Cardiovascular Diseases
Cardiomyopathies
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Cardiomyopathy

Types and Etiology

There are **three types** of cardiomyopathy based on anatomic and functional features:

1. Dilated
2. Hypertrophic
3. Restrictive

- **Dilated cardiomyopathies** are the most common. They are often idiopathic, but they may be due to infection (echovirus or coxsackie B virus) or be postinfectious, familial, or secondary to systemic disease or to cardiotoxic drugs, and metabolic.
- **Hypertrophic cardiomyopathies** are usually familial with autosomal dominant inheritance but may occur sporadically.
- **Restrictive cardiomyopathies** are rare; they may be **idiopathic** or associated with **systemic diseases** (amyloidosis,sarcoidosis,hemochromatosis).

CLINICAL MANIFESTATIONS

Dilated cardiomyopathies result in enlargement of the left ventricle only or of both ventricles. Myocardial contractility is variably decreased.

Children with dilated cardiomyopathy present with signs and symptoms of inadequate cardiac output and **heart failure**. Tachypnea and tachycardia are present **on examination**. Peripheral pulses are often weak because of a **narrow pulse pressure**. Rales may be audible on auscultation. The heart sounds may be muffled, and an S3 is often present. Concurrent infectious illness may result in circulatory collapse and shock in children with dilated cardiomyopathies.

CLINICAL MANIFESTATIONS

Hypertrophic cardiomyopathy is initially difficult to diagnose.

Infants, but not older children, frequently present with signs of heart failure. Sudden death may be the initial presentation

In older children: Dyspnea, fatigue, chest pain, syncope or near-syncope, and palpitations may be present. A murmur is heard in more than 50% of children referred after identification of an affected family member.

Restrictive cardiomyopathies

Presenting symptoms usually include dyspnea exacerbated by a respiratory illness, syncope, hepatomegaly, and an S4 heart sound on examination

IMAGING STUDIES

Cardiomegaly usually is seen on **chest radiographs** for all three types of cardiomyopathies.

The electrocardiogram (ECG) in

dilated cardiomyopathy may have nonspecific ST-T-wave changes and left ventricular hypertrophy. ECG evidence of right ventricular hypertrophy is present in 25% of children with cardiomyopathy

hypertrophic cardiomyopathy

is universally abnormal, but changes are nonspecific. Primary hypertrophic cardiomyopathy is associated with a prolonged QT interval.

Children with **restrictive cardiomyopathies** may show atrial enlargement on the ECG.

IMAGING STUDIES

Echocardiography features vary by type of cardiomyopathy.

Dilated cardiomyopathies result in left atrial and ventricular dilation and depressed contractility.

hypertrophic cardiomyopathies Asymmetric septal hypertrophy and left ventricular outflow tract obstruction.

restrictive cardiomyopathies Massive atrial dilation is seen.

Endomyocardial biopsy specimens, obtained while the patient is hemodynamically stable, identify histologic type and allow tests for mitochondrial or infiltrative diseases.

TREATMENT

- Supportive therapy, including **diuretics**, **inotropic medications**, and **afterload reduction**, is provided for all three types of cardiomyopathy.
- If a specific etiology can be identified, treatment is directed at the etiology.
- Symptomatic therapy with close monitoring and follow-up is crucial.
- Because of the high mortality rate associated with all forms of cardiomyopathy, **cardiac transplantation** must be considered.

TREATMENT

Dilated

1-positive inotrops

2-Diuretics.

3- Afterload reducing agents.

4-B-Blockers.

5-Antiarrhythmics.

6-Anticoagulants.

7-Cardiac Transplant.

Hypertrophic

1-B-Blockers

2-Ca-Channels blocker

Restrictive

1-Diuretics

2-Anticoagulants

3-Cardiac Transplant

Cardiac Tumors

Cardiac tumors rare in pediatric patients. The vast majority of tumors of the heart are **benign.**

Rhabdomyomas

- Are the **most common** pediatric cardiac tumors and are associated with tuberous sclerosis in 70–95% of cases.
- Rhabdomyomas may occur at any age, from fetal life through late adolescence.
- They are often multiple, can occur in any cardiac chamber, and originate within the myocardium, often extending into the atrial or ventricular cavities. Depending on their location and size, rhabdomyomas can result in inflow or outflow obstruction, leading to cyanosis or cardiac failure; many are asymptomatic.
- Atrial and ventricular arrhythmias have been reported with rhabdomyomas and on occasion, ventricular preexcitation (Wolff-Parkinson-White syndrome) is present on electrocardiogram (ECG).

