PEDIATRIC CARDIAC EMERGENCIES

Abstract: *With major advances in the field of pediatric cardiology over the past few decades, outlook for children with congenital heart disease has improved immensely. Congenital heart defects, which were once considered incompatible with life, are now regularly managed immediately after birth with good survival. Better understanding of basic cardiovascular physiology behind such emergencies, has led to more focused and etiology oriented management resulting in better outcome. Through this article we aim to present some of the recent advances; both medical and interventional, which are changing the way we approach common pediatric cardiac emergencies.*

Keywords: *Cyanotic spell, Tachyarrhythmias, Heart failure, Neonatal emergencies.*

Pediatric cardiology has witnessed a number of major advances in the last three decades. This has translated into a vastly improved outlook for children with heart disease. Congenital heart defects, which were once considered incompatible with life are now regularly managed immediately after birth with good survival. Today, most cardiac emergencies are approached very differently. This is the direct result of a number of specific advances. More importantly, there is much more optimism in the eventual outcome of a child who presents with a cardiac emergency.

Through this article we aim to present some of the recent advances; both medical and interventional, which are changing the way we approach common pediatric cardiac emergencies.

We will focus on advances in the following common conditions presenting as cardiac emergencies in the pediatric age group

1. Cyanotic spell
2. Heart failure
3. Neonatal emergencies: duct dependant circulation, transposition
4. Tachyarrhythmias

A. Cyanotic spell

Classically seen in patients with tetralogy of Fallot, these are characterized by episodes of paroxysmal dyspnea with marked cyanosis especially during infancy. These episodes result from drastic reduction of pulmonary blood flow, with an accompanying increase in the right-to-left shunt and a drop in systemic arterial oxygen saturation. The key factors incriminated in the etiology include increased infundibular contractility,[1] peripheral vasodilation[2] and hyperventilation.[3] Kothari hypothesized that stimulation of right ventricular mechanoreceptors secondary to either increased contractility (due to endogenous catecholamines) or a decrease in right ventricular cavity size (such as with valsalva-like maneuver) may trigger a reflex response resulting in hyperventilation, some peripheral vasodilation, without bradycardia and perpetuates the vicious cycle.[4] Using this hypothesis he was able to correlate most of the precipitating events leading to cyanotic spells.

Traditional management included knee chest position, sedation with morphine, correction of acidosis with sodium bicarbonate and use of β-blockers (Propranolol), which was thought to relieve the infundibular spasm. But with better understanding of the underlying pathophysiology, volume loading with IV fluids to increase right ventricular preload; increasing systemic vascular resistance along with sedation with the help of ketamine and antagonizing the vasodilatory effects of β adrenergic stimulants with the use of β blockers[5] have become the cornerstone of

management of cyanotic spells. Recently, dexmedetomidine, a newer sedative agent has been shown to be of use in control of cyanotic spell.[6]

Keeping a low threshold for mechanical ventilation in refractory cases may help by breaking the vicious cycle of hyperpnoea and further cyanosis and would also prevent the adverse outcomes associated with severe cyanotic spells.[7-9] Preoperative extra corporeal membrane oxygenation therapy has also been used in patients with severe refractory spells.[10]

Long-standing cyanosis and repeated episodes of cyanotic spells have been shown to correlate with adverse neurodevelopment.[11] With advances in peri operative cardiac care, the trend has been towards early primary correction of TOF in the infancy itself. In fact many centers are performing primary repair within the first six months of age. But there is significant peri-operative morbidity, which includes prolonged mechanical ventilation, increased inotrope requirement and end organ dysfunction.[12-15] The alternative to complete corrective surgery includes palliation with a BT shunt, which is especially true for centers from developing countries with limited resources. Advances in cardiac catheterization techniques have helped in performing a potential alternative form of palliation with balloon pulmonary valvotomy (BPV) in carefully selected cases of Tetralogy with severe valvular PS or PDA stenting in those with TOF and pulmonary atresia.[16] Interim palliation with BPV has been regularly performed at our center for selected infants less than 3 months of age with predominantly valvar pulmonic stenosis. Data from our institution showed significant increase in the saturations, pulmonary annulus size and the branch pulmonary artery size after BPV for such cases.[17] Thus it can be considered as a safe and effective interim palliative procedure for symptomatic young infants with TOF and predominant valvar stenosis. Our experience with palliative PDA stenting in selected cases (> 18months) with congenital cyanotic heart disease with reduced pulmonary blood flow, with no immediate prospect of definitive surgical correction because of unsuitable anatomy or economic considerations showed statistically significant alternative form of palliation, with no complications during the procedure and also led to significant improvement in oxygen saturations. However PDA stenting can only be offered to the minority of patients with a PDA in association with TOF.

B. Heart failure

Heart failure (HF) refers to a clinical state of systemic and pulmonary congestion resulting from inability of the heart to pump as much blood as required for the adequate metabolism of the body. The clinical picture of HF results from a combination of "relatively low output" and compensatory responses to increase it.

