Introduction

Over the last 60 years pediatric cardiac anesthesia has developed as a subspecialty of pediatric anesthesia or, a subspecialty of cardiac anesthesia, depending on one’s perspective. It is impossible to describe the evolution of pediatric cardiac anesthesia without constantly referring to developments in the surgical treatment of congenital heart disease (CHD) because of the great interdependency of the two fields. As pediatric anesthesia developed over the years surgical treatments of children with CHD were invented, starting with simple surgical ligation of a patent ductus arteriosus (PDA) to sophisticated, staged repair of complex intracardiac lesions in low birthweight neonates requiring cardiopulmonary bypass (CPB) and circulatory arrest. Practically every advance in surgical treatment of CHD had to be accompanied by changes in anesthetic management to overcome challenges that impeded successful surgical treatment or mitigated morbidity associated with surgical treatment.

This history will mostly be organized around the theme of how anesthesiologists met these new challenges using the then-available anesthetic armamentarium. The second theme running through this story is the slow change of interest and focus from just events in the operating room (OR) to perioperative care in its broadest sense, including perioperative morbidity. The last theme is the progressive reduction in the age of patients routinely presenting for anesthesia and surgery from the 9-year-old child undergoing the first PDA ligation in 1938 to the fetus recently reported in the New York Times in 2002 who had aortic atresia repaired in utero. Interestingly, both patients had had their cardiac procedures at the same institution!

This story will be told working through the different time frames. The first years, 1938–54; CPB and early repair, 1954–70; deep hypothermic circulatory arrest (DHCA) and the introduction of prostaglandin E1 (PGE1), 1970–80; hypoplastic left heart syndrome (HLHS), 1980–90; refinement and improvement in mortality/morbidity, 1990–2000.

The first years, 1938–54

These years began with the ligation of the PDA and continued with palliative operations. The first successful operation for a CHD occurred in August 1938 when Robert E. Gross ligated the PDA of a 9-year-old girl. The operation and the postoperative course were smooth, but because of the interest in the case the child was kept in the hospital until the 13th day. In the report of the case Gross mentions that the operation was done under cyclopropane anesthesia, and continues: “The chest was closed, the lung being re-expanded with positive pressure anesthesia just prior to placing the last stitch in the intercostal muscles.”

A nurse using a “tight fitting” mask gave the anesthetic. There was no intubation and of course no postoperative ventilation. The paper does not mention any particular pulmonary complications so it cannot have been much different from an ordinary postoperative course of the day.

In 1952 Dr Gross published a review of 525 PDA ligations where many, if not all, of the anesthetics were administered by the same nurse anesthetist, under surgical direction. Here he states, “formerly we employed cyclopropane anesthesia for these cases, but since about half of the fatalities seemed to have been attributable to cardiac arrest or irregularities under this anesthetic, we have now completely abandoned cyclopropane and employ ether and oxygen as a routine.” It is probably correct that cyclopropane under these circumstances with insufficient airway control were more likely to cause cardiac arrhythmias than ether. An intralaryngeal airway was used which also served “to facilitate suction removal of any secretions from the lower airway” (and we may add, the stomach). Dr Gross claimed that the use of this airway reduced the incidence of postoperative pulmonary complications. Without having a modern, rigorous review of this series, it is hard to know what particular anesthetic challenges other than these confronted the anesthetist, but we may assume that intraoperative desaturation from the collapsed left lung, postoperative pulmonary complications,
McQuiston described the anesthetic technique used at the procedures and the anesthetic management. In 1948 CHD were added and a number of papers appeared describing for congenital malformations of the heart “in which there is pulmonary artery stenosis or atresia.” They reported in 1946 on their anesthetic experience with 100 operations. Whether patients 1 and 3 were intubated is unclear, but it is noted that in all three cases positive pressure ventilation was used to reinflate the lung. Interestingly, in this early kinder and gentler time, the surgical and pediatric authors reporting the Blalock-Taussig operation acknowledged by name the pediatricians and house officers who took such good care of the children postoperatively but still did not acknowledge in their paper the contribution of the anesthesiologists Lamont and Harmel!

Although intubation of infants was described by Gillespie as early as 1939 it is difficult to say exactly at what time intubations became routine. Doctors Harmel and Lamont, who were anesthesiologists, reported in 1946 on their anesthetic experience with 100 operations for congenital malformations of the heart “in which there is pulmonary artery stenosis or atresia.” They reported 10 anesthetic-related deaths in the series, so it is certain that they encountered formidable anesthetic problems in these surgical procedures. This is the first paper we know of published in the field of pediatric cardiac anesthesia.

In 1952 Damman and Muller reported a successful operation in which the main pulmonary artery was reduced in size and a band placed around the artery in a 6-month-old infant with single ventricle (SV). It is mentioned that morphine and atropine were given preoperatively but no further anesthetic agents are mentioned. At that time infants were assumed to be oblivious to pain so we can wonder what was used beyond oxygen and restraint.

Over the next 20 years many palliative operations for CHD were added and a number of papers appeared describing the procedures and the anesthetic management. In 1948 McQuiston described the anesthetic technique used at Children’s Memorial Hospital in Chicago. This is an excellent paper for its time, but a number of the author’s conclusions are erroneous, although they were the results of astute clinical observations and current knowledge at the time. The anesthetic technique for shunt operations (mostly Potts’ anastomosis) is discussed in some detail, but is mostly of historical interest today. McQuiston explained that he had no experience with anesthetic management used in other centers such as the pentothal-N₂O-curare used at Minnesota or the ether technique used at the Mayo Clinic. McQuiston used heavy premedication with morphine, pentobarbital and atropine and/or scopolamine; this is emphasized because it was important “to render the child sleepy and not anxious.” The effect of sedation with regard to a decrease in cyanosis (resulting in making the child look pinker) is noted by the authors. They also noted that children with severe pulmonary stenosis or atresia do not decrease their cyanosis “because of very little blood flow,” and these children have the highest mortality.

McQuiston pointed out that body temperature control was an important factor in predicting mortality and advocated the use of moderate hypothermia, i.e. “refrigeration” with icebags, because of a frequently seen syndrome of hyperthermia. McQuiston worked from the assumption that hyperthermia is a disease in itself, but did not explore the idea that the rise in central temperature might be a symptom of low cardiac output with peripheral vasoconstriction. Given what we now know of shunt physiology, it is interesting to speculate that this “disease” was caused by pulmonary hyperperfusion after the opening of what would now be considered as an excessively large shunt, stealing a large portion of systemic blood flow.

In 1950 Harris described the anesthetic technique used at Mount Zion Hospital in San Francisco. He emphasized the use of quite heavy premedication with morphine, atropine and scopolamine. The “basal anesthetic agent” was Avertin (tribromoethanol). It was given rectally and supplemented with N₂O/O₂ and very low doses of curare. Intubation was facilitated by cyclopropane. The FIO₂ was changed according to cyanosis, and bucking or attempts at respiration were thought to be due to stimulation of the hilus of the lung. This was treated with “cocainization” of the hilus.

