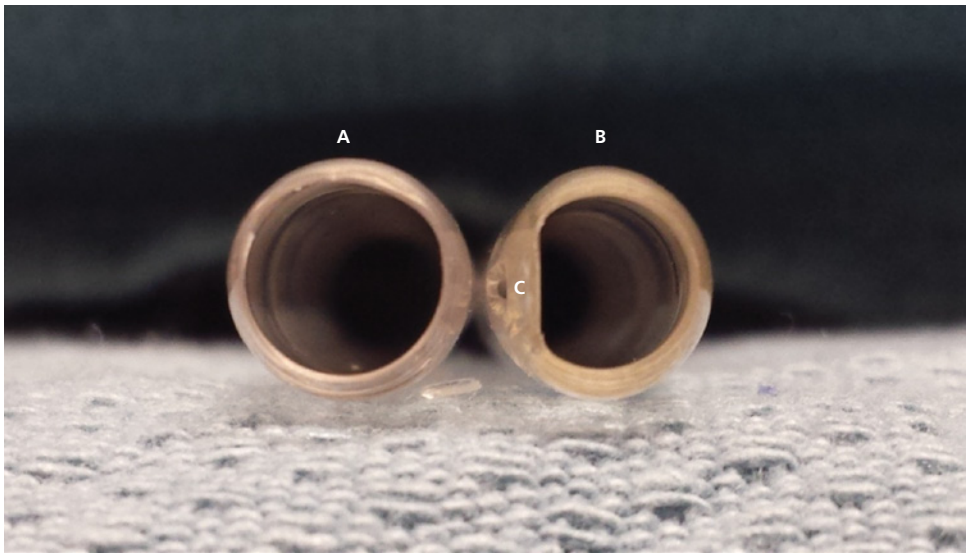


Figure 1.20 Luminal variation between types of arterial cannulae. *A-Standard round lumen for a 22 Fr. Medtronic arterial cannula. B-D-shaped lumen for a 22 Fr. Medtronic arterial cannula with central aortic pressure (CAP) monitoring capability. C-Left of "C" is the central aortic pressure monitoring port.* (*See insert for color representation of the figure.*)

Table 1.13 Arterial cannulae for femoral cannulation.

Weight (kg)	Size (Fr.)	Brand	Water chart flow (LPM)
<4.5	8	Edwards Lifesciences Fem-Flex	0.8
4.5–10	10	Edwards Lifesciences Fem-Flex	1.3
10–14	12	Edwards Lifesciences Fem-Flex	2.0
14–28	14	Edwards Lifesciences Fem-Flex	2.4
28–40	15	Biomedicus	3.0
40–55	17	Biomedicus	4.0
55–75	19	Biomedicus	5.3
>75	21	Biomedicus	6.5

*Most arterial cannulae can be used for femoral cannulation with a cut down procedure.
†The cannulae listed here come with appropriate supplies for Seldinger technique insertion.

used arterial cannulae. Furthermore, some arterial cannulae have a port at the tip which can be used to monitor central aortic pressure (not cannula lumen pressure) before and after bypass. If the monitoring line for this feature runs through the lumen, overall flow capabilities will be lower since the effective lumen size is decreased. This means that two arterial cannulae with the same French size (same OD) can have different flow capabilities (see Figure 1.20). Some arterial cannulae come with a side port allowing for stopcock placement. This is useful for monitoring patient arterial pressure post bypass (on bypass, it only reflects bypass circuit pressure) and as a means for deairing during cannulation (prebypass and emergently). Finally, Table 1.13 lists weight-based arterial cannula options for femoral cannulation.

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CHAPTER 2

Priming the bypass circuit

Prime constituents

The prime constituents and priming process for congenital cardiac surgery vary considerably among institutions. Adult cardiac surgery programs most often use primes that are referred to as "clear," "crystalloid," or "colloidal," whereas pediatric centers are more likely to use "blood" primes. This difference is primarily due to the desire to maintain a defined hematocrit on bypass and the greater relative hemodilution younger patients experience due to the priming volume of a bypass circuit. An average adult with a 5 L circulating blood volume, a 1 L clear bypass prime, and an initial hematocrit of 36 will be hemodiluted on bypass down to a hematocrit of 30%. A 3.6 kg neonate with a circulating blood volume of 0.3 L, a 0.36 L prime, and an initial hematocrit of 40 would be hemodiluted on bypass down to an unacceptably low hematocrit of 18% (*see section "Hematocrit Management" in Chapter 3*). Since the circuit prime volume tends to more closely approximate the patient's own blood volume in pediatric cases, pediatric patients are hemodiluted to a greater degree, and blood is more commonly used in the prime.

Clear primes can be defined as any prime solution that does not use blood. Crystalloid and colloidal primes are types of clear primes. Crystalloid solutions range from basic normal saline (0.9% NaCl) to balanced electrolyte solutions such as Plasma-Lyte A 7.4 (Baxter Healthcare, Deerfield, IL), Isolyte S 7.4 (B Braun Medical, Irvine, CA), and Normosol-R (Hospira, Inc., Lake Forest, IL). Lactated Ringer's solution (Hospira, Inc., Lake Forest, IL) and Hartmann's Solution (MacoPharma, Middlesex, UK) are crystalloid solutions uncommonly used for bypass primes since they contain a lactate load. Balanced electrolyte solutions mirror the concentrations of many normal plasma electrolytes. The properties of these solutions are listed in Table 2.1 for comparison.

Colloidal primes are similar to crystalloid primes in that they do not use blood. However, they differ in that protein or starches are included to more closely match the body's normal oncotic pressure. Albumin (5 and 25%), hydroxethyl starch ("Hextend," BioTime, Berkeley, CA and "Hespan," B Braun Medical, Irvine, CA), Gelofusine (B Braun Medical, Irvine, CA), and Dextran (Sigma-Aldric, St. Louis, MO) are colloidal solutions that may be used. It may appear on the surface that colloidal priming solutions are superior to crystalloid solutions since they have the advantage of more physiologic oncotic pressures and, perhaps, will result in less edema during and after cardiopulmonary bypass. However, it is important to note that they carry an increased risk of anaphylactoid reactions on bypass and are more expensive without adequate proof of effectiveness [1–6]. Additionally, commercially prepared colloidal starch solutions have been shown to carry an increased risk of causing renal impairment and post bypass coagulopathy [3]. Furthermore, if there is capillary leak on bypass, these solutions may more easily extravasate and actually promote edema. Albumin has been used as a colloidal priming solution to add oncotic pressure and additionally as a method to coat the bypass circuit and oxygenator. Theoretically, an albumin coating may preserve platelet function and increase functional oxygenator life by helping to prevent "capillary leak" within a microporous membrane oxygenator [7]. However, studies justifying albumin in the prime and its associated cost are lacking.

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Table 2.1 Crystalloid solution properties.

	Plasma-Lyte A 7.4	Isolyte S 7.4	Normosol-R	Lactated Ringer's	Hartmann's solution
Sodium (mEq/L)	140	141	140	130	131
Potassium (mEq/L)	5	5	5	4	5
Magnesium (mEq/L)	3	3	3	0	0
Chloride (mEq/L)	98	98	98	109	111
Acetate (mEq/L)	27	27	27	0	0
Gluconate (mEq/L)	23	23	23	0	0
Phosphate (mEq/L)	0	1	0	0	0
Lactate (mEq/L)	0	0	0	28	29
Calcium (mEq/L)	0	0	0	1.5–3.0	2
Calculated osmolarity (mOsm/L)	294	295	295	273	279
pH range	6.5–8.0	7.0–7.8	6.5–7.6	6.0–7.5 (though lactate metabolized by the liver can be alkalinizing)	5.0–7.0 (though lactate metabolized by the liver can be alkalinizing)

Additionally, surface modifying agents (SMAs) for bypass tubing and oxygenators, when incorporated, likely provide these same proposed benefits in a more cost-effective manner [8]. Pediatric facilities commonly use whole blood or a combination of packed red blood cells and plasma (commonly referred to as reconstituted whole blood, though technically inaccurate), which negates the need for albumin as a separate additive to the prime.

A blood prime consists, at least in part, of whole blood or packed red blood cells. However, it is important to note that the bypass circuit must first be primed with a clear solution to appropriately deair and prebypass filter (to remove potential particulate emboli created during the manufacturing process). Once deaired and with the prebypass filter removed, blood may be added and a portion of the clear prime removed. The amount of clear versus blood prime is normally determined based on the patient's diagnosis, prebypass hematocrit, desired hematocrit on bypass, hematocrit of the product to be used, and the bypass circuit volume.

Common prime constituents

- Plasma-Lyte A 7.4, Isolyte S 7.4, or Normosol-R
- Heparin
- Calcium chloride or calcium gluconate
- Sodium bicarbonate
- A broad spectrum antibiotic
- Methylprednisolone for select case types and patients
- Mannitol
- Albumin
- Whole blood or packed red blood cells and plasma, as required to maintain an acceptable hematocrit once on bypass. Some centers use red blood cells only, with or without albumin.

The bypass prime is ideally determined on an individual patient basis with the goal of acceptable hemodilution, acid–base balance, and electrolyte composition. Antibiotics are normally given before surgical incision and may also be added to the circuit at the onset of bypass to account for the circuit's dilutional effect on its plasma levels. Steroids may be given on bypass to attenuate the inflammatory response to bypass (especially for moderate-to-profound hypothermia cases) and for heart or lung transplant patients, as an immunosuppressive agent. Mannitol may be added to the circuit prime, but some centers only administer it at the onset of rewarming or before cross clamp removal. Dosing at this time prevents the possibility of an excessively high prime osmolarity causing neurologic injury [9]. Institutional prime protocols vary greatly, but all aim to provide reasonably physiologic prime values with acceptable hemodilution.

Steps for priming

Where and when to prime?

The bypass circuitry is selected based on the patient's cardiac diagnosis, body surface area, planned surgery, and expected maximum pump flow for bypass. The circuit is ideally primed with crystalloid before the patient arrives in the operating room. This practice allows the perfusionist to verify that the heart–lung machine components and OR equipment are working. It also results in immediate availability of circulatory support in the event of a patient crisis. Blood priming can commence once the perfusionist has a sense of what the hematocrit will be immediately before

bypass. This estimation is important when determining whether or not to use a blood prime; and if so, in what quantity. First though, the oxygenator must be inspected for defects and then water tested.

Water testing the oxygenator's integrated heat exchanger

The oxygenator's integrated heat exchanger must be water tested prior to the addition of crystalloid solutions. It is important to run water through the integrated heat exchange system at a flow and pressure comparable to the operating room parameters (if done outside of the OR) and to inspect for leaks to the blood compartment. A small water-to-blood leak on bypass may be difficult to diagnose (at least initially) and can cause significant patient harm (metabolic acidosis with lysis of blood elements), so it is imperative not to skip this step. If the blood compartment remains water free after several minutes of water testing, carbon dioxide flushing and crystalloid priming may ensue. If water enters the blood compartment, a replacement oxygenator should be prepared and the defective unit should be sent to the manufacturer for evaluation. Additionally, if possible, the affected oxygenator manufacturing lot should be quarantined until the manufacturer evaluates the heat exchanger defect.

Carbon dioxide flush

Once the circuit has been assembled and the oxygenator water tested, it is common to flush the entire system with carbon dioxide (CO_2). A carbon dioxide flush of the entire system fills the tubing and components with CO_2 and inhibits bacterial growth in case of inadvertent contamination [17, 18]. To note, the CO_2 flush may be coincident with water testing of the oxygenator's heat exchanger. A CO_2 flush ideally displaces air (primarily nitrogen) in the circuit with carbon dioxide, which is more soluble than nitrogen. Neurocognitive dysfunction has been associated with the gaseous embolic load created during cardiopulmonary bypass [10–16]. Carbon dioxide flushing of the circuit (as well as the surgical field) is a simple method of reducing nitrogenous gaseous microemboli (GME) from bypass circuits and arterial line filters (ALFs) in particular [10, 11, 19]. After completion of the clear prime process, the circuit is ideally free of macro- and microbubbles. CO_2 flushing greatly aids this. If bubbles do remain (hidden in the oxygenator, ALF or tubing, etc.), they should preferentially be carbon dioxide, and thus should readily dissolve with time [11, 19].

The inside of the bypass circuit should be sterile. A carbon dioxide flush of the entire system fills the tubing and components with CO_2 and inhibits bacterial growth in case of inadvertent contamination. This is more of a benefit when a circuit is assembled several hours in advance of crystalloid priming. This advantage of CO_2 flushing is often overlooked with oxygenator manufacturers increasingly indicating in their product information-for-use (IFU) pamphlets that CO_2 flushing is optional or unnecessary.

One caveat to CO_2 flushing is worth mentioning. It is important that when the system is CO_2 flushed and then primed with fluid, there must be a method to off-gas excess carbon dioxide. An oxygenator naturally does this, and the perfusionist can verify an appropriate PCO_2 value in the prime before placing a patient on bypass with an in-line blood gas monitor or by sending a sample to the lab. Closed recirculating cardioplegia systems do not have a "natural" way to off-gas CO_2; and if flushed with CO_2 before a fluid prime, the cardioplegia solution could have an extremely high PCO_2 with a resultant marked respiratory acidosis.

Crystalloid prime

After water testing and CO_2 flushing, the circuit is primed with a crystalloid solution only (and always with constant flow—never with pulsatile flow that can entrain air from the gas phase within microporous membrane oxygenators). Blood, buffering agent, and medications are added later. The crystalloid solution is added to the venous reservoir and is ideally circulated through the oxygenator, ALF, and a recirculation line back to the venous reservoir. It is preferred to exclude the arterial and venous lines (AV loop) until the primary components are properly deaired. The perfusionist follows the flow of the prime, circulated at or above the manufacturer recommended priming flow rate, from the venous reservoir through the circuit and inspects and taps all tubing, connectors, and components along the way. Tapping components helps move trapped air along. All components need to be visually inspected. This may require removal of the oxygenator (just the oxygenation membrane and not the reservoir) from its holder to fully inspect the entire cylinder. However, not all units allow this. External ALFs will also require removal, inversion, tapping, and inspection. High flow and tapping of all components during crystalloid priming are essential components for perfusion safety.

Once the primary circuit components are deaired, the arterial head occlusion is set. Arterial head occlusion can be set by meniscus drop or via a pressure method. The meniscus method has the perfusionist bring the crystalloid prime solution in the bypass arterial limb 75 cm above

the venous reservoir fluid level. Then, the arterial head occlusion is adjusted for each roller to be minimally non-occlusive at the same location within the raceway. This usually translates to a meniscus drop of 1 cm/min. Alternately, the system may be pressurized to a static level. Then, the occlusion for each roller of the arterial head can be backed off to allow for a pressure drop of 0.25–1 mmHg/s. There are other variations to these techniques. Either way, it is important to remember that a fully occlusive head will cause physical trauma to the blood, while an under occlusive head will allow for retrograde flow (and inaccurate system display flow on bypass) and possible physical trauma as well due to sloshing in the raceway [20, 21].

After the occlusion is set, the prime can be circulated through the AV loop, and that part of the circuit should be tapped and inspected to ensure it is properly deaired. The crystalloid prime should also be circulated through an in-line prebypass filter (0.2–10 μm) at this time. A prebypass filter helps ensure any residual particles from the tubing, oxygenator, and ALF production processes get removed before placing the patient on bypass. The prebypass filter is always removed before blood priming and before bypass.

Some patients will receive a crystalloid-only prime since it results in an acceptable dilutional hematocrit on bypass. The prime for these patients should be buffered (usually with sodium bicarbonate) and be kept recirculating. Constant flow through the circuit at the desired temperature for commencement of bypass is important in preventing the formation of gas bubbles in the circuit. A warm room, especially during anesthetic induction and access line placement, can promote gas coming out of solution if a prime is not recirculated for an extended period of time. A brief period of increased system flow before the division of the arteriovenous loop at the surgical field helps ensure air-free AV tubing.

Some centers try to maintain a certain heparin level in the prime and for any fluids added to the bypass circuit. With this constant heparin concentration technique, the crystalloid prime solution contains heparin in a standardized amount (i.e., 3–5 units heparin per milliliter prime). Any blood added to the prime or during bypass also contains heparin in the standardized concentration with this method.

Other additives, such as antibiotic, steroids, and antifibrinolytics, can be added just before the onset of bypass once the AV loop has been divided. It is not advisable to add these to the circuit sooner since part of the prime solution may be suctioned to the wall or lost to the field or pump suckers once the bypass lines are divided.

Blood prime

A blood prime may consist of traditional whole blood, reconstituted whole blood (red blood cells and plasma) or just red blood cells. The product chosen will be determined by institutional practice. It is common to use "fresh" blood (<7–10 days old) for neonatal and infant patients. Many centers also irradiate the blood for these patients or for all of their patients. Some clinicians believe that the clotting factors present in whole or reconstituted whole blood are important in mitigating bleeding issues after bypass.

The prebypass filter must be removed before blood priming. It is also preferable to add heparin to the blood unit before administering any volume to the bypass circuit. This step is especially important if calcium chloride or calcium gluconate is used in the final prime since calcium chelating anticoagulants (i.e., citrate) can be overwhelmed and allow blood to clot in the bypass circuit if an additional anticoagulant is not present.

There are several values needed to make the determination as to whether blood is needed in the pump prime and how much: institutional and\or surgeon protocol, pump prime volume, estimated patient blood volume, patient hematocrit, and the hematocrit of the product used for priming. The dilutional hematocrit on bypass if a clear prime is used can be calculated with the following equation:

$$\text{Dilutional Hct on bypass} = \frac{\begin{array}{c}(\text{patient blood volume}) \\ \times(\text{patient Hct})\end{array}}{\begin{array}{c}(\text{patient blood volume} \\ + \text{circuit prime volume})\end{array}}$$

Institutional and/or surgeon protocol will determine the lowest acceptable dilutional hematocrit at the onset of bypass. If the predicted hematocrit is less than this desired value, blood must be added to the prime. A simple equation to estimate the amount of blood required in the pump prime is as follows:

$$\text{prime blood needed (ml)} = \frac{(\text{PBV} + \text{circuit volume})(\text{desired bypass Hct}) - (\text{PBV})\,(\text{patient Hct})}{\text{Hct of prime blood product}}$$

PBV=Patient Blood Volume

- A negative value would indicate that the bypass hematocrit will be above the desired value with a crystalloid prime.
- If the volume of blood needed exceeds the prime volume, there are three options:
  - If whole blood is being used, prebypass ultrafiltration to raise the hematocrit can be employed. If RBCs are used, the hematocrit cannot practically be raised above the hematocrit in the blood bank unit.
  - Additional blood may be administered to the circuit resulting in a higher than normal prime volume (the reservoir level will be higher than normal at the onset of bypass).
  - Blood may be administered early in the bypass run with early and aggressive hemoconcentration until the desired hematocrit is achieved.
- Patient blood volume may be estimated with the values in Table 2.2.
  - Patients with significant valve regurgitation or single-ventricle physiology in particular may have higher blood volumes than predicted with these estimates.
- Circuit volume is calculated from components used.
- Patient hematocrit is determined from an intraoperative specimen with consideration given to how much time will elapse before bypass (during extended dissection, the patient may require crystalloid or colloid volume administration).
- The hematocrit of the blood product used is based on institutional practice:
  - Whole blood may be estimated to have a hematocrit of 35–40%.
  - Red blood cells may be estimated to have a hematocrit of 65–75%.

If a blood prime is needed, specific steps are taken to replace a portion of the crystalloid prime with blood. First, the venous reservoir should be emptied to near its outlet. This is often done by clamping the venous line inlet to the venous reservoir and expelling volume from a side port proximal to this clamp. Then, the desired amount of blood is added to the venous reservoir using the reservoir gradations to quantify the blood volume added. Next, the blood is slowly transfused through the circuit while expelling crystalloid volume from the venous port proximal to the venous clamp. This is continued until the reservoir level reaches the desired prebypass level. This process allows the perfusionist to visualize the flow of blood through the circuit while performing an additional inspection for leaks and flow abnormalities in the circuit. Since this process is generally slow, owing to the low flow capabilities of the venous side port stopcock, the perfusionist can also take the time to ensure that positional elements of the circuit are correctly attached.

Table 2.2 Patient estimated blood volume.

Patient weight (kg)	Blood volume (mL/kg)
<10	85
10 to <20	80
20 to <30	75
30 to <40	70
>40	65

- Venous saturation/hematocrit cell or in-line blood gas monitor should be oriented down to minimize interference from possible venous line air.
- Venous sampling ports should exit down to minimize interference from possible venous line air.
- Post ALF ports should be secured higher than their circuit connections. This is to prevent micro air embolism due to abrupt stoppage of pump flow due to servoregulation, which can allow trapped air to rise from these connections into the circuit arterial limb. The circuit is ideally free of air but micro air may be present in these connections (especially stopcocks) and may be impossible to visualize. Therefore, it is a safe practice to secure these lines higher than their circuit attachment points. Then, there is no chance of air rising into the circuit at these points during abrupt flow stoppages. This is an underappreciated safety issue with oxygenators that contain integrated ALFs. Ports that were formerly pre-ALF are now post-ALF!
- If an external ALF is used, it should be secured below the level of the reservoir. If an external ALF is positioned higher than the reservoir, an open purge line (failed one way valve) or a cracked ALF can allow air to enter the filter during no flow situations (i.e., when the lines are clamped before or after bypass or when stoppages occur due to servoregulation).

Once the proper amount of blood has been added to the prime and the proper amount of crystalloid has been expelled from the circuit, the venous line clamp is removed and the prime is recirculated. A buffer such as sodium bicarbonate and a calcium source are then added based on institutional protocol. The recirculated prime can be gassed at this time as well. Ideally, this buffered prime is also run through an in-line blood gas monitoring device and circuit values can be adjusted. These values should be checked in the lab, and this check also allows for calibration of the

in-line blood gas unit. It is important that when adding calcium and sodium bicarbonate to a prime (clear or blood) that they be added cautiously. If added together into a cardiotomy reservoir, precipitation of calcium carbonate/sodium chloride can occur. This is because oftentimes, the cardiotomy reservoir, to which drugs are added, is not in the prime blood flow path, and so the two medications can mix in a near-static setting (some flow to the cardiotomy may occur through a filter purge line or manifold line). The venous reservoir is in the recirculating blood flow path. If there is concern for precipitation, it may be prudent to add sodium bicarbonate and calcium to the venous reservoir or one to the venous reservoir and the other to the cardiotomy reservoir. Additionally to note is that clear primes are more apt to see precipitate in an alkaline environment that may be present when the oxygenator sweep gas does not contain CO_2 before bypass.

Finally, circuit prime values should ideally be within ranges specified by department protocol. If not, adjustments should be made. Gas and pH values can be adjusted rather quickly by changing the gas source, FiO_2, and/or sweep rate. Electrolyte value adjustment may require prebypass ultrafiltration that takes more time (*see section "PBUF" in Chapter 3*). It is important to remember that when using a blood prime to effect a prescribed bypass hematocrit, it is also important to provide the patient with a reasonable glucose, blood gas, acid–base, and electrolyte profile.

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CHAPTER 3

The bypass plan

The perfusionist must be familiar with the patient's original diagnosis, catheterization lab interventions, previous surgeries, and current needs when developing the bypass plan. The following are major considerations for management of the patient during cardiopulmonary bypass. Please refer to *Chapter 6* for defect-specific considerations.

Communication agreement for case

The care team must have a reasonably standardized communication plan for the bypass run [1–3]. This includes standardizing to closed-loop communication for certain anesthesia, surgical, and cardiopulmonary bypass (CPB) commands, notifications, and/or concerns. Each communication should be verbally acknowledged per the team standard (either repeated back or with another affirmation) to ensure the information is properly delivered and handled. This technique is essential for bypass safety and is used at most centers to announce checkpoints during the case.

Common bypass-related checkpoints where closed-loop communication among the team is essential include the following:

- Heparin in
- ACT running
- Bypass lines clamped and ready for division at the field
- Pump suckers on (with suckers and vent line tested under saline at field)
- Arterial cannula in (continuity from bypass arterial limb to arterial cannula verified)
- Venous cannula(e) in (continuity from bypass venous limb to venous cannula(e) verified)
- Is the perfusionist ready to go on CPB?
- ACT and/or heparin level adequate
- Going on bypass
- Taking venous return
- Full-flow/target index achieved
- Ventilator off (after ejection ceases)
- Anesthetic gas on at pump
- Cooling started to the target temperature
- Cardioplegia system ready
- LV vent in and on
- Caval tapes applied, drainage remains adequate
- Cross clamp on
- Cardioplegia running
- Cardioplegia dose delivered, cardioplegia flow off or recirculating
- Cross-clamp interval (Q20 min or other standard)
- Caval tapes off
- LV vent off
- Cross clamp off
- LV vent on
- Rewarming patient
- Index flow during warming as prescribed by the surgeon
- SVC cannula clamped/removed (with bicaval cannulation)
- Ventilator on

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

- Drips running, dosages
- Pacer settings, if applicable
- Pump flow during weaning for desired filling pressures
- LV vent clamped/removed
- Anesthesia team ready for separation from bypass
- Perfusionist ready for separation from bypass
- Pump off
- Surgical repair verified by imaging
- Modified ultrafiltration started/completed
- Venous cannula(e) removed
- Protamine started
- Pump suckers turned off
- Protamine completed
- Is it acceptable to remove the arterial cannula (are the volume requirements manageable by the anesthesiologist without use of the heart–lung machine)?
- Arterial cannula out
- Surgical instrument and needle counts verified
- Chest closure

Anticoagulation management

Anticoagulation is required for cardiopulmonary bypass to prevent clotting of blood in both the patient and bypass circuit. Clots in the patient or circuit can lead to the inability of the bypass oxygenator and pump to properly function, patient embolic events, stroke, and death. Two primary methods of monitoring and managing anticoagulation exist for cardiopulmonary bypass patients with congenital heart disease; the activated clotting time (ACT) and heparin concentration management (Hepcon HMS, Medtronic Inc., Minneapolis, MN).

Anticoagulation is required to prevent, at least in a significant way, activation of the coagulation pathway. Blood exposed to nonendothelial surfaces, such as bypass tubing, activates factor XII (fXII). Blood exposed to tissue factor (TF) activates factor VII (fVII), allowing a complex to form between the two while also activating platelets. Both mechanisms of activation lead to a series of common proteolytic reactions that work toward clot formation on activated surfaces. Classical descriptions of the coagulation system referred to *intrinsic* (foreign surface contact), *extrinsic* (exposure to damaged tissue), and *common* pathways in the process of clot formation, which are shown in Figure 3.1 [4]. Table 3.1 lists the coagulation factor numbers and names commonly used for them.

Modern descriptions of the coagulation system refer to a single "pathway" with phases of *initiation, amplification, propagation*, and *stabilization*. These reactions are localized to different cell surfaces. Hoffman and Monroe referred to this description of the coagulation system as the "cell-based model of hemostasis" [4]. This cell-based theory, based on their work and that of others, does not view the coagulation system as a cascade with different pathways. Rather, it describes coagulation as having one primary activator (TF) which initiates a process of concurrent reactions on different cell surfaces. If the primary procoagulant stimulus is adequate, additional cell surface-based reactions occur, particularly on the platelets. These reactions promote thrombin formation, which eventually leads to a stabilized fibrin clot.

In this cell-based model, initiation begins with activation of tissue factor, platelets and Factor VII which when combined produce a small amount of thrombin. Amplification continues with more thrombin formation and additional activation of platelets. Platelet activation and binding helps localize the clot formation. Concurrently, propagation increases thrombin and fibrin formation with the stabilization phase ultimately resulting in an organized fibrin clot. This more modern explanation of coagulation better exemplifies how there are numerous and simultaneous parallel processes working toward properly localized formation of a cross-linked fibrin clot. Figure 3.2 illustrates this model.

In consideration of these coagulation processes, the goal of bypass anticoagulant management then is to ultimately prevent thrombin production, which leads to fibrin clot formation. Efforts to minimize overall circuit exposure, to limit the nonphysiologic appearance of nonendothelial

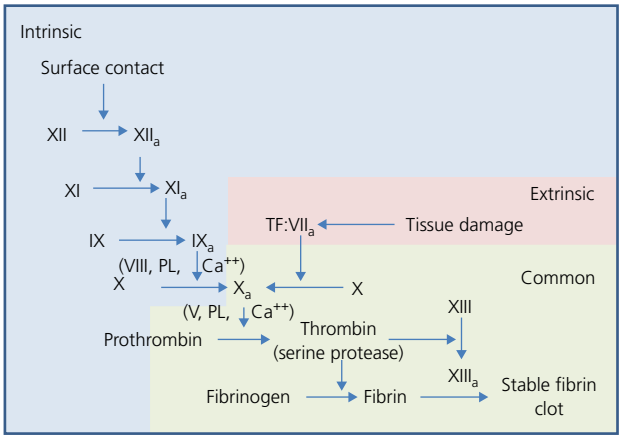


Figure 3.1 Classical description of the blood coagulation pathways. VII-stabile factor, XII-Hageman factor, IX-Christmas factor, XI-plasma thromboplastin, XIII-fibrin stabilizing factor, Ca++ ionized calcium, PL-platelets, TF-tissue factor. http://commons.wikimedia.org/wiki/File:Classical_blood_coagulation_pathway.png. Reproduced with permission from Graham Beards and Wikimedia Commons. (*See insert for color representation of the figure.*)

Table 3.1 Coagulation factors.

Primary coagulation factors	Also known as
Platelets	Thrombocytes
I	Fibrinogen
II	Prothrombin
III	Tissue factor, tissue thromboplastin, platelet tissue factor
IV	Calcium
V	Proaccelerin, labile factor
VI	This factor is no longer assigned. It was at one time but then discovered to be the activated form of factor V.
VII	Proconvertin, stabile factor, prothrombin conversion accelerator
VIII	Antihemophilic factor A
IX	Christmas factor, antihemophilic factor B, plasma thromboplastin component
X	Stuart-Prower factor, prothrombinase
XI	Plasma thromboplastin antecedent
XII	Hageman factor
XIII	Fibrin stabilizing factor

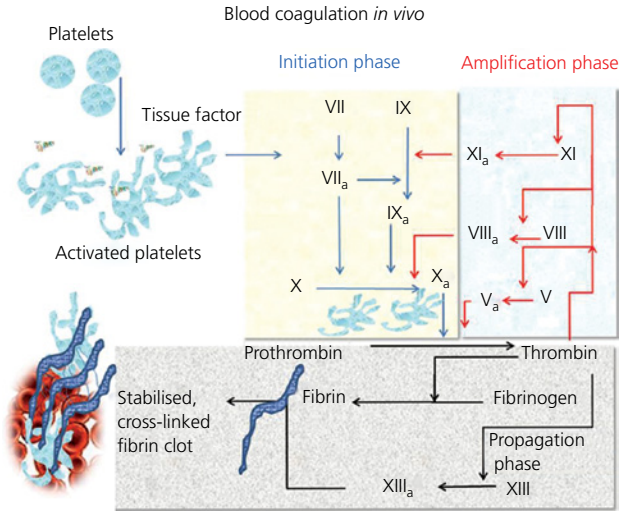


Figure 3.2 Cell-based model of hemostasis *in vivo*. http://commons.wikimedia.org/wiki/File:Coagulation_in_vivo.png. Reproduced with permission from Graham Beards and Wikimedia Commons. (*See insert for color representation of the figure.*)

circuit surfaces, and to limit activation of procoagulant factors contribute to this goal. Heparin is the primary medication administered to provide anticoagulation for cardiopulmonary bypass.

Minimizing circuit exposure

Tremendous strides have been made over the past decade in regards to decreasing circuit prime volumes for pediatric perfusion cases. This decrease in prime volume parallels an overall decrease in exposure of the blood to nonendothelial circuit surfaces. As a case in point, infants commonly were exposed to bypass circuit primes of 400–550 mL just 10–15 years ago. Today, these same patients routinely receive prime volumes of 180–300 mL. Oxygenator and arterial line filter design have played a major role, especially when the two are integrated. Techniques including vacuum-assisted venous drainage, retrograde autologous priming, venous autologous priming, and maximizing the use of smaller tubing diameter lines have also played a part. However, it should be noted that activation of procoagulant systems, and the inflammatory response to bypass, is still primarily due to the use of cardiotomy suction.

Limiting the nonphysiologic appearance of bypass circuitry

Perfusion circuits have been improved with surface-modifying agents (SMAs) although their use is not universal. The goal of SMAs is to limit the nonphysiologic appearance of nonendothelial surfaces that activate procoagulant systems, cause protein denaturation, platelet activation, and platelet adhesion. Heparin (Medtronic Trillium, Carmeda BioActive Surface) and other SMAs (Terumo X-coating, Sorin P.h.i.s.i.o., and Jostra Safeline/Bioline/Softline) are currently in use. All can be bonded to the bypass tubing and components during manufacturing. The presence of heparin chains on bypass circuit surfaces decreases activation of the coagulation systems by binding antithrombin and thrombin, which decreases the circulating concentration of procoagulant thrombin. Other SMAs create a super smooth surface which, similar to heparin coatings, limits activation of the coagulation system and the inflammatory response to bypass. All SMA circuits decrease platelet activation and better maintain platelet function after bypass. Platelet activation is undesirable because it allows for conformational changes whereby they expose phospholipids allowing procoagulant attachment and further platelet-to-platelet activation, attachment, and clumping.

Limiting activation of procoagulant factors

Antithrombin III is a protein that normally inactivates several parts of the coagulation cascade. It achieves this by binding procoagulant thrombin and preventing activation of factors VII and X in particular. Procoagulant and anticoagulant factors are normally appropriately balanced in the body. The balance normally shifts toward coagulation with exposure to nonendothelial surfaces, tissue factor, and activation of platelets and factors VII and X. This balance is shifted in favor of anticoagulation by several means, including the presence of heparin. Heparin efficacy requires an adequate concentration of antithrombin III. Heparin dramatically increases the effectiveness of antithrombin III which prevents or limits the activation of these factors. Heparin binds with antithrombin III and thrombin to form a complex that inhibits activation of platelets, factors VII, X, and XIII (fibrin stabilizing factor), and limits free thrombin. This inhibition leads to anticoagulation and is commonly quantified with a prolongation of the ACT. Heparin is normally given in bolus form before bypass with additional doses on bypass as needed.

Monitoring the effect and dosing of heparin

Monitoring of the ACT and/or heparin concentration in the blood are methods used for managing anticoagulation on bypass. The traditional activated clotting time simply measures the number of seconds required for a blood clot to form when blood is activated in a sampling tube. An activating agent such as celite (silica, glass beads, and kaolin are

also used as activators) shifts the hemostatic balance in favor of procoagulation in the sampling tube. A specific volume of blood is added to a tube and warmed to 37 °C and continually rotated in a test well with a freely moving magnetic element resting on the bottom, or 6 o'clock position, within the tube. When a sufficient clot forms, the rotation of the sampling tube brings the magnetic element that is within the clot to the top, or 12 o'clock position, signaling the timer to stop counting. An alternate system quantifies clot formation by measuring the impedance of a plunger in a blood sample. Both methods result in a real-time counting of the number of seconds required for organized clot formation. An ACT of greater than 400–480 s is commonly considered to be therapeutic for cardiopulmonary bypass. Other methods of ACT measurement focus on blood flow within a capillary tube and/or use proprietary algorithms to accelerate the test in some way. An accelerated test reports results in seconds like the standard ACT but can complete the tests in less time. For example, a reported ACT value of 480 s may only take 300 s to calculate. There are numerous factors that affect all of these tests, and these include room temperature (and the unit's ability to adequately warm the sample), hematocrit, platelet count, and a precise sample size without air entrained (if in a capillary tube). Traditional real-time ACT measurement is used nearly universally at centers performing bypass for congenital cardiac patients. Some centers rely solely on the ACT, while others also monitor heparin concentration in the blood [5–7].

Monitoring the heparin concentration is thought to be more predictive of how well activation of the clotting system has been suppressed. This may be especially true during hypothermia and hemodilution, both of which are common for congenital cardiac surgery patients. A primary supporting theory for heparin concentration monitoring is that while hypothermia and hemodilution (dilution of clotting factors including platelets) are enough to elevate the ACT, procoagulant systems may still be at work. This can lead to consumption of clotting factors (resulting in an elevated ACT) during bypass with excessive bleeding after bypass (consumptive coagulopathy). The only way to more assuredly prevent activation then is to provide an adequate concentration of heparin in the blood to maximize the effect of the heparin–antithrombin–thrombin complex. Heparin concentration in whole blood, like the ACT, can be measured with a point-of-care device. The heparin concentration test works off of the principle that heparin is neutralized by protamine in a known ratio with too much or too little protamine resulting in a prolonged clotting time. By exposing blood samples to several different concentrations of protamine and detecting which sample clots first, the heparin concentration can be determined. Heparin concentrations are generally run in the 2.5–6 units/mL range for cardiopulmonary bypass. To note, neonates and infants have immature hemostatic systems and notably lower thrombin and antithrombin levels. While this may manifest as apparent heparin resistance after heparin bolus (ACT <400–480 s), the reduced potential for thrombin to activate procoagulant factors in this setting may actually mean that lower ACT values and lower heparin concentrations provide an adequate anticoagulant effect [7].

Anticoagulation management will vary by institution, but it usually includes maintaining the ACT above 400–480 s and, if measured, a heparin concentration of 2.5–6 units/mL. Samples are usually drawn after heparin bolus and every 20 min on bypass with an increased sampling rate during rewarming or when values are marginal. ACT and heparin concentration management will depend in part on the equipment selected for bypass and whether SMAs are utilized.

Heparin dosing is commonly empiric for congenital cardiac cases. A prebypass heparin bolus of 300–400 units/kg is commonly used to achieve the target prebypass ACT value, but more may be required to achieve the target heparin concentration. Figure 3.3 illustrates the use of a heparin–dose response (HDR) curve for, theoretically, more precise heparin dosing. Additional heparin boluses to raise the ACT and/or heparin concentration are frequently 50–150 IU/kg and will vary by patient factors, operative progress, and clinician practice.

As noted previously, an adequate antithrombin III level is required for heparin to adequately prolong the ACT. Normal functional AT III levels are usually stated in the range of 80–120%. Inadequate levels are generally defined as less than 60%. Newborns have less functional AT III with levels approaching adult values by 6 months of age. If AT III levels are known to be inadequate preoperatively, fresh plasma, reconstituted whole blood, or commercially available AT III concentrate (from human sources or recombinant from animal sources) may be given to facilitate achieving an adequate ACT value prebypass.

Finally, it is important to clarify that while some clinicians equate more heparin with more bleeding, the opposite may in fact be true. Higher heparin doses may better attenuate procoagulant systems and prevent consumption of factors during bypass [8]. This would support less bleeding after bypass once protamine is administered.

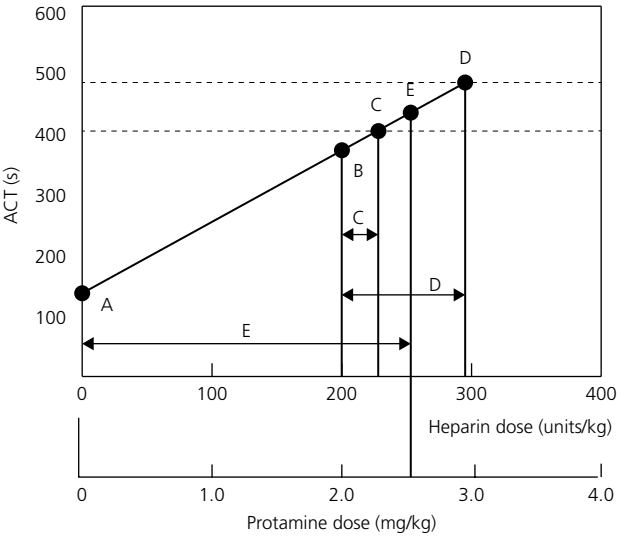


Figure 3.3 Heparin dose response curve for quantifying heparin and protamine doses. Graph of a heparin (and protamine) dosing algorithm. In the graph, the control activated clotting time (ACT) is shown as point A, and the ACT resulting from an initial heparin bolus of 200 units/kg is shown in point B. The line connecting A and B then is extrapolated and a desired ACT is selected. Point C represents the intersection between this line and a target ACT of 400 s, theoretically requiring an additional heparin dose represented by the difference between points C and B on the horizontal axis (arrow C). Similarly, to achieve an ACT of 480 s (higher horizontal dotted line intersecting the ACT versus heparin dose line at point D), one would administer the additional heparin dose represented by arrow D. To estimate heparin level and calculate protamine dose at the time of heparin neutralization, the most recently measured ACT value is plotted on the dose–response line (point E in the example). The heparin level present theoretically is represented by the difference between point E and point A on the horizontal axis (arrow E). The protamine dose required to neutralize the remaining heparin then may be calculated. Protamine 1.0 mg/kg is administered for every 100 units/kg of heparin present. From Hensley FA, Gravlee GP (2003). A Practical Approach to Cardiac Anesthesia. Philadelphia: Wolters Kluwer. © 2012, Wolters Kluwer Health.

Reversing the anticoagulation effect of heparin

The anticoagulant effect of heparin is reversed with protamine. Protamine forms a complex with heparin which neutralizes its effect. There exists a 1:1 ratio for the complex formed between these two substances (some sources report a ratio of 1.1:1). This equates to a reversal dose of essentially 1 mg protamine for each milligram of heparin (100–140 IU) estimated to be in the patient's circulatory system.

There exist numerous variables when calculating the amount of heparin in the patient which needs to be reversed. There are three commonly used and accepted methods for dosing protamine: the use of empiric, HDR curve, and protamine titration dosing.

Empiric protamine dosing

Empiric dosing has the clinician give 1 mg (range 1–1.5 mg) protamine per 100 units of heparin estimated to be in the patient. Since heparin is commonly dosed before bypass at 300–400 units/kg, the empiric dose can also be stated as 3–4 mg protamine/kg. The estimate of heparin remaining in the patient may additionally consider the total amount of heparin administered during cardiopulmonary bypass. However, most clinicians simply give a higher protamine dose (i.e., 4 mg/kg) to account for additional heparin on bypass and protamine loss to bleeding during administration.

Protamine administration is commonly followed up with an ACT. An ACT that approaches baseline and/or a normal value at the very least indicates that protamine (and not some other medication) was administered and has had an effect (and didn't end up on the floor via a line disconnect!). However, one must account for the potential anticoagulant effects of thrombocytopenia, platelet dysfunction, hypothermia, and the dilution of other procoagulant factors which may be present after bypass. It is important therefore to consider that after bypass, the ACT may not be at baseline or at a near normal value for reasons other than the presence of heparin.

HDR curve protamine dosing

An HDR curve, shown in Figure 3.3, is another way to calculate the protamine dose. An HDR utilizes the data points of baseline ACT and the ACT after heparin bolus for bypass. These values provide a line and slope, which can then be used after bypass. The ACT value before protamine administration can be plotted on this line and the residual units of heparin in the patient can be calculated. Protamine is dosed based on this residual heparin value. Again, an ACT after protamine administration is measured and understood in the context that the ACT may not be at baseline or at a near normal value for reasons other than the presence of heparin.

Protamine titration dosing

Protamine titration dosing is another option for the reversal of heparin. A point-of-care device such as the Medtronic Hepcon HMS is used. This machine estimates the amount of heparin present in a post-bypass sample by exposing it to varying amounts of protamine. The protamine and blood sample that clots first is assumed to best match the heparin concentration. This concentration value, along with an estimated patient circulating blood volume can then be used to determine the amount of heparin present, which must be neutralized. Protamine is then administered at a

predetermined ratio (e.g., 1–1.5 mg protamine/100 units of heparin). Again, an ACT after protamine administration is measured and understood in the context that the ACT may not be at baseline or at a near normal value for reasons other than the presence of heparin.

Protamine administration

The administration rate of protamine is another important consideration [9]. Rapid administration of protamine may cause hypotension, anaphylaxis, pulmonary hypertension, and pulmonary edema. Protamine is preferably given at a rate that prevents adverse effects. Administration rate varies by individual practice with administration over 5–10 min generally considered prudent.

Finally, accidental administration of protamine on bypass has been reported [10]. This is likely an underreported complication that can have lethal outcomes. Protamine immediately and effectively reverses heparin's anticoagulant effect. Therefore, the protamine dose should not be drawn up in advance. Protamine vials must be kept well away from the medications normally administered by the anesthesiologist and perfusionist. It is a safe practice to draw up the protamine dose only and immediately before administration.

Blood gas management

Blood gas management during cardiopulmonary bypass for patients with congenital heart defects primarily varies in regards to carbon dioxide management and oxygenation strategy. Most often, when clinicians refer to "blood gas management," they are talking more specifically about carbon dioxide management and not necessarily oxygenation strategy, which is also important. "Blood gas management" and "carbon dioxide management" therefore tend to be interchangeable terms. The various strategies have advantages and disadvantages based on patient diagnosis and age. Adherents to respective strategies cite neurologic and myocardial outcome studies as a basis for their approach.

Blood gas management for congenital cardiac surgery must consider the fact that a majority of cardiopulmonary bypass in this population is conducted at some degree of hypothermia. Hypothermia shifts the oxyhemoglobin dissociation curve to the left resulting in oxygen being bound to hemoglobin more tightly and, therefore, less likely to be released at the tissue level. Hypothermia causes vasoconstriction and increases the viscosity of blood, which theoretically may impair microvasculature blood flow. These concerns have increasingly important consequences under deep hypothermia or with low flow bypass. Blood gas management strategies address these concerns. Since carbon dioxide management varies in regard to $PaCO_2$ value correction for temperatures less than 37 °C, it is important to note that there are no differences between strategies at normothermia. The primary techniques for blood gas management include alpha-stat versus pH-stat and normoxic versus hyperoxic, with crossover options for both.

Carbon dioxide management

Alpha-stat blood gas management

The term "alpha-stat" refers to keeping the ionized (alpha) imidiazole of histidine constant (static). The imidazole of histidine is an intracellular protein buffer involved in maintaining intracellular pH. Maintaining the natural balance of hydrogen ions (H^+) relative to hydroxide ions (OH^-) with the imidazole of histidine and other systems is important in maintaining enzymatic function and cellular life. Simply put, the alpha-stat strategy aims to maintain intracellular function by maintaining intracellular pH (not blood pH) and enzymatic function. This is an important consideration with increasing levels of hypothermia. The solubility of gases increases with hypothermia which leads to overall alkalinity in the blood since less gaseous CO_2 is present. This alkalinity is acceptable since the ratio of hydrogen and hydroxide ions is maintained intracellularly by histidine. Enzymatic function is preserved. Maintenance of this electrochemical neutrality is also important in regards to cerebral blood flow. Preservation of enzymatic function and other cellular and vascular systems additionally maintains cerebral blood flow autoregulation where flow is matched to metabolic need. This may be particular important in older patients with acquired vascular disease who are more prone to embolic events and downstream sequelae on bypass, especially in the cerebral vasculature. Alpha-stat limits cerebral blood flow to that which is necessary to meet metabolic demand and no more. The risk of embolic events is thus kept to a minimum by limiting potential delivery of emboli. However, at deep hypothermic temperatures of 22 °C or less, autoregulation is likely lost [11].

Alpha-stat blood gas management in a practical sense involves maintaining the PCO_2 at 40 mmHg and the pH at 7.40 as measured at 37 °C, regardless of patient body temperature. This method is most commonly achieved on bypass with air and oxygen delivered through a gas blender and graduated flow meter. The perfusionist adjusts the FiO_2 per protocol for independent management of PaO_2 on bypass. The gas flow rate is adjusted, much like the ventilator rate and tidal volume for intubated patients, to affect the $PaCO_2$. An inverse relationship between sweep flow and $PaCO_2$ exists where an increased gas flow rate generally decreases $PaCO_2$ and vice versa. This is the predominant blood gas strategy for adult cardiac surgery patients with acquired cardiac disease.

pH-stat blood gas management

A blood gas machine analyzes all samples at a uniform 37 °C. Blood gas analyzers by default use the alpha-stat method since results are reported at 37 °C regardless of patient temperature. Of course, most patients having blood gas values quantified are normothermic. The term "pH-stat" refers to keeping the blood pH constant (static) at the prescribed level of hypothermia. This technique simply adjusts the blood gas values (PaO_2 and $PaCO_2$) using corrective equations based on the temperature the blood exists within the patient. The addition of carbon dioxide is generally needed to achieve pH-stat management because of the increased solubility of gas at hypothermia. Increased gas solubility decreases the gaseous form in the blood (i.e., decreased $PaCO_2$) while increasing the

dissolved form (i.e., increased plasma CO_2). This dissolved CO_2 either remains as CO_2 in the blood or interacts with water (H_2O) to equilibrate with carbonic acid (H_2CO_3). The carbonic acid may then dissociate into hydrogen ions (H^+) and bicarbonate (HCO_3^-) and then back to H_2O and CO_2. These are the primary reactions responsible for the carbonic–acid–bicarbonate buffer system in the body.

Marked differences in temperature corrected $PaCO_2$ and pH values result as compared to the temperature uncorrected values at hypothermia. During deep hypothermia, it is common and in fact the aim, to see temperature corrected pH-stat $PaCO_2$ and pH values of 40 mmHg and 7.40, respectively. These deep hypothermia values, as shown in Table 3.2, correlate with measurements of $PaCO_2$ greater than 80 mmHg and pH less than 7.20 if evaluated in the temperature-uncorrected alpha-stat method.

On the surface, it may seem quite undesirable to have such a high $PaCO_2$ and low pH (for the uncorrected values). However, there are important considerations for patients on hypothermic cardiopulmonary bypass. Carbon dioxide shifts the oxyhemoglobin dissociation curve to the right, which helps counteract the left shift caused by hypothermia. This is thought to increase the amount of oxygen available to the tissues. Carbon dioxide is also a powerful cerebral vasodilator and pulmonary vasculature vasoconstrictor. Patients receiving hypothermic bypass with pH-stat management may therefore benefit from improved off-loading of oxygen at the tissue level, increased cerebral blood flow and improved cooling of the brain. A decreased run off of blood to the pulmonary circulation is especially important for patients with known or even undiagnosed aortopulmonary collaterals. Additionally, pH-stat management, with its low pH at deep hypothermia, suppresses the cerebral metabolic rate which may prolong oxygen availability during periods of circulatory arrest [12].

Several animal lab and human studies were performed at Boston Children's Hospital which looked at blood gas management techniques under pediatric models of hypothermic bypass, with and without circulatory arrest. The conclusions for the human trials can be summarized such that pH-stat blood gas management resulted in decreased inotropic support in the immediate post-bypass period, decreased incidence of epileptiform activity, decreased time to first EEG activity, decreased early postoperative mortality and improved developmental outcome scores [13–15]. These findings supported earlier animal studies which found that greater cerebral blood flow during cooling for circulatory arrest resulted in improved recovery of cerebral ATP and intracellular pH with improved cerebral oxygen delivery and uptake [16, 17].

pH-stat blood gas management in a practical sense involves maintaining the $PaCO_2$ and pH within normal limits as measured at the patient temperature. This is usually quantified with a reliable temperature probe location, which approximates the brain temperature. A tympanic temperature may be ideal, but the venous blood temperature may be more practical to use for temperature correction since it is not affected by user placement and other artifact as much and is therefore more reliable.

pH-stat is most commonly achieved on bypass with air and oxygen delivered through a gas blender and graduated flow meter with the addition of low flow 100% CO_2. An electronic gas blender and flow meter is another option. Other methods of CO_2 delivery to the oxygenator gas have been developed since errors in systems using 100% CO_2 can quickly result in massive respiratory acidosis. A carbogen delivery system is one alternative method where carbogen (2–10% CO_2 with balance oxygen) may be used as the sole gas source [18]. As with all blood gas management strategies, an in-line blood gas monitor that supplies real time data is essential for tight control of values during bypass. To note, the carbogen method limits one to the hyperoxic oxygen strategy since blended gas is not utilized. However, carbogen (2–10% $CO_2$with balance oxygen) and carbair (2–10% CO_2 with balance air) supplied to a traditional gas blender with equal CO_2 percentages allows one to have independent control of oxygen and carbon dioxide. Additionally, it may be possible to provide pH-stat management simply by using very low sweep gas flow on bypass. However, this method is not recommended since the sweep flow required may be outside of the oxygenator manufacturer's recommended range.

The gas flow rate during pH-stat management is adjusted in the traditional manner to affect the $PaCO_2$where the inverse relationship between the flow of blended oxygen or carbogen and $PaCO_2$ still exists. If 100% CO_2 is utilized,

Table 3.2 Typical pH-stat blood gas values with equivalent alpha-stat values.

	30 °C pH-stat (alpha-stat equivalent)	24 °C pH-stat (alpha-stat equivalent)	18 °C pH-stat (alpha-stat equivalent)
pH	7.40 (7.29)	7.40 (7.22)	7.40 (7.15)
$PaCO_2$ mmHg	40 (54)	40 (70)	40 (91)
PaO_2 mmHg normoxic strategy	200 (230)	200 (255)	200 (277)
PaO_2 mmHg hyperoxic strategy	500 (549)	500 (594)	500 (642)

a direct relationship between its flow and $PaCO_2$ exists where increased CO_2 flow increases $PaCO_2$. It is important to note that when carbogen systems are utilized in neonates and for deep hypothermia cases in particular, this sweep flow relationship may change. In some cases, an increased carbogen gas flow may be needed to load more carbon dioxide onto the blood (even after maximizing the available carbogen CO_2 concentration). pH-stat blood gas management is the predominant strategy for congenital cardiac surgery patients [19–22].

Finally, it is worth noting that patients who chronically retain carbon dioxide have a cerebral blood flow normalized to their elevated $PaCO_2$. This should be taken into consideration regardless of the blood gas strategy utilized. NIRS monitoring may be particularly helpful for these cases. Lower than expected NIRS values may be improved by raising the $PaCO_2$ to the patient's normal level.

Crossover strategy for carbon dioxide management

Some clinicians believe that the primary benefits of pH-stat blood gas management are increased cerebral blood flow and improved brain cooling. Additionally, the primary benefits of alpha-stat blood gas management are preservation of intracellular buffering and enzymatic function. These divergent strategies are most pronounced during deep hypothermia with or without circulatory arrest. Some institutions try to extract the benefit of both techniques by using pH-stat to cool the patient with a crossover to alpha-stat for maintenance at low flow bypass or prior to planned circulatory arrest. Animal studies have suggested this strategy to be optimal [23]. However, prospective randomized human clinical trials supporting this crossover strategy have not been performed. And, pH-stat with hyperoxia has been shown to produce the least amount of acid suggesting it best attenuates anaerobic metabolism [24, 25].

Oxygenation strategy

The arguments concerning oxygenation strategy for patients with congenital cardiac defects center on reducing the introduction of air (primarily nitrogen) emboli into the bypass circuit, preventing oxygen toxicity, limiting reperfusion injury secondary to oxygen-free radical formation, maximizing oxygen delivery (especially during hypothermia), and with that, minimizing the potential for hypoxic brain injury [24–32]. The normoxic strategy has variants based on the FiO_2 used or desired PaO_2 on bypass. The normoxic strategy can be simply qualified as running the PaO_2 between 100 and 300 mmHg during bypass. The hyperoxic strategy, on the other hand, has its variants as well. It can be qualified as running the PaO_2 greater than 300 mmHg or with the highest FiO_2 possible within a system (i.e., 4% CO_2 with balance oxygen of 96%). On top of that, as with carbon dioxide management on bypass, a crossover strategy may be employed.

First in regards to oxygenation strategy is the issue of microembolic events on bypass. Several investigators have abundantly shown that even with current oxygenator and arterial line filter technology, there are still gaseous microemboli that are delivered to the patient [33–38]. These emboli may be generated due to venous line air, high vent and sucker flow, CVR-oxygenator design or characteristics of a microporous oxygenator which may allow ventilating gas source to be drawn across the oxygenator's micropores into the blood phase during abrupt events where blood flow stops and fluid momentum pulls gas across the membrane. Considering that these causes can only be minimized on bypass and not entirely eliminated, one argument for a hyperoxic strategy is that any emboli formed *in* the circuit will consist primarily of soluble oxygen (and possibly some carbon dioxide) which is readily absorbed and less likely to cause significant microvasculature blockages. Additionally, any air emboli *introduced into* the system (from the suckers, vent, or venous line) will exist in a system with the maximal gradient for off-gassing the relatively insoluble nitrogen both at the tissue level and at the oxygenator.

Second in regards to oxygenation strategy is the issue of oxygen toxicity on bypass. Oxygen toxicity may develop with extended exposure to high levels of oxygen. Retinopathy of prematurity (ROP) in neonates is a commonly noted disease related to exposure to high levels of oxygen. Toxicity can present in several ways and is due to the formation of oxygen-free radicals that can harm DNA, RNA, and cell structures and prevent normal vascular development. Oxygen-free radicals are naturally occurring and are normally countered enzymatically within the body. However, excess oxygen can theoretically overload the body's ability to scavenge oxygen radicals leading to cellular disruptions and possibly even cellular death. One must consider that oxygen toxicity is more likely to occur with extended exposure to high levels of oxygen over considerable hours or days. The theoretical risks of high oxygen exposure over an extended period of time must be weighed against the relatively short duration of bypass where significant physiologic perturbations occur, namely hemodilution, hypothermia, nonpulsatile flow, and possibly even circulatory arrest. Oxygen toxicity is not normally a consideration with the relatively short duration of bypass.