Table 1. Management of cyanotic spells: Key advances

1. Greater emphasis on fluids for the initial resuscitation.
2. Intravenous β blockers in addition to conventional medications (morphine, soda-bicarbonate)
3. Lower threshold for mechanical ventilation
4. Low threshold for surgery during the emergency: Corrective surgery preferred if anatomy suitable
5. A single episode of cyanotic spell is sufficient to warrant consideration for surgical repair.

Broadly, heart failure results either from an excessive volume or pressure overload on normal myocardium (left to right shunts, aortic stenosis) or from primary myocardial abnormality (myocarditis, cardiomyopathy). Arrhythmias, pericardial diseases and combination of various factors can also result in HF. The resultant decrease in cardiac output triggers a host of

physiological responses aimed at restoring perfusion of the vital organs.[18] Important among these are renal retention of fluid, renin-angiotensin mediated and sympathetic over-activity. Excessive fluid retention increases the cardiac output by increasing the end diastolic volume (preload), but also results in symptoms of pulmonary and systemic congestion. Vasoconstriction (increase in after load) tends to maintain flow to vital organs, but it is disproportionately elevated in patients with HF and increases myocardial work. Similarly, sympathetic overactivity results in increase in contractility, which also increases myocardial requirements. An understanding of the interplay of the four principal determinants of cardiac output - preload, alter load, contractility and heart rate is essential in optimizing the therapy of HF. It is clinically useful to consider HF in different age groups separately.

Etiology of heart failure by age (Table.2)

The most common causes of heart failure in infancy are congenital heart defects.

The single most significant advance in management of heart failure in children is the recognition of the fact that a specific cause is identifiable in the vast majority of children with heart failure. Most of these causes are correctable (Table.3) and can be addressed through surgery, catheter interventions or very specific medications. The term cardiomyopathy is now reserved for a small proportion of patients and this diagnosis should only be made after thorough evaluation. A number of conditions can masquerade as cardiomyopathy and these need to be recognized.

Emergency management of heart failure includes a rapid assessment to define the possible etiology and severity assessment directed investigations to confirm the same. But greater emphasis needs to be placed on rapid early stabilization irrespective of the underlying etiologic factor. Conventionally therapy is directed towards:

I. Medical management

The initial management involves the usual assessment of the patient's airway, breathing and circulation (ABCs). This is followed by more specific therapeutic measures.

Table 2. Etiology of heart failure in various age groups.

Birth	1.	**Myocardial**
		Asphyxia; transient myocardial ischemia
		Sepsis and or myocarditis
		Hypoglycemia
		Hypocalcemia
	2.	**Neonatal hematological abnormalities**
		Anemia or hyper viscosity syndrome
	3.	**Neonatal heart rate abnormalities**
		SVT or congenital complete AV block
	4.	**Structural abnormalities**
		Volume overload lesions -
		Tricuspid regurgitation
		Pulmonary regurgitation
		Systemic arteriovenous fistula
		Hypoplastic left heart syndrome
1 week	1.	**Structural abnormalities**
		Critical aortic stenosis or pulmonic stenosis

Coarctation or interrupted aortic arch
Hypoplastic left-heart syndrome
TAPVC (with obstruction)
PDA (preterm infants)
Duct-dependent lesions with a large PDA

2. **Heart muscle dysfunction or arrhythmias**
3. **Renal abnormalities**
   Renal failure or systemic hypertension
4. **Endocrine disorders**
   Adrenal insufficiency

2 weeks - 2 months

1. **Shunt defects**
   Septal defects (ASD, VSD, AVSD)
   Aortopulmonary shunt (PDA, AP window, Truncus)
2. **Single ventricle**
3. **Obstructive lesions (see above)**
4. **Myocardial dysfunction**
   Cardiomyopathy
   Anomalous origin of the left coronary artery
   Metabolic diseases
5. **Pulmonary disease**

Table.3. Correctable causes of left ventricular dysfunction in children

Condition	Diagnostic clues	Specific treatment
	Congenital cardiovascular disease	
Anomalous left coronary artery from pulmonary artery	ECG changes of myocardial infarction typically in I, aVL, V4-6. 2D and color doppler echocardiography are usually diagnostic	Surgery (Coronary translocation or the Takeyuchi operation)
Severe coarctation of aorta	Weak femoral pulses, echocardiography	Surgery, balloon angioplasty
Critical aortic stenosis	Auscultation, echocardiography	Balloon angioplasty
	Acquired cardiovascular diseases	
Takayasu's arteritis	Careful palpation of all pulses; abdominal bruit, renal ultrasound, renal perfusion scans, doppler evaluation of the thoracic and abdominal aorta, aortography, high baseline renal parameters or extreme elevation following ACE inhibitors	Treatment of active disease (usually indicated by high ESR) may require steroids, immunosuppresants or both. After activity subsides, affected vessels may be treated by balloon angioplasty / stenting or surgery
Tachyarrhythmias		
Any long-standing ectopic atrial tachycardia, permanent junctional re-entrant tachy-cardia, chronic atrial flutter	Disproportionate tachycardia that is not readily explained by the condition of the child. Careful ECG evaluation, esophageal or invasive electrophysiologic testing	Antiarrhythmic drugs Radiofrequency ablation when appropriate
	Metabolic and nutritional causes	

