



NCC Pediatrics Continuity Clinic Curriculum: Heart Murmurs

Goals & Objectives: *To perform a proper assessment of a child with a murmur and list appropriate steps in the evaluation:*

- Understand the components of a complete H&P in a child with a murmur.
- Describe a murmur in terms of location, quality, timing, intensity, and radiation.
- Differentiate pathologic from non-pathologic murmurs, based on the H&P.
- List appropriate initial steps in the evaluation of a child with a pathologic murmur.
- Describe circumstances in which pediatric cardiology consultation is indicated.

Pre-Meeting Preparation:

Please read the following enclosures:

- “Evaluation and Management of Heart Murmurs in Children” (AAFP, 2011)

Conference Agenda:

- Review Murmurs Quiz
- Complete Murmurs Cases
- **Optional:** Play “Innocent or Not That Innocent” Audio-Clip Game

Post-Conference: Board Review Q&A

Extra-Credit:

- [“The Cardiac Murmur: When to Refer”](#) (MD Consult: Pediatrics Clinic of N Am; 1998)
- [“Does the Presence of a ‘New’ Murmur Predict Cardiac Pathology in Otherwise Asymptomatic Children and Adolescent”](#) (Gupta & May, unpublished abstract, 2015)
- Modules at www.easyauscultation.com/heart-sounds

Evaluation and Management of Heart Murmurs in Children

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Heart murmurs are common in healthy infants, children, and adolescents. Although most are not pathologic, a murmur may be the sole manifestation of serious heart disease. Historical elements that suggest pathology include family history of sudden cardiac death or congenital heart disease, in utero exposure to certain medications or alcohol, maternal diabetes mellitus, history of rheumatic fever or Kawasaki disease, and certain genetic disorders. Physical examination should focus on vital signs; age-appropriate exercise capacity; respiratory or gastrointestinal manifestations of congestive heart failure; and a thorough cardiovascular examination, including features of the murmur, assessment of peripheral perfusion, and auscultation over the heart valves. Red flags that increase the likelihood of a pathologic murmur include a holosystolic or diastolic murmur, grade 3 or higher murmur, harsh quality, an abnormal S_2 , maximal murmur intensity at the upper left sternal border, a systolic click, or increased intensity when the patient stands. Electrocardiography and chest radiography rarely assist in the diagnosis. Referral to a pediatric cardiologist is recommended for patients with any other abnormal physical examination findings, a history of conditions that increase the likelihood of structural heart disease, symptoms suggesting underlying cardiac disease, or when a specific innocent murmur cannot be identified by the family physician. Echocardiography provides a definitive diagnosis and is recommended for evaluation of any potentially pathologic murmur, and for evaluation of neonatal heart murmurs because these are more likely to be manifestations of structural heart disease. (*Am Fam Physician*. 2011;84(7):793-800. Copyright © 2011 American Academy of Family Physicians.)

Hear murmurs are common in asymptomatic, otherwise healthy children. These murmurs are often innocent and result from the normal patterns of blood flow through the heart and vessels.¹ However, a heart murmur may be the sole finding in children with structural heart disease; therefore, a thorough evaluation is necessary.

Incidence and Prevalence

Congenital heart disease (CHD) may occur in the presence or absence of a heart murmur. The incidence of CHD varies between four and 50 per 1,000 live births.² One review found an incidence of 75 cases per 1,000 live births; of these, six cases per 1,000 were moderate or severe.³

History

Certain historical features suggest possible structural heart disease (*Table 1*).^{1,2,4-11} Cardiovascular signs and symptoms can be non-specific (e.g., poor feeding, failure to thrive) or specific (e.g., chest pain, palpitations),

and can help identify children who are likely to have structural heart disease (*Table 2*).^{4,7,10}

In infants, feeding difficulties may be the first sign of congestive heart failure, which is present in approximately one-third of infants and children with CHD.⁴ The most common symptoms in a series of children presenting to the emergency department with acute heart failure included dyspnea (74 percent), nausea and vomiting (60 percent), fatigue (56 percent), and cough (40 percent).¹²