Third in regards to oxygenation strategy is the issue of ischemic reperfusion damage on bypass. This damage occurs when blood flow and oxygen are restored to an ischemic tissue bed. The sudden increase in oxygen and blood allows for oxidative damage secondary to formation of oxygen-free radicals. Whereas oxygen toxicity has free radical formation in an environment abnormal only in regards to oxygen content, ischemic reperfusion injury generally occurs in a hypoxic and acidotic environment. The end result is similar in that free radicals overload the cellular system and cause oxidative stress and damage. It can be argued that a hyperoxic strategy exacerbates this issue. However, one major qualification is important to note. Increased ischemic reperfusion injury may be possible in some tissue beds with a hyperoxic strategy, but the decreased hypoxic brain injury noted experimentally may supersede concerns for such. Clinician opinion varies between these competing concerns.

This leads to the final concern regarding oxygenation strategy which is overall hypoxic brain injury. An animal study of circulatory arrest by Nollert *et al*. concluded that a hyperoxic strategy decreases histologically assessed brain damage [32]. Dexter *et al*. showed that the brain utilizes more dissolved oxygen than hemoglobin-bound oxygen during deep hypothermic cardiopulmonary bypass [27]. And Pearl *et al*. demonstrated that a hyperoxic strategy, when paired with pH-stat blood gas management, produces significantly less acid than normoxic and alpha-stat strategies suggesting better attenuation of anaerobic metabolism [25]. Hypoxic brain injury is related to several factors including oxygen delivery and usage, hematocrit, temperature, perfused capillary density, cerebral blood flow, and PaO_2. Current evidence and research supports hyperoxia during cardiopulmonary bypass to prevent inadequate oxygen delivery whether HCA is utilized or not [30, 31]. Many centers have adopted a hyperoxic strategy for their congenital cardiac surgery practice. A hyperoxic strategy is beneficial in preventing or delaying the onset of neuronal hypoxia, especially during circulatory arrest. However, there have not been adequate prospective studies comparing oxygenation strategy for more routine congenital cardiac repairs (without deep hypothermia or circulatory arrest).

Normoxic technique

A normoxic strategy for bypass involves maintaining the arterial PaO_2 in the 100–300 mmHg range and varies by institution. The only equipment needed is an oxygen and air blender in the gas supply system. Tight control of the PaO_2 additionally requires the use of an in-line blood gas monitor since the PaO_2 may also vary with changes in patient temperature and pump flow. To note, as long as the sweep gas flow to the oxygenator is within manufacturer limits, it will not affect the PaO_2 to a significant degree. The PaO_2 is primarily manipulated with adjustments to the sweep gas FiO_2.

Hyperoxic technique

The hyperoxic strategy for bypass involves maintaining the arterial PaO_2 greater than 300 mmHg or simply managing the sweep gas with the highest FiO_2 possible within a system. This can be achieved with an oxygen and air blender as with the normoxic strategy. Again, the PaO_2 is simply manipulated with adjustments to the sweep gas FiO_2. However, some centers simply keep the blender at 100% oxygen at all times negating the need for FiO_2 manipulations. Alternately, a carbogen system may be employed. Here, 2–10% carbon dioxide with balance oxygen (i.e., 4% CO_2 with 96% O_2) is supplied as the sweep gas. This system does not allow for independent management of the FiO_2. Either method has the ability to achieve PaO_2 values of 300–700 mmHg. While the absolute oxygen content does not significantly increase after the hemoglobin is 100% saturated, the dissolved content increases and nonetheless may be important during hypothermic low flow perfusion and/or circulatory arrest [27].

Crossover technique for oxygenation strategy

Another concept to consider in regards to oxygenation strategy is a crossover technique. Some studies have shown beneficial effects of hyperoxic preconditioning of myocardium before bypass and cardioplegic arrest. If one believes hyperoxia on bypass has either an overall neutral effect or that it is detrimental, hyperoxia before bypass with normoxia on bypass is an option to consider for improved myocardial preservation, as suggested in a literature review by Young in the *Journal of Extracorporeal Technology* [29]. His review suggested that prebypass hyperoxia may provide protection against oxygen free radicals for cases that include cardioplegic arrest.

Carbair/Carbogen option

A carbogen system, as noted previously, is one method of safely providing pH-stat blood gas management. However, a carbogen system of 2–10% CO_2 with balance oxygen does not allow for independent management of the patient PaO_2. One way around this limitation is to utilize carbogen

(2–10% CO_2 with balance oxygen) and carbair (2–10% CO_2 with balance air) supplied to a traditional gas blender with equal CO_2 percentages. Such a system allows one to have independent control of oxygen and carbon dioxide. Here, the carbon dioxide percentage would be equal regardless of FiO_2 setting on the traditional gas blender. Adjustments to the blender would only affect the FiO_2 and ultimately the patient PaO_2. The traditional flow meter and blender will not be completely accurate with these nontraditional gas supplies but will be sufficient for trending while an in-line blood gas monitor verifies desired values. The carbair/carbogen option may be useful for those who utilize carbogen and desire PaO_2 manipulation on bypass, especially for cyanotic congenital cardiac patients for whom some clinicians believe the hyperoxic strategy may be particularly harmful.

Summary

Blood gas management strategies have increasingly important differences as one approaches deep hypothermic target temperatures. There is little significant difference in carbon dioxide strategies for temperatures greater than 30 °C. Adult cardiac surgery patients with acquired disease are more likely to suffer from sequelae of atherosclerotic plaques and emboli. For these reasons, alpha-stat blood gas management is preferred in adults since it maintains autoregulatory matching of cerebral blood flow and metabolism (above 22 °C) limiting the embolic load. Congenital cardiac surgery patients are more likely to suffer from sequelae related to uneven brain cooling and inadequate oxygen delivery. For these reasons, pH-stat blood gas management is preferred in congenital heart surgery since cerebral blood flow is optimized. The preferred technique for adult congenital heart surgery patients is debatable. Adult congenital patients with risk of sequelae secondary to atherosclerotic disease should likely be treated with the alpha-stat technique. Adult patients with known or possible aortopulmonary collaterals should likely be treated with the pH-stat technique. The choice of oxygenation strategy for congenital cardiac patients is critical in regards to oxygen availability and minimizing the potential for hypoxic brain injury. Oxygenation strategy is also an important consideration with gaseous microemboli on bypass [33–38]. All strategies discussed have merit, and the one employed will most often depend on institutional experience and surgeon preference.

Hematocrit management

Hematocrit management varies widely across institutions and especially within the realm of congenital heart surgery. Intensive care unit (ICU) management has a narrower spectrum of variation. The further one follows a patient retrospectively through their admission from the ICU back to the immediate preoperative period, the more variance you will find. This is due to the fact that some clinicians have standardized overall minimum acceptable hematocrits, while others specify minimum levels at given time periods (i.e., before bypass, on bypass, coming off bypass, before transfer to the ICU, in the ICU). Of course, the type of congenital heart disease a patient has and multiple other variables factor into the decision whether to transfuse a patient on or off cardiopulmonary bypass.

The most often cited studies concerning hematocrit management during cardiopulmonary bypass for congenital heart surgery occurred at Boston Children's Hospital and are commonly referred to as the Boston hematocrit trials. The findings can be summarized by looking at the conclusions of each of three key publications.

The first study by Jonas *et al.* compared hematocrit levels of 20% versus 30% for infants undergoing low-flow bypass for correction of TOF, d-TGA, truncus arteriosus, VSD, or CAVC [39]. The overall conclusion was that patients randomized to the 20% group had less desirable outcomes: lower nadir of cardiac index, higher lactate levels, and a greater increase in total body water postoperatively with lower Psychomotor Development Index scores at 1 year of age. Important to note for this trial is that after 1 year of study, ongoing enrollment for randomization was stopped when O'Brien-Fleming rules indicated that further randomization to the 20% arm was potentially harmful to patients.

The second study, by Newburger *et al.*, compared hematocrit levels of 25% versus 35% for infants undergoing low-flow bypass for correction of the same defects as the first study [40]. Here, intraoperative fluid accumulation was greater in the 25% group overall, while just the d-TGA patients in the 35% group had a shorter admission. However, the overall conclusion was that there were no significant differences in outcomes between the two groups.

The third publication, by Wypij *et al.*, was a meta-analysis of the first two [41]. There were several notable findings. First, the two initial studies looked at hematocrit levels not exactly as prescribed. The first study more precisely resulted in a hematocrit comparison of 22% versus 28%. This was due to the fact that it is difficult to maintain an exact hematocrit on bypass due to the many variables involved. Fluid administration pre-CPB and once on CPB (valve-testing solution, cardioplegia administration, unknown exact patient blood volume, etc.) vary the hematocrit making it difficult to precisely control. The second study more precisely resulted in a hematocrit comparison of 25% versus 33% for similar reasons. Perhaps the most important overall finding was that 1-year Psychomotor Development Index scores increased as the hematocrit approached 24% but showed no significant improvement above that value. This finding has led some clinicians to conclude that an on-bypass hematocrit of greater than or equal to 24% is acceptable. Others believe there should be some latitude for variance and prescribe a hematocrit of 28–30% to ensure the nadir hematocrit doesn't acutely fall below the 24% threshold. The jury is still out on the safe acceptable lower limit for hematocrit on bypass. It is important to consider that these studies did not look at single ventricle patients and the operations occurred at a single institution. Perfusion and surgical techniques vary considerably between institutions making it difficult to simply extrapolate the data.

It is unclear what lower level of hemodilution is acceptable on cardiopulmonary bypass for congenital heart surgery patients. Data suggest a bypass hematocrit of greater than or equal to 24% is desirable for certain congenital cardiac defects. The advent of continuous near-infrared spectroscopy (NIRS) and jugular venous saturation monitoring perhaps allows clinicians to tailor their transfusion thresholds for various patients based on real-time data in the varied settings of bypass (hypothermia, hemodilution, changes in flow, non-pulsatile perfusion, etc.) and off bypass (warm and ejecting). Several groups have demonstrated and recommend transfusion-free cardiac surgery, even in neonates and infants [42–45]. While this is certainly possible to achieve, long-term follow up and neurodevelopmental outcome comparisons are lacking. Further studies are needed to qualify that transfusion-free lower hematocrit strategies have comparable long-term outcomes as compared to protocols maintaining a hematocrit of greater than or equal to 24% during cardiopulmonary bypass for congenital cardiac surgery patients.

Practical application for bypass

The perfusionist must identify the target bypass hematocrit when determining the bypass plan. This will be based on institution protocol, anesthesiologist input and surgeon preference for each unique patient. Simple calculations are then used to determine the desired pump prime to achieve

the desired hematocrit once on bypass (*see Chapter 2*). Additionally, while on bypass, the perfusionist adjusts the hematocrit using ultrafiltration and with the addition of bank blood. Ultrafiltration can only be performed within certain limits since an adequate and safe venous reservoir level must be maintained. Bank blood administration is usually necessary to increase the hematocrit when no excess volume is present in the venous reservoir.

Blood pressure management

There is little consensus regarding optimal blood pressure on bypass for congenital cardiac surgery patients. All clinicians agree that blood pressure is an important factor in maintaining adequate cerebral and other end organ blood flow. Protection of the brain, in particular, is equally important as the cardiac repair. Perhaps then, discussing adequate cerebral blood flow is the proper place to begin.

$$\text{CBF (mL/min)} = \frac{\text{CPP (mmHg)}}{\text{CVR (mmHg/mL/min)}}$$

$$\text{CPP (mmHg)} = \text{MAP (mmHg)} - \text{CVP (mmHg)}$$

Cerebral blood flow (CBF) has been equated to cerebral perfusion pressure (CPP) divided by cerebral vascular resistance (CVR) [46]. Some clinicians simplify this statement by stating that *adequate* cerebral blood flow, with "reasonable" systemic blood pressure and flow on bypass (based on temperature), is present if there is an adequate cerebral perfusion pressure. The cerebral perfusion pressure can be simplified as patient mean arterial pressure (MAP) minus central venous pressure (CVP) [46]. This concept is easy to apply clinically (an elevated CVP with systemic hypotension results in a lowered CPP and must be addressed). However, cerebral blood flow is also affected by numerous bypass strategies. It can be agreed that adequate cerebral blood flow is providing enough oxygen (and nutrients) to meet the metabolic needs of the brain. The brain is the organ with the highest oxygen demand and is responsible for 20% of total oxygen uptake under normal conditions with only 15% of cardiac output [46]. The brain's relatively greater need for oxygen is still true on bypass. Cardiopulmonary bypass interventions do not change this relative need, but they do attempt to decrease overall oxygen requirements while increasing oxygen supply. The body's overall oxygen requirement is decreased with anesthesia, paralytics, and hypothermia. The brain's overall oxygen supply is increased with dilation of cerebral blood vessels with pH-stat blood gas management, a proper systemic vascular resistance aiding blood flow to the brain (blood pressure management), oxygenation strategy, hematocrit management, and in many cases simply by increasing the pump flow. Some factors that may work against the provision of adequate cerebral blood flow are advancing patient age and the associated risk for cerebrovascular disease, cannulation strategy (including arterial inflow location and venting of the aortic root) and whether preferential aortopulmonary blood flow exists (via collaterals or a surgical shunt) on bypass which may steal from effective cerebral circulation.

An adult's normal blood pressure is often cited as 120/80 mmHg giving a normal mean pressure of 93 mmHg. Blood pressure in children increases with age. Neonates commonly have a blood pressure of 70/40 mmHg giving a normal mean of 50 mmHg. These values climb toward the adult reference range by the teen years. In regards to blood pressure, it is known that the brain autoregulates blood flow to meet metabolic need. Different investigators cite varied blood pressure ranges for which this autoregulatory mechanism is maintained. It is likely in the 50–150 mmHg range for adults and 30–110 mmHg in children and infants [47]. Adults on bypass are commonly maintained with a mean blood pressure of 60–90 mmHg, which is within the autoregulatory range. An adult patient would be maintained at the higher end if cerebrovascular disease is suspected or present. Blood pressure on bypass is commonly 30–45 mmHg with neonates and 40–60 mmHg with children. It is worth noting that the common bypass ranges for blood pressure may be below normal mean blood pressure ranges but that they are within the autoregulatory range. This is acceptable with the bypass strategies mentioned which increase supply and decrease oxygen demand on bypass. Most notably, pH-stat blood gas management is commonly used in congenital cardiac surgery with hypothermia of 30 °C and less. This strategy increases $PaCO_2$ and dilates cerebral blood vessels rendering the cerebral blood flow to be more dependent on pump flow and patient blood pressure. Deep hypothermia itself inhibits cerebral autoregulation leaving cerebral blood flow, as with pH-stat management, to be pressure and flow dependent on bypass. Autoregulatory blood pressure ranges on bypass have been shown to be different than the traditional ranges [48–50]. And, patient $PaCO_2$ alone may affect the autoregulatory range [51].

It is worth noting that cerebral blood flow and patient blood pressure simply shouldn't be maximized on bypass. Cardiopulmonary bypass is not benign and microair likely is transmitted to all patients. Adults with vascular disease are at risk for embolic plaque dislodgement with downstream ischemia, which is especially of concern in the brain. Excess cerebral blood flow in neonates increases

Table 3.3 Typical target blood pressure ranges during cardiopulmonary bypass.

Patient age	Mean blood pressure range (mmHg)
<1 month	30–45
1–12 months	40–50
1–10 years	45–60
10–16 years	50–70
>16 years	60–90

the risk of hyperperfusion injury including edema and hemorrhage.

Blood pressure management varies among institutions and even by practitioners at the same institution. The near universal use of NIRS has allowed clinicians to more assuredly test the boundaries of blood pressure on bypass for given patient populations [52]. The cardiopulmonary bypass mean blood pressure ranges listed in Table 3.3 are based on clinical experience at select institutions.

Temperature management

Some degree of hypothermia is employed for the majority of cardiopulmonary bypass cases for congenital heart surgery. Simply put, hypothermia decreases the metabolic rate during bypass. This allows for decreased pump flows with perhaps better visualization at the surgical field (decreased collateral, Thebesian, and bronchial blood flow which normally returns to the left heart even during complete bypass). Additionally, lower pump flows result in a decrease in transmission of air emboli from imperfect bypass disposables (all are imperfect) and less blood trauma (due to a decreased air:blood interface with decreased sucker and vent flow). Hypothermia improves protection of all organs and provides an improved margin of safety if the pump must be stopped suddenly due to a pump alarm, inadvertent decannulation, or other surgical issue.

Temperature ranges

Temperature ranges for bypass have been classified by depth. Terms such as normal, tepid, mild, moderate, routine, deep, and profound have been used. Table 3.4 outlines reported range classifications.

Different authors have applied different ranges to these terms [53–55]. In a practical sense, a target temperature is prescribed by the surgeon and not a range. The perfusionist simply adjusts the heater–cooler system and surface cooling unit appropriately to effect the prescribed target temperature over a safe time period. The rate of cooling and warming is commonly limited with the temperature gradient allowed between the venous blood temperature and the oxygenator heat exchanger water temperature. Textbooks and oxygenator manufacturers often cite a maximum gradient between these values of 8–12 °C. The rationale is that a higher gradient increases the risk of gaseous microemboli forming either in the pump circuit or in the patient. This can be due to cold blood with dissolved gases entering a warm region where gas solubility is decreased. An animal study by Nollert *et al.* failed to show increased GME with higher than recommended temperature gradients [56]. However, most programs do follow manufacturer guidelines. Limiting the temperature gradient additionally helps provide for more even cooling and warming on bypass.

Table 3.4 Temperature range classifications.

Term	Range (°C)*
Normal/tepid	32–35
Mild	28–32
Routine/moderate	24–28
Deep	18–24
Profound	<18

*Range listing varies by author (overlapping intentional).

Patient temperature management for cardiopulmonary bypass involves the heat exchanger as well as surface cooling. Heat-exchange blankets under the patient, adjustable air temperature circulators around the patient, and room temperature may also be used to provide hypothermia and to assist with rewarming on cardiopulmonary bypass. Insulated ice packs around the head may also be used to maintain hypothermic brain temperatures [57]. This is most often done to prevent radiant warming during low flow and circulatory arrest cases. In addition to the benefits of pH-stat blood gas management to aid in cooling of the brain, medications such as phentolamine, nitroprusside, nitroglycerin, milrinone, and phenoxybenzamine may be used to decrease systemic vascular resistance in order to aid overall cooling and warming. Isoflurane may also be utilized in the oxygenator ventilating gas to both cause systemic and cerebral vasodilation while at the same time decreasing the cerebral metabolic rate of oxygen ($CMRO_2$) [58, 59]. Some clinicians allow for a lower blood pressure on bypass if it aids in cooling/warming and the NIRS and ECG are acceptable.

The concept of Q10

The temperature range specifics for levels of hypothermia may not be critical, but it is important to note that increasing levels of hypothermia do affect the body differently. The concept of Q10 is often cited in explaining this phenomenon. A Q10 value can be defined as the factor by which biologic functions are affected by a 10 °C temperature change. Many biologic reactions in the body have a Q10 of 2–3, which during warming on bypass means the metabolic rate double or triples when warming from 27 to 37 °C. Or, stated inversely, during cooling on bypass a temperature change from 37 °C down to 27 °C results in a metabolic rate of one-third to one-half of baseline. However, it is important to note that organ systems are not uniform in respect to their Q10 and that Q10 values increase with depth of hypothermia and are nonlinear [60–62]. The cerebral metabolic rate in particular is commonly cited as decreasing by 6–7% per degree Celsius, which is another way of considering the cerebral

effect of hypothermia [63]. Finally, infants have a higher overall Q10 relative to adults, which means that metabolic suppression is greater for a given level of hypothermia.

The change in metabolic rate is indirectly evident to the perfusionist during hypothermic bypass. The venous saturation and NIRS values normally increase during the cooling phase of bypass. Oftentimes, in congenital cardiac bypass cases, these values approach 100%. This, of course, is in light of a cardiac output (pump flow during total bypass) which is lower than the patient's normal cardiac output. A decreased metabolic rate with hypothermia decreases oxygen consumption and decreases the work which must be done by the artificial lung (membrane oxygenator) and normally results in increased SvO_2 and NIRS values but will vary depending on bypass strategies and anatomic variations. The effects of hypothermia on biologic processes are also evident with the activated clotting time (ACT). The ACT normally increases with hypothermia, in part due to changes in the proteins (clotting factors and platelets) involved with the clotting cascade. The ACT test is performed at 37° since the machine uniformly warms samples to this temperature but some of the changes hypothermia causes remain as evidenced with an elevated ACT.

Considering the benefits of hypothermia, the question then arises as to why all patients are not cooled to deep or profound levels hypothermia to take full advantage of its benefits. The answer to this primarily lies with the fact that not all biologic processes behave the same or linearly with increased levels of hypothermia. Cell membrane functions including osmosis (water transfer) and diffusion (electrolyte transfer), as well as enzymatic reactions, are increasingly disrupted with increased levels of hypothermia. The ability of the body to recovery from such perturbations varies. Therefore, it is up to the prescribing clinician to decide how to balance the benefits of hypothermia with its disadvantages relative to regaining homeostasis once rewarmed and separated from bypass.

Cooling and warming

Cooling and warming times are a function of many factors including patient size relative to heat exchanger size, heat exchanger efficiency, attributes of the thermocirculator, manufacturer limits on temperature gradients between the venous blood and water temperature, pump flow rates, and systemic vascular resistance. These factors are discussed in *section "Oxygenators" in Chapter 1* and *section "Flow rates, regional perfusion, and hypothermic circulatory arrest" in Chapter 3*.

The time allowed for cooling on bypass is usually not a safety issue. Most cardiopulmonary bypass cases for congenital cardiac surgery simply have the patient cooled at a rate well within recommendations for gradients since there is no urgency to reach the target core temperature. Circulatory arrest cases are quite different. Minimum safe cooling times to allow for homogenous brain cooling for DHCA are usually in the 20–25 min range [64]. Inadequate cooling time before DHCA has been associated with worse neurologic outcomes including choreoathetosis [65].

Warming is thought of in a much different manner. With rewarming a patient on bypass, there can be an urgency to reach normothermia to allow separation from bypass. Additionally, there is commonly a desire to reach a target warm temperature for separation to prevent after-drop and prevent hypothermia induced coagulopathy. While the studies on the neuroprotective effect of hypothermic bypass are inconclusive, there are clearly issues that must be considered when rewarming a patient in order to separate from bypass [66–69]. Rapid warming has been associated with a transient cerebral oxygen demand greater than supply [67]. Aggressive rewarming and hyperthermia have been linked to greater neuropsychologic dysfunction after bypass [68]. Further, oxygenator outlet temperature of greater than 37 °C has been correlated with acute kidney injury [69]. These potential complications must be considered in light of the fact that oxygenator temperature systems at normothermia are known to underreport outlet values by 0.5 °C or more [70]. This may not seem like much; but if temperatures greater than 37 °C lead to dysfunction and the perfusionist allows or is directed to increase the oxygenator outlet temperature to hasten rewarming, the risk of hyperthermic injury may be greater. The ideal rate of warming has not been identified. An animal study of DHCA illustrated well the balance which is desired when rewarming. This pig study by Alam *et al.* showed that a rewarming rate of 0.5 °C/min was superior to rewarming rates of either 0.25 or 1 °C/min [71]. Rewarming is likely ideal at a rate that doesn't unnecessarily prolong bypass (which is not a benign modality) and that doesn't lead to cerebral oxygen supply/demand issues due to hyperthermia [72]. Some authors report that overall rewarming should be limited. The neuroprotective effect of hypothermia was shown to be beneficial in an animal study by de Lange *et al.* only when coupled with warming to 35 °C

and maintenance of this mild hypothermia for 6 h postoperatively [72]. Much of the data concerning rewarming and hyperthermia has been gathered in animal and adult models that are more homogenous than congenital cardiac populations. But, considering that congenital cardiac patients have preexisting deficits and vulnerabilities [73], intuitively it seems prudent to avoid both excessive rewarming rates and hyperthermic oxygenator outlet temperatures (real and as displayed). And certainly, one must be cognizant that the varied patient temperature monitoring sites used during pediatric cardiopulmonary bypass likely underestimate actual core brain temperature [74].

Hypothermia for myocardial protection

Myocardial protection during open cardiac procedures nearly universally involves hypothermia. Cardioplegia solution is frequently delivered at 4–12 °C. The heart tolerates this depth of hypothermia very well and hypothermia may be particularly important with single dose cardioplegic strategies where the heart may not be perfused or redosed for 1–3 h. It is worth noting that the differing level of hypothermia between the body and the heart must be appreciated by the surgical team. If hypothermic myocardial arrest is employed, several factors may unexpectedly rewarm the heart. These include collateral flow to the heart (particularly to the left side), single venous cannulation allowing blood flow through the right atrium, and radiant warming due to the heart's proximity to major blood flow sources. This myocardial rewarming may influence the effectiveness of myocardial protection. Some clinicians use a thermal pad under the heart to prevent radiant warming of the myocardium. Saline slush on or around the heart may also be used to aid with myocardial hypothermia and protection.

Flow rates, regional perfusion, and hypothermic circulatory arrest

Flow rates

The range commonly cited for normal cardiac index is 2.5–4.2 L/min/m^2 [75, 76]. Patients with some forms of congenital cardiac disease exist with normal values which may well exceed 4.0 L/min/m^2 [77]. These normal values compare with common cardiopulmonary bypass values for normothermic congenital cardiac patients of 2.4–3.5 L/min/m^2. It could be argued that these patients on bypass are regularly being underperfused, but over 50 years of clinical experience has shown that the values commonly used for bypass are well tolerated in the context of bypass conditions [78]. Additionally in support of this, several studies in various models have shown that the use of real-time arteriovenous and near-infrared saturation monitoring and the monitoring of oxygen uptake, venous oxygen tension, and lactic acid production support the safe use of these lower flow rates during bypass [79–83]. Data do not currently exist outlining specific safe cardiopulmonary bypass flow ranges for various lesions [83]. In a practical sense, the perfusionist determines flow rates during cardiopulmonary bypass primarily by patient temperature, anatomic considerations, achievable venous return, and institutional/surgeon preferences, particularly for systemic vascular resistance and mean arterial blood pressure management. The initial target flow rate is normally achieved shortly after commencing bypass with verification of expected values for venous return, central venous pressure (CVP), NIRS and arteriovenous saturations to follow. Pump flow rates are then normally decreased in step with patient temperature and operative progress and needs. The use of real-time NIRS as a trending device is useful in guiding flow rate requirements during bypass and tolerance to temporary low flows required to improve visualization at the surgical field [84–88]. Table 3.5 lists commonly used index flows for bypass in congenital heart surgery. These values are estimates only as actual flow rates will depend on numerous real-time factors.

Table 3.5 Common index flows during cardiopulmonary bypass.

Patient temperature (°C)	Bypass flow rate (L/min/m^2)
≥35	2.4–3.5
32	2.2
30	2.0
28	1.8
26	1.6
24	1.4
22	1.2
20	1.0
<20	0.7

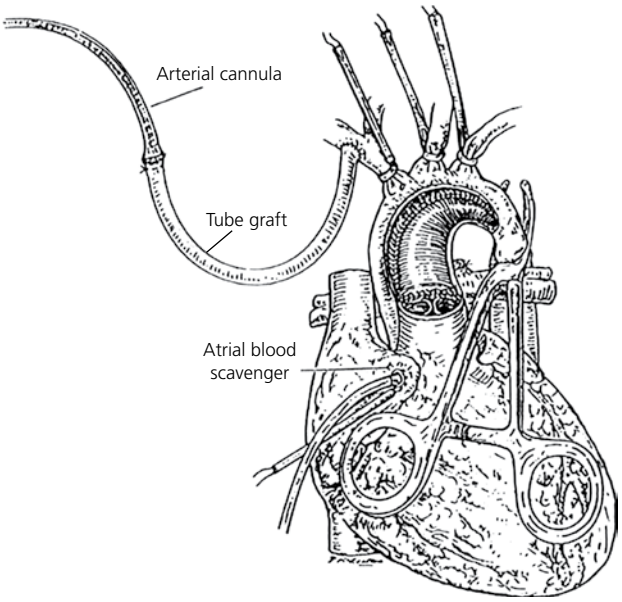


Figure 3.4 Depiction of the operative field during aortic arch reconstruction with RLFP. Arterial inflow is through the cannulated shunt after the anastomosis to the innominate artery is performed. Exposure is maintained by the brachiocephalic snares, a clamp on the descending aorta, and the right atrial blood scavenger. The 3.5-mm polytetrafluoroethylene graft will admit an 8 F or 10 F arterial cannula (Medtronic Bio-Medicus, Eden Prairie, Minn). On completion of the neoaorta, deairing is accomplished by removing the aortic clamp. From Pigula *et al.* [89]. Reproduced with permission of Elsevier.

Regional Perfusion and Hypothermic Circulatory Arrest

Regional low-flow perfusion (RLFP) normally refers to selective antegrade cerebral perfusion and may also be referred to as regional cerebral perfusion (RCP). RLFP is undertaken to allow for aortic arch repairs while avoiding, or more commonly limiting, circulatory arrest. It is commonly achieved with direct arterial cannulation of the innominate artery or with a tube graft to the innominate artery into which the arterial cannula is inserted, as depicted in Figure 3.4. Patients are placed on traditional bypass with either method and cooled to a target temperature of 18–24 °C. Once sufficiently cooled, pump flows are reduced to 20–40 mL/kg/min and brachiocephalic snares are placed, which allows for selective perfusion of the brain with drainage to the superior vena cava or right atrium where blood is then returned to the bypass pump. RLFP rates of 20–40 mL/kg/min have been shown to

adequately perfuse the brain while preventing sequelae from hyperperfusion [89]. Flows may be adjusted during RLFP based on NIRS values (and cerebral blood volume readings if available).

RLFP is an effective bypass strategy and is utilized because studies have shown that outcomes are improved when hypothermic circulatory arrest (HCA) is limited [90–94]. However, it is worth noting that patients with congenital heart disease have preexisting deficits and vulnerabilities that importantly affect neurodevelopmental outcomes after cardiopulmonary bypass making comparisons to the normative population challenging [73].

Some recent investigations have shown that patients receiving circulatory arrest *can* have similar outcomes as those receiving regional low-flow perfusion and that factors such as the cardiopulmonary bypass techniques used (hematocrit, oxygenation and blood gas strategy, hypothermia protocol, flow rates, etc.), technical surgical variations, socioeconomic status, parental IQ, and even birth order may affect outcomes [94–97]. However, most data do show that duration of DHCA greater than 33–41 min is associated with less desirable neurodevelopmental outcomes [94–96]. A safe duration of DHCA has not been defined though a proposal by Whittaker and Grist provides a theoretical time limit based on prevention of anaerobic metabolism with adequate tissue oxygen loading [97]. Most clinicians have concluded that circulatory arrest is certainly a technique where less is more; less time on HCA likely results in more favorable neurodevelopmental outcomes.

Other regional perfusion strategies have been described, which limit whole body or myocardial and/or splanchnic circulatory arrest times [98–100]. Regional cerebral and coronary circulation (RCCP) has an RCP-type arterial cannulation but differs in that the right brachiocephalic vessel is not controlled during part of the arch repair. With RCCP, the distal and transverse arch can be repaired with coronary perfusion allowed with control of the aortic arch just distal to the right brachiocephalic takeoff. The proximal arch is subsequently repaired under myocardial arrest, though repair without cardioplegic arrest has been described [99]. A supplemental strategy to RCCP adds a second arterial cannula to the arterial limb of the bypass circuit which is directed to the descending aorta to provide at least intermittent splanchnic perfusion during aortic arch repair. Pump flows are adjusted accordingly for the areas that are being perfused at any given time. Figure 3.5 illustrates a general timeline for these regional perfusion

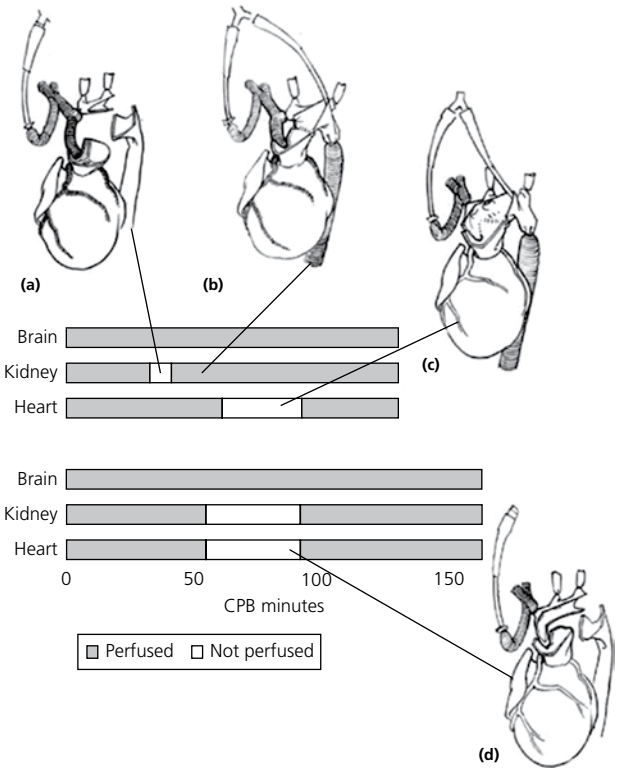


Figure 3.5 Schematic depiction of three-region perfusion strategy during neoaortic arch reconstruction. Shaded bars depict the timeline of direct perfusion (shaded) or ischemia (white) to the cerebral (brain), splanchnic (kidney), and coronary (heart) circulations. Upper panel: three-region perfusion strategy described in this article, with drawings **(a)**, **(b)**, and **(c)** representing the corresponding stages of surgery. The cerebral and coronary circulations are perfused throughout the distal arch reconstruction **(a)**. Distal arch complete, direct perfusion of the splanchnic circulation is resumed **(b)**. Cardioplegia is administered and the coronary circulation is interrupted during only the proximal neoaortic arch completion **(c)**. The direct perfusion of the cerebral circulation is uninterrupted throughout. Lower panel: perfusion and ischemia timeline of the standard approach, with continuous cerebral perfusion throughout, but cardioplegia and interruption of the coronary and splanchnic circulations during the entire neoaortic arch reconstruction **(d)**. Typically, this results with corresponding cooling, rewarming, and overall cardiopulmonary bypass time longer than that of the three-region strategy. From Karavas *et al.* [98]. Reproduced with permission of Elsevier.

strategies as compared to an RCP-only strategy. Regional cerebral, coronary, and splanchnic perfusion (RCCSP) techniques may improve not only neurodevelopmental outcomes, but myocardial and other end organ function after cardiopulmonary bypass where HCA techniques have traditionally been used [98]. However, the technique for regional perfusion and its monitoring vary widely across institutions, and studies have yet to prove its superiority over limited periods of circulatory arrest [101].

Hypothermic circulatory arrest still plays a role in congenital heart surgery. It is commonly used, at least in a

limited fashion, even when alternative strategies are employed. The technique is of great benefit to the surgeon, especially when collateral or other unique circulation prevents a sufficient field of view to allow for a technically accurate repair. In a practical sense for total body circulatory arrest, after cardioplegia is given the perfusionist simply stops arterial inflow, clamps the arterial limb, allows the patient to exsanguinate into the cardiotomy venous reservoir and then clamps the venous line. The patient's normal blood volume cannot remain in the patient since it would still "bleed" into the surgical field preventing adequate visualization. The bypass circuit is then maintained in a limited recirculating mode until bypass is reinitiated.

Finally, it is interesting to consider future therapies that may allow for safer and longer whole body and transplant organ circulatory arrest times by limiting oxygen and substrate requirements on a molecular level with novel therapies such as hydrogen sulfide-induced hypometabolism, vitrification, and other techniques [102–105]. It is likely that hypothermic circulatory arrest will always play a role in congenital cardiac surgery, and so studies investigating enhancements to the strategy are warranted.

Methods of ultrafiltration

The process of running blood through a device with a semipermeable membrane to remove "free water" (water, electrolytes, and substances with a molecular size smaller than the membrane pore size) is termed "ultrafiltration." Excess crystalloid volume on cardiopulmonary bypass may be removed using *ultrafiltration*. The term *hemofiltration* is similar, but it implies that fluid removed with the filter is replaced with some other fluid. Dialysis is another related therapy. In dialysis, a dialysate is used on one side of a semipermeable membrane to remove waste products through diffusion. Dialysis is rarely used during bypass. The terms *ultrafiltration* and *hemofiltration* tend to be used interchangeably in the realm of cardiac surgery and are used without distinction in this text.

The term *ultrafiltration* probably gets more use for two reasons. First, adult programs began using ultrafiltration to remove excess crystalloid volume for years before widespread use in pediatric perfusion. Second, the many acronyms associated with filtration work better with the vowel "u" rather than the consonant "h"! As a case in point, there are several types of filtration worthy of defining: PBUF, CUF, ZBUF, DUF, MUF, and SMUF.

Before bypass

Prebypass ultrafiltration (PBUF)

This form of ultrafiltration is used to help balance the electrolytes and remove undesirable products from blood primed circuits before going on bypass. Blood prime values should be as physiologic as possible. Blood bank preservatives, anticoagulants, and storage may create primes with unacceptably high values for glucose, sodium, potassium, lactate, and so on. Ultrafiltering the circuit volume and adding a balanced electrolyte solution to replace the volume lost in the process is an effective means to control the prime values [106, 107]. PBUF may be performed as needed to achieve acceptable values as measured by a lab or under a more experience-based system based on volumes. A volume-based system may define the amount of filtrate to be removed and replaced based on the institutional system (type of preservative used in blood, age of blood, whether the blood has been irradiated or washed, amount of blood in the prime relative to crystalloid volume, etc.). For example, a volume-based PBUF protocol may state that 2 mL of Plasma-Lyte A per milliliter of prime blood should be ultrafiltered through the circuit before bypass. The goal is the same with either method; the resulting prime should approach physiologic values.

On bypass

Conventional ultrafiltration (CUF)

This form of ultrafiltration is the most basic form on cardiopulmonary bypass. Excess crystalloid volume on bypass can accumulate in the venous reservoir. This volume may be secondary to prebypass fluid administration, cardioplegia delivery, valve testing solution or the need to add crystalloid during periods of decreased venous return (loss to priming the vent or suckers, temporary loss to the chest, etc.). Here, excess volume exists in the circulating volume. The perfusionist simply diverts a portion of the pump blood flow, actively with a roller head or passively based on the bypass circuit pressure, through the hemofilter. Excess fluid is removed and the ultrafiltrate volume, and decreased venous reservoir volume, both indicate the total volume removed. The hematocrit also increases in step with the amount of free water removed. No additional fluid is administered with CUF.

Zero-balance ultrafiltration and dilutional ultrafiltration

The terms "zero-balance ultrafiltration" (ZBUF) and "dilutional ultrafiltration" (DUF) tend to be used interchangeably and refer to procedures on bypass. DUF originally implied incrementally adding crystalloid volume on bypass (diluting the patient a little) and removing the same amount of volume with the ultrafilter. ZBUF was essentially the same except that the crystalloid volume added to the system and the volume removed by the ultrafiltrate line was controlled in tandem so that there was no dilutional effect during the procedure. In practical use, the term ZBUF best captures the idea of the overall process whereby volume is ultrafiltered off and replaced in identical amounts resulting in zero net effect on fluid balance.

ZBUF may be beneficial when there is not excess volume to remove from the bypass circuit, as there is with CUF, but when there may be undesirable electrolyte values or presumed mediators of inflammation in the circulating volume. To improve these undesirable values, the perfusionist adds aliquots of crystalloid and removes equal volumes of ultrafiltrate. This process results in a circulating volume whose electrolyte values approach that of the crystalloid replacement solution. If the circuit

potassium is 6 mEq/L with a lactate of 6 mmol/L, ZBUF with normal saline will bring the circulating volume values for potassium and lactate increasingly towards zero. Again, since equal amounts of volume are added and then removed, the net effect on fluid balance is zero.

After bypass

Modified ultrafiltration and simplified modified ultrafiltration

Modified ultrafiltration (MUF) refers to an after bypass procedure where an ultrafilter is incorporated into a bypass circuit system to remove "free water" after bypass and increase the hematocrit. Ultrafiltration after bypass is thought to have several benefits, though there are conflicting reports in the literature [108–112]. Proponents of MUF usually cite several benefits such as decreased myocardial edema, decreased inotropic requirement, decreased circulating mediators of inflammation, improved systolic blood pressure, increased hematocrit (with decreased homologous blood transfusions), improved coagulation profile, and improved pulmonary function. While these potential benefits are debatable, at the very least, most clinicians agree that MUF efficiently increases the hematocrit while salvaging residual bypass circuit whole blood after bypass (cell saver salvage post bypass only returns red blood cells).

MUF perhaps has the largest set of institutional variations in regards to types of ultrafiltration. As mentioned, MUF implies a system by which blood is processed through an ultrafilter after bypass. The first group to describe such a process for pediatric cardiac surgery was Naik and colleagues at The Hospital for Sick Children in London, commonly referred to as Great Ormond Street Hospital. In 1991, they described a process where after bypass, arterial blood was drawn from the aorta using a roller head pump, pumped through an ultrafilter, and then reinfused to the right side of the heart via the venous cannula [113, 114]. To note, this method resulted in blood flow through portions of the MUF circuit which were opposite of the flow during cardiopulmonary bypass. Their method could be described as arteriovenous MUF (AVMUF) since the source for their system was arterial and return was to the venous system. This original method of MUF may be referred to as The Great Ormond Street Hospital method (GOSH method).

Numerous centers around the world have made modifications to the GOSH method specific to where the blood is drawn from (aortic root needle, arterial cannula side port, down the bypass arterial limb, or a venous source). There have also been modifications to how the blood is warmed (through a separate IV heater, a cardioplegia heat exchanger, or the original oxygenator heat exchanger) and to where it is infused (the right atrial cannula, caval cannula, the aorta, or retrograde up a portion of the bypass venous limb). The various forms of MUF are classified by the source (A, arterial or V, venous) and the return location (A, arterial or V, venous) resulting in four possible sets: AAMUF, AVMUF, VAMUF, and VVMUF. All the possibilities have been described in the literature with numerous variations for each. Figure 3.6 shows a simple schematic of a bypass circuit. Figure 3.7 provides a schematic of a typical GOSH method MUF circuit.

The most significant advancement in MUF technique, in my opinion, came out in the year 2000 and was published by the Halifax, Nova Scotia perfusion group [115]. They titled their VAMUF technique "simplified modified ultrafiltration"(SMUF). Their system truly was simplified because of several concepts. Most importantly, the blood flow through the bypass circuit did not change (remained antegrade), no intervention by the surgeon was required after bypass, filtration-heat exchange-oxygenation was maintained with the standard bypass circuit and oxygenator, no additional heating system or circuitry was required, and no special interventions were needed to reinstitute

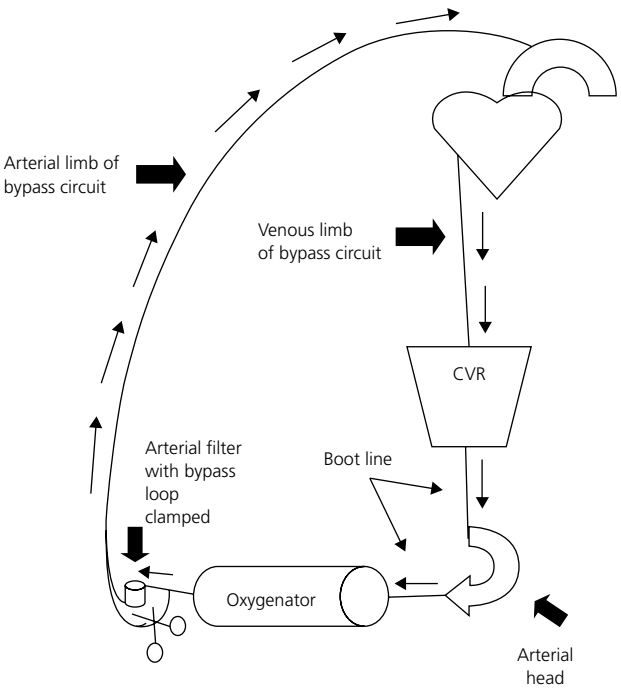
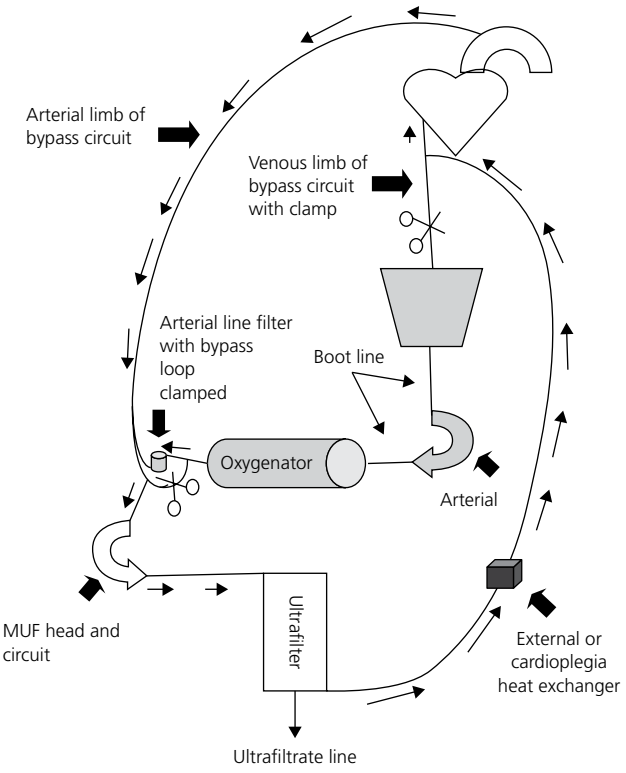
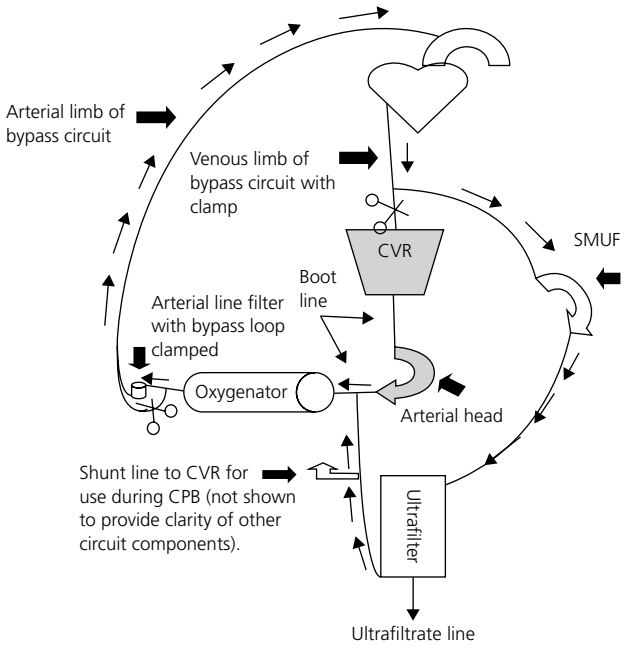


Figure 3.6 Typical bypass circuit.



- The ultrafilter is positioned with the inlet at the top and outlet at the bottom. This orientation allows the ultrafilter to serve as an additional air-handling device.
- Shaded areas indicate the portion of the circuit that may contribute volume for ultrafiltration after bypass.

Figure 3.7 Typical AVMUF flow path through bypass circuit (GOSH method).



- The ultrafilter is positioned with the inlet at the top and outlet at the bottom. This orientation allows the ultrafilter to serve as an additional air-handling device.
- Shaded areas show the portion of the circuit which may contribute volume for ultrafiltration after bypass.
- The SMUF circuit can be used during bypass without affecting overall pump output by using a shunt from the ultrafilter outlet line directed back to the CVR.

Figure 3.8 Typical SMUF flow path through bypass circuit.

bypass or to give cardioplegia once SMUF was complete. These important advancements were achieved by using the bypass venous line for the SMUF source and using a roller head to draw venous blood. This blood was then pumped through an ultrafilter and then the oxygenator (with integral heat exchanger) and arterial line filter with reinfusion to the aorta via the bypass circuit arterial limb. The SMUF system also had the added advantage of continuous hematocrit monitoring since it incorporated blood flow down the bypass venous line where the venous saturation/hematocrit sensor is normally located. This advantage allows the perfusionist to perform ultrafiltration after bypass based on traditional time or volume end-points, or in this system, based on the hematocrit achieved (within the limits of available circuit volume or with the addition of RBCs to the circuit). Figure 3.8 shows a diagram of a SMUF circuit with the addition of a shunt line to the CVR, which was not incorporated in the original SMUF description. This additional line allows for active ultrafiltration *on* bypass without affecting the arterial limb blood flow, as well a method for deairing the system if venous air inadvertently enters the SMUF circuit. The original SMUF circuit, if used during bypass for active conventional ultrafiltration, *added* to the overall arterial pump output since it provided flow to the oxygenator in addition to the arterial head output.

A 3/16″ SMUF circuit such as the one depicted in Figure 3.8 can be added to a pump set-up with a prime volume under 70 mL including the hemoconcentrator itself. This additional prime can be recovered at the end of SMUF by chasing that portion of the circuit through with saline (or air). In this way, the effective overall additional prime volume of the SMUF circuit is negligible.

The original GOSH MUF has been used successfully, and remains in use, at numerous centers around the world. The primary issues to be aware of with GOSH MUF are that air can enter the system if drawn around the arterial cannula purse strings (due to the negative pressure generated in the MUF system), air can be cavitated out of solution or drawn across the oxygenator membrane if the bypass arterial limb or cannula becomes obstructed, the patient may cool if a heating system is not incorporated,

excess potassium may be delivered to the patient if a cardioplegia system is retrofitted after bypass to provide heat exchange during MUF, cerebral blood flow may be compromised if excess flow is taken from the aortic root (cerebral steal), and reinstitution of bypass (and subsequent cardioplegia delivery) may be complicated due to several changes to the circuit which need to be made. I believe the Halifax SMUF technique is undeniably safer, simpler, and just as effective as the GOSH MUF technique. However, there are two caveats. First, some clinicians advocate for an aortic root vent as the source of MUF flow to help ensure deairing after bypass. An aortic root vent source may be helpful, but such deairing comes at the expense of running a circuit that likely has air without a cardiotomy venous reservoir which has been shown to greatly reduce GME transmission (*see Figure 1.7*). A second caveat in favor of traditional AVMUF is that some clinicians argue that returning oxygenated blood to the right side of the heart lowers pulmonary vascular resistance which may be important for some patients.

Regardless of the technique used for MUF, there are identical phases to the process. These three phases are termed normovolemic, hypovolemic, and transfusion. There are some minor differences between MUF techniques regarding *which parts* of the circuit can "contribute" volume to the MUF system and how much *overall volume* can be contributed. Additionally, the technique for flushing volume through the MUF system at the end of the procedure may vary. It is decidedly easier to MUF with plenty of venous reservoir volume after bypass. However, this is not always possible and in fact it may not be necessary. Even zero-balance MUF (adding crystalloid and removing it with no change in hematocrit) can theoretically have beneficial effects by removing mediators of inflammation. However, as stated earlier, the data are inconclusive.

SMUF can be performed after procedures with one or two venous cannulae. With bicaval cannulation, it is common for the SVC cannula to be removed before separation from bypass. The IVC cannula then serves as the circuit source for this form of VAMUF. Circuit clamps and stopcocks are adjusted so that the SMUF blood source will come down the venous line, pass through the SMUF roller head and ultrafilter, on to the arterial pump boot line, through the oxygenator and arterial line filter, and then back to the bypass circuit arterial limb and arterial cannula. With this arrangement, SMUF may be considered a form of partial bypass since the oxygenator-heat exchanger and ALF are incorporated. The ultrafilter is turned on at a flow of 10–30 mL/kg/min within a range of 100–350 mL/min. Flow rates above 350 mL/min through a system incorporating high-flow stopcocks may produce excessively high pressures and risk damaging the blood. Although it is not common to MUF/SMUF larger patients, it could be accomplished with "Y" connectors instead of stopcocks to handle higher flow rates. Vacuum is applied to the ultrafilter at a pressure up to 100 mmHg (and only with active flow through the device). The oxygenator heat exchanger remains on and importantly maintains the patient temperature or even completes the rewarming process after traditional bypass.

Since ultrafiltration removes "free water" from the blood, normovolemic SMUF requires the perfusionist to replace the ultrafiltrate volume loss to maintain an appropriate filling pressure. This volume comes from the venous reservoir and boot line up to where it meets the SMUF circuit. By flushing this volume through with crystalloid to the patient, the hematocrit increases since circuit blood is being added to the system while only ultrafiltrate is removed. During this phase, the arterial head flow essentially should equal the ultrafiltrate loss. However, it is common to see required filling pressures fall while blood pressure rises during SMUF. This is at least in part due to the increased viscosity of the blood. It is debatable as to how much of this effect is due to the removal of mediators of inflammation or decreased total body and myocardial water. The normovolemic phase ends when there is no more available reservoir and circuit volume to contribute (or be flushed through with crystalloid) to the ultrafiltration circuit.

The next phase of SMUF, if desired, is the hypovolemic phase. Here, ultrafiltration continues but without available circuit volume to make up for ultrafiltrate loss, the patient's filling pressures fall. This phase is carried out until "safe" but lower filling and blood pressures, as prescribed by the surgeon, are achieved. The goal here is to efficiently utilize the circuit volume before transitioning to the transfusion phase. The venous cannula is normally removed after the hypovolemic phase with the transfusion phase immediately following. Protamine can be started once the venous cannula is removed. SMUF commonly requires 7–15 min post bypass. While some may consider this additional time before protamine can be administered to be a disadvantage of the technique,

the fact that SMUF can be performed during the postoperative echocardiogram means that the delay is usually quite limited.

The transfusion phase of SMUF simply involves flushing the SMUF circuit volume through in order to transfuse the patient back up to the desired filling pressure. Here, the SMUF circuit blood is chased through with a venous cannula in a saline basin while ultrafiltration continues. The vacuum line is clamped when flow ceases through the SMUF circuit. Volume can be given as needed through the circuit. When the SMUF circuit is flushed through to the hemoconcentrator, the ultrafilter is effectively clamped out of the system. This leaves the perfusionist with the hemoconcentrated volume in the oxygenator and bypass circuit arterial limb. This volume can be flushed through with saline or crystalloid and transfused to the patient as needed or sequestered after the arterial cannula is removed. The anesthesiologist then can use this final high hematocrit circuit volume primarily or it can first be processed in the cell saver.

SMUF specifications

- The heater–cooler unit stays on to maintain patient temperature during SMUF.
- SMUF circuit flow of 10–30 mL/kg within a range of 100–350 mL/min.
  - Neonates under 3 kg may be particularly sensitive to the SMUF circuit flow since it is a steal from the right heart. This will be evident if the pulse pressure flattens. If this occurs, the vacuum pressure should be turned off and the SMUF circuit flow reduced to a level which permits adequate ventricular ejection.
- Apply vacuum pressure of 50–10 mmHg to the ultrafilter.
  - This makes the process more efficient.
  - This step may be omitted for unstable or very small patients who may be exceptionally sensitive to small intravascular volume changes.
- ***Normovolemic phase:***
  - Arterial head flow should equal ultrafiltrate loss taking into account improved hemodynamics as the procedure progresses.
  - The cardiotomy venous reservoir may be flushed through with a crystalloid solution to recover as many red blood cells as possible from the filters.
  - This phase ends when there is no more reservoir and boot line volume (up to the SMUF circuit inlet) to contribute.
- ***Hypovolemic phase:***
  - The surgeon prescribes a filling or blood pressure nadir to ultrafiltrate down to.
  - This phase ends when the prescribed low blood pressure is achieved.
- ***Transfusion phase:***
  - Normally, the venous cannula is removed after the hypovolemic phase with the transfusion phase immediately following.
  - The SMUF circuit blood is chased through with the venous cannula in a saline basin while SMUF continues and the "good" circuit volume is transfused to the patient.
  - The vacuum line is clamped before flow is stopped in the SMUF circuit.
  - The transfusion phase ends when the filling and blood pressures are back to the prescribed levels.
  - Residual volume can be transfused as needed or sequestered and given to the anesthesiologist for administration during chest closure and transport to the ICU.

Ultrafiltration is utilized nearly universally on bypass for congenital heart surgery, and it is warranted for every case to incorporate a hemoconcentrator. Its use for CUF and DUF/ZBUF are undeniably useful. Its use for MUF/SMUF is also useful, and some would argue it to be immensely beneficial. Reports in the literature regarding the effect of MUF/SMUF on total body water, mediators of inflammation, cardiac function, pulmonary function, morbidity and mortality are conflicting. However, at the very least, the use of MUF/SMUF is an effective means of recovering circuit whole blood volume after bypass which improves the post-bypass hematocrit and reduces the need for autologous blood transfusions.

