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Pediatric Cardiac Emergencies

Maria C Yates[1] and P Syamasundar Rao[2]*

[1]*Pediatric Cardiology Fellow, University of Texas/Houston Medical School and Children's Memorial Hermann Hospital, Houston, TX, USA*
[2]*Professor of Pediatrics & Medicine, Director, Pediatric Cardiology Fellowship Programs and Emeritus Chief and Interim Director of Pediatric Cardiology, University of Texas/Houston Medical School, and Children's Memorial Hermann Hospital, Houston, TX, USA*

Abstract

With the advent of and advances in diagnostic methods and transcatheter and surgical treatment modes, patients with Congenital Heart Defects (CHD) are surviving longer. Cardiac defects that were once fatal in infancy are now treatable and patients are surviving into adulthood. However, these patients are at increased risk of acute events and therefore, likely to frequent emergency rooms. These patients should now be added to the group of CHD patients that would ordinarily come thru' the emergency rooms. Presented in this review is a selection of the more common life-threatening pediatric cardiac emergencies likely to be encountered in the emergency department. The identification and management of supraventricular tachycardia, hyper cyanotic spells, congestive heart failure, emergencies in the palliated patients with a functional single ventricle and neurologic events related to congenital heart disease are discussed.

Keywords: Pediatric cardiac emergencies; Arrhythmia; Supraventricular tachycardia; Cyanotic spells; Congestive heart failure; Cerebrovascular accidents; Tetralogy of Fallot; Hypoplastic left hear syndrome; Single ventricle; Modified Blalock-Taussig shunt; Bidirectional Glenn; Fontan surgery; Neurologic events

Abbreviations: CHD: Congenital Heart Disease; CHF: Congestive Heart Failure; CVA: Cerebro Vascular Accident; ECMO: Extracorporeal Membrane Oxygenation; IV: Intravenous; PGE_1: Prostaglandin E_1; SVT: Supra Ventricular Tachycardia; TF: Tetralogy of Fallot; VSD: Ventricular Septal Defect; WPW: Wolff-Parkinson-White syndrome

Introduction

There are many types of cardiac emergencies in neonates, infants, and children. While some of these may eventually require treatment at a center staffed with specialists in pediatric cardiology and pediatric cardiovascular surgery, the initial recognition of the emergency, stabilization and management do fall on the emergency physician when a child presents to the emergency room.

While any child may have a cardiac emergency, patients with structural Congenital Heart Disease (CHD) are at increased risk for developing acute life-threatening problems. The incidence of CHD in the general population is approximately 1% of live births [1]. In the past few decades, advances have been made in the medical and surgical management of the more severe forms of CHD. Because of this, there are now more survivors with severe forms of CHD. These patients may frequent the emergency rooms, and their distinctive physiology requires understanding by the care-givers so as to provide adequate and appropriate therapy. Moreover, they may have emergencies related not only to the cardiovascular system, but also neurological, gastrointestinal, infectious and other systems.

There are many pediatric cardiac emergencies as reviewed elsewhere [2,3] and are listed in Table 1. The purpose of this review is to discuss the recognition and management of the more common, selected, life-threatening pediatric cardiac emergencies that are likely to be encountered in the emergency rooms. Discussion of management of pediatric cardiac arrest will not be included; the reader is referred to the American Heart Association, 2010 Guidelines for Pediatric Basic Life Support [4] for discussion of this subject. Similarly discussion of cyanotic newborn is not included since these babies typically present in the hospital setting and not to an emergency room. Interested reader is referred elsewhere [3] for management of neonatal cyanosis.

Arrhythmias

Several types of arrhythmias are seen; however, supra-ventricular tachycardia (SVT) is the most frequent arrhythmia both in the neonatal period and childhood and will be discussed in this review. SVT is the most common arrhythmia of the pediatric age group requiring medical treatment [5]. Although patients with CHD may develop SVT, it is more commonly seen in children with structurally normal hearts. The heart rate in SVT frequently exceeds 250 beats per minute in the neonates, above 220 in infants and over 150 beats per minute in older children. It is usually characterized by a narrow QRS complex (Figure 1), unless there is aberrant ventricular conduction (Figure 2). Most SVTs (70%) are reentrant in type, involving an accessory pathway (Wolff-Parkinson-White syndrome {WPW}); if the pathway manifests antegrade conduction of the impulse from the atria to the ventricle, a short PR interval and a delta wave (Figure 3) are seen on a surface electrocardiogram; however, it should pointed out that WPW pattern is not seen while the child is in SVT. It is seen only after conversion to sinus rhythm. If the pre-excitation is not present on ECG, the pathway may be concealed or there is an alternate mechanism.

