

**Etiology of Syncope in the Pediatric Patient**

**CARDIOVASCULAR: CONGENITAL OR ACQUIRED**

**Cardiac**

- Dysrhythmias
- Myocardial disorders
- Valvular disease
- Pericardial disease

- Congenital heart disease
- Pulmonary hypertension
- Cardiac tumors (primary or metastatic): Atrial myxoma, rhabdomyosarcoma

**Vascular**

- Aortic disease: Severe coarctation, aortic dissection
- Coronary artery disease / disorders

- Disease / disorders of great vessels (other than aorta)
  - Subclavian steal
  - Thoracic outlet

**NEUROLOGIC**

- Central nervous system disorder
  - Hemorrhage
  - Stroke, transient ischemic attack
  - Migraine headaches

- Peripheral nervous system
  - Neuropathies

**RESPIRATORY**

- Hypoxia from respiratory failure or pulmonary disease
- Pulmonary emboli (can cause syncope by mechanism other than hypoxia)
- Cough (post-tussive)
- Breath-holding: Cyanotic, pallid, mixed (likely from autonomic dysregulation)

**Disorder of Oxygen Transport**

- Carbon monoxide poisoning
- Cyanide poisoning
- Other toxins
- Severe anemia

**METABOLIC / ENDOCRINE**

- Hypoglycemia

**DRUGS / DRUG OVERDOSE**

- Poisons/toxins (*see also* Disorders of Oxygen Transport, *above*)
- Drugs of abuse: Cocaine, opiates, alcohol, others
- Prescription drug abuse (intentional): Benzodiazepine, opioids, others
- Prescription drugs (unintentional): Antihypertensives, cardiac especially vasodilators (calcium channel blockers, beta blockers, nitrates), psychiatric drugs especially tricyclics, phenothiazines
- Nonprescription
- Over-the-counter drugs/herbals/vitamins

**HYPOVOLEMIC / HEMORRHAGIC**

- Decreased intravascular volume (from blood or volume loss)

**POSTURAL (ORTHOSTATIC)**

- Postural orthostatic tachycardia syndrome (POTS)
- Dysautonomic syndromes: Primary or secondary autonomic failure

**NEUROCARDIOGENIC**

- Vasovagal
- Situational
- Carotid sinus syncope

**PSYCHIATRIC**

- Anorexia nervosa, bulimia
- Hyperventilation (anxiety, panic disorders)

**History and Physical Examination Findings Suggestive of Serious Etiology of Syncope**

**HISTORY**

	<b>BENIGN</b>	<b>SERIOUS</b>
Position	Occurs with change in position (from lying to sitting or standing, or sitting to standing) (implies orthostatic syncope)	Unrelated to position (occurs while recumbent or sitting)
Exercise	Not related to exercise	Occurs during exercise or exertion (occurs when CO fails to meet increased demands), ↑CO needed with exercise
Onset	Gradual	Rapid (suggests dysrhythmias)
Prodrome	With prodrome	None (suggests dysrhythmia with sudden onset)
Isolated vs. recurrent	Isolated single event	Recurrent over a short time frame (usually hours, days, or few weeks)
Injury: Secondary to syncope	No injury from syncope (no injuries occur with psychogenic syncope)	Injury (bruises, lacerations, fractures, etc.) from syncope
Associated symptoms: CV	No associated CV symptoms	Chest pain, shortness of breath, palpitations
Associated signs: Skin	No cyanosis, no pallor; or, short-lived, mild (transient) pallor	Severe pallor or cyanosis

**FAMILY HISTORY**

	<b>BENIGN</b>	<b>SERIOUS</b>
Sudden death	No	Yes
Myocardial Infarction at an early age	No	Yes
Cardiomyopathy	No	Yes
Neuromuscular disorders	No	Yes
Congenital deafness	No	Yes (Consider Jervett-Lange-Nielsen Syndrome)
Marfan syndrome	No	Yes (Consider aortic dissection)
Autoimmune disease (maternal lupus)	No	Yes (Complete AV block in infant from maternal lupus)

**PAST MEDICAL HISTORY**

	<b>BENIGN</b>	<b>SERIOUS</b>
Prior cardiac surgery	No	Yes (damage to conduction system may occur months, even years, later)
History of congestive heart failure	No	Yes (may have poor cardiac output and / or dysrhythmias)
Congenital heart disease	No	Yes
Acquired heart disease	No	Yes