Hypocalcemia	Typically occurs in newborns. Low calcium levels are universal. Adolescents with previously undiagnosed hypoparathyroidism. Chvostek and Trousseau signs may be positive. Prolonged QTc on ECG	Rapid response to restoration of calcium levels
Infantile beri-beri	1-4 month old infant who has been breast-fed by a thiamine deficient mother. Prominent edema, diarrhea and vomiting.	Rapid response to thiamine given intravenously
Carnitine deficiency	Hypoglycemia, coma and congestive heart failure in the infant. Ventricular hypertrophy may be seen. High ammonia levels, low serum carnitine and increased urinary excretion of carnitine	Rapid and sustained response to oral carnitine supplementation.
Hypophosphatemia	Poorly controlled diabetes, alcoholism, following hyper alimentation nutrition recovery, syndrome and following recovery from severe burns, hyperparathyroidism, hypomagnesaemia, Fanconi syndrome, malabsorption and vitamin D deficiency	Careful restoration of phosphorous levels
Selenium deficiency	Well described as Keshan disease. Formerly endemic in parts of China. Outside China selenium deficiency has been described in individuals on chronic parenteral nutrition and in those with AIDS. Focal myocyte necrosis on biopsy	Selenium administration can result in partial or complete resolution

Table.4. Various inotropes with their mechanism of action and adverse effects

Drug	**Mechanism**	**Dose**	**Special comment**
Dopamine	Increases cardiac outpatient, BP and improves peripheral perfusion. Characterized by dose dependent pharmacodynamic response, <10μg/kg/min, β_1 stimulation by Norepinephrine release, >10μg/kg/min, alpha receptor stimulation.	5-15 μg/kg/min	Renal dosing of dopamine is not recommended currently. Caution while using in patients with PAH as it may further increase pulmonary resistance, and hence pressure.
Dobutamine	Potent inotropic effect with vasodilation	5-15 μg/kg/min	Chronic usage over several days should be avoided as it causes down regulation of β receptors
Epinephrine	α and β agonist. Effects are dose-dependent: at low doses, if can cause vasodilation (β2-receptors); at high doses, it may produce vasoconstriction (α-receptors) of skeletal and vascular smooth muscle, with a subsequent increase of myocardial oxygen consumption.	0.1-1μg/kg/min	Tachycardia, arrhythmias, hyperglycemia, bronchospasm, vasoconstriction at very high doses leading to end organ injury.
Sodium Nitroprusside (a less expensive alternative to milrinone)	An NO donor that induces vascular smooth muscle relaxation and, thus, vasodilation. Nitroprusside seems to cause more systemic arterial (at the arteriolar level) dilation than systemic venous dilation. Therefore, it causes more reduction of after load than preload. Cardiac output	Initially 0.5 to 1μg/kg/min by continuous I.V. infusion. Usual dose is 3 μg/kg/min; maximum dose	Hypotension, reflex tachycardia, cyanide or thiocyanate toxicity; especially in those with hepatic or renal dysfunction respectively.

	increases and aortic and left ventricular impedance are decreased.	is 5μg/kg/min
Phospho-diesterase inhibitors; Milrinone	Selectively inhibit PDE III, increasing intracellular concentrations of cyclic adenosine monophosphate (cAMP). Increased cAMP in myocardial tissue results in increased intracellular calcium ion concentration and enhanced myocardial contractility. In vascular smooth muscle the increase in cAMP results in smooth muscle relaxation causing vasodilation.	Loading dose = 50 μg/kg administered over 15 minutes Maintenance dose: 0.35-0.75μg/kg/min	Improve myocardial function without increasing myocardial oxygen consumption and have a lower risk of arrhythmias than catecholamines. Watch for hypotension, thrombocytopenia and dysrhythmias. Hypotension responds to volume infusion.

l. Correction of low cardiac output state

Use of inotropes has been a well-established modality for stabilization of patients in heart failure irrespective of the underlying etiology. Inotropes other than digoxin are used for short-term support of circulation or to tide over the crisis (Table.4). Commonly used inotropes include