1. Cardiopulmonary arrest
2. Shock
3. Arrhythmias
4. Hypercyanotic ("Tet") spells
5. Cyanosis in the newborn
6. Congestive heart failure
7. Cardiac emergencies in the patient with a functional single ventricle
8. Cerebrovascular accidents
9. Brain abscess

Table 1: Commonly encountered pediatric cardiac emergencies.

***Corresponding author:** P Syamasundar Rao, Professor and Emeritus Chief, Division of Pediatric Cardiology, University of Texas/Houston Medical School, 6410 Fannin, UTPB Suite# 425, Houston, TX 77030, USA, Tel: 713-500-5738; Fax: 713-500-5751; E-mail: P.Syamasundar.Rao@uth.tmc.edu

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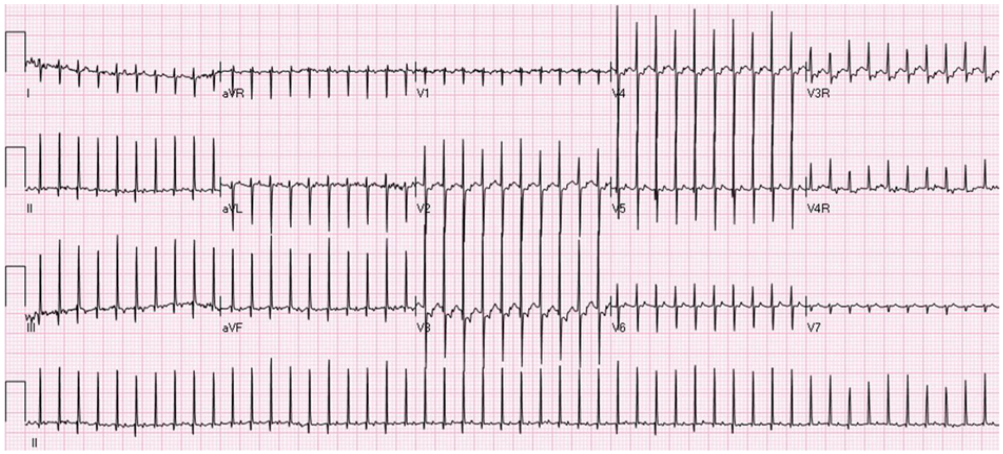

Figure 1: Electrocardiogram of an infant with Supraventricular tachycardia. Note the heart rate is approximately 260 beats per minute and the QRS duration is very short (narrow) and no definitive P waves were seen.

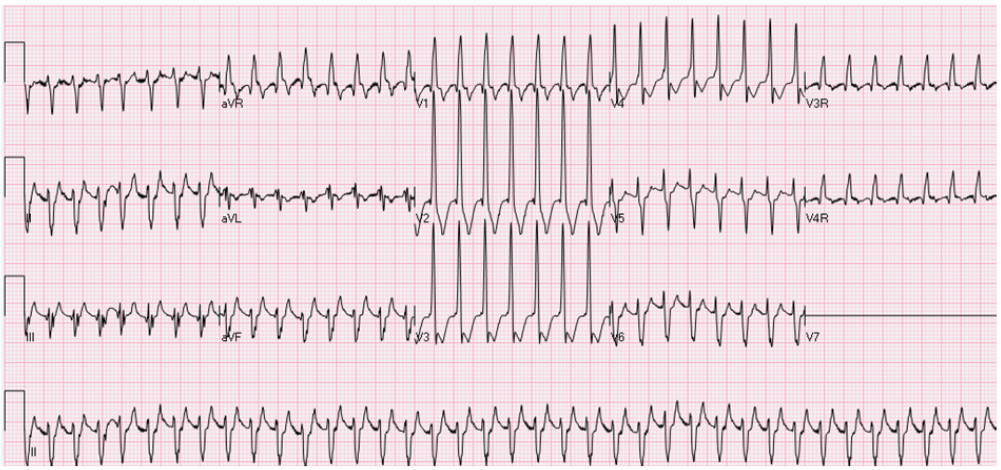

Figure 2: Electrocardiogram on another infant with Supraventricular tachycardia, but with aberrant ventricular conduction; note the wide QRS complexes.

