Management of the postoperative pediatric cardiac surgical patient

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Objective: To review the salient aspects and latest advances in the management of the postoperative pediatric cardiac patient.

Data Source: A Medline-based literature source.

Conclusion: The practice of pediatric cardiac intensive care has evolved considerably over the last several years. These efforts are the result of a collaborative effort from all subspecialties involved in the care of pediatric patients with congenital heart disease. Discoveries and innovations that are representative of this effort include the extension of cerebral oximetry from the operating room into the critical care setting; mechanical circulatory devices designed for pediatric patients; and surgery in very low birth weight neonates. Advances such as these impact postoperative management and make the field of pediatric cardiac intensive care an exciting, demanding, and evolving discipline, necessitating the ongoing commitment of various disciplines to pursue a greater understanding of disease processes and how to best go about treating them. (Crit Care Med 2011; 39:1974–1984)

Key Words: pediatric cardiac surgery; postoperative management; cardiopulmonary bypass; hemodynamic monitoring; ventricular dysfunction; cardiopulmonary interaction; vasoactive support

The field of pediatric cardiac intensive care continues to evolve, which is in large part the result of collaborative efforts from anesthesia, surgery, cardiology, critical care, and other subspecialties, including neonatology, neurology, and endocrinology. Examples include an increasing number of surgeries in very low birth weight infants; the extension of technology such as cerebral oximetry from the operating room into the intensive care setting; and innovations in mechanical circulatory devices. Industry-sponsored studies and initiatives from the National Institutes of Health such as the Pediatric Heart Network have contributed to the evolution of pediatric cardiac intensive care. These collective efforts are demonstrated in the Prophylactic Intravenous Use of Milrinone After Cardiac Operation in Pediatrics (PRIMACORP) study and more recently the single ventricle reconstruction trial and thyroid supplementation study (to be discussed).

The increase in complexity of disease, innovations in technology, and evolving therapeutic strategies as well as national quality initiatives, individually and collectively, place a tremendous demand on the team, necessitating a focused, concerted effort by all members to challenge current practices while practicing state-of-the-art care. In this review, we discuss the salient aspects and latest advances in the primary challenge after surgery, which is to maintain adequate cardiopulmonary function and to ensure adequate tissue oxygenation.

The Inflammatory Response to Cardiopulmonary Bypass

Exposure of the blood components to the nonendothelialized circuit, pulmonary and myocardial reperfusion injury, and the formation of heparin–protamine complexes stimulate the release of systemic and local proinflammatory mediators and activate the coagulation/fibrinolytic pathways (1). Virtually every inflammatory pathway and parenchymal cell contributes to the inflammatory cascade. The magnitude of the inflammatory response to cardiopulmonary bypass (CPB) has been associated with preoperative conditions such as shock and heart failure (2); intraoperative factors such as the duration of bypass and cardioplegic arrest (3); and genetic variances have been identified that affect the inflammatory response to several stimuli, including CPB (4, 5).

Several studies have found a relationship between the inflammatory response to bypass and the development of multi-organ dysfunction and postoperative morbidity (3, 6). Prospective, randomized studies in children found the administration of glucocorticoids before bypass significantly reduced the proinflammatory and augmented the compensatory anti-inflammatory responses to bypass, reduced the extent of myocardial injury, and improved the postoperative course (7–10). Two prospective studies failed to demonstrate benefit from the use of glucocorticoids however they limited enrollment to older children (11, 12). A majority of pediatric cardiac centers use glucocorticoids to ameliorate the inflammatory response to bypass (13).

