

Neonatal and Pediatric Guidelines for Arrhythmia Management

What the Neonatal and Pediatric Critical Care Nurse Needs to Know

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Introduction to the Problem:

Arrhythmias are relatively common in the pediatric cardiac intensive care unit. Grosse-Wortmann et al studied 494 patients revealing 59% of neonates and 79% of older children have arrhythmias within 24 hrs of surgery. Of these arrhythmias, junctional ectopic tachycardia (JET) was seen in 9% of neonates and 5% of older children. Ventricular tachycardia was found in 3% of neonates and 15% of older children (Gross-Wortmann, 2010).

In terms of specific arrhythmias, sinus tachycardia is the most frequently seen arrhythmia, with supraventricular tachycardia being the next most common, followed by sinus bradycardia (Hanash, 2010). Reentrant tachycardia is common in infants and children with congenital heart disease (CHD). Some arrhythmias in the early post operative period like premature atrial contraction's (PAC's) and premature ventricular beats (bigeminy) are usually transient and well tolerated. Others like junctional ectopic tachycardia (JET) and atrial flutter may cause significant hemodynamic instability and compromise or even sudden cardiac death.

Primary arrhythmias occur in children without structural heart disease, although they may be secondary to ion channel diseases that are still being elucidated. Risk factors that predispose children for secondary arrhythmias include congenital cardiac malformations, surgical repair and scarring, long cardiopulmonary bypass times, or exposure to chronic hemodynamic stress (Brugada, 2013). Electrolyte and acid-base imbalance and the use of vasoactive drugs also predispose children to arrhythmias (Jhang, 2010). Inflammation/carditis seen in diseases such as acquired heart diseases like Kawasaki disease, rheumatic fever and myocarditis may produce arrhythmogenic foci (Curley, 2001). Conditions of ventricular volume overloading, valvular regurgitation, congestive heart failure and pulmonary hypertension are other secondary reasons (Huh, 2010). Regardless of the cause of the arrhythmia, there are certain common signs, symptoms and treatment options that are ultimately based on the rhythm more than on the etiology with certain very important exceptions. Symptoms may vary depending upon age and include feeding intolerance, lethargy, irritability, pallor, diaphoresis, syncope, fatigue or palpitations.

Mechanisms of tachyarrhythmias can be enhanced automaticity with triggered foci or enhanced conduction with the presence of reentrant circuits. Similarly bradycardia can result from suppressed automaticity or suppressed conduction, where normal conduction is delayed or blocked (Allen, 2011). Understanding the mechanism informs the optimal treatment choice.

This review will describe the variety of arrhythmias that occur in pediatric patients, how they are characterized, and the associations with congenital heart disease, cardiomyopathies and ion channel diseases. Clinical manifestations, etiologic considerations, diagnostic measures and treatment strategies will be discussed.

Arrhythmias to Consider/Address:

CARDIAC ARRHYTHMIA	CHARACTERISTICS
Sick sinus syndrome (SSS)/ Tachy-Brady Syndrome	Sinoatrial (SA) node becomes dysfunctional and is no longer a reliable pacemaker, most commonly manifested as bradycardia, although there can also be tachycardia. When the sinus rate is slower than another potential pacemaker in the heart, it may no longer be the dominant pacemaker. SSS can also cause an alternating bradycardia and tachycardia. A number of rhythms result including sinus bradycardia, sinus arrest and junctional rhythm, and ectopic atrial and nodal rhythms.

	<p>The term SSS includes SA node dysfunction plus symptoms of dizziness, syncope or sudden cardiac death (Park, 2010).</p>
Bradycardias	<p>Often caused by hypoxia, vagal tone, hypothyroidism, cardiac surgery, endocarditis and myocarditis (Hanash, 2010); hyperkalemia, sleep, hypothermia, sedation and anesthesia. (Curley, 2001).</p> <p><u>Sinus bradycardia:</u> Sinus node slower than normal for age related normal values.</p> <p><u>Slow junctional escape rhythm/nodal rhythm:</u> Spontaneous depolarization of the AV node. The sinus node has either failed to fire or is slower than the AV node. Rates 50-80 beats/min in children less than 3 yrs and 40-60 beats/min for children older than 3yrs. Can be common after atrial surgery and are usually transient.</p> <p><u>Ventricular escape rhythm or ideoventricular rhythm:</u> Origin of impulse is from the ventricle and presents with rates slower than from the AV node. QRS have wide complex morphology. This is a secondary phenomenon vs. a primary arrhythmia and occurs when the sinus node and/or the AV node are dysfunctional. An example of this is complete heart block with a ventricular escape rhythm. The ventricle itself is working well, and the escape rhythm is a symptom of another problem.</p>
Premature Beats/Extra-Systoles	<p>Premature atrial, junctional and ventricular ectopic beats are common and may occur in patterns of bigeminy, trigeminy, quadrageminy or couplets. These are generally benign.</p>
Wandering Atrial Pacemaker	<p>Shifting of the pacemaker site from the SA node to alternate sites in the atria and junction (AV node). P-wave configuration changes as the site changes.</p>
Supraventricular Tachycardias (SVT) SVT is used as a collective term.	<p>Originates above the bundle of His. Reentrant circuits generally have an abrupt onset and termination i.e. are paroxysmal (Hanash, 2010).</p> <p><u>Sinus tachycardia:</u> Sinus node is faster than age-related normal values due to enhanced automaticity. Usually due to fever, pain, anxiety, anemia, medications, hypovolemia or in the presence of increased catecholamines. While not generally an indication of conduction system pathology, sinus tachycardia may be an important indicator of significant cardiovascular compromise.</p> <p><u>Reentrant tachycardias:</u> Reentrant tachyarrhythmias require the presence of two possible conduction pathways with different conduction and refractory properties. The tachycardia uses both pathways; one as an antegrade limb and one as a retrograde limb of the reentry circuit.</p> <p>a) Within the atria: atrial flutter, atrial fibrillation; intra-atrial reentrant tachycardia (IART) atrial flutter - or incisional tachycardia represents macroreentry within the atrial muscle and may be slower than atrial flutter (Walsh 2007).</p> <p>b) Atrioventricular reentrant tachycardias include:</p> <ol style="list-style-type: none"> 1) atrioventricular reentrant tachycardia (AVRT): commonly associated with Wolff-Parkinson-White. Accessory pathway present allowing impulses that entered via the AV node to enter the atria 2) atrioventricular nodal reentry tachycardia (AVNRT): uses a “slow-fast AV nodal pathway”. Antegrade conduction limb is the slow pathway and retrograde limb fast one. Simulation of the atria by the retrograde pathway produces inverted p-waves if visible. Concurrent stimulation of the ventricles. 3) permanent junctional reciprocating tachyarrhythmia (PJRT). These are reentrant circuits in which one limb includes the AV node.