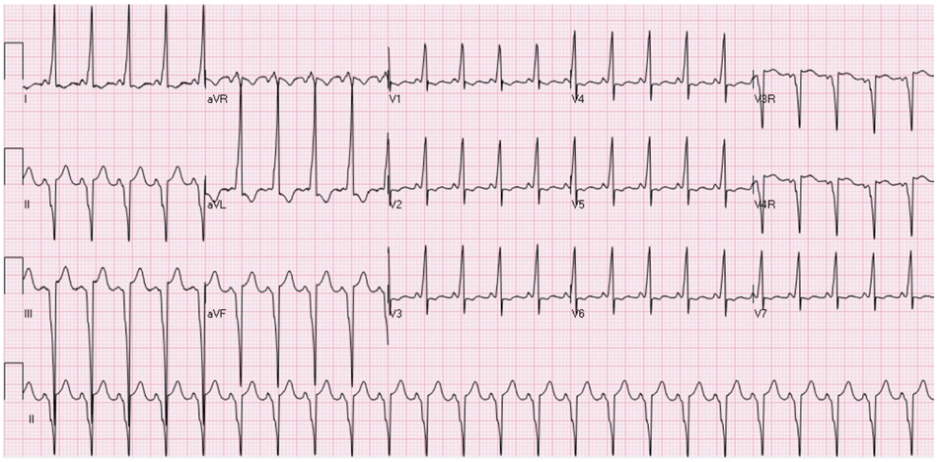

Figure 3: Electrocardiogram on an infant following conversion of Supraventricular tachycardia to sinus rhythm exhibiting Wolff-Parkinson-White syndrome with short PR interval and delta waves in most leads.

After confirmation of the diagnosis of SVT with ECG, treatment should be instituted; the type of treatment depends on the patient's hemodynamic status at the time of presentation.

For the hemodynamically stable patient with adequate cardiac output, vagal maneuvers may be attempted prior to intervention with medications. In the neonate and infant, this involves placing ice (crushed

ice in a plastic bag) gently on the face, covering the eyes but taking care not to occlude the airway. Another option is rectal stimulation with a rectal thermometer or insertion of nasogastric tube to simulate Valsalva. In the older child, vagal maneuvers are similar to adults and may include Valsalva maneuver, placing the face in a tub of ice water, or carotid massage. We do not recommend eye ball massage. If the arrhythmia does not resolve within 10-15 seconds of vagal maneuvers and the patient remains stable, intravenous adenosine may be administered. The intravenous line used for adenosine administration should be as centrally located as possible; an upper extremity line is preferred over a lower extremity line. The dose of adenosine is 100 mcg/kg; administered by rapid IV push. If not effective, the dose may be increased by increments of 50 mcg/kg (maximum 6 mg) until conversion. Adenosine's mechanism of action is to cause a brief blockade of conduction at the AV node, lasting approximately 10-15 seconds [6]. If there is a wide QRS pattern to the SVT, this may indicate antegrade conduction via an accessory pathway. If the AV node is blocked, there is a small risk of ventricular tachycardia due to rapid ventricular activation after AV nodal blockade. Therefore, whenever adenosine administered it is prudent to have resuscitation equipment available. If adenosine is not effective, other medications may be administrated; the options include: (a) Esmolol at bolus dose of 200 to 400 micrograms/kg over 10 minutes, followed by 75 mcg/kg; (b) Verapamil 0.1-0.2 mg/kg by very slow intravenous infusion (Verapamil is not recommended in infants less than 1 year. One may consider prior calcium gluconate 100 mg/kg IV slowly); (c) Procainamide (loading dose of 7 to 15 mg/kg given over a 30 to 45 minutes followed by a maintenance dose of 40 to 50 mcg/kg/minute) and (d) Amiodarone (bolus infusion of 5 mg/kg infused over a 20 to 60 minutes followed by continuous infusion of 10 to 15 mg/kg/day). Item a followed by item d seem to be preferred by most at the current time.

For the hemodynamically unstable SVT patient, or in the stable patient in whom the above interventions were unsuccessful, synchronized cardioversion with 0.5 to 2 watt-second/kg should be attempted (Table 2).

After conversion to normal rhythm, starting medications to prevent recurrence is important. In the past, oral digoxin (10 mcg/kg/day in two divided doses) for 6-12 months for neonates and infants used to be the standard practice [7,8]; however, more recently, because of concern for reentrant tachycardia, Propranolol 1-4 mg/kg/day by mouth in 3 to 4 divided doses has become the norm. In children, long acting beta blockers such as Atenolol (1 mg/kg/day as a single dose) for 6-12 months

1. Hemodynamically stable
a. Vagal maneuvers (ice to face, Valsalva)
b. Adenosine 100 mcg/kg rapid IV push; may be increased to 200 mcg/kg in 50 mcgincrements until response
c. Esmolol at bolus dose of 200 to 400 micrograms/kg over 10 minutes, followed by 75 mcg/kg
d. Amiodarone (bolus infusion of 5 mg/kg infused over a 20 to 60 minutes followed by continuous infusion of 10 to 15 mg/kg/day)
e. Procainamide (loading dose of 7 to 15 mg/kg given over a 30 to 45 minutes followed by a maintenance dose of 40 to 50 mcg/kg/minute)
f. Verapamil* 0.1 mg/kg slow IV push; may increase to 0.2 mg/kg in 15 minutes if no response (maximum dose 5 mg)
2. Hemodynamically unstable
a. Immediate synchronized DC Cardioversion at 0.5 to 2 Joules/kg
b. Transesophageal pacing§

*Verapamil should not be used in patients under 1 year of age, those taking beta-blockers, or signs of congestive heart failure
§Should only be done by an experienced cardiologist

Table 2: Management of supraventricular tachycardia.