Standard and augmented venous return

Standard venous return with gravity siphon drainage

Sufficient venous return to the heart–lung machine is essential to provide adequate cardiac output to the patient during cardiopulmonary bypass. Traditionally, and commonly today, venous return occurs via gravity siphon drainage (GSD) which requires no additional energy input once set in motion. Siphon drainage allows for the movement of a fluid (blood) through a tube (cannulae and venous limb of bypass circuit) from a higher area (patient on OR table) to a lower area (venous reservoir) as long as the fluid column is not interrupted with air (air lock in venous limb). The fluid may actually flow uphill (from the right atrium or cavae up into the venous cannulae) if the continuous column of fluid flowing downhill exerts enough force. An oft-cited explanation for this effect has one consider the uphill and downhill fluid columns as connected chain links rolling over a spindle at the high point in the circuit. When set in motion, the longer length of downhill chain exerts enough force to pull the shorter uphill length of chain along. While this explanation may not be completely accurate from a physics standpoint [116], it does help conceptualize the process of gravity siphon drainage during cardiopulmonary bypass.

Gravity siphon drainage for bypass is dependent on the relative heights of the patient versus the venous reservoir (specifically, the venous straw outlet), the length and diameters of the venous cannuale and tubing, maintenance of a continuous fluid column, patient volume status, and characteristics of the venous reservoir [117, 118]. Increasing the OR table height or lowering the venous reservoir (if possible) both increase the kinetic potential for GSD (the downhill chain length and pull is increased). Increasing the venous cannulae and tubing diameters increases the kinetic potential for GSD (the downhill chain weight and size are increased) and decreases flow resistance in the system. Additionally, GSD for the most part depends on a continuous fluid column, or else the siphon effect will cease (consider the downhill chain cut in half decreasing the pulling force on the uphill chain). Venous reservoirs have a tube that extends low into the reservoir (the venous straw) near the venous filter outlet to help maintain this continuous column. Of course, the patient must have sufficient intravascular volume returning to the venous cannulae to allow for the siphon effect as well (continuous chain links must be supplied). Finally, the venous reservoir may be constructed in varying ways which affects inflow resistance to venous return.

From a practical standpoint, gravity siphon drainage simply has the perfusionist commence bypass by releasing a clamp on a fluid filled venous line shortly after starting arterial inflow. Venous return is visually monitored throughout the bypass run and quantified by changes in the venous reservoir level. Considering the factors that GSD is dependent on, techniques have been developed to assist venous return to mitigate the requisites and limitations of GSD. Roller pump, kinetic, and vacuum-assisted venous drainage have been reported, each with unique considerations for cardiopulmonary bypass [119–133].

Augmented venous return

Roller pump-assisted venous drainage

Roller pump-assisted venous drainage has limited usage in cardiac surgery today. It essentially has the perfusionist insert a roller head pump as a parallel circuit shunt in the venous line. The roller head pump can be turned on; and with the line that bypasses the roller head at least partially clamped, negative pressure can be exerted on the venous system. Negative pressure in the venous line must be measured. This type of system requires careful monitoring to not impart excessive negative pressures on the vasculature. Roller pump-assisted venous drainage may decrease dependence on the factors associated with gravity siphon drainage. However, kinetic-assisted venous drainage (KAVD) and vacuum-assisted venous drainage (VAVD) systems are much more commonly utilized and are likely easier to use while being safer. A novel use of a roller head pump to apply negative pressure to a cardiotomy venous reservoir via its sucker port to simulate the vacuum-assisted venous drainage technique has also been reported [119].

Kinetic-assisted venous drainage

KAVD may also be referred to as kinetic venous-assisted drainage (KVAD). The pronunciation of KVAD ("K-Vad") rolls offs the tongue much more easily than KAVD ("Kav-Da"), which likely explains its more common use in discussion. Another option is kinetic assisted venous return, or KAVR ("Kav-Er"), which is also easy to pronounce. The term "KVAD" is used throughout this text.

KVAD is a simple technique to augment venous return. Kinetic assist to venous return has the perfusionist insert a centrifugal pump into the venous line. This pump may be placed in series with the venous line or be added as a small

parallel shunt. A parallel shunt insertion ensures that the centrifugal head does not impede venous return when it is not in active use or if there is a failure with the head. If a shunt line is utilized, it must be clamped when the centrifugal head is in use. The perfusionist can commence bypass with traditional gravity drainage and activate KVAD as needed. When turned on, the centrifugal head exerts a negative pressure on the portion of the venous line which extends to the venous cannulae [120]. This pressure can be monitored and titrated to effect during bypass. Pressure in the system is adjusted by changing the head RPMs. It is common to exert a negative pressure in the range of −30 to −50 mmHg though adult minimally invasive systems have reported values up to −100 mmHg [121–123]. Figure 3.9 depicts a centrifugal head insertion into the bypass circuit for KVAD.

As with gravity drainage, a centrifugal head for assisted venous return requires a reasonably continuous fluid column since significant air entrainment will affect performance of the centrifugal head. RPMs may only be adjusted within reasonable limits since high negative pressures may entrain excessive amounts of air from the cannulation sites or elsewhere increasing overall GME transmission and decreasing venous drainage performance [123, 129, 131]. Other drawbacks of KVAD include higher cost relative to other drainage techniques and the influence of head prime volume. Since centrifugal head prime volume may lead to an unacceptable level of hemodilution, it is not common for the technique to be used in children under 35 kg. In addition to improving venous return, major advantages of KVAD include preload and afterload sensitivity which limit its ability to exert extreme pressures, ease of use, the ability to use smaller venous cannulae and line tubing, and the fact that the technique does not increase the possibility of air embolization via a pressurized venous reservoir. KVAD does not change the dynamics of the cardiotomy venous reservoir which remains vented to the atmosphere.

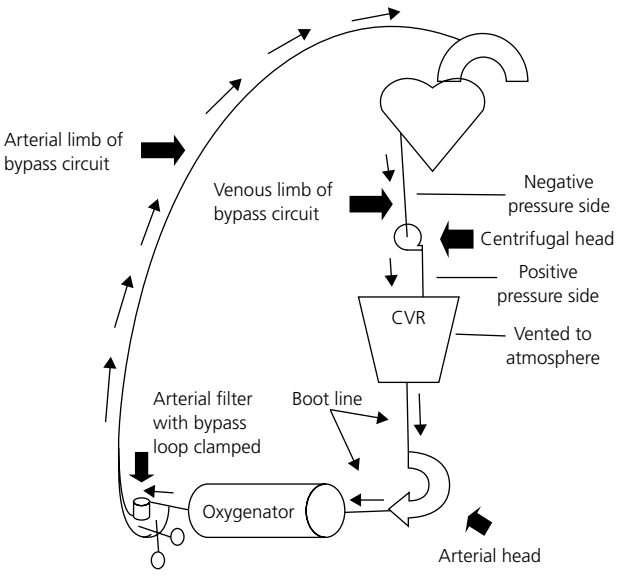


Figure 3.9 Centrifugal head inserted into venous limb of bypass circuit to allow for kinetic-assisted venous drainage. The CVR remains vented to atmosphere.

Vacuum-assisted venous drainage

VAVD is another rather simple technique to augment venous return during cardiopulmonary bypass. It is commonly referred to simply as "vacuum" or "vacuum-assist". With VAVD, the bypass venous reservoir is specialized in that it is "sealed" and not vented to the atmosphere around its top circumference as many oxygenators in years past were. Instead, this sealed reservoir has a single vent port. The vent port can be kept open to allow for use with gravity, roller head-assisted, or kinetic-assisted systems. The vent port can be attached to a vacuum system to allow for VAVD.

A vacuum system consists of basic tubing, commonly 1/4″ or suction, attached to the vent port which then splits into two limbs. One limb is open to atmosphere, and the other is attached to a vacuum regulator with a moisture trap to prevent fouling of the regulator [124]. The regulator is set to the desired vacuum level and the limb open to atmosphere is left unclamped. This allows the perfusionist to commence bypass with traditional gravity drainage. If VAVD is desired, the perfusionist clamps the limb that is open to atmosphere and the regulated vacuum level is applied to the cardiotomy venous reservoir and system. One can switch between vacuum and GSD throughout the bypass run as needed. Figure 3.10 depicts a typical VAVD setup.

VAVD allows for the use of smaller venous cannulae and tubing aiding surgical access while limiting hemodilution [125]. It may also allow for smaller incisions for minimally invasive procedures while at the same time improving visibility at the surgical field and better decompressing the heart. Additionally, it quickly relieves air in the venous line preventing air locks. Finally, VAVD has the advantage of being less expensive to implement than KVAD.

The negative pressure in the cardiotomy venous reservoir during VAVD requires the perfusionist to consider variables not present with GSD or KVAD (KVAD creates negative pressure in the venous line but not in the reservoir!). VAVD increases the external arterial line filter

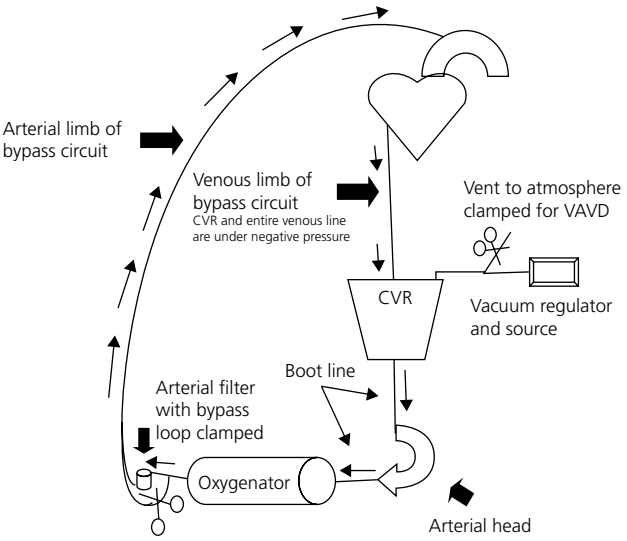


Figure 3.10 Vacuum-assisted venous drainage with the single CVR vent port attached to the vacuum regulator and source with the vent-to-atmosphere limb clamped.

purge and/or sampling line shunt flow and increases the rate of transfusion when adding volume to the reservoir (blood, crystalloid and medications) [124]. One must be careful not to overtransfuse or overadminister since negative pressure is applied to all line attachments to the CVR. Further, the displayed roller head arterial pump output may be inaccurate with negative pressure values below −50 mmHg [126]. And, excessive negative pressure may damage the formed elements of the blood [127].

VAVD is known to accelerate arterial line GME transmission during bypass [35, 128–132]. Increased GME transmission may occur even when the heart is not surgically entered. This can be due to entrainment of venous air from any opening in the vasculature including around the purse string sutures which transits the pulmonary vasculature to the left side of the heart [132]. More commonly, increased GME has been reported simply due to increased air entrainment into a bypass circuit venous system that cannot adequately remove that air [131]. All methods of assisted drainage may also suffer from this phenomenon leading some to conclude that gravity drainage may better protect patients from the neurologic complications of an increased embolic load [132]. It is worth considering that VAVD has been added to the perfusionist's armamentarium with equipment that wasn't specifically designed with its use in mind [128].

Another unique and notable safety concern with VAVD is pressurization of the cardiotomy venous reservoir. Gross air embolism via the bypass arterial line has been virtually eliminated with the advent of numerous safety systems [35]. Gross air embolism via the bypass venous line was unheard of decades ago but exists today and sealed reservoirs designed for VAVD increase the risk [133]. A sealed reservoir with a single vent port has the ability to become pressurized. Positive pressure in the reservoir can force air retrograde through the bypass venous line as air pressure builds and forces blood through the venous straw and venous limb to the patient with air following once the reservoir volume is exhausted. The venous system in effect becomes the path of least resistance for the pressure gradient that develops. This can be catastrophic. To prevent this, VAVD systems normally incorporate a positive pressure relief valve on the reservoir [122]. This is essential because pressure in the reservoir is affected by more than just the vacuum setting. It is actually a balance between the vacuum setting, pump output, venous return, and the positive flow to the reservoir from the pump suckers and vent line. Automated vacuum pressure regulators help ensure the desired pressure is applied under varying conditions. The atmospheric vent and vacuum limbs should branch off close to the reservoir to decrease the chance of kinking. Any obstruction in these lines can result in pressurization and subsequent MAE. As with KVAD, VAVD negative pressures are commonly in the −30 to −50 mmHg range though higher pressures are sometimes used. A negative pressure relief valve may also be used on the reservoir as an additional safety measure to prevent pressure values greater than −100 mmHg [122].

Summary

Augmented venous return is in widespread use for acquired and congenital cardiac surgery but is not without limitations and complications. Augmented return allows for the use of smaller diameter cannulae and venous tubing, may decrease circuit prime volume and limit hemodilution and transfusion, may improve venous drainage, cardiac decompression and visualization at the surgical field, and allow for minimally invasive surgical techniques. Clinicians must be familiar with the limitations of each system and be particularly cautious regarding the potential for increased arterial GME transmission and the risk of massive air embolism via the venous line. There is an increased risk of MAE with VAVD but it is probably underappreciated that even systems not utilizing VAVD can be at an increased risk for MAE as manufacturers have redesigned their venous reservoirs to allow them to be sealed [133].Proper understanding of drainage techniques and careful consideration of the pitfalls of each modality are essential to prevent neurologic complications due to air transmission in cardiac surgery patients [122, 123, 132].

The prebypass checklist

The prebypass checklist is an essential aspect of perfusion safety and should be performed before every bypass run. It should also be part of the permanent patient medical record [134]. There are commonly dozens of items that are checked utilizing the list. Figure 3.11 is an example of a complete prebypass checklist. It may appear that a good number of these items are not essential to check before bypass since they do not directly affect implementation of bypass. However, it is important to check all items since a primary goal of the prebypass checklist is to prevent unnecessary distractions once on bypass. Some centers have amended their checklist to differentiate standard versus expedited setup items. This distinction may be useful during emergencies. Additionally, institutional checklists may be used for going on bypass, termination from bypass and emergent reinstitution of bypass, and so on. A prebypass worksheet outlining pump flow rates and cannulae options may also be useful and Figure 3.12 is included as an example. It is imperative that each perfusion practice modify their department's checklist and worksheet to accommodate the institution's work flow and equipment.

Checklists are only as good as their thoughtful design and use. There is a risk of complacency when doing a checklist often. It is important that the perfusionist read and check every item, every time. Consideration should be given to a challenge format whereby an assistant reads the check points and the primary perfusionist actively verifies each item.

Other Considerations:

_____
_____
_____
_____
_____
_____
_____
_____
_____

__ Patient data reviewed
__ Circuit components patient specific
__ Anticipated cannulae and connectors available
__ Oxygenator heat exchanger water tested
__ Circuit and cannulae expiration dates checked
__ Drug dosages verified
__ Arterial boot size entered
__ Sweep gas flow to oxygenator verified
__ Oxygen analyzer functioning
__ Heater–cooler flow within normal limits
__ Patient blanket water/air flow verified
__ Head rotation and tubing direction verified for all pumps (sucker/vent tested under saline)
__ One-way valve in vent line correctly positioned
__ Circuit CO_2 flushed and crystalloid primed
__ Arterial pressure linked, deaired, zeroed, with alarms set
__ Reservoir properly vented/pressure relief valve in use
__ Pump head occlusions set
__ Tubing connections banded
__ Raceways clear of debris
__ BSA info entered into HLM computer
__ Alarm volume appropriate (must be loud enough to alert team)
__ KVAD/VAVD system checked
__ Reservoir light functional
__ Flashlight available and working
__ Sat/HCT/Venous blood gas monitor connected
__ Level sensor linked and tested
__ Bubble detector linked and tested
__ Hand crank(s) present
__ Isoflurane filled and off, waste anesthetic gas system connected
__ HLM Console battery charged
__ Phone to call for back up personnel available
__ Circuit temp probes connected, alarms set
__ Patient temp probes connected
__ Venous occluder calibrated
__ Additional crystalloid volume available
__ ACT EQC / Hepcon calibrated (Q8 hrs.)
__ Suction canister available for ultrafiltration
__ Spare tubing clamps available
__ Cardioplegia system primed, iced/cooled, linked, boot size correct, pressure zeroed with alarms set
__ Prebypass worksheet completed (see example in Figure 3.12)
__ Replacement components available
__ Sweep gas source verified (FiO_2/carbogen)
__ Circuit blood primed with pH, gas and electrolyte values verified
__ In-line blood gas monitor calibrated and connected, pH/alpha selected
__ All blood in room checked
__ Arterial flow probe connected and zeroed
__ Prime ACT verified >720 s (for blood primes)
__ HLM console locked in place
__ Arterial and venous lines attached to field, properly placed, long enough for bed movement
__ Arterial and venous clamping areas secured (for quick access)
__ Post ALF ports deaired and higher than circuit entry (to prevent air entry in case of abrupt pressure change due to servoregulation)
__ Heparin bolus given and ACT acceptable before bypass

Figure 3.11 The Prebypass Checklist.

Prebypass worksheet

Target HCT_____ End-CPB HCT_____(S)MUF HCT _____ alpha/pH stat Temp _____

MAP range_____ CPG dose full_____(___/____) blood/crys 1/2 _____(___/___) blood/crys

Prebypass values

SaO2	Left _____	Right _____
NIRS	Left _____	Right _____ Somatic _____
Heart rate		
Blood pressure	_____/_____	Left/Right Rad/Fem
CVP	Baseline _____	Post SVC cannula _____On CPB _____

Emergent cannulation options

Arterial	#1 Fr. /flow _____/_____	#2 Fr. /flow _____/_____
Venous	#1 Fr. /flow _____/_____	#2 Fr. /flow _____/_____
	#3 Fr. /flow _____/_____	#4 Fr. /flow _____/_____

Pump flow rates

cc/kg/min	**= cc/min**	**Index flow L/min/m2**	**= cc/min**
10		0.25	
20		0.5	
30		0.75	
40		1.0	
50		1.25	
60		1.5	
75		1.75	
100		2.0	
125		2.25	
150		2.5	
175		3.0	
200		3.5	

Figure 3.12 Prebypass worksheet example.

The surgical safety checklist for congenital heart surgery

The World Health Organization (WHO) began an initiative in 2007 to create a surgical safety checklist that could be used around the world to decrease surgical errors and improve surgical outcomes. Their efforts produced a surgical safety checklist that was released in 2008 [135]. According to a 2010 *New England Journal of Medicine* article regarding implementation of the surgical checklist in the Netherlands, surgical complications were reduced by a third, while surgical deaths were nearly halved [136]. These findings were attributed to checklist implementation. Additional studies have shown a decrease in surgical morbidity and mortality when the WHO surgical checklist, or a modified version, has been implemented.

Boston Children's Hospital and The Society of Thoracic Surgeons (STS) developed a congenital heart surgery checklist that has been implemented, and modified, at institutions around the world. The single most important tool the checklist provides is a formalized

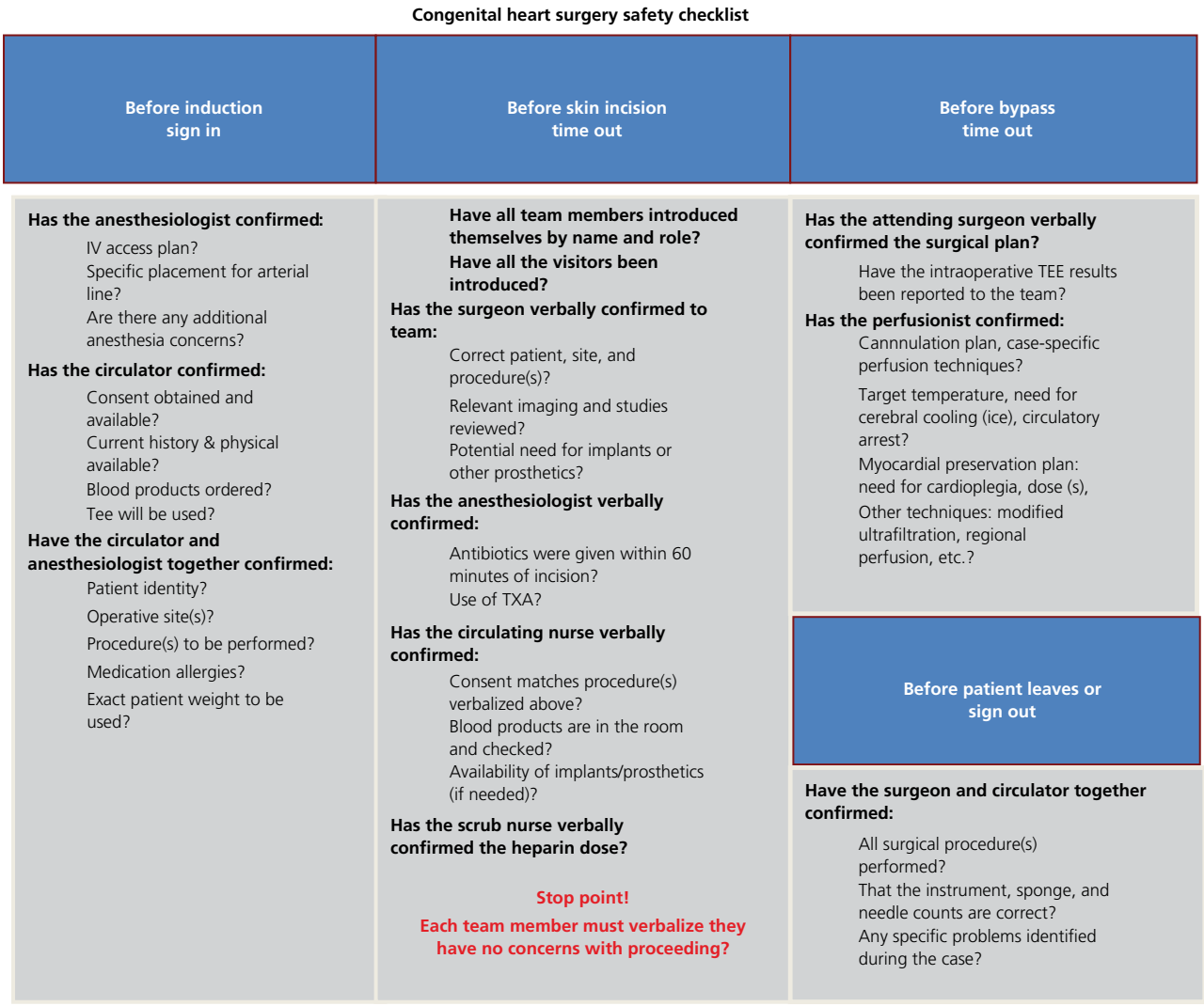


Figure 3.13 Surgical safety checklist example.

communication standard with all operating room caregivers present where the patient specifics and planned procedure are discussed. This effectively places all team members on the same page, so to speak, and gives each member a time to ask questions and raise concerns.

Figure 3.13 is an example of a modified surgical safety checklist for congenital cardiac surgery based on WHO and STS publications [135, 137]. This checklist identifies four timeouts during the patient's operating room experience where critical information should be verified and discussed among the entire surgical team.

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Team communication

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CHAPTER 4

Typical phases of cardiopulmonary bypass

Cardiopulmonary bypass may be roughly segmented into three phases: commencement, standard bypass, and termination. It is important for the perfusionist to have a relatively standardized practice but one that is also flexible to deal with the unique demands of an individual case. A relatively standardized practice better allows one to identify nonroutine events and other issues that may be important to the case. The following are examples of standardized steps and time points for a cardiopulmonary bypass run. Postbypass considerations and bypass time definitions are listed as well

Commencement of bypass

Commencing bypass is a critical moment during the operation where full attention needs to be aimed at ensuring venous drainage, verifying a proper system pressure at full flow, and seeing that gas exchange appears adequate at an expected ventilation–perfusion (V:Q) ratio. Before bypass, the perfusionist should document available values such as heart rate, blood pressure, central venous pressure, near infrared spectroscopy (NIRS), blood gas, and electrolyte values (*see section "The prebypass worksheet" in Chapter 3*). These values can then be compared with the on-bypass values during the initial assessment of bypass.

The pump suckers are normally turned on once the heparin bolus has been administered and an adequate ACT value has been achieved. An exception to this is that suckers may be turned on prior to an adequate patient ACT in order to recover heparinized prime blood from the discarded portion of the arteriovenous pump loop.

Commencing bypass is commonly one of the highest workload periods for the perfusionist. A proper setup, prebypass checklist, and bypass plan all ensure that attention at this time can be focused on the patient's response to bypass and proper function of the equipment used. If medications such as antibiotics, antifibrinolytics, steroids, and so on are to be used, consideration should be given to delivering these to the cardiotomy venous reservoir immediately before bypass to decrease the workload when commencing bypass (and to prevent loss to the field or suckers if administered to the circuit at an earlier time point). The perfusionist should also follow various time points to know when certain interventions are possible: arterial cannula in (can transfuse from pump), ACT > 350–400 (can go on sucker bypass), one venous cannula in (can go on partial bypass), and two venous cannulae in (full bypass likely possible if cavae are dissected free, etc.).

Here is a sample sequence of events and commands for commencing bypass.

- While cannulation proceeds:
  - The arterial and venous pump lines should be visualized to their respective cannulae on the field. This is especially critical with circuits using same-sized arterial and venous pump lines.
  - Once the arterial cannula is in, one can test the bypass circuit arterial line pressure at an index flow of 0.5–1.0, but this is not always possible in patients sensitive to volume, especially before venous cannulation. The bypass arterial line pressure should approximate patient arterial pressure in a static setting and should not spike during volume delivery prebypass.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

- The first time the arterial line clamp is removed after cannulation is a critical moment of the operation. If a recirculation line clamp at the level of the oxygenator/reservoir is inadvertently left off, releasing the arterial line clamp can result in rapid exsanguination retrograde via the bypass arterial limb. All system clamps should be verified before releasing the pump arterial line clamp.
- The perfusionist's arterial and venous line clamps should be on at all times the lines are not in use. If the arterial clamp is left off after arterial cannulation but before bypass, blood may bleed back through the pump arterial line since roller heads are usually set to a minimally nonocclusive standard. Furthermore, if the arterial line clamp is off, accidental infusion of blood can occur with the potential for over transfusion (i.e., arterial head knob accidentally nudged).
- The change, if any, in CVP value with venous cannulation (especially SVC cannulation for bicaval cases) should be noted so that it can be compared with the on-bypass value.

- Surgeon asks perfusionist if they are ready for bypass.
  - Survey the setup and double-check console settings before announcing "Ready." It is a preferred practice to quickly verify at this point that safety features such as the reservoir-level sensor and/or bubble detection systems are attached and turned on. Once on, these devices should remain so for the duration of the case.
- Surgeon instructs perfusionist to go on bypass.
- Perfusionist announces, "Going on."
  - The room should ideally be quiet (no music or unnecessary conversations).
  - The experienced perfusionist can be audibly cued by different sounds in the pump heads, heating/cooling system, and with comments at the field.
  - The electronic record and pump timers are turned on.
- Gas flow is set to expected flow rate.
  - A gas flow for an expected V/Q of 1:1 is a common starting point.
- Arterial clamp is removed and flow is increased, while ensuring the system pressure does not spike to an unexpectedly high level (due to improper arterial cannula placement or aortic dissection, etc.).
- Within seconds of the arterial clamp removal, the venous clamp is removed and held near the clamp site while ensuring adequate drainage while the pump flow is increased to the target flow.
  - The venous clamp may be reapplied quickly in case of emergency, thus preventing unnecessary exsanguination (inadvertent arterial decannulation, aortic dissection preventing adequate arterial flow, or arterial pump head failure). Quickly applying the venous clamp may allow the patient to eject and support their own circulation while the emergency is dealt with. Exsanguination needlessly adds volume resuscitation to any emergency situation the team must deal with.
  - The venous clamp may also be rapidly applied if the reservoir empties suddenly (and past the level sensor point), which may indicate a pressurized cardiotomy. It is always better to delay bypass if there is any concern regarding proper reservoir venting (*see section "Massive air embolism" in Chapter 8*).
  - Achieving full drainage with a single venous cannula generally takes longer than with bicaval cannulation. Be patient to empty with a single venous cannula before ramping flows up to assess overall adequate drainage.
  - Once the target flow is achieved with an acceptable system pressure, the venous clamp can be put aside.
  - The arterial and venous clamps, along with several other tubing clamps, should be readily available for use.
  - The change in CVP pressure once on bypass should be noted. Generally, the CVP falls to zero or a negative number. Positive values should be discussed with the surgeon. Consideration to bed position and transducer height must be accounted for.
  - In-line blood gas and venous saturation monitors will give an early indication to the state of bypass. The color of the arterial and venous lines within the first minute of bypass is also a useful indicator to the state of bypass. (However, erythrocytotic patients and those operated on at altitude may present with relatively dark arterial blood color even when properly oxygenated.)
- If more than one venous cannula is used, the surgeon usually opens just one to establish bypass and then announces when all are open to the bypass venous line. Opening multiple venous cannulae before bypass can lead to air in the cavae/atrium. This is due to the fact that venous cannulae are usually not fully deaired before bypass and because a pressure differential may exist between the cavae that can push air along the pressure gradient.

- "Full flow" (commonly 2.4–3.0 L/min/m^2) is announced to room.
  - The anesthesiologist will normally turn the ventilator off and the pulse oximeter audible tone off at this point, as long ventricular ejection has ceased.
  - Initial pump flow may need to be higher with a patent ductus arteriosus, palliative shunt, or significant aortopulmonary collaterals before those shunts are clipped or controlled. Aortic insufficiency patients may also need higher pump flows if there is significant loss of forward flow to the ventricle and LV vent. NIRS monitoring may be particularly helpful in ensuring adequate cerebral blood flow.
- "Cooling" is announced to room.
  - The heat exchanger, patient blanket/wrap, and room temperature should be adjusted appropriately.
  - Ice may be placed around the patient's head for target temperatures of 25°C or less (to assist with cooling but to also prevent rewarming during any low-flow or circulatory arrest periods).
- Full scanning of available information is started.
  - There should be no interruptions to the perfusionist during the first minutes of bypass while all available settings and values are assessed.
  - Anesthetic agents for the pump, nursing and visitor questions, and so on should wait until the perfusionist is certain the patient is safely on bypass.
  - Standard monitoring throughout the case includes MAP, pump flow, bypass system pressures, CVP, NIRS, in-line blood gas monitoring, venous return level, the presence of venous air, verification that alarm systems are on, operative progress, myocardial ischemic time/temperature, and table and transducer height changes and their effect on CVP, MAP, etc.

Standard support phase of bypass

Commencement of bypass is a high workload time period with critical events and checks occurring over the course of seconds. The standard support phase ideally has a lower workload and involves events and changes occurring over the course of minutes. One exception to this is the cardioplegia delivery period that may take several minutes and involve a high workload much like the commencement phase. While all monitoring is critical throughout the bypass run, the standard bypass period generally allows the perfusionist to verify pump case documentation via an electronic or written record, send regular blood gases to calibrate monitoring equipment, and prepare the patient for eventual termination of bypass in a controlled methodical manner.

Here is a sample sequence of events and commands during the standard support phase of bypass:

- Full scanning of available information is continued.
  - The perfusionist should develop their own scan standard based on monitoring and equipment location and personal preference. Developing a solid scan overviewing all aspects of cardiopulmonary bypass is essential.
  - Standardizing the location of items such as clamps and bypass drugs enables the perfusionist to notice variations readily (i.e., noticing back-up clamps not in their normal location and then discovering they were borrowed by another service; this could hamper circuit interventions during an emergency).
  - Many centers now have overhead or surgeon headlight-mounted camera systems. These systems add great value to the team by enabling everyone to see the surgical field at least in part. The perfusionist can use this information to follow the procedure progress and to minimize disruptions to the case. If the surgeon requests "down on your flow," the perfusionist may be able to view the reason (significant bleeding from a vessel), and not have to continually remind the surgeon of the flow status (still down on my flow), at least until the issue is resolved.
- The rate and effectiveness of cooling must be monitored. The bypass arterial and venous temperatures can be assessed relative to several patient temperatures (rectal, nasopharyngeal, esophageal, tympanic, bladder). Uneven or slow cooling may be indicative of an anatomic issue (coarctation of the aorta with central aortic cannulation may result in slow lower body cooling) or a physical issue (central arterial cannula positioned to favor head vessels may also result in slower than expected lower body cooling—along with a higher than expected bypass system pressure).
- Anesthetic medications and pump meds (antibiotics, steroids, sodium bicarbonate, and pressor meds) are administered as needed.
- Verify the electronic record is capturing data/maintain written record.

  - Electronic records generally capture data intervals of 1 min or several seconds.
  - Written records should document patient data and pump parameters in intervals of 10 min or less. Significant changes and events should be documented as they occur.
- Prepare cardioplegia and ensure the system is ready to deliver.
- "LV vent in/on" announced to room.
- Surgeon places caval tapes (if necessary) and perfusionist verifies that venous drainage, CVP value and NIRS are still acceptable. To note, if the CVP line is within the caval snare, the value may become unreliable.
- "Cardioplegia is ready" should be announced to room.
- "Cross-clamp on" should be announced to room.
- "Cardioplegia going in" verified.
- "Cardioplegia dose is in" announced.
- "Cardioplegia recirculating" or "cardioplegia off" announced.
- The cross-clamp time, time since cardioplegia, and/or myocardial temperature should be announced regularly per surgeon preference (commonly every 20 min).
- The induced ventricular fibrillation time, circulatory arrest time, low flow time, myocardial reperfusion time, cooling/rewarming time, and so on should be relayed to the surgical team per protocol and surgeon preference.
- Temperature appropriate and case-progress adjustments to pump flow and gas flow/source are made as necessary and documented.
- Hematocrit monitored and adjusted per protocol and case progress.
  - Consideration should be given to early administration of blood products, if needed, for younger or critically ill patients more sensitive to acute changes in electrolytes and acid–base status. It may be advisable to administer the necessary products before cross-clamp removal so that desirable base and electrolyte values are present during initial myocardial reperfusion.
- Conventional ultrafiltration (CUF) should be used during bypass for excess circuit volume.
- Zero-balance ultrafiltration (ZBUF) may be considered during rewarming to remove mediators of inflammation and to help balance electrolyte values.
- Urine output should be verified during bypass. The commonly cited minimum value for acceptable output of 0.5 mL/kg/h is debatable. However, most clinicians would agree that urine output should be present and if not, it must be explained (kinked catheter, leak in tubing system, deep hypothermia with low flow/circulatory arrest, etc.). Furthermore, hemoglobinuria should be discussed among the team. Consideration must be given to alkalinizing the urine when hemoglobinuria is present or suspected to prevent cast formation in, and possible damage to, the renal tubules. To note, redness of the urine may also be caused by hematuria (whole blood in the urine) due to urinary catheter insertion or other trauma that would not necessarily require alkalinization.
- "Rewarming" should be announced to the room.
  - Mannitol (0.25–0.5 g/kg) may be given shortly before cross-clamp removal, or at the onset of rewarming, as an osmotic diuretic and free radical scavenger. For this latter reason, the mannitol concentration is ideally highest during initial myocardial reperfusion. Rapid administration of mannitol can cause transient hypotension due to increased skeletal muscle blood flow [1]. This should be a consideration for administration to prevent unwanted hypotension immediately after cross clamp removal (though some clinicians prefer a short duration of hypotension during early myocardial reperfusion).
  - Additional anesthetic agents may be administered to the pump per anesthesiologist preference.
  - The operating room table blanket temperature should be increased.
  - If ice is placed on the head, it should be removed.
  - If circulatory arrest is used with head vessel loops applied, the perfusionist should make sure these loops are removed before rewarming. The surgeon often announces this to the room.
  - Additional temperature devices, such as topical air blankets and room temperature should be adjusted per protocol.
  - Temperature gradients are monitored and consideration should be given to matching the rewarming rate to operative progress.
  - If the patient is warming up well but the necessary surgical procedures required before cross-clamp removal are not completed, rewarming should be appropriately slowed to prevent radiantly rewarming the arrested heart.
  - Gradients between the venous blood and heat exchanger water of 8–12°C are often cited as the maximum safe values to prevent gas from coming out of solution and forming microemboli (whether in the patient or in the circuit). These values should not be exceeded.

  - Oxygenator manufacturers may list temperature gradient maximums of up to 12–15°C, but these limits are not commonly used.
  - It is debatable as to the absolute temperature needed to safely separate from bypass. However, consideration should be given to normothermic values if bleeding will be more of a concern (extensive suture line, etc.).
  - Consideration should be given to not over warming the patient, especially neonates and those who have undergone circulatory arrest. This may include warming to 35–36°C and not exceeding 36°C for any monitored site.
  - Adequate warming helps prevent after-drop and concomitant risks of coagulopathy and cardiac effects. However, some clinicians believe mild hypothermia, and not increasing the bypass time solely for rewarming purposes, improves cognitive and renal outcomes (see section "Temperature Management" in Chapter 3).
- When an LV vent is used, the surgeon may request it be turned off at various points during the surgery, most commonly when the left side of the heart or aorta is being closed. This prevents entraining air into the left side. It is recommended that the vent be clamped in addition to simply turning the pump head off. This is a safeguard against inadvertently turning the vent on (which may occur simply by bumping the controller knob) and causing air to enter the left side (which may or may not be identified by the team before cross clamp removal).
- "Cross-clamp off" should be announced to the room.
- "LV vent back on" should be announced to the room.
- "Caval tapes off" should be announced to the room.
- Flow should be increased in a temperature appropriate manner.
- Consideration may be given to allowing for ejection, once the state of the repair allows, to aid in the rate and quality of rewarming.
- If the patient is in a tachyarrhythmia, consider decreasing the arterial inflow temperature.
- The overall volume status of the patient must be assessed in conjunction with the hematocrit. The reservoir must have enough volume to allow for precise weaning at various flow rates without an untoward amount of crystalloid administration during separation from bypass. The perfusionist must make this assessment based on the case history. If the hematocrit is marginal or even acceptable, but there is not enough volume for weaning, crystalloid administration during weaning may send the hematocrit unacceptably low for smaller patients. The venous reservoir volume necessary for weaning varies between defects and is difficult to globally quantify. However, a rough estimate for many congenital defects would be to have 1/3 of the estimated patient blood volume in the reservoir before weaning. This value may additionally be affected by the reservoir used (soft vs. hard shell, dynamic holdup, minimum operating volume, etc.). Additional reservoir volume should be considered for smaller patients expected to have more immediate postoperative bleeding since the pump suckers may be primed and holding volume for a period of time after weaning off pump. Single ventricle patients tend to require more volume to separate from bypass, and so 1/2 of the estimated patient blood volume should be in the reservoir.
- Crystalloid used at the field to help identify "bleeders" can lower the hematocrit on neonates and small infants if returned to the pump. Ultrafiltration should be used to aggressively remove this volume before separation from bypass.
- The perfusionist should be confident that all bypass and patient values are optimized before the surgeon initiates weaning and termination of bypass.

Termination of bypass

Cardiopulmonary bypass may be terminated rather acutely for relatively healthy patients per surgeon preference. Termination may simply involve clamping the venous line and transfusing volume into the patient to effect the prescribed CVP/LA/systolic BP. However, it is more common in congenital heart cases to proceed with an evaluative weaning period. This involves decreasing the arterial pump flow in a step-wise fashion while gradually occluding the venous return to the reservoir. A finely graduated electronic venous occluder is ideally utilized when weaning neonates and infants. This process may last one to several minutes as the surgical team evaluates function with real-time imaging, ventilator requirements, and pharmacologic support with a decreasing level of support from the heart–lung machine.

- All bypass and patient values are confirmed to be optimized before weaning.

- A blood gas and electrolyte sample measured in the lab should be done shortly before the weaning process, in consideration with lab turnaround times.
- The anesthesia team should review these values in particular, as a baseline before weaning.
- The reservoir volume and patient hematocrit must be adequate to begin weaning.

- A CVP or LA line is generally used to assess volume status during weaning.
- The surgeon generally asks what the exact pump flow is before weaning.
  - To note, the pump flow may be less than "full flow" (2.4–3.0 L/min/m^2) if one of the caval cannulas was removed during the rewarm period, and the patient is ejecting and being ventilated.
  - It may be especially helpful for very small or critically ill patients, or those with small left-sided structures, to watch the leading edge of the waveforms on the hemodynamic monitor while adjusting arterial inflow and venous return. For example, during weaning the LA pressure may average over a few seconds, but the leading waveform may sooner indicate the response to changing pump parameters.
- The surgeon requests decreasing pump flows for a given filling pressure.
  - If augmented venous drainage is in use, it is normally stopped before weaning.
  - Flow and pressure are prescribed by the surgeon and the perfusionist makes appropriate adjustments to arterial inflow and venous line occlusion. Example, "go to an index of 1.5 with an LA of 5" and then, "go to an index of 1.0 with an LA of 6," and so on.
  - The perfusionist verifies the request and announces when the prescribed parameters are achieved.
  - To note, the left ventricular vent flow (if still in use) may need to be decreased in order to finely control the weaning process. Alternately, the surgeon announces vent removal to the room if taken out before weaning. The vent may be removed with its purse string used for left atrial line placement before weaning.
- Weaning continues to an index flow of 0.25–0.5, and the surgeon and anesthesiologist verify acceptable heart rate, rhythm, pacemaker settings (if applicable), blood pressure, ventilation settings, core temperature, and medication drips.
- The anesthesia team verifies that they are ready to separate from bypass.
- The surgeon then requests the perfusionist to "come off bypass."
- The perfusionist clamps the venous line and announces, "pump is off" when the arterial pump head is turned off and the bypass arterial line is clamped.
  - The perfusionist should double check to make sure there is no left ventricular vent flow once off bypass. If the vent is inadvertently left in and on, there will be a matching requirement for transfusion.
  - Ideally, traditional surgical clamps are used to clamp the arterial and venous lines. These clamps have been tried-and-true for decades and are less likely to experience failure or user error as with electronic clamps that are currently available.
  - If a traditional clamp is used for the venous line, the electronic venous occluder should be set to full open (if it was used for weaning). This prevents inadvertently impeding venous return if the patient needs to go back on bypass.
  - The perfusionist's arterial and venous line clamps should be applied at all times the lines are not in use. If the arterial clamp is left off after bypass, blood may bleed back through the pump arterial line since roller heads in the arterial position are set to a minimally nonocclusive standard.
  - Volume is generally transfused via the pump arterial line to maintain a prescribed filling pressure or only at the request of the surgeon if imaging and reparative evaluation are ongoing.
  - If bleeding requires frequent transfusions from the pump, consideration should be given to an appropriate continuous transfusion instead of frequent boluses (i.e., transfusing 20 cc/min and adjusting as needed). This method gives the team a precise value that can be monitored objectively, in addition to observations at the field. Furthermore, this allows for euvolemia during the interventions to correct the bleeding as opposed to only giving when the blood pressure or CVP falls to a set low value. To note, the perfusionist may back off from the requirement, in concert with the anesthesiologist, who may be transfusing blood products (platelets, cryoprecipitate) and who needs "room" in the patient for such transfusions. The surgical team must be aware of a slow continuous transfusion from the pump during assessment of the repair since pump blood may alter the arterial oxygen saturations and possibly affect myocardial function.

Post bypass

The perfusionist should continue to monitor patient needs after bypass. If a method of modified ultrafiltration is used, it is performed after bypass (and before protamine administration) once the surgeon is reasonably confident with the surgical repair and the patient is stable.

Active cardiotomy suction blood can be used for transfusion until the protamine is administered by the anesthesia team. Ideally, the pump suckers are turned off when the protamine starts to absolutely ensure none gets into the bypass circuit. This is preferred since protamine in the bypass circuit effectively ruins the circuit and results in a situation where an emergent bypass run would require assembly of another circuit. Emergent assembly of an additional circuit unnecessarily places the patient at risk. Some cardiac centers turn the pump suckers off once half (or some other percentage) of the protamine is administered in order to salvage as much field suction blood as possible. This method generally works since field suction blood has limited amounts of protamine or none at all. This method does not work if the bleeding preferentially has protamine in it as with protamine administered through a central line with right-atrial back-wall bleeding, which may be difficult to visualize at the field. Small amounts of such cardiotomy suction blood can clot a bypass circuit instantly resulting in a gelatinous reservoir, an unusable circuit, and needless patient risk. Cell saver suction, if in use, is preferred during protamine administration.

Transfusion from the pump can continue until the arterial cannula is removed. Then, the perfusionist can hemoconcentrate the residual circuit blood or run it through a cell saver. The anesthesia team can use this blood as necessary. Alternately, the perfusionist can keep the residual blood (or replacement crystalloid) recirculating in the pump. With either method, it is still important to maintain the circuit for use in case of the emergent need to reinstitute bypass.

Finally, at some time post bypass, the perfusionist should discuss with the surgeon any issues on bypass which were not fully addressed during the operation. Systems, equipment, and other issues identified should be reported through an established event surveillance and quality assurance program. Such a system goes a long way toward preventing team members from repeating the mistakes and unforeseen events of others. The perfusion record should be finalized and entered into the hospital's electronic database or physical patient chart before the patient leaves the operating room. National registry data points should be documented and submitted per departmental policy.

Bypass times

The Society of Thoracic Surgeons (STS) database was rolled out in 1989 as a quality improvement initiative. It is now an international database with over 1000 reporting member sites including more than 100 congenital heart surgery centers. A major benefit of the program is that reporting centers can compare their data with that of others around the world. The database includes numerous data points including bypass times and techniques, blood product utilization, patient demographics, and surgical techniques. The bypass times commonly reported for congenital cardiac surgery cases are defined in the following text.

- Cardiopulmonary bypass time (CPBT)
  - The total time while on bypass including any sucker bypass or regional perfusion times. This value excludes any circulatory arrest or modified ultrafiltration periods.
- Cross-clamp time (CCT)
  - The total time without continuous myocardial perfusion provided by aortic root blood flow. This value includes the circulatory arrest period.
  - For heart and heart/lung transplants, the CCT represents the ischemic time of the donor heart.
  - Donor lungs for transplantation have a cross-clamp time documented in a separate section of the STS database.
- Circulatory arrest time (CAT)
  - The total time within the cardiopulmonary bypass period that the patient is without bypass pump flow.
- Regional perfusion time (RPT)
  - The total time regional cerebral perfusion techniques are utilized. This value includes antegrade and retrograde selective cerebral perfusion techniques.
- Induced fibrillation time (IFT)
  - The total time that myocardial perfusion is maintained while the heart is electively fibrillated. This value is not currently in the STS database, but some centers utilize this technique and track its use.

Reference

1 Cote CJ, Greenhow DE, Marshall BE. The hypotensive response to rapid intravenous administration of hypertonic solutions in man and in the rabbit. Anesthesiology. 1979;50(1):30–5.

CHAPTER 5

Additional notes based on bypass tasks

As noted in previous chapters, the perfusionist must continually monitor dozens of aspects of bypass during the pump run. This chapter includes additional considerations based on specific bypass time points and the operative period.

Prebypass

Heart–lung machine (HLM)

- The HLM should be placed in a location where it can be easily wheeled to the field once needed. Until then, it should be locked in place to prevent movement by anyone except the perfusionist. This is an important consideration to prevent unintentional, and perhaps unnoticed, damage to parts of the bypass circuit.
- Ideally, the lines connecting the bypass circuit to the heater-cooler and gas systems and electrical supply are positioned such that OR personnel need not walk over them.

Reoperations

- Reoperative patients are at an increased risk of emergently requiring femoral or alternate cannulation strategies.
- Supplies for alternate cannulation strategies should be available in the operating room before the chest is opened.
- If emergent cannulation is required, augmented venous return should not be used until the surgeon verifies there are no vascular openings. If vascular openings are present (i.e., hole in the right ventricle (RV) or back wall of right atrium or loose purse strings), the addition of augmented drainage could entrain air into the vasculature where it can then enter the heart. Ejection across the pulmonary bed and/or septal defects could then allow for left sided and aortic/systemic air emboli [1].

Cannulation

- The pump circuit lines should be clamped during cannulation, and the system should be pressurized to approximately the patient blood pressure. This helps identify a proper occlusion setting (for roller head pumps) and allows an initial transfusion at a pressure similar to the patient's blood pressure after arterial cannulation. If the system pressure falls dramatically during the initial pressurization with the AV lines clamped, the pump boot line can be clamped in an attempt to isolate the problem. If normal circuit pressurization can be maintained with the pump boot line clamped, a head occlusion problem is likely present. If not, the source of pressure loss is likely elsewhere (sampling or recirculation line open, pressure monitoring system leak, etc.).
- The arterial cannula is generally inserted first before the venous cannula(e). This is an important consideration in case volume is needed. An arterial cannula connected to the bypass circuit can be described as "the world's largest IV line." While technically incorrect, this functional aspect is important for transfusing quickly and efficiently, or even slowly, for volume loss in the prebypass period.
- The perfusionist should visually verify that the pump arterial and venous lines are connected to their respective cannulae on the field. The surgeon verifies this during cannulation and the perfusionist should double

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check this. This is especially critical when the AV lines are of the same caliber.
- Some centers verify proper arterial cannula placement by transducing the aortic cannula pressure directly and immediately after cannulation. This can be done through an arterial cannula stopcock with a pressure line passed off to the perfusionist or anesthesiologist for measurement. Or, the pump arterial line can be transduced and compared directly to the patient arterial line on the OR monitor (statically with the mean pump pressure or dynamically if attached to a monitor source providing a waveform with systolic and diastolic values).
- The perfusionist maintains clamps on the arterial and venous pump lines during cannulation. The pump lines are also clamped at the field to keep air out. The field clamp on the arterial limb is removed after arterial cannulation and generally stays off for the rest of the case. The field clamp on the venous limb should not be removed until the perfusionist initiates bypass. This is an important consideration. If the pump lines are connected incorrectly (i.e., the arterial limb is connected to the venous cannula(e) and vice versa), the field venous clamp will alert the perfusionist to the problem. With this error, the perfusionist would get an immediate high pressure alarm if volume was transfused before bypass or when commencing bypass. The surgeon usually instructs the perfusionist to go on bypass and then releases the venous clamp at the field.

Transfusion during cannulation

- Volume from the bypass circuit can be transfused as soon as arterial cannulation is complete with connection to the pump arterial limb.
- Rarely, the arterial head occlusion setting, which was properly set during priming, may need adjustment immediately before bypass. This may be due to temperature change in the pump loop tubing or tubing which seats differently after circulating for an extended period of time before bypass. This issue may be identified by an unusual patient response when trying to give volume prebypass (lack of a response) or by the inability of the pump system to hold pressure with the lines clamped. The issue can be remedied by reassessing the pump occlusion via a pressure method.
- Volume from the circuit must be cautiously transfused to the patient during cannulation for two reasons. First, the volume in the pump arterial limb is likely to be at room temperature. Second, the prime is rarely truly physiologic. It may be crystalloid only or blood with slightly elevated potassium, for example. With either situation, pump blood can enter the coronary circulation (or the left ventricular (LV) cavity with aortic insufficiency) and cause an arrhythmia or fibrillation, if transfused too rapidly. A slow transfusion, especially important in neonates, helps prevent these untoward issues.

On bypass

Verification of adequate drainage when caval tapes are used

- Bicaval cannulation alone provides the patient with partial bypass since a portion of the venous return still may enter the right heart. Complete or "full" bypass occurs when caval tapes are applied to the SVC and IVC and results in all of the venous return (except of course coronary sinus, collateral, and bronchial flow, etc.) being diverted to the venous reservoir.
- The surgeon generally announces when the caval tapes are applied and the perfusionist should verify that venous return is still adequate under these conditions. The venous reservoir level and CVP are ideally unchanged (or improved) once the caval tapes are applied. The surgeon must be notified if this is not the case.
- Caval tapes may be applied during cardioplegia delivery. This is not unusual. However, the cardioplegia volume being returned to the pump through the vent (with septal defects) or field suckers may mask decreased venous return to the reservoir. The perfusionist must account for this possibility. In this situation, if a CVP is in use, it may be particularly useful.

Blood gas management

- CO_2 may vary significantly and acutely on bypass as vent/sucker flow varies, when air is intermittently entrained into the venous line, and if collateral circulation is significant.
- Whether diagnosed or not, aortopulmonary collaterals may result in a high CO_2 requirement. The perfusionist should be suspicious of significant collaterals with a higher than expected CO_2 requirement (*see section "Aortopulmonary collaterals" in Chapter 6*, and *section "$PaCO_2$ lower than expected" in Chapter 7*).

- Arterial blood gases are normally drawn from a manifold connected to a post oxygenator source. The system pressure is routinely 100–250 mmHg, and the perfusionist can actively pull arterial samples from the manifold. However, if system pressures are particularly low during low-flow bypass or regional perfusion, blood gas samples should be drawn passively (allow the syringe to fill based on system pressure). This will prevent the possibility of creating an instantaneous negative pressure in the arterial system, which has the risk of drawing air into the aorta around the purse string or across the microporous oxygenator membrane.
- Cautiously treat metabolic acidosis after deep hypothermia (with or without circulatory arrest) during the rewarm period when pH-stat is used. Some patients will have increased base compensating for the increased dissolved carbon dioxide used for pH-stat during deep hypothermia. During rewarming, as the CO_2 is blown off and blood gas management is transitioned to alpha-stat, the patient may become more basic since it takes more time for base correction than $PaCO_2$ correction.

Cardioplegia delivery

- Application of the aortic cross clamp is a critical time point during bypass. The perfusionist must make sure the bypass system pressure does not spike (indicating a possible problem with the aortic cross clamp position); while at the same time, ensuring initial and overall cardioplegia delivery flow and pressure are within expected ranges with an appropriate myocardial response (widened QRS with a slowed rate quickly progressing to asystole).
- For recirculating systems, the table lines should be labeled for flow direction. This should be verified during the prebypass phase. The table line connections must be tightened and deaired before bypass. Allowing unprimed cardioplegia lines on the field poses the risk of air embolization if, for example, the surgeon connects the unprimed line in an emergency and then instructs the perfusionist to flow through that line.
- Try to load and/or give cardioplegia before administering vasoconstrictors to the bypass circuit to prevent vasoconstrictor delivery directly to the coronaries.
- The cardioplegia solution expiration date must be checked before use. Cardioplegia commercially prepared or made in-house generally has a shelf life of 7–14 days.
- Recirculating systems maintain a flow when not in delivery mode. The perfusionist must make sure this recirculating flow is low enough to not cause injury in the event the cross clamp is applied without forewarning.
- Cardioplegia flow and system pressure may be significantly higher than normal during initial root administration for AI patients to aid in coronary perfusion and to prevent loss to the LV through the regurgitant aortic valve. The surgeon may attempt to decrease loss to the ventricle by physically manipulating the aortic annulus during root administration.
- For AI patients, if a recirculating cardioplegia system is in use, be sure to turn the recirculation flow down after root administration and before direct coronary administration to prevent excessive pressure in the coronary arteries.
- For AI patients, if a nonrecirculating cardioplegia system is in use, the deadspace volume may be flushed out between cardioplegia doses. AI patients are more likely to receive additional cardioplegia doses, even within normally single dose cardioplegia systems.
- Single venous cannulation increases the rate of rewarming of the arrested heart, while bicaval cannulation better isolates the heart. Similarly, an increased left heart return via major aortopulmonary collateral arteries (MAPCAs), bronchial flow, and so on, will increase the rate of rewarming the arrested heart.
- Cardioplegia may be given through an arterial cannula side port at the onset of planned circulatory arrest. Additional cardioplegia volume should be given in these instances to account for the increased dead space volume in the arterial cannula.

Planned circulatory arrest

- An insulated bag of ice may be placed on or around the head to prevent rewarming during low flow bypass and circulatory arrest.
- Planned circulatory arrest generally has the perfusionist clamp the arterial line first with the venous clamp applied only after the patient is fully drained, and at the direction of the surgeon.
- If bicaval cannulation is used with caval tapes applied, the venous line may be kept open, at the discretion of the surgeon.
- The circuit volume should be kept recirculating during the arrest period.
- If circulatory arrest was used with head vessel loops applied, the perfusionist should make sure these loops are removed before rewarming. The surgeon ideally announces to the room that the head vessel loops are off.