	<p>Wolff-Parkinson-White Syndrome (WPW): Baseline resting ECG is characterized by a short PR interval, wide QRS and delta wave which is a manifestation of the accessory on sinus rhythm. WPW is marked by the delta wave on the <i>resting</i> ECG (Allen, 2001). Atrial flutter or atrial fibrillation in the presence of this type of accessory connection can result in VF.</p> <p>The QRS complex in SVT is wide if there's aberrant conduction, in which the antegrade limb is the accessory connection. If the AV node is the antegrade limb, the QRS is a narrow complex.</p> <p><u>Automatic tachycardia – AET and JET:</u> local enhanced automatic focus of certain cardiac myocytes in the atria or AV node. AET and JET are non-reciprocating tachycardias that originate from a single focus unlike reentrant rhythms.</p> <p>AET/JET are seen more commonly in neonates and usually observed within the first several days after cardiopulmonary bypass (Grosse-Wortmann, 2010). They are refractory arrhythmias that are relatively resistant to treatment. The goal is rate control and restoring AV synchrony. These are often transient arrhythmias lasting 24-72 hours. Rapid rates lead to early contraction of the atria against closed AV valves resulting in <i>cannon A waves</i> on hemodynamic monitoring lines (CV, RA, and LA) (Schwartz, 2014).</p> <p>(AET) When this occurs at an ectopic site within the atria, it is called atrial ectopic tachycardia. AET occurs as a result of irritation of tissues during cardiac surgery, with placement of intracardiac lines, application of sutures, or cutting tissue. Any reason for dilated atria, cardiomyopathy or diseased AV valves, ventricular dysfunction can result in this rhythm disorder. Rates are usually above 170-180 beats/min and beyond 200 beats/min. A block at the AV node can cause AV dissociation, further contributing to hemodynamic instability in addition to the rapid atrial rate. The rhythm may be variable, and may be interspersed with periods of sinus rhythm. The rate can ramp up or slow down over minutes in contrast to the sudden onset and offset of SVT (Schwartz, 2014).</p> <p>(JET) When the ectopic focus initiates at or near the AV node then the arrhythmia is junctional ectopic tachycardia. JET is usually caused by surgery around the AV node and rates often range between 160 beats/min to as high as 280 beats/min. Characteristics include inverted P-waves in lead II and an R-P interval which is short or absent. Primarily seen post re-warming from cardiopulmonary bypass and within 3 days of the surgery (Schwartz, 2014).</p>
Ventricular tachycardia (VT)	<p>Three or more consecutive ventricular complexes are by definition VT. Wide QRS complex morphology and a different QRS morphology than the usual QRS waveform characterize VT. Morphology may be monomorphic (uniform), polymorphic (multiform); or Torsades de Pointes where the points seem to twist around the isoelectric line.</p> <p>Often associated with structural heart disease, particularly late (years) after repair. Other common clinical situations in which one might see VT include dilated and hypertrophic cardiomyopathy, metabolic alterations including severe hypoxia, acidosis, hyper/hypokalemia, and drug toxicity such as cocaine, digoxin, and tri-cyclic antidepressants. Other conditions include myocarditis and long Q-T syndrome. Whenever VT occurs in a paediatric patient one must also consider ischemia or infarction (Allen, 2001). Patients may present hemodynamically stable or in cardiac arrest.</p>
Ventricular Fibrillation (VF)	<p>Completely uncoordinated depolarization of heart muscle mass resulting in inability to maintain any global excitation contraction coupling. The myocardium fails to squeeze and cardiac arrest occurs.</p>

1 st Degree Heart Block	Slowed conduction through the AV node resulting in prolonged duration of PR interval.
2 nd Degree Heart Block (Mobitz I, Wenckebach)	Intermittent block of conduction of atrial beats to the ventricle resulting in dropped QRS complexes. Progressive lengthening of the PR interval until a QRS is dropped and the cycle starts again with a shorter PR interval that progressively lengthens.
2 nd Degree Heart Block (Mobitz II; Classical type)	<p>Patterned dropping of QRS complex with a fixed ratio of atrial depolarizations (P waves) to conducted beats with a consistent PR interval throughout.</p> <p>Higher risk than Mobitz I. Intermittent block of conduction of some beats to the ventricle without progressive prolongation of the PR interval. Potentially may progress to complete heart block.</p> <p>Related to His bundle or bundle branch dysfunction (Curley, 2001).</p>
3 rd Degree Heart Block/ Complete Heart Block	Complete block of AV node resulting in AV dissociation between atrial and ventricular events. No relationship between the P waves and QRS complexes.