Cardiac Tumors

Fibromas

- Are the 2nd most common pediatric cardiac tumor and, in contrast to rhabdomyomas, are usually **solitary and intramyocardial**.
- The size and location of fibromas can lead to heart failure, cyanosis, or rhythm disturbances.
- There is an increased incidence in patients with **Gorlin syndrome** (3%).

Myxomas

- The most common cardiac tumor seen in adults, occur infrequently in the pediatric population.
- Myxomas are predominantly intraatrial, appear pedunculated, and are rather mobile. They may cause obstruction to inflow or outflow and may present with a murmur, heart failure, or syncope.
- On occasion, atrial myxomas are associated with systemic symptoms of fever, malaise, and arthralgia.

Cardiac Tumors

Other benign tumors include **hemangiomas**, **Purkinje cell tumors**, **papillomas, lipomas, and mesotheliomas**.

Depending on their location, these benign tumors can result in valvular function abnormalities, myocardial dysfunction, or heart block and other arrhythmias.

Malignant Tumors

Malignant pediatric cardiac tumors are much less common than benign tumors (75% vs. 25%), and the vast majority of such malignancies are sarcomas, including **angiosarcomas, rhabdosarcomas**, or **fibrosarcomas. while Lymphomas and pheochromocytomas** are rare. Tumors originating from non cardiac sources that invade, extend, or metastasize to the heart are more frequently seen than primary malignant cardiac tumors.

In pediatric patients, **Wilms tumor** and **lymphoma/leukemia** are the most common causes of such secondary tumors.

Diagnosis of Cardiac Tumor

- Noninvasive imaging with **echocardiography and/or MRI** : may be diagnostic and can determine tumor type, location, extent, and hemodynamic impact.
- **ECG and Holter** studies are valuable adjuncts when rhythm abnormalities are suspected.
- **Cardiac catheterization:** is rarely indicated but may be used to confirm tumor location, assess intra cardiac hemodynamics, and perform biopsy for histologic assessment.

Treatment of cardiac tumors

Treatment of the majority of cardiac tumors in pediatric patients is usually unnecessary.

- ***Everolimus,*** an inhibitor of the mammalian target of rapamycin (mTOR), may enhance resolution in symptomatic patients with cardiac rhabdomyomas.
- **Careful clinical follow-up and imaging are important.**
- **Antiarrhythmic medications** may be prescribed to control rhythm disorders.
- **Surgical removal of a cardiac tumor** may be indicated to relieve obstruction, improve myocardial or valve function, or control arrhythmias.
- **Heart transplantation** has been performed in cases of un resectable tumors with significant hemodynamic compromise.
- **Radiation or chemotherapy** can improve cardiac function in rare cases of lymphoma or leukemia compressing the heart with hemodynamic compromise.
- Bibliography is available at Expert Consult.

Dysrhythmias

Cardiac dysrhythmias or abnormal heart rhythms are uncommon in pediatrics.

Many pediatric dysrhythmias are normal variants that do not require treatment or even further evaluation.

Dysrhythmias

Etiology of dysrhythmias

Drugs

- Intoxication (cocaine, tricyclic antidepressants, and others)
- Antiarrhythmic agents (proarrhythmic agents [quinidine])
- Sympathomimetic agents (caffeine, theophylline, ephedrine, and others)
- Digoxin

INFECTION AND POSTINFECTION

- Myocarditis
- Lyme disease
- Endocarditis
- Diphtheria
- Guillain-Barré syndrome
- Rheumatic fever

Etiology

METABOLIC-ENDOCRINE

- Electrolyte disturbances (↓↑K+, ↓↑Ca2+, ↓Mg2+)
- Cardiomyopathy
- Thyrotoxicosis
- Uremia
- Pheochromocytoma
- Porphyria
- Mitochondrial myopathies

STRUCTURAL LESIONS

- Congenital heart defects
- Ventricular tumor
- Ventriculotomy
- Arrhythmogenic right ventricle (dysplasia)

Etiology

OTHER CAUSES

- Adrenergic-induced
- Prolonged QT interval
- Maternal systemic lupus erythematosus
- Idiopathic
- Central venous catheter

Sinus rhythm : originates in the sinus node and has a normal axis P wave (upright in leads I and aVF) preceding each QRS complex. Because normal rates vary with age, sinus bradycardia and sinus tachycardia are defined based on age.

