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# Tetralogy of Fallot

## The evolution of congenital heart surgery

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According to the American Heart Association (AHA), congenital heart defects (CHDs) remain the number one birth defect.<sup>1</sup> Each year, approximately 35,000 neonates are born with a CHD.<sup>1</sup> It's estimated that 1.3 million children and adults in the United States are diagnosed with a CHD.<sup>1</sup> Prior to 1940, neonates born with cyanosis were referred to as "blue babies."<sup>2</sup> These neonates either died shortly after birth or survived for a short period. Today, survival of "blue babies" isn't only possible, it's expected. The progression of management and treatment of the cyanotic neonate diagnosed with Tetralogy of Fallot has moved from palliation to complete repair. Anatomical repair is now promoted in neonates and infants with 95% survival into adulthood.<sup>3</sup> Advances in diagnosis and treatment of CHDs over the last 60 years, including diagnostic imaging, surgical techniques, anesthesia management, and cardiopulmonary bypass (CPB), have improved surgical outcomes. This article will present an overview of the evolution of cardiac surgery and current care for the neonate diagnosed with Tetralogy of Fallot (TOF).

### What's a CHD?

A CHD is a structural anomaly that develops during the embryological development of the heart and blood vessel structures within the first trimester of fetal development. The anatomical structures involved in a patient diagnosed with a CHD include the interior

walls of the heart (septa), heart valves (aortic, tricuspid, mitral, and pulmonic), or the external vessels carrying blood to and from the heart and lungs. Anatomical descriptions and pathology of CHDs date back to 1671.<sup>4</sup> Postmortem examination enabled anatomists and naturalists to describe normal cardiac anatomy and defects.<sup>4</sup>

CHDs can be described as cyanotic or acyanotic. Cyanosis results from any decrease in blood flow to the lungs. Heart failure results if there's too much blood flow to the lungs. The neonate may be symptomatic or asymptomatic depending on the severity and complexity of the defect. A complex combination of defects usually requires immediate medical and/or surgical intervention.<sup>5</sup>

### TOF

TOF is a cyanotic defect with four specific components: pulmonary outflow tract obstruction, ventricular septal defect, overriding aorta, and right ventricular hypertrophy (see *Tetralogy of Fallot*). TOF occurs in 3 of every 10,000 live births and is one of the most common forms of cyanotic heart disease.<sup>4</sup> First described in 1671, a series of reports published by Etienne-Louis Fallot in 1888 outlined the anatomy and described the nonrandom pathologic association of the four anatomic components.<sup>4</sup> He referred to this defect as "la maladie bleue." In 1924, the term "Tetralogy of Fallot" was first used by Maude Abbott, who based this nomenclature on Fallot's descriptions.<sup>4</sup>

In TOF, the level of pulmonary obstruction can occur at the infundibular muscle, pulmonary valve, main pulmonary artery, and branch pulmonary arteries. In severe forms of TOF, the pulmonary valve and the pulmonary artery structures are narrowed and/or underdeveloped (atretic). The degree of cyanosis correlates directly to the amount of blood flow that reaches the lungs for oxygenation. In the severest form of TOF, immediate medical and surgical interventions are required. According to Starr, 25% of infants born with severe pulmonary obstruction die in the first year of life without surgical intervention, and 40% die within 3 years.<sup>4</sup> The cause of death in surgically untreated patients is primarily related to periods of extreme cyanosis, usually caused by stress. These hypercyanotic spells are treated with knee chest position or oxygen and morphine (when available).<sup>4</sup>

### The early years

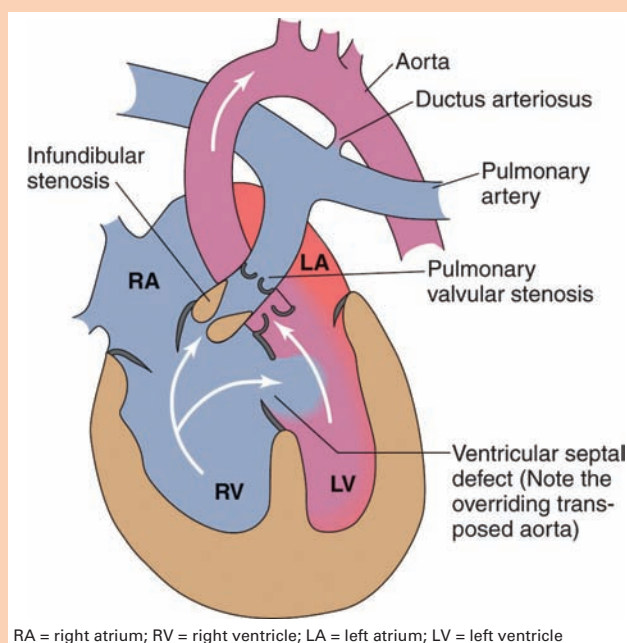
Surgery as a potential cure for CHD was once believed to be impossible. In 1896, the suturing of a stab wound to the right ventricle demonstrated that the heart muscle could hold sutures without tearing,