Specific Etiologies/Categories of Cardiac Arrhythmias:

As noted earlier, there are many reasons for arrhythmias. Increased end diastolic pressures resulting in atrial or ventricular stretch, valvular dysfunction, tumors, multiple surgeries, scarring and ischemia all play a significant role in arrhythmia generation. Cardiac swelling, pro-arrhythmic drugs, acid/base and electrolyte imbalance are also frequent etiologies of rhythm issues (Shaddy, 2011).

- **Neonatal Arrhythmias**

Common arrhythmias in neonates with structurally normal hearts are premature atrial contractions (PAC's), atrial flutter, atrioventricular reentry tachycardia (AVRT), permanent junctional reciprocating tachycardia (PJRT), ventricular tachycardia, and heart block.

Neonatal heart block is associated with maternal autoimmune disease, i.e. systemic lupus.

- **Congenital Heart Disease**

Early post operative arrhythmias usually seen are sinus tachycardia, sinus bradycardia, SVT, JET, complete AV block, and less frequently ventricular tachycardia (Allen, 2011).

Post-operative arrhythmias result from manipulation or injury of the conduction system. The site of surgical repair may increase the risk of certain types of arrhythmias observed.

Late post operative arrhythmias such as atrial flutter, and/or intra-atrial reentrant tachycardia are seen months to years after surgery. These arrhythmias are observed more often with Fontan, Mustard, Senning and tetralogy of Fallot repairs (Brugada, 2013). These tachyarrhythmias can result in poor ventricular function and decreased quality of life. Common late post operative arrhythmias include atrial tachycardia which may be seen in 50% of Fontan patients and tend to recur after a period of time (Shaddy, 2011).

- **Arrhythmias Associated with Specific Congenital Cardiac Malformations:**

CONGENITAL	EARLY POST OPERATIVE ARRHYTHMIAS
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HEART DEFECT	
Aortic Arch with VSD	JET (Zampi 2012).
Severe Aortic Stenosis Aortic Valve Surgery	Myocardial ischemia from severe left ventricular outflow obstruction, LV hypertrophy and strain resulting in ventricular arrhythmias (Allen, 2001). Conduction abnormalities and complete heart block may be seen post surgical resection of sub-aortic obstructive tissue (May, 2005). Although remote to the conduction system, junctional tachycardia may occur (Grosse-Wortmann, 2010). Prone to VT (Shaddy, 2011).
Atrial Septal Defect (ASD)	Sinus node dysfunction and transient atrial arrhythmias, atrial flutter, atrial fibrillation, ventricular tachycardias (Brugada, 2013; Hanash, 2010).
Atrioventricular Septal Defect (AVSD)	Transient and permanent sinus node dysfunction, supraventricular arrhythmias (Keane, 2006); JET (Zampi 2012); AV block; and VT (May, 2005). Grosse-Wortmann et al found that complete AV block was more common post operative repair of <i>complete</i> AVSD. (Grosse-Wortmann, 2010).
Congenitally Corrected Transposition of the Great Arteries (cc-TGA /L-TGA)	Accessory pathways (Huh, 2010); AV Block: 2 nd & 3 rd degree (Aziz 2013, Deal, 2008); ventricular ectopy. Congenital AV block may preexist due to intrinsic structural malformation (Walsh, 2007)..
Cor–Triatriatum	Sinus bradycardia, atrial tachyarrhythmias, AV conduction disturbances (May, 2015).
D-Transposition of the Great Arteries (D-TGA)	Sinus bradycardia, sinoatrial block, junctional rhythm, JET, premature atrial contractions, Mobitz 1, VT (Brugada, Decker 2012; Zampi 2012; Aziz 2013). Prone to VT if repaired with atrial level switch procedures, Senning or Mustard (Shaddy, 2011). Late complications of arrhythmias in the Jatene arterial switch procedure are rare. Late complications of atrial switch (Senning/Mustard) are that greater than 50% of patients have serious arrhythmias (Williams, 2005).
Ebstein’s Anomaly of the Tricuspid Valve	Common to have rhythm disturbances related to atrial and ventricular dilatation and conduction disturbances (May, 2005): accessory pathways (Huh, 2010); WPW and VT (Curley, 2001), SVT, atrial fibrillation, atrial flutter (Hanash, 2010); 1 st degree heart block and rarely 3 rd degree heart block (Park, 2006). Congenital accessory pathways such as WPW may preexist due to intrinsic structural malformation (Walsh, 2007).
Heart Transplant	Intraatrial reentrant tachycardia (LaPage, 2010), AET. Sinus bradycardia, AV block in a small percentage of children. Supraventricular and ventricular arrhythmias are relatively uncommon and may indicate rejection (Allen, 2001).
Pulmonary Atresia with Intact	Rare rhythm disturbances observed (May, 2005). Ventricular arrhythmias if coronary sinusoids with ischemia.