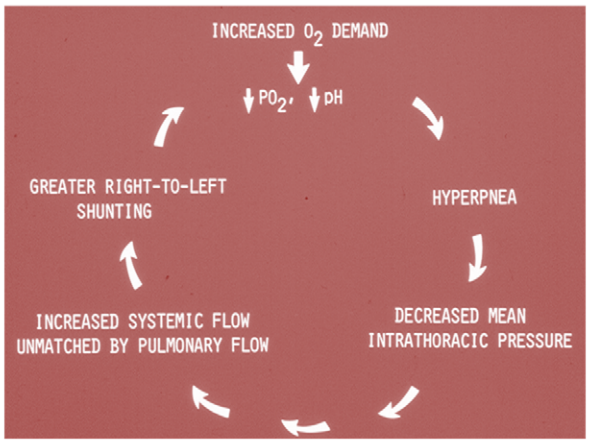

Figure 4: The mechanism of hyper cyanotic spells as proposed by Guntheroth [11]. Hyperpnea reduces mean Intrathoracic pressure, which decreases systemic and pulmonary resistances. Decreased systemic resistance is not matched with increased pulmonary flow because of dominant right ventricular outflow tract obstruction.Thus, there is even greater right-to-left shunt, further decreasing the PO_2 and pH and thus a vicious cycle.

may be employed. Discussion of the use of other medications such as Flecanide, Sotalol and Amiodarone either alone or in combination and the need for and timing of radiofrequency ablation for prevention of recurrences is beyond the scope of this presentation.

Hypercyanotic (Tet) Spell

Hypercyanotic or "Tet" spell may occur in infants and children with unrepaired tetralogy of Fallot and other cardiac defects with large inter-ventricular communication and pulmonary outflow tract obstruction. The spell is manifested by increase in rate and depth of respiration (hyperpnea) and cyanosis and may progress to limpness and syncope; however the babies usually recover subsequently. These spells are usually self-limited but may rarely lead to serious complications such as syncope, seizure-like episodes, cerebrovascular accidents, or even death [9]. The spell may last for a few minutes to a few hours. These spells are most usually seen between the ages of 1 and 12 months, with a highest frequency between the ages of 2 and 3 months. The spells may occur at any time of the day, but are most frequent in the morning after waking from sleep; defecation, crying and feeding are usual precipitating factors. Examination reveals an agitated and cyanotic infant and a previously heard cardiac murmur of right ventricular outflow obstruction is either absent or markedly decreased in intensity during the spell.

The mechanism causing the spells is not completely understood, but is believed to be an acute spasm of the right ventricular outflow tract (infundibulum) [10], resulting in markedly decreased pulmonary blood flow along with right-to-left shunting across the ventricular septal defect (VSD); the infundibular spasm may be precipitated by acute increase in endogenous catecholamines. An alternative and more plausible explanation is paroxysmal hyperpnea [11]. During sleep oxygen consumption is low and normal acid base balance is maintained. When the baby wakes up, the O_2 consumption increases rapidly and may produce a slight acid base imbalance. Normally the respiratory center brings the imbalance back to normal. However, when a sudden increase in activity with resultant marked increase in oxygen consumption occurs prior to the above described modification takes place, decrease in PO_2 and pH and increase in PCO_2 trigger a hyperpnea

response from the respiratory center and enter a vicious cycle (Figure 4) [11,12]. Whatever is the mechanism, the inciting factors appear to be agitation, intercurrent illness, dehydration and invasive procedures without adequate prior sedation.

For a cyanotic spell, it may seem reasonable to administer oxygen. Since the mechanism of hypoxemia is limited pulmonary blood flow and not alveolar hypoxia, oxygen may be of limited benefit. Oxygen via simple face mask may be administered. However, if this further agitates the child, then the oxygen should be removed as the agitation will worsen the spell more than the oxygen will help.

The first maneuver to attempt is the "knee-chest" position. Take the child's legs and bend the knees to be up against the chest. This will increase systemic vascular resistance and decrease right to left shunting at the VSD level.