Sinus arrhythmia : is a common finding in children and represents a normal variation in the heart rate associated with breathing.

The heart rate increases with inspiration and decreases with expiration, producing a recurring pattern on the electrocardiogram (ECG) tracing. Sinus arrhythmia is normal and does not require further evaluation or treatment.

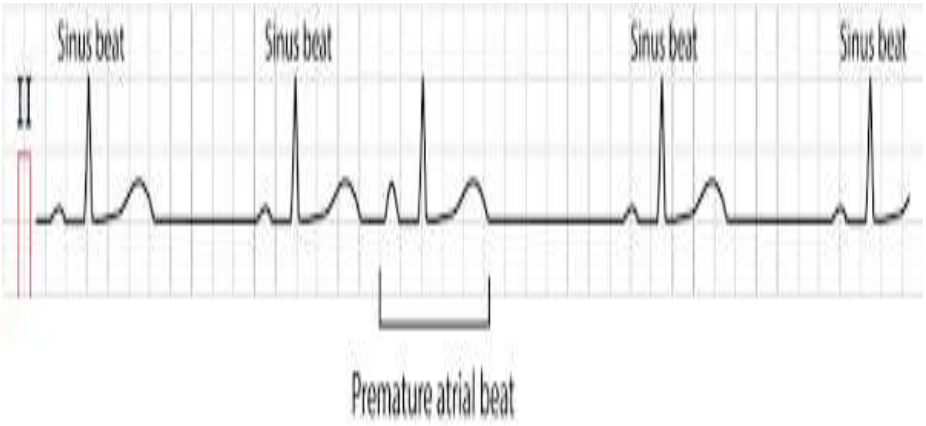
Atrial Dysrhythmias

A **wandering atrial pacemaker:** is a change in the morphology of the P waves with variable PR interval and normal QRS complex. This is a benign finding, requiring no further evaluation or treatment.

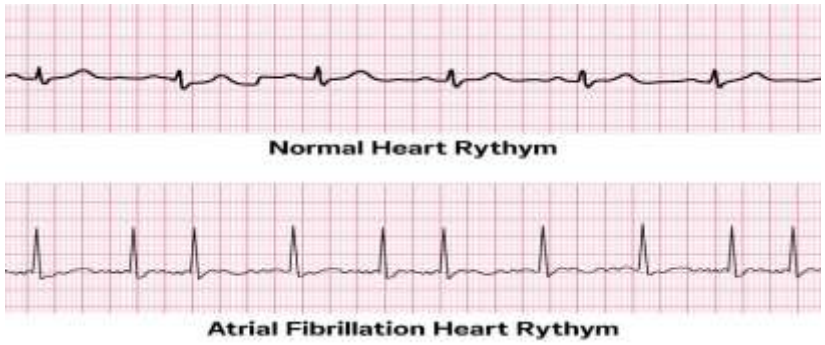
Premature atrial contractions: are relatively common prenatally and in infants. A premature P wave, usually with an abnormal axis consistent with its ectopic origin, is present. The premature atrial activity may be blocked (no QRS following it), conducted normally (normal QRS present), or conducted aberrantly (a widened, altered QRS complex). are usually benign and, if present around the time of delivery, usually disappear during the first few weeks of life.

Atrial flutter and **atrial fibrillation:** are uncommon dysrhythmias in pediatrics and usually present after surgical repair of complex congenital heart disease. They may also be seen in patients with myocarditis or in association with drug toxicity.