Newer agents

Levosimendan : It is a pyridazinone-dinitrate that belongs to a new class of drugs, the calcium sensitizers. In contrast to other inotropic agents, levosimendan is deemed to improve myocardial contractility without increasing intracellular calcium. It acts by binding to myocardial troponin C, causing a configuration change in tropomyosin that exposes actin and myosin elements, allowing for a more effective contraction. It offers the advantage of increasing systolic force without compromising coronary perfusion. Moreover levosimendan opens adenosine triphosphate (ATP)-sensitive vascular potassium channels, causing vascular hyperpolarization and relaxation, coronary artery dilation and myocyte mitochondrial activation. Levosimendan is used in the treatment of decompensated cardiac failure and as an elective drug in patients with perioperative risk of ventricular failure. It has also been used in the rescue therapy of patients who have difficulty weaning from cardiopulmonary bypass or from mechanical circulatory support. There arc also reports documenting its favorable effect in reducing pulmonary vascular resistance and endothelin-1 levels and in improving right ventricular failure.[19,20]

Calcium infusion as an inotrope : Inotropic state of left ventricle can be altered by alteration of circulatory calcium level. Calcium supplementation plays an essential role in augmenting left ventricular function in pediatric patients. Calcium, either in the form of gluconate or chloride salt have a significant inotropic effect in the ionised form but can cause increased resistance in coronary and systemic vascular beds leading to decrease in oxygen supply to the myocardium and increased left ventricular after load. The usual recommended dose of calcium chloride in low cardiac output syndrome (LCOS) in infants and children is 10-20 mg/kg slowly into a central vein. The dose of calcium gluconate is three times more compared to calcium chloride.

2. Correction of congestive state

This is achieved with the use of diuretics (to reduce pulmonary or systemic congestion), and after load reducing agents (ACE inhibitors).

Diuretics afford quick relief in pulmonary and systemic congestion. 1-2mg/kg of furosemide is the agent of choice. Secondary hyper-aldosteronism does occur in infants with CHF and addition of spironolactone 1 mg/kg single dose to other diuretics conserves potassium.

3. Role of mechanical ventilation

A lower threshold for early elective mechanical ventilation especially for cases not responding to above line of therapy needs to be emphasized. Mechanical ventilation helps by reducing the work of breathing and thus translating into reduction of myocardial oxygen demand. Hypoventilation and low FiO_2 help in reducing the amount of left to right shunt by increasing the pulmonary artery pressures.[21]

4. Role of neurohormonal modulation

ACE inhibitors: Several studies in infants and children with left-to-right shunts or dilated cardiomyopathy have demonstrated improved hemodynamics or significant clinical improvement after the introduction of an ACE inhibitor.[22-27] Three studies demonstrated a decrease in the pulmonary-systemic blood flow ratio and the left-to-right shunt following the administration of an ACE inhibitors.[24,26,28] Infants with a left-to right shunt and increased systemic vascular resistance showed the greatest response.[26] Oliguria, acute renal failure and hypotension are the most common side effects. Renal function should be monitored carefully when initiating therapy with ACE inhibitors. Hypotension is more frequent in patients who have elevated plasma renin activity and in patients who are volume depleted; therapy in these patients should be initiated with low doses. Captopril is the most commonly used ACEI. Dosage involves a test dose of 0.1mg/kg with monitoring of blood pressure, followed by gradual increment in doses up to l mg/kg/dose every 8 hourly.

βblockers: Like ACE inhibitors, (β-blockers interfere with the endogenous neurohormonal system. ACE inhibitors interrupt the renin-angiotensin system, whereas β-blockers inhibit the effects of the sympathetic nervous system. Although the use of β-blockers in CHF may seem counterintuitive, low doses titrated slowly in adults with systolic dysfunction decrease symptoms of CHF and decrease both, the risk of mortality and the combined risks of hospitalization or death.[29,30] The rationale perhaps relates to down grading of β-receptors due to chronic catecholamines stimulation. It is important to note that β-blockers are only used in stable patients. The therapy is best undertaken in hospital as careful monitoring is required. Carvedilol, a nonselective βblocker with alfa-1 blocking and anti-oxidative properties has proven to be beneficial in infants with dilated cardiomyopathy and there is significant improvement in their functional status.[31]

4. Correction of precipitating factors

Almost always, the worsening in clinical state of a patient with CHF can be traced to a precipitating event, the treatment of which leads to significant improvement. The checklist includes intercurrent infections, anemia, electrolyte imbalances, rheumatic activity, infective endocarditis, arrhythmia, pulmonary embolism, drug interactions, drug toxicity or non-compliance and other system disturbances.

5. Role of prostaglandin E1

Neonates with transposition of great arteries, coarctation of aorta, aortic stenosis in failure or hypoplastic left heart syndrome, etc., improve remarkably with PGE_1. The therapy is initiated at 0.05 μg/kg/min and may be gradually raised or lowered depending, on the response. Apnea may

occur during the infusion and ventilatory support should he available. Irritability, seizures hypotension and hyperpyrexia are rare.