In 1952 Dr Robert M. Smith discussed the circulatory factors involved in the anesthetic management of patients with CHD. He pointed out the necessity to understand the pathophysiology of the lesion and also “the expected effect of the operation upon this unnatural physiology.” That is, he recognized that the operations are not curative. The anesthetic agents recommended were mostly ether following premedication.

While most of these previous papers had been about tetralogy of Fallot (TOF), Dr Smith also described the anesthetic challenges of surgery for coarctation of the aorta. He emphasized the hypertension following clamping of the aorta and warned against excessive bleeding in children operated on at older ages using ganglionic blocking agents. This bleeding was far beyond what anesthesiologists now see in patients operated on at younger ages, before the development of substantial collateral arterial vessels.
From 1954 to 1970 the development of what was then called the “heart–lung machine” opened the heart to surgical repair of complex intracardiac congenital heart defects. At the time, the initial high morbidity of early CPB technology seen in adults was even worse in children, particularly smaller children weighing less than 10 kg. Anesthetic challenges multiplied rapidly in association with CPB coupled with early attempts at complete intracardiac repair. The lung as well as the heart received a large share of the bypass-related injuries leading to increased postoperative pulmonary complications. Brain injury began to be seen and was occasionally reported, in conjunction with CPB operations, particularly when extreme levels of hypothermia were used in an attempt to mitigate the morbidity seen in various organ systems after CPB.

In Kirklin’s initial groundbreaking report of intracardiac surgery with the aid of a mechanical pump–oxygenator system at the Mayo Clinic, the only reference to anesthetic management is a brief remark that ether and oxygen were given. In Lillehei’s description of direct vision intracardiac surgery in humans using a simple, disposable artificial oxygenator, there is no mention of anesthetic management. What strikes a “modern” cardiac anesthesiologist in these two reports is the high mortality: 50% in Kirklin’s series and 14% in Lillehei’s series. All of these patients were children with CHD ranging in age from 1 month to 11 years. Clearly such mortality and the associated patient care expense would not be tolerated today.

At that time, pediatric anesthesia was performed with open drop ether administration and later with ether using different non-rebreathing systems. Most anesthetics were given by nurses under the supervision of the surgeon. The first physician anesthetist to be employed by a children’s hospital was Robert M. Smith in Boston in 1946. The anesthetic agent to come into widespread use after ether was cyclopropane; in most of the early textbooks it was the recommended drug for pediatric anesthesia. Quite apart from being explosive, cyclopropane was difficult to use. It was obvious that carbon dioxide absorption was necessary with cyclopropane to avoid hypercarbia and acidosis, which might precipitate ventricular arrhythmias. However administration of a Waters’ absorber could be technically difficult especially as tracheal intubation was considered dangerous to the child’s small, delicate airway.

In all the early reports it is noted or implied that the patients were awake (more or less) and extubated at the end of the operation. In the description of the postoperative course, respiratory complications were frequent, either in the form of pulmonary respiratory insufficiency or airway obstruction. This latter problem was probably due to the fact that the largest tube which would fit through the larynx was used. Another reason may have been that the red rubber tube was not tissue tested. The former problem was probably often related to the morbidity of early bypass technology on the lung.

Arthur S. Keats, working at the Texas Heart Institute and Texas Children’s Hospital with Denton A. Cooley, had much experience with congenital heart surgery and anesthesia from 1955–60, and provided the most extensive description of the anesthetic techniques used in this era. He described anesthesia for congenital heart surgery without bypass in 150 patients, the most common operations being PDA ligation, Potts’ operation, atrial septectomy (Blalock–Hanlon operation), or pulmonary valvotomy. Premedication was with oral or rectal pentobarbital, chloral hydrate per rectum, intramuscular meperidine, and intramuscular scopolamine or atropine. Endotracheal intubation was utilized, and ventilation was assisted using an Ayre’s T-piece, to-and-fro absorption system, or circle system. Cyclopropane was used for induction, and a venous cutdown provided vascular access. Succinylcholine bolus and infusion were used to maintain muscle relaxation. Light ether anesthesia was used for maintenance until the start of chest closure, and then 50% N₂O used as needed during chest closure. Of note is that the electrocardiogram, ear oximeter, and intra-arterial blood pressure recordings were used for monitoring during this period, as well as arterial blood gases and measurements of electrolytes and hemoglobin. The next year he published his experiences with 200 patients undergoing surgery for CHD with CPB, almost all of whom were children. Ventricular septal defect (VSD), atrial septal defect (ASD), TOF, and aortic stenosis were the most common indications for surgery. The anesthetic techniques were the same as above, except that d-tubocurare was given to maintain apnea during bypass.

Perfusion rates of 40–50 mL/kg/minute were used in infants and children, and lactic acidemia after bypass (average 4 mmol/L) was described. No anesthetic agent was added during bypass, and “patients tended to awaken during the period of bypass,” but apparently without recall or awareness. Arrhythmias noted ranged from frequent bradycardia with cyclopropane and succinylcholine, to junctional or ventricular tachycardia, ventricular fibrillation (VF), heart block, and rapid atrial arrhythmias. Treatments included defibrillation, procainamide, digitalis, phenylephrine, ephe-drine, isoproterenol, and atropine. Eleven of 102 patients with VSD experienced atrioventricular block. Epicardial pacing was attempted in some of these patients but was never successful. Fresh citrated whole blood was used for small children throughout the case, and transfusion of large amounts of blood was frequently necessary in small infants. Mortality rate was 13% in the first series (36% in the 42 patients <1 year of age), and 22.5% in the second series (47.5% in the 40 patients <1 year of age). Causes of death included low cardiac output after ventriculotomy, irreversible VF, coronary air emboli, postoperative atrioventricular block, hemorrhage,
pulmonary hypertension, diffuse atelectasis, and aspiration of vomitus. No death was attributed to the anesthetic alone. Reading these reports provides an appreciation of the daunting task of providing anesthesia during these pioneering times.

Tracheostomy after cardiac operations was not unusual and in some centers it was done “prophylactically” a week before the scheduled operation. These practices were certainly related to primitive (in present terms) techniques and equipment used for both endotracheal intubation and CPB. Postoperative ventilatory support did not become a routine until later when neonatologists and other intensive care specialists had proven it could be done successfully. Successful management of prolonged respiratory support was first demonstrated in the great epidemics of poliomyelitis in Europe and the USA in 1952–54.15

Halothane was introduced in clinical practice in the mid-1950s and it became rapidly the most popular agent in pediatric anesthesia, mostly because of the smooth induction compared to the older agents. Halothane was also widely used for pediatric cardiac anesthesia in spite of its depressive effect on the myocardium and the significant risk of arrhythmias. Halothane continues to be used in some places as the anesthetic agent of choice for pediatric cardiac cases, although newer inhalational agents like isoflurane and sevoflurane are now more widely used. Sevoflurane is probably the inhalational agent most often used for induction of anesthesia in pediatric cardiac cases in US academic centers.