Atrial Dysrhythmias



Premature atrial beat



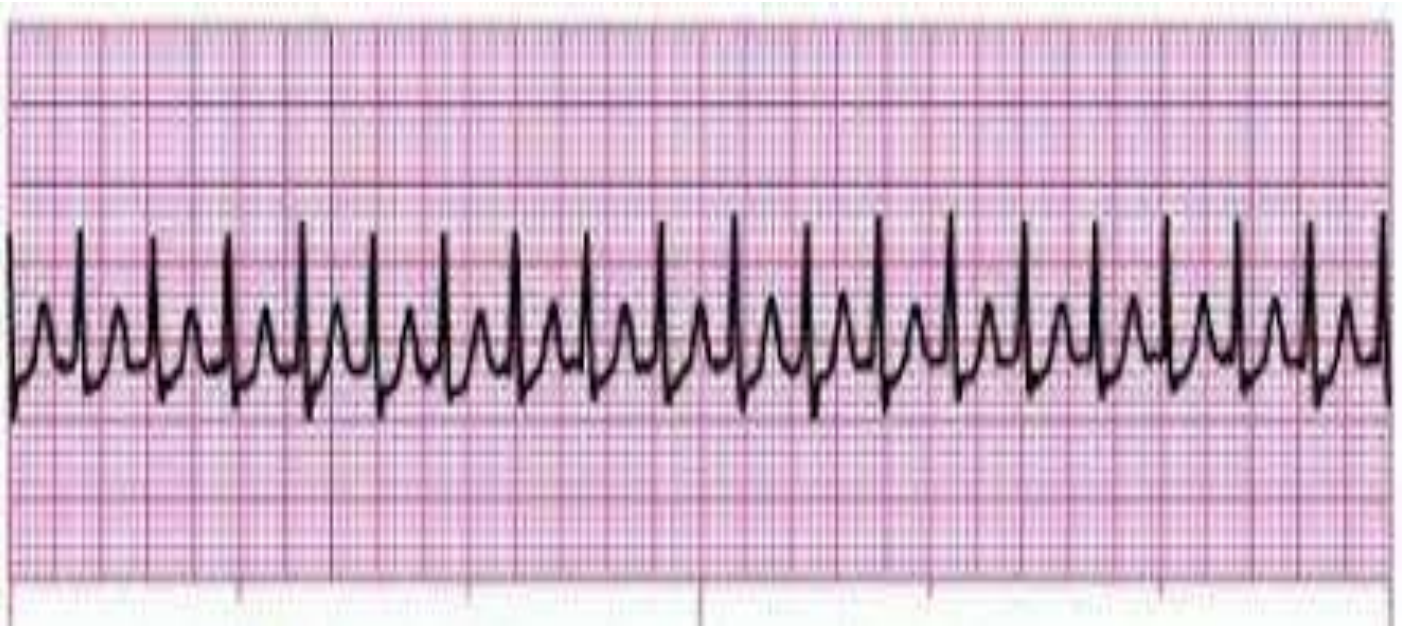
Atrial Fibrillation

Atrial Dysrhythmias

Supraventricular tachycardia (SVT) is the **most common** symptomatic dysrhythmia in pediatric patients.

- The rhythm has a rapid, regular rate with a narrow QRS complex.
- SVT in infants is often 280−300 beats per minute with slower rates for older children and adolescents. The tachycardia has an abrupt onset and termination.
- In a child with a structurally normal heart, most episodes are relatively asymptomatic other than a pounding heartbeat.
- If there is structural heart disease or the episode is prolonged (>12 hours), there may be an alteration in the cardiac output and development of symptoms of heart failure.
- Although most patients with SVT have structurally normal hearts and normal baseline ECGs, some children have Wolff-Parkinson-White syndrome or preexcitation as the cause of the dysrhythmia.

Supraventricular tachycardia



Ventricular Dysrhythmias

Premature ventricular contractions (PVCs):

are less common than premature atrial contractions in infancy but more common in older children and adolescents.

The premature beat is not preceded by a P wave, and the QRS complex is wide and bizarre. If the heart is structurally normal, and the PVCs are singleton, uniform in focus, and disappear with increased heart rate, they are usually benign and require no treatment.

Any deviation from the presentation (history of syncope or a family history of sudden death) requires further investigation and possibly treatment with antiarrhythmic medications.