Exercise tolerance should be assessed in an age-appropriate fashion. Parents of infants should be asked about their child's ability to play and the duration and vigor of feeding; parents of older children should compare their child's ability to participate in team sports with that of peers.⁴ Chest pain is rarely a presenting symptom of cardiac disease in children.^{13,14} In a pediatric cardiology clinic, chest pain or syncope prompted consultation in approximately 10 percent of children; only 11 percent of those with chest pain and 5 percent of those with syncope had cardiac disease.¹⁴ A high degree of suspicion is

Table 1. Historical Findings Suggesting Structural Heart Disease in Children with Heart Murmurs

<i>Historical finding</i>	<i>Significance</i>
Family history	
CHD	More common in children with a first-degree relative who has CHD (three- to 10-fold increased risk ⁷); high penetrance with ventricular septal defect and mitral valve prolapse
Sudden cardiac death or hypertrophic cardiomyopathy	Increased risk of hypertrophic cardiomyopathy (autosomal dominant pattern)
Sudden infant death syndrome	Can be secondary to undiagnosed CHD lesions ⁸
Personal history	
Conditions that may coexist with CHD:	Certain genetic disorders (e.g., DiGeorge syndrome, velo-cardio-facial syndrome) are associated with cardiac malformations
Aneuploidy (e.g., trisomy 21, Turner syndrome)	Trisomy 21 is associated with an increased risk of atrioventricular septal defects, atrial septal defects, ventricular septal defects, patent ductus arteriosus, and tetralogy of Fallot
Connective tissue disorder (e.g., Marfan syndrome)	Turner syndrome is associated with increased risk of coarctation of the aorta, aortic valve stenosis, and left ventricular hypertrophy
Inborn error of metabolism	Marfan syndrome is associated with mitral valve prolapse, aortic root dilation, and aortic insufficiency
Major congenital defects of other organ systems	
Syndrome with dysmorphic features	
Frequent respiratory infections	Respiratory symptoms may be attributable to heart disease (i.e., congestive heart failure); enlarged vessels may lead to atelectasis or difficulty clearing respiratory secretions, thereby promoting infection
Kawasaki disease	Leading cause of acquired cardiac disease in children; can cause coronary artery aneurysm and stenosis ⁹
Rheumatic fever	Associated with development of rheumatic heart disease
Prenatal or perinatal history	
In utero exposure to alcohol or other toxins	Fetal alcohol syndrome is associated with an increased risk of atrial and ventricular septal defects, and tetralogy of Fallot ¹⁰
In utero exposure to selective serotonin reuptake inhibitors or other potentially teratogenic medications	Selective serotonin reuptake inhibitor exposure is associated with a small but statistically significant increased risk of mild heart lesions, including ventricular septal defects and bicuspid aortic valve (although not all studies found an increased risk ¹¹) Lithium exposure is associated with Ebstein anomaly of the tricuspid valve Valproate (Depacon) exposure is associated with coarctation of the aorta and hypoplastic left heart syndrome
Intrauterine infection	Maternal infections may increase risk of structural heart lesions (e.g., maternal rubella infection is associated with patent ductus arteriosus and peripheral pulmonary stenosis)
Maternal diabetes mellitus	Increased risk of CHD, including transient hypertrophic cardiomyopathy, tetralogy of Fallot, truncus arteriosus, and double-outlet right ventricle
Preterm delivery	CHD is associated with other conditions (e.g., genetic disorders, in utero exposure to toxins) that can result in preterm birth; 50 percent of newborns weighing less than 3 lb, 5 oz (1,500 g) at birth have CHD (most commonly patent ductus arteriosus) ⁷

CHD = congenital heart disease.