Ventricular Septum	
Pulmonary Atresia with a VSD	Sometimes AV conduction abnormalities observed (May, 2005).
Single Ventricle - Hypoplastic Left Heart Syndrome (HLHS)	Atrial arrhythmias
Single Ventricle – Bidirectional Cavopulmonary (Glenn) Connection	Transient sinus node dysfunction (Allen, 2001).
Single Ventricle - Fontan	Sinus node dysfunction (May, 2005), atrial reentrant tachycardia: atrial flutter, atrial fibrillation, intra-atrial tachycardia (Huh, 2010), VT (Park, 2006). Early or late SVT, junctional rhythm, accelerated junctional rhythm and VT (Grosse-Wortmann, 2010).
Tetralogy of Fallot (TOF)	Sinus node dysfunction, supraventricular tachycardias – atrial flutter, accelerated junctional rhythm, JET, AV blocks, VT. (Brugada, 2013; Aya, 2012; Zampi, 2012; Deal, 2008). Right bundle branch block (RBBB) (Keane, 2006; May 2005; Allen, 2001). Prone to VT due to the volume loading on the RV causing RV dilation, failure, and increased right sided pressures. Predisposes patient to SCD (Shaddy, 2011).
Total Anomalous Pulmonary Venous Return (TAPVR)	Atrial arrhythmias, JET (Zampi, 2012), sinus bradycardia, AV conduction disturbances (May, 2005).
Truncus Arteriosus	AV conduction disturbances (May, 2005); ventricular arrhythmias due to the right ventriculotomy.
Tricuspid Atresia	Supraventricular arrhythmias (Keane, 2006); atrial ectopy, flutter, fibrillation (Curley, 2001).
Ventricular Septal Defect (VSD)	Junctional rhythm, accelerated junctional rhythm, JET, VT (Grosse-Wortmann, 2010), AV conduction block.

- **Inherited Cardiomyopathies:**

Genetic predisposition to cardiac arrhythmias with an increased risk of sudden cardiac death (SCD)

CARDIO-MYOPATHIES	PATHOPHYSIOLOGY	CARDIAC ARRHYTHMIAS
Hypertrophic Cardiomyopathy (HCM)	Hypertrophic myocardium with asymmetric septal hypertrophy.	VT, SCD.
Dilated Cardiomyopathy (DCM)	Dilated poorly contractile ventricles.	SVT, VT (Hanash, 2010), SCD.

Arrhythmogenic Right Ventricular (RV) Cardiomyopathy (ARVC) or Dysplasia	A form of dilated cardiomyopathy. Fibrofatty replacement of the RV wall myocytes and patchy areas of fibrosis with progressive RV dysfunction and enlargement (Allen, 2001).	RV tachyarrhythmias with variable response to beta blockers and to catheter ablation.
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- **Channelopathies – Electrical Myopathies:**

CARDIO-MYOPATHIES	PATHOPHYSIOLOGY	CARDIAC ARRHYTHMIAS
Long QT Syndrome (LQTS)	Identified by prolonged QT interval corrected for heart rate (QTc). QT interval greater than 0.46 seconds, with upper normal limit of 0.44 seconds (Park, 2008) Acquired or congenital Can result secondary due to: - drugs (i.e. amiodarone, procainamide, sotalol, tricyclic antidepressants) and/or - electrolyte imbalance (hypomagnesaemia, hypocalcaemia, hypokalemia)	High risk of bursts of VT such as runs of Torsades de Pointes, progressing to VF and SCD. May present with syncope, seizures or SCD.
Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)	Polymorphic ventricular tachycardia. CPVT is initiated by stimulation of the adrenergic receptors from stress, emotion or exertion/physical activity. Found in normal hearts with normal coronary arteries and normal ECG's.	VT, ventricular fibrillation, and SCD.
Brugada Syndrome (BrS)	Autosomal dominant genetic In 20% of cases, mutations in the sodium channel are thought to be causative (Park, 2008).	History of ventricular arrhythmias – ventricular fibrillation, syncope and SCD. Marked by RBBB and striking ST elevation in V1-V3. ECG manifestation and arrhythmias most likely during times of fever (Keane, 2006).

- **Inflammatory Disease:**

ACQUIRED	PATHOPHYSIOLOGY	CARDIAC ARRHYTHMIAS
Myocarditis	Viral myocarditis is a cell mediated immunologic reaction. Myocardium may have lymphocyte infiltration, necrosis and scarring (Park, 2006). Myocarditis may lead to cardiomegaly and congestive heart failure, hemodynamic compromise, shock and death. Cells undergo lymphocyte infiltration, necrosis and	Risk SCD from VT and AV block (Hanash, 2010).

Clinical Assessment:

Review history: family history (heart disease, death at young age, sudden death, and seizures); neonatal history; child's personal history of syncope, palpitations, racing heart beat, seizures, exercise intolerance, family, feeding intolerance; genetics, congenital cardiac malformations; diagnostic investigations, previous surgical repair and post surgical anatomy. Inquire about events preceding rhythm disturbance.