Induced ventricular fibrillation

- Induced fibrillation with tactile stimulation or electrical induction may be used instead of, or as an adjunct to, cardioplegic arrest.
- Induced fibrillation is always performed in conjunction with hypothermia. Myocardial metabolic requirements during hypothermic fibrillation remain higher than the requirements during cardioplegic arrest.
- Induced fibrillation provides a reasonably relaxed heart with coronary blood flow and allows intracardiac surgical work.
- Induced fibrillation can be used to decrease or even eliminate cardioplegic arrest time. This is particularly helpful in patients who are difficult to cross clamp.
- The fibrillator device may be kept on during the fibrillation time or may be shut off if fibrillation can be maintained without it.
- Pay particular attention to the ECG for resumed activity, especially if the fibrillator device is used for induction but not maintenance of fibrillation. Alert the surgeon immediately if a rhythm resumes.
- Be aware of whether or not the fibrillator pads are kept in place during the fibrillation period when used only for induction. If the pads are in place and activity resumes, the fibrillator device can be turned on immediately as the surgeon is informed. Otherwise, the surgeon must intervene to reestablish fibrillation with proper fibrillator pad placement.
- The perfusion pressure, hematocrit, patient temperature, patency of the coronary vessels, and blood gas management strategy (pH-stat helps counteract the leftward shift of the oxyhemoglobin dissociation curve) are important considerations when coronary perfusion is being maintained during fibrillation [2–8].
- Myocardial oxygen consumption during hypothermia is more profoundly decreased in fibrillating hearts than in beating hearts and the coronary vasodilation capacity is reduced at hypothermia [4, 6, 7].
- There will be an increased amount of volume in the suckers if the right atrium is open. Additional circuit volume may be needed to accommodate this.
- Do not begin rewarming the patient during fibrillation, especially if the pads are not in place, since this increases the chance of the heart resuming a rhythm and increases the metabolic requirements of the myocardium.
- Induced fibrillation is not an option for patients with aortic regurgitation since coronary perfusion could be compromised (with preferential flow retrograde across the aortic valve) and LV dilation may result.
- Induced fibrillation is not an option for hypertrophied ventricles since inadequate coronary blood flow is likely for parts of the myocardium.
- Fibrillator devices run off of battery or wall power. Battery units typically have an auto-off feature to prevent accidental full discharge when not in use. If the induced fibrillation period exceeds the time limit of this auto-off feature, the unit will need to be cycled off/on to maintain its electrical output.
- Battery powered units may also have an indicator light. This light turns on under no-load situations to verify output. When in use with fibrillator pads, the light typically turns ***off*** when there is a good connection between the pad electrodes and the heart and turns ***on*** when the connection between the pad electrodes and the heart is lost (i.e., surgeon lifts the heart away from the pads or removes them altogether).

Administration of blood products

- It is common to administer blood during bypass for congenital cardiac patients, especially in neonates in infants.
- When feasible, products should be added to the circuit slowly to prevent sudden pH and electrolyte changes, especially with a beating heart that may be more susceptible to these acute changes.
- Rapid blood administration may cause hypotension, especially in neonates and infants.
- If the need for blood while on bypass is certain, consideration should be given to administering blood before cross clamp removal. This allows for acid–base and electrolyte correction before reperfusing a heart that has recently undergone arrest and ischemia.
- Blood should be considered for volume replacement, even if the hematocrit is near or at the target value, to prevent acute hemodilution while weaning, maintain the colloid osmotic pressure, and improve oxygen-carrying and buffering capabilities.
- Frequent administration of blood should be followed with increased blood gas sampling and monitoring of electrolytes.

Atrial line placement

- When the surgeon places an RA, LA, or common atrial line into a beating heart, make sure there is sufficient volume in the heart before insertion to prevent possible air entrainment.

LV vent placement

- When the surgeon places a vent into a beating heart, make sure there is volume in the heart before the RUPV/LA/LV is entered to prevent possible air entrainment.

Bed rotation during bypass

- It is common for the bed position to be adjusted during surgery. When a position change is announced, the perfusionist must monitor the pump lines to ensure they are properly positioned and not stretched or kinked in the process. Stretched pump lines increase the risk of accidental decannulation.
- The relative effect of transducer height change on pressure measurements (CVP, MAP, etc.) must be considered with bed position changes.

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CHAPTER 6

Bypass considerations based on diagnosis

Congenital cardiac defects may or may not present in the prenatal or neonatal periods depending on lesion, severity, and prenatal surveillance. Congenital heart disease may present as feeding intolerance, failure to thrive, ECG abnormalities, murmurs, recurrent respiratory infections or transient cyanosis. For many lesions, early intervention is preferred to prevent pulmonary hypertension and/or ventricular dysfunction leading to congestive heart failure. Some defects are quite incompatible with life (TGA/IVS with restrictive ASD or small PDA) and are repaired within days of birth or diagnosis. Other lesions may go undetected until later in life (L-TGA, ALCAPA) and therefore do not have surgical intervention until much later.

Congenital cardiac defects exist over a wide spectrum, and it is important to recognize that each cardiopulmonary bypass plan must take into account the specifics of each planned repair. One of the better examples of how one diagnosis can vary over a wide spectrum is the patient diagnosed with double outlet right ventricle (DORV). On one end of the spectrum, these patients may essentially be a variant of the Tetralogy of Fallot (TOF) diagnosis with repair requiring a well-placed VSD patch (due to the overriding aorta) and perhaps some subpulmonary muscle bundle resection. On the other end of the spectrum, these patients may have transposed great vessels and a VSD. This would require an arterial switch operation and VSD closure. The basic explanations and bypass notes provided in this chapter must therefore be molded to the specific diagnosis, planned surgical procedure, and other pertinent anatomic characteristics to be useful.

The perfusionist must review the available diagnostic studies when devising a bypass plan. Certainly, this review should identify the original diagnosis, surgical and cath lab history, planned repair, status of the ductus arteriosus, presence of aortopulmonary collaterals or palliative shunts, status of the aortic valve (regurgitation may require additional cardioplegia while stenosis requires the circuit to be prepared particularly early since CPR is ineffective in this population), and current level of medical support required (intubated, on ECMO, vasopressor support, etc.). It is essential that the bypass plan consider that the provided medical history is not absolute as well. There are times when diagnostic studies list, for example, "no significant septal defects" or "no significant collaterals identified." Certainly, these important qualifications may impact cannulation strategy and bypass management, and the perfusionist must be prepared for such (a PVR with planned single venous cannulation may require bicaval cannulation if the operative echocardiogram identifies a septal defect). Or, a high level of prebypass vasopressor support may indicate the need for a selection of medications to lower blood pressure soon after commencing bypass (and thus aid in efficient cooling of the patient for hypothermic cases). Finally, complex congenital cardiac procedures may require the perfusionist to consider several diagnoses for a single bypass run. An excellent example of this is in the patient diagnosed with TOF and complete atrioventricular canal defects. The perfusionist must ultimately have a discussion with the anesthesiologist and cardiac surgeon to ensure the best possible bypass plan for the patient.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

Anomalous coronary arteries

Anomalous coronary arteries primarily present in two distinct forms and each have their own unique considerations for bypass. The two most common diagnoses for anomalous coronary arteries are anomalous left coronary artery from the pulmonary artery (ALCAPA) and anomalous aortic origin of a coronary artery (AAOCA).

Anomalous left coronary artery from the pulmonary artery

Basics:

The left coronary artery arising from a venous source such as the PA has obvious repercussions. The myocardium fed by the left coronary may fail without adequate oxygen delivery. Recall that oxygen extraction by the myocardium is normally maximal, and so increased oxygen delivery must be via an increased heart rate. ALCAPA patients may not present until the preteen years or later if adequate collateral vessels develop. Coronary steal with loss of some left coronary blood flow retrograde to the low pressure PA can be particularly troublesome. This can spiral into failure and cardiomyopathy. Surgical correction is aimed at translocating the coronary artery to the aorta. Mitral regurgitation (MR) is commonly associated with ALCAPA with mitral valve repair depending on the degree of regurgitation. The diagnosis of anomalous *right* coronary artery from the PA also exists with surgical correction again aimed at translocating the artery to the aorta.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required. The cardioplegia system must be able to provide flow to the aortic and pulmonary roots in order to adequately arrest and protect the heart.
- Cardioplegia is frequently given soon after going on bypass. The perfusionist should prepare for cardioplegia immediately after establishing bypass. Additional cardioplegia may be needed to account for delivery into the pulmonary root.
- The left and right pulmonary arteries will be controlled before cross clamping to ensure the portion of the cardioplegia delivery into the pulmonary root reaches the anomalous coronary artery.
- Anomalous coronary artery blood flow once on bypass (and before cardioplegia delivery) can be achieved with different strategies.
  - The heart may be emptied when commencing bypass if a side arm off of the aortic cannula is used to perfuse the MPA and anomalous coronary artery (with the RPA and LPA snared). This can be achieved with an "aortic root" needle placed in the MPA and connected to a side arm from the arterial cannula. This root needle can then be connected to the cardioplegia delivery system at the time of cross clamping.
  - The heart can be maintained in an ejecting state.
  - The branch pulmonary arteries can be snared allowing for antegrade flow across the pulmonary valve and into the anomalous coronary artery as long as some blood is allowed to enter the right side of the heart.
- These patients may be particularly arrhythmic. Consideration must be given to commencing bypass slowly to prevent ventricular fibrillation and LV distention, especially before the LV vent is placed. A cannulation strategy including PA perfusion off of the arterial cannula is preferable since coronary blood flow to the PA is still maintained if the patient fibrillates when placed on bypass.
- Single venous cannulation (dual stage if available) is commonly used.
- MR and/or septal defects require the use of bicaval cannulation.
- Left ventricular vent common if bicaval cannulation is used.
- Target temperature 25–32 °C.
- Minimum dilutional hematocrit is 30–35%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- The surgeon may request a higher than normal blood pressure during rewarming and myocardial reperfusion to aid in coronary blood flow. However, it is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

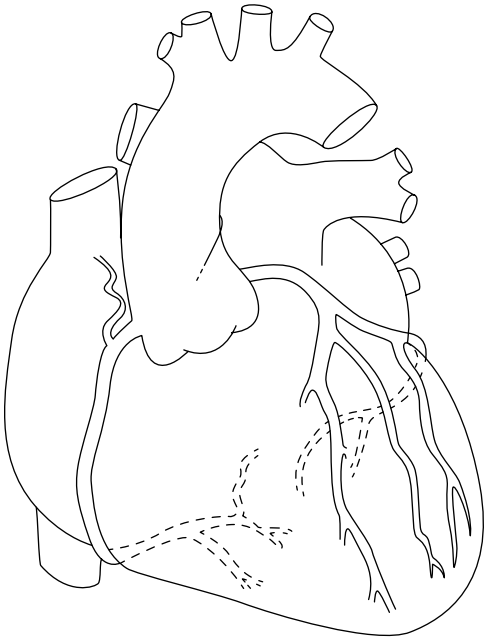

Figure 6.1 Normal coronary artery connections. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

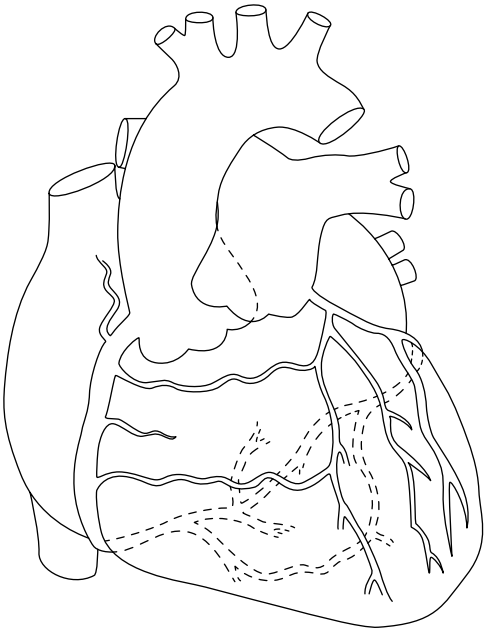

Figure 6.2 Anomalous left coronary artery arising from the pulmonary artery. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Anomalous arotic origin of a coronary artery

Basics:

Anomalous aortic origin of a coronary artery most often has a coronary that arises from a nonstandard sinus of Valsalva. This may present with an intramural course (the take-off runs within the aortic wall before branching normally) or an interarterial course (a segment runs between the aorta and pulmonary artery). An intramural course may restrict blood flow in the lumen or at the ostia. Surgical correction for an intramural course is aimed at "unroofing" the coronary artery, which is essentially removing a portion of aortic wall from the ostia to where the coronary travels away from the aortic root. An interarterial course may present as a slit-like flattened lumen easily obstructed by external pressure. Here, coronary blood flow may be especially compromised during exercise since the coronary artery may be compressed with elevated PA pressures. Surgical correction for an interarterial course may require translocation of the coronary to a more suitable location in the aortic root.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required. The surgeon may request a higher cardioplegia delivery pressure to ensure distribution down a coronary with a slit-like lumen.
- Single venous cannulation (dual stage if available) is commonly used.
- The presence of septal defects normally requires the use of bicaval cannulation.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 30–35%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- The surgeon may request a higher than normal blood pressure during rewarming and myocardial reperfusion to aid in coronary blood flow. However, it is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

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Surgeon preferences:

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Aortic regurgitation/insufficiency

Basics:

A sufficiently regurgitant aortic valve increases the workload on the heart and may lead to failure. Significant regurgitation can lead to a low diastolic pressure that can negatively affect coronary artery perfusion. The valve needs to be made competent either by repairing/rebuilding the leaflets or with valve replacement. Occasionally, an annuloplasty or root reduction (if abnormally enlarged) is enough to make the valve competent. In pediatric patients, aortic valve repair or homograft valve replacement are both favored over mechanical replacement when feasible since it allows for growth and may preclude the need for lifelong anticoagulation, especially in active young patients. Mechanical replacement is more common as one progresses through the adult years.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia is required. There should be 1.5–2 times the arresting dose available to account for volume lost to regurgitation and the need for direct administration if the initial root dose is not effective.
- Cardioplegia flow and system pressure may be significantly higher than normal during initial root administration to aid in coronary perfusion and to prevent loss to the left ventricle through the regurgitant aortic valve. The surgeon may attempt to decrease loss to the ventricle by physically manipulating the aortic annulus during root administration.
- If a recirculating cardioplegia system is in use, be sure to turn the cardioplegia head flow down after root administration and before direct coronary administration to prevent excessive pressure in the coronary arteries.
- If a nonrecirculating cardioplegia system is in use, the dead space volume should be flushed out between cardioplegia doses. AI patients are more likely to receive additional cardioplegia doses, even within normally single dose cardioplegia systems.
- Single venous cannulation (dual stage if available) is most commonly used.
- The presence of septal defects normally requires the use of bicaval cannulation.
- Left ventricular vent is required. The amount of LV vent return is a rough surrogate for the degree of regurgitation. If the vent flow is adjusted so that it is full (without air entrainment from a suction-limiting one-way valve), the percentage of loss through the regurgitant valve can be estimated. It is expected that the relative amount would be significantly less after successful repair during reperfusion of the heart. If it is not, the surgeon should be informed.
- Aortic insufficiency increases the likelihood that blood from the aortic cannula will flow retrograde to the left ventricular cavity. Consideration should be given to going on bypass slowly and cooling slowly to prevent LV distention secondary to the AI and\or ventricular fibrillation due to a cool prime directly perfusing the coronaries. This can be an issue, especially with younger patients.
- Patients with aortic regurgitation are at an increased risk of systemic hypoperfusion if a significant amount of arterial pump head output is lost retrograde across the aortic valve to a left ventricular vent.
- One must be extra vigilant in monitoring venous return and preventing LV distension (due to AI), particularly until the LV vent is placed.
- Consider upsizing the vent for significant AI since the vent flow may be much greater than normal for a particular size patient.
- If the patient fibrillates before an LV vent is placed, immediately decrease the pump flow to prevent LV distention and consider 1–1.5 mg/kg of lidocaine delivered to the pump.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- The perfusionist must consider that once the aortic cross clamp is on (and loss to the ventricle is prevented), the pressure may increase suddenly and significantly.
- Increased pump flows, in consideration for the vent flow, should be used first line to treat hypotension before the aortic cross clamp is placed.
- When vent flow is high due to significant AR, there will be an increased need for CO_2 administered to the circuit since the blood CO_2 is effectively being scrubbed with an increased air:blood interface in the vent line and cardiotomy reservoir. The $PaCO_2$ may vary greatly until the cross clamp is applied. Decreasing the sweep flow alone may not be sufficient to raise the $PaCO_2$ to the desired

value. An increased CO_2 gas flow rate or increased carbogen concentration may be needed.

- Surgical work from the aortic sinus to transverse arch, or any other procedure that intermittently stimulates the aortic baroreceptors (pressure/stretch receptors) in this region, may produce acute changes in blood pressure on bypass. The perfusionist should be cautious treating such fluctuations that may present as quite temporary. Patience in treatment for an acutely fluctuating pressure may prevent rapid sequential administration of vasodilators and vasoconstrictors.
- Valve irrigation solution should ideally be scavenged with either wall or cell saver suction.
- The surgeon may request a higher than normal blood pressure during rewarming and myocardial reperfusion to aid in coronary blood flow and to prevent ventricular fibrillation if the ventricle is hypertrophied. However, it is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

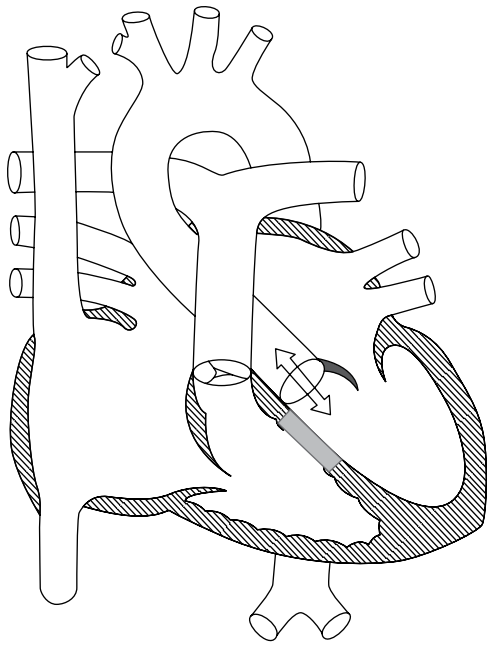

Figure 6.3 Aortic regurgitation. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Aortic stenosis

Basics:

Aortic stenosis (AS) can be congenital (*see also section "Critical aortic stenosis"*) or acquired. It is commonly classified by location: supravalvar, valvar, and subvalvar (subaortic). AS can lead to ventricular hypertrophy and eventual heart failure. Surgical correction in its simplest form is aimed at excising the obstructing tissue, muscle, or membrane. More extensive involvement in the subaortic region (left ventricular outflow tract) may require excision of the ventricular septum and placement of a ventricular septal patch (*see section "Konno procedure" in Chapter 9*). Supravalvar stenosis may require enlargement of the aortic sinuses.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- The perfusionist must be exceptionally prepared for these cases. Patients with AS do not respond well to CPR since either compression of the chest (closed CPR) or heart (open CPR) is met with significant outflow obstruction. Systemic blood flow and coronary blood flow are insufficient with CPR for AS patients; and if cardioversion is difficult, bypass is the safest treatment. Therefore, the bypass pump should be primed as soon as possible to ensure there is no perfusion delay in case of the emergent need for bypass.
- Cardioplegia is required. Consideration must be given to higher cardioplegia delivery pressures/flows if significant ventricular hypertrophy exists. This will help ensure proper myocardial distribution of cardioplegia.
- If ventricular hypertrophy exists, consider having more than the standard cardioplegia dose available.
- Single venous cannulation (dual stage if available) is most commonly used.
- The presence of septal defects normally requires the use of bicaval cannulation.
- Subaortic stenosis increases the likelihood of bicaval cannulation since there exists the possibility of entering the right heart through the ventricular septum (unintentionally or with a Konno procedure).
- Left ventricular vent is common.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- Valve irrigation solution should ideally be scavenged with either wall or cell saver suction.
- These patients are normally hypercontractile after bypass since the LV has been pumping against significant obstruction. Such patients may require less venous reservoir volume to separate from bypass.
- If the heart was in failure, the LV may not be hypercontractile.
- AS patients are likely to exhibit high blood pressure once ejection is established after the repair. This must be controlled, in particular, because the aorta has been entered and now has a suture line. It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection. However, the surgeon may elect for a somewhat higher blood pressure to prevent ventricular fibrillation if there is hypertrophy.
- The perfusionist needs to be extra cautious when increasing the pump flow immediately after cross clamp removal since the blood pressure may go to an unacceptably high level.
- The anesthesia team will have medications to treat hypertension, but the perfusionist may need to intervene while the drips take effect (bolus nitroprusside, nitroglycerin, phentolamine, etc.).
- When the repair is anticipated to require a short bypass run, as with an uncomplicated subaortic membrane resection, the anesthesiologist may request that anesthetic gas be delivered through the circuit for the duration of CPB.

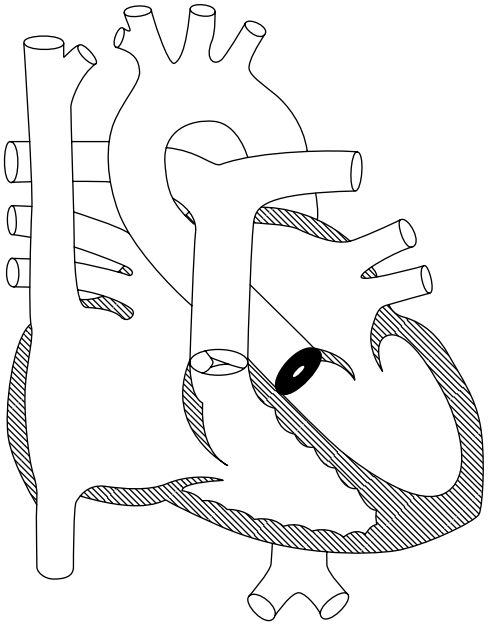

Figure 6.4 Valvar aortic stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

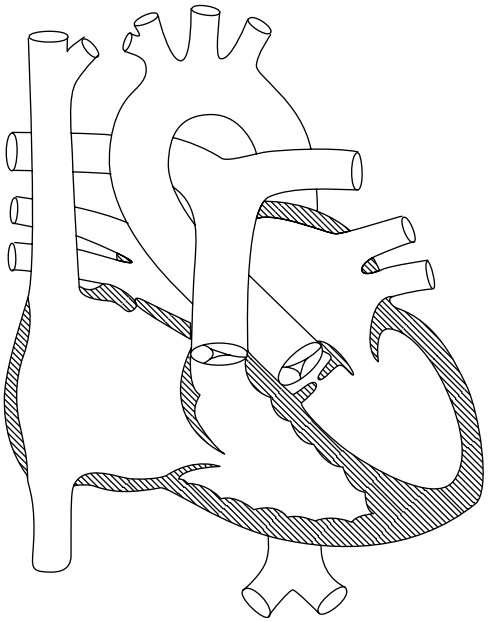

Figure 6.5 Discrete membranous subaortic stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

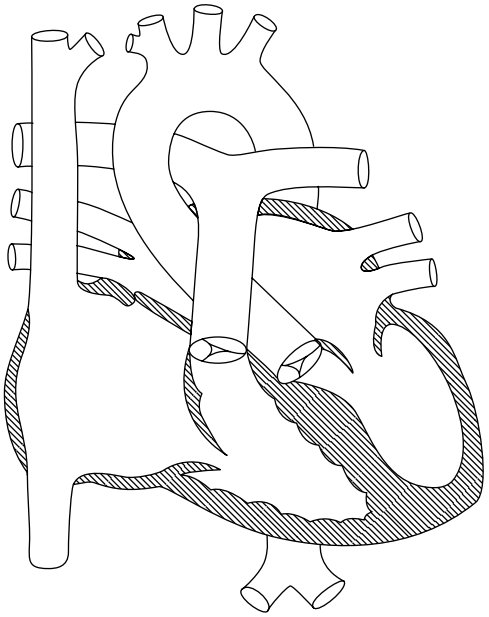

Figure 6.6 Hypertrophic subaortic stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

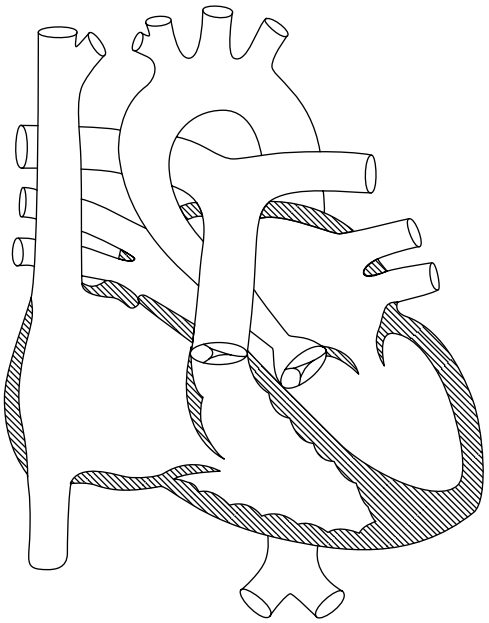

Figure 6.7 Supravalvar aortic stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Surgeon preferences:

Aortopulmonary collaterals

Basics:

Aortopulmonary collaterals (APCs) can develop between the aorta (or other systemic arteries) and the pulmonary circulation, particularly when there is insufficient pulmonary artery blood flow. These APCs can develop in a variety of scenarios, but most commonly they are associated with pulmonary atresia (PA or TOF/PA) or discontinuous pulmonary arteries. They can also develop with other defects, especially in patients with single ventricle physiology (Fontan circulation). These collaterals may present as an important source of pulmonary blood flow before surgical repair but can stenose with time. APCs of the head and neck vessels are of particular concern because they may result in inadequate cerebral blood flow due to systemic flow being diverted to the pulmonary bed. These vessels are more likely to be unifocalized. This involves surgically placing their proximal ends to a right-sided structure such as the pulmonary artery or a right ventricle-to-pulmonary artery conduit. This offloads work from the left side of the heart by reducing the Qp:Qs ratio.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5–5 L/min/m^2 depending on the anticipated collateral blood flow.
- APCs are frequently corrected before bypass (unifocalized) or at least controlled with vessel loops. MAPCAs must be corrected or controlled when possible since effective systemic blood flow may be compromised once on bypass. This is due to a portion of the aortic blood flow being diverted to the pulmonary system. Pump flows may need to be increased (sometimes significantly) to provide adequate effective systemic flow.
- Patients with significant collateral blood flow are at an increased risk of ventricular distension upon initiating bypass. This can occur due to the heart ejecting less forcefully in the setting of a prime temperature which is less than patient temperature and if the prime ionized calcium level is lower than the patient value, particularly in neonates and infants.
- Head and neck collaterals are more likely to be unifocalized since they have implications for cerebral perfusion.
- The need for cardioplegia is dependent on associated defects.
- Cannulation is dependent on associated defects.
- A left ventricular vent is required if bicaval cannulation is used since the collateral flow increases return to the left side of the heart via the pulmonary circulation.
- Consideration should be given to a lower target core temperature on bypass to help offset a potentially compromised cerebral blood flow.
- Consideration should be given to a higher hematocrit (>30–35%) to help ensure adequate oxygen delivery to the brain in the setting of potentially compromised cerebral blood flow.
- pH-stat blood gas management is preferred. This method increases the CO_2 in the blood, which provides dilation of the cerebral vessels aiding cooling and oxygen delivery while helping prevent overall inadequate cerebral blood flow. Inadequate cerebral blood flow, despite an "appropriate" set pump flow, can lead to significant morbidity including stroke, choreoathetosis, and seizures. Increased $PaCO_2$ levels also constrict the pulmonary bed which favors cerebral and systemic blood flow.
- Consideration should be given to the anticipated bypass pump flow, which might be much greater than normal, when selecting equipment. If a patient is placed on bypass emergently before the APCs are controlled, the patient may need an index flow of 3.5–5 L/min/m^2. The oxygenator and filter system must be able to accommodate this.
- Consideration should be given to the amount of blood products on hand. Increased pump flows and sucker flows may increase the dynamic hold up and prime of the system (particularly the volume in the vent line, sucker tubing, and cardiotomy) requiring additional products early in a pump run. The perfusionist should be cognizant that this need can be masked as poor venous return when the reservoir level drops but in fact may simply result from priming other parts of the circuit.
- The use of alpha agonists is debatable in this patient population. On one hand, some clinicians believe that the blood pressure should be within its normal range regardless of diagnosis. On the other hand, some clinicians feel that alpha agonists simply increase the peripheral vascular resistance and thus increase pulmonary blood flow for patients with APCs (the number may look good on the monitor, but effective systemic and cerebral blood flow may be suffering). The use of NIRS may be helpful in determining adequate cerebral blood flow.
- Flow should be used first line to increase blood pressure but will be limited by venous return (and vent/sucker return) or possibly even the rated flow for the selected equipment.
- When bypass and/or sucker/vent flows are high (due to APC flow back to the left heart), there will be an increased need for CO_2 administered to the circuit since the blood CO_2 is effectively being scrubbed with an

increased air:blood interface in the cardiotomy reservoir, vent, and suckers. This need may vary greatly throughout a bypass. Decreasing the sweep flow alone may not be sufficient to raise the CO_2 to a normal value.

- An in-line blood gas monitor is essential to properly provide pH-stat in this patient population.

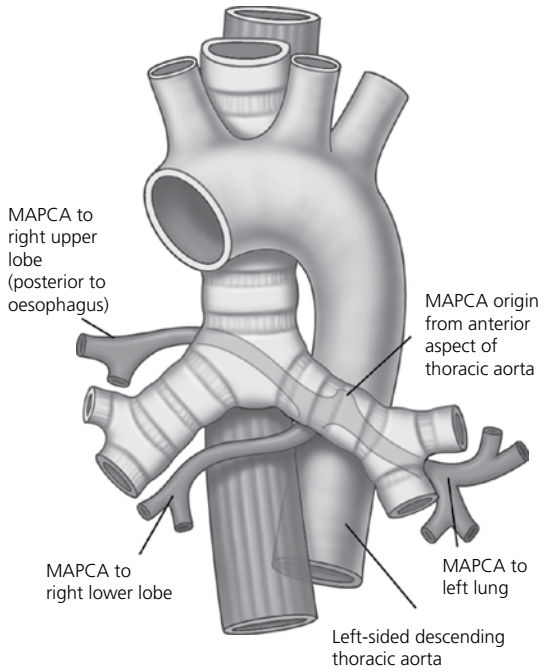

Figure 6.8 Major aortopulmonary collaterals from the descending aorta. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Aortopulmonary window

Basics:

An aortopulmonary window is a congenital condition whereby there is communication between the aorta and main pulmonary artery. The size of the communication determines the symptomology. Basically, the larger the communication, the greater the symptoms. Excess blood flow from the aorta to pulmonary artery, though the aortopulmonary window, can lead to pulmonary hypertension and heart failure. The clinical presentation may be similar to other conditions that result in a significant left-to-right shunt (ASD, VSD, PDA). The surgical repair is aimed at closing the communication or "window." The window is typically closed by dividing the great vessels and patching each one separately.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2. Pump flow may need to be higher to compensate for pulmonary runoff until the window is controlled.
- Cardioplegia is required.
- Single venous cannulation (dual stage if available) may be used.
- The presence of septal defects normally requires the use of bicaval cannulation.
- The branch pulmonary arteries are usually controlled before or soon after commencing bypass since runoff to the pulmonary circulation can compromise systemic perfusion (much like a PDA, BTS, or MAPCAs which need to be controlled).
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

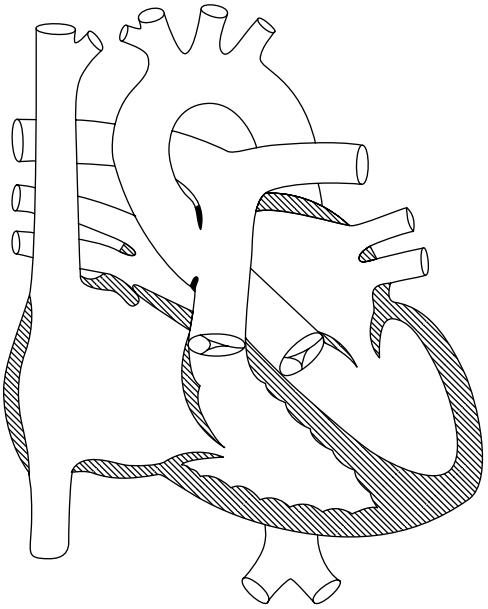

Figure 6.9 Aortopulmonary window. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Atrial septal defect

Basics:

Atrial septal defects (ASDs) can increase the workload on the heart by increasing the cardiac output that needs to be maintained due to the volume recirculated to the lungs with the left-to-right shunt (increased Qp:Qs). Correction is aimed at closing the "hole(s)" and ensuring the pulmonary veins drain properly to the left side.

Secundum defects are centrally located in the atrial septum and may be closed primarily or with a patch.

Primum ASDs have deficient tissue lower in the septum, which may extend further to create a cleft mitral valve. This would require cleft closure and/or mitral valve repair in addition to the septal repair.

A third type of atrial septal defect is termed "sinus venosus". A defect in the SVC RA junction can allow normally placed right pulmonary veins to drain into the SVC or RA. Sinus venosus defects may be found either at the right atrial-SVC junction (superior sinus venosus defect) or by the right atrial-IVC junction (inferior sinus venosus defect). Correction requires routing the pulmonary venous blood flow to the left atrium and closing any septal defects. Scimitar syndrome is named for the X-ray appearance of the enlarged curved sword (scimitar) shaped pulmonary venous drainage from the right lung to the IVC seen with some inferior sinus venosus defects. With Scimitar syndrome, the right lung is commonly hypoplastic and its blood supply is via MAPCAs.

A fourth type of left-to-right shunt at the atrial level is the unroofed coronary sinus. An unroofed coronary sinus may also be referred to as a coronary sinus septal defect (CSSD). It results in a similar physiology as an ASD with similar perfusion strategies for the bypass run. The coronary sinus normally drains coronary venous blood flow to the right atrium. A coronary sinus septal defect allows left atrial blood to enter the RA via the coronary sinus defect. If a persistent left superior vena cava (PLSVC) to coronary sinus is present, surgical correction is required to route the coronary sinus blood flow to the right atrium. If no LSVC is present, simple closure of the CSSD may be performed.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required (induced ventricular fibrillation may be used alone or in addition).
- These cases tend to have short bypass runs, which means one needs to be extra vigilant and quick to establish bypass and prepare for cardioplegia since the surgeon may progress to cross clamping within minutes of going on pump.
- Try to load and/or give cardioplegia before administering vasoconstrictors to the bypass circuit to prevent vasoconstrictor delivery directly to the coronaries via the cardioplegia.
- Bicaval cannulation required; if a PLSVC is present without a bridging vein, a third venous cannula may be needed. Alternately, a low flow sucker can be placed in the LSVC for drainage. The LSVC drainage to the heart can also simply be handled with a drop sucker.
- Some surgeons may use an endotracheal tube (ETT) to cannulate the SVC. This precludes the need to dissect out the SVC, especially with a mini-sternotomy. The patient can then be placed on full bypass by inflating the cuff to internally seal the SVC (with a caval tape on the IVC cannula).
- Alternately, a straight venous cannula can be placed through the atrium and positioned in the SVC, which may be preferable with a mini-sternotomy. A caval tape would then be required for full bypass.
- Left ventricular vent is common; sinus venosus cases may have a vent placed directly through the LA so that it does not impair the surgical correction.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25%.

Table 6.1 Endotracheal tube sizing for SVC.

Estimate for ETT in SVC	
Kilograms	**ETT size**
<9	4.0
9–20	4.5
20–35	5.0
35–50	5.5
>50	6.0

≤5.0 ETT 3/16 × 1/4 connector
≥5.5 ETT 1/4 × 1/4 or 1/4 × 3/8 connector

- These patients, especially those allowed to have a lower hematocrit to prevent blood transfusion, may require increased flow early in the case (for hypotension or low NIRS values) and the use of vasoconstrictors may be needed.

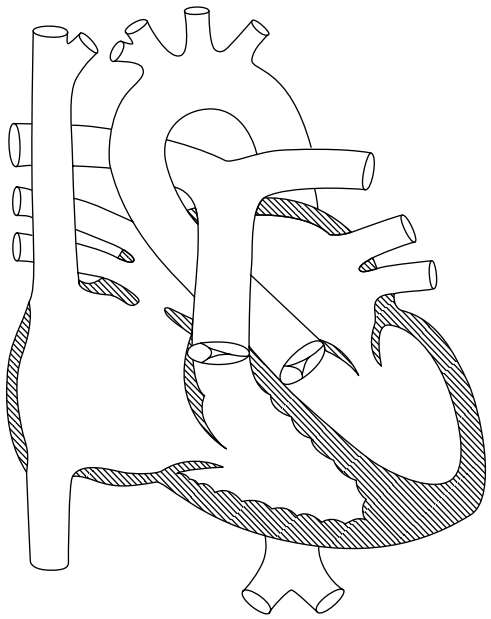

Figure 6.10 Secundum atrial septal defect. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

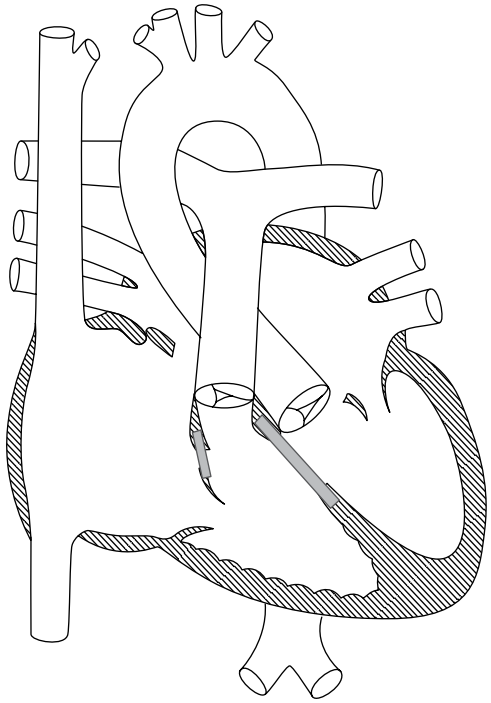

Figure 6.11 Primum ASD with cleft mitral valve. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- When the repair is anticipated to require a short bypass run, the anesthesiologist may request that anesthetic gas be delivered through the circuit for the duration of CPB.

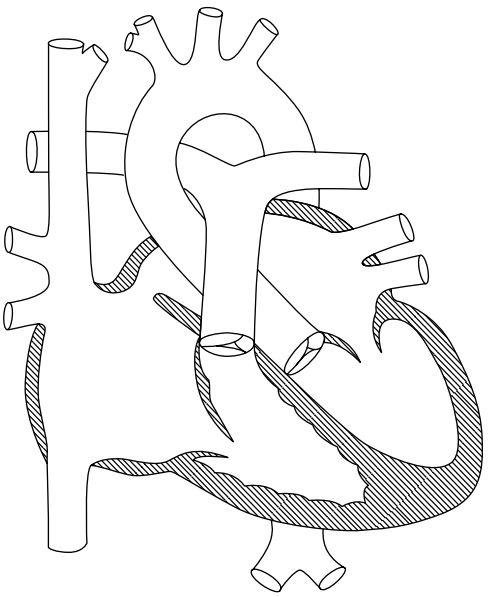

Figure 6.12 Sinus venosus ASD with PAPVR (right pulmonary veins returning to the SVC/RA junction). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

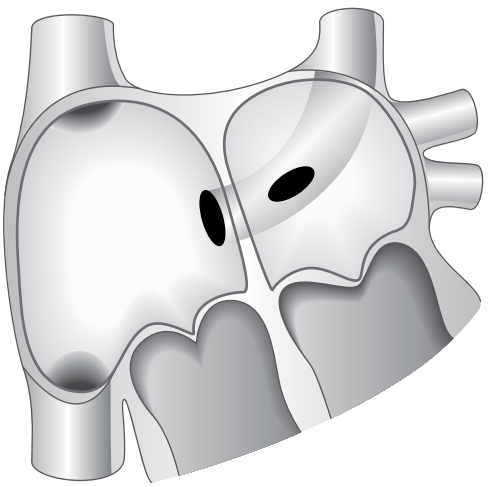

Figure 6.13 Example of persistent LSVC with unroofed coronary sinus. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Cardiomyopathy requiring orthotopic heart transplantation

Basics:

Heart transplantation may be indicated for unrepaired complex congenital cardiac defects, failed primary complete or palliative surgical procedures and heart failure caused by various forms of cardiomyopathy (CMO). Restransplantation for early and late organ failure also remains as an indication. Transplantation involves excising the native heart and implanting the donor organ. The new heart is commonly implanted by sewing the following vessels in this particular order: left atrium, inferior vena cava, superior vena cava, aorta, and pulmonary artery. Some cardiomyopathy patients receive an LVAD or BiVAD as a bridge to transplantation.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- FFP may be administered either preoperatively or in the prime for VAD patients on anticoagulant therapy.
- Cardioplegia is not normally needed. The donor heart receives cardioplegia immediately before harvesting. If the time before reperfusion of the donor heart in the recipient is greater than 4–5 h, the surgeon may elect to administer cardioplegia, either cold or warm, prior to reperfusion of the donor heart.
- The cross-clamp time for heart transplants refers to the time from cross clamping the donor heart to reperfusion of the donor heart in the recipient. Here, the cross-clamp time may be longer than the cardiopulmonary bypass time.
- Cardioplegia is not administered to the native heart but the perfusionist must still be vigilant of the bypass system pressure during application of the aortic cross clamp.
- Steroids are frequently administered to the circuit at the beginning of the bypass run and immediately before reperfusion of the donor heart.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 26–32 °C. Lower target temperatures may be used to help prevent rewarming the donor organ before reperfusion.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- It is prudent to monitor the myocardial reperfusion time since it is common for the surgeon to request updates during the rewarming/assessment period.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.
- Heart failure frequently negatively impacts the pulmonary vasculature. Inhaled nitric oxide is commonly available for the post-bypass period to treat pulmonary hypertension.

Case notes:

Surgeon Preferences:

Coarctation of the aorta

Basics:

Coarctation of the aorta (CoA) is a condition where the aortic blood flow path is restricted, usually in the area of the ductus arteriosus insertion (or ligamentum arteriosum for older patients). This restriction can impair lower body perfusion and increase the workload on the heart. If this defect requires surgical correction in isolation, it is frequently done without cardiopulmonary bypass. Other associated cardiac anomalies increase the probability of using bypass for correction. Surgical correction on bypass may include excision of the affected area with end to end anastomosis of the aorta, patch repair (especially when associated with arch hypoplasia), or subclavian flap repair (the subclavian artery is transected and folded down over the affected area that is opened to receive the SCA take-off).

Severe CoA may be associated with arch hypoplasia requiring long-segment repair of the aortic arch. Here, regional perfusion may be used. Regional perfusion can be provided with several different strategies. The overall goal with regional perfusion is to provide, at least, cerebral blood flow during the aortic arch repair. Additionally, continuous or intermittent myocardial and/or lower body perfusion can be incorporated as well (*see section "Flow rates, regional perfusion, and hypothermic circulatory arrest" in Chapter 3*).

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia common with traditional bypass. Alternately, left heart bypass may be used with the circuit bypassing the affected area and running from the LA to the descending aorta for isolated coarctation repair.
- Single venous cannulation (dual stage if available) may be used.
- The presence of septal defects normally requires the use of bicaval cannulation.
- Left ventricular vent common if bicaval cannulation is used.
- Target temperature is 24–28 °C. Target temperature must consider that lower body perfusion may be interrupted during the repair. Arch augmentation commonly requires a target temperature of 18 °C.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- The pump flows during traditional bypass may need to be reduced during the repair if the aorta is clamped around the coarctation since the pump will only be providing flow to the upper body (with limited descending aortic flow from collateral vessels). Regional perfusion techniques generally have pump flows of 20–60 mL/kg/min depending on what is being perfused, the NIRS value, and blood pressure.
- An isolated coarctation repair may employ a simple left atrial-to-femoral artery pump circuit. Here, the heart provides pulsatile blood flow to the upper body, while the LA-FA circuit provides continuous flow to the lower body during the coarctation repair. Ideally, upper and lower body arterial blood pressure lines are used to monitor and control flow between the two systems.
- A right radial arterial line is preferred if only one line is to be used since it allows for pressure monitoring during the repair since femoral and left sided pressures will not be useful during the coarctation repair in most cases (some regional perfusion techniques may provide lower body flow).
- If bypass is needed, extra attention must be paid to the rate of cooling for the lower body since flow may be impeded depending on the degree of coarctation. Higher pump flows may be helpful to assist with cooling.
- Some level of metabolic acidosis is expected if a cross clamp is used on the descending aorta during the repair without lower body perfusion.
- Increased vigilance during rewarming applies as well since any impediment to cooling the lower body that existed should be alleviated after the repair. The surgeon should be notified if the rate of lower body rewarming is below expected values.
- Topical cooling with a recirculating blanket may be especially helpful for these patients.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

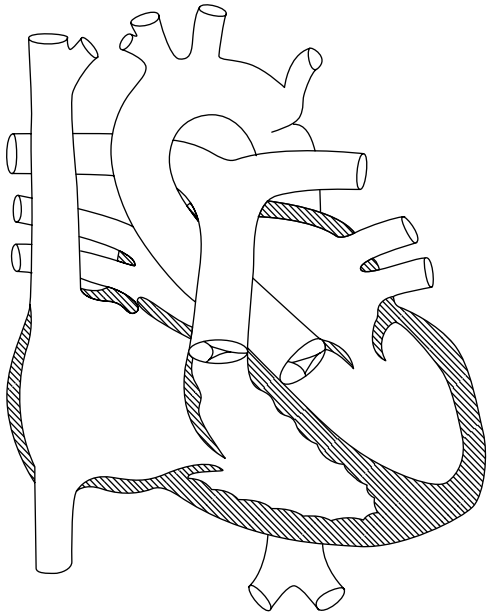

Figure 6.14 Coarctation of the aorta. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Common atrioventricular canal defect; same as complete atrioventricular canal, atrioventricular septal defect or endocardial cushion defect

Basics:

A common atrioventricular canal (CAVC) results when there is insufficient formation of endocardial tissue (cushions), which normally comes together to form both AV valves (tricuspid and mitral) and the atrial and ventricular septa. This insufficiency creates a single or "common" pathway between both atria and both ventricles with essentially one valve between the two, an atrioventricular valve (AV valve). The AV canal defect is additionally classified as balanced or unbalanced. Canal defects are balanced when the common AV valve is seated roughly evenly above the ventricular chambers. Canal defects are unbalanced when the common AV valve is seated such that it favors one ventricle over the other. This is an important distinction since an unbalanced anatomy is more likely to result in underdevelopment of the unfavored ventricle. Ventricular size is an important consideration with unbalanced defects since significant hypoplasia of one ventricle may preclude biventricular repair. CAVC can lead to heart failure due to a significant left-to-right shunt. Surgical correction is aimed at creating two functional AV valves (tricuspid and mitral) and closing the atrial and ventricular septal defects.

Partial atrioventricular canal defect (PAVC) exists as well. Here, there is insufficient primum septum that can extend to the mitral valve creating both a primum ASD and MR due to a cleft in the mitral valve. PAVC is synonymous with a Primum ASD.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 24–28 °C.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- PAVC may simply require suture closure of the mitral cleft and patch closure of the primum ASD.
- CAVC repair requires the common AV valve to be divided into two. This usually requires valve testing solution to be used during the reconstruction. The valve testing solution is usually saline and should primarily be sent to a cell saver or wall sucker.
- Excessive valve testing solution returned to the pump will require sodium bicarbonate for buffering. The patient's sodium may rise if bicarbonate administration is regular.
- Consideration should be given to informing the surgeon of the sodium load when the patient's sodium level rises above 150 mmol/L.
- Aggressive conventional ultrafiltration may be helpful in maintaining the hematocrit.
- Dilutional ultrafiltration (DUF) or zero-balance ultrafiltration (ZBUF) may be helpful in adjusting the sodium level when large volumes of test solution are returned to the pump.

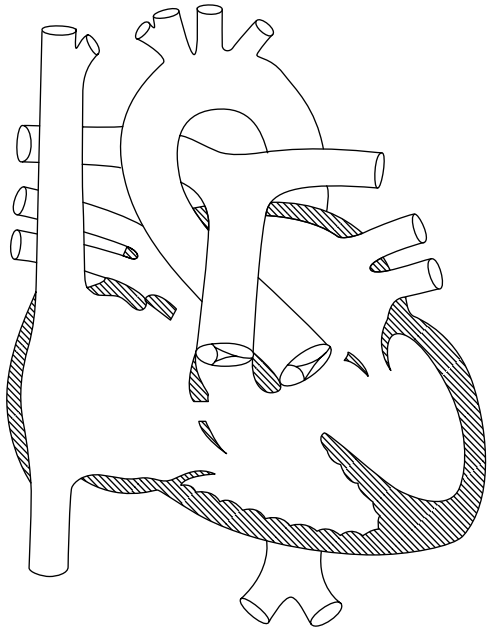

Figure 6.15 Complete atrioventricular canal. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Cor triatriatum

Basics:

Cor triatriatum literally means a heart with three atria. Here, there is a membrane within one of the atria that separates it into two. This occurs more commonly in the left atrium, resulting in a pulmonary venous (upper/proximal) chamber and a supramitral (lower/distal) chamber. The size of the communication between the chambers determines the severity of the disease. A significant membrane impeding pulmonary venous return to the left ventricle can lead to heart failure. If the membrane is small and the communication to the left ventricle adequate, cor triatriatum may be an incidental finding during the repair of other defects. Cor triatriatum less commonly is found on the right side. Surgical correction is aimed at excising the membrane within the affected atrium.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–30 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

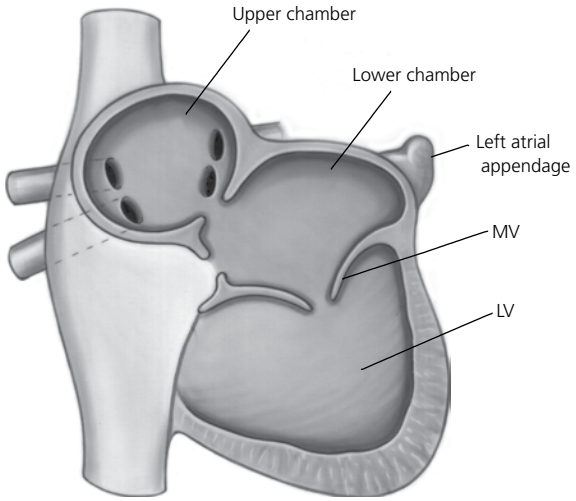

Figure 6.16 Cor triatriatum. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Corrected transposition of the great arteries (L-TGA, Levo-TGA, or C-TGA) or congenitally corrected TGA

Basics:

Corrected TGAs (CCTGAs) can be confusing at first study because the great arteries are indeed not transposed as with dextro-TGA. The aorta arises on the left side and the pulmonary artery arises on the right side as one would normally expect. The difference with L-TGA is that the ventricles rotated differently (leftward or levo) in utero with the result being that the morphologic left ventricle is right sided and the morphologic right ventricle is left sided. Therefore, the right-sided left ventricle pumps venous blood to the lungs and the left-sided right ventricle pumps arterial blood to the body. The ventricular position and discordance with its associated great vessel is how this defect is classified. The Van Praagh segmental classification for this defect would be the set {S,L,L) since the atria are situs, the ventricles rotated left, and there is ventriculoarterial discordance.

Associated defects (VSD, TR) with cyanosis may lead to early identification of this defect. However, L-TGA may exist with no symptomology until later in later life. The left-sided right ventricle can fail because it is not designed to provide systemic support. Surgical correction is aimed at making the morphologic right ventricle provide pulmonary blood flow and the morphologic left ventricle provide systemic blood flow. Most commonly, this is done with a double-switch procedure: atrial and arterial-level switches. This results in right-sided venous blood flowing to the left-sided right ventricle and on to the pulmonary artery. The left-sided pulmonary venous blood flows to the right-sided left ventricle and on to the aorta.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia is required. An additional cardioplegia dose should be available since this procedure may have an extended cross clamp time. Induced ventricular fibrillation may be used during the initial intracardiac assessment and the atrial switch procedure.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 22–30 °C.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

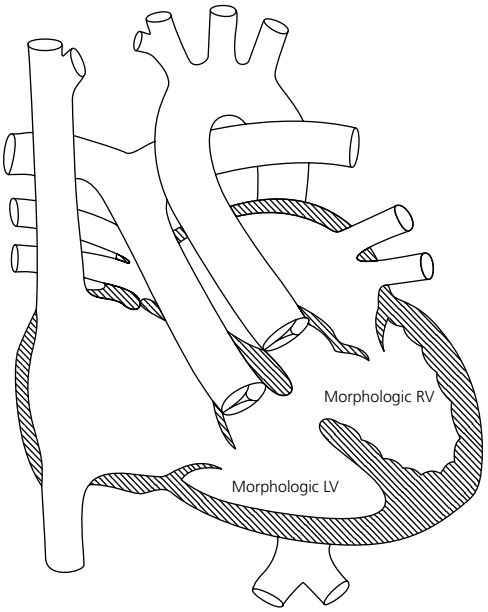

Figure 6.17 Congenitally corrected transposition of the great arteries with VSD. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

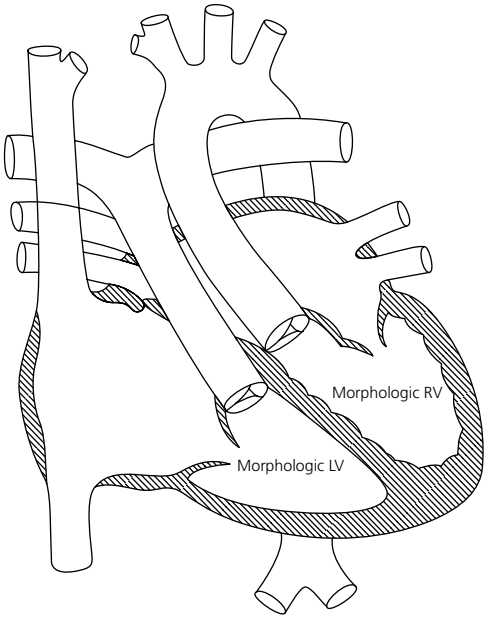

Figure 6.18 Congenitally corrected transposition of the great arteries with IVS. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Critical aortic stenosis

Basics:

Critical AS is the term normally applied to significant congenital aortic stenosis in the neonatal period. Aortic stenosis in older patients is simply referred to as AS. Critical AS can be due to valvar, subaortic, or supraaortic stenosis. Critical AS in the newborn may also be associated with small left-sided heart structures. A neonatal heart with critical AS can quickly progress to failure after birth. In utero and in the immediate postnatal period, a patent ductus arteriosus provides adequate systemic flow. If this vessel begins to close in the neonatal period, the heart progresses toward failure as the LV (which may be underdeveloped) has to pump against significant LVOTO. For this reason, PGE1 may be maintained to keep the PDA open. The aortic valve may also be balloon dilated in the catheterization lab before surgical repair. Surgical correction is aimed at opening the left ventricular outflow tract and providing a functional aortic valve. Less commonly, critical AS is associated with inadequate left heart structures and the patient receives staged operations towards a Fontan circulation.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- The perfusionist must be exceptionally prepared for these cases. Patients with critical AS do not respond well to CPR since either compression of the chest (closed CPR) or heart (open CPR) is met with significant outflow obstruction. Systemic blood flow and coronary blood flow are insufficient with CPR for AS patients and if cardioversion is difficult, bypass is the safest treatment. Therefore, the bypass pump should be primed as soon as possible to ensure there is no perfusion delay in case of the emergent need for bypass.
- Cardioplegia required. Consideration must be given to higher cardioplegia delivery pressures/flows if significant ventricular hypertrophy exists. This will help ensure proper myocardial distribution of cardioplegia.
- Single venous cannulation may be used.
- Most often, the presence of associated defects requires the use of bicaval cannulation for defect closure.
- Left ventricular vent is common.
- Target temperature is 24–28 °C.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 30–35%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management is performed.
- These patients may be hypercontractile since the LV has been pumping against significant obstruction. Such patients may require less venous reservoir volume to separate from bypass.
- The perfusionist needs to be extra cautious when increasing the pump flow immediately after cross clamp removal since the blood pressure may go to an unacceptably high level.
- If the heart was in failure, the LV may not be hypercontractile.
- Patients may exhibit high blood pressure once ejection is established. This must be controlled, in particular, because the aorta has been entered and now has a suture line. However, the surgeon may elect for a somewhat higher blood pressure to prevent ventricular fibrillation if there is hypertrophy.
- The anesthesia team will have medications to treat hypertension (if need be), but the perfusionist may need to intervene while the drips take effect (nitroprusside, nitroglycerin, phentolamine, etc.).
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

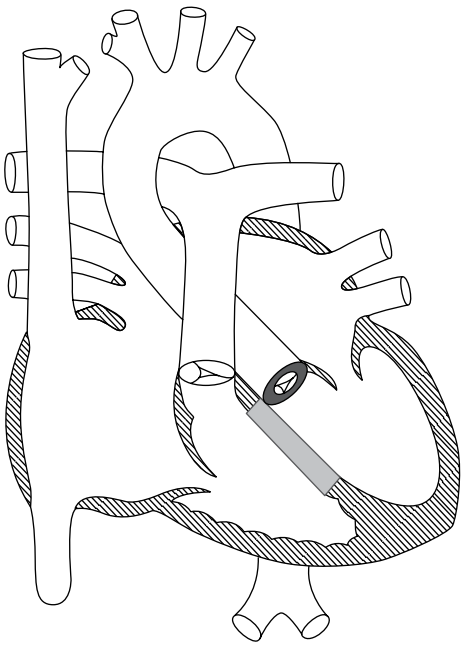

Figure 6.19 Critical aortic stenosis. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Double chambered right ventricle

Basics:

A double chambered right ventricle (DCRV) usually results from the presence of anomalous or hypertrophied muscle bundles in the RVOT. The presence of these bundles essentially creates two chambers in the RVOT. The more apical chamber creates high pressure with RV contraction and the chamber closest to the pulmonary valve tends to be lower pressure due to pressure loss across the significant muscle bundles. This impairs pulmonary blood flow and can lead to heart failure primarily due to right-sided hypertrophy. A VSD, or other congenital cardiac anomaly, is commonly associated. Surgical correction is aimed at resecting the muscle bundles while preserving tricuspid valve function. Associated defects are repaired as needed.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required. Consideration must be given to higher cardioplegia delivery pressures/flows if significant ventricular hypertrophy exists. This will help ensure proper myocardial distribution of cardioplegia.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

Case notes:

Surgeon preferences:

Double inlet left ventricle

Basics:

Double inlet left ventricle (DILV) usually presents with severe right ventricular hypoplasia. Commonly, there is ventricular inversion, discordant ventriculoarterial relationship, ASD, and VSD (bulboventricular foramen or BVF). Both the mitral and tricuspid valves empty into the right-sided left ventricle. Palliation usually provides for single ventricle physiology (Fontan circulation) as the presentation of two functional ventricles is rare. Operations may be staged from a Glenn shunt or hemi-Fontan toward an eventual Fontan procedure. However, some effectively single ventricle patients may receive a PA band as a first operation to better balance the pulmonary and systemic circulations during early infancy when pulmonary vascular resistance precludes passive pulmonary blood flow. A pulmonary artery banding procedure usually does not require bypass.