Assessment of Hemodynamics and Tissue Oxygenation

An accurate and timely determination of cardiac output (CO), systemic O₂ delivery (DO₂), and tissue oxygenation is essential to optimizing outcomes. Studies have demonstrated that it is not uncommon for there to be discordance between measurements of these parameters and estimations based on the physical examination and interpretation of conventional hemodynamic parameters such as heart rate, blood pressure, and right atrial pressure (14, 15). Compensatory increases in systemic vascular resistance maintain arterial blood pressure as CO falls and central venous/right atrial pressures may not correlate well with right
ventricular filling or left atrial pressures (16, 17). The primary determinants of ventricular filling are the ventricular diastolic transmural pressure (ventricular diastolic pressure – surrounding pressure) and ventricular compliance (Fig. 1). With pericardial disease, increases in intrathoracic pressure, and impaired diastolic function, the optimal filling pressure increases.

Echocardiography provides an assessment of ventricular filling and function and quantification of valvular regurgitation. Doppler measurements assess obstructions and pulmonary arterial pressures. Trending serial lactate levels has prognostic value (18, 19). A low serum HCO3− level and base deficit are reflective of tissue hypoxia to the extent that serum lactate levels are elevated, producing an anion gap; otherwise, these abnormalities are consistent with a hyperchloremic metabolic acidosis (20–23).

The routine placement of central venous catheters readily enables the clinician to use venous oximetry to assess oxygen transport balance (Table 1). The critical O2 extraction ratio is defined by the onset of shock and ranges from 50% to 60% (Table 2) (24). The critical DO2 changes in parallel with changes in oxygen consumption (VO2); however, the critical O2 extraction ratio remains constant, which is an important consideration in the postoperative period when oxygen demand changes over time and with therapy (25). When mixed venous oxygen saturations are unavailable, alternative sampling sites include the right atrium, superior vena cava, jugular vein, and inferior vena cava–right atrial junction (Table 2) (26–28). As CO falls, blood flow is redistributed to maintain vital organ perfusion. As a result, inferior vena cava saturations fall first, which is accompanied by a less severe drop in mixed venous oxygen saturations. As CO falls further, superior vena cava and jugular saturations begin to decrease (28, 29). Venous oximetry has been shown to improve outcomes in pediatric patients at risk for developing shock, including severe sepsis and the hypoplastic left heart syndrome (30, 31).

Cerebral near-infrared spectroscopy noninvasively assesses cerebral oxygenation. Cerebral oximetry relies on the relative transparency of biological tissue to near-infrared spectroscopy light where oxy- and deoxy hemoglobin have distinct absorption spectra (32). The oximeter monitors the nonpulsatile signal reflecting the microcirculation where 75–85% of the blood volume is venous. Thus, the O2 saturation is used as an indicator of O2 extraction for the area of the brain immediately beneath the probe (frontal cortex). Because of technical constraints, the technology is limited to relative quantitation and is thus useful for tracking changes for a given patient. Studies have demonstrated a good correlation between cerebral O2 saturations and jugular bulb and superior vena cava saturations (33, 34). A study by Li and colleagues (35) evaluated the relationship between cerebral oxygenation and hemodynamic and oximetric parameters in neonates following the Norwood procedure. They found that cerebral oxygen saturations closely and negatively correlated with the O2 extraction ratio. Cerebral oximetry is also used as a surrogate for central and mixed venous oxygen saturations; however, the correlation between these readings is marginal (36, 37). The reasons for this include the technical constraints described; the fact that in a low CO state, blood flow is redistributed to maintain perfusion of vital organs; and cerebral blood flow is sensitive to changes in arterial PCO2.

**Maintenance of Tissue Oxygenation**

The primary goal after surgery is to maintain adequate tissue oxygenation, which is accomplished by optimizing oxygen transport balance (i.e., minimizing oxygen demand and consumption and maintaining adequate oxygen delivery (Table 3) (38). Underlying respiratory and circulatory dysfunction, the inflamma-
Infant cardiovascular reserve

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Increased circulatory demands due to greater respiratory load
Neuromuscular competence

Table 4. Factors responsible for diminished infant respiratory reserve

<table>
<thead>
<tr>
<th>Decreased functional residual capacity</th>
<th>Decreased subglottic cross-sectional area</th>
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<tr>
<td>Decreased ventilatory reserve</td>
<td>Highly compliant chest wall</td>
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<tr>
<td>Less diaphragmatic contractile reserve</td>
<td>Increased respiratory demand</td>
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<td>Higher O2 consumption and minute ventilation</td>
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Table 5. Impaired respiratory mechanics