Information from references 1, 2, and 4 through 11.

necessary to detect underlying cardiac disease in children who report exertional syncope or chest pain, or who have a family history of hypertrophic cardiomyopathy.^{1,13,14}

Physical Examination

The patient's vital signs should be compared with age-established norms (available at <http://www.cc.nih.gov/ccc/pedweb/pedstaff/age.html>), and a focused examination of the respiratory, cardiovascular, and gastrointestinal systems should be performed⁵ (Table 3^{2,5-7,10,15,16}). Congenital anomalies of other organ systems may be associated with CHD in up to 25 percent of patients.⁶ The

child's appearance, activity level, color, and respiratory effort should be assessed, and the neck should be examined for prominent vessels, abnormal pulsations, and bruits.¹ Jugular venous distension is rare in children.⁴ The chest wall should be inspected for abnormalities of the sternum, which can be associated with CHD,¹⁵ and for abnormal cardiac impulses or thrills.¹ The lungs should be auscultated for abnormal breath sounds such as crackles, which may indicate pulmonary congestion, or wheezing, which may indicate cardiac asthma. Abdominal examination should focus on the liver location (seeking abdominal situs) and evaluation for liver

Table 2. Symptoms Suggesting Cardiac Disease

<i>Symptom or sign</i>	<i>Significance</i>
Cardiovascular	
Chest pain	May be related to aortic stenosis or hypertrophic cardiomyopathy
Cyanosis	Structural heart lesion with restricted pulmonary blood flow
Dizziness	Multiple potential causes, including hypoxia and CHF
Near-syncope or syncope	May be related to aortic stenosis or hypertrophic cardiomyopathy
Palpitations	May be related to arrhythmias secondary to structural heart lesions
Constitutional	
Developmental delay	Congenital heart lesions are more common in children with certain genetic disorders and syndromes
Diaphoresis	May indicate CHF or poor cardiac fitness
Easily fatigued	May indicate CHF, hypoxia, or poor cardiac fitness
Poor exercise tolerance or capacity for play	May indicate CHF, hypoxia, or poor cardiac fitness
Poor growth or failure to thrive	May indicate CHF, poor cardiac fitness, or a genetic disorder or syndrome; poor weight gain most commonly reflects decreased cardiac output or left-to-right shunts with pulmonary hypertension
Respiratory	
Asthma-like symptoms	Cardiac asthma resulting from pulmonary congestion
Chronic cough	Atelectasis or difficulty clearing secretions because of pulmonary vascular congestion
Dyspnea on exertion	May indicate CHF, hypoxia, or poor cardiac fitness

CHF = congestive heart failure.

Information from references 4, 7, and 10.

enlargement or ascites, which may signal congestive heart failure.⁵

The peripheral pulses should be examined for rate, rhythm, volume, and character, and capillary refill time should be less than three seconds.⁴ The heart should be auscultated over the tricuspid, pulmonary, mitral, and aortic areas with the bell and diaphragm of the stethoscope while the patient is supine, sitting, and standing¹⁷ (Figure 1¹⁸). Innocent murmurs are produced by the normal flow of blood through the heart. Changing the flow by changing the patient's position (for example, decreasing flow to the heart with the Valsalva maneuver) will change the intensity of the murmur. Young children should be prompted to push out their abdomen against the examiner's hand.¹ The physician should listen for normal S₁ and S₂; a wide fixed split S₂ is characteristic of an atrial septal defect.¹⁹ Gallops can be a normal finding in adolescents.¹