Clinical assessment: irritability, feeding intolerance, respiratory distress, tachycardia/bradycardia for age, irregular heart rate/pulse, decreased capillary refill time, lethargy, congestive heart failure, decreased level of consciousness, syncope, absent pulses/cardiac arrest.

Heart Rate: Need to be familiar with normal heart rate for different ages. Infants generally have heart rates greater than 80 beats/min and less than 170 beats/min. Children usually have heart rates greater than 60 beats/min and less than 140 beats/min. Heart rates above these ranges are concerning and warrant further assessment. Consider the appropriate heart rate response for physiology. Cardiac assessment includes auscultation of heart sounds for murmurs, extra heart sounds, abnormal activity of the precordium palpation for heaves and thrills, assessment of perfusion, pulses, capillary refill time, blood pressure and assessment of vital signs. Cutaneous saturation or pulse oxymetry is part of the cardiorespiratory assessment and should be assessed. Identify tolerance of the arrhythmia through assessment of clinical symptoms.

Profound hemodynamic effects may result from loss of AV synchrony such as JET, AET or AV block, heart rate that is too slow or too fast, VT or VF. This is worse in the context of preexisting myocardial dysfunction or palliated physiology. A rapid heart rate results in decreased diastolic and coronary artery filling times.

Critical Thinking Points:

- Infants and children experience early post operative arrhythmias more frequently than neonates after repair of congenital heart disease. Neonates have a higher risk of JET and this is increased with VSD closure procedures. Children who were older had greater incidence of PAC's, PJC's, PVC's and VT (Grosse-Wortmann, 2010). In general, surgeries that involve the conduction tissue around the membranous septum and AV node such as VSD and AVSD can be at increased risk of JET, heart block. Tetralogy of Fallot and surgeries that involve the coronary arteries can be at increased risk of ventricular arrhythmias. Any surgery that requires incision into the ventricular myocardium has an increased risk of ventricular ectopy. Neonatal surgeries such as Truncus arteriosus, HLHS and arterial switch procedures are at higher risk.
- Patients with pre-operative arrhythmias are more likely to continue to have post operative arrhythmias at the time of discharge. Pre-operative arrhythmias are often benign and thought to be caused by injury to/edema of the conduction system, enlargement of chambers or hypoxia (Grosse-Wortmann, 2010).
- The first 24 hours is the period of time with the highest prevalence for post operative arrhythmias. Myocardial injury related to ischemia and reperfusion injury in combination with mechanical surgical injury can lead to a pro-arrhythmic state.
- Cardiac arrhythmias may cause low cardiac output states. Decision to treat an arrhythmia should be based on hemodynamic necessity not just for making the rhythm appear normal (Decker, 2012).
- Syncope with genetic arrhythmogenic disorders is a predictor of risk (Priori, 2013).

Tachycardias increase myocardial oxygen consumption and may alter the balance between oxygen delivery and oxygen consumption. Consider monitoring mixed venous oxygen saturation (central venous saturation) and lactate levels in critically ill children. Lactate levels should be less than 2.0 mmol/L. There is worse prognosis with lactate levels above 4 mmol/L or the presence of high lactate levels that are not decreasing. The normal mixed venous

oxygen saturation range is 70-80% in children with normal hearts. Children with single ventricle physiology or cyanotic heart disease usually have a mixed venous oxygen saturation of 45-55%. The difference between arterial O₂ saturation and the central venous saturation, should normally be between 25-30%.

- Wide complex tachycardia is treated as VT unless proven otherwise.
- Cardiopulmonary bypass may lower the levels of antiarrhythmic medications that were therapeutic prior to surgery, placing the child at an increased risk for complications (Decker, 2012).
- Consultation with cardiology and follow-up may be warranted for investigation of structural heart disease, cardiomyopathy, or thrombus formation (Hanash, 2010).

Diagnostic Evaluation:

This section outlines diagnostic tests that may be considered in order to ensure an accurate diagnosis and impact of arrhythmia.

- Recording baseline pre-operative and post-operative rhythm strips is optimal. Any Abnormal ECG's should be compared to baseline.
- Document the rhythm disturbance by a 12 or 15 lead paediatric electrocardiogram. Paediatric 15 Lead ECG includes right sided leads V4R, V5R, V6R. This can be invaluable in accurate identification of the type of arrhythmia.
- The patient should be monitored continuously. A Holter electrocardiogram may be of value in identification of the arrhythmia events.
- Perform an atrial electrocardiogram using the atrial pacing wire in post cardiac surgical patients, where P-waves cannot be clearly identified.
- It can be helpful to capture electrical evidence of termination of the tachycardia on a 15 lead ECG or rhythm strip.
- Test blood levels of K⁺, Ca⁺⁺, Mg⁺⁺; thyroid function tests; complete blood count, toxicology screen (Hanash, 2010). Electrolyte imbalances are often associated with rhythm disturbances. If suspicious of myocarditis or with worsening cardiac function check viral etiologies (Hanash, 2010). Lab testing may include cardiac enzymes: troponin levels and CPK-MB – markers of myocardial injury.
- A chest X-ray may demonstrate enlargement of the heart.
- ECHO: Echo provides a qualitative and quantitative evaluation of cardiac function to rule out underlying structural heart disease, thrombus formation and ventricular dysfunction. A quantitative value of ejection fraction can be reported.
- Use of pharmacological agents such as adenosine and procainamide can assist with diagnosis of arrhythmias
- Exercise testing may be used to provoke and diagnose arrhythmias and associated symptoms.
- A catecholamine challenge or transoesophageal pacing can also be used to provoke arrhythmias in a controlled environment.
- Invasive electrophysiology studies with cardiac catheterization help to identify ectopic foci and accessory pathways which can be mapped and ablated (Curley, 2001).