6. Miscellaneous

Extra corporeal membrane oxygenation, left ventricular assist device (LVAD) and the intra aortic balloon pump (IABP) have also found a place in the management of pediatric patients with heart failure. There is a 74% survival rate and the long-term outcome has been excellent in most cases.[32] But the high cost of equipments and availability at select centers limits the use of these modalities for regular management of heart failure.

II. Catheter based management

With the recent advancements in pediatric interventional cardiology, it has become possible to treat certain congenital cardiac conditions without waiting for complete resolution of heart failure. Classic examples include transcatheter coil or device closure of PDA, especially in preterm newborns stuck on mechanical ventilation and balloon dilatation of critical aortic stenosis or severe coarctation of aorta presenting with severe heart failure. Balloon dilatation of critical aortic stenosis has now been achieved even in smaller neonates through the umbilical artery route with the availability of smaller sized catheters.[33]

III. Emergency surgical management

Advances in cardiac surgical techniques and preoperative cardiac critical care have aided early primary repair of many congenital heart defects irrespective of the weight and age and with underlying refractory heart failure or respiratory tract infections. Classic examples include closure of heart defects especially large PDA in small newborns not amenable to transcatheter closure. ALCAPA repair at the time of diagnosis, repair or replacement for regurgitant valvular lesions, etc. Surgically treating critically ill cases with large VSD and respiratory infection has shown a success rate of 91.6% amongst the operated cases.[34]

C. Neonatal cardiac emergencies

These constitute a specific group of conditions with certain common presentations (Table.6). The three common presentations include:

1. Neonate with cyanosis
2. Neonate with cardiovascular collapse
3. Neonate with heart failure

I. Medical management

A relatively well child presenting dramatically around 3 days to 1 week of life with either cyanosis or cardiovascular collapse strongly suggests duct dependent lesion. Oxygen may precipitate duct closure and therefore should be used with caution in duct dependent lesions. It may also worsen left to right shunts by decreasing pulmonary vascular resistance. Pulmonary blood flow can increase at the cost of systemic blood flow. Thus tailoring the FiO_2 to achieve a SpO_2 of 80-85% may be adequate to balance systemic and pulmonary circulations in patients with a single ventricle physiology. Cyanosis not associated with acidosis need not be corrected. PGE1 infusion can be life saving. PGE1 sensitive lesions may present with cyanosis and murmur or mild /no cyanosis with abnormal pulses. Clinical suspicion of obstructed TAPVC should be high, in situations with worsening of cyanosis after initiation of PGE1 infusion. Dosage needs to be titrated according to the saturation.

Ideal ABG target should be to achieve a PO_2 and PCO_2 level of about 40 with a pH of 7.4. Adverse effects of PGE1 include apnea, tachycardia, bradycardia, fever, NEC, seizures. thrombocytopenia. Major limiting factor for initiation of treatment is its high cost.

II. Catheter based interventions

Balloon atrial septostomy: Neonates with transposition of the great vessels with intact ventricular septum and an inadequate sized foramen ovale have severe cyanosis due to poor intercirculatory mixing. Balloon atrial septostomy helps by improving this admixture between the systemic and the pulmonary venous blood to achieve a reasonable saturation and improve the hemodynamics till a time that an arterial switch operation can be performed. The advantage of this procedure is that it can be even performed on the bedside with the help of echocardiographic guidance especially in sick newborns.

Balloon aortic valvotomy: Neonates with critical AS suffer from low cardiac output and shock secondary to poor left ventricular function and/or mitral insufficiency. Outcome is usually fatal in most of these patients within the first weeks of life with medical treatment alone. The use of percutaneous balloon aortic valvuloplasty was first introduced in 1984 and has become the first-line treatment for critical aortic valve stenosis in neonates.[35-38] Most studies show that at 5-year follow-up, around 85% of patients are alive and 60% remain free of re-intervention.[39-41]

Table 5. Cardiac emergencies in the newborn

Physiologic Category	conditions	Manifestation
Duct dependent systemic blood flow*	Hypoplastic left heart syndrome, critical coarctation, interruption of aortic arch, critical aortic stenosis	Heart failure
Duct dependent pulmonary blood flow*	Pulmonary atresia, critical pulmonary stenosis, Ebstein anomaly	Cyanosis, hypoxia,
Obstruction of pulmonary venous return	Obstructed total anomalous pulmonary venous return, mitral atresia with a restrictive patent foramen ovale	Cyanosis, hypoxia, heart failure
Parallel circulation with poor mixing	D transposition with intact ventricular septum	Cyanosis, hypoxia
Valve regurgitation	Congenital mitral valve regurgitation	Heart failure
High-output state	AV malformations (usually intracranial)	Heart failure
Myocardial dysfunction	Myocardial diseases (inflammatory and metabolic)	Heart failure
Tachyarrhythmia	Atrial flutter, neonatal atrio-ventricular reentrant tachycardias, ectopic atrial tachycardia	Tachycardia, heart failure
Bradyarrhythmia	Complete heart block	Bradycardia, heart failure

*Some of the duct dependent conditions (critical PS, AS, coarctation) manifest with varying severity. The most severe forms manifest early (in the first few days) with absolute

dependence on the duct for survival. Others may manifest later in the neonatal period (first few weeks) with heart failure or cyanosis and may not be strictly "duct dependent".