Table 3. Physical Examination of Children with Heart Murmurs

<i>Finding</i>	<i>Significance</i>
Abnormal growth (height and weight plotted on growth chart)	Feeding difficulties may be a sign of cardiac disease in newborns and infants (decreased exercise capacity) Certain genetic disorders may increase risk of delayed growth and CHD
Abnormal vital signs (compared with age-adjusted norms)	Arrhythmia, tachycardia, hypoxia, and tachypnea may indicate underlying structural heart disease Blood pressure discrepancy between upper and lower limbs may indicate coarctation of the aorta (pressure gradient of > 20 mm Hg with low blood pressure in the lower extremities)
Adventitious breath sounds (e.g., wheezing, rales, ronchi, pleural rub)	Wheezing may be associated with cardiac asthma; rales may be associated with pulmonary congestion secondary to congestive heart failure
Chest contour signaling maldevelopment of the sternum ¹⁵	Defective segmentation of the sternum may occur in children with CHD
Dysmorphic features	Certain genetic or congenital conditions increase risk of CHD
Cardiovascular findings	
Abnormal S ₂	Classic finding of wide split fixed S ₂ with atrial septal defects; abnormal S ₂ may be present in other types of CHD
Capillary refill	Normal peripheral perfusion is less than 2 to 3 seconds; delay may indicate poor perfusion secondary to diminished cardiac output
Displaced point of maximal impulse; precordial impulses (heaves, lifts, thrills)	Possible structural abnormality or ventricular enlargement
Edema	Congestive heart failure
Left-sided precordial bulge	Cardiac enlargement
S ₃ or S ₄	Can indicate structural heart disease; S ₃ can be a normal finding but usually disappears when patient is upright
Substernal heave	Right ventricular hypertension
Systolic ejection click	Semilunar valvular stenosis
Weak or absent femoral pulses	Coarctation of the aorta
Gastrointestinal findings	
Ascites	Congestive heart failure
Hepatomegaly	Congestive heart failure
Location of liver signals abdominal situs	High rate of CHD with abdominal situs

CHD = congenital heart disease.

Information from references 2, 5 through 7, 10, 15, and 16.

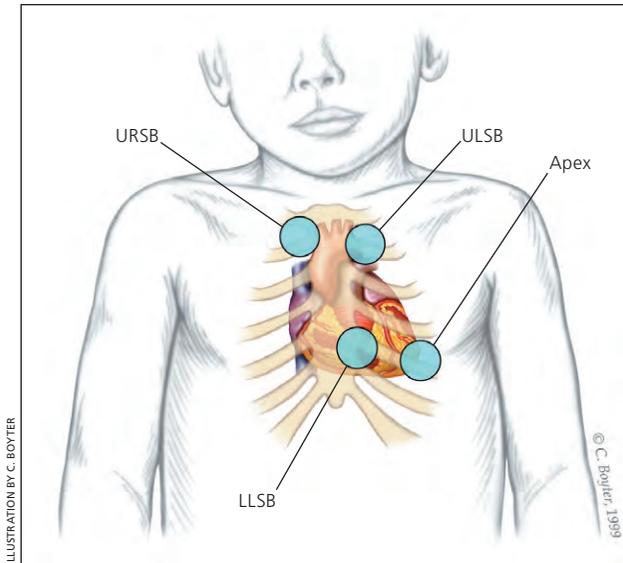


Figure 1. Listening areas for clicks: upper right sternal border (URSB) for aortic valve clicks; upper left sternal border (ULSB) for pulmonary valve clicks; lower left sternal border (LLSB), or the tricuspid area, for ventricular septal defects; and the apex for aortic or mitral valve clicks.

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The heart murmur is characterized by its timing during the cardiac cycle; its location, quality, intensity, and pitch (how it sounds); and the presence or absence of clicks¹ (Table 4^{5,7,17} and Table 5²⁰⁻²³). The intensity of heart murmurs is graded from 1 to 6. Grade 1 murmurs are barely audible; grade 2 murmurs are faint but can be heard immediately; grade 3 murmurs can be heard easily and are moderately loud; grade 4 murmurs can be heard easily over a wide area but do not have a palpable thrill; grade 5 murmurs are loud and have a precordial thrill; and grade 6 murmurs are loud enough to hear with the stethoscope raised off the chest.^{17,24} Certain characteristics of the murmur may be considered red flags, prompting stronger consideration for structural heart disease. These include a holosystolic murmur (odds ratio [OR] of pathologic murmur = 54), grade 3 or higher (OR = 4.8), harsh quality (OR = 2.4), an abnormal S₂ (OR = 4.1), maximal intensity at the upper left sternal border (OR = 4.2), a systolic click (OR = 8.3), diastolic murmur, or increased murmur intensity with standing.^{6,10,25} A decrease or lack