During this period, adult cardiac anesthesiologists following the practice reported by Edward Lowenstein in 197016 began to use intravenous anesthesia based on opiates. Initially morphine in doses up to as much as 1 mg/kg was given with 100% oxygen and this technique became the anesthetic of choice for adult cardiac patients, but vasodilation and hypotension associated with its use slowed the incorporation of this technique into pediatric cardiac anesthesia until the synthetic opiates became available.

Before CPB was yet developed, or when it still carried high morbidity and mortality, a number of modalities were used to improve the outcome for infants. One was inflow occlusion (IO), another was the hyperbaric chamber. Inflow occlusion was useful and, if well managed, an elegant technique. The secret was the organization of the efforts of the entire operative team, and the technique required the closest cooperation between surgeon and anesthesiologists. The technique was as follows.

The chest was opened in the midline. After pericardiotomy, a side-clamp was placed on the right atrium (RA) free wall and an incision made in the RA or proximal on the pulmonary artery prior to placing the vascular clamps used to occlude caval return. Prior to application of the clamps, patients were hyperventilated with 100% oxygen. During IO, the superior vena cava (SVC) and inferior vena cava (IVC) inflow were occluded, ventilation held, the RA or the pulmonary artery clamp released; the heart was allowed to empty, and the septum primum excised or the pulmonic valve dilated. After excision of the septum or valvotomy, one caval clamp was released initially to de-air the atrium. The RA side-clamp or the pulmonary artery clamp was then reapplied and the other caval clamp released. The heart was resuscitated with bolus calcium gluconate (range 30–150 mg/kg) and bicarbonate (range 0.3–3.0 mEq/kg). Occasionally inotropes were administered, most often dopamine. It was important to titrate the inotropes so as not to aggravate rebound hypertension caused by endogenous catecholamines. The duration of the IO was between 1 and 3 minutes—terrifying minutes for the anesthesiologist, but quickly over.

Another modality used to improve the survival after shunt operations, pulmonary artery banding (PAB) and atrial septectomy, was to operate in the hyperbaric chamber, thereby benefiting from the increased amount of physically dissolved oxygen. It was a cumbersome affair operating in crowded and closed quarters. There was room for only two surgeons, two nurses, one anesthesiologist, and one baby, as the number of emergency oxygen units limited access. Retired navy divers ran the chamber and kept track of how many minutes the personnel had been in the hyperbaric chamber in the previous week. Help was not readily available because the chamber was buried in a sub-basement and people had to be sluiced in through a side arm that could be pressurized. The chamber was pressurized to 2–3 atmospheres so it was unpleasantly hot while increasing the oxygen pressure and cold while decreasing the pressure; people with glasses were at a disadvantage. It did not seem to add to survival and was abandoned circa 1974.

Anesthesia was a challenge in the hyperbaric chamber. The infants were anesthetized with ketamine and nitrous oxide. As the pressure in the chamber increased, the concentrations of N₂O had to be decreased to avoid the hypotension and bradycardia that occurred rapidly.

Also in this era, the first infant cardiac transplant was performed by Kantrowitz in 1967.17 The recipient was an 18-day-old, 2.6 kg patient with severe Ebstein’s anomaly, who had undergone a Potts’ shunt on day 3 of life. The donor was an anencephalic newborn. The anesthetic technique is not described, and the infant died 7 hours postoperatively of pulmonary dysfunction.

The era of deep hypothermic circulatory arrest and the introduction of prostaglandin E₁, 1970–80

About 1970 physiological repair of CHD or “correction” had begun to come of age. In the adult world, coronary bypass operations and valve replacement spurred interest in cardiac anesthesia, which centered increasingly on the use of high dose narcotics and other pharmacological interventions. As
synthetic opiates with fewer hypotensive side effects became available, their use spread into pediatric cardiac anesthesia late in the 1970s and 1980s.

Children were still treated as “small adults” because major physiologic differences were not yet well appreciated, particularly as they related to CPB morbidity. Cardiopulmonary bypass was rarely employed during surgery on children weighing less than 10 kg because of the very high mortality and morbidity that had been experienced in the early years. The notion of repairing complex CHD in infancy was getting attention but was hindered by technical limitations of surgical techniques, CPB techniques, and anesthetic challenges in infants. Theoretically, physiological repair early in life provides a more normal development of the cardiovascular and pulmonary systems and might avoid palliation all together. The advantage of this was that the sequelae after palliation, for instance, distorted pulmonary arteries after shunts and PAB, might be avoided. Pulmonary artery hypertension following Waterston and Potts’ shunts occurred as a result of increased pulmonary blood flow and resulted in pulmonary obstructive disease. This would not develop if the defect were physiologically repaired at an early age. Furthermore, parents could be spared the anxiety of repeated operations and the difficulties of trying to raise a child with a heart that continued to be impaired.

The perceived need for early repair, together with the high mortality of bypass procedures, in infants and small children led to the introduction of DHCA. It was first practiced in Kyoto, Japan, but spread rapidly to Russia, the West coast of the USA at Seattle, and from there to midwestern and other US pediatric centers. As an example of the difficulties this presented to anesthesiologists, the introduction of DHCA in practice at the Children’s Hospital in Boston is useful. The newly appointed chief of cardiovascular surgery at the Children’s Hospital in Boston was Aldo R. Castaneda, MD, PhD, one of the first supporters of early total correction of CHD, who quickly embraced DHCA as a tool to accomplish his goals for repair in infants. In 1972, he immediately introduced DHCA into practice at the Children’s Hospital in Boston and the rather shocked anesthesia department had to devise an anesthetic technique to meet this challenge, aided only by a couple of surgical papers in Japanese which Dr Castaneda kindly supplied to the anesthesia department. These papers had, of course, little reference to anesthesia.

The first description of the techniques of DHCA from Japan in the English literature was Horiuchi’s from 1963. They used a simple technique with surface cooling and rewarming during CPB, using ether as the anesthetic agent, without intubation. In 1972 Mori reported details of their technique for cardiac surgery in neonates and infants using deep hypothermia, again in a surgical publication. Their anesthetic technique was halothane/N₂O combined with muscle relaxant; carbon dioxide was added to the anesthetic gas during cooling and rewarming (pH stat) to improve brain blood flow. The infants were surface cooled with ice bags and rewarmed on CPB.