Recommendations for Acute Treatment:

A clear diagnosis should be documented by electrocardiogram. Priority is providing hemodynamic stability if patient is symptomatic. The primary goal is to establish rhythm and rate control (Hanash, 2010). Treatment depends upon numerous factors: type of arrhythmia, age, duration, frequency, clinical presentation, tolerance and cardiac function. Determine with each arrhythmia whether the patient is hemodynamically stable or unstable. Manage arrhythmias that present with hemodynamic instability aggressively. Treatment choices (pharmacotherapy, electrical pacing, D/C cardioversion or defibrillation, ablative therapy) depend upon understanding the mechanism of arrhythmia (Allen, 2011). For patients that present with life threatening tachyarrhythmias, cardioversion or defibrillation should be administered emergently without delay.

Post operative arrhythmias may be responsive to correction of electrolytes, inducing hypothermia, decreasing inotropic and/or vasoconstrictor infusions and the use of appropriate antiarrhythmic medications (Jhang, 2010). The severity of symptoms can range from mild, non-specific such as decreased appetite and nausea to cardiac arrest. In case of cardiac arrest or significant cardiac instability cardiopulmonary resuscitation is started immediately, followed by D/C cardioversion/defibrillation for shockable rhythms and the administration of epinephrine according to the American Heart Association’s (AHA) Paediatric Advanced Life Support (PALS) Guidelines (Kleinman, 2010). There should be simultaneous treatment of the cause if known.

Use electrical energy to convert shockable rhythms in hemodynamically unstable patients. Recommend limited use of electrical energy in stable patients.

- D/C Cardioversion (synchronized delivery of electrical energy to the myocardium); usual range 0.5-1 J/kg, and may increase to 2 Joule/kg.
- Defibrillation (unsynchronized delivery of electrical energy to the myocardium); start with 2 J/kg, then increase to 4 Joule/kg, then can increase to 4-10 Joule/kg. Refer to AHA PALS Guidelines (Kleinman, 2010).

ARRHYTHMIA	MANAGEMENT [Refer to AHA PALS Guidelines (Kleinman, 2010)]
Bradycardias	<p>Treat the underlying cause: hypothermia, hypoxia, vasovagal stimulation, drug toxicity, increased intracranial pressure, hyperkalemia, drugs i.e. β-blockers, infections i.e. Lyme disease.</p> <p>Administer intravenous (IV) atropine, epinephrine, and isoproterenol. Follow with transoesophageal, transvenous, transcutaneous, or through atrial or ventricular pacing wires for fresh post operative. (Park, 2010).</p> <p>For rates less than 60 beats/min and poor perfusion begin cardiopulmonary resuscitation.</p>
<p>Narrow Complex Tachycardias:</p> <p>SVT is used as a collective term</p>	<p><u>Sinus tachycardia:</u> Treat the underlying cause: fever, shock, hypovolemia, pain, anxiety, myocarditis, congestive heart failure, drugs.</p> <p>The following rhythms with stable hemodynamics can be treated with medications. For hemodynamically unstable patients electrical energy should be applied using cardioversion at 0.5-2 Joules/kg.</p> <p><u>Atrial Flutter and Atrial Fibrillation:</u> Treat with amiodarone, digoxin, or β-blockers such as metoprolol, esmolol propanolol. D/C cardioversion may be required if unstable. Cardioversion may be performed without anticoagulation if patient presents within 48 hours of initiation of tachycardia and there is no clot in the heart. Anticoagulation with heparin or warfarin if arrhythmia duration greater than 48 hrs in duration or uncertain onset to prevent thrombus movement from the atrium. Rapid atrial overdrive pacing via esophagus or atrial wire may be effective (Park, 2008; Allen, 2001).</p> <p><u>AV nodal and AV re-entrant tachycardia:</u> Stable: initial vagal maneuvers i.e. ice to face for younger children elicits the dive reflex,</p>