Balloon dilatation/Stenting of coarctation of aorta: Newborns with coarctation of aorta present in a critical state with left ventricular dysfunction, shock, pulmonary hypertension and end organ dysfunction. Emergency surgical correction may not always be feasible at short notice and may be associated with significant mortality and morbidity. Percutaneous balloon dilatation or stenting of coarctation of aorta achieves immediate relief of heart failure by reducing left ventricular pressure overload and allows postponing of surgery to a more appropriate time. The drawbacks of balloon dilatation are the high incidence of restenosis due to recoil and unfavorable scarring and aneurysm formation due to damage to the media of the vessel. Placement of a stent is said to interfere with the growth of the vessel. In spite of this, these procedures are life saving measures and may be considered for interim palliation

PDA stenting for duct dependant pulmonary circulation: Conventional management of children with duct dependent pulmonary circulation (DDPC) includes prostaglandin infusion and/or surgically created aorto pulmonary shunts. Surgical shunts have significant mortality and morbidity. Maintaining the duct patency by stenting has been shown to be feasible with low complication rates[16].

II. Surgical Approach

Emergency surgical intervention is indicated for certain congenital cardiac defects like total anomalous pulmonary venous connection with obstruction after basic stabilization (mechanical ventilation, inotropes, diuretics, correction of acidosis). This has shown to reduce the post-operative mortality.[42]

D. Tachyarrhythmias

Confronting a new patient with a sustained tachycardia in the emergency room can be an intimidating experience for the uninitiated. However, prior knowledge of the finite list of diagnostic possibilities and familiarity with a standard sequence of therapy, can easily transform these encounters into an intellectually stimulating exercise with an optimal patient outcome. It is therefore imperative that the attending physician be well versed with the approach to tachyarrhythmias, which results in a rapid treatment response that is appropriately tailored to the underlying mechanism and severity of the arrhythmia.

Though there are several mechanisms for tachycardia, the clinician must be able to narrow the field to the one or two most likely possibilities in the acute setting in order to guide acute therapy and design a long-term management plan. This can be accomplished with reasonable accuracy on the basis of standard ECG recordings. A full 12-lead ECG should be obtained on all patients with a tachyarrhythmia whenever possible. A single lead rhythm strip or the monitor is a very poor substitute, because it lacks definition of the QRS and P-wave axis that can be the key to pinpointing the correct mechanism. In addition, the P-wave may not always be visible on a single given lead, nor can the maximum width of QRS complex be determined with certainty.

The only clinical situation where a full ECG should be deferred is when the suspected tachyarrhythmia is associated with hemodynamic compromise. This situation is seldom seen with narrow QRS tachycardia. However selected children with wide QRS tachycardia may have serious hemodynamic compromise. Examples include, ventricular tachycardia, ventricular fibrillation and atrial fibrillation in the presence of pre-excitation. In all other settings, a 12 lead ECG should be obtained in the presenting tachycardia, after which a rhythm strip should be run

during each therapeutic maneuver (especially while giving adenosine), followed by a repeat ECG after restoration of the patient's normal rhythm.