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<tr>
<th>Upper airway dysfunction</th>
<th>Postextubation group/subglottic edema</th>
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<tr>
<td>Neurologic dysfunction</td>
<td>Injury to the recurrent laryngeal nerve</td>
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<tr>
<td>Central nervous system injury</td>
<td>Excessive sedation/analgesia</td>
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<tr>
<td>Lower airway dysfunction</td>
<td>Bronchiolar compression due to interstitial edema from left–right cardiac shunting, cardiopulmonary bypass and pulmonary venous hypertension</td>
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<tr>
<td>Parenchymal lung disease</td>
<td>Atelectasis</td>
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<td>Edema (hydrostatic and permeability)</td>
<td>Pneumonia</td>
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<td>Chest wall edema</td>
<td>Pleural effusion</td>
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<td>Abdominal load increased (visceromegaly, ascites)</td>
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Table 6. Ventilatory failure

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<tr>
<th>Neuromuscular competence</th>
<th>Central nervous system depression</th>
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<tr>
<td>Diaphragmatic paralysis</td>
<td>Disuse atrophy (compounded by muscle relaxants and glucocorticoids)</td>
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<td>Malnutrition</td>
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<td>Respiratory load</td>
<td>Compliance of lung/chest wall</td>
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<td></td>
<td>Resistive load</td>
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<td>Respiratory muscle energetics</td>
<td>Muscle oxygen consumption</td>
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<td></td>
<td>Respiratory muscle perfusion</td>
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<td>Cardiac output state</td>
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Table 7. Infant cardiovascular reserve

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<tr>
<th>Decrease contractile reserve</th>
<th>Cardiomyocyte contains fewer and poorly organized contractile proteins</th>
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<tr>
<td>Immature sarcoplasmic reticulum</td>
<td>Decrease myocardial compliance</td>
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<tr>
<td>Increase circulatory demands due to greater oxygen consumption</td>
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Respiratory Dysfunction

Infants in particular are at risk for developing respiratory insufficiency after cardiac surgery as a result of diminished respiratory reserve (Table 4) (41). Respiratory mechanics are impaired to varying degrees (Table 5). The inflammatory response to CPB increases vascular permeability and extravascular lung water, leading to interstitial and alveolar edema and impaired surfactant function (42). As a result, lung compliance decreases and if an adequate end-expiratory lung volume is not maintained, pulmonary venous admixture and arterial hypoxemia result. Ventilatory failure after cardiac surgery may result from a number of causes (Table 6), some unique to patients undergoing repair of structural heart disease. Ventilatory failure results from a combination of increased respiratory load, decreased neuromuscular competency, and impaired respiratory muscle energetics (i.e., V̇O2/ḊO2 relationship) (Table 6) and is manifested by hypercapnia with a normal arterial to end tidal CO2 gradient (43, 44). An additional cause of impaired gas exchange is low CO and abnormally distributed pulmonary blood flow, which produce an increase in ventilation-perfusion ratios, wasted ventilation, and hypercapnia with an elevated arterial to end-tidal CO2 gradient (45–48).

Cardiovascular Dysfunction

There is invariably some degree of postoperative myocardial diastolic and systolic dysfunction, in part the result of the inflammatory response and ischemia–reperfusion injury (49, 50). Improving CO involves optimizing ventricular loading conditions, myocardial conduction, and ventricular performance. These challenges are even greater in neonates as a result of their diminished cardiovascular reserve (Table 7) (51, 52).

Systolic Dysfunction

Nitric Oxide Donors. The primary strategy for treating systolic dysfunction is to optimize ventricular loading conditions. Nitroglycerin in low to moderate doses (<3 μg/kg/min) increases venous capacitance without much effect on the arterial resistance vessels (64). As a result, ventricular filling pressures fall, whereas stroke volume is unaffected. High-dose nitroglycerin and nitroprusside increase venous capacitance and reduce systemic vascular resistance and thus filling pressures fall and stroke volume and CO rise significantly (65).