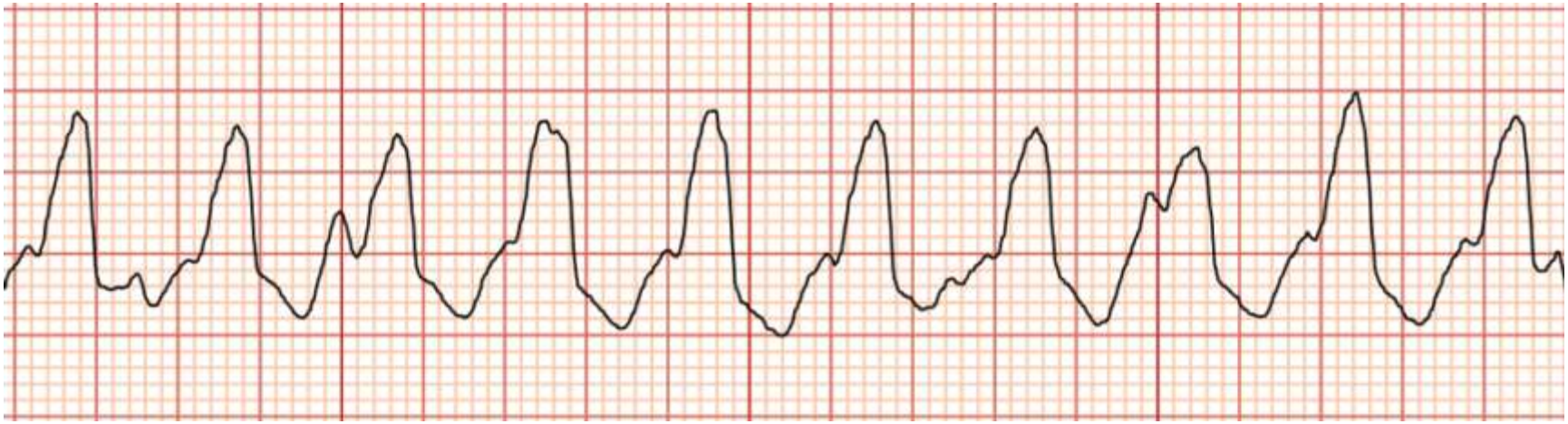
Ventricular Dysrhythmias

Ventricular tachycardia :

defined as **three or more consecutive PVCs**, is also relatively rare in pediatric patients.

- Although there are multiple causes of ventricular tachycardia,
- it usually is a sign of serious cardiac dysfunction or disease.
- Rapid-rate ventricular tachycardia results in decreased cardiac output and cardiovascular instability.
- **Treatment:** in symptomatic patients is synchronized cardioversion.
- Medical management with lidocaine or amiodarone may be appropriate in a conscious asymptomatic patient.
- Complete evaluation of the etiologic picture is necessary, including electrophysiologic study.

Ventricular Tachycardia



Heart Block

First-degree heart block: is the presence of a **prolonged PR interval**. It is asymptomatic and, when present in otherwise normal children, requires no evaluation or treatment.

Second degree heart block is when some, but not all, of the P waves are followed by a QRS complex.

Mobitz type I (also known as Wenckebach) is characterized by a progressive prolongation of the PR interval until a QRS complex is dropped. It is often seen during sleep, usually does not progress to other forms of heart block, and does not require further evaluation or treatment in otherwise normal children.

Mobitz type II is present when the PR interval does not change, but a QRS is intermittently dropped. This form may progress to complete heart block and may require pacemaker placement.

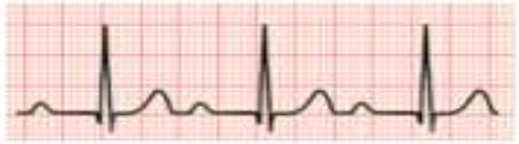
Heart block

Third-degree heart block

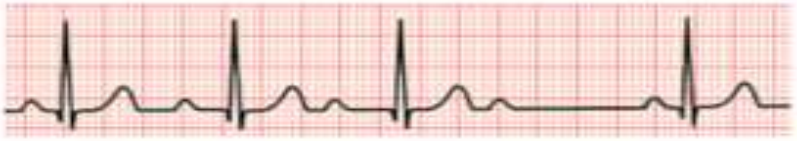
- whether congenital or acquired, is present when there is no relationship between atrial and ventricular activity. The ventricular rate is much slower than the atrial rate.
- **Congenital complete heart block** is associated with maternal collagen vascular disease (such as systemic lupus erythematosus or Sjogren syndrome) or congenital heart disease.
- The **acquired form** most often occurs after cardiac surgery but may be secondary to infection, inflammation, or drugs.