Intravenous fluid boluses of normal saline at 10 cc/kg may increase right ventricular preload and therefore might increase right ventricular output. Care should be taken, however, not to cause fluid overload. Also, correct any metabolic acidosis that may be present. If the spell persists, then giving medications to calm the child may help. Morphine maybe given to help with sedation but also with decreasing the respiratory drive and therefore decrease patient's hyperpnea which may be further exacerbating the spell. If the spell continues to persist, then increasing systemic vascular resistance with phenylephrine or norepinephrine may be attempted.

If the patient remains hypoxemic, then intravenous beta blockade may be attempted. The mechanism behind the action of this class of drugs is not completely understood, but is believed to be a combination of relief of the infundibular spasm causing decreased right ventricular outflow obstruction, prevention of decrease in systemic vascular resistance and/or prevention of ventilatory response (hyperpnea) to hypoxia, all through beta adrenergic blockade. Propranolol may be given by a slow intravenous infusion at a dose of 0.1 mg/kg/dose. Alternatively, Esmolol may be given as a loading dose of 500 mg/kg/dose followed by a maintenance infusion of 50-100 mcg/kg/min.

Rarely, emergency systemic-to-pulmonary artery shunt (Blalock-Taussig) or even extracorporeal membrane oxygenation (ECMO) may be required for refractory hypercyanotic spells (Table 3).

1. Knee-to-chest position
2. Oxygen via face mask (discontinue if causes agitation in the patient)
3. Morphine 0.1 mg/kg intramuscular or intravenous
4. Normal saline bolus 10 mL/kg intravenous
5. Esmolol loading dose of 500 mcg/kg/dose followed by an infusion of 50-100 mcg/kg/min.
6. Phenylephrine 5-10 mcg/kg intravenous (maximum dose 5 mg) or Norepinephrine as a continuous infusion of 0.05 to 0.5 mcg/kg/min (the rate of infusion should be adjusted to increase the systolic blood pressure by 15 to 20% of the control value)
7. Correct any acidosis with sodium bicarbonate 1-2 mEq/kg intravenous
8. Correct any electrolyte imbalance
9. Persistent or refractory spells may require emergency Blalock-Taussig shunt or ECMO

Table 3: Management of hypercyanotic spells.

Congestive Heart Failure

There are numerous conditions that may precipitate Congestive Heart Failure (CHF) in children; the most common are large left to right shunts, such as VSD, patent ductus arteriosus, single ventricle lesions without pulmonary stenosis, among others. Additional causes are primary myocardial dysfunction, as seen in myocarditis or cardiomyopathy and ductal dependent lesions such as hypoplastic left heart syndrome, severe coarctation of the aorta or interruption of the aortic arch. The child in CHF may demonstrate tachypnea, retractions, poor feeding, diaphoresis particularly with exertion or feeding, and failure to thrive. Examination may reveal tachypnea, tachycardia, cardiomegaly, hepatomegaly and poor perfusion reflected by cool extremities and delayed distal capillary refill. Cardiac murmurs indicative of the basic cardiac lesion may be present in some patients.

1. Furosemide 1 mg/kg/dose; may repeat up to every 6 hours
2. Respiratory support as needed
3. Correct acidosis or metabolic derangements
4. Milrinone 0.25-0.75 mcg/kg/min if not hypotensive
5. Dopamine 5-10 mcg/kg/min
6. Prostaglandin E1 (PGE1)for babies with ductal dependent systemic perfusion (0.05 to 0.1 mcg/kg/min; reduce to 0.025 mcg/kg/min once the baby is stabilized)

Table 4: Management of acute congestive heart failure.

For a child with a large left to right shunt, the initial presentation with signs of CHF may be at 6 to 8 weeks of age. It is at this time that the pulmonary vascular resistance naturally drops and, therefore, the left-to-right shunt increases. In addition, any inter current respiratory or gastrointestinal illness (usually viral) may precipitate CHF. A child with myocarditis or cardiomyopathy may present at any age. Ductal dependent lesions usually present within the first few days to weeks after discharge from birth admission.

For the patient who presents with signs of CHF with pulmonary edema, initiation of diuresis is the first step. Furosemide at 1mg/kg/dose intravenously may be given. For pulmonary edema associated with respiratory distress, respiratory support may be needed. Nasal CPAP or, for severe distress, endotracheal intubation may be needed. Caution should be used when administering oxygen, however. Since oxygen is a potent pulmonary vasodilator, when given in large concentrations, it can increase the magnitude of the left-to-right shunting across a large defect, thus accentuating CHF. In patients with single ventricle lesions (to be discussed in the next section), O2 saturations in the range 80% are satisfactory and one should not aim to reach 100%.