Table 4. Characteristics of Innocent Heart Murmurs

Type	Description	Age at detection	Can sound like
Aortic systolic murmur	Systolic ejection murmur best heard over the aortic valve	Older childhood into adulthood	—
Mammary artery soufflé*	High-pitched systolic murmur that can extend into diastole; best heard along the anterior chest wall over the breast	Rare in adolescence	Arteriovenous anastomoses or patent ductus arteriosus
Peripheral pulmonary stenosis	Grade 1 or 2, low-pitched, early- to mid-systolic ejection murmur heard over axilla or back	< 1 year	Pulmonary artery stenosis or normal breath sounds
Pulmonary flow murmur	Grade 2 or 3, crescendo-decrescendo, early- to mid-systolic murmur peaking in mid-systole; best heard at the left sternal border between the second and third intercostal spaces; characterized by a rough, dissonant quality; loudest when patient is supine and decreases when patient is upright and holding breath	All	Atrial septal defect or pulmonary valve stenosis
Still murmur	Grade 1 to 3, early systolic murmur; low to medium pitch with a vibratory or musical quality; best heard at lower left sternal border; loudest when patient is supine and decreases when patient stands	Infancy to adolescence, often 2 to 6 years	Ventricular septal defect or hypertrophic cardiomyopathy
Supraclavicular/brachiocephalic systolic murmur	Brief, low-pitched, crescendo-decrescendo murmur heard in the first two-thirds of systole; best heard above clavicles; radiates to neck; diminishes when patient hyperextends shoulders	Childhood to young adulthood	Bicuspid/stenotic aortic valve, pulmonary valve stenosis, or coarctation of the aorta
Venous hum	Grade 1 to 6 continuous murmur; accentuated in diastole; has a whining, roaring, or whirring quality; best heard over low anterior neck, lateral to the sternocleidomastoid; louder on right; resolves or changes when patient is supine	3 to 8 years	Cervical arteriovenous fistulas or patent ductus arteriosus

*—Mammary artery soufflé murmur is caused by blood flow in the arteries and veins leading to and from the breasts.

Information from references 5, 7, and 17.