Heart Block

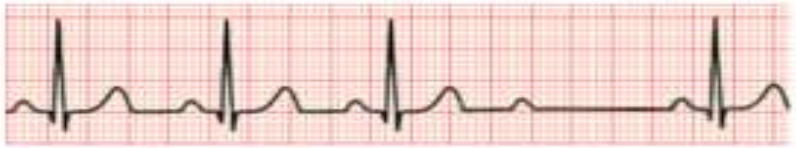
First degree AV block



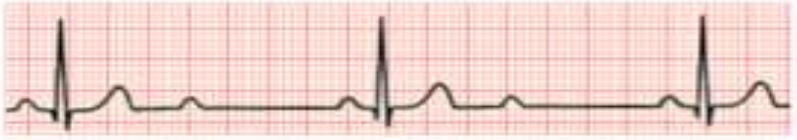
Second degree AV block (Mobitz I or Wenckebach)



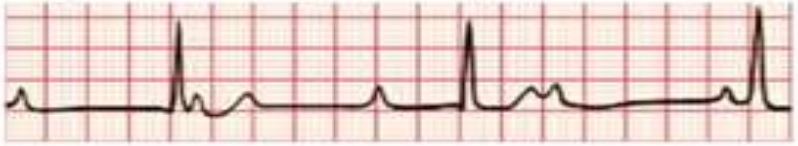
Second degree AV block (Mobitz II)

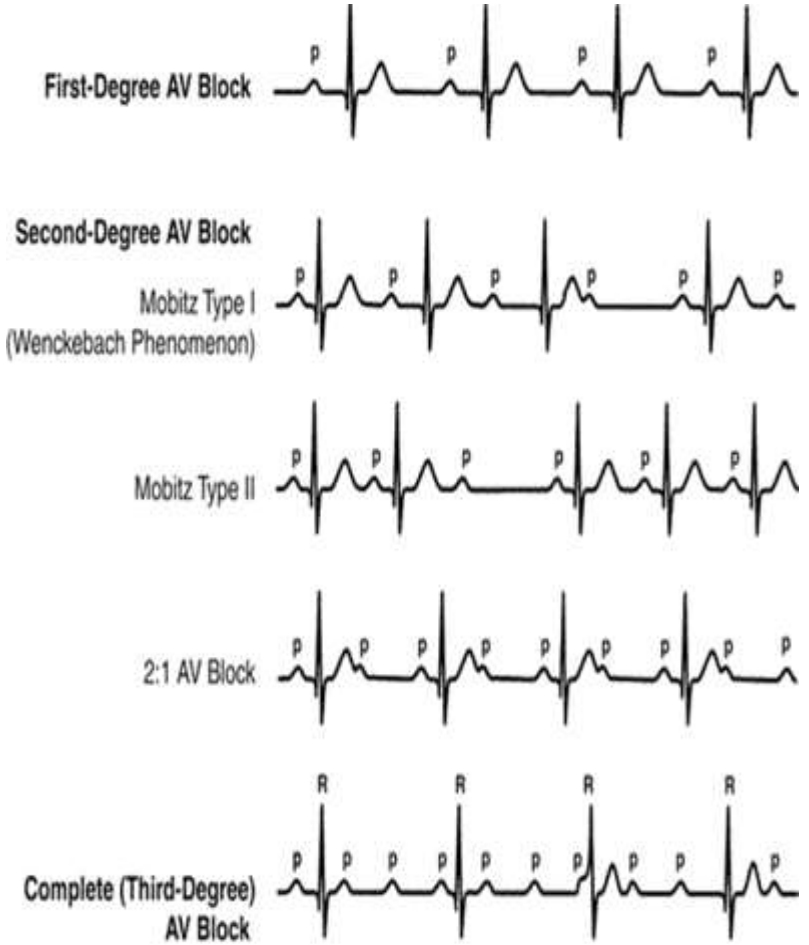


Second degree AV block (2:1 block)



Third degree AV block with junctional escape





TREATMENT

Most **atrial dysrhythmias** require no intervention.

Treatment of SVT depends on presentation and symptoms.