**VITAL SIGNS**

	<b>BENIGN</b>	<b>SERIOUS</b>
Pulse/heart rate	No	Yes
Tachycardia or bradycardia		
Regular	Yes	No
Respirations: Bradypnea	No	Yes (Consider serious CNS or respiratory disease)
Respirations: Tachypnea	Usually no, occasionally yes from hyperventilation secondary to pain / anxiety	Yes (Consider respiratory disease)
Blood pressure (low)	No	Yes (Consider hypovolemia or hemorrhage)
Blood pressure (high)	No	Yes (Consider hypertensive crisis / encephalopathy)
Positive orthostatic vital signs	No	Yes (Consider hypovolemia, hemorrhage, autonomic nervous system disorders)

**PHYSICAL EXAMINATION**

	<b>BENIGN</b>	<b>SERIOUS</b>
General appearance	Normal	Abnormal; Unusual facies (may have syndrome with CV disease, such as Down's syndrome, Williams disease, Marfan syndrome)
Respiratory	Normal	Abnormal; Rales, wheezing suggest underlying CV or pulmonary disease
Mental status	Normal	Abnormal; may have inadequate CNS perfusion or CNS disease
Neurologic	Normal	Abnormal; especially focal abnormalities, suggest CNS disease / injury
Neck exam	Normal	JVD suggests CHF
Musculoskeletal	Normal	Abnormal; weakness, decreased strength, tone, or muscle mass imply neuromuscular disorder
Extremity	Normal	Abnormal; calf pain (consider deep vein thrombosis), edema (Is there CHF or other systemic disease?)
Dermatologic	Normal	Abnormal; decreased turgor / tenting or other signs of inherited disease; café-au-lait, von Recklinghausen's disease

**Key:** CO = cardiac output; CV = cardiovascular; CNS = central nervous system; JVD = jugular venous distention

# Differential Diagnosis of Syncope in Pediatric Patients

NEUROLOGIC	CLINICAL FEATURES
Breath-holding Spells* — Three Categories <b>Incidence</b> <i>Cyanotic</i> 52%–62% Hold breath in expiration → apnea / cyanosis <i>Pallid</i> 19%–28% Apnea → pallor <i>Mixed or unclassified</i> 19%–20%	Age 6–24 months, resolved by age 4–5 years, + family history 20%–35%, Key: inciting event: crying / emotional upset (cyanotic) or pain, fall, hit head (pallid) → loss of tone/apnea, ± seizure, ± posturing Normal EEG, short time frame; common cause of infant syncope, if severe can cause LOC, seizure, posturing Etiology: Autonomic dysregulation, treatment of iron deficiency anemia
Apnea*	Common, especially in premature infants, due to brain stem immaturity, resolves with age
Hyperekplexia* (“Stiff baby syndrome,” “Startle disease”)	Stiffness when awake, nocturnal myoclonus, exaggerated startle reflex, toddler sudden falls in response to surprise / stimuli / stress / emotion; rare genetic disease
Seizures	Postictal period, aura, bladder / bowel incontinence, automatisms
Migraine (Basilar)	Visual symptoms, aura, headache
Vertigo	No LOC, dizziness, or spinning sensation
Transient ischemia attack	Neurologic symptoms (weakness, aphasia, etc.) that resolve
Sleep disorders: Cataplexy, narcolepsy	
<b>VASCULAR</b>	
Aortic dissection	BP and / or pulse differences between arms
Subclavian steal	BP and / or pulse differences between arms, symptoms with arm exercise or arm movements
<b>RESPIRATORY</b>	
Hyperventilation	History of tachypnea ± paresthesias, ± carpopedal spasm
Pulmonary emboli	May cause syncope; Symptoms: dyspnea, chest pain; Diagnosis: spiral CT chest or VQ scan
Pulmonary hypertension	May cause syncope / sudden death, loud S2, ECG ± RVH; Symptoms: SOB, DOE, exercise tolerance; Diagnosis: echocardiogram
<b>METABOLIC</b>	
Hypoglycemia	May cause syncope; Associated symptoms: diaphoresis, ± history of DM / glucose disorders, ↓ oral intake, alcohol ingestion (especially in infants/young children); Diagnosis: check glucose
<b>PSYCHIATRIC</b>	
Hysteria (Conversion disorder) Factitious disorders: Malingering, Munchausen’s syndrome	No associated neurologic / cardiovascular changes, no injury occurs, patient may describe event thus, no LOC, may have secondary gain / audience
Panic disorder / anxiety	Hyperventilation

\* = Unique to pediatric population

**Key:** ECG = electrocardiogram; EEG = electroencephalogram; LOC = loss of consciousness; BP = blood pressure; CT = CAT scan; VQ = ventilation perfusion scan; RVH = right ventricular hypertrophy; SOB = shortness of breath; DOE = dyspnea on exertion; DM = diabetes mellitus