Surprisingly, given the enormity of the physiological disturbances and challenges presented by DHCA, very few articles describing an anesthetic technique for DHCA were published, perhaps because DHCA and early correction was not widely accepted. A paper from Toronto described an anesthetic regime with atropine premedication occasionally combined with morphine. Halothane and 50% N₂O were used, combined with d-tubocurare or pancuronium. Carbon dioxide was added to “improve tissue oxygenation by maintaining peripheral and cerebral perfusion.” The infants were cooled with surface cooling (plastic bags with melting ice) and rewarmed on CPB. It was noted that six of the 25 infants had VF when cooled to below 30°C.

Given the lack of any scientific data or studies to guide anesthetic management of such cases, a very simple technique with ketamine-O₂-N₂O and curare supplemented by small amounts of morphine (0.1–0.3 mg/kg) was used at the Children’s Hospital in Boston. This was the way infants were anesthetized for palliative cardiac surgical procedures in the hyperbaric chamber at Boston Children’s Hospital. The infants were surface cooled in a bathtub filled with ice water to a core temperature of approximately 30°C. The bathtub was a green plastic bucket (for dishwashing!) bought at a Sears–Roebuck surplus store, keeping things as simple as possible (Fig. 1.1). This method was used in hundreds of infants over the next couple of years and only one infant developed VF in the ice water bathtub. This was an infant with TOF who suffered a coronary air embolus either from a peripheral i.v. or during an attempted placement of a central venous line. In retrospect, it is amazing that so few papers were published about the anesthetic management of this procedure that rapidly was seen to be lifesaving. The little material that was published about these techniques was restricted to surgical journals and did not describe or make any attempt to study the anesthetic techniques used for DHCA. The published surgical articles were largely unknown to cardiac and pediatric anesthesiologists.

It was during these 10 years that the “team concept” developed with cardiologists, cardiac surgeons and anesthesiologists working together in the OR and the intensive care unit (ICU) in the larger centers. These teams were facilitated by the anesthesiologists’ “invasion” of weekly cardiology–cardiac surgeon conferences where the scheduled operations for the week were discussed. Dr Aldo Castaneda, the chief surgeon at Boston Children’s Hospital, was a leader in the creation of a cardiac team concept for pediatric cardiac surgery.

During the first year of using DHCA in Boston, it was noticed that a number of the infants had “funny, jerky” movements of the face and tongue. A few also had transient seizures during the postoperative period, but as they had normal electroencephalographs (EEGs) at 1-year follow up, it was felt that significant cerebral complications were not a
problem. In view of knowledge developed subsequently, these clues to neurologic damage occurring during and after pediatric cardiac surgery involving DHCA were overlooked. In hindsight maybe it will be more correct to say these clues were ignored, thus a great opportunity to study this problem was delayed for almost two decades. The issue of neurologic damage with DHCA was raised repeatedly by surgeons such as John Kirklin, but was not really studied until the group at Boston Children’s Hospital led by Jane Newburger and Richard Jonas systematically followed a cohort of infants who had the arterial switch operation in the late 1980s using DHCA techniques.21 In the late 1980s and early 1990s, Greeley and coworkers at Duke University performed a series of human studies delineating the neurophysiologic response to deep hypothermia and circulatory arrest.22 These studies provided the crucial data in patients from which strategies for cooling and rewarming, length of safe DHCA, blood gas management, and perfusion were devised to maximize cerebral protection.

Those ongoing studies were followed by a number of studies comparing DHCA with hypothermic low flow perfusion, with different hematocrit in the perfusate and with different pH strategies during hypothermic CPB, pH-stat vs. α-stat.

During those years, the ketamine–morphine anesthetic technique had been supplanted by fentanyl-based high dose narcotic techniques. For the neurologic outcome studies the anesthetic technique was very tightly controlled, using fentanyl doses of 25 µg/kg at induction, incision, onset of bypass, and on rewarming, in addition to pancuronium. From the beginning of this period, surgical results as measured by mortality alone were excellent with steady increases in raw survival statistics. Because anesthetic techniques were evolving over this period of time, it was difficult to definitely ascribe any outcome differences to different anesthetic agents. A 1984 study of 500 consecutive cases of cardiac surgery in infants and children looked at anesthetic mortality and morbidity. Both were very low, so low in fact that they were probably not universally believed.23

As the new synthetic opioids such as fentanyl and sufentanil were developed, they replaced morphine to provide more hemodynamic stability in opiate-based anesthetic techniques for cardiac patients. In 1981, Gregory and his associates first described the use of “high dose” fentanyl, 30–50 µg/kg, combined with pancuronium in 10 infants undergoing PDA ligation. It is noteworthy that transcutaneous oxygen tension was measured as part of this study. This paper was in fact the introduction of high dose narcotics in pediatric cardiac anesthesia.24

The technique was a great success; one potential reason for that success was demonstrated 10 years later in Anand et al.’s paper showing attenuation of stress responses in infants undergoing PDA ligation who were given lesser doses of fentanyl in a randomized, controlled study.25

During this same period, synthetic opioids were replacing morphine in adult cardiac surgery. This technique slowly and somewhat reluctantly made its way into pediatric anesthesia,26 replacing halothane and morphine, which had previously been the predominant choice of pediatric anesthesiologists dealing with patients with CHD. In the years from 1983 to 1995 a number of papers were published showing the effect of different anesthetic agents on the cardiovascular system in children with CHD. Ketamine, nitrous oxide, fentanyl, and sufentanil were systematically studied. Some
misconceptions stemming from studies of adult patients were corrected, like the notion that N₂O combined with ketamine raises pulmonary artery pressure (PAP) and pulmonary vascular resistance (PVR). On the other hand, the role of increased PCO₂ or lower pH in causing higher PVR was also demonstrated and that subsequently became important in another connection. A number of studies done at this time demonstrated in a controlled fashion the earlier clinical observation (Harmel and McQuiston in the late 1940s) that in cyanotic patients the oxygen saturation would rise during induction of anesthesia, almost irrespective of the agent used. These events only reinforce the value of acute clinical observation and provide an example of how the interpretation of such observations may well change as new knowledge is discovered.

**Patent ductus arteriosus and the introduction of prostaglandin E₁**

In the mid-1970s several discoveries were made and introduced into clinical practice that turned out to be of great importance to the pediatric cardiac anesthesiologist and the rest of the cardiac team, the most important being the discovery that PGE₁ infused intravenously prevented normal ductal closure. These developments revolved around the role of the PDA in the pathophysiology of both cyanotic and acyanotic CHD. The critical role of PDA closing and opening in allowing early neonatal survival of infants with critical CHD began to be appreciated and clinicians sought methods of either keeping the PDA open or closing it, depending on what type of critical CHD the neonate was born with and the role of patency of the ductus arteriosus in the CHD pathophysiology. In some cases, particularly in very small neonates, the importance of closing the PDA was increasingly appreciated and in other cases, the critical importance of maintaining the patency of a PDA was appreciated.