Table 5. Prevalence and Characteristics of Pathologic Heart Murmurs

Type of structural heart lesion	Prevalence among children with congenital heart disease (%)	Symptoms and clinical course	Characteristics
Ventricular septal defect	20 to 25	Small defects: usually asymptomatic Medium or large defects: CHF, symptoms of bronchial obstruction, frequent respiratory infections	Small defects: loud holosystolic murmur at LLSB (may not last throughout systole if defect is very small) Medium and large defects: increased right-to-left ventricular impulses; thrill at LLSB; split or loud single S ₂ ; holosystolic murmur at LLSB without radiation; grade 2 to 5; may also hear a grade 1 or 2 mid-diastolic rumble
Atrial septal defect	8 to 13	Usually asymptomatic and incidentally found on physical examination or echocardiography; large defects can be present in infants with CHF	Grade 2 or 3 systolic ejection murmur best heard at ULSB; wide split fixed S ₂ ; absent thrill; may have a grade 1 or 2 diastolic flow rumble at LLSB
Patent ductus arteriosus	6 to 11	May be asymptomatic; can cause easy fatigue, CHF, and respiratory symptoms	Continuous murmur (grade 1 to 5) in ULSB (crescendo in systole and decrescendo into diastole); normal S ₁ ; S ₂ may be "buried" in the murmur; thrill or hyperdynamic left ventricular impulse may be present
Tetralogy of Fallot	10	Onset depends on severity of pulmonary stenosis; cyanosis may appear in infancy (2 to 6 months of age) or in childhood; other symptoms include hypercyanotic spells or decreased exercise tolerance	Central cyanosis; clubbing of nail beds; grade 3 or 4 long systolic ejection murmur heard at ULSB; may have holosystolic murmur at LLSB; systolic thrill at ULSB; normal to slightly increased S ₁ ; single S ₂
Pulmonary stenosis	7.5 to 9	Usually asymptomatic but may have symptoms secondary to pulmonary congestion	Systolic ejection murmur (grade 2 to 5); heard best at ULSB radiating to infraclavicular regions, axillae, and back; normal or loud S ₁ ; variable S ₂ ; systolic ejection click may be heard at left sternal border and may vary with respiration
Coarctation of the aorta	5.1 to 8.1	Newborns and infants may present with CHF; older children are usually asymptomatic or may have leg pain or weakness	Systolic ejection murmur best heard over interscapular region; normal S ₁ and S ₂ ; decreased or delayed femoral pulse; may have increased left ventricular impulse
Aortic stenosis	5 to 6	Usually asymptomatic; symptoms may include dyspnea, easy fatigue, chest pain, or syncope; newborns and infants may present with CHF	Systolic ejection murmur (grade 2 to 5) best heard at upper right sternal border with radiation to carotid arteries; left ventricular heave; thrill at ULSB or suprasternal notch
Transposition of the great arteries	5	Variable presentation depending on type; may include cyanosis or CHF in first week of life	Cyanosis; clubbing of nail beds; single S ₂ ; murmur may be absent or grade 1 or 2 nonspecific systolic ejection murmur; may have a grade 3 or 4 holosystolic murmur at LLSB and mid-diastolic murmur at apex
Total anomalous pulmonary venous connection	2 to 3	Onset of CHF at 4 to 6 weeks of age	Grade 2 or 3 systolic ejection murmur at ULSB; grade 1 or 2 mid-diastolic flow rumble at LLSB; wide split fixed S ₂
Tricuspid atresia	1.4	Early-onset cyanosis or CHF within the first month of life	Cyanosis; clubbing of nail beds; normal pulses; single S ₂ ; holosystolic murmur at LLSB or midsternal border; murmur may be absent; mid-diastolic flow murmur at apex may be present
Hypoplastic left heart syndrome	Rare	May be asymptomatic at birth, with cyanosis and CHF developing with duct closure	Hyperdynamic precordium; single S ₂ ; nonspecific grade 1 or 2 systolic ejection murmur along left sternal border
Truncus arteriosus	Rare	Onset of CHF in first few weeks of life; minimal cyanosis	Increased cardiac impulses; holosystolic murmur (ventricular septal defect); mid-diastolic rumble

CHF = congestive heart failure; LLSB = lower left sternal border; ULSB = upper left sternal border.

Information from references 20 through 23.

Table 6. The Seven S's: Key Features of Innocent Murmurs

Sensitive (changes with child's position or with respiration)
Short duration (not holosystolic)
Single (no associated clicks or gallops)
Small (murmur limited to a small area and nonradiating)
Soft (low amplitude)
Sweet (not harsh sounding)
Systolic (occurs during and is limited to systole)

Information from reference 27.

of change in the murmur intensity with passive leg elevation (likelihood ratio [LR] = 8.0) or when the child moves from standing to squatting (LR = 4.5) increases the likelihood of hypertrophic cardiomyopathy.²⁶

Characteristics that are more likely to be associated with an innocent murmur include a systolic (rather than diastolic) murmur; soft sound; short duration; musical or low pitch; varying intensity with phases of respiration and posture (louder in supine position); and murmurs that become louder with exercise, anxiety, or fear^{17,24} (Table 6²⁷). The most common innocent murmur is a Still murmur, which is characteristically loudest at the lower left sternal border and has a musical or vibratory quality that is thought to represent vibrations of the left outflow tract.^{1,5}

Auscultation may be less accurate in younger patients, when other signs or symptoms of cardiovascular disease

are present, and when findings on radiography or electrocardiography (ECG) are abnormal.²⁸ Online libraries of digital heart sounds are available to familiarize physicians with the characteristics of abnormal heart sounds (Table 7).