and touching the heart didn't automatically cause a lethal disturbance to the heart rhythm.<sup>6</sup> Opening the chest cavity without collapsing the lungs was an obstacle solved in 1900 with the advent of closed positive pressure ventilation.<sup>7</sup> Many surgeons were discouraged from pursuing cardiac repairs due to poor success rates.<sup>7</sup> Children with cyanotic heart disease had no other treatment option.

Postmortem descriptions of the heart led to the theoretical development of possible clinical syndromes associated with certain anatomical findings.<sup>2</sup> Limited methods of cardiac imaging made early diagnosis difficult to impossible. CHD diagnosis during the 1940s was primarily dependent on physical examination, electrocardiography, and fluoroscopy.<sup>6</sup> When the development of early fluoroscopy allowed a visual examination of the heart structures, Dr. Helen Taussig theorized that cyanosis was caused by an obstruction in the pulmonary artery.<sup>2</sup>

Once pulmonary structures could be visualized, surgical palliation became a possibility. Several surgical pioneers are credited with developing proposals for surgical correction of CHD. Dr. Robert Gross developed an operation to close a blood vessel in young children, known as ligation of the patent ductus arteriosus (PDA).<sup>8</sup> Dr. Taussig believed that the reverse, opening a blood vessel by means of creating a shunt, could relieve cyanosis.<sup>8</sup> Dr. Taussig's history of caring for cyanotic children over a period of 10 years afforded her the opportunity to witness the progression of cyanosis with this disease.<sup>4</sup> Her observations were that cyanotic children improved as long as their ductus arteriosus remained open versus those children whose ductus arteriosus closed early, causing death.<sup>8</sup> Dr. Alfred Blalock researched the bypass of occluded blood vessels. Together, Dr. Taussig and Dr. Blalock performed the first surgical palliation for TOF, a vascular bypass to relieve cyanosis, at Johns Hopkins Medical Center in Baltimore, M.D.<sup>2</sup> In 1944, after the success of the "blue baby" operation, the Blalock-Taussig Shunt (BTS) became a defining moment in the treatment of TOF and the development of surgical palliation of CHD. Today, neonates with severe

### Tetralogy of Fallot



Source: Rubin R, Strayer DS, eds. *Rubin's Pathology: Clinicopathologic Foundations of Medicine*. 6th ed. Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins; 2012:494.

cyanosis are treated with a BTS, often as an emergency intervention within the first month of life.<sup>8</sup>

In addition to the BTS, other surgeons began to look at additional surgical approaches to increase pulmonary blood flow. In 1946, Dr. Willis Potts developed a method for completing a side-to-side anastomosis of the descending aorta and left pulmonary artery.<sup>8</sup> Dr. David Waterston developed a similar approach with a side-to-side anastomosis of the ascending aorta and right pulmonary artery.<sup>8</sup> Because these shunts were created with direct anastomoses, both Potts and Waterston shunts were difficult to take down at the time of corrective surgery. Both of these surgical approaches were abandoned in favor of the BTS. Dr. William Rashkind developed a technique of inserting a balloon catheter across the patent foramen ovale in the atrial septum, thus, creating an opening for unoxygenated blood flow to the lungs and increasing oxygenation.<sup>8</sup> The balloon atrial septostomy (BAS) continues to be used early in the preoperative period to facilitate mixing of blood at the atrial level. Today, this sterile procedure is performed at the patient's bedside in the cardiac intensive care unit (CICU).