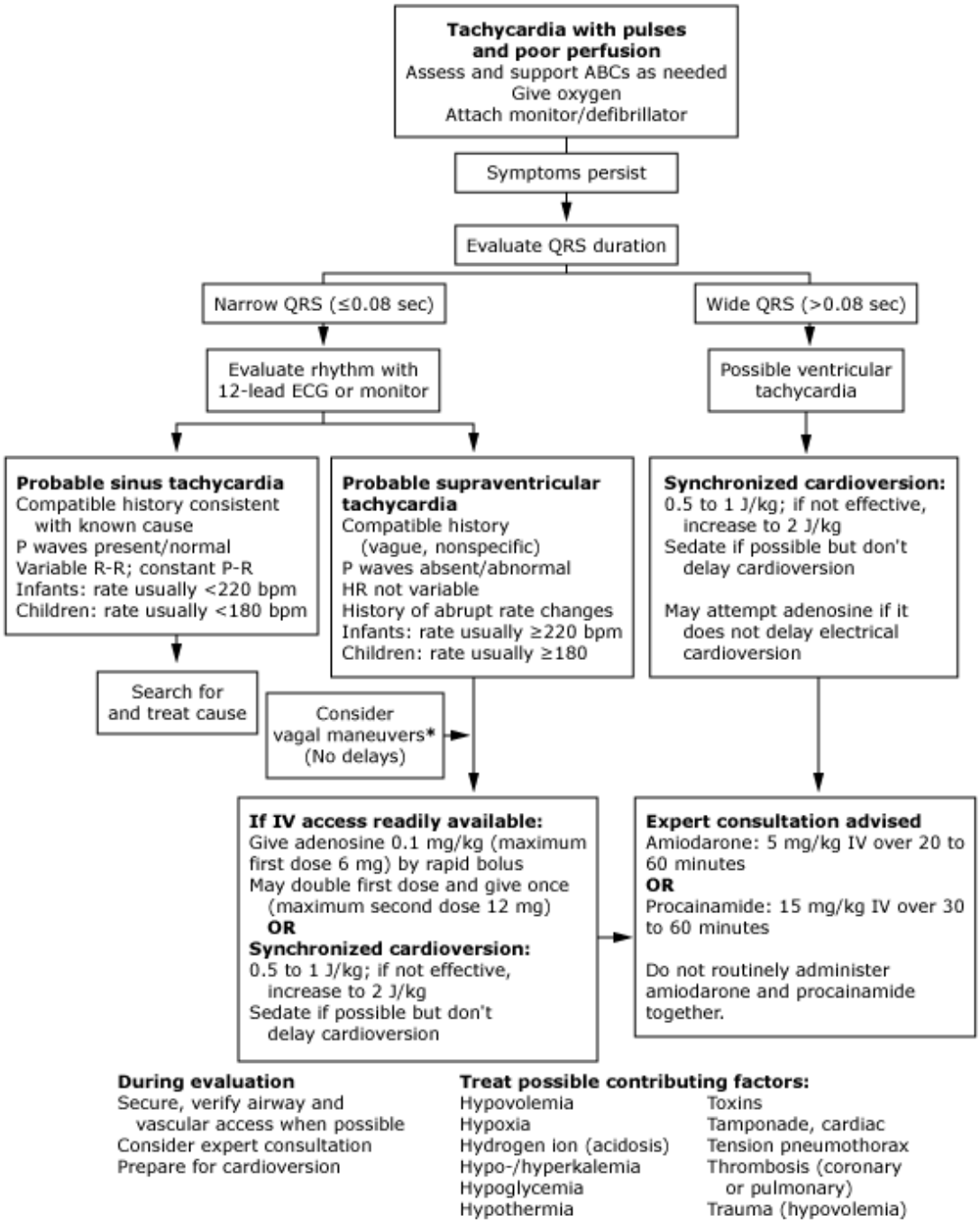
Emergency management

The initial point to address from the ECG is whether the QRS complex in tachycardia is narrow or wide. This step is intended to provide a gross discrimination between supraventricular tachycardia (SVT) and ventricular tachycardia (VT). The QRS complex is labeled narrow only if its duration falls within the normal range for age in all[43] 12 ECG leads. Though SVT is more common in children, a wide QRS complex suggestive of VT requires urgent evaluation and conversion.

An algorithmic approach to management of tachyarrhythmias is given below (Fig.1).

This basic approach is for the emergency management of arrhythmias without taking into consideration the underlying mechanism for tachycardia.

Fig.1. Algorithmic approach to management of tachyarrhythmias in children



* Vagal maneuvers: In infants or young children, use a bag filled with ice and cold water over the face for 15 to 30 seconds or rectal stimulation with a thermometer. In older children, encourage bearing down (Valsalva maneuver) for 15 to 20 seconds. Carotid massage and orbital pressure should not be performed in children. Adapted from: American Heart Association. 2005 American Heart Association (AHA) guidelines for cardiopulmonary resuscitation (CPR) and emergency cardiovascular care (ECC) of pediatric and neonatal patients: pediatric advanced life support. Pediatrics 2005; 117:e1005.

With advances in cardiac electrophysiology, it has become imperative to diagnose the underlying mechanism of the tachyarrhythmia to guide further management strategy. For e.g. SVT due to accessory pathway is amenable to treatment with radiofrequency catheter ablation with high success rates. Certain mechanisms like ectopic atrial tachycardia and permanent junctional reciprocating tachycardia are well known to be associated with a pattern of heart failure known as tachycardia induced Cardiomyopathy or tachycardiomyopathy.[44] Heart failure and cardiomyopathy may improve completely or partially after control of such arrhythmias.

In certain SVT with very high rate, the underlying mechanism may be uncovered only after administration of I.V. Adenosine; thus highlighting the importance of recording an ECG while giving adenosine.

Concept of tachycardiomyopathy

Coined by Gallagher JJ,[45] the term tachycardiomyopathy refers to impairment in left ventricular function secondary to chronic tachycardia, which is partially or completely reversible after normalization of heart rate and/or rhythm irregularity.