Natriuretic Peptides. The natriuretic peptides act primarily as counterregulatory hormones to the renin–angiotensin–aldosterone system. Atrial and B-type natriuretic peptides are produced primarily by myocardium in response to chamber wall stress (66). Stimulation of natriuretic peptide receptors causes vasodilation of venous capacitance and arterial resistance vessels, a dose-dependent natriuresis/diuresis, and improved ventricular relaxation. Nesiritide (Natrecor, Scios, Inc., Fremont, CA) is the human recombinant form of B-type natriuretic peptide. In contrast to nitroglycerin and
milrinone, tachyphylaxis does develop to the hemodynamic effects of nesiritide (67) and its use is not associated with reflex stimulation of systemic and cardiac adrenergic activity (68).

Studies in adults have demonstrated its efficacy in treating congestive heart failure, whereas studies in children have been limited (69). A study by Jefferies et al (70) evaluated the safety and efficacy of nesiritide in pediatric decompensated heart failure by prospectively monitoring 55 separate infusions in 32 patients. They found results similar to those found in adults: a significant increase in diuresis; reduction in ventricular filling pressures and increase in CO; and a marked improvement in the mean New York Heart Association functional class. No hypotension or arrhythmias were noted during 478 cumulative days of therapy and serum creatinine levels trended downward after therapy.

Catecholamines. The use of catecholamines is limited because they are chronotropic, arrhythmogenic, and increase global and myocardial VO$_2$ (68). Dopamine and dobutamine provide modest inotropic support. In contrast to dobutamine, dopamine decreases venous capacitance and as a result ventricular filling pressures do not decrease (72, 73). At higher doses (>10 µg/kg/min), systemic vascular resistance begins to increase as a result of β-agonist activity. Dobutamine decreases systemic vascular resistance, which is thought to be the result of β$_2$ agonist activity. Epinephrine provides unparalleled inotropic support and in low doses (<0.05–0.10 µg/kg/min) reduces systemic vascular resistance (74, 75).

Phosphodiesterase Inhibitors. Milrinone, and its predecessor amrinone, act through selective inhibition of phosphodiesterase III. The advantages of milrinone over catecholamines are that it is not chronotropic and arrhythmogenic, it does not increase myocardial VO$_2$, and it is unaffected by adrenergic receptor desensitization. Milrinone provides modest inotropic support, vasodilates pulmonary and systemic vessels, and exerts a lusitropic effect (76). As a result, ventricular filling pressures decrease while stroke volume and CO increase (77). A study by Hoffman and colleagues (78) evaluated the efficacy and safety of prophylactic milrinone in pediatric patients undergoing cardiac surgery. Patients (n = 238) were randomized to low-dose (25 µg/kg/min) or high-dose (75 µg/kg/min) milrinone or placebo. High-dose milrinone significantly reduced the risk of developing low CO syndrome compared with placebo, a 64% relative risk reduction, and there was no significant difference in the incidence of hypotension or arrhythmia compared with placebo.

Levosimendan. Levosimendan represents the first of a new class of agents, the calcium sensitizers, which are in use in Europe (79). These agents provide modest inotropic support by enhancing the sensitivity of the myofilaments to the prevailing cytosolic calcium concentration. Because less energy is consumed in the cycling of calcium, myocardial VO$_2$ remains unchanged. By stimulating adenosine 5-triphosphate-dependent K+ channels in vascular smooth muscle cells, levosimendan decreases systemic and pulmonary vascular resistance. In contrast to other vasoactive agents, levosimendan has a long duration of action as a result of the generation of active metabolites with an elimination half-life of 3–4 days.