As the survival of very small premature infants began to improve, mostly because of technical improvements with the use of a warmed isolette and improved mechanical ventilation for very small newborns, it became apparent that in many of these infants the PDA would not undergo the normal closure over time. As the understanding of these infants’ physiological problems improved and more infants survived, the role of continued patency of the PDA in neonates needing mechanical ventilation was appreciated. This led to medical therapy directed at promoting ductal closure using aspirin and indomethacin.

When such attempts failed, it was increasingly understood that necrotizing enterocolitis in the preemie was associated with decreased mesenteric blood flow secondary to the “steal” of systemic blood flow into the pulmonary circulation through a PDA. Thus, in cases when the PDA failed to close in premature infants, the need for operative treatment of the PDA in preemies arose as prophylaxis for necrotizing enterocolitis.

Pediatric and cardiac anesthesiologists were now faced with the task of anesthetizing these tiny preemies safely. This involved maintaining body temperature in infants of 1 kg or less with very large surface area/volume ratios. Intraoperative fluid restriction was important and low levels of Fio₂ were used to decrease the risk of retinopathy of prematurity. As the decade progressed these issues emerged and were addressed. In 1980, Neuman and Hansen described the anesthetic management of 70 such infants using an O₂/N₂O-muscle relaxant anesthesia technique with no mortality. Low Fio₂ was used to reduce the risk of retrolental fibroplasia, and precautions were taken to prevent heat loss. In those days before acquired immunodeficiency syndrome (AIDS) became a wide concern, 40% of the infants received blood transfusion. Interestingly, the question of whether to operate in the neonatal intensive care unit (NICU) or the OR for closure of the PDA in the very small newborns was discussed at that time and remains unsettled today!

The PDA lesion presents an interesting story. In 1938 it was the first of the CHD lesions to be successfully treated surgically. In the mid-1970s it was closed with medical therapy, first with aspirin and later with indomethacin. It was the first CHD lesion to be treated in the catheterization laboratory using different umbrella devices or coils. Presently, if surgical closure is necessary, it is often done using a minimally invasive, thoracoscopic video-assisted technique. This has the user-friendly, more benign effect of using four small incisions, avoiding an open thoracotomy, limiting dissection and trauma to the left lung. At the same time, this latest development of surgical technique required the anesthesiologist to again change the anesthetic approach to these patients. Unlike adult anesthesiologists who can use double-lumen endotracheal tubes for thoracoscopic procedures, pediatric anesthesiologists caring for 1–3 kg infants undergoing PDA ligation do not have the luxury of managing the left lung. Another problem posed by thoracoscopic PDA ligation in the infant is the emerging need for neurophysiologic monitoring of the recurrent laryngeal nerve’s innervation of the muscles of the larynx to avoid injury, a known complication of PDA surgery. The last issue is tailoring the anesthetic so that the children are awake at the end of the operation, can be extubated and spend an hour or so in the post-anesthesia care unit, bypassing the cardiac intensive care unit (CICU). In fact, in 2001 a group led by Hammer at Stanford published the first description of true outpatient PDA ligation in two infants aged 17 days and 8 months. These patients were managed with epidural analgesia, extubated in the OR, and discharged home 10 hours postoperatively. This report brings PDA closure full circle from a 13-day hospital stay following an ether mask anesthetic for an open thoracotomy to a day surgery procedure in an infant undergoing an endotracheal anesthetic for a thoracoscopic PDA ligation!
numerically and in terms of being life-sustaining in neonates with critical CHD. The introduction of PGE₁ suddenly improved the survival rate of a large number of neonates with CHD having lesions that require ductal patency to improve pulmonary blood flow, or to improve systemic blood flow distal to a critical coarctation of the aorta. The introduction of PGE₁ into clinical practice for therapy of neonatal CHD substantially changed the life of the pediatric cardiac surgeon and the pediatric cardiac anesthesiologist, as frequent middle-of-the-night shunt operations with extremely cyanotic infants almost immediately became a thing of the past. These operations were particularly daunting when one realizes that these procedures were most common before the availability of pulse oximetry; the only warning signs of impending cardiovascular collapse were the very dark color of the blood and preterminal bradycardia. To get an arterial blood gas with a Pao₂ in the low teens was not uncommon, and Pao₂ measurements in single digits in arterial blood samples from live neonates during such surgical procedures were recorded! Even more dramatic was the disappearance of the child with critical post-ductal coarctation. These infants were extremely acidic with a pH of 7.0 or less at the start of the procedure (if it was possible to obtain an arterial puncture); they looked mottled and almost dead below the nipples. With the advent of PGE₁ therapy, they were resuscitated medically in the ICU and could be operated on the following day in substantially better condition than previously.

But the introduction of PGE₁ had an effect that was not clearly foreseen except maybe by some astute cardiologists. Survival of a number of these neonates presented pediatric cardiologists and cardiac surgeons (and then anesthesiologists) with rare and severe forms of CHD that had hitherto been considered a “rare” pathological diagnosis. Foremost among these were the infants with HLHS and some forms of interrupted aortic arch (IAA). As further experience was gained, it became obvious that these forms of disease were not so rare, but infants who had survived with those forms of CHD were very rare.

The story of hypoplastic left heart syndrome, 1980–90

As mentioned above, the introduction of PGE₁ brought major changes to pediatric cardiac anesthesia, solving some problems and at the same time bringing new challenges for the cardiac team. New diagnoses of CHD presented for treatment and were recognized; some had been known previously but had until then presented insurmountable obstacles to any effective therapy.

One of these was HLHS. It had been accurately described in 1958 by Noonan and Nadas but only as a pathological diagnosis. The syndrome is a ductus dependent lesion and had 100% mortality within a few days to weeks when the ductus underwent physiological closure. Hypoplastic left heart syndrome was therefore of no practical interest from a therapeutic standpoint until ductal patency could be maintained. When it became possible to keep the ductus arteriosus patent with PGE₁, these neonates rapidly became a problem that could not easily be ignored. In the beginning, most of the infants were misdiagnosed as having sepsis and being in septic shock—few babies reached the tertiary center without a tell-tale Band-Aid indicating a lumbar puncture to rule out sepsis.

But even with the ability to diagnose the defect in a live neonate temporarily kept alive with a PGE₁ infusion, the outlook was not much better. There was no operation devised, and in some centers such neonates were kept viable on a PGE₁ infusion for weeks and even months in the (usually) vain attempt to get them to grow large enough for some surgical procedure to be attempted.

In the next years several centers tried different approaches with ingenious conduits, trying to create an outlet from the right ventricle to the aorta and the systemic circulation.

Those were also the years where President Ronald Reagan’s Baby Doe regulations were in effect. Anyone who thought an infant was being mistreated, i.e. not operated upon, could call a “Hot Line Number” which was posted in all neonatal ICUs to report the physicians “mislreatment” of the infant. Fortunately this rule died a quiet death after a few chaotic years.