Correct any metabolic acidosis that may be present. If the above measures have been undertaken and the patient is still in distress, then inotropic agents, such as Milrinone or Dopamine, or after load reducing agents, such as Nitroprusside or Enalapril, may be required. Milrinone has been previously shown to improve the cardiac index of patients as well as reducing both right and left ventricular after load in pediatric patients after cardiac surgery [13], and has also been used in other low cardiac output states not related to surgery, such as myocarditis or severe sepsis (Table 4). For babies with ductal dependent systemic perfusion, Prostaglandin E_1 (PGE_1) should be administered intravenously. The current dosage recommendations are for infusion of PGE_1, 0.05 to 0.1 mcg/kg/min; reduce to 0.025 mcg/kg/min once the baby is stabilized.

Cardiac Emergencies in the Patient with a Functional Single Ventricle

Precise understanding of the unique physiology of patients born with complex congenital heart lesions with single functioning ventricle is required to adequately address their care. There are several types of cardiac defects which have single functioning ventricle and these include hypoplastic left heart syndrome, tricuspid atresia, single ventricle, unbalanced atrioventricular canal defects and others. The common finding among these defects is that the heart has only one functioning ventricle, and this ventricle pumps blood to both the body

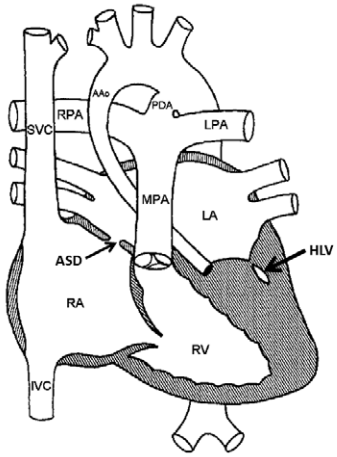

Figure 5: Diagrammatic illustration of hypoplastic left heart syndrome in the neonatal period prior to surgical intervention. Note the markedly Hypoplastic Left Ventricle (HLV) and a small Atrial Septal Defect (ASD), AAo, ascending aorta; IVC, inferior vena cava; LA, left atrium; LPA, left pulmonary artery; MPA, main pulmonary artery; PDA, patent ductusarteriosus; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava.

and lungs [14-17]. Treatment of these defects requires a three-staged surgical approach, beginning in the neonatal period and completed between 2 and 4 years of age [14-17].

Stage I

Single ventricle physiology patients born with obstruction of pulmonary blood flow receive an aortic to pulmonary artery shunt [18], usually a modified Blalock-Taussig shunt [19]. This shunt (Gore-Tex graft) is typically placed from the subclavian artery to the ipsilateral pulmonary artery. In patients who have single ventricle physiology with unrestricted pulmonary blood flow, the blood will preferentially flow to the lungs since the pulmonary vascular resistance is lower than the systemic vascular resistance. This results in severe congestive heart failure and decreased systemic perfusion. These patients receive a pulmonary artery band [20], which is a surgically induced constriction of the main pulmonary artery to protect the pulmonary vasculature and decrease pulmonary blood flow. Patients with hypoplastic left heart syndrome (classically consists of mitral atresia, aortic atresia, a very hypoplastic left ventricle, and a hypoplastic ascending aorta and aortic arch (Figure 5) undergo an operation in the neonatal period called the Norwood procedure [21,22]. The Norwood procedure (Figure 6) consists of surgically augmenting the hypoplastic aortic arch by anastomosing it with the pulmonary artery, creation of an atrial septal defect to allow for egress of pulmonary venous return into the right heart and provide pulmonary blood flow by placement of a Gore-Tex graft (Blalock-Taussig shunt described earlier) connecting the aorta to the pulmonary artery, or alternatively, a Sano shunt [23] which connects the right ventricle to the pulmonary artery. Some single ventricle patients require connection of the aorta to the pulmonary artery (Damus-Kaye-Stansel) along with a Blalock-Taussig shunt.

Stage II

No matter which surgery was done for palliation in the neonatal period, all patients will undergo a bidirectional Glenn procedure [24,25]. This consists of anastomosing the superior vena cava to the right pulmonary artery, end-to-side (Figure 7). The previously created Blalock-Taussig shunt is ligated.

Stage III

These patients eventually undergo Fontan procedure [26,27], usually one year following the bidirectional Glenn procedure. The Fontan, as performed at the current time, consists of connecting the inferior vena cava to the pulmonary artery via an extra cardiac conduit (Figure 8); the superior vena cava has previously been connected to the pulmonary artery during stage II, bidirectional Glenn. After the Fontan procedure, the single functioning ventricle pumps blood only to the body and the blood flow to the lungs is directly from the systemic veins, completely passive, and the two circulations are finally separated. Most patients receive a fenestration between the extra cardiac conduit and the atrial mass, the so called fenestrated Fontan (Figure 8).