In the 1970s, the introduction of prostaglandin E-1 provided increased opportunities for early primary surgical repair in infancy.<sup>4</sup> The use of prostaglandin E-1 facilitates pulmonary blood flow to increase oxygenation and/or prevent metabolic acidosis.<sup>5</sup> Prostaglandin E-1 is administered in the immediate newborn period to maintain fetal circulation through the PDA. Prostaglandin E-1 is useful when there is a risk for premature closure of the ductus arteriosus. Many cyanotic and acyanotic lesions are dependent on ductal blood flow for oxygenation.

Surgical correction of CHD would not be possible without the development of CPB techniques. Prior to 1952, congenital heart surgery focused on extracardiac vascular structures.<sup>4</sup> The understanding of the relationship of the heart and blood oxygenation along with the discovery of heparin as an anticoagulant was crucial to the development of an



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extracorporeal machine that would support a patient during heart surgery.<sup>6</sup> In 1930, Dr. John Gibbons was a surgical fellow assigned to monitor a patient who had undergone an uneventful cholecystectomy 2 weeks earlier. The patient suffered a pulmonary embolism, became cyanotic, and slipped into unconsciousness. Dr. Gibbons believed that if there was a machine that could take over the function of the heart and lungs for a short period of time, life-saving surgery could be performed. Over the next 20 years, Dr. Gibbons

dedicated his career to the research and development of a heart-lung machine.<sup>6</sup> In 1952, his CPB machine allowed the successful closure of an atrial septal defect on an 18-year-old.<sup>6</sup> At that time, all cannulas and tubing were nondisposable. Assembly and disassembly, cleaning, and sterilizing the equipment were laborious tasks. Effective blood anticoagulation, development of oxygen membranes, and learning how best to drain the blood from the body and return the blood safely without air were some of the early challenges to CPB that led to successful patient outcomes. Anticoagulation, cannulation of the aorta for blood inflow, and cannulation of the superior and inferior vena cava to drain blood from the body remain the foundation of CPB. Once CPB machines and techniques were refined, the number of cardiac surgery procedures increased.<sup>6</sup> Research, innovation, and technical advances have enabled CPB use in smaller patients with lower morbidity. Smaller oxygenators, minimizing pump prime volumes, and new filtration techniques are all innovations that support safe neonatal surgical care. Today, patients as small as 4.4 lbs (2 kg) have been successfully placed on CPB for open-heart surgery.<sup>8</sup>

At the same time, Dr. Lillehei and his team were developing the use of cross-circulation as a way to perform surgery on intracardiac structures. This method involved a human donor with the same blood type on an adjacent operating table serving as a biological oxygenator.<sup>9</sup> Dr. Lillehei's initial results with controlled cross-circulation were better than results

using the first heart-lung machine.<sup>9</sup> Forty-five patients had surgery performed with controlled cross-circulation, and just under 50% of those patients survived 30 or more years.<sup>4</sup> The first successful repair of TOF was performed by Dr. Lillehei in 1954 using cross-circulation.<sup>6</sup>

Cardiac surgical procedures for management of TOF (developed in the 1960s) focused on a staged approach.<sup>8</sup> Early management of the patient diagnosed with TOF included one or two BTS procedures followed by complete repair in early childhood.<sup>3</sup> The early outcomes for complete repair depended on the severity of cyanosis, symptoms, and the effectiveness of the shunt performed to increase pulmonary outflow.<sup>8</sup> The mortality of surgical repair and CPB at that time was around 60%.<sup>4</sup> As experience with shunt procedures grew and complications related to CPB decreased, the awareness of morbidity and mortality associated with distortion of the anatomy, thrombosis formation, and difficulty taking down anastomoses made primary repair more appealing in select patients. Many early repairs required a right ventriculotomy and a patch on the right ventricular outflow tract to relieve stenosis.<sup>4</sup> Improvements in surgical technique including myocardial protection, preservation of coronary anatomy, prevention of coronary embolism, and maintenance of adequate cardiac output in the postoperative period not only improved outcomes but allowed for increased survival in patients under 5 years of age.<sup>4</sup> During the 1970s, complete repair of simple TOF during the neonatal period was accomplished with relatively low mortality using hypothermia and deep hypothermic cardiac arrest.<sup>4</sup> The benefits of early primary repair minimized potential abnormal development of the heart and lungs exposed to the altered blood circulation and gas exchange of TOF.<sup>8</sup> Today, both early and late survival rates for complete primary repair have improved to greater than 98%.<sup>8</sup>

The evolution of cardiac catheterization and imaging techniques to facilitate diagnosis of CHD occurred in parallel to developing surgical techniques.<sup>7</sup> Many of



**Adding the fetal US to prenatal care increases early diagnosis of CHD by 50%.**

the first cardiac surgical procedures were performed without a complete diagnosis or an understanding of the anatomical structures. Without the more precise diagnostic techniques of cardiac catheterization and ultrasound (US), initial surgical mortality was high in congenital heart surgery. Historical advances in these technologies won't be discussed in detail here; however, the contributions of diagnostic imaging CHD cannot be understated.