Bypass notes:

Effective single ventricle repair will involve a bypass plan similar to the staged operations for hypoplastic left heart syndrome.

See section "Hypoplastic left heart syndrome".

Notes:

Double outlet left ventricle

Basics:

Double outlet left ventricle (DOLV) usually presents with VSD, pulmonary stenosis, and right ventricular hypoplasia. The surgical correction will be aimed at biventricular repair if the right ventricle is deemed adequate, while single ventricle repair (Fontan pathway) is optioned if the right ventricle is inadequate.

Bypass notes:

The bypass plan will be dictated by the surgical plan for one or two ventricle repair.

Biventricular repair will likely involve a bypass plan similar to a TOF/PA repair.

See section "Tetralogy of Fallot".

Single ventricle repair will involve a bypass plan similar to a Stage 1 procedure.

See section "Hypoplastic left heart syndrome".

Notes:

Double outlet right ventricle

Basics:

Double outlet right ventricle (DORV) in its most basic form exists as a VSD with aortic override into the right ventricle. This may exist much like a TOF diagnosis (VSD, overriding aorta but without RVOTO or RVH), and the great arteries may be side by side. Here, surgical correction may simply involve a well-placed VSD patch that does not cause left ventricular outflow tract obstruction (LVOTO). At the other end of the spectrum, there is a VSD and the great vessels are rotated in such a way that the patient requires an arterial switch operation (Jatene procedure). The symptomology will be determined by the presentation spectrum.

Bypass notes:

The bypass plan will be dictated by the surgical plan.

A presentation with a VSD, PS, and overriding aorta will have a bypass plan similar to a TOF/PS patient.

See section "Tetralogy of Fallot".

A presentation with great vessels rotated such that the aorta is rightward in relation to the pulmonary artery will have a bypass plan similar to a d-TGA patient.

See section "d-Transposition of the great arteries".

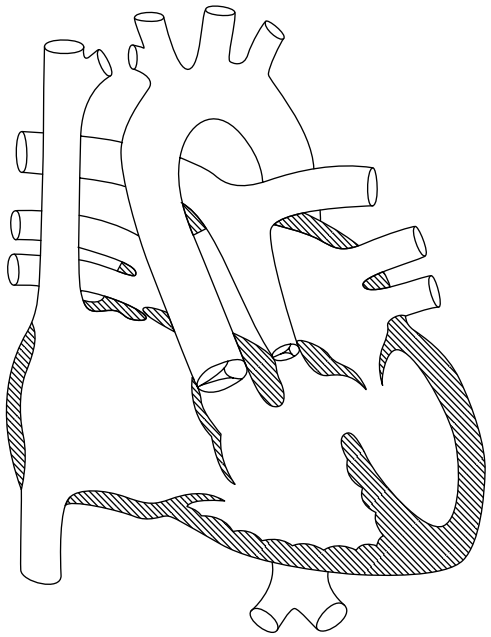

Figure 6.20 DORV, VSD, PS, Side by Side Great Arteries. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

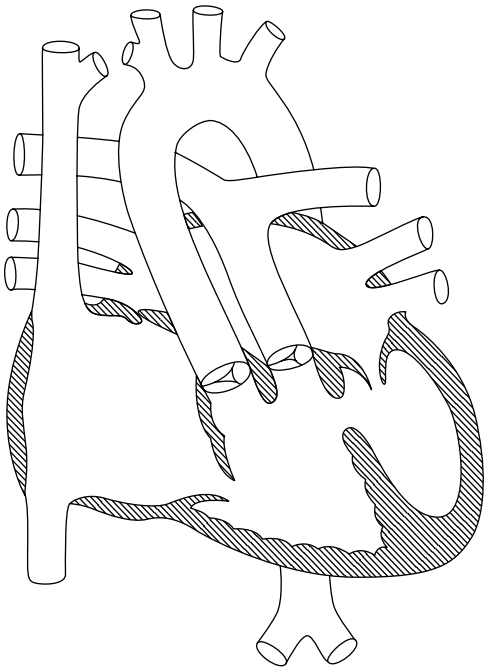

Figure 6.21 DORV, VSD, TGA. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Notes:

Ebstein's anomaly

Basics:

This defect is characterized by an apically displaced tricuspid valve. This results in an atrialized portion of the right ventricle (above the tricuspid valve) and an overall smaller effective right ventricular cavity. This tends to produce tricuspid regurgitation (TR) due to valve deformity or by leaflet attachment to the RV. Commonly associated findings are an atrial septal defect and electrophysiologic abnormalities including Wolff–Parkinson–White (WPW) syndrome. Surgical correction is aimed at repairing the tricuspid valve in its current location or by moving the valve basally and restructuring it (the cone procedure). Septal defect closure, annuloplasty ring placement and cryoablation for rhythm abnormalities are performed as needed.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required (induced ventricular fibrillation may be used during the assessment phase with cardioplegia during the repair).
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit of 30 during bypass with a HCT of 35 at termination of bypass.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- Surgical repair of the tricuspid valve requires valve testing solution as the procedure progresses. The valve testing solution is usually saline and should primarily be to a cell saver or wall sucker. Excessive valve testing solution returned to the pump will require sodium bicarbonate for buffering. The patient's sodium may rise if bicarbonate administration is regular.
- Consideration should be given to informing the surgeon of the sodium load when the patient's sodium level rises above 150 mmol/L.
- Conventional ultrafiltration may be helpful in maintaining the hematocrit.
- DUF or ZBUF may be helpful in adjusting the sodium level when large volumes of test solution are returned to the pump.

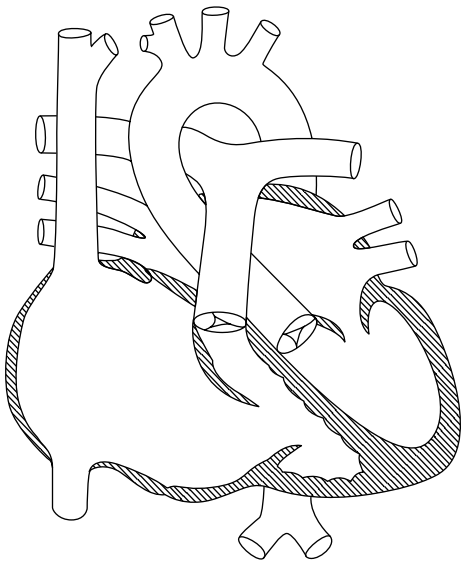

Figure 6.22 Ebstein's anomaly. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

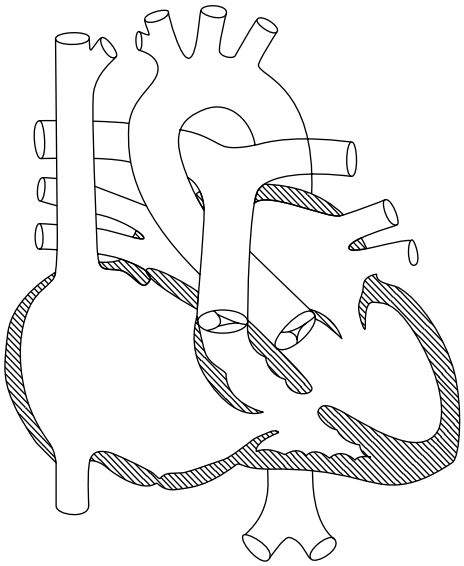

Figure 6.23 Ebstein's anomaly with VSD. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome (HLHS) is the most common cardiac defect that may lead a patient down the pathway of single ventricle palliation. However, other forms of single ventricle anatomy such as tricuspid or pulmonary atresia with an intact ventricular septum may lead to single ventricle palliation as well. The underlying principle is that only a single functional ventricle is present to provide systemic blood flow and pulmonary blood flow must be managed in staged procedures to optimize patient outcomes. The hypoplastic aortic arch is augmented and attached to the native pulmonary artery that becomes the neoaorta. The pulmonary arteries (left and right) form a continuous connection once the MPA insertion is oversewn. Pulmonary blood flow is most often provided from a Blalock–Taussig or Sano shunt. The atrial septum is excised to allow all heart return to flow to the pumping chamber. Next, as pulmonary vascular resistance sufficiently decreases by 4–6 months of age, the first-stage palliation is taken down and replaced with a cavopulmonary anastomosis (bidirectional Glenn shunt). Here, PVR has fallen enough to allow for some passive pulmonary blood flow. Finally, when PVR allows in the toddler years, the majority of pulmonary blood flow is routed through the pulmonary bed without direct ventricular support in what is most commonly referred to as the Fontan procedure. There are several variations to the staged procedures leading one down the Fontan pathway or what may also be termed the single ventricle pathway. The most common are discussed in the following text:

- Stage 1 procedure (with BTS or Sano)
- Hybrid Stage 1 palliation
- Stage 2 or BDG procedure
- Fontan procedure

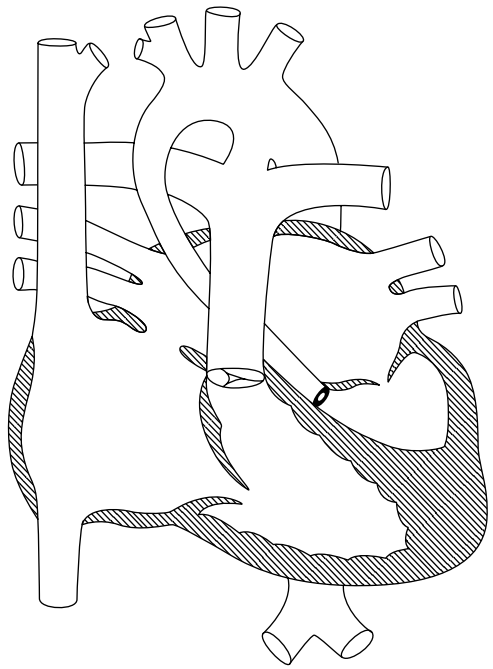

Figure 6.24 Hypoplastic left heart syndrome. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Stage 1 (Norwood) procedure

Basics:

This palliation is most often used as the first-staged operation for hypoplastic left heart syndrome. It is commonly performed during the first week of life. Arterial cannulation is variable but commonly, the pulmonary artery (for flow to the ductus arteriosus) is cannulated for arterial inflow since the true ascending aorta is inadequate. Alternately, the innominate artery is cannulated directly or a tube graft is anastomosed to the innominate artery into which the arterial cannula is inserted. This technique is more common for regional perfusion strategies. The main pulmonary artery branches are controlled once on bypass to prevent runoff to the pulmonary bed. Surgical correction is aimed at providing a single ventricular outflow tract for systemic flow and a smaller source for pulmonary blood flow. The hypoplastic aortic arch is augmented and attached to the native pulmonary artery which becomes the neoaorta. The pulmonary arteries (left and right) form a continuous connection once the MPA insertion is oversewn. Pulmonary blood flow is most often provided from a Blalock–Taussig or Sano shunt. The atrial septum is excised to allow all heart return to flow to the pumping chamber.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Arterial cannulation is variable as there are several options. The arterial inflow may be with the arterial cannula in the ductus arteriosus, the innominate artery, or a tube graft attached to the innominate artery (particularly for patients getting a BTS).
- Venous cannulation is most commonly with a single venous cannula in the right atrium.
- Bicaval cannulation may be used when circulatory arrest is not planned.
- An LV vent is not typically required.
- Target temperature is 18 °C if low flow and circulatory arrest are planned.
- Target temperature is 22–28 °C if selective cerebral (and possibly coronary) perfusion is planned.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Cardioplegia is required. It may be given through a traditional aortic root needle or by direct needle insertion in the aortic root. It is also possible to give cardioplegia at the onset of circulatory arrest through the arterial cannula (via a sidearm attached to the cardioplegia delivery system) with a cross clamp temporarily applied distal to the arterial cannula insertion site.
- Circulatory arrest is commonly used but is not an absolute.
- Regional perfusion techniques may limit the need for circulatory arrest. Generally, pump flows of 20–60 cc/kg/min through the arterial cannula with flow directed to the cerebral and possibly the coronary and splanchnic circulations (*see section "Flow rates, regional perfusion, and hypothermic circulatory arrest" in Chapter 3*).
- Cerebral and somatic NIRS monitoring is commonly used to evaluate the perfusion techniques utilized.
- Minimum dilutional hematocrit 30–35% during bypass and 40–45% coming off bypass.
- pH-stat and hyperoxic blood gas management are preferred. A crossover technique may be considered for circulatory arrest.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

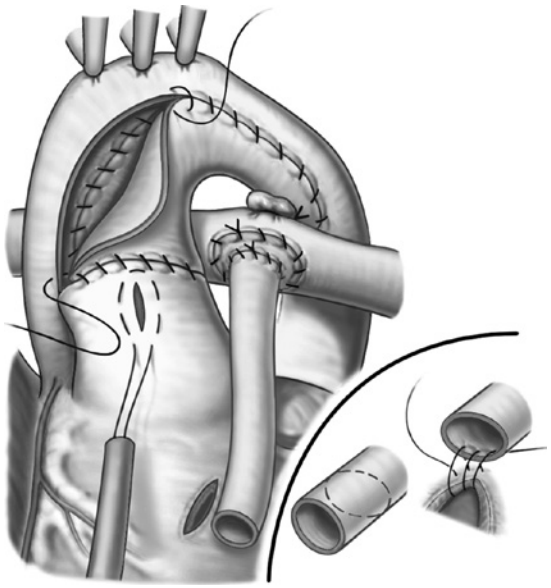

Figure 6.25 Stage 1 repair of HLHS with arch reconstruction, anastomosis of the proximal aorta to the pulmonary artery (neoaortic root), and a tube graft placed between the right ventricle and pulmonary arteries (Sano shunt). The atrial septum is also excised (not shown). From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

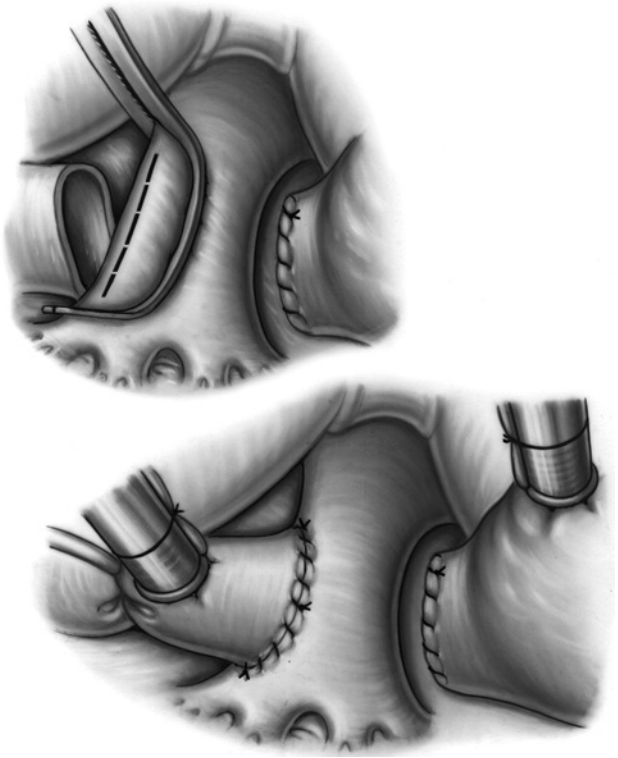

Figure 6.26 Bidirectional Glenn Shunt.

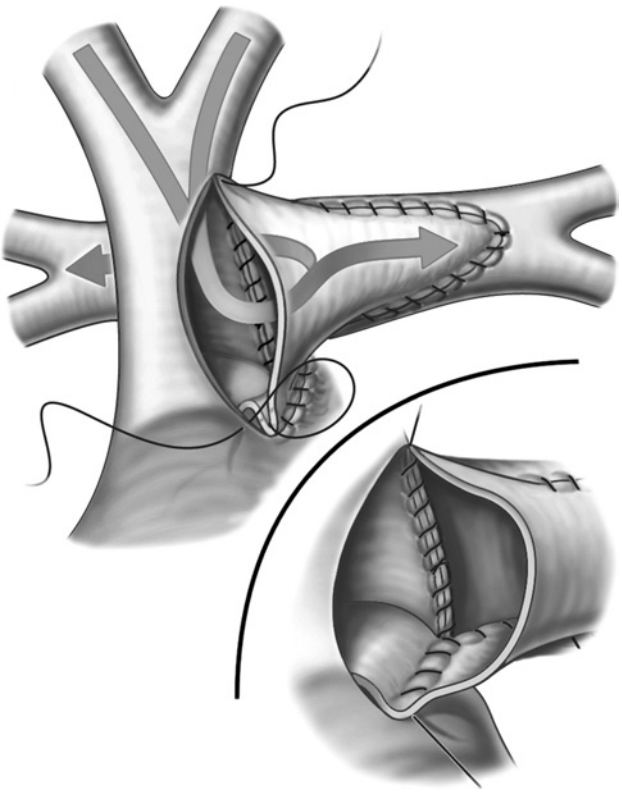

Figure 6.27 Hemi-Fontan Procedure.

Case notes:

Surgeon preferences:

Hybrid stage 1 palliation

High-risk patients with hypoplastic left heart syndrome may receive a hybrid stage 1 procedure instead of a Norwood operation as the first operation in life. Different institutions have their own definitions of high-risk patients that may include those with low birth weight, borderline right ventricular function, or coronary anomalies. The hybrid stage 1 is performed off bypass and commonly in a catheterization lab setting. Here, the ductus arteriosus is stented open and the right and left pulmonary arteries are banded. This arrangement provides an adequate amount of pulmonary blood flow while maintaining systemic output to the aorta (partially antegrade and partially retrograde via the ductus arteriosus). If a hybrid stage 1 is used as the first operation, a comprehensive stage 2 is performed on cardiopulmonary bypass as the next operation. The comprehensive stage 2 transitions the patient to the standard stage 2 (BDG) but requires arch repair, removal of the PA bands, and generally some pulmonary artery repair as well. Thereafter, the patient would receive a Fontan operation to complete single ventricle palliation.

Stage 2 or bidirectional Glenn shunt

Basics:

This palliation is most often used as the second staged operation for hypoplastic left heart syndrome. It is commonly performed a several months after the first-staged operation when pulmonary vascular resistance allows for a portion of the pulmonary blood flow to be passive (without a direct pumping chamber). This operation usually involves taking down the first stage's Blalock–Taussig shunt (BTS) or Sano shunt and connecting the superior vena cava to the right pulmonary artery in an end to side fashion. The superior aspect of the RA is oversewn. Some patients may present with a PLSVC that requires a second cavopulmonary connection as well. This would then be referred to as a bilateral BDG (BBDG). The BDG may also be termed a "superior cavopulmonary anastomosis."

An alternative to the BDG operation is the hemi-Fontan procedure. Here, the SVC-PA-RA is patched to divert SVC blood to the PA with an additional patch (also called a dam) preventing communication with the RA. This second patch facilitates an eventual lateral tunnel Fontan since it can simply be removed at the next staged intervention.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia is not normally required; it may be needed for tricuspid valve repair or to modify the atrial communication (usually to make it larger).
- Bicaval cannulation is required. The SVC cannula will be placed in the innominate vein or high in the SVC. The lower cannula may be placed in the atrium instead of the IVC if no additional intracardiac work is needed.
- Left ventricular vent normally is not needed.
- Target temperature is 30–34 °C.
- Minimum dilutional hematocrit is 30–35% on bypass and 40% coming off bypass.
- The heart is commonly kept beating for the procedure, but it is not normally necessary to maintain ejection.
- Be aware of the status of the BTS or Sano. If it is not controlled immediately on bypass, increased pump flows will be required to compensate for pulmonary runoff.
- Consideration should be given to higher pump flows since these cases are normally performed at greater than or equal to 30 °C on patients who normally are erythrocytotic.
- Adjustments to a higher hematocrit may be made earlier than normal during bypass since the pump run is generally short. Blood administration and the need for sodium bicarbonate and ultrafiltration to correct for electrolyte imbalances should occur well before the cavopulmonary anastomosis is complete since it is common to come off bypass within 10 min of this time point.
- Don't be alarmed if you hear the surgeon request a cross clamp (and cardioplegia has not been prepared). Cross clamps may be used to clamp the SVC below the venous cannula, to clamp the RA below the area to be oversewn, and/or to clamp the proximal RPA (or other pulmonary segment) during the repair.
- The displayed CVP value may become unusable if an applied SVC clamp includes the catheter.

Case notes:

Surgeon preferences:

Fontan procedure (total cavopulmonary anastomosis)

Basics:

This palliation is most often used as the third-staged operation for hypoplastic left heart syndrome. It is commonly performed in the early toddler years. The Fontan pathway results in the majority of venous blood flowing to the lungs without direct ventricular support. The SVC blood already flows to both lungs (bidirectional) via the RPA insertion with the Glenn procedure. The IVC blood flow can be routed to the lungs primarily in two ways. It can be "extracardiac" with a conduit to the pulmonary artery or "intracardiac" with a lateral tunnel to the RPA Glenn site. The lateral tunnel directs IVC blood up to the RPA with an anastomosis of the atrium to the RPA, just inferior to the SVC anastomosis. Both the lateral tunnel and extracardiac Fontan procedures can be fenestrated as well. Here, a small communication between the right-sided circulation and the direct pumping chamber circulation is made. The Fontan fenestration serves as an important pressure relief in case of acutely high pulmonary vascular resistance.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Larger patients presenting for a Fontan may be significantly erythrocytotic. These patients may not need blood in the prime from a hematocrit perspective. However, they may receive FFP preoperatively, in the bypass prime, or on bypass to aid in the correction of factor deficiencies.
- Cardioplegia required for lateral tunnel technique (induced ventricular fibrillation may be used during the assessment phase with cardioplegia commonly given during the repair).
- Bicaval cannulation is required. The upper cannulation is usually higher than normal (in the innominate vein) with the IVC cannulation lower than normal to facilitate creating the intra-atrial baffle or extracardiac conduit.
- Left ventricular vent is not always needed and will depend on surgeon's preference.
- A competent tricuspid valve is important in the Fontan circulation. A tricuspid valve repair may be included as part of the operation. Valve testing solution returned to the pump will require sodium bicarbonate for buffering.
- When the SVC is clamped in order to open the RPA, watch the NIRS and CVP as this is analogous to placing a caval tape.
- The displayed CVP value may become unusable if an applied SVC clamp includes the catheter.
- Target Temperature is 28–32 °C.
- Minimum dilutional hematocrit is 30–35% during bypass and 40% coming off bypass.
- Don't be alarmed if you hear the surgeon request a cross clamp (and cardioplegia has not been prepared). A cross clamp may be used to clamp the SVC below the venous cannula to facilitate establishing RA/RPA continuity for lateral tunnel Fontans.

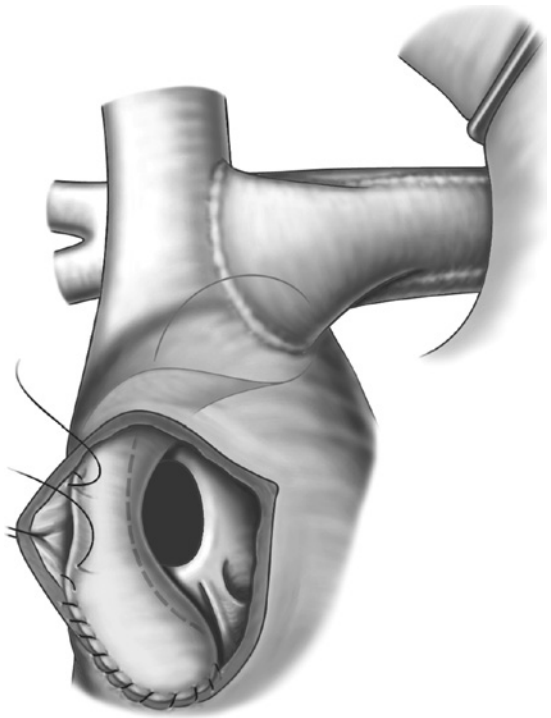

Figure 6.28 Lateral tunnel Fontan with baffle directing IVC blood flow to the lungs incorporating a prior Glenn Shunt. A fenestration in the baffle is also common (not shown). From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Interrupted aortic arch

Basics:

An interrupted aortic arch (IAA) requires a patent ductus arteriosus. The proximal arch and the coronaries are supplied normally via the left ventricular outflow tract and aortic root. The distal arch is supplied via the PDA. Septal defects (VSD and/or ASD) are always present. The type of IAA is classified by where the interruption occurs:

- Type A is interrupted distal to the left subclavian artery.
- Type B is interrupted between the left common carotid and left subclavian arteries.
- Type C is interrupted between the innominate and left common carotid arteries.

Surgical correction is aimed at connecting the two separate arch segments. Since both arch segments need blood flow on bypass, dual arterial cannulation is performed. An interrupted arch may require regional perfusion. Regional perfusion can be provided with several different strategies. The overall goal with regional perfusion is to provide, at least, cerebral blood flow during the arch repair. Additionally, continuous or intermittent myocardial and/or lower body perfusion can be incorporated as well (see section "Flow rates, regional perfusion, and hypothermic circulatory arrest" in Chapter 3).

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia required. Cardioplegia may be given with a traditional aortic root needle, direct needle insertion into the aortic root, or via the proximal arterial cannula via a sidearm. Sidearm cardioplegia delivery through the proximal aortic cannula is more common when circulatory arrest is used.
- Two arterial cannula (ascending aorta and PDA) are needed for true interrupted arches (as opposed to severe coarctation whereby one arterial cannula may be sufficient).
- Bicaval venous cannulation is common to deal with an associated VSD and/or ASD. However, some patients may require circulatory arrest for correction whereby single venous cannulation is used.
- Left ventricular vent common with bicaval cannulation.
- Target temperature 24–28 °C for bicaval cannulation without circulatory arrest.
- Target temperature 18–22 °C for single venous cannulation and circulatory arrest.
- Regional perfusion techniques may limit the need for circulatory arrest. Generally, pump flows of 20–60 cc/kg/min through the arterial cannula with flow directed to the cerebral and possibly the coronary and splanchnic circulations (*see section "Flow rates, regional perfusion, and hypothermic circulatory arrest" in Chapter 3*).
- Cerebral and somatic NIRS monitoring is commonly used to evaluate perfusion techniques utilized.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management is preferred for cases performed at 30 °C or below.
- Some level of metabolic acidosis is expected if a cross clamp is used on the descending aorta during the repair without lower body perfusion.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

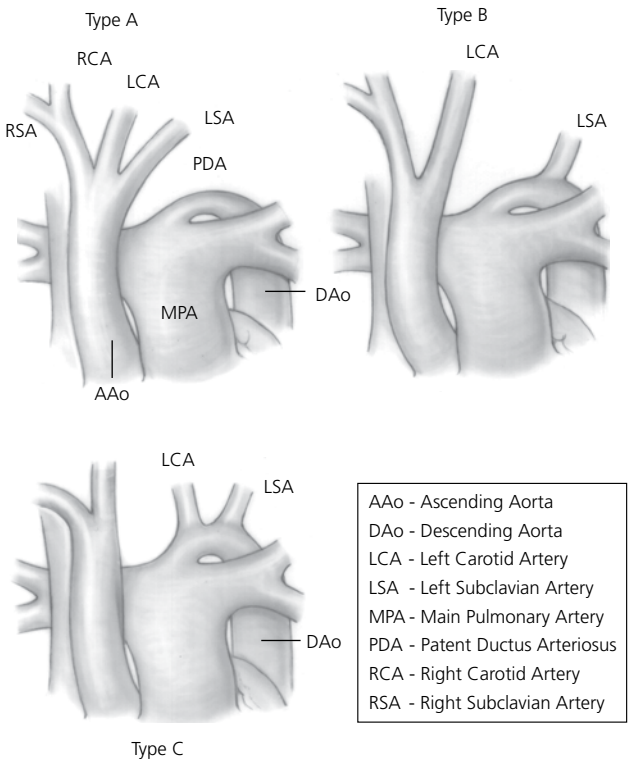

Figure 6.29 The three types of interrupted aortic arches. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Left superior vena cava

Basics:

An LSVC may be present when normal fetal development is altered for this vessel. It may persist in isolation with drainage to the coronary sinus or the left atrium. An additional bridging vein connecting this drainage to the right sided SVC may be present. Surgical correction is not normally needed for this defect in isolation. However, it may be addressed in conjunction with other congenital cardiac defects.

Bypass notes:

- The bypass plan for equipment selection, cannulation, cardioplegia, LV vent use, target temperature, hematocrit, and blood gas management will be determined by the associated cardiac defects.
- If a bridging vein is present or if the LSVC flow is deemed to be minimal, the vessel may not be addressed during cannulation, depending on the other surgery the patient requires. Here, additional right-sided flow (if drainage is to the coronary sinus) or left-sided flow (if drainage is to the left atrium) may simply be addressed with a pump sucker.
- The LSVC may be ligated below the bridging vein if such a pathway is deemed adequate for drainage.
- If a persistent LSVC is present without a bridging vein, a third venous cannula may be needed.
- A low flow sucker can be placed in the LSVC for drainage. This may make cannulation easier than adding a third venous cannula in consideration of limited surgical field space.

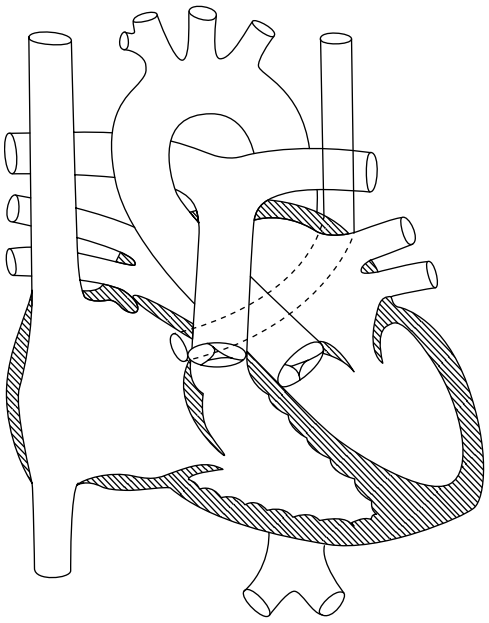

Figure 6.30 Left superior vena cava returning to the coronary sinus (without a bridging vein). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

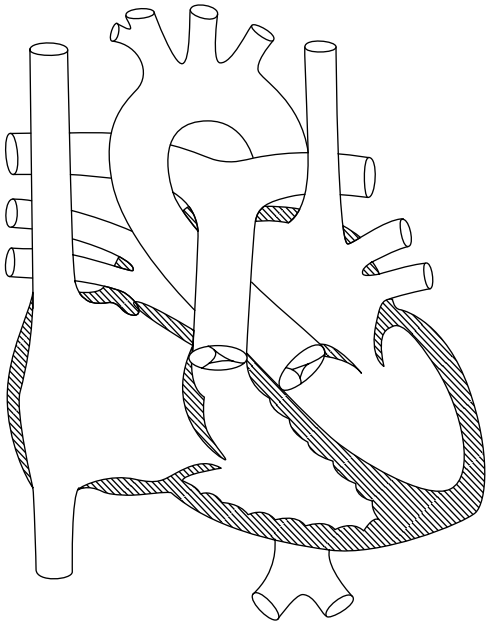

Figure 6.31 Left superior vena cava returning to the left atrium. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Lung transplantation

Basics:

Lung transplants are frequently for the diagnoses of cystic fibrosis, pulmonary hypertension, graft-versus host disease post bone marrow transplant and redo lung transplants. Repair at pediatric centers most often involves double lung transplantation.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is not normally needed.
- The cross-clamp time for these cases refers to the ischemic time of each lung. Each lung will have its own ischemic time since each is normally perfused as soon as possible once implanted.
- Single venous cannulation is common (dual stage if available).
- Left ventricular vent is not required.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- These patients may live with $PaCO_2$ values much higher than normal. If the goal is to normalize the blood gas values on bypass, the sweep gas flow rate will initially be higher than one would normally expect with a given oxygenator. If NIRS values are low, consider raising the $PaCO_2$ toward the patient's normal value.
- The antirejection drug antithymoglobulin (aka antithymocyte globulin or ATG) may be given after the recipient's lungs are removed.
- Immunosuppressive steroids are commonly given at the initiation of bypass and also once the first donor lung is perfused.
- Postoperative bleeding can be of particular concern, especially in patients with a primary diagnosis of cystic fibrosis. The addition of thawed FFP in the bypass circuit prime or during bypass is not uncommon.

Case notes:

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Surgeon preferences:

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Mitral regurgitation/insufficiency

Basics:

Mitral regurgitation/insufficiency (MR/MI) results from a dilated mitral annulus, diseased leaflets, or abnormal leaflet coaptation (including a leaflet cleft). Severe MR can lead to left atrial enlargement, atrial fibrillation and heart failure since the heart must pump more to maintain adequate systemic flow. Left ventricular hypertrophy and impaired myocardial perfusion can result. Surgical correction is aimed at repairing the physical valve or replacing it with a tissue valve or mechanical valve. Mitral valve repair in pediatric patients is favored over mechanical replacement since it allows for growth and may preclude the need for lifelong anticoagulation, especially in active young patients. Repair of the valve may include placing an annuloplasty ring that helps control the annulus size and provide for improved leaflet coaptation. Mechanical replacement is more common as one progresses through the adult years.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required. Consideration must be given to higher cardioplegia delivery pressures/flows if significant ventricular hypertrophy exists. This will help ensure proper myocardial distribution of cardioplegia.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- Valve irrigation solution is ideally scavenged with cell saver or wall suction.
- Valve testing solution may be scavenged with pump suckers or the cell saver suction.
- Excessive valve testing solution returned to the pump will require sodium bicarbonate for buffering. The patient's sodium may rise if bicarbonate administration is regular.
- Consideration should be given to informing the surgeon of the sodium load when the patient's sodium level rises above 150 mmol/L.
- Aggressive conventional ultrafiltration may be helpful in maintaining the hematocrit.
- DUF or ZBUF may be helpful in adjusting the sodium level when relatively large volumes of test solution are returned to the pump.
- Severe and/or long-standing MR may negatively impact the pulmonary vasculature. Inhaled nitric oxide is commonly made available when there is concern for pulmonary hypertension after bypass.
- It is not uncommon to see reoperative mitral valve repairs (*See section "Reoperations" in Chapter 5*).

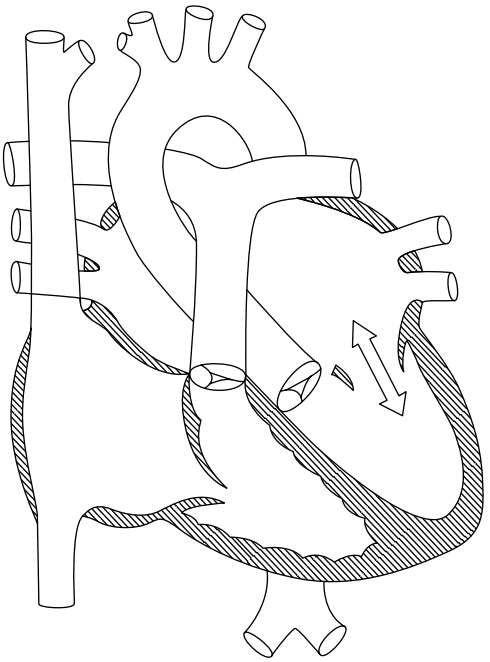

Figure 6.32 Mitral regurgitation. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Mitral stenosis

Basics:

Mitral stenosis (MS) is a condition where flow from the left atrium to the left ventricle is impaired across the mitral valve. In congenital cardiac patients this is often due to calcified mitral valve leaflets. In adults, MS is frequently associated with rheumatic fever. Regardless of cause, pulmonary congestion and subsequent heart failure can result. Surgical correction is aimed at repairing the physical valve (common in children) or replacing it with a tissue valve or mechanical valve. Mechanical replacement is more common as one progresses through the adult years.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required. Consideration must be given to higher cardioplegia delivery pressures/flows if significant ventricular hypertrophy exists. This will help ensure proper myocardial distribution of cardioplegia.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%. An increased hematocrit before coming off bypass must be considered for sick myocardium.
- Valve irrigation solution is ideally scavenged with cell saver or wall suction.
- Valve testing solution may be scavenged with pump suckers or the cell saver suction. It is not common for MS cases to require much valve testing solution. Therefore, elevated sodium levels due to sodium bicarbonate administration are not normally seen.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

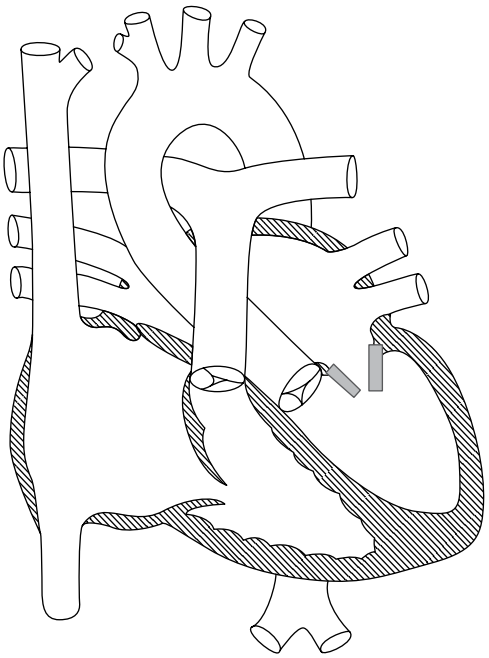

Figure 6.33 Mitral stenosis. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Patent ductus arteriosus

Basics:

The ductus arteriosus is a communication between the aorta and left pulmonary artery, which is normal and required for fetal circulation. It normally closes shortly after birth. The structure becomes the ligamentum arteriosum after ductal closure. Patients may present with a patent ductus arteriosus in isolation and may require a PDA closure without the use of bypass through a thoracotomy. Patients presenting with additional congenital cardiac defects, especially those requiring surgery in the first days or weeks of life, may have a ductus arteriosus which is maintained open with prostagladin E1. This may actually be quite beneficial in patients with limited pulmonary blood flow (hypoplastic left heart syndrome, pulmonary stenosis, etc.). However, once surgical correction for associated defects is performed, the PDA is generally ligated.

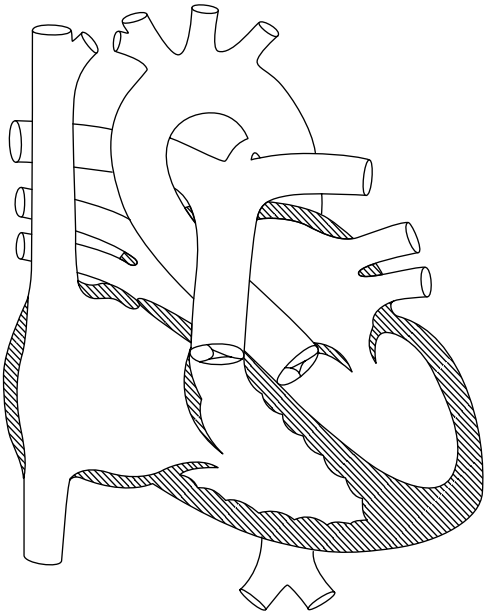

Figure 6.34 Patent ductus arteriosus. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Bypass notes:

- Cardioplegia, cannulation, and target temp depend on the nature of the associated defect(s) since isolated PDA closure is not normally performed on bypass.
- Initial pump flow will need to be higher with a patent ductus arteriosus before it is controlled at the field, usually within the first minute of bypass. The NIRS value may be particularly helpful in ensuring adequate pump flow if the ductus is not immediately controlled on bypass.
- The perfusionist should verify (verbally or visually if possible) that the ductus is closed before pharmacologically treating low blood pressure. If an alpha agent is given before ductal closure, the blood pressure may be unacceptably high once the duct is ligated.
- The rectal temperature will not cool normally until the ductus is controlled. This is important to consider when there is a concern regarding a possible coarctation of the aorta. The rectal temperature may initially cool slowly due to runoff through the duct. Cooling should resume normally after ductal ligation. If it does not, this may be contributing evidence to the presence of a coarctation.

Case notes:

Pulmonary artery abnormalities

Basics:

Stenotic or tortuous path pulmonary arteries may need to be surgically corrected on bypass. Most often, correction is performed at the time of repair for other defects although isolated pulmonary artery surgery does occasionally present. It is important that pulmonary blood flow is not impeded through one or both main pulmonary arteries since inadequate oxygenation and lung development may result. Preferential flow to one lung may also result in an unacceptably high PVR in the affected lung.

A pulmonary artery sling is where the left pulmonary artery arises anomalously from the right pulmonary artery and passes behind the trachea and esophagus. The course of the LPA may cause tracheal compression and significant respiratory symptoms. Bypass is commonly required for tracheal repair or for surgical correction at the main pulmonary artery site.

Bypass notes:

- Cardioplegia is not normally needed for isolated pulmonary artery repair. Pulmonary valve involvement increases the likelihood of using cardioplegia. An associated septal defect (PFO, ASD, VSD) will require cardioplegia and bicaval cannulation.
- Single venous cannulation may be used.
- Bicaval cannulation may improve visualization at the surgical field and decrease venous return problems on bypass (venous air locks).
- Left ventricular vent is common with bicaval cannulation.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

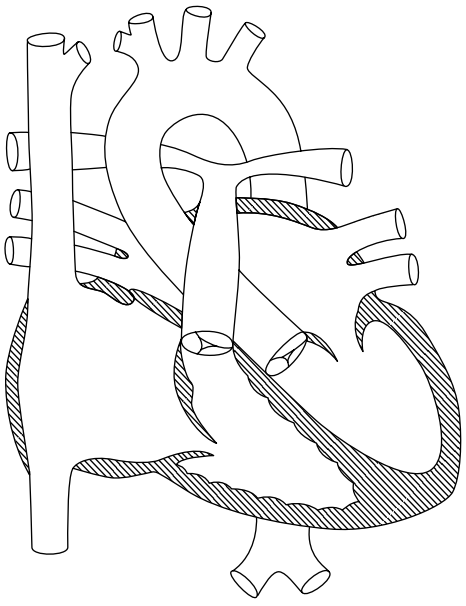

Figure 6.35 Branch PA stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

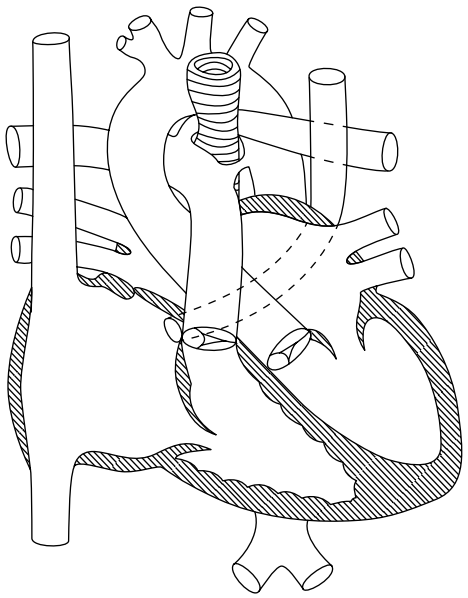

Figure 6.36 PA sling with tracheal compression (LSVC to CS also shown). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

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Pulmonary atresia

Basics:
Pulmonary atresia is normally diagnosed in the neonatal period and is classified either with an intact ventricular septum (PA/IVS) or with a ventricular septal defect (PA/VSD).

Pulmonary atresia with an intact ventricular septum

PA/IVS usually presents with small right heart structures due to inadequate right heart blood flow in utero. An ASD and patent ductus arteriosus are associated. If the right heart is functionally adequate, repair may be staged. Some patients may be treated with ductal stenting first. Others receive a balloon valvotomy first with or without a BTS off of cardiopulmonary bypass. Later, a surgical intervention is performed to open up the RVOT pathway and the pulmonary valve. Pulmonary blood flow from the RV may remain inadequate after relief of RVOTO due to poor RV compliance. If not already performed, a BTS is placed to provide sufficient pulmonary blood flow. The final stage would involve taking down the BTS and ensuring a clear RVOT with a competent pulmonary valve. Alternately, a BDG may be placed to provide pulmonary blood flow and the decision to pursue a one or two ventricle repair can be made at a later time.

To note, patients with PA/IVS may present with right-ventricle-dependent coronary circulation (RVDCC) with sinusoids in the RV supplying a portion of the coronary circulation. If at least some of the coronary blood flow is dependent on right ventricular pressure, the heart should not be decompressed on bypass since it would result in inadequate coronary perfusion. Relief of RVOTO in patients with RVDCC may be contraindicated. These patients normally receive a BTS to provide sufficient pulmonary blood flow. If the RVDCC is deemed minimal, cardioplegia may be given immediately before decompressing the right heart for surgical repair but adequate myocardial protection is not guaranteed.

If the right heart in PA/IVS cannot adequately support pulmonary circulation, the patient may be palliated with staged operations resulting in a Fontan circulation (*See section "Hypoplastic left heart syndrome"*).

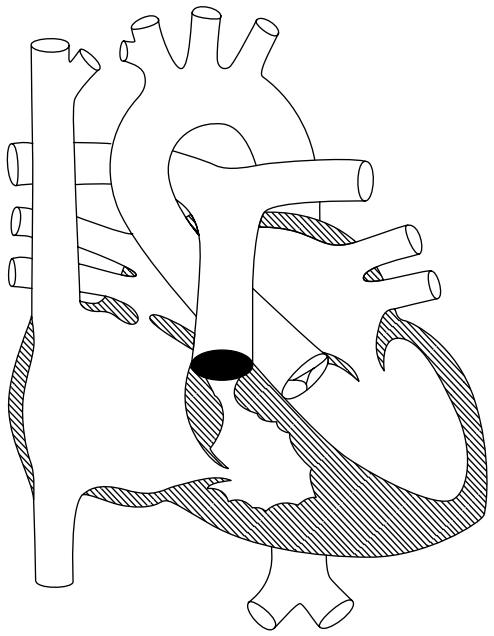

Figure 6.37 Pulmonary atresia with intact ventricular septum. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

PA/IVS bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2. Less commonly, if MAPCAs are present, an increased anticipated pump flow should be accounted for.
- A BTS may be placed off bypass as a first operation (along with a balloon valvotomy).
- Cardioplegia is required to repair the RVOT and close the ASD (possible fenestrated closure).
- The presence of RVDCC will determine if the heart can be decompressed normally when commencing bypass.
- Bicaval cannulation is required for intracardiac work.
- Left ventricular vent is common.
- Target temperature 32–35 °C for support during BTS placement only (though, most often, the BTS is placed off bypass).
- Target temperature is 24–28 °C for repair on bypass.
- Target temperature is 18 °C for planed circulatory arrest which may be used for significant RVDCC to aid in myocardial protection.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 30–35%.
- pH-stat blood gas management preferred.

Pulmonary atresia with ventricular septal defect

PA/VSD is very similar to TOF/PA, but there is no overriding aorta. PA/VSD can present with various anomalies such as atrial septal defect, absence of the main pulmonary artery, discontinuous pulmonary arteries, major aortopulmonary collaterals (MAPCAs) and hypoplastic pulmonary arteries. A patent ductus arteriosus is frequently present. Surgical correction with an adequate right ventricle is aimed at closing the VSD and providing pulmonary blood flow with a homograft and unifocalization of collateral vessels if possible. Pulmonary artery issues are also addressed. Much less commonly, the right ventricle may not allow for two ventricle repair and a staged approach for single ventricle palliation is taken (an inadequate RV is more likely to be seen with PA/IVS than PA/VSD).

PA/VSD bypass notes:

- Maximum anticipated flow for equipment selection: 3–5 L/min/m^2 depending on the anticipated collateral blood flow lost to the pulmonary system. If a patient is placed on bypass emergently before the APCs are controlled, the patient may need a high index flow. The oxygenator and filter system must be able to accommodate this.
- Control of associated aortopulmonary collaterals is paramount.
- APCs are frequently addressed before bypass (unifocalization) or at least controlled with vessel loops. MAPCAs must be addressed or controlled when possible since effective systemic and cerebral blood flow may be compromised once on bypass (*see section Aortopulmonary Collaterals on page 93*).
- Bicaval cannulation is required for RVOT repair and VSD closure.
- Cardioplegia is required for RVOT repair and VSD closure.
- A left ventricular vent is required if bicaval cannulation is used since the collateral flow increases return to the left side of the heart via the pulmonary circulation.
- Target temperature varies depending on associated defects.
- Consideration should be given to a lower target core temperature on bypass to help offset a potentially compromised cerebral blood flow due to collaterals.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Consideration should be given to a higher hematocrit (>30–35%) to help ensure adequate oxygen delivery to the brain in the setting of potentially compromised cerebral blood flow due to the collaterals.
- pH-stat blood gas management should be used. This method increases the CO_2 in the blood which provides dilation of the cerebral vessels aiding cooling and oxygen delivery while helping maintain adequate cerebral blood flow. Inadequate cerebral blood flow, despite an "appropriate" set pump flow, can lead to significant morbidity including stroke, choreoathetosis, and seizures.
- Consideration should be given to the amount of blood products on hand. Increased pump flows and sucker flows may increase the dynamic hold up and prime of the system (particularly the volume in the sucker tubing and cardiotomy) requiring additional products early in a pump run. The perfusionist should be cognizant that this need can be masked as poor venous return when the reservoir level drops but in fact may simply result from priming other parts of the circuit (the sucker and vent lines can become full of blood).
- The use of alpha agonists is debatable in this patient population. On the one hand, some clinicians believe that the blood pressure should be within its normal range regardless of diagnosis. On the other hand, some clinicians feel that alpha agonists simply increase the peripheral vascular resistance, and thus increase pulmonary blood flow for patients with APCs (the number may look good on the monitor, but effective systemic and cerebral blood flow may be suffering). The use of NIRS may be helpful in determining the effectiveness of alpha agents.
- Flow should be used first line to increase blood pressure but will be limited by venous return (and vent/sucker return) or possibly even the rated flow for the selected equipment.
- When bypass and/or sucker/vent flows are high (due to APC flow back to the left heart), there will be an increased need for CO_2 administered to the circuit since the blood CO_2 is effectively being scrubbed with an increased air:blood interface in the venous reservoir, vent, and suckers. CO_2 requirements may vary greatly throughout a bypass.
- An in-line blood gas monitor is essential to properly provide pH-stat in this patient population.

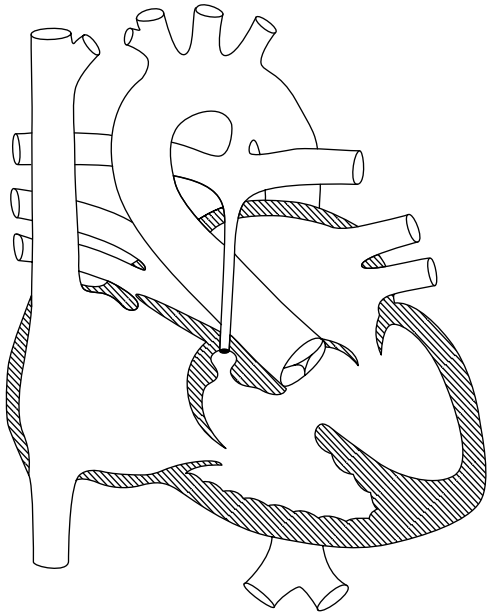

Figure 6.38 PA/VSD, PDA, confluent PAs. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

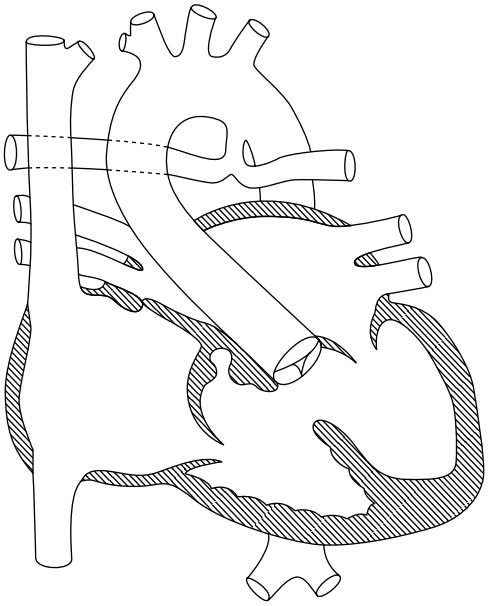

Figure 6.39 PA/VSD, PDA, abnormal PAs. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Pulmonary regurgitation/ insufficiency

Basics:

Pulmonary regurgitation/insufficiency (PR/PI) in congenital cardiac patients is frequently associated with TOF patients months to years after primary corrective surgery. However, it may be seen in isolation or in relation to other congenital defects. A certain degree of PR is tolerable, but significant regurgitation may lead to right heart failure. Surgical correction is aimed at repairing the pulmonary valve (common in children) or with valve replacement (more common in adults).

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia not required, but some surgeons utilize it to provide a more blood free surgical field.
- Induced ventricular fibrillation may be used.
- The presence of a septal defect (PFO, ASD, VSD) requires cardioplegia and bicaval cannulation.
- Single venous cannulation is common if there are no septal defects (dual stage if available for patient size).
- Left ventricular vent is usually only used with bicaval cannulation.
- Target temperature is 30–34 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

See section "Reoperations" in Chapter 5.

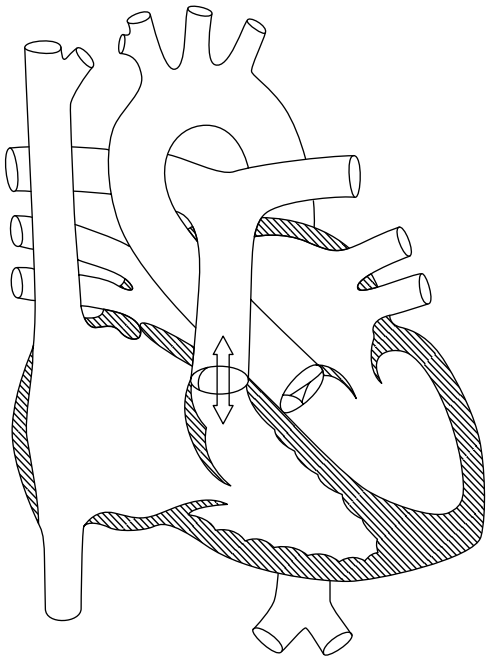

Figure 6.40 Pulmonary regurgitation. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Pulmonary stenosis

Basics:

Pulmonary stenosis is commonly classified by the location of the stenotic area. It can be classified as subvalvar, valvar, or supravalvar (just as aortic stenosis is classified). Significant pulmonary stenosis will result in right ventricular hypertrophy and right heart failure. Pulmonary stenosis can sometimes be treated in the catheterization lab. Significant stenosis not amenable to catheterization lab procedures requires removing the source of obstruction and ensuring the pulmonary valve is competent. Pulmonary stenosisis is commonly associated with TOF.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.