Namachivayam and colleagues evaluated levosimendan in 15 children with severe ventricular dysfunction who were catecholamine-dependent (80). A combination of a loading dose and continuous infusions (24–48 hrs) allowed for a substantial reduction in catecholamine infusions with discontinuation in a majority of patients. A prospective randomized trial was performed by Momeni and colleagues (81) of children (n = 36) undergoing cardiac surgery in which patients received either milrinone or levosimendan with initiation of CPB for up to 48 hrs. There was no difference between groups in terms of serum lactate levels (primary aim) or oxygen extraction; however, the rate pressure index was significantly greater in those receiving milrinone.

Cardiopulmonary Interaction. In systemic heart failure, PPV improves CO. As discussed, PPV may compromise ventricular filling (82). However, so long as the dilated ventricle resides on the flat portion of its pressure volume curve, a decrease in venous return will not affect stroke volume. PPV reduces systemic venous afterload and by unloading the respiratory pump respiratory muscle VO$_2$ decreases, allowing for a redistribution of a limited CO (83, 84).

Other Strategies for Improving Circulatory Function

Arginine Vasopressin. Arginine vasopressin (AVP) plays a vital role in the defense of arterial blood pressure, because physiological levels are required for normal vascular tone of venous capacitance and arterial resistance vessels. The rationale behind the use of exogenous AVP in critical illness is that neurohypothesis stores of AVP may become exhausted, leading to vasodilatory shock (85). Prospective, randomized studies in adults have yielded equivocal results (86, 87). There have been a few reports on the use of AVP in children (88, 89). A retrospective analysis was performed by Rosenzweig et al of their experience with AVP in 11 children after cardiac surgery. All had refractory hypotension and all but two had normal or mildly depressed left ventricular function. During the first hour of AVP, there was a significant improvement in blood pressure and the inotropic score.

Glucocorticoids. A similar rationale exists for the use of glucocorticoids (90). Cortisol is essential for maintaining vascular tone and may affect myocardial function (91). Particularly in premature and stressed term neonates, a relative adrenal insufficiency may develop as a result of the effects of inflammation on an immature hypothalamus–pituitary axis (92). Although the definition of a relative adrenal insufficiency remains unclear, most would agree that a normal or near-normal cortisol level in a critically ill patient is indicative of an inadequate hypothalamus–pituitary axis response (93). Prospective randomized studies of critically ill premature infants with refractory hypotension demonstrated improved hemodynamics in those treated with glucocorticoids (94, 95). Studies in children with refractory hypotension after cardiac surgery have been limited to several retrospective studies and one small prospective randomized study. All studies demonstrated a significant improvement in arterial blood pressure over the first several hours after the initiation of therapy that coincided with a weaning of vasoactive support (96–99).

Thyroid Hormone. After CPB, serum thyroid hormone levels are depressed (100). Because of the known effects of thyroid hormone on ventricular function, several studies have evaluated the role of thyroid replacement therapy in children after cardiac surgery (101–104). A rela-
tively large prospective randomized study (n = 193) was performed by Portman and colleagues of T3 supplementation in children undergoing cardiac surgery. Analysis using age stratification found those patients <5 months of age who received T3 experienced a significantly shortened time to extubation (primary end point), improved cardiac function, and required less vasoactive support. A study by Mackie and colleagues randomized 42 infants undergoing the Norwood procedure or repair of interrupted aortic arch to T3 supplementation or placebo. There was no difference in the primary end points of clinical outcome score and CO between groups; however, those receiving therapy did have a significantly higher systolic blood pressure. A study by Chowdhury and colleagues demonstrated an improved postoperative course including reduced need for vasoactive support in children (n = 28) randomized to T3 as did the study by Bettendorf and colleagues that demonstrated an improved postoperative course and systolic function in those children (n = 40) randomized to treatment.