At the time of early operations, anesthesia was accom-

plished using an open drop ether mask. Endotracheal tube sizes made intubation for a neonate or small child nearly impossible.<sup>7</sup> Intraoperative bronchospasm was treated with direct tracheal intubation. Preoperative hematocrit levels for patients with cyanosis could reach as high as 80%, thus, placing them at risk for thrombosis. Without pulse oximetry, auscultation of breath and heart sounds was the only means of assessing pulmonary blood flow and oxygen saturation.<sup>7</sup>

The early development of techniques used by intradisciplinary surgical teams needs to be appreciated. Each innovation is significant in the advancement of congenital heart surgery to primary repair in the neonatal period. The focus of these early efforts was as much on preventing mortality as on improving techniques. In addition, the costs associated with medical treatment and observation before and after palliative procedures had both financial and psychosocial impact on the family.<sup>8</sup>

### **Prenatal care**

Improvements in US imaging now allow for early prenatal diagnosis of CHD. Nearly 80% of patients diagnosed with a CHD have no associated risk factors.<sup>10</sup> A routine US that includes a four-chamber view of the fetal chest is recommended as a standard of care during the middle of the second trimester (16 to 20 weeks).<sup>10</sup> Adding the fetal US to prenatal care increases early diagnosis of CHD by 50%.<sup>10</sup> Fetal intervention that may improve long-term outcomes for the child may be an option to consider in a certain few diagnoses. If a CHD is suspected,



a referral for a fetal echocardiogram will provide additional details of cardiac anatomy, function, and rhythm.<sup>10</sup> At birth, understanding the presence of a CHD can improve postnatal outcomes. Early diagnosis allows for development of birth plans, discussion with a cardiologist about treatment options, and parent education with nursing before the child's birth.<sup>10</sup>

If heart structures are too small to be clearly visualized by prenatal US, the time of diagnosis is delayed into the first days and/or weeks of life. Late diagnosis of CHD carries the added burden of increased associated morbidity and mortality. In this circumstance, parents/guardians need to make decisions during a very stressful time for the family.

### **Preoperative management**

The preoperative condition of the neonate will directly impact postoperative outcomes. The goal of preoperative medical management focuses on maintaining adequate cardiac output and peripheral perfusion, balancing pulmonary and systemic blood flow, and maintaining normal sinus rhythm without signs of heart failure. Limited cardiorespiratory reserves and immature organ function—coupled with the pathophysiology of CHD—need to be considered in care and treatment of the neonate diagnosed with TOF.

The transition of fetal circulation during the neonatal period is critical. Using prostaglandin E-1 to maintain a PDA and using an echocardiography-guided BAS are two advances that support adequate oxygenation preoperatively.<sup>5</sup> Dysrhythmias can result in poor cardiac output, since A-V synchrony contributes to 30% of cardiac output. Symptoms of heart failure may be managed with the use of diuretics and inotropic support. Hypercyanotic episodes or “tet-spells,” beginning in infancy throughout toddler age, are managed with knee chest positioning, oxygen administration, and morphine. Tet-spells are caused by an imbalance of pulmonary to systemic blood flow, and if left untreated, can result in syncope and death.<sup>4</sup> Tet-spells can be precipitated by agitation and dehydration. In more severe cases, additional circulating blood volume and endotracheal intubation to improve ventilation and oxygenation may be needed. Other treatment options include neosynephrine to increase peripheral vascular resistance, correction of metabolic acidosis, and, ultimately, surgical palliation and/or correction. This level of critical care for a neonate/infant who is diagnosed with TOF is

reserved for the CICU preoperatively. CICU nursing staff have additional education and patient care management experience, including anatomy and physiology of CHDs, repairs, effects of cyanosis, and pulmonary hypertension on patient outcomes, medical therapies, and the unique challenges of patients living with CHDs across the lifespan.<sup>11</sup> The preoperative management of the newborn diagnosed with a CHD has added new challenges to the CICU setting.