In the meantime the search for a palliative operation went on, also spurred by the increasing success of the Fontan operation, which had been introduced in 1970. This meant that there now was a theoretical endpoint for HLHS as well as for other forms of single ventricle (SV) physiology. It was William Norwood at Boston Children’s Hospital who was the first to devise not only a viable palliation but also to complete the repair with a Fontan operation the following year. The publication of this landmark paper spurred considerable discussion. Many cardiologists and surgeons took the position that this operative procedure represented experimental and unethical surgery, and that these infants “were better off dead.”

The current approach to these infants varies from multistage physiological repair with palliation followed by Fontan operation. Another alternative is neonatal transplantation as proposed by the group at Loma Linda in California.

Some cardiologists are still advocates of conservative “comfort care” for neonates with HLHS.

With eventual survival of about 70% being achieved in many centers, these infants can no longer be written off as untreatable. Now the question is more about quality of survival, especially intellectual development. It is also recognized that many have both chromosomal and non-chromosomal anomalies both in the cerebral and gastrointestinal systems.

As was the case from the beginnings of pediatric cardiac surgery, this new patient population presented a management dilemma for the anesthesiologists; they posed a new set of problems that required solution before acceptable operative results could be achieved. It was obvious that
patients with HLHS were hemodynamically unstable before CPB because of the large volume load on the heart coupled with coronary artery supply insufficiency. The coronary arteries in HLHS are supplied from the PDA retrograde through a hypoplastic ascending and transverse aorta that terminates as a single “main” coronary artery. A common event at sternotomy and exposure of the heart was VF secondary to mechanical stimulation. This fibrillation was sometimes intractable, necessitating emergent CPB during internal cardiac massage. This was not an auspicious beginning to a major experimental open heart procedure.

It was during these years that the transition from morphine–halothane–N₂O to high dose narcotic technique with fentanyl or sufentanil combined with 100% oxygen took place. This technique seemed to provide some protection against the sudden VF events. Despite this modest progress in getting patients successfully onto CPB, it soon became painfully clear to us that we had not made much progress in treating this lesion when we tried to wean the patients from bypass. The infants were still unstable coming off bypass and severely hypoxic, and it took some time before we discovered a way to deal with the problem.

A chance observation led us to a solution. We noticed that infants who came off bypass with low PaO₂ (around 30 mmHg) after the HLHS repair often did well, while the ones with immediate “excellent gases” (PaO₂ of 40–50 mmHg or better) became progressively unstable in the ICU a couple of hours later, developing severe metabolic acidosis and dying during the first 24 h.

This observation combined with discussions with the cardiologists about PVR and systemic vascular resistance (SVR) made us attempt to influence these resistances to assure adequate systemic flow. In retrospect, infants with low PaO₂ after bypass had smaller pulmonary artery shunts and adequate systemic blood flow, while those with larger pulmonary shunts and higher initial PaO₂ levels after weaning from bypass tended to “steal” systemic blood flow through the pulmonary artery shunt. This would occur in the postoperative period, as the PVR remained elevated as a result of CPB before returning to more normal levels. These observations led to the technique of lowering the FiO₂ sometimes as low as 0.21 and to allow hypoventilation to increase PVR in patients that had larger size shunts placed to supply adequate systemic blood flow as part of what became known as the Norwood operation. A different technique used at other institutions to deal with this problem was to add carbon dioxide to the anesthetic gas flow, increasing PVR and continuing to use “normal ventilation” in children that had larger shunts placed and excessive pulmonary blood flow. Both techniques represented different approaches to the same problem: finding ways of dealing with the need to carefully balance PVR and SVR after bypass in a fragile parallel circulation in the post-bypass period where dynamic changes were taking place in ventricular function.

These observations and the subsequent modifications in anesthetic and postoperative management improved the survival for the stage I palliation (Norwood procedure). It should be noted that the pediatric cardiac anesthesiologist was a full, contributing partner in the progressive improvement in outcome of this very complex and challenging lesion. More important, the techniques developed and the knowledge gained in this process also simplified the management of other patients with parallel circulation and SV physiology. The obvious example is truncus arteriosus where the “usual” ST depression and frequent VF that occurred intraoperatively almost always can be avoided. Any decrease in PVR during anesthesia in a child with unrepaired truncus arteriosus can lead to pulmonary “steal” of systemic blood flow and decreased diastolic pressure through the common trunk to the aorta and pulmonary artery, resulting in hypotension and insufficient systemic blood flow expressed initially as coronary insufficiency and ST depression (or elevation).

During the same decade the surgical treatment of transposition of the great arteries (TGA) underwent several changes. The Mustard operations (as one type of atrial switch procedure) were feared because of the risk of SVC obstruction as a complication of this surgical procedure. At the end of a Mustard procedure, it was not uncommon to see a child with a grotesquely swollen head who had to be taken back to the OR for immediate reoperation. Many of those children suffered brain damage, especially when reoperation was delayed. This resulted from low perfusion pressure during bypass because of venous hypertension in the internal jugular veins and SVC. The extent and prevalence of such damage was never systematically studied. The arterial pressure during bypass and in the immediate post-bypass period in the OR tended to be low and the pressure in SVC high. An article from Great Ormond Street Hospital for Children in London demonstrated arrested hydrocephalus in Mustard patients. The Senning operation (another variant of the atrial switch approach to TGA) was better, but those children could develop pulmonary venous obstruction acutely in the OR, after the procedure, or progressively after hospital discharge. When the diagnosis was not promptly made and acted upon, these infants were often quite sick by the time they came to reoperation.

The successful application of the arterial switch procedure described by Jatene et al. then began to revolutionize operations for TGA. It eliminated the risk of obstruction of the pulmonary and systemic venous return seen after the Mustard and Senning procedures. It also diminished the incidence of the subsequent sick sinus syndrome, a complication that might develop in the first 10 years postoperatively resulting from the extensive atrial suture lines and reconstructions required by these “atrial” switch procedures. The introduction of the arterial switch operation again involved anesthesiologists. The initial attempts at arterial switch operations in many institutions resulted in substantial numbers of
infants who had severe myocardial ischemia and even frank infarcts. This resulted from a variety of problems with the coronary artery transfer and reimplantation into the “switched” aorta that had been moved to the left ventricle outflow tract. Pediatric cardiac anesthesiologists gained extensive experience with intraoperative pressor and inotropic support, and nitroglycerine infusions. They were expected by surgeons to provide support to get infants through what later turned out to be iatrogenically caused myocardial ischemia. As surgeons learned to handle coronary artery transfers and reanastomoses well, these problems largely disappeared, along with the need for major pressor and inotropic support, and for nitroglycerine infusion inappropriately directed at major mechanical obstructions in the coronary arterial supply. The arterial switch operation has now been refined at most centers to the point where it is largely “routine” and presents, for the most part, no unique anesthetic challenges.