**Postoperative Management: Lesion-Specific**

*Single Ventricle Physiology.* Patients with single ventricle physiology are at risk for developing shock, the result of systolic dysfunction compounded by the inefficiencies of a parallel circulation. The aortic-pulmonary artery shunt allows for continuous flow throughout the cardiac cycle, leading to excessive pulmonary perfusion (Qp), which occurs at the expense of systemic perfusion (Qs), diastolic hypotension, and potentially inadequate coronary perfusion. The distribution of total CO (Qp + Qs) is determined by the systemic to pulmonary vascular resistance ratio, which is much greater than one. Pulmonary overcirculation increases as this ratio increases; as Qs wanes, neurohormonal systems are activated, which further increases the systemic to pulmonary vascular resistance ratio, and a vicious cycle ensues culminating in shock.

Management begins with an accurate assessment of Qs. Studies have demonstrated the limitations of inferring Qs and systemic DO2 from arterial oxygen saturations and studies have found improved outcomes using venous oximetry from the superior vena cava (31, 105). The focus on postoperative management is to not increase pulmonary vascular resistance but rather to lower systemic vascular resistance, which improves stroke volume, Qs, and Qp, producing a marked increase in systemic DO2 (106–108). Milrinone is an ideal agent for this strategy. Low-dose epinephrine (<0.05–0.10 μg/kg/min) provides greater inotropic support while reducing systemic vascular resistance (109). Dopamine provides modest inotropic support but does not reduce systemic vascular resistance and has been shown to worsen oxygen transport balance by increasing VO2 without an appreciable effect on systemic DO2 (110, 111).

An alternative strategy for the initial palliation of the hypoplastic left heart syndrome is the use of a right ventricle to pulmonary artery conduit. Because shunting occurs only during systole, the Qp/Qs ratio tends to be lower and the diastolic blood pressure higher compared with the standard approach (112, 113). The single ventricle reconstruction trial prospectively randomized (n = 549) infants with the hypoplastic left heart syndrome or a related single, morphologic right ventricular anomaly and a planned Norwood procedure to one of these two strategies (114). The rate of transplantation-free survival at 12 months was significantly better in those infants receiving the right ventricular to pulmonary artery conduit (74 vs. 64%, p = .01); however, after 12 months, there was no difference in transplantation-free survival between groups (p = .06). The rate of composite “serious adverse events” (death, acute shunt failure, cardiac arrest, extracorporeal membrane oxygenation, or necrotizing enterocolitis) during the Norwood hospitalization was significantly lower in the right ventricle to pulmonary artery conduit group (36 vs. 48%, p = .02).

**Bidirectional Glenn.** The primary challenge after the bidirectional Glenn is hypoxemia. This may be the result of pulmonary venous admixture, systemic venous collaterals, pulmonary arteriovenous malformations, or inadequate Qp (115–116). One strategy to improve Qp and oxygenation is permissive hyperventilation, which uncouples cerebral blood flow from cerebral metabolism allowing for an increase in superior vena cava flow and Qp (118). A study by Hoskote et al (119) demonstrated that progressive increases in Paco2 led to increases in Qp and oxygenation without increasing pulmonary vascular resistance. These changes were accompanied by a decrease in systemic vascular resistance and increase in Qs and a marked increase in DO2. In the absence of systemic hypotension, manipulation of systemic blood pressure will not affect cerebral blood flow and oxygenation because cerebral pressure autoregulation is intact (120).

**Fontan Procedure.** The primary challenge after the Fontan procedure is a low CO state, which results from inadequate ventricular volume. Diastolic dysfunction is present to varying degrees. In contrast to the bidirectional Glenn, all systemic venous return must overcome the resistance of the pulmonary circulation without a subpulmonic pumping chamber, which further compromises ventricular filling (60, 61, 121). Inotropic support provides marginal support as a result of limited venous return and ventricular filling (54). Negative pressure ventilation improves ventricular filling and CO by increasing venous return and the ventricular diastolic transmural pressure and is in part the rationale for an early extubation after surgery (Fig. 1) (122, 123). Volume administration increases the upstream driving pressure for venous return, whereas venodilators increase venous capacitance compromising venous return (59). Even modest increases in pulmonary vascular resistance may not be tolerated and normalizing the functional residual capacity, pH, and alveolar PO2 may improve Qp (124); some patients may respond to inhaled nitric oxide (125).