Having a newborn diagnosed with a CHD places an enormous stress on the family.<sup>12</sup> Disruption to the parenting role, feeling powerless, and delayed parent-infant bonding adds to parental anxiety.<sup>12</sup> Family teaching begins on admission based on the assessment of learning needs and readiness to learn. Stress can prevent the parents from completely understanding the child's diagnosis and surgical plan.<sup>12</sup> Caring for an infant at home with complex medical needs can lead to parents becoming fearful and over-protective of their child. Multiple follow-up visits may be required. These stressors can have a significant impact on a young family, including divorce.<sup>12</sup>

### **Intraoperative nursing care**

The role of the cardiac perioperative nurse has adapted to evolving patient care needs. During the 1960s and 1970s, the primary concern was whether or not a child would survive surgery for a CHD. The evolution of CPB improved surgical outcomes, and the possibility of novel surgical approaches was recognized. As patient survival improved, preventing complications related to patient care in the OR became an integral part of intraoperative care. With each surgical success, the boundaries of potential surgical repairs expanded, and the patients undergoing open-heart repairs were younger with more complex defects. During the 1980s, the need for formalization of specialized teams for intraoperative care became evident.<sup>13</sup> Nurses relied on available knowledge related to perioperative care. New surgical procedures and techniques with considerations for the patient and family led nurses to rely on interactions with surgeons, anesthesiologists, and perfusionists to advance knowledge.<sup>13</sup> The consistency of performing cardiac operations is critical to develop the ability to anticipate needs of the surgical team and support safe patient care. The cardiac perioperative nurse needs to maintain up-to-date knowledge of acute and critical patient care needs.

Highly-skilled, knowledgeable, specialized professional nurses are now expected to have an understanding of complex cardiac defects, anomalies, and repairs that support coordinated quality care and a systematic plan for error reduction.<sup>13</sup> Ongoing education related to CHD and certification in their respective specialty (certified nurse in OR) reinforces knowledge, enhances quality patient care, and strengthens new experiences. A dedicated team in the cardiac OR ensures patient safety through standardization of practice.

As the number of patients undergoing congenital heart surgery in the newborn period increased, the complexities and focus of care for the cardiac perioperative nurse transformed from a task-oriented approach to an evidence-based role. Cardiac perioperative nurses assume primary care of the patients and collaborate with the interdisciplinary team of surgeons, anesthesiologists, and CPB perfusionists. Their role involves patient assessment, implementing processes, and evaluating outcomes. The cardiac perioperative nurse is a clinical leader, charge nurse, data collector, and facilitator of new innovation involving equipment and innovative surgical procedures. Critical thinking allows the cardiac perioperative nurse to anticipate potential surgical and anesthetic challenges that impact patient outcomes.

The cardiac perioperative nurse provides safety and comfort for the patient and family by humanizing the surgical environment and establishing a therapeutic nurse-family relationship within a limited time frame. Parents of a child diagnosed with a CHD are under stress related to the cardiac diagnosis and fear of the unknown. Depending on the timing of diagnosis and severity of the defect, parents face a loss of control over their child's life and the possibility of losing their child. Even though caring for families is part of the daily routine for cardiovascular nurses, it's essential for the cardiac perioperative nurse to support parents and gain their trust. Answering questions, educating parents on the plan for care on the day of surgery, and maintaining a connection throughout the procedure helps provide ongoing family support. Their responsibilities also rest with a continuous, ongoing communication strategy with parents and families. The cardiac perioperative nurse needs to be positioned to facilitate difficult information given to families at a minute's notice. The cardiac perioperative nurse

supports education and information relative to the CHD, interprets and clarifies patient and family questions, and empowers patients/families to make informed decisions. Provision and facilitation of information is one of the most common skills of any cardiac perioperative nurse.<sup>14</sup>

Along with these responsibilities, the cardiac perioperative nurse is accountable for the quality of care he or she provides for the patient and family and the technical knowledge required. The anesthetic management of neonates undergoing cardiac surgery requires a clear understanding of neonatal respiratory and cardiac physiology.<sup>5</sup> Many neonates arrive at the OR intubated with indwelling monitoring lines and inotropic support. The cardiac perioperative nurse continually assesses the patient's hemodynamic status and assists the anesthesia team during patient transfer, induction, and line placement. In the neonate and small child, hypotension at any point during surgery can be related to the amount of circulating blood volume. Hemodynamic changes need to be addressed quickly and effectively with volume replacement.