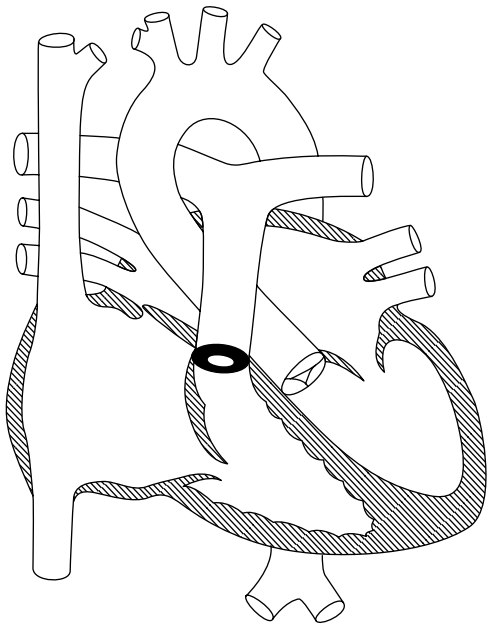

Figure 6.41 Valvar pulmonary stenosis. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

- Cardioplegia normally used though it isn't always necessary. An associated septal defect (PFO, ASD, VSD) normally requires cardioplegia.
- Single venous cannulation is possible (especially for supravalvar stenosis), but bicaval cannulation is more common.
- Left ventricular vent is common with bicaval cannulation.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25–30%.
- Valve irrigation solution should ideally be scavenged with either wall or cell saver suction.
- pH-stat blood gas management for cases performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

Case notes:

Surgeon preferences:

Pulmonary vein stenosis or pulmonary venous obstruction

Basics:

Pulmonary vein stenosis (PVS) may present as either a primary diagnosis or after an intracardiac repair for conditions such as total anomalous pulmonary venous return (TAPVR). It presents as a narrowing of the pulmonary veins, which impedes pulmonary blood flow from the lungs to the left atrium. This can result in pulmonary hypertension and right heart failure. PVS is commonly a progressive disease requiring surgical and catheterization lab interventions. Lung transplantation becomes an option for some patients. Surgical correction is aimed at relieving the stenosis. Most often, surgical correction involves a "sutureless" technique. This involves opening the stenotic veins with an incision extending as far back as practical into the pericardial reflection. A pericardial patch is then sutured to the left atrium over the entire area that prevents placement of suture line directly on the pulmonary veins (since suture here is thought to promote restenosis).

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required.
- Bicaval cannulation is common. Single venous cannulation is possible with planned circulatory arrest.
- Left ventricular vent is common with bicaval cannulation.
- Target temperature is 22–28 °C with bicaval cannulation. Target temperature is 18 °C for planned circulatory arrest.
- Lower target temperatures may be used to allow for low-flow bypass during the repair.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Increased drop sucker flow should be expected. This can result in priming of the sucker lines and an increased need for volume during the repair. Aggressive ultrafiltration during rewarming may be required.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management preferred.
- PVS frequently negatively impacts the pulmonary vasculature. Inhaled nitric oxide is commonly available for the post-bypass period to treat pulmonary hypertension.

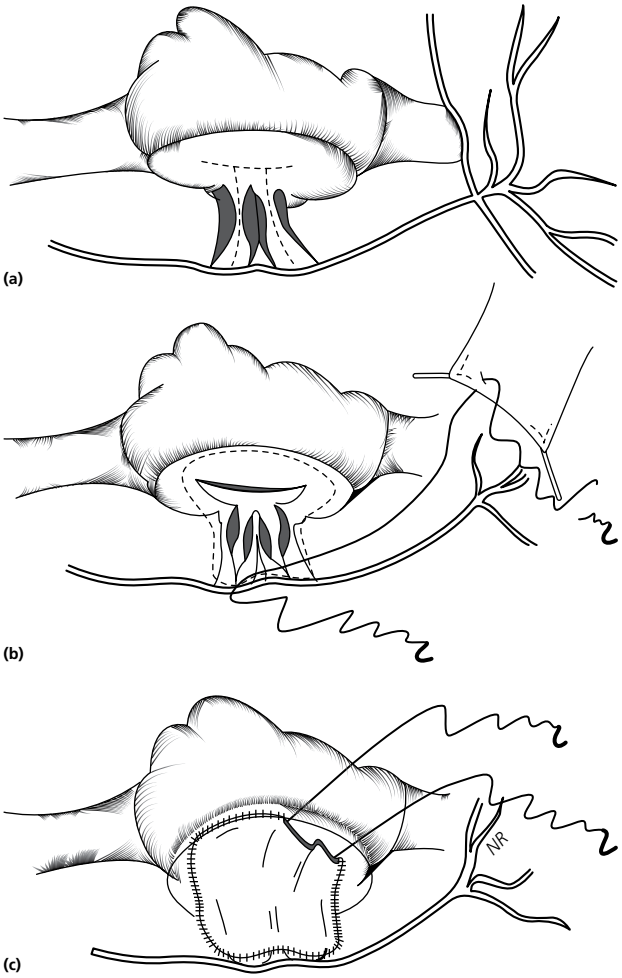

Figure 6.42 Repair of right PVO by using a "sutureless" free, pericardial patch. (a) PVO involves the right-sided PVs. (b) The obstructed PVs are widely incised. The incision is extended peripherally well beyond the obstructed segments. Proximally, it is carried across the PVs–LA junction. The dotted line indicates the line of suturing of the free pericardial patch. The suture line is kept away from the pulmonary venous endothelium. (c) The pulmonary venous pathway is reconstructed by using a free pericardial patch as shown. Care is taken to avoid injury of the right phrenic nerve and sinus node. From Ricci M, Elliott M, Cohen GA, *et al*. Management of pulmonary venous obstruction after correction of TAPVC: risk factors for adverse outcome. *Eur J Cardiothorac Surg*. 2003;**24**(1):28–36. Reproduced with permission of Oxford University Press.

Case notes:

Surgeon preferences:

Tetralogy of Fallot

Basics:

The TOF diagnosis encompasses a spectrum of severity with four primary findings: VSD, overriding aorta, right ventricular hypertrophy, and right ventricular outflow tract obstruction with either pulmonary stenosis (TOF/PS) or pulmonary atresia (TOF/PA). TOF with absent pulmonary valve is another variation, albeit much less common. Patients with an atrial septal defect may be classified as having pentalogy of Fallot. Patients with TOF/PS most commonly need VSD closure with relief of RVOTO via balloon dilation and/or a transannular patch across the pulmonary annulus. Patients with TOF/PA or those with an absent pulmonary valve (TOF/APV) most commonly need an RV-to-PA conduit in addition to the VSD closure, usually with a pulmonary arterioplasty. TOF patients, especially those with pulmonary atresia or absent pulmonary valve, and/or those existing outside of the neonatal period, are more likely to have aortopulmonary collateral vessels (*See section "Aortopulmonary collaterals*").

Bypass notes:

- Cardioplegia is required.
- Bicaval cannulation is common.
- Single venous cannulation is more likely in very small neonates (<3 kg) and those with additional defects, especially LSVC to coronary sinus where tricaval cannulation would be difficult and single venous cannulation would have the advantage of capturing all of the venous return. Single venous cannulation cases for TOF are more likely to utilize circulatory arrest, but it's not an absolute. The single venous cannula can be directed to the IVC during the intracardiac repair with a field sucker accounting for the balance of venous return under low flow conditions.
- A single venous cannula increases the likelihood of large swings in PCO_2 values during the repair. This is due to increased sucker flow and an increased chance of air in the venous line during the on-bypass intracardiac repair.
- Left ventricular vent common with bicaval cannulation. The presence of MAPCAs will increase left heart return and vent flow on bypass. Priming of the vent line may require the addition of volume to compensate, especially with smaller patients.
- Target temperature is 24–28 °C, and it is 18 °C for circulatory arrest.
- Minimum dilutional hematocrit is 30% on bypass, 30–35% coming off bypass; higher for neonates and those with APCs.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- pH stat blood gas management is preferred.
- Be particularly cautious with older unrepaired TOF patients who are more likely to have MAPCAs (*See section "Aortopulmonary collaterals*").
- Patients with exceptionally thickened right ventricles (primarily those outside of the neonatal period) may be sensitive to volume during weaning and in the immediate post-bypass period. Be prudent not to overfill the heart during weaning off bypass and when transfusing post-bypass.

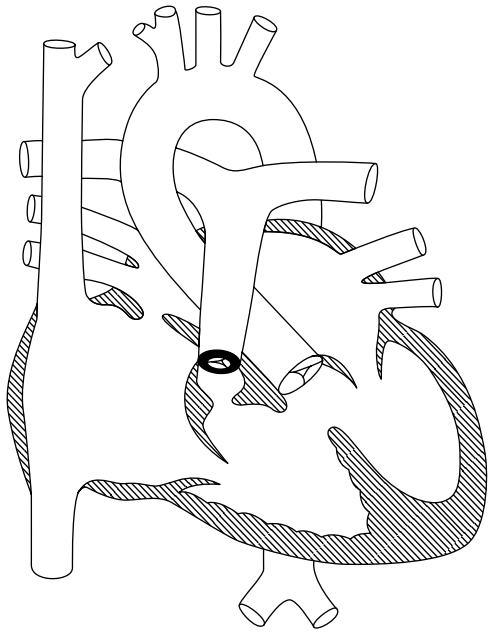

Figure 6.43 Tetralogy of Fallot. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

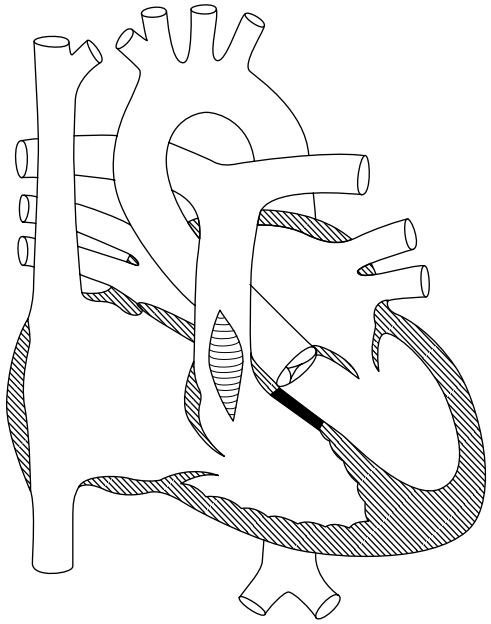

Figure 6.44 TOF repair with VSD patch and transannular outflow patch. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Total anomalous pulmonary venous return and partial anomalous pulmonary venous return

Same as total/partial anomalous pulmonary venous connection (TAPVC/PAPVC)

Basics:

TAPVR can be classified by where the pulmonary venous confluence is located. It may be supracardiac (commonly to a confluence behind the heart, then to a vertical vein and then the innominate vein and RA), cardiac (commonly to the coronary sinus), or subdiaphragmatic (draining to the IVC or portal vein) or mixed between these classifications. When the pulmonary veins empty into the right heart, a septal defect must be present to allow for right to left shunting and left-sided blood flow. To note, if the pulmonary veins are obstructed enough, a true crisis exists, and the patient will require immediate surgery since medical management is ineffective. Surgical correction for TAPVR is aimed at creating a pathway for the pulmonary veins to the left atrium. Hypothermic circulatory arrest is commonly used.

Partial APVR (PAPVR) may also present where some of the pulmonary veins return normally and some do not (as with sinus venosus ASDs with right-sided pulmonary veins returning to the RA).

Bypass notes:

- Cardioplegia is required.
- Bicaval venous cannulation may be used in older children and adults. In this population, circulatory arrest is less likely, and a target temperature of 24–28 °C is common.
- Single venous cannulation is more likely in neonates and infants. However, bicaval cannulation may be used in this population since it allows for intracardiac surgery for associate defects during the cooling phase. A single venous cannula can then be used during rewarm if desired. The target temp is 18 °C if the surgeon wants circulatory arrest to be a readily available option. Otherwise, 20–28 °C is used.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- The venous line is usually clamped during circulatory arrest when a single venous cannula is used. This allows the surgeon to remove the cannula to facilitate visualization at the field.
- If bicaval cannulation is used and caval tapes are applied, the venous line may be left unclamped during circulatory arrest.
- A left ventricular vent is not normally used with single venous cannulation but is common with bicaval cannulation.
- A drop sucker in the left atrium may be used with bicaval cannulation.
- The LA may be undersized if flow through it has been limited due to the anatomy. These patients will likely be more difficult to precisely wean off bypass. These patients may see large variations in filling pressures with minimal change in volume status during weaning. The perfusionist must be exceptionally careful not to overfill these patients during separation from bypass.
- Scimitar syndrome is a form of PAPVR and is named for the X-ray appearance of the enlarged curved sword (scimitar) shaped pulmonary venous drainage from the right lung to the IVC. The right lung is commonly hypoplastic and its blood supply is via MAPCAs.

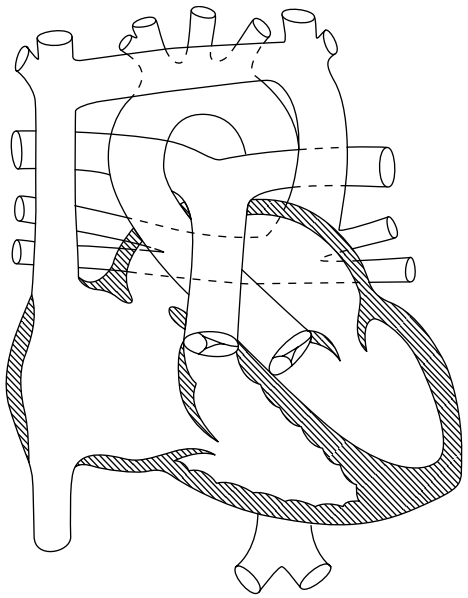

Figure 6.45 Supracardiac total anomalous pulmonary venous return to the left vertical vein. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

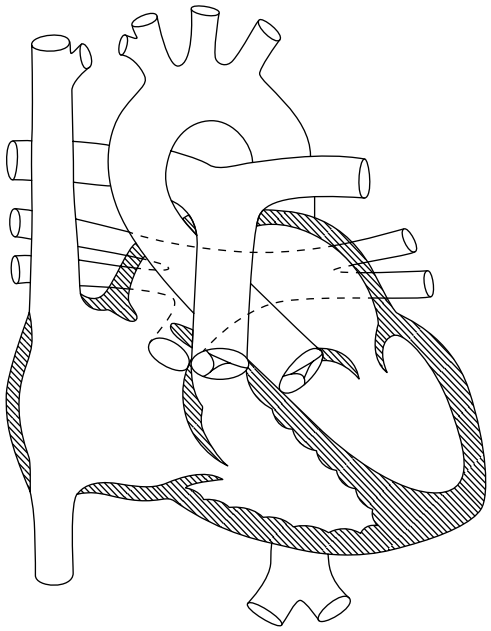

Figure 6.46 Cardiac total anomalous pulmonary venous return to the coronary sinus. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

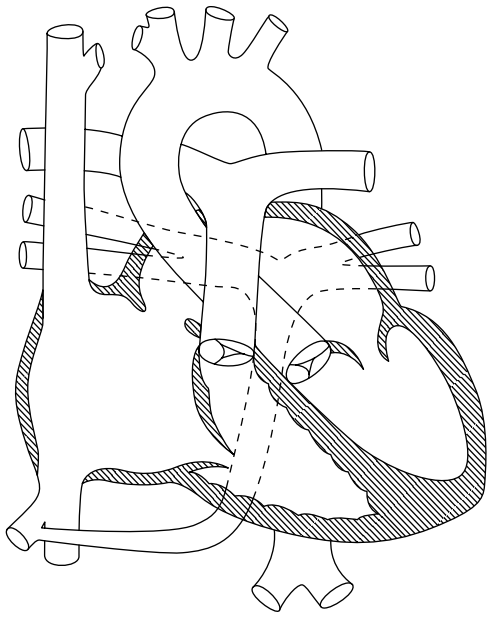

Figure 6.47 Infracardiac total anomalous pulmonary venous return to the ductus venosus. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

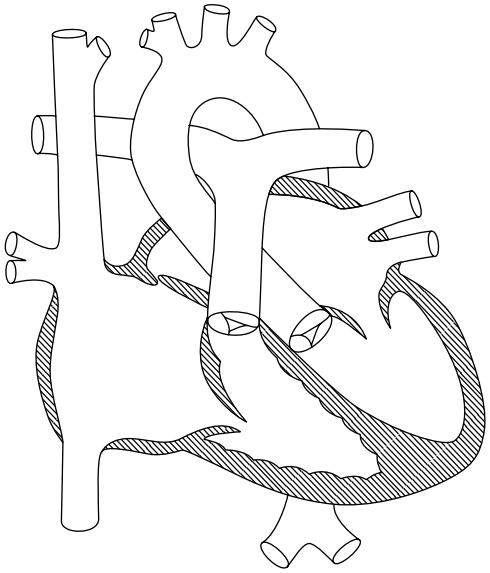

Figure 6.48 Partial anomalous pulmonary venous return with right pulmonary veins returning to the junction of the superior vena cava and right atrium. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

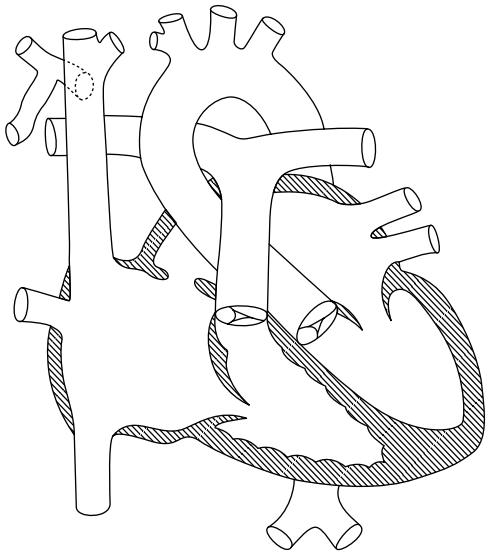

Figure 6.49 Partial anomalous pulmonary venous return with RUPVs returning to the superior vena cava and the right lower pulmonary vein returning to the right atrium. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

d-Transposition of the great arteries

Basics:

Transposition of the great arteries is just that; the aorta and pulmonary artery are transposed to the other's ventricle. The atria and ventricles are morphologically normal and located appropriately. The right ventricular outflow is to the aorta, while left ventricular outflow is to the pulmonary artery. This relationship is classified as discordant. The Van Praagh segmental classification set is {S,D,**D**}. The term "d-TGA" originates from the last letter in the segmental classification which indicates the ventriculoarterial relationship.

TGA is always associated with at least one septal defect (ASD/PFO/VSD) and at times more (ASD/PFO and VSD). A PDA may be present as well. In fact, adequate shunting at the septal level (or PDA) is required to support life. Patients are generally classified as TGA/IVS (TGA with intact ventricular septum) or TGA/VSD (TGA with VSD). Patients with inadequate ductal and/or septal shunting frequently receive prostaglandins and a balloon atrial septostomy early in the postnatal period to ensure that the two parallel circulations created by d-TGA mix well enough to support life. Collateral blood flow to the lungs may be present with d-TGA and can affect the bypass cannulation strategy.

In the normal heart, the left coronary arises from the left/anterior sinus of Valsalva and the right coronary artery arises from the right/anterior sinus of Valsalva. In d-TGA, the relationship between the great vessels is altered. Coronary arteries in d-TGA are frequently termed "normal for d-TGA." Normal for d-TGA is for the left coronary artery to come off of the left/anterior sinus of Valsalva (which is the same as a normal heart) and for the right coronary artery to come off the right/posterior sinus of Valsalva (in the normal heart the RCA comes off the right/anterior sinus of Valsalva). There are numerous other branching patterns that can be seen as well as single coronary artery.

Surgical correction is normally an arterial switch operation (ASO). The aorta and pulmonary artery are transected and moved to their respective ventricle. The coronary arteries are removed in buttons from the right-sided aortic root, mobilized, and then transferred to the left-sided pulmonary root (which becomes the neoaortic root). Significant stenosis of either outflow tract may preclude the switch operation and require an alternate surgical approach with a Nikaidoh or Rastelli type repair.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia is required.
- Left ventricular vent is common.
- Target temperature is 24–28 °C with bicaval cannulation. Here, the septal defects are repaired on bypass. This is more common in patients with a VSD.
- Target temperature 18 °C with single venous cannulation. Here, the septal defects are repaired under circulatory arrest. Deep hypothermia may be especially helpful with multidose cardioplegia protocols as temperature alone can have an arresting effect and direct redosing of cardioplegia in neonates is difficult.
- IVS patients are more likely to have single venous cannulation. Here, the ASD can be closed with the venous cannula directed down the IVC. The target temperature in this case would be 24–28 °C. Alternately, the ASD can be closed under circulatory arrest with a target temperature of 18 °C.
- Single venous cannulation may be preferred if significant collateral flow is expected. Bicaval cannulation may result in difficult intracardiac visualization with collaterals causing significant left heart return during ASD and/or VSD closure.
- An ice wrap applied to the head to prevent warming is common for procedures 25 °C and below.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management is preferred.
- The coronary transfer is critical with arterial switch operations. Early signs of coronary issues during rewarming are usually addressed surgically. The perfusionist should have additional cardioplegia ready in case an additional period of myocardial arrest is required.
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

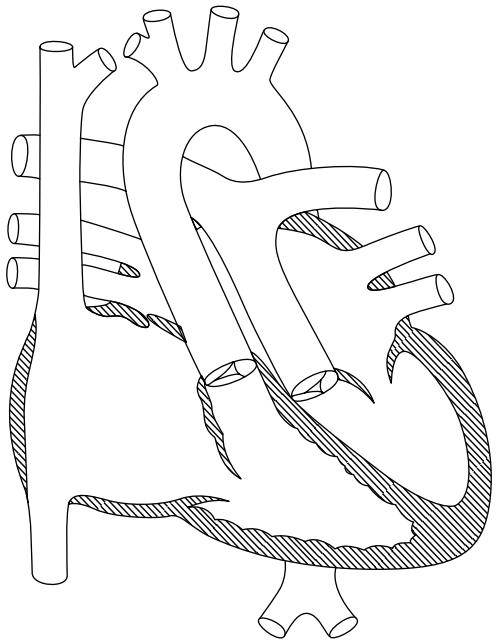

Figure 6.50 d-TGA with IVS (before atrial septostomy). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

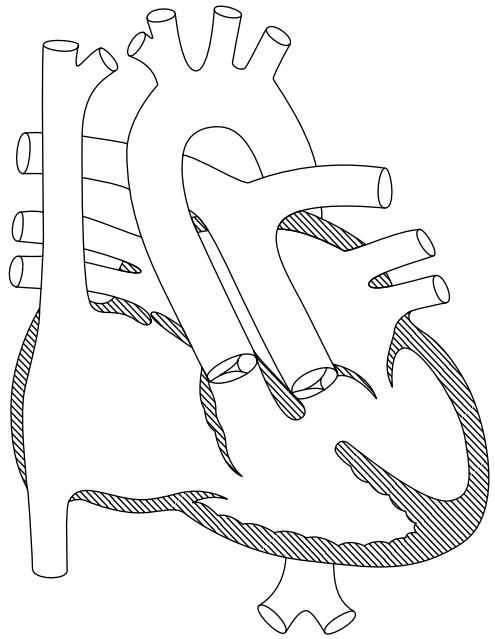

Figure 6.51 d-TGA with VSD. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

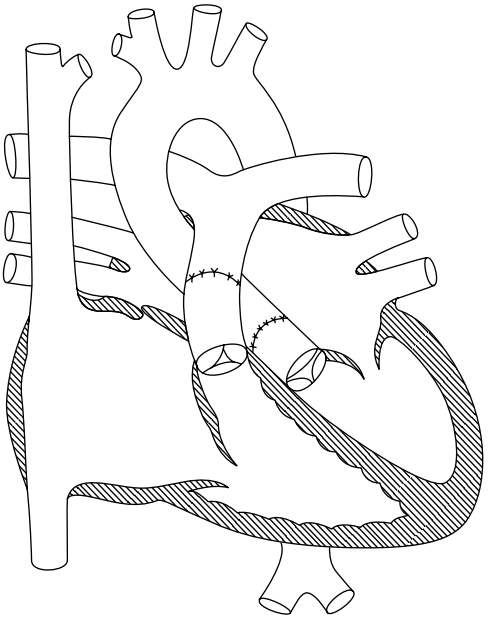

Figure 6.52 Arterial switch operation for d-TGA/IVS (coronaries are moved from the right-sided aortic root to the left-sided pulmonic/neo-aortic root). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

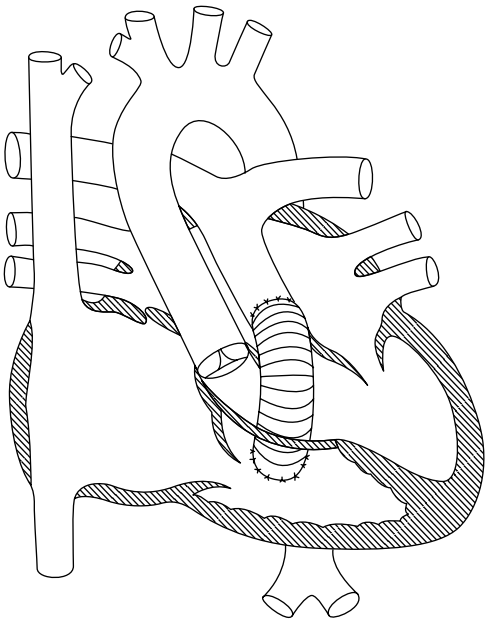

Figure 6.53 Rastelli repair for d-TGA/VSD/PS (VSD is patched to the aorta and a conduit is placed from the RV to the PA). From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Tricuspid atresia

Basics:

Tricuspid atresia in the neonate is normally seen with inadequate right ventricular size (due to the lack of blood flow through the RV in utero), an ASD, VSD, and a PDA. The pulmonary valve in tricuspid atresia can be atretic, dysplastic, or unrestrictive. Surgical repair is dedicated to ensuring a stable source of pulmonary blood flow, which may require a BTS, PAB, or no intervention at all if the pulmonary and systemic circulations are balanced with septal defects and sufficient antegrade flow through the PA. If the right heart is unable to provide pulmonary blood flow, the patient is usually led down the pathway of staged procedures toward a Fontan circulation. A Damus–Kaye–Stansel procedure may be performed when tricuspid atresia is associated with TGA and a restrictive VSD.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.5 L/min/m^2.
- Cardioplegia is not normally needed for BTS or BDG procedure. It is normally required for intracardiac work.
- Single venous cannulation is used for BTS procedure.
- Bicaval cannulation is used for BDG and Fontan procedures.
- Left ventricular vent is not normally needed.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 30–40% for single ventricle palliation.
- pH-stat blood gas management for cases is performed at 30 °C or below.

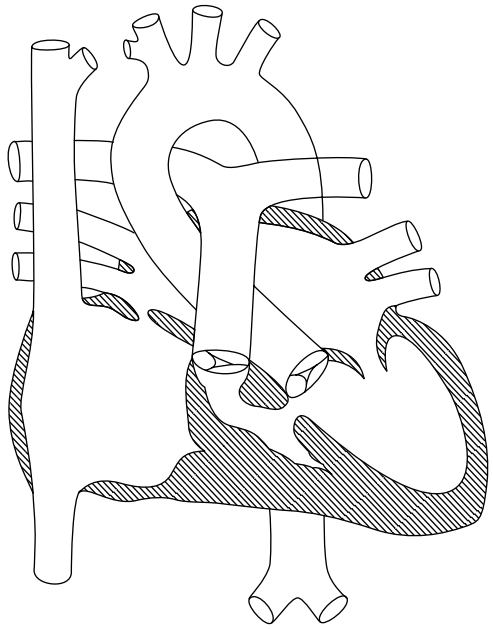

Figure 6.54 Tricuspid atresia with BVF type ventricular septal defect and unrestrictive pulmonary artery. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Tricuspid regurgitation/insufficiency

Basics:

Tricuspid regurgitation/insufficiency (TR/TI) is commonly due to right ventricular dilation that can be caused by numerous conditions. Significant TR can lead to right heart failure. TR is commonly associated with Ebstein's Anomaly and as a late finding after repair of complete atrioventricular canal defect. Repair of the valve may include placing an annuloplasty ring which helps control the annulus size and provide for improved leaflet coaptation.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia is required.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.
- Valve irrigation solution is ideally scavenged with cell saver or wall suction.
- Valve testing solution may be scavenged with pump suckers or cell saver suction
- Excessive valve testing solution returned to the pump will require sodium bicarbonate for buffering. The patient's sodium may rise if bicarbonate administration is regular.
- Consideration should be given to informing the surgeon of the sodium load when the patient's sodium level rises above 150 mmol/L.
- Aggressive conventional ultrafiltration may be helpful in maintaining the hematocrit.
- DUF or ZBUF may be helpful in adjusting the sodium level when relatively large volumes of test solution are returned to the pump.

See section "Reoperations" in Chapter 5.

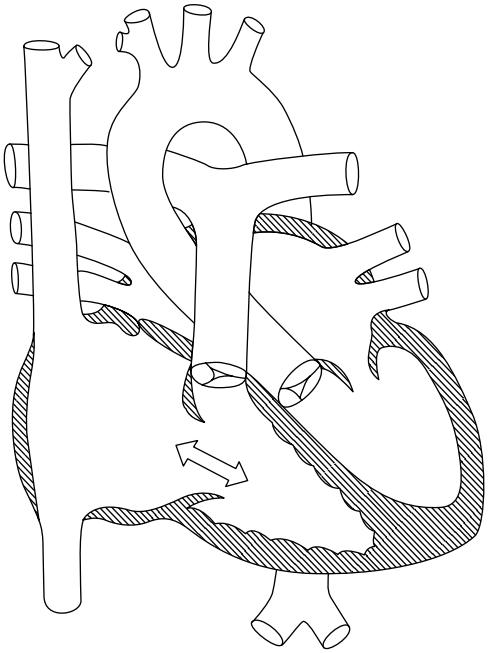

Figure 6.55 Tricuspid regurgitation. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Truncus arteriosus

Basics:
Truncus arteriosus (TA) can be described as a single great artery arising from the ventricular outflow tracts in the presence of a large VSD. From this common trunk emerges the aorta and pulmonary arteries. TA can be classified by how the pulmonary arteries arise from the aorta:

- TA type I has a main pulmonary artery coming off of the aorta.
- TA type II has right and left pulmonary arteries arising separately from the posterior aorta but close together.
- TA type III has right and left pulmonary arteries arising separately from the aorta from different sides and perhaps different levels.

Surgical correction is aimed at closing the VSD, ensuring a competent truncal valve, providing pulmonary blood flow with a right ventricle to pulmonary artery conduit incorporating the PAs removed from the truncus and patching the aorta where the PAs formerly arose.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia required.
- Truncal valve regurgitation may require direct coronary delivery of cardioplegia. The perfusionist should prepare 1.5–2 doses of cardioplegia.
- Truncal valve regurgitation may require cardioplegia delivery early in the case. The perfusionist should prepare the cardioplegia as soon as bypass parameters are deemed acceptable.
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–32 °C.
- Consideration should be given to going on bypass slowly and cooling slowly to prevent LV distention secondary to truncal valve regurgitation and\or ventricular fibrillation due to a cool prime rapidly entering the heart.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management is preferred.
- The pulmonary arteries must be controlled before or soon after commencing bypass since runoff to the pulmonary circulation can compromise systemic perfusion (much like a PDA or MAPCAs which need to be controlled).
- It is important to know the surgeon's preference for maximum aortic pressure whenever the aorta has been opened surgically, especially when allowing for ejection.

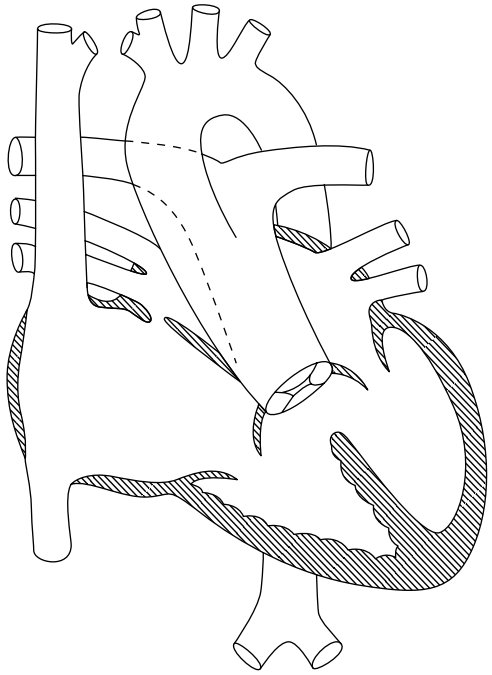

Figure 6.56 Truncus arteriosus Type I. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

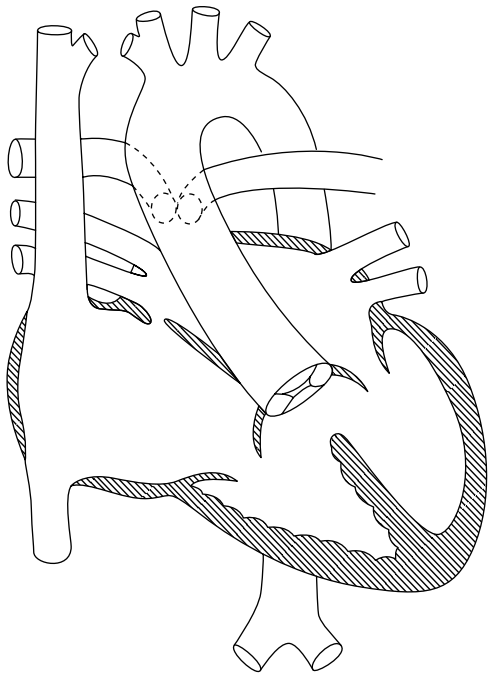

Figure 6.57 Truncus arteriosus Type II. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

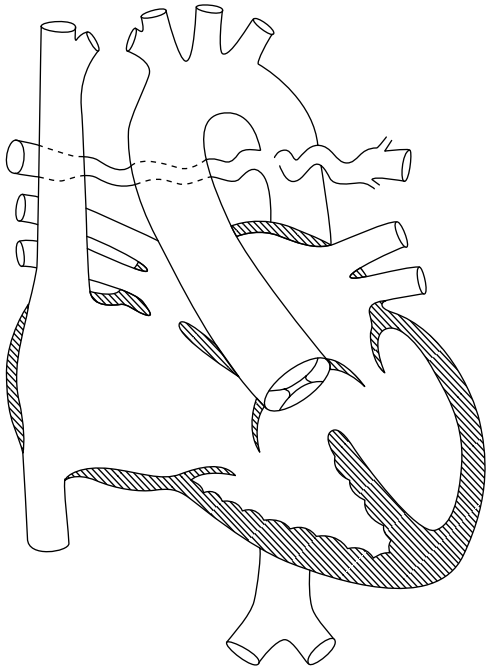

Figure 6.58 Truncus arteriosus Type III. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

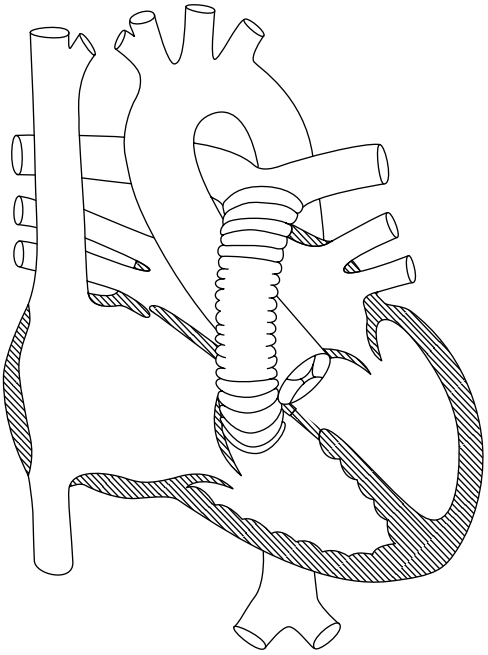

Figure 6.59 Truncus arteriosus Type 1 Repair. RV to PA conduit, ascending aorta repair and VSD patch. From Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

Ventricular septal defect

Basics:

A VSD is a communication between the right and left ventricles. While conceptually these are simpler defects, they can be varied in presentation. They may present as a singular small hole centrally located in the septum requiring only primary closure or present as multiple holes in different parts of the septum, which may be difficult for the surgeon to visualize and address. VSDs are commonly classified by location in the ventricular septum:

- Membranous VSD—located in the membranous septum.
- Outlet, subarterial, subpulmonary, or conoventricular VSD—located below the aortic and pulmonary valves, high in the infundibular septum.
- Inlet VSD (canal type)—located near the "inlet" to both ventricles, between the mitral and tricuspid valves, generally more posterior than membranous VSDs.
- Muscular VSD—located low in the septum approaching the apex of the heart.
- The communication between ventricles which exists in patients with a hypoplastic ventricle (as with DILV or tricuspid atresia) is referred to as the BVF. The BVF is the only inlet connection to the rudimentary ventricle. This interventricular communication is often important for the systemic ventricular outflow tract. It is treated quite differently than other septal communications. This communication is usually not closed but rather it is left as is or enlarged to prevent outflow tract obstruction.
- Surgical correction for true VSDs is aimed at closing the hole(s) primarily (suture only) or with a patch. The patch material is most commonly synthetic (Dacron or Gore-Tex, etc.), but autologous pericardium may also be used.

Bypass notes:

- Maximum anticipated flow for equipment selection: 3.0 L/min/m^2.
- Cardioplegia required (induced ventricular fibrillation may be used alone or in addition).
- Bicaval cannulation is required.
- Left ventricular vent is common.
- Target temperature is 28–34 °C.
- Minimum dilutional hematocrit is 25–30%.
- pH-stat blood gas management for cases is performed at 30 °C or below. Alpha-stat blood gas management is performed for adults and patients with suspected vascular disease.

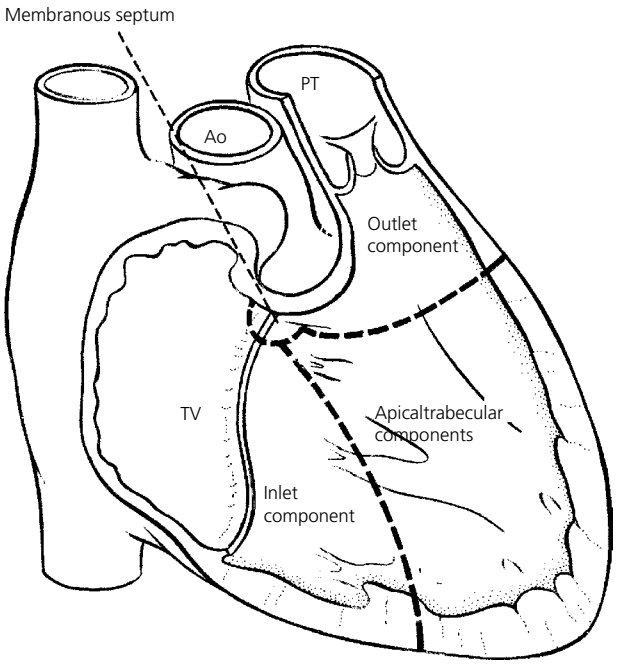

Figure 6.60 Location of ventricular septal defects. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

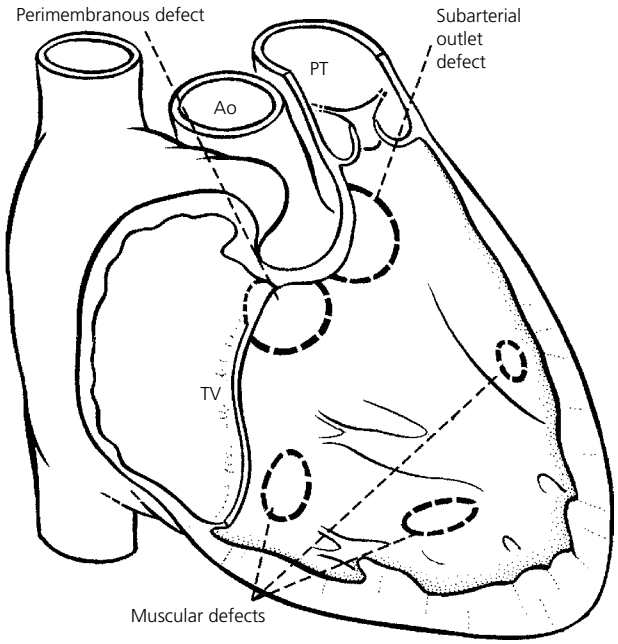

Figure 6.61 Location of ventricular septal defects. From Stark et al. [2]. Reproduced with permission of John Wiley & Sons.

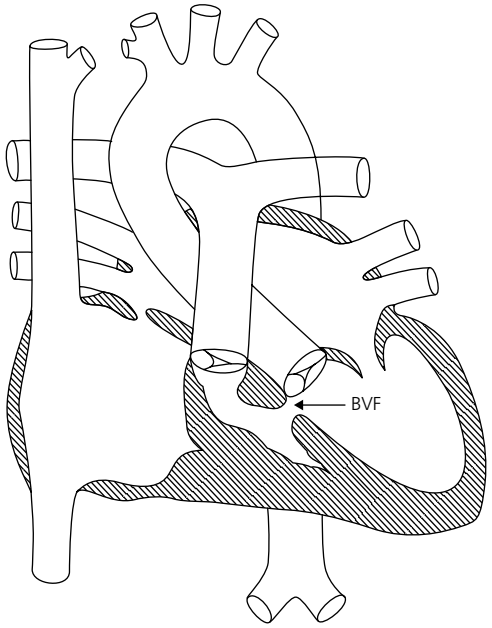

Figure 6.62 Bulboventricular foramen in tricuspid atresia. Adapted from Mullins and Mayer [1]. Reproduced with permission of John Wiley & Sons.

Case notes:

Surgeon preferences:

References

1 Mullins CE, Mayer DC eds. Congenital Heart Disease: A Diagrammatic Atlas. Hoboken: John Wiley & Sons, Inc.; 1992.
2 Stark JF, de Leval MR, Tsang VT eds. Surgery for Congenital Heart Defects, 3rd edition. Chichester: John Wiley & Sons, Ltd; 2006.

CHAPTER 7

Notes on select issues during bypass

Blood pressure higher than expected

- Is the value accurate? Request that the patient arterial line be flushed and zeroed.
- Has the transducer been moved lower (bed rolled to one side) causing an artifactually high blood pressure reading?
- Is the bypass circuit pressure also high?
  - If the bypass circuit and patient pressures are both high, the pressure is more likely to be real.
  - If the bypass circuit pressure is normal, the problem is more likely to be with the patient arterial line system or with cannula position (the flow may be preferential to the right arm with a right radial line, for example).
- Is the pump flow appropriate?
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the heart–lung machine (HLM) computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
    - The use of a flow probe can quickly diagnose whether calculated flow and delivered flow are disparate.
- Is the patient blood pressure high because the patient is still ejecting?
  - Increase left ventricular vent flow if one is in use.
  - Significant collateral/shunt flow to the left heart in the presence of restrictive or absent septal defects may result in continued ejection. Significant aortic regurgitation can lead to a similar result.
- Is the systemic vascular resistance high?
  - Are any medications running into the patient which may increase SVR? Patients on cardiac meds normally have those medications turned off once on bypass.
  - Consult with the anesthesiologist and consider isoflurane, phentolamine, nitroglycerin, nitroprusside or milrinone, etc.

Other considerations:

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

Blood pressure lower than expected

- Is the value accurate? Request that the patient arterial line be flushed and zeroed.
- Has the transducer been moved higher (bed rolled to one side) causing an artifactually low blood pressure reading?
- Is the bypass circuit pressure also low?
  - If the bypass circuit and patient pressures are both low, the problem is more likely to be flow related or patient related (SVR).
  - If the bypass circuit pressure is unchanged or as expected with a displayed low patient pressure, the problem is more likely to be with the patient arterial line system itself.
- Is the pump flow adequate?
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - External shunts
    - Are there any inadvertently open circuit shunts (oxygenator prime/purge line)?
    - Is the sampling manifold open (if not needed for in-line blood gas monitoring)?
    - If hemoconcentrator flow is passively supplied, turn it off to ensure the unit is not allowing excess steal from the arterial circuit.
    - If active hemoconcentrator flow from an arterial source is being used, ensure it is flowing at the intended rate (correct tubing size entered into head controller).
  - Internal shunts
    - Does the patient have an uncontrolled PDA, surgically placed systemic-to-pulmonary shunt (BTS/Sano), aortopulmonary collaterals or an AP window?
    - Is there excessive "steal" to the LV vent?
    - Internal shunts will require higher than predicted pump flows until they are surgically controlled. Some aortopulmonary collaterals may not be accessible, and therefore increased pump flows will be required throughout the bypass run. The use of NIRS may be particularly helpful in determining an adequate pump flow to maintain cerebral blood flow when internal shunts are not controlled. The pump flow should ideally result in adequate NIRS values while not flooding the surgical field. This is not always possible, and so aggressive field suction (and venting) may be needed to compensate.
  - Physical obstruction to flow
    - Check for kinked tubing in the arterial circuit, especially where the tubing enters and exits the roller head. A reservoir lamp or flashlight can be helpful with peering into the tubing guides to check this.
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
    - The use of a flow probe can quickly diagnose whether calculated flow and delivered flow are disparate.
    - Was the occlusion properly set (roller pumps)? There is the possibility that once the arterial head tubing changes temperature during the case (warming with constant flow, cooling with low flow hypothermic bypass), the occlusion may change. During a planned circulatory arrest or in consultation with the surgeon, one can come off bypass, clamp the arterial limb, and pressure check the system. If the system pressure falls dramatically, the occlusion can be adjusted appropriately. This can be accomplished in just several seconds off bypass.
- Was cardiotomy suction blood from the chest introduced to the circuit?
  - Suctioned mediastinal blood may contain activated agents that cause transient hypotension.
- Was stagnant blood in the hemoconcentrator introduced into the circuit?
  - Stagnant circuit blood may contain activated agents that cause transient hypotension.
- Is the systemic vascular resistance low? Consult with anesthesia.
  - Maximize pump flow in consideration of the operative needs and patient temperature.
  - Are any blood pressure medications running into the patient? Some surgeons prefer to have nitroglycerin empirically run after cross clamp removal and/or during rewarming. This drip may need adjustment.
  - Is isoflurane in use? Consult with the anesthesiologist and consider decreasing or terminating its use.
  - Consult with the anesthesiologist and consider an alpha agonist such as phenylephrine.

- Is the patient flushed (red faced)? Epinephrine and diphenhydramine may be considered for a suspected allergic reaction, especially when phenylephrine doesn't work.

- Is the hematocrit adequate?
  - Some patients may normally exist with a high hematocrit. A relatively low hematocrit may result in a low systemic blood pressure. Consider increasing the hematocrit.

- Is there a possible aortic dissection?

Other considerations:

Bypass circuit pressure higher than expected

- Is the value accurate? Zero the circuit pressure transducer.
- Is the patient arterial blood pressure also high?
  - If the bypass circuit and patient pressures are both high, the problem is more likely to be real.
- Is the patient arterial blood pressure normal or low with a high circuit pressure?
  - Is there a kink in the circuit arterial limb?
  - Is the arterial cannula kinked?
  - Is the arterial cannula appropriately sized?
  - Is the arterial cannula properly positioned (if no bumper is used on the cannula, it is more likely to migrate from its original location)?
  - Is there a possible aortic dissection?
- Is the pump flow appropriate and accurate?
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
- Is the SVR high?
  - Consult with anesthesia and consider lowering SVR with isoflurane or another medication.
- What is the status of the arterial line filter?
  - Is there evidence of a filter defect or clot?
  - Oxygenators with integrated arterial line filters generally have system pressure monitored post-filter. If this is the case, filter clots may only be evident, pressure-wise, with a significant change in pre-membrane pressure (which isn't always monitored).

Other considerations:

Bypass circuit pressure lower than expected

- Is the value accurate? Zero the circuit pressure transducer.
- Is the patient blood pressure also low?
  - If the bypass circuit and patient pressures are both low, the problem is more likely to be flow related or patient related (SVR).
  - If the bypass circuit pressure is low with a normal or high patient pressure, the problem is more likely to be with the bypass circuit transducer system. Check the zero and considering changing the transducer. You can also briefly attach the bypass circuit pressure dome to a different transducer (i.e., the cardioplegia transducer) to verify the value.
- Pump flow
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - External shunts
    - Are there any inadvertently open circuit shunts (oxygenator prime/purge line)?
    - Is the sampling manifold open (if not needed for in-line blood gas monitoring)?
    - If hemoconcentrator flow is passively supplied, turn it off to ensure the unit is not allowing excess steal from the arterial circuit.
    - If active hemoconcentrator flow from an arterial source is being used, ensure it is flowing at the intended rate (correct tubing size entered into head controller).
  - Internal shunts
    - Does the patient have an uncontrolled PDA, surgically placed systemic-to-pulmonary shunt (BTS/Sano), aortopulmonary collaterals, or an AP window?
    - Is there excessive "steal" to the LV vent?
    - Internal shunts will require higher than predicted pump flows until they are surgically controlled. Some aortopulmonary collaterals may not be accessible and so increased pump flows will be required throughout the bypass run. The use of NIRS may be particularly helpful in determining an adequate pump flow to maintain cerebral blood flow when internal shunts are not controlled. The pump flow should ideally result in adequate NIRS values while not flooding the surgical field. This is not always possible and so aggressive field suction (and venting) may be needed to compensate.
  - Physical obstruction to flow
    - If the arterial pump head display is your flow indicator, check for kinked tubing in the arterial circuit proximal to the pressure monitoring site, especially where the tubing enters and exits the roller head. A reservoir lamp or flashlight can be helpful with peering into the tubing guides to check this.
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
    - The use of a flow probe can quickly diagnose whether calculated flow and delivered flow are disparate.
    - Was the occlusion properly set (roller pumps)? There is the possibility that once the arterial head tubing changes temperature during the case (warming with constant flow, cooling with low flow hypothermic bypass), the occlusion may change. During a planned circulatory arrest or in consultation with the surgeon, one can come off bypass, clamp the arterial limb, and pressure check the system. If the system pressure falls dramatically, the occlusion can be adjusted appropriately. This can be accomplished in just several seconds off bypass.
- Is the systemic vascular resistance low? Consult with anesthesia.
  - Maximize pump flow in consideration of the operative needs and patient temperature.
  - Are any blood pressure medications running into the patient? Some surgeons prefer to have nitroglycerin empirically run after cross clamp removal and/or during rewarming. This drip may need adjustment.
  - Is isoflurane in use? Consult with the anesthesiologist and consider decreasing or terminating its use.
  - Consult with the anesthesiologist and consider an alpha agonist such as phenylephrine.
  - Is the patient flushed (red faced)? Epinephrine and diphenhydramine may be considered for a suspected allergic reaction, especially when phenylephrine doesn't work.
- Is the hematocrit adequate?
  - Some patients may normally exist with a high hematocrit. A relatively low hematocrit may result in a low systemic blood pressure. Consider increasing the hematocrit.

Other considerations:

Central venous pressure elevated

- Is the value accurate? Request that the central line be flushed and zeroed.
  - Is the central line inaccurate because it is in the caval snare?
- Is the bed rotated with the CVP transducer positioned lower?
  - Rotation of the bed which affects relative transducer height can artifactually affect the CVP reading.
- Was the bed lowered decreasing the kinetic potential between the heart and venous reservoir? Consider raising the bed or lowering the venous reservoir if possible.
- Is the CVP acutely elevated?
  - Check venous line for physical obstructions on and off the surgical field.
  - Check for an air lock.
  - Ask surgeon about venous cannula(e) position.
    - Venous cannula side ports against chamber or vessel wall?
    - SVC cannula inserted too far? Innominate blocked?
- Has the CVP progressively become elevated?
  - Ask surgeon about cannula position (per above considerations).
  - Consideration should be given to the possibility of a luminal obstruction caused by something in the cava/atrium or clot formation within the cannula [1, 2].
- Have the NIRS values also changed?
  - Stable NIRS may favor artifact in the CVP reading.
  - Decreasing NIRS may indicate the elevated CVP is real.
- Is the pump flow appropriate?
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
    - The use of a flow probe can quickly diagnose whether calculated flow and delivered flow are disparate.
- Consider decreasing pump flow in a temperature appropriate manner. Target temperature may need adjustment as well.

Other considerations:

Heat exchange issue (slow cooling or warming)

- Are the water lines properly connected at the heater-cooler ***and*** the oxygenator?
  - Verify heater-cooler water supply is directed to oxygenator water inlet.
  - Are the water lines appropriately sized so as not to impede flow?
  - Are the water lines of appropriate length? Longer lines will decrease performance in a system.
- Is the patient obese?
  - Fat does not warm or cool as efficiently as muscle which has more blood flow. Obese patients will take longer to cool and warm.
- Is there a coarctation of the aorta?
  - This will hamper lower body cooling (with central cannulation), but rewarming should be normal after repair.
- Is the patient significantly vasoconstricted?
  - Consult with anesthesia and consider lowering SVR with isoflurane or another medication.
  - Phentolamine, nitroprusside, nitroglycerin, milrinone, and other agents may be used to decrease SVR in order to aid cooling and warming. Some clinicians allow for a lower blood pressure if it aids in cooling/warming and the NIRS are acceptable.
- What is the pump flow rate?
  - Increased pump flow rates are a first-line treatment to increase thermal exchange.
  - Heat exchange efficiency drops off as flow rate increases. If an oxygenator is being used near its maximum rated flow, heat exchange efficiency will decrease.
- What is the water flow rate through the heater-cooler system?
  - Heat exchange performance is in part driven by the counter flow principle (water and blood flows are opposing to maximize thermal transfer) and by the absolute water flow through the system. If the water flow rate is below normal, check for kinks in the water tubing.
    - Consider a backup unit if the water flow rate is below normal.

Other considerations:

NIRS values lower than expected

The perfusionist should monitor the NIRS values during the prebypass period. These values should be compared to bypass values throughout the run and are most often an excellent indicator of cerebral perfusion. If the NIRS values are lower than expected on bypass, or even worse, lower than prebypass values, consideration should be given to the following:

- Is the value accurate? Verify NIRS probe placement with the anesthesiologist and concern about the current value.
- Is the pump flow adequate?
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - External shunts
    - Are there any inadvertently open circuit shunts (oxygenator prime/purge line)?
    - Is the sampling manifold open (if not needed for in-line blood gas monitoring)?
    - If hemoconcentrator flow is passively supplied, turn it off to ensure the unit is not allowing excess steal from the arterial circuit.
    - If active hemoconcentrator flow from an arterial source is being used, ensure it is flowing at the intended rate (correct tubing size entered into head controller).
  - Internal shunts
    - Does the patient have an uncontrolled PDA, surgically placed systemic-to-pulmonary shunt (BTS/Sano), aortopulmonary collaterals or an AP window?
    - Is there excessive "steal" to the LV vent?
    - Internal shunts will require higher than predicted pump flows until they are surgically controlled.
  - Physical obstruction to flow
    - If the arterial pump head display is your flow indicator, check for kinked tubing in the arterial circuit, especially where the tubing enters and exits the roller head. A reservoir lamp or flashlight can be helpful with peering into the tubing guides to check this.
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
- Blood pressure
  - Maintain at prescribed level with consideration for trying a higher pressure.
  - Is the blood pressure accurate (zeroed and not influenced by cannulation or surgical clamps)?
- Gas supply
  - Is the proper gas source selected?
    - Increase FiO_2 if possible.
  - Gas flow rate (this is less likely to affect oxygenation but should be verified nonetheless)
    - Is the set gas flow reaching the patient? Verify the gas system is intact from the source to the oxygenator.
    - Consider changing the flow meter. Some systems may accumulate debris in the gas tubes leading to inaccurate readings.
    - Consider trialing a standalone source of 100% oxygen to readily verify source and flow on a different system.
    - Is the pump anesthetic gas vaporizer sealed with tight inlet and outlet connections?
    - Is the gas flow rate within the manufacturer recommended range?
  - Is the PaO_2 as expected? Maximize PaO_2 while identifying the cause of the low NIRS values.
  - Is the $PaCO_2$ as expected? Consider pH-stat if not in use or at least try increasing the $PaCO_2$ temporarily. Some congenital patients may have bypass-significant aortopulmonary collaterals that are not always documented by imaging studies. Increasing cerebral blood flow with pH-stat management may improve the NIRS.
  - Chronic CO_2 retainers may require a $PaCO_2$ value closer to their norm. Consult with anesthesiologist.
- Has the CVP progressively become elevated?
  - Ask surgeon about venous cannula(e) position.
    - Venous cannula side ports against chamber or vessel wall?
    - SVC cannula inserted too far? Innominate blocked?
  - How does the current CVP compare to the prebypass and post cannulation values?
  - Empirically, increasing pump flow to improve NIRS values may be counterproductive. If drainage of the head is the issue, increasing pump flow may increase the CVP and possibly decrease the cerebral perfusion pressure, resulting in worsening NIRS values.

- Consideration should be given to the possibility of a luminal obstruction caused by something in the atrium or clot formation within the cannula [1, 2].