Hypoxemia may also complicate the postoperative course and may be the result of systemic venous collaterals (126). Transudative and chylos pleural and pericardial effusions are not uncommon after the Fontan procedure and may result from traumatic injury to a lymphatic tributary or elevated systemic or pulmonary venous pressure impeding lymphatic drainage (127, 128). Pleural fluid accumulation may also be the result of systemic–pulmonary artery collaterals (129). Any space-occupying thoracic lesion will increase intrathoracic pressure and compromise the gradient for venous return; a pericardial effusion will further reduce the ventricular diastolic transmural pressure; either lesion will further compromise an already under filled ventricle.

**Tetralogy of Fallot.** After repair of tetralogy of Fallot, a low CO state may develop as a result of right ventricular diastolic failure (130); this most likely
results from exposure of the hypertrophied right ventricle to CPB and reperfusion injury. Because systolic function is intact, inotropic agents are of little benefit (55, 56). Low CO output results from inadequate right ventricular filling, which is compounded by a reduction in the effective compliance of the left ventricle. During diastole, the hypertensive right ventricle alters the normal transpapillary gradient, causing the ventricular septum to bow into the left ventricle (130, 131). As discussed, a fall in intrathoracic pressure as occurs with the use of a cuirass or after extubation has been shown to improve hemodynamics and tissue oxygenation (132, 133). Invariably some degree of right bundle branch block is present after surgery and cardiac resynchronization therapy has been shown to improve hemodynamics in some patients (134–136).

Mechanical Circulatory Support. Mechanical circulatory support devices may be used as a bridge to recovery, like in fulminant myocarditis and the postcardiotomy syndrome, or as a bridge to transplantation. The mainstay of short-term pediatric mechanical circulatory support has been and remains extracorporeal membrane oxygenation and the centrifugal ventricular assist device (VAD). By and large, long-term mechanical circulatory support in pediatrics has relied on extracorporeal membrane oxygenation with a 47% to 57% success rate as a bridge to transplantation (134, 135). Over the last several years, several long-term VADs have been designed for or applied to the pediatric patient, leading to improved outcomes. A study of the Pediatric Heart Transplant Study database found a survival to transplantation rate of 77% using VADs from 1993–2003 with an even higher success rate from 2000–2003 of 86% (139). Despite improved overall outcomes, the success rate is significantly lower in patients with congenital heart disease and in the smaller, younger patients.

The Berlin Heart Excor (Berlin GmbH, Berlin, Germany) is the first commercially available VAD designed for the pediatric population (Fig. 2). It has specialized cannulae and a miniaturized pneumatically driven pump that produces pulsatile flow. The device is suitable for patients with a body weight of >2.5 kg and may be implanted as a bilateral VAD, left VAD, or right VAD. The Berlin Excor has been in use in Europe since 1992 and recently was approved by the Food and Drug Administration under a limited investigational device exemption. A prospective multicentered trial is underway in the United States as part of an effort to obtain Food and Drug Administration approval. The MEDOS HIA VAD (MEDOS Medizintechnik GmbH, Stolberg, Germany) is a paracorporeal pneumatically driven pump that produces pulsatile flow. The device is approved for patients with a body surface area of 0.7–1.5 m². The pediatric experience has been limited.

Pulmonary Hypertension. Patients with cardiac disease are prone to developing pulmonary hypertension, which may be exacerbated by the adverse effects of CPB and reperfusion injury on pulmonary vascular, respiratory, and right ventricular function (141, 142). Initial therapies include normalization of blood pH; correction of alveolar hypoxia with titration of end-expiratory pressure to achieve a normal functional residual capacity; and right ventricular inotropy. Pharmacologic strategies to reduce pulmonary vascular resistance are included in Figure 3. Nonselective vasodilators may precipitate systemic hypotension if systemic vascular resistance falls to a much greater extent than pulmonary vascular resistance and may impair oxygenation by releasing hypoxic pulmonary vasoconstriction.