Fenelon, et al[46] further classified tachycardiomyopathy into two categories, namely "pure type" and "impure type". In the former, chronic tachycardia causes LV dysfunction in a normal heart and completely recovers after termination of the tachycardia. In the latter, such a condition occurs in patients with structural heart diseases and the cardiac dysfunction may only recover incompletely after termination of the tachycardia. The incomplete recovery of LV function might be the result of long term tachycardia inducing irreversible myocardial injury.

Tachycardiomyopathy is induced by various supraventricular and ventricular arrhythmias. Ectopic atrial tachycardia (EAT) and permanent junctional reciprocating tachycardia (PJRT) are the two most common arrhythmias associated with tachycardiomyopathy in children. This is due to their incessant nature and refractoriness to anti-arrhythmic drugs.

The basic concept for the treatment of tachycardiomyopathy is in controlling the heart rate, either by pharmacological means or non-pharmacological procedures like surgical or radiofrequency ablation.

Table 6. Key advances in management of tachyarrhythmias in children

1. Most tachyarrhythmias in children in the absence of structural heart diseases are well tolerated.
2. An episode of tachyarrhythmia should be looked upon as an opportunity to identify the mechanism (re-entrant or automatic) and arrive at a specific diagnosis.
3. Unless the tachyarrhythmia is associated with hemodynamic compromise, it is imperative to record a baseline ECG and document the response to adenosine or other forms of treatment.
4. Persistent tachyarrhythmia can manifest as heart failure and masquerade as cardiomyopathy.

Thus it becomes imperative that any tachyarrhythmia presenting to the emergency room be worked up completely along with evaluation of cardiac function after initial stabilization and restoration of sinus rhythm.

Radiofrequency ablation

In a very short period of time, there has been a revolution in the treatment of arrhythmias in both adults and children. This revolution in arrhythmia therapeutics has been facilitated largely by refinement of catheter ablation techniques, which allow arrhythmias to be diagnosed and "cured"

in one session in the catheterization laboratory. In many circumstances drug therapy has been pushed down as a second line alternative therapy.

Catheter ablation is a procedure during which a patient's cardiac conduction system is tested for its ability to sustain an arrhythmia. Once the arrhythmia is diagnosed, its substrate is electrically identified and anatomically localized. Subsequently, a critical component of the arrhythmia's substrate is destroyed using energy delivered through the tip of a catheter. Currently, the energy most commonly used is radiofrequency energy. The electrophysiology study, which is performed in conjunction with this technique, identifies and localizes the arrhythmogenic substrate of a pathologic tachycardia. This substrate may be an accessory connection responsible for atrio-ventricular (AV) reciprocating SVT or Wolff-Parkinson-White (WPW) syndrome; an irritable focus in the atrium or ventricle responsible for an ectopic atrial tachycardia (EAT) or ventricular tachycardia (VT); a scar in the atrium or ventricle responsible forming a component of a re-entrant tachycardia circuit (atrial flutter or VT); or an area of slow AV node conduction that is critical for the perpetuation of the AV node re-entry tachycardia.

Almost all common pediatric arrhythmias can be treated with catheter ablation. The most commonly treated substrate is the accessory atrio-ventricular pathway (AP). The overall success rate regardless or the pathway location, the presence of multiple pathways, catheter approach, or patient age, can be as high as 98% and typically range 85% to 95%.[47-49]

On the basis of data from 1999 Pediatric RF Ablation Registry, AVNRT accounts for approximately 24% of ablations performed in children, with success rates of up to 97%.[50,51]

Major complications are minimal, but include complete AV block when ablating septal pathways,[47,49] cardiac perforation and tamponade,[50,51] inadvertent coronary damage,[40] and vascular or embolic injury.[51,52]

Conclusion

Pediatric cardiac emergencies require very specific treatment in the emergency room setting. Considering the possibility of a cardiac problem as the cause for the presenting symptoms is the initial step in successful management. With advancements in the understanding of basic cardiovascular physiology behind such emergencies, their handling has become more focused and etiology oriented; resulting in better outcomes. Cyanotic spells can now be readily managed with various medications and if required urgent surgical intervention. Management of cardiac failure is now more focused on unveiling the underlying cause along with modification of derangements of the various compensatory mechanisms that have taken place. Newborn cardiac emergencies are being regularly handled with the availability of prostaglandin E1 and with the advancements in transcatheter procedures to tide over the crisis. Approach to arrhythmias requires more focus on finding the underlying mechanism to provide a more specific pharmacological or interventional therapy.

To conclude, common pediatric cardiac emergencies are now managed more efficiently with better outcomes seen every in the worst case scenarios.

Points to Remember

- ***Cyanotic spell: 1V fluids, low threshold for mechanical ventilation, early surgery***
- ***Heart failure: Emphasis on identifying underlying etiology***
- ***Neonatal cardiac emergencies: Early detection, role of prostaglandin E1 and transcatheter procedures.***

- ***Tachvarrythmias: Importance of 12 lead ECG at presentation and rhythm strip during therapeutic maneuvers.***

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