Preventing skin and tissue injury is a key phenomenon of concern for the cardiac perioperative nurse.<sup>15</sup> Patients undergoing cardiac surgery are at risk for developing pressure-related tissue injury. A head-to-toe, front and back skin assessment is completed prior to patient positioning and prior to patient transfer to the CICU. Skin integrity and circulatory compromise are noted and documented. In addition to occiput, elbows, and heels, all intravenous lines, tubing, and other devices are protected with gel pads to prevent pressure on the skin surface. Endotracheal tube, nasogastric tube, and blood pressure cuffs are a few examples of devices that can cause pressure-related injury during the intraoperative phase of care.

A comprehensive, interdisciplinary team approach to prevent surgical site infections includes standardization of practice to optimize outcomes.<sup>16</sup> Preoperative antibiotics are administered within 60 minutes of the skin incision. Appropriate preoperative surgical scrub is performed the night before surgery, and infants over 2 months of age are prepped with 2% chlorhexidine gluconate and 70% alcohol.<sup>16</sup> The neonate will often need a delayed sternal closure to manage post-operative hemodynamic instability.<sup>17</sup> Post-bypass edema and post-surgical bleeding can cause cardiac tamponade, leading to cardiac arrest.<sup>17</sup> Maintenance of an occlusive dressing around the sternum and strict

adherence to sterile technique continues after transfer to CICU care. Education of the CICU staff related to aseptic practice is the responsibility of the cardiac perioperative nurse. Patients are continuously monitored for breaks in sterile technique and signs of infection.

Normal sinus rhythm is essential to provide adequate cardiac output in neonates. Any alteration in heart rhythm will diminish cardiac output. Heart block, ectopic atrial tachycardia, and junctional heart rhythms are a few examples of rhythm irregularities seen in the neonate/infant diagnosed with CHD. A temporary pacemaker, temporary wires with pacing cables, and defibrillator are checked and readily available in the OR. If a patient has episodes of low cardiac output, persistent hypoxia, or cardiac arrest, extracorporeal membrane oxygenation (ECMO) may need to be initiated in the CICU or the OR as part of cardiac resuscitation.<sup>18</sup>

Hypothermia reduces metabolic rate and decreases tissue oxygen consumption. Body surface cooling is used as an adjunct to bypass induced hypothermia. Myocardial preservation, cerebral perfusion, and renal and hepatic function are of particular concern related to CPB. During the 1970s, the use of hypothermic circulatory arrest resulted in improved outcomes in neonates and infants.<sup>8</sup> An ice bath was used to reduce core temperature to 86° F (30° C).<sup>5</sup> Refinements in current OR equipment facilitates cooling on bypass. Cooling and rewarming of surface temperature is achieved through the use of ice packs, cooling/warming water flow blankets, ambient air temperature regulation, and convective warming devices. Warmed I.V. fluids and irrigating solutions are used to aid in return to normothermia. The patient's temperature is monitored using rectal, esophageal, or tympanic temperature probes.

Early primary repair led to high morbidity and mortality in the neonatal age group. It was believed that complete repair of heart defects that have no probability of resolution in the neonatal



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period is preferred.<sup>3</sup> However, a controversy regarding the timing of a complete repair versus palliation remains. The degree of cyanosis in a patient with TOF is directly related to the amount of pulmonary blood flow. The trajectory and degree of cyanosis has direct implications for surgical timing.<sup>3</sup> If the patient is coming from home, knowing the patient's response to feeding and agitation will provide insight into potential tet-spells. Parents know their child better than anyone and are the healthcare team's greatest resource. The cardiac perioperative nurse continuously

assesses oxygen saturations and visual notation of signs of cyanosis. In extreme cases, the older child with an unrepaired TOF may demonstrate significant clubbing of fingers and toes. The stress of parental separation, premedication, and anesthesia induction can all lead to a tet-spell. Patients with high hematocrit levels who have no fluid intake preoperatively are also at high risk for increased cyanosis and tet-spells. The cardiac perioperative nurse needs to be vigilant with ongoing assessments until bypass is initiated.