- Hematocrit
  - Consider raising the hematocrit to the patient's normal or even above to see if there is a response with the NIRS.
- Temperature
  - Is the pump flow rate appropriate for the current temperature?
- In-line blood gas monitoring
  - Do the in-line values reasonably match the lab values?
  - Consider recalibrating the in-line blood gas monitor?
- Anesthesia considerations
  - Have anesthesiologist check the face for signs of inadequate drainage of the head.
  - Is the patient shivering due to inadequate neuromuscular blockade? Is the level of anesthesia adequate? Did the medications get to the patient? Consider an additional dose to rule this out.
  - If isoflurane is integral with the circuit, is there a chance of malignant hyperthermia? Is the lactate level increased with rising patient temperature and a low venous saturation (at least lower than expected given the pump parameters)? Even if the vaporizer is off, consider removing the vaporizer from the system and changing the downstream tubing.
  - Did the anesthesiologist expect anesthetic gas on bypass for primary anesthesia?
  - Is there anesthetic agent in the vaporizer?

Other considerations:

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$PaCO_2$ higher than expected

The perfusionist continually monitors $PaCO_2$ throughout the bypass run. There is a common range for the V/Q ratio given the oxygenator, gas supply (carbogen, traditionally blended gas, the addition of 100% CO_2, etc.) target temperature, and patient size. When the $PaCO_2$ value is higher than expected or within normal range with increased ventilatory requirements, the perfusionist must investigate the possible causes. Consideration should be given to the following:

- Is the value accurate?
  - Verify in-line value with lab value.
  - Does the in-line blood gas monitor temperature reading approximate the pump arterial line temperature indicating adequate cuvette blood flow?
- Is the oxygenator appropriately sized?
  - An undersized oxygenator may not be able to adequately clear carbon dioxide.
  - Is the pump flow at the upper limit of the oxygenator in use?
  - Was the oxygenator primed several hours in advance of the surgery? Or, has the unit been in use for a time approaching the manufacturer recommended limit?
    - Consider increasing the sweep flow significantly for a brief period of time if there is suspicion of the oxygenator wetting out. Maximum gas flow values and time limits for such are usually specified by the manufacturer. For example, the manufacturer may recommend increasing the gas flow to 10 LPM for 10 s when a wetted out oxygenator bundle is suspected. The manufacturer recommended actions should not be exceeded since air embolism to the blood phase may result (Tables 1.1 to 1.4).
- Pump flow
  - What is the cardiac index?
    - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - External shunts
    - Are there any inadvertently open circuit shunts (oxygenator prime/purge line)?
    - Is the sampling manifold open (if not needed for in-line blood gas monitoring)?
    - If hemoconcentrator flow is passively supplied, turn it off to ensure the unit is not allowing excess steal from the arterial circuit.
    - If active hemoconcentrator flow from an arterial source is being used, ensure it is flowing at the intended rate (correct tubing size entered into head controller).
  - Physical obstruction to flow
    - If the arterial pump head display is your flow indicator, check for kinked tubing in the arterial circuit, especially where the tubing enters and exits the roller head. A reservoir lamp or flashlight can be helpful with peering into the tubing guides to check this.
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
- Gas supply
  - Is the proper gas source selected?
  - Is the proper gas cylinder hooked up to the selected source? For example, if carbogen is in use and a 10% CO_2/90% O_2 cylinder is connected to the 3% CO_2/97% O_2 source line, a higher than expected $PaCO_2$ may result.
  - Gas flow rate
    - Is the set gas flow reaching the patient? Verify the gas system is intact from the source to the oxygenator.
    - Consider changing the flow meter. Some systems may accumulate debris in the gas tubes leading to inaccurate readings.
    - Consider trialing a standalone source of 100% oxygen to readily verify source and flow on a different system (especially an alternate flow meter).
    - Is the pump vaporizer sealed with tight inlet and outlet connections?
    - Is the gas flow rate within the manufacturer recommended range?
    - Maximize V/Q ratio as recommended by the oxygenator manufacturer.
  - Is the PaO_2 lower than expected as well?
    - This may indicate overall delivered gas flow is lower than the set values. Switch to a standalone 100% oxygen tank with its own flow meter while verifying the problem and evaluating the overall gas delivery system. Verify that there are no leaks in the gas delivery system.
    - Ensure the V/Q ratio is within manufacturer specifications.
    - Consideration should be given to maximizing the V/Q ratio per manufacturer recommendation.

- Additions to the circuit
  - Was sodium bicarbonate recently administered? A percentage of this will immediately form carbon dioxide. A transient increase in $PaCO_2$ lasting less than 2 min is expected.
  - Is bicarbonate buffered saline being used at the field and being sucked back to the bypass circuit causing transient increases in the $PaCO_2$?
  - Was blood recently administered? Bank blood will frequently cause a transient increase in $PaCO_2$.
  - Is there a chance of a slow water-to-blood leak causing metabolic acidosis and a concomitant need for bicarbonate administration?
- Field Flooding with Carbon Dioxide
  - Significant changes in $PaCO_2$ can be expected if CO_2 field flooding is in use. A field sucker may be intermittently in the well of CO_2 gas. Consideration should be given at the field to only using the field suckers as needed and not leaving them in the pericardial well if CO_2 control becomes difficult on bypass.
- Anesthesia considerations
  - Have anesthesiologist check the face for signs of inadequate drainage of the head.
  - Is the patient shivering due to inadequate neuromuscular blockade? Is the level of anesthesia adequate? Did the medications get to the patient? Consider an additional dose to rule this out.
  - If isoflurane is integral with the circuit, is there a chance of malignant hyperthermia? Is the lactate level increased with rising patient temperature and a low venous saturation (at least lower than expected given the pump parameters)? Even if the vaporizer is off, consider removing the vaporizer from the system and changing the downstream tubing.
  - Did the anesthesiologist expect anesthetic gas on bypass for primary anesthesia?
  - Is there anesthetic agent in the vaporizer?
- Target temperature
  - Is the target temperature appropriate for the pump flows that are achievable during the bypass run? Consider decreasing the target temperature.
- Consideration should be given to priming a backup oxygenator if values become increasing unacceptable, especially in conjunction with a low PaO_2.

Other considerations:

$PaCO_2$ lower than expected

The perfusionist continually monitors $PaCO_2$ throughout the bypass run. There is a common range for the V/Q ratio given the oxygenator, gas supply (carbogen, traditionally blended gas, the addition of 100% CO_2, etc.) target temperature, and patient size. When the $PaCO_2$ value is lower than expected, the perfusionist must investigate the possible causes. Consideration should be given to the following:

- Is the value accurate?
  - Verify in-line value with lab value.
  - Does the in-line blood gas monitor temperature reading approximate the pump arterial line temperature indicating adequate cuvette blood flow?
- Is the oxygenator appropriately sized?
  - Using an oxygenator made for larger patients may require greatly reduced sweep gas flow or increased added CO_2.
  - Is the gas flow rate within the manufacturer recommended range?
- Does the patient have an uncontrolled PDA, surgically placed systemic-to-pulmonary shunt (BTS/Sano), aortopulmonary collaterals or an AP window? Is the LV vent flow high?
  - Increased LV vent and sucker flow rates will increase CO_2 loss through the system due to mixing of air in the tubing and cardiotomy reservoir. Consideration should be given to the following:
    - Decreasing vent and sucker flow rates if it is acceptable at the surgical field.
    - Decreasing the sweep flow rate within manufacturer specifications.
    - Increasing the CO_2 flow rate.
    - Increasing the carbogen CO_2 concentration.
    - With carbogen systems, an *increased* sweep flow rate may be needed to add more carbon dioxide to the system in light of the efficiency seen under the current conditions (normally, increased sweep flow decreases $PaCO_2$).
- What is the status of the venous line?
  - Venous air will increase CO_2 loss through the system due to air mixing in the venous line and reservoir.
  - Notifying the surgeon of the venous air to see if it can be eliminated.
- Gas Supply System?
  - Decreasing the sweep flow rate within manufacturer specifications.
  - Increasing the CO_2 flow rate.
  - Increasing the carbogen CO_2 concentration.
  - With carbogen systems, an *increased* sweep flow rate may be needed to add more carbon dioxide to the system in light of the efficiency seen under the current conditions (normally, increased sweep flow decreases $PaCO_2$).
- What is the target temperature?
  - Increasing levels of hypothermia result in increased dissolved CO_2 levels which may decrease $PaCO_2$ values. Consideration should be given to the following:
    - Decreasing the sweep flow rate with manufacturer specifications.
    - Increasing the CO_2 flow rate.
    - Increasing the carbogen CO_2 concentration.
    - With carbogen systems, an *increased* sweep flow rate may be needed to add more carbon dioxide to the system in light of the efficiency seen under the current conditions (normally, increased sweep flow decreases $PaCO_2$).
- Is the pump flow rate in use at the lower end for an oxygenator's rated flow?
  - When an oxygenator is being used at its low end of rated performance, it may be extremely efficient at CO_2 removal. Consideration should be given to increasing the CO_2 flow rate or increasing the carbogen concentration in use. With carbogen systems, an *increased* sweep flow rate may be needed to add more carbon dioxide to the system in light of the efficiency seen under the current conditions (normally, increased sweep flow decreases $PaCO_2$).
- Field flooding with carbon dioxide
  - Significant changes in $PaCO_2$ can be expected if CO_2 field flooding is in use. The $PaCO_2$ may suddenly decrease if less CO_2 is being suctioned back to the pump via the vent or field suckers.

Other considerations:

PaO_2 lower than expected

- Is the in-line value accurate?
  - Verify in-line value with lab value.
  - Does the in-line blood gas monitor temperature reading approximate the pump arterial line temperature indicating adequate cuvette blood flow?
- Oxygenator
  - Is the oxygenator appropriately sized?
    - An undersized oxygenator may not be able to provide adequate oxygen transfer.
  - Is the pump flow at the upper limit of the oxygenator in use?
  - Was the oxygenator primed several hours in advance of the surgery? Or, has the unit been in use for a time approaching the manufacturer recommended limit?
    - Consider increasing the sweep flow significantly for a brief period of time if there is suspicion of the oxygenator wetting out. Maximum gas flow values and time limits for such are usually specified by the manufacturer. For example, the manufacturer may recommend increasing the gas flow to 10 LPM for 10 s when a wetted out oxygenator bundle is suspected. The manufacturer recommended actions should not be exceeded since air embolism to the blood phase may result (see Tables 1.1 to 1.4).
- Is the gas flow rate within the manufacturer recommended range?
- What is the FiO_2?
  - Maximize the sweep gas oxygen concentration until the reason for the low PaO_2 is identified.
  - Consider switching to a standalone 100% oxygen tank with its own flow meter while verifying the problem and evaluating the overall delivery system.
- What is the cardiac index?
  - Are the values for patient weight, height, and BSA entered into the HLM computer correctly?
  - Is the proper tubing size selected on the arterial pump head (roller pumps)?
  - Physical obstruction to flow
    - If the arterial pump head display is your flow indicator, check for kinked tubing in the arterial circuit, especially where the tubing enters and exits the roller head. A reservoir lamp or flashlight can be helpful with peering into the tubing guides to check this.
  - Is there an available flow probe to ensure arterial pump head flow matches delivered flow?
- What is the venous saturation?
  - If the venous oxygen saturation is exceptionally low, the oxygenator may need several minutes to catch up, providing the cause of the low venous oxygen saturation is temporary (very low flow required for an unexpected surgical intervention).
- Is the $PaCO_2$ higher than expected as well?
  - This may indicate overall delivered gas flow is lower than the set values. Switch to a standalone 100% oxygen tank with its own flow meter while verifying the problem and evaluating the overall delivery system. Verify that there are no leaks in the delivery system.
  - Ensure the V/Q ratio is within manufacturer specifications.
  - Consideration should be given to maximizing the V/Q ratio per manufacturer recommendation.
- Anesthesia considerations
  - Have anesthesiologist check the face for signs of inadequate drainage of the head.
  - Is the patient shivering due to inadequate neuromuscular blockade? Is the level of anesthesia adequate? Did the medications get to the patient? Consider an additional dose to rule this out.
  - If isoflurane is integral with the circuit, is there a chance of malignant hyperthermia? Is the lactate level increased with rising patient temperature and a low venous saturation (at least lower than expected given the pump parameters)? Even if the vaporizer is off, consider removing the vaporizer from the system and changing the downstream tubing.
  - Did the anesthesiologist expect anesthetic gas on bypass for primary anesthesia?
  - Is there anesthetic agent in the vaporizer?
- Target temperature
  - Is the target temperature appropriate for the pump flows that are achievable during the bypass run? Consider decreasing the target temperature.
- Close monitoring of the PaO_2 should be employed. Consideration should be given to priming a backup oxygenator if values become increasing unacceptable.

Other considerations:

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Reservoir volume acutely low

- Is the CVP acutely elevated?
  - Check venous line for physical obstructions on and off the surgical field.
  - Is this electronic venous occluder properly set to full flow?
  - Check for an air lock in the venous line, on and off the surgical field.
  - Ask surgeon about venous cannula(e) position.
    - Venous cannula side ports against chamber or vessel wall?
    - SVC cannula inserted too far? Innominate blocked?
    - IVC cannula inserted too far? Portal vein cannulation? Eustachian valve blocking IVC cannula drainage?
- Has the CVP progressively become elevated?
  - Ask surgeon about venous cannula(e) position.
    - Venous cannula side ports against chamber or vessel wall?
    - SVC cannula inserted too far? Innominate blocked?
    - IVC cannula inserted too far? Portal vein cannulation? Eustachian valve blocking IVC cannula drainage?
  - How does the current CVP compare to the prebypass and post cannulation values?
  - Empirically increasing pump flow to improve NIRS values may be counterproductive. If drainage of the head in particular is the issue, increasing pump flow may increase the CVP and possibly decrease the cerebral perfusion pressure, resulting in worsening NIRS values.
  - Consideration should be given to the possibility of a luminal obstruction caused by something in the atrium or clot formation within the cannula [1–2].
- If the CVP is unchanged?
  - Is the vent line relatively more full with blood?
  - Are the sucker lines relatively more full of blood?
  - Is volume being lost to wall suction?
  - Is volume being lost to the chest/pericardial space?
  - Is volume being lost to the floor?
    - Leaking from IV line, open chest, peripheral cannulation site, or bypass circuit?
- Has the hematocrit changed?
  - A constant hematocrit may indicate overt circulating volume loss.
  - An increasing hematocrit may indicate third spacing within the patient or significant urine output.
  - A less acute cause of apparent volume loss would be an increasing hematocrit with a hemoconcentrator inadvertently left on.
- Did the cross clamp just come off?
  - Circulating volume may have shifted to the LV vent or heart and lungs.
- Was the patient transitioned to partial bypass?
  - Circulating volume may have shifted to the heart and lungs.
- What is the status of the cardiotomy reservoir?
  - Fouling of the cardiotomy filter system can lead to increased dynamic hold up in the system.
    - The venous reservoir level should be maintained higher if cardiotomy fouling is present. This helps compensate for a possible decreased filtering ability and allows more time in the reservoir for microair to rise.
    - Consideration should be given to adding a stand-alone cardiotomy to the system to handle sucker and vent flow. Rarely, the integrated cardiotomy and venous reservoir would need to be changed. A secondary cardiotomy normally is sufficient to complete the bypass run.
- Is volume somehow being diverted to the cardioplegia system?
- Consideration should be given to urine output, third spacing, unidentified bleeding into the chest and increased wall suction use with volume loss over time (not acute).
- Is the SVR significantly low causing volume to remain in the patient?
- Is the reservoir positioned as low as possible to maximize kinetic potential?
- If augmented venous return is in use, is the intended negative pressure being applied to the venous system?

Other considerations:

References

1 Matte GS, Howe RJ, Pigula F. A single-center experience with luminal venous cannulae obstruction caused by clot formation during bypass. J Extra Corpor Technol. 2013;45:55–7.
2 Ozdirik JE, Grant PW, Cheruvu C, et al. An unusual case of cannula obstruction resulting from venous thromboembolism. J Extra Corpor. 2014;46:267–9.

CHAPTER 8

Notes on select emergency procedures during bypass

A perfusionist is faced with nearly countless possibilities for failure or other issues with the hardware and disposables used for cardiopulmonary bypass. Issues before or after bypass can normally be dealt with in a calm and controlled manner. Issues on bypass require expeditious diagnosis and action which can at times be stressful, especially if the patient cannot be safely removed from bypass "warm and ejecting" or under circulatory arrest at an appropriate hypothermic temperature. Four life-threatening issues that the perfusionist must be prepared to deal with on bypass are arterial pump head failure, oxygenator failure, massive air embolization, and acute aortic dissection at the initiation of bypass. Fortunately, these are extremely rare and the perfusionist's lack of first-hand experience in dealing with these issues can be made up for with simulation training and "wet" lab experiences. Other issues that may be seen on bypass include dealing with air locks in the venous line (a major issue if recurrent and adequate pump flow cannot be maintained) and inadvertent arterial or venous decannulation. *Chapter 7* reviewed some issues that are more common to be seen by the perfusionist. Here, high-risk/low-frequency bypass emergencies that put the patient at considerable risk are discussed.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

Arterial pump failure (roller head)

Failure of the arterial roller head pump is almost always sudden and can be caused by several factors including loss of electrical power (modular unit inadvertently turned off, modular power cable disconnect, loose internal wires, circuit board malfunction, etc.), mechanical failure (with motor, bearings, etc.) or even an improper pump head assignment (the arterial head may shut off once the cardioplegia is delivered if it is improperly assigned electronically as the cardioplegia pump). Regardless of the cause, the initial reaction by the perfusionist is to quickly assess the cause. For example, if there is no power and the power switch is off, simply turn the unit back on. This may sound simplistic, but it can certainly happen as some heart–lung machines (HLMs) do not have safety covers over these switches. After an initial assessment of just several seconds, if the cause and immediate resolution cannot be determined, the perfusionist must initiate hand cranking. Then, with the entire team being aware of the pump emergency, additional personnel can aid the perfusionist by either hand cranking, retrieving supplies, or helping diagnose and correct the problem.

Hand cranking is ideally performed at the same rate as most recent bypass flow. If a standalone flow indicator is in use and still working, it can give guidance to hand-cranking speed. It is never practical to count revolutions per minute (RPMs) to gauge flow. Rather, it is more effective for the perfusionist to monitor venous return and to crank such that the venous reservoir level (easily assessed in open reservoir systems) is maintained at the level it was previous to the failure.

Hand cranking stabilizes the situation and allows for controlled diagnosis and management. If a simple intervention fixes the problem (loose power cable), it is performed and hand cranking can stop for the several seconds it takes for the pump head to initialize and be ready for use. If it is determined that a new arterial head is required, there are several options. In modular systems, a sucker head may be sacrificed and moved close to the oxygenator to become the new arterial head. Alternately, a pump head may be taken from a backup HLM. Either way, the team must determine proper positioning to allow for transfer of the boot line (the segment that passes through the roller head), how to properly assign the new head as the arterial head for servoregulation purposes, and how to quickly assess occlusion in the replacement head. Arterial head failure poses a significant risk to the patient on cardiopulmonary bypass. The perfusionist must be familiar with the process for hand cranking and arterial head change-out. Below is *an example* of an algorithm for arterial head failure.

1. Verify power button is on for the controller and that power cords are secure.
2. If controller power switch is on with no power supplied, hand crank and have second perfusionist obtain a pump head off of another console.
3. Place replacement pump head next to failed unit.
4. Connect replacement pump head to HLM console power supply. Disconnect failed unit from HLM if need be (an alternate power supply *location* is preferred to rule out a receptacle issue).
5. Assign the replacement pump head as the arterial pump on the pump console/controller.
6. Discuss with surgeon and anesthesiologist procedure for switching heads.
7. Stop hand cranking and clamp arterial and venous lines to come off bypass. Either fill or drain patient per surgeon preference and patient status.
8. Ensure adequate reservoir volume before unclamping recirculation line. (The fluid level in post-oxygenator recirculation lines will equilibrate with the CVR level during the transition. A low CVR level may allow for air to enter the arterial limb if the connecting wye on the recirculation line is raised above the reservoir fluid level.)
9. Move boot to replacement head.
10. Verify occlusion with recirculation line and clamp recirculation line (occlusion may also be verified via a pressure method with the recirculation line clamped).
11. Commence bypass with replacement head controller.

The specific steps taken will vary by institution and the equipment in use. An additional option with some circuits is cutting in a centrifugal head to provide support since this can be done rather quickly without the need for occlusion setting.

Other considerations:

Failure to oxygenate

Oxygenator failure most often is a gradual process developing over minutes or even hours on bypass. However, it can also be sudden, especially if the bundle is cracked or compromised in some way. Since most oxygenator failure is gradual, the perfusionist generally has ample time to diagnose the problem and devise an action plan. I prefer to use the term "failure to oxygenate" to describe this emergency since there are several causes with true oxygenator failure generally being last on a long list. Failure to oxygenate can occur simply because the sweep gas flow was not turned on, there was a leak in the gas supply system, or due to a disconnected gas supply line. Such causes are easily remedied once diagnosed. Failure to oxygenate due to true oxygenator failure may not be as easy to diagnose. Declining oxygenator performance may present with the need for increasing sweep flow and FiO_2 on bypass. However, it is important to note that with the levels of hypothermia commonly used in congenital heart surgery, declining performance can be masked by the decreased metabolic needs at low temperatures. In these instances, failure to oxygenate may become quite evident only upon rewarming toward normothermia. Below is *an example* of an algorithm for failure to oxygenate (*see also section "PaO_2 lower than expected" in Chapter 7*).

1 Confirm in-line blood gas value with lab specimen if possible. Inspect the arterial and venous line color difference as a quick assessment.

2 Confirm proper gas source and blender setting.

3 Confirm integrity of gas system all the way to the oxygenator exhaust port.

4 Confirm function of flow meter. Change to a spare unit if available.

5 Confirm proper blood flow with:
- **(a)** Appropriate pump system pressure.
- **(b)** Appropriate patient arterial pressure.
- **(c)** Correct tubing size selected on pump controller.
- **(d)** Flow probe value congruent with calculated display flow.

6 Change to 100% oxygen if not already on that source
- **(a)** May change to standalone E cylinder with its own flow meter to rule out issues with the current gas supply system.

7 Consult with the anesthesiologist and confirm proper anesthesia/muscle relaxants are in use. Consider malignant hyperthermia if the CO_2 is significantly elevated with a low SvO_2, if an isoflurane source is in the gas supply system.

If failure is still evident,

8 Request second perfusionist to prime a replacement oxygenator and prepare it with clamped tubing and connectors for quick insertion into current system.

9 Inform surgeon of issue and discuss change out plan with team.
- **(a)** Decide on warm/ejecting and ventilated patient versus deep hypothermia with circulatory arrest.
- **(b)** Decide the trigger value for change out. It may be safer to limp off bypass with a lower than normal PaO_2 than to subject the patient to circulatory arrest.

10 Prepare for sterile procedure: scissors, betadine/alcohol, towels, flush solution to deair connections, and extra clamps.

11 Decide if oxygenator unit will be changed out alone versus change-out of entire oxygenator/reservoir combination.

12 Come off bypass and clamp arterial and venous lines (drain patient for circulatory arrest vs. fill patient for warm beating change-out).

13 Replace with new primed oxygenator using precut/clamped segments of tubing with connectors already attached.

14 Verify circuit is deaired.

15 Commence bypass in the standard fashion.

Some perfusion programs advocate for a simpler on-bypass oxygenator change-out system and include circuit modifications in all of their tubing packs for this purpose. One option is the parallel oxygenator insertion, or PRONTO procedure [1].

Other considerations:

Massive air embolization

Massive air embolus (MAE) with cardiopulmonary bypass can occur with transmission of gross air through the pump arterial system, the pump venous system or the vent line. Perfusionists who practiced in the early days of the profession may cite that their primary job was to constantly stare at the venous reservoir level to prevent arterial air embolism. Imperfect alarm systems or a lack of servoregulation could allow air to be pumped to the patient via the pump arterial line if venous return suddenly decreased and the reservoir emptied. This method of massive air embolism has largely been eliminated with reliable reservoir level and bubble-sensing servoregulating systems. Additionally, before widespread use of one way valves in the left ventricular vent system, a reversed vent line could inject air directly into the heart.

In the current era, massive air embolus is more likely to occur through the venous system. The reason for this is twofold. For one, vacuum-assisted venous drainage (VAVD) is more commonly used for bypass. Venous air embolization can result if the cardiotomy venous reservoir gets pressurized. The pressure in this case is vented through the path of least resistance which may be the pump venous line to the patient. With VAVD, the cardiotomy pressure is normally negative to provide vacuum to the venous system. This pressure is regulated and balanced against venous inflow and cardiotomy suction which is constantly adding air and blood to the reservoir. If this balance is sufficiently altered, as with a kinked vacuum source line or significantly increased cardiotomy sucker flow, pressure can form in the cardiotomy with subsequent transmission of venous blood and air retrograde through the pump venous line. Fortunately VAVD systems commonly employ pressure relief valves on the cardiotomy venous reservoir to prevent massive air embolization. Though, failure of these safety devices and unforeseen circumstances can still lead to massive air embolization.

A second reason why MAE is more likely to occur through the venous system in the current era is that the advent of VAVD has lead manufacturers to produce oxygenators with a singular venting port, which is needed for VAVD. This obligates clinicians to use this singular venting port style reservoir even when VAVD is not used. The singular venting port is vastly different from former production methods that incorporated more venting surface area on the reservoir top. This former design was developed to prevent MAE in the first place! The cardiac surgical team must be familiar with how to diagnose and treat MAE regardless of the causative factor [2]. Below is *an example* of an algorithm to treat MAE.

Operative measures

1. Stop the pump and clamp the arterial and venous lines. Turn off all pump heads until reservoir venting and proper head rotation/raceway tubing direction is confirmed.
2. Set oxygenator heat exchange and cooling blanket systems to 15 °C.
3. Place ice on head.
4. Have patient positioned into Trendelenburg.
5. Verify circuit integrity. Deair circuit and add volume as necessary to provide retrograde cerebral perfusion via sucker bypass. The surgeon will direct arterial inflow to the superior vena cava.
6. Ensure carbogen mix or 100% oxygen as the sweep gas source. Set a 2:1 V/Q ratio for the anticipated retrograde cerebral perfusion flow to maximize nitrogen off gassing.
7. At the direction of the surgeon, initiate retrograde cerebral perfusion flow at 0.4 L/min/m^2 and titrate to a central venous pressure of 40–50 mmHg (consideration should be given to the effect of Trendelenburg on the transducer height and reading). Retrograde cerebral perfusion is performed for 2 min or until air ceases to exit the aorta as determined by inspection and echocardiography. The anesthesia team should provide intermittent bilateral carotid compression to aid in removal of air from the vertebral arteries.
8. Reinstitute traditional bypass with pH stat blood gas management as instructed by the surgeon once cannulae are in their proper locations. Cool to 18 °C and maintain for 30–60 min to maximize reabsorption of residual air in the microvasculature.
9. Consider a higher than normal blood pressure to shrink air emboli and promote clearance.
10. Give 0.5 g/kg mannitol and 30 mg/kg methylprednisolone regardless of whether it was already administered at the onset of CPB.
11. Consult with surgeon to maintain a higher than normal pump flow and pressure to facilitate air removal from the microvasculature and at the oxygenator.
12. Rewarm to 34 °C when directed by surgeon.

Postoperative plan

1 Consider maintaining the patient paralyzed and sedated for an extended time period.
2 Consider 100% FiO_2 for 6–12 h to maximize nitrogen off gassing.
3 Consider maintaining the patient at 34–35 °C for 36 h.
4 Maintain the head of the bed up 30°.
5 Provide continuous near-infrared spectroscopy (NIRS) monitoring and consider serial transcranial Doppler studies, electroencephalograms, and eventually a head computed axial tomography (CAT) scan.

Each institution will have its own algorithm for treating this rare but potentially catastrophic bypass emergency. To note, if air is still present in the vasculature post bypass, there are two options for treatment. Some clinicians would favor hyperbaric oxygen therapy if it is technically feasible and readily available. Other clinicians would advocate for an additional deep hypothermic bypass run with retrograde cerebral perfusion. This option may be preferable since it is a readily available and effective modality the care team has experience with.

Other considerations:

Acute aortic dissection at the initiation of bypass

Cannulation of the ascending aorta can cause acute dissection. While this is uncommon in pediatric patients, the surgical team must have a defined protocol for treatment. The dissection must be identified quickly and handled expeditiously to prevent morbidity and mortality. The perfusionist may note an exceptionally high bypass circuit pressure during transfusion through the aortic cannula before bypass. Or, once on bypass, the bypass circuit pressure may spike while the patient's arterial blood pressure falls precipitously. The bypass circuit and patient pressure may normalize if the distal end of the dissection opens into the aorta. Venous return to the pump may be severely limited if the aorta ruptures. Anesthesiologist evaluation (pupil and facial changes, blood pressure-monitoring locations and changes, and TEE) and surgeon evaluation (visual inspection and palpation) will determine the presence of a dissection. Treatment of an acute dissection requires maintaining patient hemodynamics while the surgeon cannulates an alternate site. Then, the surgical repair can proceed. The following is a list of considerations for the perfusionist when acute aortic dissection at the initiation of bypass occurs.

1 The bypass circuit pressure will be unacceptably high. Alert surgical team.

2 Verify patient and bypass circuit pressures and patency of arterial limb.

3 If dissection is suspected, the surgeon will ask for bypass to be terminated. Ideally, the patient is left ejecting since the surgeon will need time to relocate the arterial cannula. Alternately, bypass may be continued with lower pump output and patient blood pressure until the arterial cannula can be relocated.

4 Provide a new arterial cannula to the surgical field. Femoral, neck or axillary arterial cannulation may be required.

5 Bypass will commence once arterial inflow is available.

6 The dissection will require repair on bypass. This frequently involves hypothermic circulatory arrest. Follow deep hypothermic circulatory arrest (DHCA) protocols (other medications, ice on head, adjust cooling blanket and room temperature, etc.).

7 Prepare cardioplegia system. This is especially important if the original surgical plan did not include cardioplegia.

8 Prepare for selective cerebral perfusion techniques at the surgeon's request.

Other considerations:

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Venous air lock

Small amounts of air entrained into the venous line may be difficult to visualize. The perfusionist may be alerted to the presence of continuous air entrainment with a decreasing $PaCO_2$ as evidenced with an in-line blood gas monitor. The surgeon should be alerted to this since venous air almost universally has been shown to result in microembolic air transmission to the patient, even with a properly functioning oxygenator and arterial line filter [3–8].

Large amounts of venous air are readily evident to the perfusionist visually. Large amounts of venous air will affect the in-line blood gas $PaCO_2$ value. The presence of large boluses of air down the venous line should be dealt with expeditiously by the team. Aside from the real risk of microembolic air transmission to the patient, large boluses of air can cause an air lock in the venous line ceasing drainage by eliminating the gravity siphon drainage effect. Here, venous blood flow to the reservoir is interrupted. This presents to the perfusionist as an acute loss of venous return. This is a pump emergency if it results in the inability to maintain an adequate pump flow for the current patient temperature.

The perfusionist must diagnose the acute loss of venous return and if it is due to an air lock caused by a large bolus of air, the remedy from the perfusionist's standpoint is quite simple. If air is trapped in the venous line, the perfusionist must "walk" the trapped air segment through the venous limb by raising a portion of the venous line and incrementally maintaining a different segment with this air as a high point while moving it toward the venous reservoir inlet. A single air bolus causing an air lock is easily dealt with, but continuous venous air causing regular air locks is more difficult to manage. If air is regularly occurring, the surgeon must diagnose the issue at the field and fix the problem. A right atrial back wall tear or one on the posterior surface of a cava may be difficult to find due to its location and since blood is not bleeding from the site (air is being entrained!).

A venous air lock can also present as a deprimed venous line. This is more likely to occur just before bypass if the venous line clamps are removed before connection to a venous cannula (though the perfusionist's and field clamps *should* be on at this point) or during planned circulatory arrest if the venous cannula is removed before the perfusionist fully empties the patient and appropriately clamps the venous line. In both instances, simply walking the air through the circuit is rarely effective. Most often, the venous line must be reprimed. Below is *an example* of the steps to treat air lock progressing to a deprimed venous line.

1 If VAVD is available, activate it to clear the venous air.

If VAVD not available, then:

2 Provide bypass with available return (sucker bypass).

3 Clamp the venous line between the reservoir and the repriming source (this can be a venous sampling line connected to the sampling manifold or a retrograde cerebral perfusion circuit arteriovenous connection).

4 Adjust clamps and stopcocks as needed to reprime the venous line. Ensure venous line and/or cannula(e) are open to atmosphere and retrograde fill via sampling manifold or other arteriovenous connection. MAE can result if the perfusionist transfuses retrograde through the venous line to a connected venous cannula!

OR

Have the field assistant fill the venous line with saline using a bulb syringe. This method may be more efficient with 3/8" and 1/2" venous lines than with smaller caliber lines.

5 Adjust clamps and stopcocks as needed to resume traditional bypass.

Other considerations:

Inadvertent arterial decannulation

Inadvertent arterial decannulation is always sudden and readily apparent at the surgical field. Here, the pump blood continues to flow through the arterial cannula onto the surgical field or worse, up and around the surgical field in a hose-like fashion! Arterial decannulation must be quickly communicated to the perfusionist so that the pump can be shut off with the venous and arterial pump lines immediately clamped. Clamping these lines and stopping any active venting prevents blood loss from the arterial cannula and aorta, and just as importantly, maintains blood in the patient by preventing exsanguination. An exsanguinated patient, those with calcified vessels or those with minimally invasive incisions, may be particularly difficult to recannulate. Recannulating the aorta (or another vessel) is vastly easier with a blood pressure. Below is *an example* of the steps to treat arterial decannulation.

1 Stop the arterial head.

2 Clamp venous and arterial pump lines.

3 Clamp the vent (aortic root or left ventricular).

4 Ensure the pump arterial line is free of air. If not, run volume through for deairing at the field (this line can get air in it if the pump is stopped, and the cannula tip is the low point allowing blood to drain out). Add volume to the circuit as necessary.

5 Commence bypass once recannulated and instructed by the surgeon.

Other considerations:

Inadvertent venous decannulation

Inadvertent venous cannulation presents to the perfusionist as a sudden and complete loss of venous return with an air-filled venous line. This can occur at any time. However, this is more likely to occur just before bypass if the venous line clamps are removed before connection to a venous cannula (though the perfusionist's and field clamps *should* be on at this point) or during planned circulatory arrest if the venous cannula is removed before the perfusionist fully empties the patient and appropriately clamps the venous line. In both instances, simply walking the air through the circuit is rarely effective. Most often, the venous line must be reprimed with retrograde flow from the HLM. The safest method for retrograde priming is for the venous cannula to be reprimed before recannulation. The perfusionist can transfuse volume up the venous line through the sampling manifold by adjusting clamps and stopcocks. Alternately, some circuits include a larger bore arterial-to-venous line connection (as with some retrograde cerebral perfusion circuits) which can be used. It is absolutely essential that with either method, the team verifies that the venous line can be safely filled retrograde. MAE can result if the perfusionist reprimes the venous line retrograde with an open venous cannula in the patient allowing the transfusion of air. Alternately, the field may fill the venous line directly with a bulb syringe if sufficient volume is readily available (with a clamp on the perfusionist's venous limb). The bulb syringe method may be more efficient for 3/8″ and 1/2″ venous lines, while smaller venous lines fill more efficiently with retrograde priming. Cardiopulmonary bypass can be provided via sucker bypass during repriming. If the patient is on planned circulatory arrest, arrest is usually prolonged for the very short duration of venous repriming. Below is *an example* of the steps to treat inadvertent venous decannulation.

1 Clamp the venous line between the reservoir and the repriming source (this can be a venous sampling line connected to the sampling manifold or a retrograde cerebral perfusion circuit arteriovenous connection).

2 Provide bypass with the available return (via suckers) and current reservoir volume.

3 Adjust clamps and stopcocks as needed to reprime the venous line. Ensure venous line and/or cannula(e) are open to atmosphere and retrograde fill via sampling manifold or other arteriovenous connection. MAE can result if the perfusionist transfuses retrograde through the venous line to a connected venous cannula! This reprimed venous line can be connected to the replaced venous cannula(e).

OR

Have the field assistant fill the venous line with saline using a bulb syringe. This method may be more efficient with 3/8″ and 1/2″ venous lines than with smaller caliber lines. This reprimed venous line can be connected to the replaced venous cannula(e).

4 Adjust clamps and stopcocks as needed to resume traditional bypass.

Other considerations:

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Massive air embolism

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CHAPTER 9

Brief overview of named procedures and terms

Alfieri stitch

- Mitral regurgitation may be caused by annular dilation. The Alfieri stitch involves suturing the free edges of opposing leaflets together to control annulus size and limit regurgitation.
- This may also be termed a "double-orifice technique" since it results in two flow paths through the annulus.
- The term may also be used for a similar repair of the tricuspid valve.

Batista procedure

- This operation is used to treat dilated cardiomyopathy but has been used for other diseases with dilated and/or hypertrophied ventricles.
- This is a form of ventricular reduction surgery that aims to improve cardiac mechanics by removing excess hypertrophied myocardium. It may also be referred to as ventricular modification or ventriculoplasty.

Bentall procedure

- This operation is used to treat severe aortic root dilation and aneurysms.
- It includes an aortic root replacement with a tube graft and mechanical or biological valve. The coronary arteries are most commonly reimplanted to the tube graft via coronary buttons (modified Bentall). The original procedure directly anastomosed the coronaries in an end-to-side fashion but was more prone to coronary issues.

Bidirectional Glenn shunt

- This shunt is commonly used for the second-stage procedure for patients with hypoplastic left heart syndrome or other single ventricle diseases. It may also be called a second-stage procedure or a cavopulmonary anastomosis.
- Today's bidirectional Glenn (BDG) shunt involves an end-to-side anastomosis of the superior vena cava to the right pulmonary artery (less commonly, it is performed on the left side for patients with persistent LSVC). Patients with bilateral superior venous cavae have the procedure on both sides in what is termed a bilateral bidirectional Genn shunt (BBDG).
- The original Glenn anastomosis (classic Glenn) connected the right superior vena cava directly to the right pulmonary artery with an end-to-end anastomosis (the proximal RPA would be oversewn). This provided blood flow only to the right lung and may be termed a unidirectional Glenn anastomosis. The classic Glenn is rarely performed now.

Blalock–Hanlon procedure

- This procedure was used in the 1940s and 1950s to treat d-transposition of the great arteries and other conditions with limited septal communication to increase pulmonary blood flow by increasing the atrial level shunt.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

- Surgical creation of an atrial septal communication off bypass.
- It is no longer performed.

Blalock–Taussig shunt (BTS)

- This shunt is commonly used as part of the Norwood operation/Stage 1 procedure for patients with hypoplastic left heart syndrome. It may also be used to provide pulmonary blood for conditions where the level of pulmonary vascular resistance requires an arterial pressure source (most commonly for a period of weeks to months before other surgery can be performed).
- The BTS can also be used as a palliative procedure for TOF (full 2 V repair is most common now).
- Classic BTS—right subclavian artery end-to-side anastomosis to right pulmonary artery. This version is no longer performed.
- Modified BTS (mBTS)—right subclavian artery to right pulmonary artery connection via tube graft.
- Most clinicians use the term "BTS" when referring to the mBTS.

Brock procedure

- Pulmonary valvotomy with special instruments off bypass. This was first used to treat pure pulmonary stenosis and tetralogy of Fallot patients in the 1940s and 1950s before the advent of open procedures performed on cardiopulmonary bypass. Such therapy is now carried out in the catheterization lab.

Central shunt

- It is the communication created with a short tube graft between the aorta and pulmonary artery.
- It is similar to an mBTS physiologically but different anatomically.

Cone procedure

- It is used for treatment of dysplastic tricuspid valves and is frequently associated with Ebstein's anomaly.
- The apically displaced tricuspid valve apparatus is detached, rotated some (creating a conical shape since the papillary muscles are still attached) and moved into a more basilar position. This results in a more functional and competent valve.
- A tricuspid valve suture annuloplasty or tricuspid ring/band placement may also be performed.

Cox maze procedure

- It was originally developed by Dr. James Cox and colleagues.
- It is used to treat atrial fibrillation and other cardiac arrhythmias. The traditional Cox maze involved precisely placing surgical incisions in the atrium to encourage non-conducting scar tissue growth. This would help eliminate accessory and reentrant conduction pathways.
- The term "maze" commonly refers to any operation that eliminates reentrant or accessory pathways by cryoablation, laser, or radiofrequency techniques.
- The term "maze" originally referred to the appearance of the endocardium after numerous incisions were made in a maze-like pattern.

Damus–Kaye–Stansel anastomosis

- It is a commonly used term when referring to connecting both ventricular outlets (aorta and PA) together to provide systemic blood flow in patients with single ventricle physiology. Pulmonary blood flow is supplied by a systemic-to-pulmonary artery shunt or via Fontan circulation.
- It was originally used in d-transposition of the great arteries with aortic valve stenosis patients as part of the Rastelli procedure. In these patients, the ventricles and great vessels are discordant. The left-sided LV is connected to the right-sided aorta via the left-sided pulmonary artery trunk. A right ventricle-to-pulmonary artery conduit is then placed for pulmonary blood flow. Right ventricular flow to the aorta is limited by the aortic stenosis.

Double switch procedure

- Most commonly used for repair of L-TGA (corrected transposition) patients.

- L-TGA patients have a right-sided LV and left-sided RV which results in ventriculoarterial discordance. The operation is undertaken since the RV in L-TGA is supporting the systemic circulation and will eventually fail.
- The operation involves switching the venous and arterial arrangements. The venous blood is routed to the contralateral ventricle (pulmonary venous blood routed to the right-sided LV and systemic venous blood routed to the left-sided RV). An arterial switch operation is also performed.

Fontan procedure

- It is a term that is most commonly used to refer to the third-stage operation for hypoplastic left heart syndrome. It may also be called a Stage 3 procedure.
- Here, one functional ventricle pumps blood to the body, and sufficiently low pulmonary artery pressures allow for pulmonary blood flow without a direct pumping chamber. Most often, the SVC and IVC blood flow is directed to the right pulmonary artery (and can flow to both lungs). This is often termed a "modified Fontan." Additionally, an atrial septal-type communication (fenestration in the graft material directing systemic venous blood to the pulmonary arteries) is normally created between the Fontan pathway and the pumping chamber's atrium. Combined, the procedure can then be termed the modified fenestrated Fontan (MFF) procedure. It may also be referred to as a total cavopulmonary anastomosis.
- A lateral tunnel Fontan redirects IVC blood through an intracardiac pathway to the RPA. Alternately, an external conduit can be placed to incorporate IVC blood up to the pulmonary arteries. This may be referred to as an external conduit Fontan or an extracardiac Fontan.
- The original Fontan procedure connected the right atrial appendage to the main pulmonary artery. This is no longer done due to improved flow dynamics with the Fontan procedures performed today.

Gott shunt

- This is a tube graft used to bypass the surgical site. It is most commonly used in the aorta to bypass an aneurysmal area during surgical repair. The main benefit of this approach is that the surgery can be performed off bypass while providing lower body perfusion throughout the repair.

Hemi–Fontan procedure

- This procedure may be used as the second-stage procedure for patients with single ventricle physiology.
- The procedure is very similar to the bidirectional Glenn procedure in that superior vena cava blood is directed to the pulmonary arteries. A main difference is that the right atrial–SVC connection is maintained and an atrial patch, or "dam", is used to direct SVC blood to the lungs. This simplifies an eventual Fontan procedure since the RA–SVC connection remains intact.

Holmes heart

- This is a form of single ventricle anatomy. It is characterized by hypoplastic right ventricle, double inlet left ventricle, and pulmonary stenosis.

Jatene operation

- This operation is commonly referred to as the arterial switch operation (ASO).
- It is used for d-transposition of the great arteries (with an intact ventricular septum or VSD), and it anatomically and physiologically corrects the circulation.
- The aorta (right-sided) and pulmonary artery (left-sided) are transected. The coronary arteries are removed as buttons from the right side and incorporated into the left-sided pulmonic trunk (which becomes the neoaortic root). The aorta and pulmonary artery are attached to their respective ventricles and associated septal defects are repaired.
- A LeCompte maneuver bringing the pulmonary arteries anterior to aorta is commonly performed.

Kawasaki disease

- Autoimmune disorder affecting blood vessels and organs throughout the body.
- Myocarditis with and without valve regurgitation may be found.

- Coronary artery aneurysms can be associated. These may be fatal without early medical treatment.
- Surgical treatment may be warranted. Retrograde cardioplegia may be indicated to provide adequate myocardial protection for patients with significant coronary artery aneurysms.

Kawashima procedure

- This operation is generally used to treat single ventricle patients who also have an interrupted IVC with azygous continuation. Here, the superior vena cava(e) receives the majority of the venous return.
- It is similar to the bidirectional Glenn in that SVC blood is diverted to the pulmonary circulation. However, a bilateral BDG is frequently required.
- The Kawashima differs from the BDG in that much more blood flow is diverted to the pulmonary arteries. The hepatic veins are commonly the only venous blood returning directly to the right side of the heart (excluding the coronary sinus). The SVC cannula for these operations is larger than normal to accommodate for this increased blood flow.

(Diverticulum of) Kommerell

- It is normally used to describe an anomalous left subclavian artery that arises from the proximal descending aorta in a patient with a right aortic arch. This vessel may course behind the esophagus resulting in a vascular ring which can cause tracheal and/or esophageal compression.
- It has also been described as an anomalous right subclavian artery in left aortic arch patients.
- The anomalous subclavian artery origin commonly has aneurysmal dilation.

Konno procedure

- Used to treat significant LVOTO by enlarging the anterior portion of the aortic root.
- A portion of the ventricular septum is incised to relieve that portion of the LVOTO, and a VSD patch is then used to separate the two ventricular chambers.
- A mechanical, bioprosthetic homograft, or autologous (Ross) aortic valve is placed in conjunction with the Konno procedure.

LeCompte maneuver

- The right pulmonary artery normally travels behind the ascending aorta. The LeCompte maneuver is used for d-TGA and other patients to improve flow characteristics and prevent obstruction to the relocated great arteries. It involves relocating the pulmonary arteries (right/main) anteriorly during the arterial switch operation.

LeCompte procedure

- This procedure has been used as part of the operation to treat d-TGA with pulmonary stenosis.
- The pulmonary trunk (vessel coming off of the LV) is implanted directly to the right ventricle without the use of a conduit (as in the Rastelli procedure). The left ventricular outflow is patched through the ventricular septum to the aorta (which comes off the right ventricle).

Manougian procedure

- This procedure is used to treat significant LVOTO by enlarging the posterior portion of the aortic root.
- It involves patch/homograft enlargement of the LVOT from the anterior leaflet of the mitral valve to the aortic root. The aortic valve is commonly replaced.
- It is similar to the Nicks procedure but involves the mitral apparatus as well.

Marfan's syndrome

- Genetic disorder affecting connective tissue throughout the body.
- Cardiac implications usually involve degeneration of connective tissue resulting in varying degrees of mitral and aortic valve regurgitation, aortic dilation, or even aortic aneurysm.

Maze procedure

- See Cox maze procedure.

Mustard procedure

- This is one of the atrial switch operations.
- It was used for isolated d-transposition of the great arteries (IVS or VSD), and it physiologically corrects the circulation. An intra-atrial pathway is created using pericardial tissue or graft material to satisfy two goals. First, right-sided venous blood is directed to the left side to be pumped out the left-sided pulmonary artery and ultimately onto the lungs. Second, the intra-atrial baffle directs left-sided pulmonary venous blood to the right side to be pumped out the right-sided aorta and ultimately the body.
- This procedure preceded the now standard ASO. The ASO anatomically and physiologically corrects the circulation. The Mustard only physiologically corrects the circulation and is less desirable because the right ventricle must then support the systemic circulation. The right ventricle eventually fails.
- The Mustard may be used in conjunction with an ASO for double switch operations for treatment of corrected transposition of the great arteries. Here, the atrial switch is performed to direct right-sided systemic venous blood to the left- sided anatomic right ventricle that pumps out to the pulmonary artery that has been moved from the right-sided ventricle (LV) to the left-sided ventricle (RV). The left-sided pulmonary venous blood is directed to the right-sided anatomic left ventricle that pumps out to the aorta that has been moved from the left-sided ventricle (RV) to the right-sided ventricle (LV).
- See Senning procedure for similarities.

Nicks procedure

- This procedure is used to treat significant LVOTO by enlarging the posterior portion of the aortic root.
- It involves patch/homograft enlargement of the LVOT across the aortic valve, but it does not involve the mitral apparatus as with the Manougian. The aortic valve is commonly replaced.
- It is similar to the Manougian procedure.

Nikaidoh procedure

- This operation is generally used to treat d-TGA with pulmonary stenosis (vessel coming off the left ventricle).
- The aortic root is removed from the right ventricular outflow tract and placed into the left ventricular outflow tract, which is remodeled as necessary. A VSD patch is placed. The right side receives a right-ventricle-to-pulmonary artery conduit or the pulmonary artery is directly attached to the RVOT.
- The primary advantage of this procedure is that it results in a more anatomically correct repair with less late LVOT complications as compared to the Rastelli procedure (also used for d-TGA with PS).

Noonan syndrome

- It is a genetic disorder that commonly includes a congenital cardiac anomaly such as an ASD, pulmonary valve issue and cardiomyopathy.
- Developmental delay, distinctive facial features, short stature, pectus excavatum, coagulation anomalies, and other defects may be found.

Norwood operation

- This operation is generally the first-stage operation to treat hypoplastic left heart syndrome. It may also be called a Stage 1 procedure.
- The diminutive aortic root is attached to the native pulmonary artery via a DKS type anastomosis. The native pulmonary artery becomes the neoaorta. This part of the operation provides systemic blood flow.
- Pulmonary blood flow is provided via a BTS or RV-PA connection (i.e., Sano shunt).

Pannus

- It is the fibrous tissue growth most commonly associated with prosthetic and biologic heart valves.
- This type of scar tissue can obstruct the valve or any portion of the outflow tract.

Pentalogy of Cantrell

- It is a collection of defects including omphalocele, diaphragmatic hernia, ectopic cordis, a lower sternal defect, and intracardiac anomalies.
- Patients usually present with several of the five associated defects. TOF is commonly associated.

Potts shunt

- AP communication created with a side-to-side anastomosis of the descending aorta to the left pulmonary artery.
- This surgical AP window was associated with LPA aneurysm formation and excessive pulmonary blood flow.
- It is rarely performed.

Rashkind procedure

- Balloon atrial septostomy. This is commonly performed on d-transposition of the great arteries patients with limited septal communication shortly after birth and before surgical correction.

Rastelli operation

- Creation of an intraventricular pathway between the left ventricle and aorta, through the ventricular septal defect, for patients with d-transposition, pulmonary stenosis (vessel coming off the LV) and a ventricular septal defect. A right ventricle-to-pulmonary artery conduit is placed for pulmonary blood flow (see LeCompte Procedure for similarities).

Ross procedure

- This operation involves replacing a diseased aortic valve (endocarditis, rheumatism, and significant regurgitation) with the patient's native pulmonary valve.
- The aortic valve may not be salvageable, and this procedure has the advantage of preventing the use of a mechanical valve on the left side of the heart. Lifelong anticoagulation may then be averted.
- A homograft is then placed in the pulmonary position.
- The Ross procedure has been modified in many important ways to increase the life of the valve and to prevent neo-aortic annular dilation with regurgitation.

Sano shunt

- The Sano shunt is a variant of the (modified) Blalock–Taussig shunt for treatment of patients with hypoplastic left heart syndrome as part of the Norwood procedure/Stage 1.
- This shunt uses a ringed PTFE conduit to connect the right ventricle to the pulmonary artery directly.

Scimitar syndrome

- A form of partial anomalous pulmonary venous return which is normally classified alongside atrial septal defects since it is a form of sinus venosus atrial septal defect.
- Named for the X-ray appearance of the enlarged curved sword (scimitar) shaped pulmonary venous drainage from the right lung to the IVC.

Senning operation

- This is one of the atrial switch operations.
- It is used for isolated d-transposition of the great arteries (intact ventricular septum or VSD), and it physiologically corrects the circulation. An intra-atrial pathway is created using primarily atrial tissue (not pericardial or graft material) to satisfy two goals. First, right-sided systemic venous blood is directed to the left side to be pumped out the left-sided pulmonary artery. Second, the intra-atrial baffle directs left-sided pulmonary venous blood to the right side to be pumped out the right-sided aorta and ultimately the body.
- This procedure preceded the now standard ASO. The ASO anatomically and physiologically corrects the circulation. The Senning only physiologically corrects the circulation and is less desirable because the right ventricle must then support the systemic circulation. The right ventricle eventually fails when required to be the systemic ventricle.
- The Senning procedure may be used in conjunction with an ASO for double switch operations for treatment of corrected transposition of the great arteries. Here, the atrial switch is performed to direct right-sided systemic venous blood to the left-sided anatomic right ventricle that pumps out to the pulmonary artery that has been moved from the right-sided ventricle (LV) to the left-sided ventricle (RV). The left-sided pulmonary venous blood is directed to the right-sided anatomic left ventricle that pumps out to the aorta that has been moved from the left-sided ventricle (RV) to the right-sided ventricle (LV).
- See Mustard procedure for similarities.

Shone's complex

- The collection of four left-sided defects of coarctation of the aorta, aortic valve stenosis, mitral ring, and mitral stenosis (due to abnormal papillary muscle attachments which may be termed "parachute mitral valve").

Takeuchi procedure

- This is an operation formerly used to treat ALCAPA. It is rarely used since coronary translocation results in more favorable outcomes.
- The left coronary artery is not translocated. It remains on the PA. An aortopulmonary communication is created to baffle aortic blood into the left coronary at its PA ostia.

Taussig–Bing anomaly

- This is a form of DORV where, due to the subaortic and subpulmonary anatomy, pulmonary venous flow preferentially crosses the VSD to the right side resulting in inadequate mixing (similar to d-TGA/IVS).

Trusler repair

- It generally refers to plication of an aortic valve cusp(s) to decrease leaflet prolapse.
- A pledgeted commissuroplasty is often done as well.

Van Praagh classification

- Drs. Richard and Stella Van Praagh and colleagues at Boston Children's Hospital developed a segmental approach to the classification of congenital cardiac defects [1].
- The classification system is an essential tool for clinicians since previous to its worldwide adoption, as the renowned cardiologist Donald Fyler commented regarding congenital heart defects, "…there are patients … who might be classified differently by two observers or even by one observer on two different occasions."
- The system uses a set of three letters denoting the status of the atria, ventricles, and great arteries. For example, a normal heart would be classified with the set {S,D,S} where the atria are situs solitus, the ventricles are normally rotated right (dextro), and the great vessels are concordant. A d-transposition of the great arteries patient would be classified with the set {S,D,D} where the atria are situs solitus, the ventricles are normally rotated right (dextro), but the great vessels are discordant.
- The system allows clinicians to more easily communicate a patient's anatomic condition by standardizing the classification of defects and the associated terminology.

Warden procedure

- It is used to treat sinus venosus atrial septal defects with partial anomalous pulmonary venous drainage.
- Before the Warden procedure, single patch techniques for baffling the anomalous pulmonary venous drainage through the right atrium and atrial septum (and part of the SVC if needed) to the left side suffered from obstruction issues.
- The Warden procedure improved flow characteristics by transecting the SVC above the anomalous drainage with insertion to the right atrial appendage and patching of the pulmonary veins through the right atrium to the left atrium through the atrial septum.
- Alternately, a two patch technique uses one patch to direct anomalous pulmonary drainage from the SVC to the left atrium while the second patch enlarges the anterior portion of the SVC-RA area to prevent obstruction.

Waterston shunt

- AP communication created with a side-to-side anastomosis of the ascending aorta to the right pulmonary artery.
- This surgical AP Window is no longer performed.

Williams syndrome

- Genetic disorder resulting in unique facial features, ophthalmic issues, developmental delay (but exceptional language skills), hypercalcemia, and cardiac defects.
- Supravalvar aortic stenosis is the most commonly associated cardiac defect.

Yasui procedure

- It is used for treatment of arch hypoplasia and aortic stenosis or other forms of significant LVOTO.
- The left ventricular outflow tract is routed from the left ventricle through the ventricular septum to the pulmonary valve (as in part of the Rastelli procedure). The pulmonic root is then attached to the aorta (as in the Damus–Kaye–Stansel procedure). The aortic arch is augmented. Additionally, a conduit is placed between the right ventricle and main pulmonary artery.

Reference

1 Van Praagh R. Transposition of the great arteries: history, pathologic anatomy, embryology, etiology, and surgical considerations. In *Cardiac Surgery: State of the Art Reviews*, vol. 5. Philadelphia: Hanley & Belfus, Inc; 1991:7–82.

CHAPTER 10

Abbreviations for congenital heart surgery

1 ventricle (single-ventricle) repair
- This term is often used when a patient has or progresses towards a Fontan circulation. The ventricular system pumps mixed systemic and pulmonary venous blood to the aorta while pulmonary blood flow is provided to a low resistance pulmonary arterial circuit via a systemic to pulmonary artery shunt or a total cavopulmonary anastomosis (Fontan).