It was during the same time period that a randomized strictly controlled study of stress response in infants undergoing cardiac surgery while anesthetized with high dose sufentanil was performed. It showed that a high dose narcotic technique would suppress but not abolish stress responses. It also seemed to show a reduction in morbidity and possibly mortality. However, when the study was repeated 10 years later, these results did not quite hold up. It must be pointed out that the patient population was older and refinement of the bypass technique had occurred.

**Fontan and the catheterization laboratory, 1990–2000**

After the anesthetic technique and preoperative management of the stage I palliation had been refined and we had been encouraged by the initial successes of stage II, problems arose. The Fontan operation became problematic as it was applied to younger patients with a great variety of SV types of CHD. Many of the patients had seemingly perfect Fontan operations but in the CICU they developed low cardiac output and massive pleural and pericardial effusions postoperatively. Many died in the postoperative period despite a variety of different support therapies; their course over the first 24–48 hours was relentlessly downward and could only be reversed by taking them back to the OR, reversing the Fontan operation, and reconstructing a systemic to pulmonary artery shunt. It was hard for the caretakers of those infants to accept such losses of children they had known from birth. They were our little friends and we knew the families too. All kinds of maneuvers were tried to avoid the above sequence of events, from early extubation to the use of a G-suit to improve venous return to the heart. In some centers a large balloon was placed tightly around the child’s lower body and intermittently inflated by a Bird respirator asynchronous with ventilation.

After a couple of years two innovations changed the outlook. Both were linked to the understanding that a major limitation of the Fontan operation was the need for a normal or near normal PVR to allow survival through the postoperative period when CPB had caused, through release of a variety of inflammatory mediators and cytokines, a marked elevation of PVR in the early postoperative period. When this bypass-related inincrease PVR was associated with younger age (<2 years of age) at the time a Fontan was attempted, the higher baseline PVR of the infant made the bypass-related PVR worse and resulted in inadequate pulmonary blood flow and (single) ventricular filling in the early postoperative period, leading to a cycle of low cardiac output, pulmonary and systemic edema, further increases in PVR, acidosis and death.

One solution was to interpose a bidirectional cavopulmonary anastomosis (bidirectional Glenn procedure, BDG) 6–12 months before completion of the Fontan operation. This procedure, increasingly known as a “hemifontan,” directed only half of the systemic venous return through the lungs at a time when the infant’s PVR had not fallen to normal levels, and preserved an alternative pathway for (single) ventricular filling through systemic venous return not routed through the lungs. This enabled the patients to maintain reasonable cardiac output, although a bit “blue”, during the early postoperative period, when the PVR had been elevated by CPB. However this made a third operation, the completion of the Fontan, necessary.

The other innovation was the “fenestrated” Fontan where a small fenestration in the atrial baffle allowed systemic venous return to bypass the lungs as a right-to-left shunt thereby maintaining ventricular filling and systemic cardiac output during the early postoperative period of high PVR. Over time the fenestration closed as PVR fell and shunting decreased. Alternatively, a device delivered during an interventional cardiac catheterization could close the fenestrations.

This whole process of testing the applicability of the Fontan principle and various modifications of the Fontan operation to a wide variety of types of severe cyanotic CHD involved another set of challenges for the pediatric cardiac anesthesiologist and collaboration between anesthesiology, cardiology and surgery. The net result of a great deal of work and collaboration among these groups was that the outlook for the HLHS patients and indeed for all children with SV defects improved locally and, as these improvements spread and were amplified by work done in other centers, the improvement became national and international. In some institutions the preferred treatment was and is neonatal transplantation. Its limits are the long waiting time for a transplant, the unavoidable mortality during the waiting period and the ongoing morbidity of neonatal heart transplants, a lifetime of immunosuppression therapy, and the accelerated risk of coronary artery disease seen in heart transplants, even in young children.
The collaboration with pediatric cardiologists around postoperative care of HLHS, Fontan patients and others spread naturally to the cardiac catheterization laboratory. As pediatric cardiologists began to develop interventional procedures, the need for more control and support of vital functions became apparent. Previously, nurses operating under the supervision of the cardiologist performing the catheterizations had sedated the children for the procedures. In many institutions this involved high volumes of cases sedated by specially trained nurses while in others with smaller pediatric case loads the practice of using general anesthesia for children undergoing cardiac catheterizations had been routine.

The interventional cardiologists turned to pediatric cardiac anesthesiologists for help in managing these patients while the cardiologists themselves were dealing with the complex demands of doing interventional procedures in infants and children with CHD. As was the case with newly devised pediatric cardiac surgical procedures, the development of interventional procedures for CHD in the cardiac catheterization laboratory posed a whole new and different set of problems and challenges for pediatric cardiac anesthesia. Not the least of these was providing anesthesia and vital function support in the dark and difficult environment of the cardiac catheterization laboratory. The introduction of dilation techniques for pulmonary arteries and veins, mitral and aortic valves, and most recently, the dilation of fetal atrial aortic valves in utero along with device closure of the PDA, ASD, and VSD all placed progressively more demands on the anesthesiologists, who became more and more involved in these procedures.

The development of another set of interventional procedures, the use of radiofrequency ablation to deal with arrhythmias in the pediatric patient, illustrates the progressive complexity and difficulty of anesthesia care in these patients. Used initially only on healthy teenagers with structurally normal hearts but having paroxysmal atrial tachycardia (PAT), anesthesia care was quite straightforward. Now, in contrast, many of these radiofrequency ablation procedures are done in children with complex CHD, repaired or unrepaired, and frequently the children (or adults) may be quite cyanotic or have low cardiac output. At present in Children’s Hospital Boston the cardiac catheterization laboratory and the cardiac magnetic resonance imaging (MRI) unit present close to 1000 anesthesia cases per year over and above cardiac surgical cases.

But with all those developments the defects remain the same. If we look at the relative distribution of cases in 1982 and 2001 we see the same diagnosis and pretty much the same numerical relationship between the major groups. As Helen Taussig remarked in her paper about the global distribution of cardiac diagnosis, only surgical interventions change the numbers. This we can see in the rise in numbers of Norwood and Fontan operations (Table 1.1).