### **Postoperative transfer of care**

The cardiac perioperative nurse is dedicated to the cardiac surgical patient's continuing care. Close monitoring of the patient continues through the postoperative period in the OR and during transfer to the CICU. Intracardiac monitoring lines are placed intraoperatively to assess for adequate cardiac output. Inotropic agents and vasoconstrictors, along with intravascular volume to support myocardial contractility and cardiac output, are included in clinical handoff.<sup>19</sup> Temporary pacing wires and pacemakers are used to manage heart rhythms that support adequate cardiac output. Patients are mechanically ventilated postoperatively until hemostasis is achieved and/or hemodynamics allow for primary and delayed chest closure. A clinical handoff is completed at the bedside with the interdisciplinary team: cardiac surgeons, anesthesiologists, cardiac perioperative nurses, ICU nurses, respiratory



therapists, intensivists, nurse practitioners, and cardiologists. A formal handoff at the time of patient transfer is essential for continuity of patient care and effective postoperative decision making for the CICU interdisciplinary team.<sup>18</sup> A brief history and synopsis of events from induction of anesthesia through surgery and bypass until postoperative transfer to the CICU is presented by the appropriate discipline. The patient's current physical status and the anticipated plan for care are discussed, and questions are encouraged. Parents are considered equal

partners in providing care to their child. Parent presence is a 24/7 part of care in the CICU. Over the past 25 years, family-centered care has evolved to include parent presence at the bedside, including intensive care units.<sup>20</sup>

Technological advancements and the ability to provide postoperative surgical treatment at the patient's bedside allow for a rapid response to changing clinical situations. The practice of performing emergency procedures during the postoperative course in the CICU eliminates the time taken for transportation to the OR during a crisis. In the case of cardiac arrest, rapid resuscitation includes the use of ECMO to support heart and lung function for a limited period of time. Despite the original intention of ECMO design to support respiratory failure in neonates, the presence of an oxygenator in the ECMO circuit provides the ability to support biventricular function and gas exchange. One of the early, successful reports of ECMO use in a post-op cardiac surgical patient was that of a 4-year-old girl suffering from hypoxemia following a TOF repair in 1973.<sup>18</sup> A better understanding of ECMO management in the 1980s led to an increased use of ECMO support for cardiac failure. Today, sterile supplies and instruments are available in the CICU for the cardiac perioperative RN to prepare for bedside surgical procedures, including rapid resuscitation with ECMO support, chest exploration for tamponade and bleeding, complications for intracardiac line removal, and secondary chest closure. Additional education, collaboration, and effective



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teamwork in expanding CICU and cardiac perioperative nursing roles are crucial for improving patient outcomes after cardiac surgery.

### Survival across the life span

Advances presented have led to increasing survival rates for neonates born with a CHD. Early survival rates for cardiac surgical patients having a primary repair is reported at 98% to 100%.<sup>4</sup> The population of adults now living with CHD increased to 1 million people in the United States in 2008.<sup>1</sup> New challenges

to the healthcare team include transitioning services from pediatric to adult cardiology, care for medical follow-up, and ongoing education for the patient relevant to their condition and impact on their adult life.<sup>11</sup> Occupational choices, health insurability, dental hygiene, appropriateness of contraception, and risks of pregnancy are issues unique to this growing population.<sup>11</sup> The natural progression of aging can add to the morbidities of later life. Coronary artery disease, dysrhythmias, pulmonary vascular disease, and declining myocardial function increase the risk of heart failure.<sup>11</sup> The most common need for reoperation is late pulmonary valve replacement.<sup>8</sup> Careful and consistent medical management by a specialized team is needed to meet the diverse needs of this growing population.

### Moving forward

While CHDs remain the number one birth defect, techniques in diagnosis, medical management, and surgical correction are credited for reducing mortality, especially in blue babies. The collaborative efforts of early innovators proved to have an essential role in the evolution of patient care and treatment of patients diagnosed with CHD. Advancements discussed have been pivotal in caring for neonates who are diagnosed with TOF. Today, patient outcomes following repair of TOF are used as a benchmark for quality.<sup>8</sup> Recognizing the patient as the center of all activity has challenged previous assumptions for care and treatment. The primary concern of cardiac surgery for patients

diagnosed with a CHD has moved from immediate survival to survival across the life span. **OR**

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