Important thing to remember about single ventricle physiology prior to completion of the Fontan procedure is that the systemic and pulmonary circulations mix with each other. Therefore, the "normal"

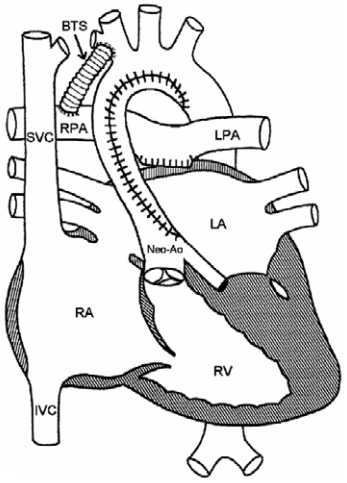

Figure 6: Staged surgical palliation of hypo plastic left heart syndrome.In stage I, Norwood procedure, the pulmonary artery and hypo plastic aorta are anastomosed to form the Neoaorta (Neo-Ao).Pulmonary blood flow is through a Gore-Tex graft shunt, here illustrated as a modified Blalock-TaussigShunt (BTS) from the right subclavian artery to the right pulmonary artery. Other abbreviations are as in figure 5.

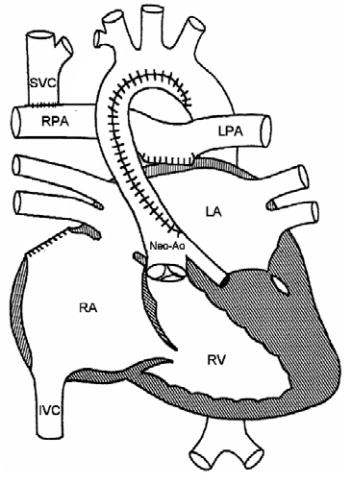

Figure 7: At the time of bidirectional Glenn procedure, the Gore-Tex shunt is taken down and the superior vena cava is anastomosed to the right pulmonary artery. Other abbreviations are as in figure 5.

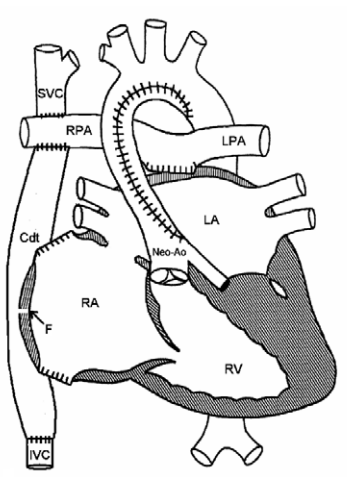

Figure 8: After completion of the Fontan circulation, the inferior vena cava is connected to the right pulmonary artery by means of a non-valved conduit (Cdt). Fenestration (F) is shown (arrow).Other abbreviations are as in figure 5.

oxygen saturations in these patients are between 75 and 85%, in contradistinction to above 96% in normal babies. In addition, the systemic and pulmonary circulations function in-parallel rather than in-series circuits of normal circulation. Excessive pulmonary blood flow produces systemic hypo perfusion and markedly reduced pulmonary blood flow produces severe hypoxemia. Consequently a delicate balance of the blood flows between the two circulations must be maintained. Conditions such as dehydration, acidosis, or fever upset the balance between the two circulations and cause a patient to become critically ill. As such, these patients are expected to utilize emergency departments with increasing frequency. These intercurrent illnesses do cause significant interstage mortality [28]; this is much higher between Stage I and II than between Stage II and III. In order to maximize successful outcomes, emergency physicians should be prepared to assess and treat acutely ill children with palliated single ventricle physiology [16]. Even minor illnesses must be treated aggressively.

The most feared, and most fatal, complication in patients with aorto-pulmonary shunts is an occlusive thrombus within the Gore-Tex graft. Because the shunt is the only source of pulmonary blood flow, rapid evaluation, diagnosis, and management are essential to ensure a successful outcome. Any patient with fever, vomiting, diarrhea, or decreased oral intake should be observed until the fever has resolved for at least 24 hours. Supplemental intravenous fluids should also be used as long as fever, vomiting or diarrhea persists, or until oral intake improves.