<table>
<thead>
<tr>
<th>Table 1.1 Cardiovascular surgery at Children’s Hospital Boston.</th>
<th>1982 (%)</th>
<th>2001 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total cases</td>
<td>n = 538</td>
<td>n = 931</td>
</tr>
<tr>
<td>Septal defects</td>
<td>27.0</td>
<td>24.0</td>
</tr>
<tr>
<td>VSD repair</td>
<td>12.0</td>
<td>10.0</td>
</tr>
<tr>
<td>ASD repair</td>
<td>9.6</td>
<td>9.6</td>
</tr>
<tr>
<td>CAVC repair</td>
<td>5.9</td>
<td>4.6</td>
</tr>
<tr>
<td>Cavopulmonary connection</td>
<td>3.0</td>
<td>11.2</td>
</tr>
<tr>
<td>Fontan procedure</td>
<td>3.0</td>
<td>5.8</td>
</tr>
<tr>
<td>Bidirectional Glenn</td>
<td></td>
<td>5.4</td>
</tr>
<tr>
<td>Systemic outflow obstruction</td>
<td>29.0</td>
<td>23.5</td>
</tr>
<tr>
<td>Coarctation</td>
<td>7.7</td>
<td>3.4</td>
</tr>
<tr>
<td>Transposition of great arteries</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Senning Arterial switch operation</td>
<td>7.0</td>
<td></td>
</tr>
<tr>
<td>LVOT repair</td>
<td>11.7</td>
<td>4.5</td>
</tr>
<tr>
<td>Norwood procedure</td>
<td>3.0</td>
<td>4.2</td>
</tr>
<tr>
<td>Pulmonary outflow obstruction</td>
<td>13.0</td>
<td>17.4</td>
</tr>
<tr>
<td>TOF repair</td>
<td>7.6</td>
<td>6.4</td>
</tr>
<tr>
<td>Conduit placement/revision</td>
<td>2.8</td>
<td>5.0</td>
</tr>
<tr>
<td>Other RVOT reconstruction</td>
<td>1.6</td>
<td>6.0</td>
</tr>
<tr>
<td>Pacemaker, AICD placement</td>
<td>5.0</td>
<td>4.3</td>
</tr>
<tr>
<td>Patent ductus arteriosis</td>
<td>8.0</td>
<td>5.1</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>15.0</td>
<td>14.1</td>
</tr>
</tbody>
</table>

AICD, automatic internal cardioverter-defibrillator; ASD, atrial septal defect; CAVC, complete atrioventricular canal; LVOT, left ventricular outflow tract; RVOT, right ventricular outflow tract; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

The future and finances

Tempora mutantur et nos in illis (time changes and we develop with time). It is 63 years since Robert Gross first ligated a PDA and we have seen amazing developments in the treatment of CHD. Concomitantly, anesthesiology has evolved and slowly defined pediatric anesthesia, then cardiac anesthesia, and now in the past two decades, pediatric cardiac anesthesia has developed as a distinct and separate area of subspecialization. There is no doubt that the current, “older” generation of pediatric cardiac anesthesiologists has played a major role in moving forward the whole field of treatment of CHD. Their contributions in adding to the knowledge of the physiology of CHD, the effects of anesthetic agents, and in enabling surgeons and cardiologists to develop new treatments are not always obvious or dramatic, but they are nonetheless important and essential parts of the progress made.

The last decade has seen many changes driven by the availability of new technology; these too provide new challenges
for the pediatric cardiac anesthesiologist to solve. Two-dimensional echocardiography has improved diagnosis both within and outside the OR and provided more challenges and opportunities for the pediatric cardiac anesthesiologist. Transesophageal echocardiography (TEE) is of special concern for the pediatric cardiac anesthesiologist. Its utility in congenital heart surgery was demonstrated in the late 1980s by the studies of several groups in Japan and the USA, including Russell and Cahalan at the University of California, San Francisco. The TEE interpretation of complex CHD and judgment of the adequacy of intraoperative repairs is considerably more challenging in CHD than in adult acquired heart disease. Many centers have called upon pediatric echocardiographers to make such judgments rather than have the pediatric cardiac anesthesiologist be responsible for that as well as for managing the patient in the post-bypass period. Also in contrast to adults, the TEE transducer may cause airway obstruction, alter left atrial pressure, or even extubate the child in the middle of an operation “under the drapes.”

Similarly, the emerging availability of cardiac MRI for diagnosis and follow-up of CHD patients has compounded the difficulties of providing anesthesia and monitoring in an intense magnetic field with limited patient access, but requiring anesthesia to be delivered to patients with severe, complex CHD under difficult conditions. Such technological advances come at a high price and it is hard to see how innovations like the long and expensive search for a method of treatment of HLHS would be justified today.

Another technical innovation of great importance driving pediatric cardiac anesthesia is extracorporeal membrane oxygenation (ECMO) (Fig. 1.2). Use of rapid response ECMO for children with CHD who suffer cardiopulmonary collapse postoperatively, cannot be weaned from CPB, or need to be supported as a bridge to heart transplantation has proven very effective in reducing mortality rates to significantly lower levels.

In the history of development of pediatric cardiac anesthesia, there is a long way between the baby in the icebath being prepared for DHCA and the complex technology necessary for ECMO resuscitation.

A significant challenge for the current generation of pediatric cardiac anesthesiologists is to help reduce the cost of care. One of the primary ways to reduce perioperative cost is limit ICU and ventilator time. This translates into increased demands and expectations for early extubation, preferably in the operation room.

Such changes in care have risks associated with them that will require careful assessment considering the advantages achieved with postoperative ventilation and sedation. For example, arrhythmias and cardiac arrest following endotracheal suctioning in the ICU postoperatively almost disappeared when heavy sedation with fentanyl prevented major swings in PAP with suctioning.51,52 Careful selection of patients for early extubation and judicious use of shorter acting anesthetic agents may allow lengths of stay to be shortened without increasing risks. In some studies early extubation after relatively simple operations has in fact proven to be safe when using new short-acting anesthetic agents like sevoflurane and remifentanil, particularly when better pain control is also employed.

Other advances such as limiting the total dose of anesthetic agents by developing ways to monitor depth of anesthesia, so as to give sufficient doses to prevent awareness and attenuate stress responses while avoiding awareness during CPB, are being explored, but remain elusive.53
In the past the outcome criterion most emphasized for treatment of CHD has been survival. Now that survival rates are very good and getting better for almost all forms of CHD, attention has turned to the quality of that survival. As part of that trend, the patient care group increasing most rapidly at most centers is the adult with CHD. This is the somewhat unexpected result as care in childhood improves and more and more of these patients survive to adulthood and even into old age. At many institutions special programs have been created to treat these patients and the problems they face. These problems include complications, reoperations, and socio-economic barriers to normal education, employment, and creation of families. The question of pregnancy and anesthetic management of delivery for these patients is also evolving. It is unclear who is most qualified to provide anesthesia for such patients during labor and delivery. But suddenly the pediatric cardiac anesthesiologist may have to care for adults.

Although much progress has been in the development of pediatric cardiac anesthesia to provide safe anesthetic care and improve outcome of treatment of CHD in the OR and catheterization laboratory for patients of all ages, much remains to be done. One can say with certainty that the intimate connection between advances in therapy, surgical or medical, and the anesthesia support services required to make those therapeutic advances possible will continue to present new challenges to the pediatric cardiac anesthesiologist. The pediatric cardiac anesthesiologists will in turn meet those challenges and in the process find ways to make still more improvements. Thus we progress in our art and science.

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