Inhaled nitric oxide (iNO) is highly effective at reducing pulmonary vascular resistance and its effects are limited to the pulmonary circulation (144). Too rapid a discontinuation of iNO may lead to downregulation of endogenous iNO and rebound pulmonary hypertension, which may be ameliorated with the use of oral sildenafil (145, 146). The use of iNO in left-sided obstructive lesions such as mitral stenosis and “obstructed” total anomalous pulmonary venous return leads to an increase in pulmonary venous pressure and may precipitate pulmonary edema without improving CO (147, 148). In patients with pulmonary hypertension secondary to left ventricular systolic heart failure, the reduction of right ventricular afterload with iNO exacerbates
left ventricular congestive heart failure if the left ventricle is not unloaded (149). A study by Argenziano and colleagues (150) demonstrated the utility of iNO in adult patients with severe left ventricular failure whose left ventricle was unloaded with a left VAD but whose flow rate was limited by elevated pulmonary vascular resistance. The use of iNO in this setting allowed for a significant increase in left VAD flow.

Arrhythmia. After cardiac surgery, temporary epicardial pacing wires are frequently attached to the right atrium and right ventricle or single ventricle enabling atrial, atrioventricular sequential, and ventricular pacing. Cardiac resynchronization therapy after surgery is a relatively new strategy to improve cardiac function. Cardiac resynchronization therapy involves nonconventional pacing strategies targeting prolonged atrioventricular and intraventricular conduction, which may improve ventricular filling and lessen the degree of discoordinate ventricular contraction (151, 152).

Several studies have evaluated cardiac resynchronization therapy after pediatric cardiac surgery. A study by Zimmerman and colleagues (153) demonstrated significant improvements in hemodynamics and CO by using multisite ventricular pacing after pediatric cardiac surgery in 29 patients with single and biventricular anatomy and prolonged QRS duration. In a follow-up study from the same center, 26 single ventricle patients regardless of electrocardiographic criteria demonstrated improved hemodynamics and CO with multisite ventricular pacing (154). A study by Janousek and colleagues (135, 136) demonstrated improved blood pressure using atrial synchronous right ventricle and biventricular pacing for primarily atrioventricular and intraventricular conduction delay after pediatric cardiac surgery. A study by Jeewa et al (155) evaluated cardiac resynchronization therapy in children undergoing biventricular repair of congenital heart disease. Patients were atrioventricular paced using right ventricle or biventricular pacing. There was no hemodynamic improvement using either strategy. A similar study conducted by Pham et al (156) demonstrated improved CO during biventricular pacing but not during conventional atrioventricular pacing. The QRS duration for each study was not prolonged. Although there are variations between these studies, including the underlying cardiac lesion, baseline conduction, and pacing strategies, the longer the baseline QRS duration, the more likely cardiac resynchronization therapy will be hemodynamically beneficial and a normal QRS duration does predict lack of benefit.

Summary

The practice of pediatric cardiac intensive care has evolved considerably over the last several years. Exciting advances are being made in other areas of perioperative care, which directly or indirectly impact postoperative management and outcomes. Industry-sponsored studies and the advent of the Pediatric Heart Network of the National Institutes of Health will contribute to the growth of the field by conducting randomized controlled studies. Discoveries and innovations in imaging, genomics, and mechanical circulatory devices are occurring; refinements in CPB are taking place, which target myocardial reperfusion injury, the inflammatory response to bypass, and neurologic sequelae. Some centers are bridging the catheterization laboratory and the operating room by exploring hybrid techniques. Fetal interventions and surgery in very low birth weight neonates are providing additional strategies for dealing with the most challenging lesions, whereas an ever-increasing population of adults with congenital heart disease is emerging, necessitating the development of a new subspecialty. The field of pediatric cardiac intensive care is an exciting, demanding, and evolving discipline, necessitating the ongoing commitment of various disciplines to pursue a greater understanding of disease processes and how to best go about treating them.

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