A patient with a severely obstructed shunt will present with severe cyanosis and respiratory distress. If the diagnosis is not made readily, rapid hemodynamic deterioration leading to cardiac arrest may occur. A continuous murmur of the shunt should normally be heard on auscultation. If the shunt is obstructed, the murmur disappears or is severely diminished in intensity. Management of an acutely obstructed shunt involves first the airway, breathing, circulation of initial assessment and basic life support. Intubation is likely to be necessary, as is cardiopulmonary resuscitation. In the case of suspected shunt obstruction, heparin may be given to prevent the progression of the thrombus. A pediatric cardiac and cardiovascular surgical consultation should be sought immediately in the case of suspected shunt occlusion. Emergency transcatheter opening of the shunt by interventional pediatric cardiologist [29] or shunt revision or placing the infant on ECMO by pediatric cardiovascular surgeon may be warranted. Unfortunately, even with the best of care, this condition carries a high mortality rate. Patients with shunts are on platelet inhibiting aspirin routinely in an effort to prevent this complication.

In summary, advances in pediatric cardiology and cardiovascular surgery have contributed to the greatly increased survival rates of patients with complex congenital heart disease. While these patients are living through the newborn period and beyond, they are most medically fragile and challenging. Increased survival of these patients means increased utilization of emergency rooms. These patients do have unique presentations and require specific treatment for common illnesses. They can also have complex cardiac emergencies which require prompt diagnosis and treatment. Emergency physicians are usually the first to be confronted with these emergency scenarios. Increased awareness and education of emergency physicians about this unique cohort of patients may help to increase the likelihood of a positive outcome.

Neurological Events

Cerebrovascular accidents (CVAs) and brain abscess may occur in association with cyanotic CHD; this is presumed to be due to intracardiac right to left shunt in the uncorrected or palliated cyanotic CHD patients. CVA used to be seen in the past in association with relative anemia in infants and children less than two years old, but is not as commonly seen in the current day and this is probably related to early surgery for the cyanotic CHD. Similarly, the CVA seen in older children with severe polycythemia has also become rare and is again related early surgical intervention incyanotic CHD. On the contrary, effective surgical palliation of complex cyanotic CHD patients leaves many infants and children at risk for development of CVA prior to complete separation of the systemic and pulmonary circuits either by complete biventricular repair or by single ventricular, Fontan type of palliation. Another group of patients with severe primary myocardial disease may have CVAs due to dislodgement of left ventricular thrombi.

Prevention of iron deficiency anemia by proper monitoring and timely correction may prevent CVA of infancy. In a similar vein, avoidance of severe hypoxemia and polycythemia by performing appropriate palliative or corrective surgery may decrease the frequency of CVA. Advice to the parents to avoid dehydration may also be useful. Timely erythropheresis may reduce the incidence of CVA in the older age group. Lastly, use of platelet-inhibiting drugs to prevent thrombi in primary myocardial disease patients may also reduce the incidence of CVA.

Early recognition and treatment of stroke is essential to minimize long term neurological sequelae. Stroke may occur in up to 30% of patients with congenital heart disease, and can be embolic or hemorrhagic in nature. Some of these may be clinically "silent" with no outward signs and only detected on brain imaging. Nevertheless, stroke should be kept in the differential diagnosis when dealing with acute onset of symptoms of any patient with CHD.

Recognition of stroke in the pediatric population requires a high degree of suspicion. While adolescents and young adults may present with more typical symptoms such as hemiparesis or dysarthria, the diagnosis can be missed or delayed in the young patient presenting with nonspecific symptoms. Atypical signs and symptoms of stroke in children may be as nonspecific as headache or seizures [30].

The management of acute stroke in children is similar to that currently in practice for adults. Combined use of reperfusion and

neuroprotection [31,32] should be considered after a pediatric neurology/stoke team consultation, especially when the time horizons are satisfied [33]. Once determined not eligible for thrombolytic therapy, the treatment is mostly symptomatic and includes adequate hydration, correction of anemia or polycythemia, anticonvulsants (if seizures are present) and physiotherapy. A detailed discussion of treatment of pediatric stroke is beyond the scope of this article; the reader is referred a recent review on this subject [30].

Conclusion

Emergencies of life-threatening nature involving the heart in children are many and complex; these cardiac emergencies can be challenging to the care-givers in the emergency rooms. In addition to the conventional emergencies of the past such as SVT, Tet spells and CHF, a new group of patients who have undergone staged palliation for single ventricle lesions should be added to this pool. With the advances in medical therapy and corrective/palliative surgery for congenital heart defects, these conditions are more likely to be seen with increasing frequency by the emergency personnel. Emergency physicians should have an understanding of the pathophysiology of these patients who present to their departments in order to promote positive outcomes. In the above presentation, a brief description of cardiac emergencies in neonates, infants and children that an emergency room physician may encounter was presented along with the management of such problems. Effective management depends upon rapid and correct diagnosis of the problem in order to institute appropriate therapeutic measures and referral to a specialized treatment center, when necessary.

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