In the past, many children born with complex congenital heart defects (CHDs) underwent palliative procedures to avoid the use of cardiopulmonary bypass, which typically is required for prolonged corrective procedures. The risk of mortality was prohibitive, especially in newborns; therefore, efforts were directed toward palliative procedures until the children grew big enough for a definitive repair. These children often were left with abnormal hemodynamics until the final complete repair at a later age.

However, the disappointing cumulative rates of morbidity and mortality for palliative operations for CHD (followed by subsequent corrective surgery at a later age) compared with early corrective procedures have become apparent during the past decade. As a result, there has been a recent trend toward early anatomic repairs of complex CHDs. Primary corrective surgery has had a significant impact on both the risk of mortality from the underlying defect and on the secondary effects of the CHD on other organ systems. This impact has been the most dramatic in neonates. Advances in our understanding of physiology and in technical skill have enabled us to consider performing reparative operations using cardiopulmonary bypass on even premature, low-birth-weight newborns.

**Neonatal Physiology and Transitional Circulation**

Care of the critically ill newborn requires an appreciation of the special structural and functional features of immature organs, the interactions of the “transitional” neonatal circulation, and the secondary effects of the CHD on other organ systems. Newborns respond more quickly and profoundly to physiologically stressful circumstances. This often results in rapid changes in pH, levels of lactate acid and blood glucose, as well as temperature and oxygen content.

Newborns have diminished fat and carbohydrate reserves as well as a higher metabolic rate and oxygen consumption. Newborns also have increased total body water compared with older children, and a higher propensity to leak fluid out of the capillary system into the interstitial space. This is especially prominent in the lungs (i.e., pulmonary edema), where the pulmonary vascular bed is nearly fully recruited at rest and lymphatic recruitment, which is required to handle increased mean capillary pressures resulting from increases in pulmonary blood flow, may not be available.

**Recent Technical Innovations Have Allowed for Drastic Reductions in Mortality Rates After Neonatal Open-Heart Surgery.**

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NEONATAL CARDIOVASCULAR PHYSIOLOGY
The newborn may be more likely to maintain blood pressure (because of the wide range in systemic vascular resistance due to the elasticity of peripheral vessels) when there exists a state of impending shock, which may lure the practitioner into a false sense of security immediately prior to circulatory collapse. Systemic blood pressure is not always a reliable indicator of adequacy of intravascular volume status (preload), cardiac output, or satisfactory oxygen delivery. The neonatal myocardium is less compliant than in the older child, less tolerant of increases in afterload, and less responsive to increases in preload (Figure 1).

The potential for sustained or labile increases in pulmonary vascular resistance is common in newborns. Concerns over inciting pulmonary hypertensive events has deterred some from pursuing a reparative approach in these patients. Finally, the extreme stress reactions demonstrated in response to cardiopulmonary bypass must be considered in the overall approach to the treatment of these patients.

These factors do not preclude intervention in the newborn but simply dictate that extraordinary vigilance must be applied in the care of these children, and that treatment plans that take into account their immature physiology must be developed.

Whereas the newborn may be more labile than the older child, there also is ample evidence that this age group is more resilient after metabolic or ischemic injury. In fact, the newborn may be particularly able to cope with some forms of stress such as ischemia or hypoxia. Tolerance of hypoxia in the newborn, specifically in the neurological system, is characteristic of many animal species and well described in the newborn human. For example, newborns with obstructive lesions on the left side of the heart frequently present with profound metabolic acidosis and shock (e.g., from impairment of blood flow to the lower body in a newborn with aortic coarctation). These children can be effectively resuscitated without persistent organ-system damage.

Neonatal CHD repair may prevent irreversible secondary organ damage that arises from unrepaired or palliated CHDs. Increasing evidence demonstrates that postoperative pulmonary hypertensive events are more common in the infant who has been exposed to
weeks or months of high pulmonary pressure and flow. This observation seems especially true in such lesions as truncus arteriosus, complete atrioventricular canal defects, and transposition of the great arteries with ventricular septal defect. For example, in the 1970s, when the repair of complete atrioventricular canal defects commonly was performed in older children, the operative mortality rate was 10 percent to 15 percent. Today, this repair typically is performed within the first 6 months of life and the operative mortality rate is 3 percent to 5 percent. Finally, cognitive and psychomotor abnormalities associated with months of hypoxemia or abnormal hemodynamics may be diminished or eliminated by early repair.

Optimal perioperative care involves: (1) initial stabilization, airway management, and establishment of vascular access; (2) a complete and thorough noninvasive delineation of the anatomic defect(s); (3) resuscitation with evaluation and treatment of secondary organ dysfunction (particularly the brain, kidney, and liver); (4) cardiac catheterization if necessary (typically for (a) physiological assessment; (b) interventional procedures such as balloon atrial septostomy or valvotomy; or (c) anatomic definition not visible by echocardiography; for example, coronary artery distribution in pulmonary atresia with intact ventricular septum or delineation of aorticopulmonary collaterals in tetralogy of Fallot with pulmonary atresia); and (5) surgical management when cardiac, pulmonary, renal, and central nervous systems are optimized. Crucial in this process is a multidisciplinary approach that combines the disciplines of pediatric cardiology, cardiac surgery, cardiac anesthesia, neonatology, intensive care, and nursing.

**RECENT TECHNICAL INNOVATIONS**

Recent technical innovations have allowed for drastic reductions in mortality rates after neonatal open-heart surgery (Figure 2). These include a greater understanding of how to conduct cardiopulmonary bypass in the newborn. Low-flow bypass now is more commonly used, as opposed to deep hypothermic circulatory arrest. Arterial blood-gas management on bypass now aims to maintain physiological temperature-corrected pH by means of carbon dioxide insufflation into the oxygenator. This has now been proven to result in fewer complications postoperatively. Similarly, it was recently shown that keeping a higher hematocrit on bypass (contrary to “traditional” cardiac surgical practice) results in less postoperative edema and improved cerebral recovery. Other improvements include the development and use of antifibrinolytics such as aprotinin or aminocaproic acid, which results in less bleeding and inflammation; the introduction of smaller tubing and bypass-circuit components such as oxygenators or filters, which result in less volume load to the patient; and ultrafiltration techniques. In addition, the development of newer instruments allows us to use minimally invasive techniques in small children, which permits markedly shorter incisions. Finally, combined work between interventional cardiology and surgery has resulted in intraoperative placement of devices such as stents or ventricular septal defect closure devices, and has resulted in shortened bypass times and simplified procedures (Figure 3, p.10). 

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The emergence and growing popularity of the arterial switch operation for transposition of the great arteries (a uniformly lethal lesion if not repaired), illustrates this trend well (Figure 4). This operation, performed within the first 3 weeks of life, currently carries an expected mortality rate of less than 5 percent for simple transposition. It also results in a considerably improved quality of life and life expectancy because the repair is anatomically correct (i.e., the left ventricle ends up pumping for the systemic circulation and the right ventricle for the pulmonary circulation).

Another good example of a recent technical innovation is the Norwood operation for hypoplastic left-heart syndrome. Previous surgical mortality rates ranged between 50 percent and 80 percent. Today, mortality rates of less than 25 percent have been achieved, mainly owing to the advances mentioned above.

Much progress has been made in the field of neonatal cardiac surgery. Anatomic repairs of complex cardiac malformations now are performed with markedly improved survival rates and quality of life. The use of palliative procedures to delay surgery has diminished considerably.

REFERENCES