- Acute treatment of SVT in infants usually consists of **vagal maneuvers**, such as the application of cold (ice bag) to the face.
- Intravenous (IV) **adenosine** usually converts the dysrhythmia.
- In patients with cardiovascular compromise at the time of presentation, **synchronized cardioversion** is indicated using 1−2 J/kg. In patients with palpitations, it is important to document heart rate and rhythm during their symptoms before considering therapeutic options.
- Ongoing pharmacologic management with **a β blocker** is usually the first choice when treatment is needed. Some practitioners still use digoxin for management of SVT, but it is **contraindicated** in patients with Wolff-Parkinson-White syndrome.

Treatment of SVT

- Additional antiarrhythmic medications are sometimes needed depending on the mechanism of the SVT.
- In patients who are symptomatic or for those not wanting to take daily medications, **radiofrequency ablation** may be performed.

Treatment

- A variety of antiarrhythmic agents are used to treat ventricular dysrhythmias that require intervention .

- Management of third-degree heart block depends on the ventricular rate and presence of symptoms. Treatment, if needed, often requires placement of **a pacemaker**.

Dysrhythmias in Children

TABLE 142.2 Dysrhythmias in Children

TYPE	ELECTROCARDIOGRAM CHARACTERISTICS	TREATMENT
Supraventricular tachycardia	Rate usually >220 beats/min (range, 180-320 beats/min); abnormal atrial rate for age; P waves may be present and are related to QRS complex; normal, narrow QRS complexes unless aberrant conduction is present	Increase vagal tone (bag of ice water to face, Valsalva maneuver); adenosine; digoxin; sotalol; electrical cardioversion if acutely ill; catheter ablation
Atrial flutter	Atrial rate usually 300 beats/min, with varying degrees of block; sawtooth flutter waves	Digoxin, sotalol, cardioversion
Premature ventricular contraction	Premature, wide, unusually shaped QRS complex, with large inverted T wave	None if normal heart and if premature ventricular contractions disappear on exercise; lidocaine, procainamide
Ventricular tachycardia	>3 Premature ventricular beats; AV dissociation; fusion beats, blocked retrograde AV conduction; sustained if >30 sec; rate 120-240 beats/min	Lidocaine, amiodarone, procainamide, propranolol, cardioversion
Ventricular fibrillation	No distinct QRS complex or T waves; irregular undulations with varied amplitude and contour, no conducted pulse	Nonsynchronized cardioversion
Complete heart block	Atria and ventricles have independent pacemakers; AV dissociation; escape-pacemaker is at atrioventricular junction if congenital	Awake rate <55 beats/min in neonate or <40 beats/min in adolescent, or hemodynamic instability requires permanent pacemaker
First-degree heart block	Prolonged PR interval for age	Observe, obtain digoxin level if on therapy
Mobitz type I (Wenckebach) second-degree heart block	Progressive lengthening of PR interval until P wave is not followed by conducted QRS complex	Observe, correct underlying electrolyte or other abnormalities
Mobitz type II second-degree heart block	Sudden nonconduction of P wave with loss of QRS complex without progressive PR interval lengthening	Consider pacemaker
Sinus tachycardia	Rate <240 beats/min	Treat cause (fever), remove sympathomimetic drugs

Classification of drugs for Antiarrhythmia

TABLE 142.3 Classification of Drugs for Antiarrhythmia

CLASS	ACTION	EXAMPLE(S)
I	Depression of phase of depolarization (velocity of upstroke of action potential); sodium channel blockade	
Ia	Prolongation of QRS complex and QT interval	Quinidine, procainamide, disopyramide
Ib	Significant effect on abnormal conduction	Lidocaine, mexiletine, phenytoin, tocainide
Ic	Prolongation of QRS complex and PR interval	Flecainide, propafenone, moricizine?
II	β blockade; slowing of sinus rate; prolongation of PR interval	Propranolol, atenolol, acebutolol
III	Prolongation of action potential; prolongation of PR, QT intervals, QRS complex; sodium and calcium channel blockade	Bretylium, amiodarone, sotalol
IV	Calcium channel blockade; reduction in sinus and AV node pacemaker activity and conduction; prolongation of PR interval	Verapamil and other calcium channel blocking agents

AV, Atrioventricular.