	<p>Valsalva maneuver for older children. If no success consider using the following antiarrhythmic medications: IV adenosine, amiodarone, sotalol, procainamide, or esmolol.</p> <p>Other options include transoesophageal, epicardial or transcutaneous atrial over-drive pacing. If hemodynamically unstable, perform D/C cardioversion.</p> <p>Calcium channel blocker verapamil is reserved for children greater than 2 year due to concerns of cardiovascular collapse. Caution with administration.</p> <p>Maintenance therapy may include β-blockers (Hanash, 2010). Examples include esmolol, propranolol, and metoprolol.</p> <p>Consider radiofrequency catheter ablation of accessory pathway if conversion fails or arrhythmia is recurrent.</p> <p><u><i>Atrial Ectopic Tachycardia (AET)/Junctional Ectopic Tachycardia (JET):</i></u> AET and (JET) usually do not respond to typical treatments used for reentrant tachycardia such as adenosine and cardioversion (Allen, 2001). The goal is to restore AV synchrony and reduce the ventricular response rate with drugs like amiodarone, digoxin or β-blockers like esmolol. (Park, 2010). Treat hyperthermia aggressively by exposure to topical ice, actively cooling and muscle relaxation. These rhythms respond to atrial overdrive pacing at a rate at about 10-20% above the ectopic rate (Grosse-Wortmann; Schwartz, 2014). Provide sedation and reduce the catecholamine infusions providing the patient tolerates a reduction in vasoactive drugs. Ensure patient is well sedated. Pull back invasive CV lines in the RA if they are contributing to the arrhythmia reducing a potential source of irritation of the atrial wall (Schwartz, 2014). A combination of strategies may slow the ectopic rate and improve hemodynamics.</p> <p><i>AET:</i> Slowing down the rate and/or the AV conduction are 2 ways of managing AET. (Schwartz, 2014) Catheter radiofrequency ablation of the His bundle or surgical mapping and resection may be required in rare situations (Allen, 2001).</p> <p><i>JET:</i> Adenosine can be used to slow the ectopic by blocking retrograde conduction through the AV node for a few beats helping to differentiate and diagnose the true origin of the rhythm although not solving the rhythm disturbance. Use of mild cooling 34 – 35 degrees centigrade in order to achieve a heart rate of less than 160 beats/minute is desirable. Application of AAI pacing above the JET rate may preserve AV synchrony. In cases of marked hemodynamic instability, extracorporeal membrane oxygenation (ECMO) may be employed (Schwartz, 2014).</p>
Wandering Atrial Pacemaker	Treatment is not usually required.
Junctional/Nodal Rhythm	Treatment ranges from doing nothing to transthoracic, transcutaneous, transoesophageal, or transvenous pacing. Monitor the hemodynamic effect and treat appropriately. May need to pace.
Wide QRS Complex Tachycardia:	<p>Any hemodynamically significantly wide QRS tachycardia should be treated as VT. VT can be difficult to differentiate from SVT with aberrancy. Rhythm may progress to VF. Establish if a pulse can be palpated differentiating VT with a pulse and pulseless VT.</p> <p><u>Stable:</u> Loading dose of amiodarone or, then a maintenance infusion. Esmolol may also convert rhythm. If hemodynamically stable, obtain a cardiology consult to determine exact type of tachycardia.</p>

	<p><u>Unstable:</u> If pulse, refer to PALS algorithm for VT with pulse. D/C shock - synchronized cardioversion</p> <p><u>Pulseless:</u> If no pulse initiate cardiopulmonary resuscitation (CPR), administer D/C shock – defibrillation and administer epinephrine. Refer to AHA PALS Guidelines (Kleinman, 2010)</p> <p><i>Torsades de Pointes:</i> Treat with magnesium sulfate.</p>
Ventricular Fibrillation	CPR and D/C shock – defibrillation; Epinephrine every 3-5 minutes. Refer to AHA PALS Guidelines (Kleinman, 2010)
Ideoventricular Rhythm	HR less than 60 beats/min and poor perfusion begin CPR. Refer to AHA PALS Guidelines (Kleinman, 2010). Administer intravenous (IV) atropine, epinephrine, or isoproterenol. Follow with transoesophageal, transvenous, transcutaneous, or transthoracic pacing. (Park, 2010).
1 st Degree Heart Block	Usually asymptomatic and does not require treatment
2 nd Degree Heart Block (Mobitz I , Wenckebach)	If heart rate < 60 b/min and symptomatic, administer intravenous (IV) atropine, epinephrine, or isoproterenol. Follow with transoesophageal, transvenous, transcutaneous, or transthoracic pacing. (Park, 2010).
2 nd Degree Heart Block (Mobitz II; Classical)	For palliated physiology or presence of myocardial dysfunction treatment may be required with heart rates > 60 b/min if hemodynamically unstable
3 rd Degree Heart Block/Complete	

Recommendations for Treatment of Arrhythmias that Recur or Fail to Respond to Other Therapies:

Invasive electrophysiological study and catheter ablation:

Radiofrequency catheter ablation may be used to cure tachyarrhythmias that are frequent or fail to respond to treatment (Brugada, 2013). Accessory pathways such as in Wolff-Parkinson-White syndrome and AV nodal reentrant tachycardia respond well. Ablation therapy of AV accessory pathways is safe and effective. Ablation of IART and VT in patients with structurally abnormal hearts may have lower success rates and may experience recurrence of the rhythm (Shaddy, 2011).

Transcatheter cryoablation. Cryoenergy necroses cells at the arrhythmogenic origin.

Surgical arrhythmia ablation may remove aggravating source of arrhythmia for patients with frequent and intractable rhythms (Shaddy, 2011).

Management of channelopathies should focus on prevention and treatment of sudden cardiac death. Lifelong β -blockers may be used as well as implantable cardioverter defibrillators for refractory arrhythmias or history of life threatening events.

β -blockers are indicated and remain first line of therapy for patients with long QT syndrome, except in those patient with asthma, where β -blockers are contraindicated (Priori, 2013).

Device therapy - pacemakers: temporary and permanent pacemakers for sinus node dysfunction and complete heart block (congenital/acquired). Consider permanent pacing after 1-2 weeks post operative for third degree heart block (Grosse-Wortmann, 2010).