1.5 ventricle (1.5V) repair
- This term may be used for the repair of a variety of congenital defects where the ventricle pumping to the lungs (pulmonary ventricle) cannot provide adequate pulmonary blood flow. Following this type of repair, the pulmonary ventricle delivers part of the systemic venous return (i.e., IVC blood) to pulmonary artery with the balance of pulmonary blood flow coming from a bidirectional Glenn. This may also be termed "partial Fontan circulation."

2 ventricle (2V) repair
- Most often, this term is used when one of the ventricles is hypoplastic and a decision must be made whether to repair (2V) or palliate (1 or 1.5V). In some patients, a hypoplastic ventricle over time and with acceptable growth can be sufficiently trained to provide the entire cardiac output. An example of this would be a patient with a hypoplastic RV that gets a modified Blalock–Taussig shunt (mBTS) as a first operation to provide adequate pulmonary blood flow for weeks to months. Then, with growth of the RV, the mBTS can be taken down and all defects surgically repaired resulting in normal two ventricle circulation.
- This broad term may encompass the majority of congenital cardiac anomaly repairs since a normal anatomy of two ventricles results. (An atrial septal defect or ASD closure patient maintains two ventricles after the repair.)

AA	aortic atresia or aortic arch
AAMI	American Association of Medical Instrumentation
AAMUF	arterio-arterial-modified ultrafiltration
aAo, AAo	ascending aorta
AAOCA	anomalous aortic origin of a coronary artery
AATS	American Association for Thoracic Surgery
ABE	acute bacterial endocarditis
ABG	arterial blood gas
ABX	antibiotics
ACE	advanced compact ECMO/ angiotensin-converting enzyme
ACT	activated clotting time
ALCAPA	anomalous left coronary from the pulmonary artery
AmSECT	American Society of ExtraCorporeal Technology
AO	aorta
AoD	descending aorta
AP	Aortopulmonary, anterior-posterior
APB	atrial premature beat
APC	aortopulmonary collateral
AR	aortic regurgitation
AS	aortic stenosis
ASA	aspirin
ASD	atrial septal defectASD1—ASD primum

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

ASD2	ASD secundum
ASO	arterial switch operation
AT 3 or ATIII	Antithrombin III
ATG	Antithymoglobulin
ATP	adenosine triphosphate
AV	aortic valve, atrioventricular or arteriovenous
AVD	augmented (or assisted) venous drainage
AVM	arteriovenous malformation
AVMUF	arteriovenous-modified ultrafiltration
AVN	atrioventricular node
AVR	aortic valve repair/aortic valve replacement
AVR/R	aortic valve repair/replacement
AVSD	atrioventricular septal defect (same as CAVC)
AVV	atrioventricular valve
AVVR	Atrioventricular valve regurgitation
BAE	bilateral atrial enlargement
BAS	balloon atrial septostomy
BAV	bicuspid aortic valve
BBB	bundle branch block
BBDG	bilateral bidirectional Glenn
BCAV	bicuspid aortic valve
BD	balloon dilation
BDF	bidirectional flow
BDG	bidirectional Glenn
BES	balanced electrolyte solution
BiVAD	biventricular assist device
BL	bilateral
BLT	bilateral lung transplant
BMI	body mass index
BMT	bone marrow transplant
BPM	beats per minute
BSA	body surface area
BT	bovine trunk
BTS	Blalock–Taussig shunt
BV	bridging vein
BVF	biventricular function, biventricular flow, or bulboventricular foramen
C/B	complicated by
C/F	circulatory failure
C-TGA	corrected transposition of the great arteries (same as L-TGA)
CA	coronary artery/cardiac arrest/ circulatory arrest/cold agglutinins/cancer
CAD	coronary artery disease
CAP	central aortic pressure
CAT	circulatory arrest time
CAVC	complete atrioventricular canal
CBF	cerebral blood flow
CCP	certified clinical perfusionist
CCT	cross-clamp time
CCTGA	congenitally corrected transposition of the great arteries
CDH	congenital diaphragmatic hernia
CF	cystic fibrosis
CHB	complete heart block
CHD	congenital heart disease
CI	cardiac index ($L/min/m^2$)
cmH_2O	centimeters of water (unit of pressure measurement)
CMO	cardiomyopathy
cMRI or CMR	cardiac magnetic resonance imaging
$CMRO_2$	cerebral metabolic rate of/for oxygen
CMV	cleft mitral valve or cytomegalo virus
CNS	central nervous system
CO	cardiac output
CO_2	carbon dioxide
CoA	coarctation of the aorta
CPAP	continuous positive airway pressure
CPB	cardiopulmonary bypass
CPBT	cardiopulmonary bypass time
CPG	cardioplegia
CPP	cerebral perfusion pressure
CPR	cardiopulmonary resuscitation
CRP	C-reactive protein
CS	coronary sinus, cell saver, or cerebrospinal
CSSD	coronary sinus septal defect
CTA	computed tomography angiogram
CTD	connective tissue disease
CUF	conventional ultrafiltration
CVA	cerebrovascular accident
CVP	central venous pressure
CVR	cardiotomy venous reservoir or cerebral vascular resistance
CXR	chest X-ray
D/T	due to
d-TGA	dextro transposition of the great arteries
dAo, DAo	descending aorta
DCM	dilated cardiomyopathy
DCRV	double chamber right ventricle
DHCA	deep hypothermic circulatory arrest
DILV	double inlet left ventricle
DIRV	double inlet right ventricle

DKS	Damus–Kaye–Stansel anastomosis
DNA	deoxyribonucleic acid
DOE	dyspnea on exertion
DOL	day of life
DOLV	double outlet left ventricle
DORV	double outlet right ventricle
DTI	direct thrombin inhibitor
DUF	dilutional ultrafiltration
ECC	extracorporeal circulation
ECD or ECCD	endocardial cushion defect (same as CAVC)
ECG	electrocardiogram
ECHO	echocardiography
ECMO	extracorporeal membrane oxygenation
ECPR	ECMO for cardiopulmonary resuscitation
EDP	end diastolic pressure
EDV	end diastolic volume
EEG	electroencephalogram
EF	ejection fraction
EFE	endocardial fibroelastosis
EGA	estimated gestational age
EKG	electrocardiogram, same as ECG
EP (S)	electrophysiology (study)
ESR	estimated sedimentation rate
EST	exercise stress test
ESV	end systolic volume
ETEA	end-to-end anastomosis
ETT	exercise tolerance test
F/B	followed by
FFP	fresh frozen plasma or fenestrated Fontan procedure
FiO_2	fractional inspired oxygen concentration
FPP	fellow of pediatric perfusion (AmSECT designation)
Fr	French size
FTT	failure to thrive
FXN	function
GA	gestational age or general anesthesia
GETA	general endotracheal anesthesia
GME	gaseous microemboli
GSD	gravity siphon drainage
GVHD	graft versus host disease
HCA	hypothermic circulatory arrest
HCM	hypertrophic cardiomyopathy
HCT	hematocrit
HD	hemodynamic
HDR	heparin dose–response (curve)
HFNC	high flow nasal cannula
HGB	hemoglobin
HIT	heparin-induced thrombocytopenia
HITT	heparin-induced thrombocytopenia with thrombosis
HLHS	hypoplastic left heart syndrome
HLM	heart–lung machine
HTN	hypertension
HTX or HT	heterotaxy syndrome or heart transplant
Hx	medical history
IAA	interrupted aortic arch
IABP	intra-aortic balloon pump
IAS	intact atrial septum
ICH	intracranial hemorrhage
IFT	induced fibrillation time
iNO	inhaled nitric oxide
INR	international normalized ratio
IU	international units
IUGR	intrauterine growth retardation
IVC	inferior vena cava
IVS	intact ventricular septum or interventricular septum
JET	junctional ectopic tachycardia
KAVR	kinetic-assisted venous return
kg	kilogram
KVAD	kinetic venous-assisted drainage
KVO	keep vein open (for intravenous infusions)
L	liter or left
L–R	left to right (to qualify a shunt)
L-TGA	levo transposition of the great arteries (same as C-TGA and Levo-TGA)
LA	left atrium or ligamentum arteriosum
LA-FA	left atrial to femoral artery
LAA	left aortic arch
LAANB	left aortic arch with normal branching
LAE	left atrial enlargement
LAVVR	left atrioventricular valve regurgitation
LBB	left bundle branch
LCA	left coronary artery, left carotid artery
LCC	left coronary cusp
LCCA	left common carotid artery
LCOS	low cardiac output syndrome (or state)
LCX	left circumflex
LD	left dominant
LFA	left femoral artery
LFV	left femoral vein
LHB	left heart bypass
LHF	left heart failure

LIJV	left internal jugular vein
LIMA	left internal mammary artery
LLE	left lower extremity
LLL	left lower lobe
LLPV	left lower pulmonary vein
LMBTS	left modified Blalock–Taussig shunt
LMCA	left main coronary artery
LOC	loss of consciousness
LOS	length of stay (hospital admission)
LP	licensed perfusionist
LPA	left pulmonary artery
LPCW (P)	left pulmonary capillary wedge (pressure)
LPM	liters per minute
LPV	left pulmonary vein(s)
LR	lactated ringers (solution) or leukocyte reduced (blood products)
LS	lung scan
LSA	left subclavian artery
LSCA	left subclavian artery
LSVC	left superior vena cava
LT	lateral tunnel (for Fontan operation)
LTR	left to right
LUL	left upper lobe
LUPV	left upper pulmonary vein
LV	left ventricle
LVAD	left ventricular assist device
LVEDP	left ventricular end diastolic pressure
LVEDV	left ventricular end diastolic volume
LVEF	left ventricular ejection fraction
LVH	left ventricular hypertrophy
LVNC	left ventricular non compaction
LVOT	left ventricular outflow tract
LVOTO	left ventricular outflow tract obstruction
LVSP	left ventricular systolic pressure
m^2	meters squared
MA	mitral atresia
MAC	minimum alveolar concentration
MAE	massive air embolus
MAP	mean arterial pressure or mean airway pressure
MAPCAs	major aortopulmonary collaterals
MB	muscle bundle
MCA	middle cerebral artery
MECC	miniaturized extracorporeal circuit
MFF	modified fenestrated Fontan
MI	myocardial infarction or less commonly, mitral insufficiency
MIB	mirror image branching
MIG	maximum instantaneous gradient
Min	minute or minimum
mL	milliliter
mmHg	millimeters of mercury
MOV	minimum operating volume
MPA	main pulmonary artery
MR	mitral regurgitation
MRI	magnetic resonance imaging
MS	mitral stenosis
MUF	modified ultrafiltration
MV	mitral valve
MVI	multivitamin
NB	normal branching (in relation to the aortic arch)
NC	nasal cannula
NEC	necrotizing enterocolitits
NIRS	near infrared spectroscopy
NKDA	no known drug allergies
NL	Normal
NPO	nil per os (Latin for nothing by mouth)
NSR	normal sinus rhythm
NSS	normal saline solution
NSVD	normal spontaneous vaginal delivery
OHT	orthotopic heart transplant
OR	operating room
OSA	obstructive sleep apnea
OSH	outside hospital
PA	pulmonary artery or pulmonary atresia
PAC	premature atrial contraction
$PaCO_2$	partial pressure (arterial) of carbon dioxide
PAH, PAHTN	pulmonary artery hypertension
PaO_2	partial pressure (arterial) of oxygen
PAP	pulmonary artery pressure or peak airway pressure
PAPVC	partial anomalous pulmonary venous connection (same as PAPVR)
PAPVR	partial anomalous pulmonary venous return (same as PAPVC)
PAVC	partial atrioventricular canal defect
PBF	pulmonary blood flow
PBUF	prebypass ultrafiltration
PCW(P)	pulmonary capillary wedge (pressure)
PDA	patent ductus arteriosus
PE	pulmonary embolus or phenylephrine
PFO	patent foramen ovale
PGE	prostaglandin E
PI	pulmonary insufficiency

PLE protein losing enteropathy
PLSVC persistent left superior vena cava
PLT(s) platelets
PM pacemaker
PMHX past medical history
PPS peripheral pulmonary stenosis
PPV positive pressure ventilation
PR pulmonary regurgitation
PRBC(s) packed red blood cells
PS pulmonary stenosis
PSEG peak systolic ejection gradient
PT prothrombin time
PTT partial thromboplastin time
PTX Phototherapy
PV pulmonary valve or pulmonary vein
PVC premature ventricular contraction
$PvCO_2$ partial pressure (venous) of carbon dioxide
PVL perivalvar leak
PVO pulmonary vein obstruction
PvO_2 partial pressure (venous) of oxygen
PVR pulmonary valve repair, pulmonary valve replacement or pulmonary vascular resistance
PVR/R pulmonary valve repair/replacement
PVS pulmonary vein stenosis or pulmonary valve stenosis
PVW (P) pulmonary vascular wedge (pressure)
Q blood flow (quantity)
QoL quality of life
Qp:Qs ratio of pulmonary (p) to systemic(s) blood flow (Q)
R right
R-L right to left (to qualify a shunt flow)
R/O rule out
RA right atrium or room air
RAA right aortic arch or right atrial appendage
RAAMIB right aortic arch, mirror image branching
RACHS risk adjustment in congenital heart surgery
RAD reactive airway disease
RAE right atrial enlargement
RAP retrograde autologous prime
RAVVR right atrioventricular valve regurgitation
RBB right bundle branch
RCA right coronary artery, right carotid artery
RCCA right common carotid artery
RCCP regional cerebral and coronary perfusion
RCCSP regional cerebral, coronary, and splanchnic perfusion
RCM restrictive cardiomyopathy
RCP regional cerebral perfusion or less commonly retrograde cerebral perfusion
RCSP retrograde coronary sinus perfuser (or perfusion)
RD right dominant
RF regurgitant fraction, radiofrequency or risk factors
RFA right femoral artery or radio frequency ablation
RFV right femoral vein
RHF right heart failure
RIJV right internal jugular vein
RIMA right internal mammary artery
RLE right lower extremity
RLFP regional low flow perfusion
RLL right lower lobe
RLPV right lower pulmonary vein
RMBTS right modified Blalock–Taussig shunt
RMCA right main coronary artery
RML right middle lobe
RMPV right middle pulmonary vein
RNA ribonucleic acid
ROP retinopathy of prematurity
ROSC return of spontaneous circulation
RPA right pulmonary artery
RPCW (P) right pulmonary capillary wedge (pressure)
RPM revolutions per minute
RPT regional perfusion time
RPV right pulmonary vein(s)
RSCA right subclavian artery
RSA right subclavian artery
RSVC right superior vena cava
RTL (F) right to left (flow)
RUL right upper lobe
RUPV right upper pulmonary vein
RV right ventricle
RVAD right ventricular assist device
RVDCC right ventricular dependent coronary circulation
RVEDP right ventricular end diastolic pressure
RVEDV right ventricular end diastolic volume
RVEF right ventricular ejection fraction
RVH right ventricular hypertrophy
RVMB right ventricular muscle bundle
RVOT right ventricular outflow tract
RVOTO right ventricular outflow tract obstruction
RVSP right ventricular systolic pressure
RVVO right ventricular volume overload

RWB reconstituted whole blood (from FFP and RBCs); same as "RBCs with plasma added"
S/P status post
SA sinoatrial
SAM systolic anterior motion (of the mitral valve which may result in LVOTO)
SAN sinoatrial node
SBP systolic blood pressure
SCA subclavian artery
SCP selective cerebral perfusion
SMA surface-modifying agent
SMUF simplified modified ultrafiltration
SOV sinus of valsalva
STJ sinotubular junction
STS Society of Thoracic Surgeons
SV single ventricle or sinus venosus (ASD)
SVASD sinus venosus atrial septal defect
SVC superior vena cava
SVR systemic vascular resistance
SVT supraventricular tachycardia
T/D take down
T21 trisomy 21 (Down's syndrome)
TA truncus arteriosus or tricuspid atresia
TAC truncus arteriosus communis
TAP transannular patch
TAPVC total anomalous pulmonary venous connection (same as TAPVR)
TAPVR total anomalous pulmonary venous return (same as TAPVC)
TAVC transitional atrioventricular canal
TAVI transcatheter (or transcutaneous) aortic valve implantation
TAVR transcatheter (or transcutaneous) aortic valve replacement
TCD transcranial Doppler
TCPC total cavopulmonary connection (same as Fontan)
TEE transesophageal echocardiography
TET tetralogy of Fallot (same as TOF)
TGA transposition of the great arteries
TGV transposition of the great vessels; same as TGA
TKO to keep open (for intravenous infusions)
TOF tetralogy of Fallot (same as TET)
TPG transpulmonary gradient
TPN total parenteral nutrition
TR tricuspid regurgitation
TRJ tricuspid regurgitation jet
TS tricuspid stenosis
TV tricuspid valve
Tx treatment or transplant
UAV unicuspid aortic valve
UCS unroofed coronary sinus
UFH unfractionated heparin
US ultrasound
V/Q ventilation to perfusion ratio
VAD ventricular assist device
VAMUF venoarterial modified ultrafiltration
VAP venous autologous prime
VATS video-assisted thoracic surgery
VAVD vacuum-assisted venous drainage
VAVR vacuum-assisted venous return
VBG venous blood gas
VI ventricular inversion
VO_2 volume of oxygen uptake; oxygen consumption
VPB ventricular premature beat
VSD ventricular septal defect
VVCs venovenous collaterals
VVMUF venovenous-modified ultrafiltration
WB whole blood
WCC well child checkup
WHO World Health Organization
WOB work of breathing
WPW Wolff–Parkinson–White syndrome
WU Wood units
***Z*-Score** standard deviation above or below the normal value for a given patient age or size
ZBUF zero balance ultrafiltration

CHAPTER 11

Recommended reference books

Cardiac anesthesia textbooks

DiNardo JA, Zvara DA. Anesthesia for Cardiac Surgery, 3rd edition. Oxford: Blackwell Publishing Ltd; 2007.

Hensley FA Jr, Martin DE, Gravlee GP. A Practical Approach to Cardiac Anesthesia, 5th edition. Philadelphia: Lippincott Williams & Wilkins; 2012.

Congenital cardiac intensive care textbook

Chang AC, Hanley FL, Wernovsky G, Wessel DL. Pediatric Cardiac Intensive Care. Baltimore: Williams & Wilkins; 1998.

Mechanical circulatory support textbook

Selke FW, del Nido P, Swanson SJ. Sabiston and Spencer Surgery of the Chest. Philadelphia: WB Saunders; 2010.

Congenital cardiac surgery textbooks

Jonas RA. Comprehensive Surgical Management of Congenital Heart Disease, 2nd edition. London: CRC Press; 2014.

Selke FW, del Nido P, Swanson SJ. Sabiston and Spencer Surgery of the Chest. Philadelphia: WB Saunders; 2010.

Stark JF, de Leval MR, Tsang VT. Surgery for Congenital Heart Defects, 3rd edition. Chichester: John Wiley & Sons, Ltd; 2006.

Cardiopulmonary bypass textbooks

Gravlee GP, Davis RF, Stammers AH, Ungerleider RM. Cardiopulmonary Bypass: Principles and Practice, 3rd edition. London: Lippincott Williams & Wilkins; 2008.

Mora C. Cardiopulmonary Bypass: Principles and Techniques of Extracorporeal Circulation. New York: Springer-Verlag; 1995.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

CHAPTER 12

Comprehensive experience-based equipment selection chart
Select medications administered during bypass

Table 12.1 is an example of a comprehensive experience-based equipment selection chart. A perfusion program should develop its own chart based on its patient population, the hardware and disposables in use, and surgeon preferences.

Table 12.2 lists medications commonly administered during bypass. Institutional dosing strategies will vary.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

Table 12.1 Comprehensive experience-based equipment selection chart.

PERFUSION FOR CONGENITAL HEART SURGERY, Matte 2015

Component	Prime	MOV	Manufacturer max flow	02 m2	Heater m2	A,V, Boot	Prime (Heparin)	BSA for 3.0 index/max flow
FX-05	43	15	1500	0.5	0.035	3/16,1/4,3/16	215 (645u)	<0.40/1200
Reservoir	75							
Art	30					3/16,1/4,1/4	230 (690u)	0.41-0.5/1500
Ven	52							
Boot	15/30					SMUF Circuit Volume = 70 ml		

Component	Prime	MOV	Manufacturer max flow	02 m2	Heater m2	A,V, Boot	Prime (Heparin)	BSA for 3.0 index/max flow
FX 15-30	144	70	4000	1.5	0.14	1/4, 1/4, 1/4	460 (1380u)	0.51-0.60/1800
Reservoir	150		(5 with KVAD)			1/4, 3/8, 1/4 1/4, 3/8, 3/8	535 (1605u) 570 (1710u)	0.61-0.70/2100 0.71-1.05/3150
Art	75/175					3/8, 3/8, 3/8	**670 (2010u)**	1.06-1.20/3600
Ven	61/136		75 cc, centrifugal head and tubing			3/8, 3/8, 3/8, KVAD	**745 (2235u)**	1.21-1.46/4400
Boot	30/65/90					3/8, 3/8, 1/2, KVAD	**770 (2310u)**	1.47-1.66/5000

Component	Prime	MOV	Manufacturer max flow	02 m2	Heater m2	A,V, Boot	Prime (Heparin)	BSA for 3.0 index/max fiow
FX 25	250	200	7000	2.5	0.2	3/8, 3/8, 1/2, KVAD	980 (2940u)	>=1.67/>5000
Reservoir	250							
Art	175					* May downsize 2-4 Fr. with KVAD		
Ven	140		75 cc, centrifugal head and tubing			* Straight Venous add 2-4 Fr. above DLP <		
Boot	90							

Kg	Blood volume
<10	85
10-20	80
20-30	75
30-40	70
>40	65

Max flows (cc/min) (max RPM 162)
3/16 boot 1200, arterial 1500
1/4 boot 2100, arterial 3150, ven 1800
3/8 boot 4400, arterial >3150, ven 3750

Tubing	cc/ft	cc/rev
3/16	5	7.2
1/4	9.7	12.5
3/8	21.7	26
1/2	38.6	44.5

Kg Arterial (Man, LPM)	
<4.5	8 **BIO** (0.7) 6 DLP (0.4)
4.5-10	10 **BIO** (1.3)
10-14	12 **BIO** (2.1)
14-28	14 **BIO** (2.8)
25-35	16 ***DLP*** (3.8)
28-50	18 *EOPA LL* 4.8 18*EOPA CAP* 2.6
50-75	18 *EOPA LL* 4.8 20 *EOPA CAP* 4.1
75-90	20 *EOPA LL* 5.7 22 *EOPA CAP* 5.4
>90	22 *EOPA LL* >5.7

Femoral Art	
<4.5	8 **RMI** (0.825)
4.5-10	10 **RMI** (1.25)
10-14	12 **RMI** (2.0)
14-28	14 **RMI** (2.4)
28-40	15 ***BIO*** (3.0)
40-55	17 ***BIO*** (4.0)
55-75	19 ***BIO*** (5.3)
>75	21 ***BIO*** (6.5)

Kg	DLP < Venous S/IVC
<3	12/12 12=0.8/T0.7/D0.6/E1.0
3-6	12/14 14=1.6/T1.1/D0.8/E1.3
6-8	12/16 16=1.9/T1.5/D1.4/E1.8
8-12	14/16 T=Terumo
12-16	14/18 18=2.6/T2.0/D1.8/E2.6
16-22	16/18 D=DLP PVC tip
22-30	16/20 20=3.0/T2.4/D2.4/E3.3
30-34	18/20 E=Edwards LS
34-46	18or20 /22
46-58	20/22 22=4.5/T2.7/D3.1
58-75	20/24 24=5.5/ T4.1/D3.5
>75	22/24

Femoral Ven (LPM, KVAD LPM)	DLP STR VEN	
8 ***BIO*** (0.47)	12	600
10 ***BIO*** (0.9) 10 **RMI** (0.8)	14	800
12 ***BIO*** (1.45) 12 **RMI** (1.3)	16	1400
14 ***BIO*** (2.0) 14 **RMI** (2.0)	18	1800
15 ***BIO*** (1.3)	20	2400
17 ***BIO*** (1.8, 2.5)	22	3100
19 ***BIO*** (2.5, 3.5)	24	3500
21 ***BIO*** (3.2, 4.5)	26	4400
23 ***BIO*** (4.0, 5.6)	28	5200
25 ***BIO*** (4.7, 6.6)		
27 ***BIO*** (5.8)		
29 ***BIO*** (6.5)		

ETT Venous	
Kg	**ETT Size**
<9	4.0
9-20	4.5
20-35	5.0
35-50	5.5
>50	6.0
<=5.0 3/16*1/4 >=5.5 1/4*1/4 or 1/4*3/8	

Single	DLP < Ven
<5	14
5-12	16
12-18	18
18-26	20 (1/4 OR 3/8)
26-55	22
>55	24-28

Dual Stage (KVAD)	
29/29	30-65 (45-85) May also use for 20-30Kg depending on surgeon preference
29/37	>65 (>85)

Table 12.2 Select medications administered during bypass.

Select medications administered during bypass	Common bolus dose	Half life	Mechanism of action/effect
Calcium chloride	5–20 mg/kg, up to 1 g/dose; contains 3× the available calcium ions relative to the gluconate form	Not applicable	Normalized ionized calcium levels are important for cardiac contractility, particularly in neonates and infants
Calcium gluconate	25–100 mg/kg, up to 1 g/dose; contains 1/3 the available calcium ions relative to the chloride form.	Not applicable	Normalized ionized calcium levels are important for cardiac contractility, particularly in neonates and infants
Cefazolin	25 mg/kg, up to 2 g/dose	108 min	First generation cephalosporin; broad spectrum
Diphenyhydramine	1–2 mg/kg, up to 50 mg/dose	8–12 h	H1 receptor antagonist; antihistamine
Epinephrine	0.05–10 μg/kg	2 min	Beta-1 agonist; vasopressor and inotrope
Furosemide	0.2–1 mg/kg, up to 20 mg/dose	60–100 min	Loop Diuretic
Heparin	Bolus for CPB: 300–400 IU/kg During CPB: 50–150 IU/kg to maintain target ACT/ heparin concentration	60–150 min	Anticoagulant; potentiates the effect of antithrombin III
Lidocaine	1–1.5 mg/kg, up to 100 mg	1–3 h	Sodium channel blocker; Class I antiarrhythmic; membrane stabilizer
Magnesium sulfate	10–25 mg/kg, up to 2 g/dose	not applicable	Calcium channel blocker
Mannitol	0.25–0.5 g/kg, up to 25 g/dose	100 min	Osmotic diuretic, free radical scavenger
Methylprednisolone	30 mg/kg, up to 3 g/dose	18–26 h	Corticosteroid
Milrinone	25–100 μg/kg	1–3 h	Phosphodiesterase inhibitor
Nitroglycerin	0.5–10 μg/kg	3 min	Coronary vasodilator; greater venous vasodilation than arterial vasodilation
Nitroprusside	0.5–10 μg/kg	10 min	Vasodilator with equal effect on arteries and veins
Phenylephrine	0.1–0.5 μg/kg	150 min	Alpha agonist
Phentolamine	0.1–0.5 mg/kg, up to 5 mg/dose	19 min	Alpha blocker
Sodium bicarbonate	0.5–2 mEq/kg or (base deficit) (kg) (0.3)	Not applicable	Alkalinizing agent
Vancomycin	10–20 mg/kg; must be given over ≥60 min to prevent red man syndrome caused by nonallergic mast cell degranulation	4–11 h	Inhibits cell wall synthesis in Gram-positive bacteria
Zosyn	75–100 mg/kg, up to 3 g/dose	42–72 min	Inhibits cell wall synthesis and septum formation during cell division

Index

Note: Page numbers in *italics* refer to Figures; those in **bold** to Tables.

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

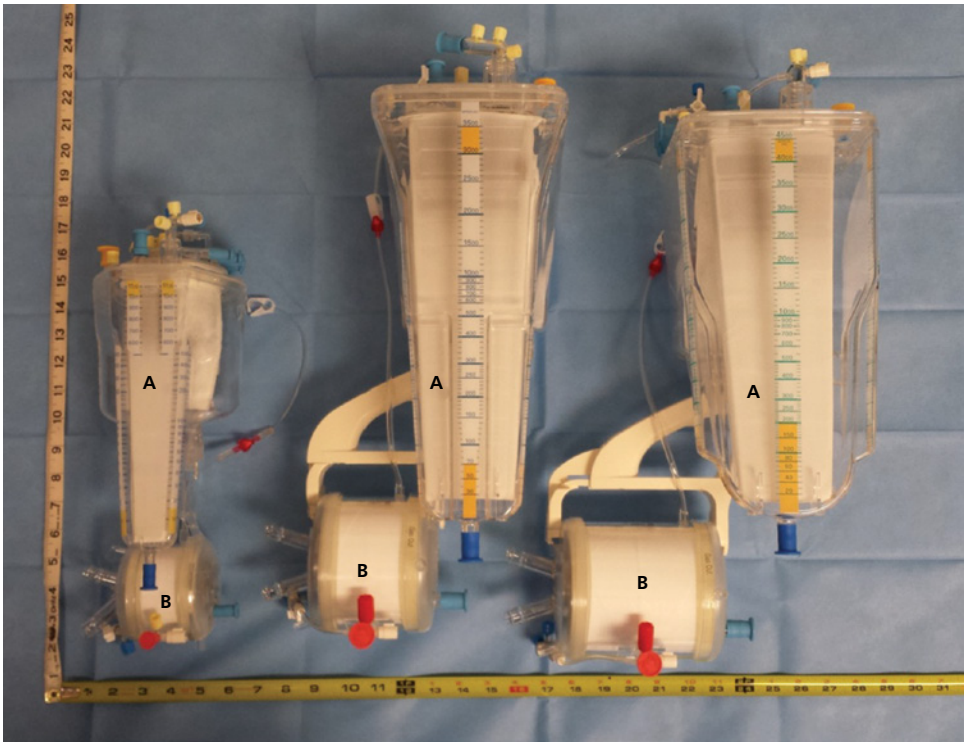


Figure 1.2 Terumo CAPIOX FX series of oxygenators. *Left to right: Terumo CAPIOX FX05, Terumo CAPIOX FX15-30, Terumo CAPIOX FX25. A—cardiotomy venous reservoir and B—oxygenator membrane with integrated arterial line filter and heat exchanger.*

Perfusion for Congenital Heart Surgery: Notes on Cardiopulmonary Bypass for a Complex Patient Population, First Edition. Gregory S. Matte.

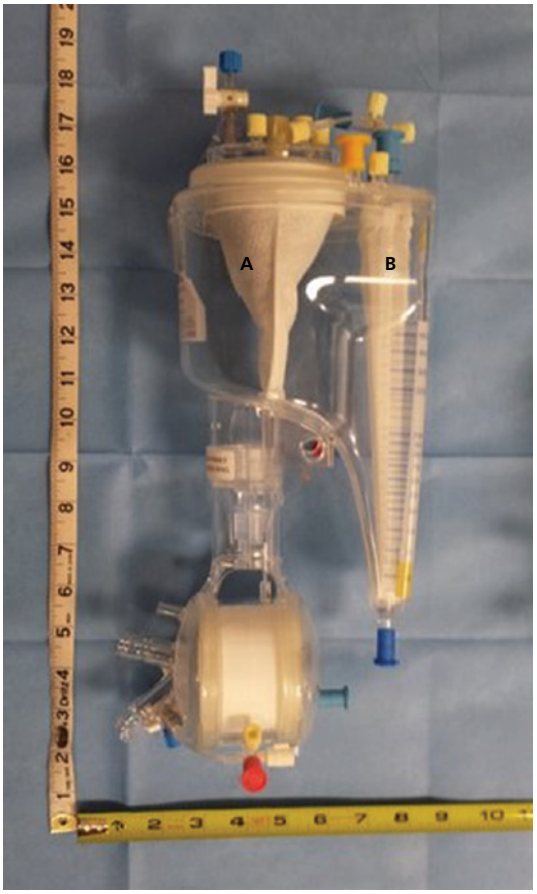


Figure 1.6 Filters in the Terumo CAPIOX FX05 oxygenator. *A—cardiotomy filter in the CVR. B—venous filter in the CVR.*

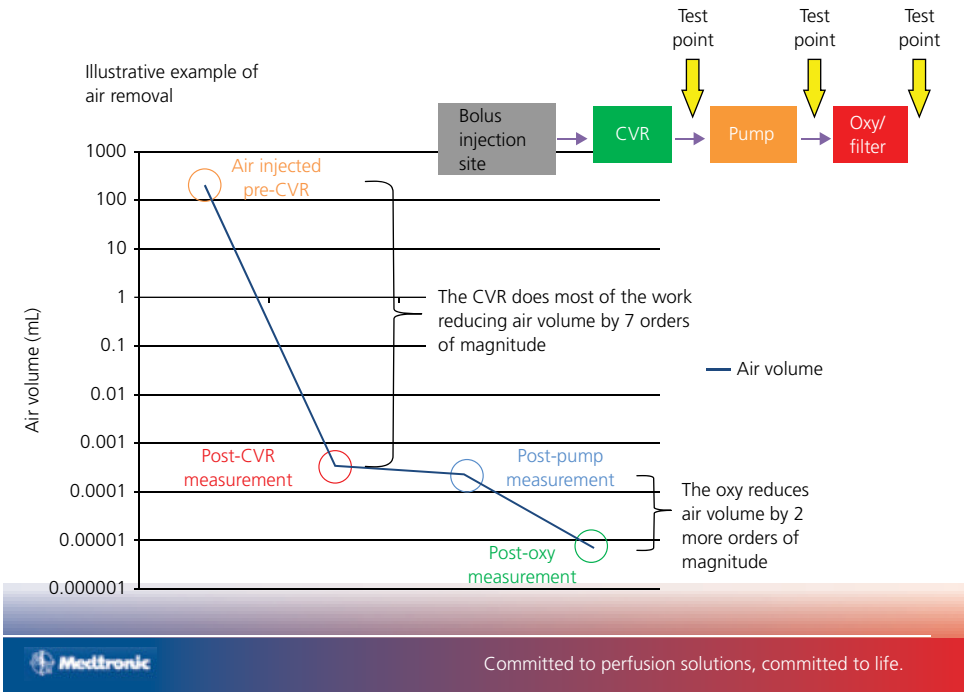


Figure 1.7 Air removal in the CVR and oxygenator. Reproduced with permission from Medtronic, Inc., Minneapolis, MN. All rights reserved.

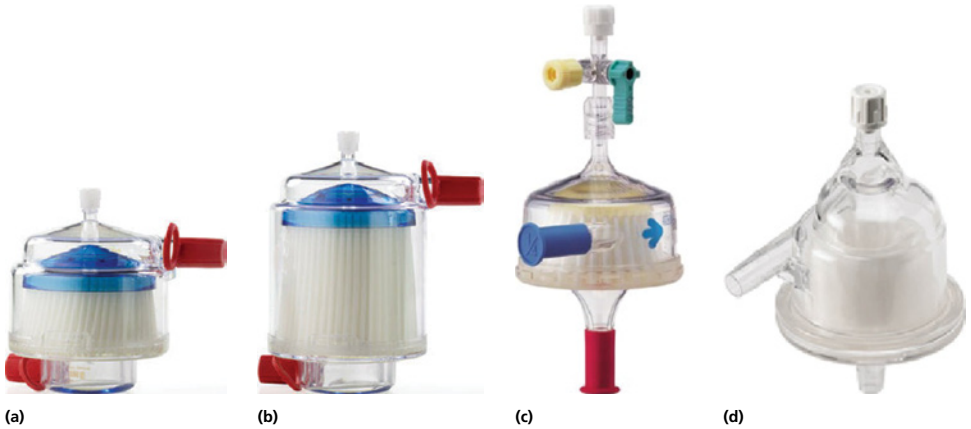


Figure 1.8 External arterial line filters. **(a)** *Sorin Group D736*. **(b)** *Sorin Group D733*. **(a)** and **(b)** Reproduced with permission from Sorin Group USA Inc., Arvada, CO. All rights reserved. **(c)** *Terumo Capiox AF02*. Reproduced with permission from Terumo Cardiovascular Group, Ann Arbor, MI. All rights reserved. **(d)** *Medtronic Affinity Pixie*. Reproduced with permission from Medtronic, Inc., Minneapolis, MN. All rights reserved.

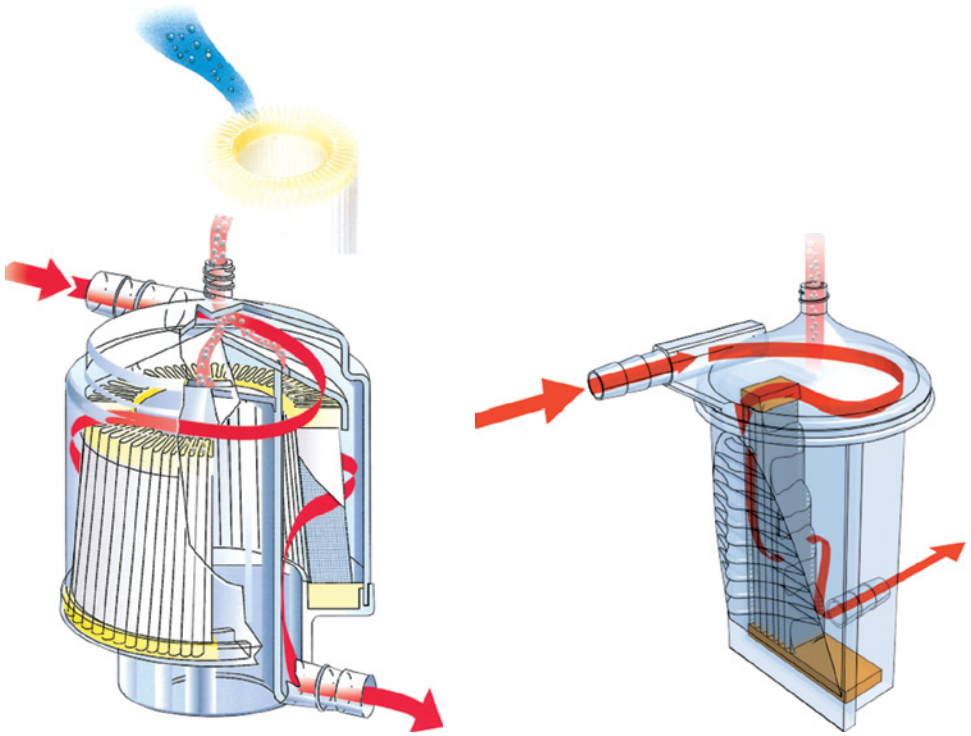


Figure 1.9 Typical flow path through external arterial line filters. The top luer connector purges continuously via a line connected to the CVR. Reproduced with permission from Sorin Group USA Inc., Arvada, CO. All rights reserved.

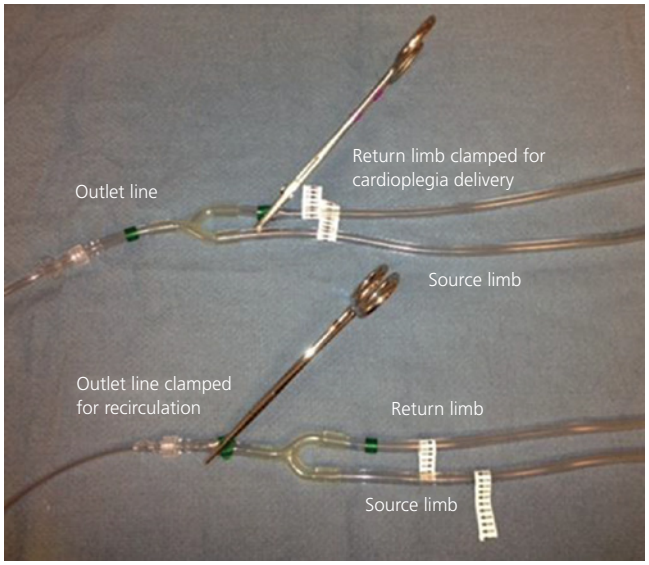
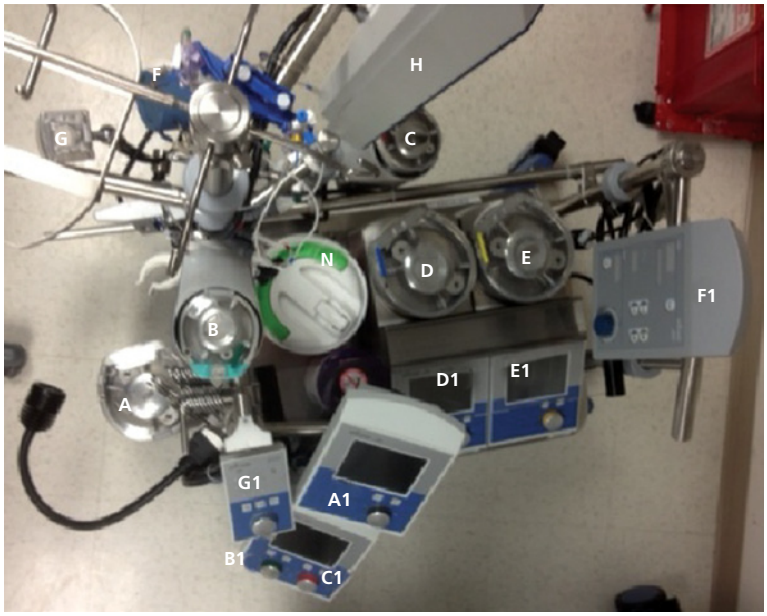


Figure 1.10 Recirculating cardioplegia system table lines. Top: Recirculating delivery system with table line return limb clamped for cardioplegia delivery. Bottom: Recirculating delivery system with table line outlet limb clamped for recirculation.



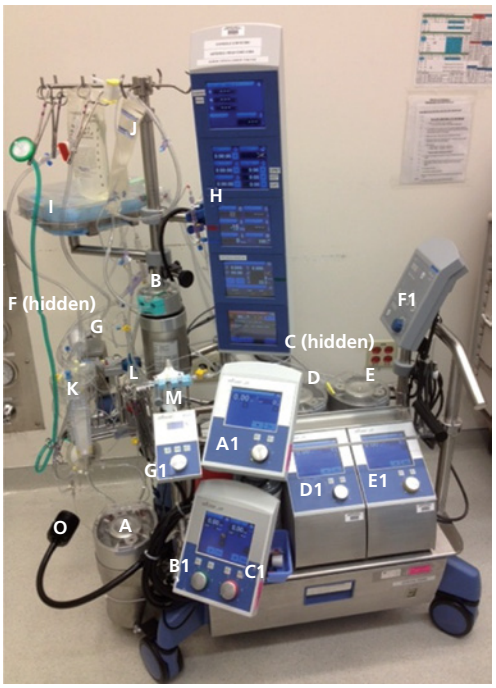


Figure 1.13 Top and side views of a customized Stockert S5 heart–lung machine. *A—Arterial head and its controller A1, B—cardioplegia head and its controller B1, C—vent head and its controller C1, D—field sucker and its controller D1, E—field sucker and its controller E1, F—centrifugal head motor (for kinetic venous-assisted drainage) and its controller F1, G—electronic venous occluder and its controller G1, H—master display tower, I—sterile custom tubing pack, J—custom cardioplegia tubing set, K—oxygenator, L—hemoconcentrator, M—blood gas sampling manifold, and N—ice bucket for cardioplegia cooling coil.*

Figure 1.14 Stockert heater-cooler system 3 T (three-tank system). *A—Cold cardioplegia circuit. The first tank instantly provides 2–10 °C water to the cardioplegia system. B—Warm cardioplegia circuit. The second tank instantly provides 15–41 °C water to the cardioplegia system. It can also be fitted for a water-based patient surface cooling/heating blanket. C—Oxygenator circuit. The third tank provides 15–41 °C water to the oxygenator heat exchanger. It has a second pump and circuit to provide water flow from the same tank to a water-based patient blanket if desired.*

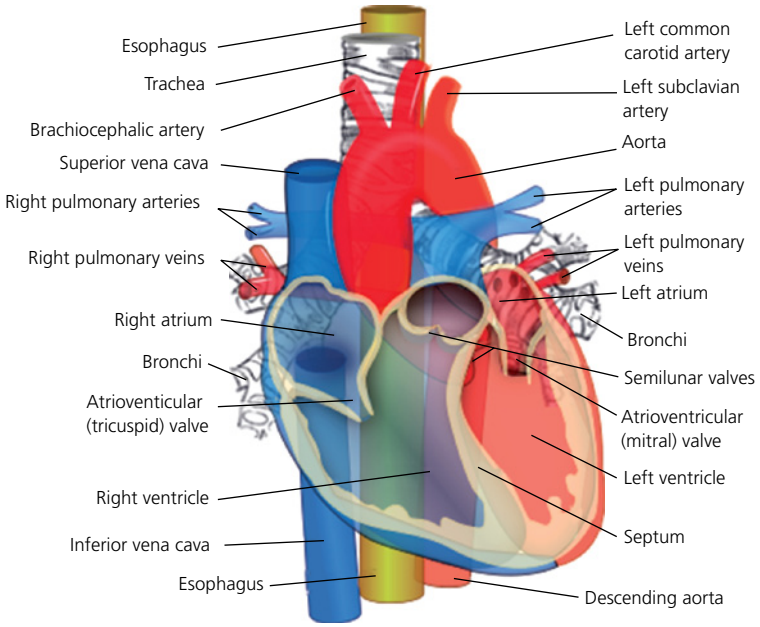


Figure 1.15 General cardiac anatomy. http://en.wikipedia.org/wiki/File:Relations_of_the_aorta,_trachea,_esophagus_and_other_heart_structures.png. via Wikipedia. Reproduced with permission from under the Creative Commons Attribution-Share Alike 3.0. Unported license (accessed January 7, 2014).

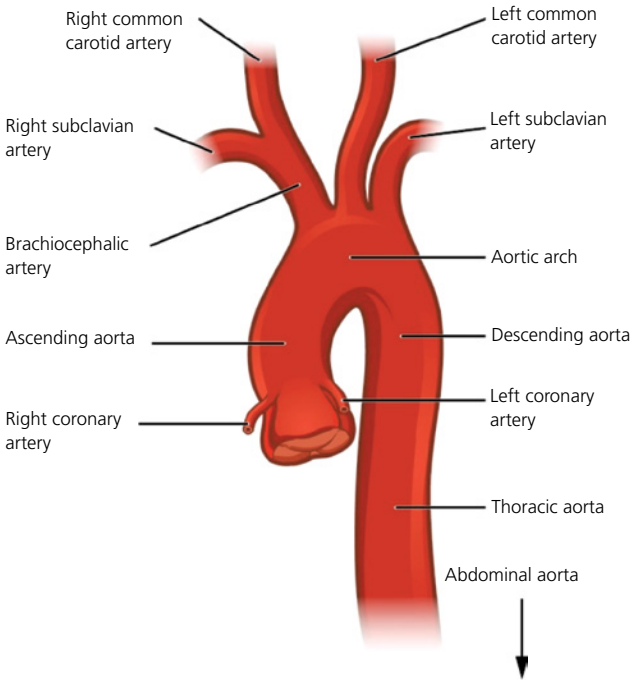


Figure 1.16 Aortic arch anatomy. http://cnx.org/content/m46646/latest/2121_Aorta.jpg. Reproduced with permission from OpenStax College on Wikimedia Commons under the Creative Commons Attribution-Share Alike 3.0. Unported license (accessed January 7, 2014).

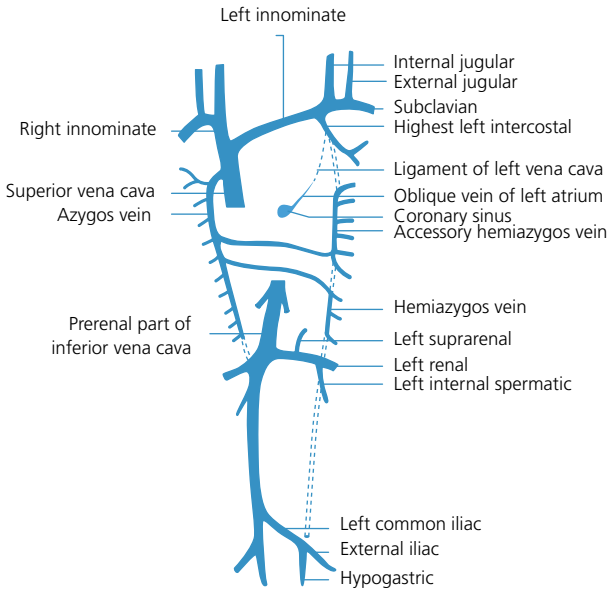


Figure 1.17 Central venous anatomy. Reproduced with permission from Gray's Anatomy of the Human Body, 1918 edition (http://en.wikipedia.org/wiki/File:Gray480.png) via Wikipedia.

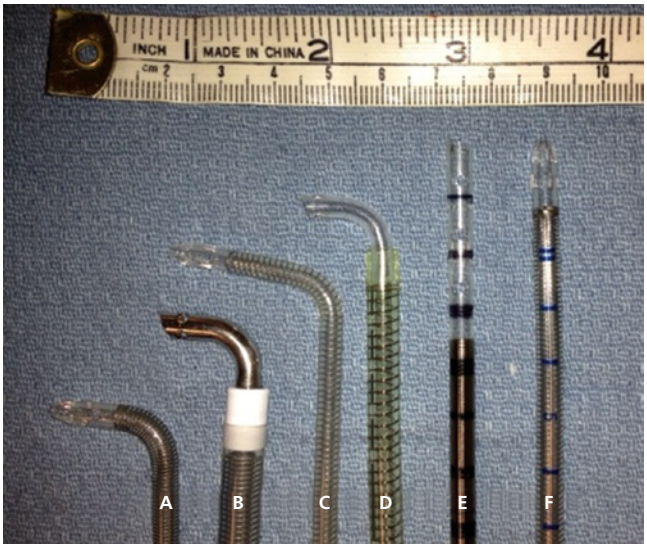


Figure 1.18 Select pediatric venous cannulae. *A—14 Fr. Terumo Tenderflow right angle PVC tip. B—14 Fr. Medtronic DLP right angle metal tip. C—12 Fr. Medtronic DLP right angle PVC tip. D—12 Fr. Edwards Lifesciences Thin-Flex right angle plastic tip. E—14 Fr. Medtronic Bio-Medicus straight with multiple side port holes. F—14 Fr. Medtronic DLP malleable straight PVC tip.*

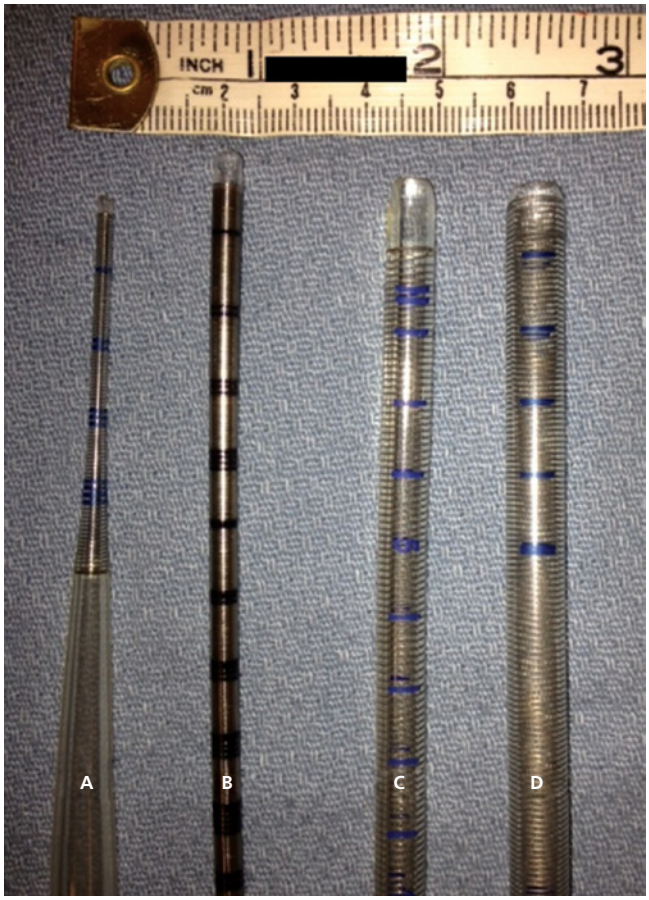


Figure 1.19 Select arterial cannulae for central cannulation. *A-6 Fr. Medtronic DLP One Piece. B-12 Fr. Medtronic Bio-Medicus. C-20 Fr. Medtronic EOPA. D-22 Fr. Medtronic EOPA CAP.*

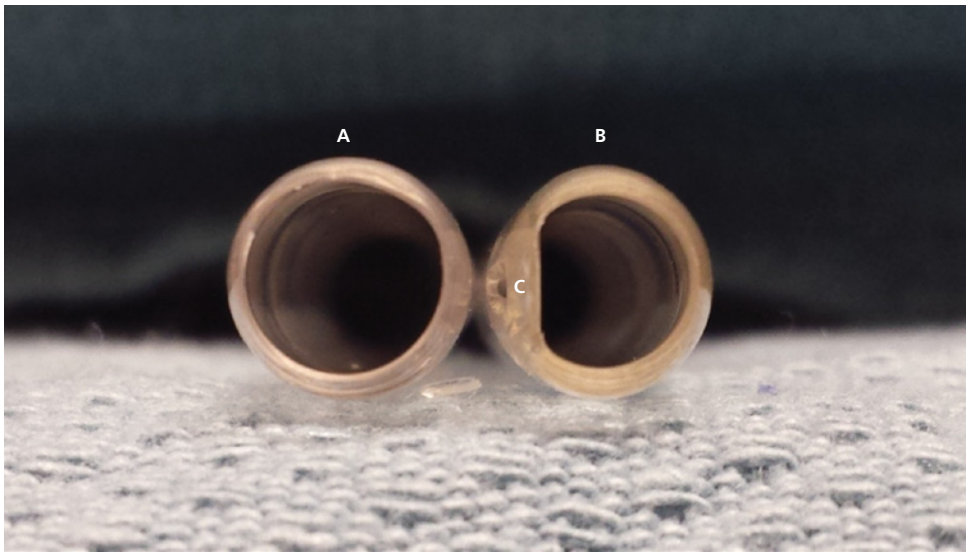


Figure 1.20 Luminal variation between types of arterial cannulae. *A-Standard round lumen for a 22 Fr. Medtronic arterial cannula. B-D-shaped lumen for a 22 Fr. Medtronic arterial cannula with central aortic pressure (CAP) monitoring capability. C-Left of "C" is the central aortic pressure monitoring port.*

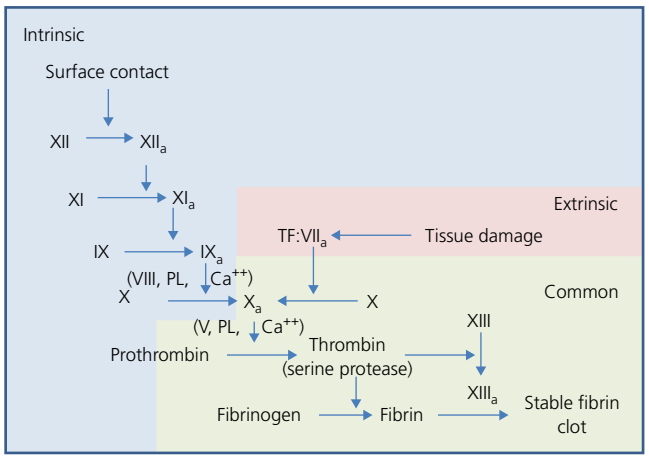


Figure 3.1 Classical description of the blood coagulation pathways. VII-stabile factor, XII-Hageman factor, IX-Christmas factor, XI-plasma thromboplastin, XIII-fibrin stabilizing factor, Ca++ ionized calcium, PL-platelets, TF-tissue factor. http://commons.wikimedia.org/wiki/File:Classical_blood_coagulation_pathway.png. Reproduced with permission from Graham Beards and Wikimedia Commons.

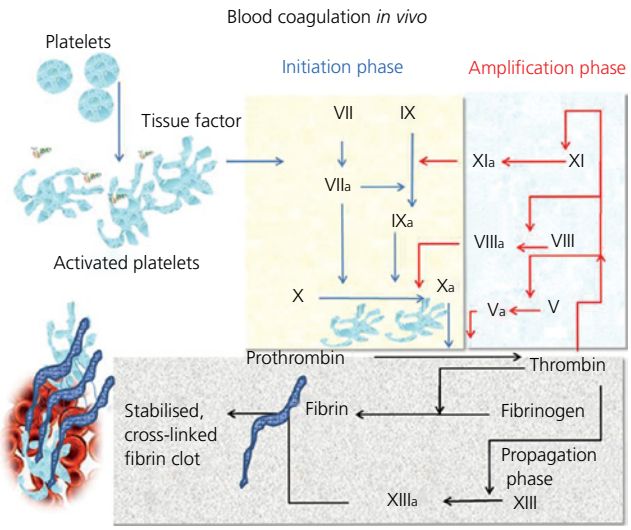


Figure 3.2 Cell-based model of hemostasis *in vivo*. http://commons.wikimedia.org/wiki/File:Coagulation_in_vivo.png. Reproduced with permission from Graham Beards and Wikimedia Commons.

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