## Cardiovascular Causes of Pediatric Syncope

<b>ARRHYTHMIAS: CONGENITAL OR ACQUIRED</b>	
<ul style="list-style-type: none"> <li><b>Tachyarrhythmias</b> <ul style="list-style-type: none"> <li>o Wolff-Parkinson-White syndrome</li> <li>o Ventricular tachycardia</li> <li>o Ventricular fibrillation</li> <li>o Arrhythmogenic right ventricular dysplasia</li> <li>o Torsades de pointes</li> </ul> </li> <li><b>QT Abnormalities</b> <ul style="list-style-type: none"> <li>o Long QT syndromes: Congenital Romano-Ward syndrome, Jervell and Lange-Nielsen syndrome</li> <li>o Acquired: Medications causing QT prolongation: Psychotropics (tricyclic antidepressants, phenothiazines, promotility drugs [cisapride] — especially in combination with other drugs [e.g., erythromycin, ketoconazole])</li> <li>o Short QT syndrome</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li><b>Device Malfunction</b> <ul style="list-style-type: none"> <li>o Pacemaker malfunction</li> <li>o AICD malfunction</li> </ul> </li> <li><b>Specific Congenital Heart Defects</b> <ul style="list-style-type: none"> <li>o High-risk patients: Ebstein anomaly, tetralogy of Fallot, others</li> </ul> </li> <li><b>Post-operative Congenital Heart Disease</b></li> </ul>
<b>STRUCTURAL DISORDERS: MYOCARDIAL DYSFUNCTION / DISEASE</b>	
<ul style="list-style-type: none"> <li><b>Primary</b> <ul style="list-style-type: none"> <li>o Myocarditis</li> <li>o Dilated cardiomyopathy</li> <li>o Idiopathic hypertrophic subaortic stenosis (IHSS)</li> <li>o Arrhythmogenic right ventricular dysplasia (<i>see also</i> Arrhythmias)</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li><b>Secondary (Acquired)</b> <ul style="list-style-type: none"> <li>o Infections:                             <ul style="list-style-type: none"> <li>– <i>Viral:</i> Coxsackie, others</li> <li>– <i>Parasitic:</i> Chagas disease</li> </ul> </li> <li>o Immunologic, vasculitis, rheumatologic diseases: Lyme disease, amyloidosis, sarcoidosis</li> <li>o Generalized neuromuscular diseases: Muscular dystrophy</li> </ul> </li> </ul>
<b>CORONARY ARTERY DISEASE</b>	
<ul style="list-style-type: none"> <li><b>Congenital:</b> Anomalous coronary artery</li> </ul>	<ul style="list-style-type: none"> <li><b>Acquired:</b> Kawasaki disease</li> </ul>
<b>VALVULAR</b>	
<ul style="list-style-type: none"> <li><b>Severe / Critical Aortic Stenosis</b></li> <li><b>Severe / Critical / Mitral / Pulmonic / Tricuspid Valve Disorder</b> (stenosis or regurgitation)</li> <li><b>Prosthetic Valve Dysfunction</b></li> </ul>	
<b>VASCULAR</b>	
<ul style="list-style-type: none"> <li><b>Aorta:</b> <i>Aortic dissection</i>, secondary to hypertension, atherosclerosis, connective tissue disorders (Marfan disease, Ehlers-Danlos syndrome)</li> </ul>	<ul style="list-style-type: none"> <li><b>Other Great Vessel Abnormalities</b> <ul style="list-style-type: none"> <li>o Subclavian steal syndrome</li> <li>o Thoracic outlet syndrome</li> </ul> </li> </ul>
<b>OUTFLOW OBSTRUCTION TO SYSTEMIC BLOOD FLOW</b>	
<ul style="list-style-type: none"> <li><b>Severe Coarctation</b></li> <li><b>Cardiac Tumor Mass</b> (atrial myxoma)</li> </ul>	<ul style="list-style-type: none"> <li><b>Also: Hypertrophic Obstructive Cardiomyopathy, Critical Aortic Stenosis</b></li> </ul>
<b>PERICARDIAL DISEASE</b>	
<ul style="list-style-type: none"> <li><b>Pericarditis / Pericardial Tamponade</b></li> </ul>	
<b>CONGENITAL HEART DEFECTS</b>	
<ul style="list-style-type: none"> <li><b>Cyanotic / Acyanotic Congenital Heart Disease</b></li> </ul>	<ul style="list-style-type: none"> <li><b>Eisenmenger Syndrome</b></li> </ul>
<b>PULMONARY HYPERTENSION</b>	
<ul style="list-style-type: none"> <li><b>Primary</b> (idiopathic)</li> </ul>	<ul style="list-style-type: none"> <li><b>Secondary</b></li> </ul>

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