Device therapy - implantable cardioverter defibrillator (ICD): Subcutaneous/submuscular placement of an ICD device for patients who are at risk of sudden death, for example patients with long QT syndrome can be life saving (Priori, 2013; Brugada, 2013). In patients with life threatening events of arrhythmogenic nature or those at risk for SCD, placement of an ICD device should be strongly considered. Risk-benefit should be considered and patient should be consented for the procedure. Devices should be programmed carefully to prevent undue shocks (Priori, 2013). ICD's have proven beneficial in adults in preventing SCD. They can be used safely with good effect in pediatric population although less well studied (Shaddy, 2011). Patient with tetralogy of Fallot is a common diagnosis for ICD placement. One adverse effect of this device therapy is inappropriate shock delivery which in the tetralogy of Fallot patients has been noted to be as high as 28% vs. cardiomyopathy patients, experiencing it in 13% (Shaddy, 2011).

Extracorporeal Membrane Oxygenation (ECMO) may be required in hemodynamically unstable patients as a means to establish effective anti-arrhythmic therapy. Stabilizing hemodynamics permits the opportunity to proceed with alternative strategies such as going to the catheterization lab for electrophysiology study and possible ablations. ECMO could be a bridge to cardiac transplantation with progressive hemodynamic instability (Jhang, 2010).

Associated Complications of Arrhythmias:

Arrhythmias can lead to myocardial dysfunction, heart failure, cardiac arrest, SCD, intra-cardiac clot formation, seizures, syncope, and acquired cardiomyopathy.

- These complications can happen any time during a course of arrhythmias. Complications may present early post op or late.
- Arrhythmias may result in heart dysfunction and failure, followed by sudden cardiac death.
- Some medication that are used to treat arrhythmias can cause side effects including prolonged QT, changes to the QRS duration; and pro-arrhythmia effects; organ injury such as liver, lung and thyroid toxicity.
- Children with decreased cardiac function because of arrhythmias are prone to clot formation. Consider anticoagulation until the arrhythmia is under control and cardiac function improved.

Complications of Treatments (Medications and Devices)

- Consider pacemaker complications such as lead failure due to epicardial lead fracture/exit block; transvenous insulation break/dislodgement, hematomas, bleeding, pneumothorax, cardiac perforation, tamponade, venous thrombosis; generator failure, battery depletion; infections (superficial vs. deep pocket). Venous occlusion rates have been noted to be as high as 15-21% with transvenous pacing or ICD's (Shaddy, 2011).
- Complications of catheter ablation are heart block, chamber perforation, thromboembolism, pericardial effusion/tamponade and thromboembolism. The arrhythmia may recur after original success of ablative therapy.
- Monitor for ICD complications such as infection and inappropriate firing. Five year lead failure or fracture has been noted to be as high as 15% (Shaddy, 2011).
- Children with CHD may experience late arrhythmias with symptoms appearing into adulthood (Huh, J, 2010). Sudden cardiac death may occur and is a late complication of repair especially for tetralogy of Fallot, TGA with Interatrial repair Mustard/Senning, Aortic Stenosis and Coarctation of Aorta. LV obstructive disorders may result in complications due to aortic aneurism that ruptures, coronary artery disease, left ventricular hypertrophy and cardiomyopathy (Allen, 2001).

Special Considerations:

- Infants and children present clinically in different ways. Neonates/infants may present with feeding intolerance, poor weight gain, lethargy or irritability, and pallor and diaphoresis. Older children can present with symptoms of dizziness, exercise intolerance, syncope, chest pain or palpitations.
- Post surgical scarring, suture lines, and conduits can be substrates for future appearance of rhythm disturbances.
- Sports participation should be discussed with the physician, as activity places high risk children/adolescents with inherited cardiomyopathies at risk of cardiac arrhythmias and SCD.
- Children and their families with a positive family history, a history of channelopathies or arrhythmogenic cardiomyopathy are referred for genetic consultation as these can lead to a lethal outcome. Genetic based pediatric arrhythmias can lead to lethal outcomes (Brugada, 2013).
- Consider timing, sedation and anticoagulation prior to administering synchronized cardioversion.
- Anticoagulation is recommended prior to conversion for sustained atrial tachyarrhythmias - atrial fibrillation, atrial flutter greater than 24- 48 hrs (i.e. Fontan). Formation of thrombi may result in emboli and stroke (Allen, 2011).
- Adenosine and B-blockers should be used cautiously with asthmatic patients due to bronchial constriction.
- Complications of implanted devices (pacemakers/antitachycardic defibrillators) are more common in pediatric patients. Consider the psychological impact of device placement.
- Cardiac Resynchronization Therapy (CRT): Biventricular synchronous pacing may be used in patients with mechanical or electrical ventricular dyssynchrony causing poor function and congestive heart failure. Biventricular synchronous pacing may improve cardiac function.

Conclusion:

Cardiac arrhythmias can affect the quality of life, and lead to morbidity and mortality of infants and children. Prompt diagnosis and appropriate early management can potentially prevent life threatening events and improve functional capacity and quality of life for the child and their family. Acute hemodynamic state of the child determines course of action. Treatment focuses on termination of the arrhythmia itself and prevention of future recurrences. The combination of pharmacotherapy, electrical therapies, antitachycardic pacing, antiarrhythmic devices and surgical or interventional catheterization lab the treatments are useful in managing patients with cardiac arrhythmias both in the short term and long term.

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