Role of Diagnostic Testing

Chest radiography and ECG rarely assist in the diagnosis of a heart murmur.^{5,6,29} Studies in newborns³⁰ and children³¹ with asymptomatic murmurs have shown that chest radiography does not influence clinical management or assist with diagnosis. A prospective study of 201 newborns who were referred to pediatric cardiologists for evaluation of a heart murmur found that the addition of ECG to clinical assessment did not improve the sensitivity or specificity of detecting structural heart lesions.³² In a study of 128 infants and children who were evaluated for heart murmurs, the addition of ECG and chest radiography to cardiac auscultation was more likely to mislead than assist the physician in making the correct diagnosis.³³

In a study of more than 900 children in a pediatric cardiology clinic who had innocent-sounding murmurs, an abnormal finding from the history, physical examination, or diagnostic tests (ECG, chest radiography, or pulse oximetry) was 67 percent sensitive but only 38 percent specific for the presence of a structural heart lesion in infants younger than six weeks, yielding positive and negative LRs very near 1.0 (i.e., no useful diagnostic information).²⁸ In infants older than six weeks, sensitivity increased to 100 percent, but specificity decreased to 28 percent (positive LR = 1.6; negative LR = 0.026). Thus, this information is helpful for ruling out structural causes of an innocent-sounding murmur in infants and children older than six weeks, but it is not helpful in younger infants.

In two separate populations geographically remote from a pediatric cardiologist, phonocardiography (i.e., digital heart sound recordings reviewed by a pediatric cardiologist) had high sensitivity and specificity, and good intraobserver agreement in distinguishing between innocent murmurs and murmurs that were potentially or probably pathologic and that required echocardiography.^{34,35}

Indications for Referral

In children and adolescents, the diagnosis of an innocent heart murmur can be made if four criteria are met: absence of abnormal

Table 7. Online Resources for Pediatric Cardiac Auscultation

The Auscultation Assistant Web site: http://www.wilkes.med.ucla.edu/inex.htm
Blaufuss Medical Multimedia Laboratories Web site: http://www.blaufuss.org
Heart Sounds and Murmurs Web site: http://www.dundee.ac.uk/medther/Cardiology/hsmur.html
Johns Hopkins University Cardiac Auscultatory Recording Database Web site: http://www.murmurlab.com/card6/ (registration required)
Texas Heart Institute Web site: http://www.texasheart.org/education/cme/explore/events/eventdetail_5469.cfm
University of Michigan Heart Sound and Murmur Library Web site: http://www.med.umich.edu/lrc/psb/heartsounds/index.htm
University of Washington Department of Medicine Demonstrations: Heart Sounds and Murmurs Web site: http://depts.washington.edu/physdx/heart/demo.html

physical examination findings (except for the murmur); a negative review of systems (i.e., child is asymptomatic); a history that is negative for features that increase the risk of structural heart disease; and characteristic auscultatory features of a specific innocent heart murmur.^{2,5} These criteria are not appropriate for newborns or infants younger than one year because these patients have a higher rate of asymptomatic structural heart disease.³⁶ When an innocent murmur cannot be definitively diagnosed, the child should be referred for echocardiography, to a pediatric cardiologist, or both.

A study in Oman found that the prevalence of abnormal findings on echocardiography was not significantly different between patients referred by pediatric cardiologists and those referred by primary care physicians.³⁷ However, pediatric cardiologists more accurately detect structural heart lesions in newborns and children with heart murmurs,^{32,38} and can assist family physicians in the assessment of a suspicious murmur. For both innocent and pathologic murmurs, referral to a pediatric cardiologist for confirmation or clarification of the diagnosis is associated with decreased parental anxiety.³⁹

Neonatal Heart Murmurs

Newborns are at higher risk of having serious structural heart disease that presents as an asymptomatic murmur.^{6,10} Approximately 1 percent of newborns have a heart murmur, and 31 to 86 percent of these infants have structural heart disease,⁴⁰⁻⁴² including asymptomatic newborns. Because of the higher likelihood of structural heart disease in asymptomatic newborns and young infants with heart murmurs, referral to a pediatric cardiologist and/or for echocardiography is recommended.^{28,42,43} Even potentially life-threatening heart defects may not be associated with any initial signs or symptoms other than a heart murmur.^{41,42}

The reported sensitivity for detection of a pathologic heart murmur in newborns ranges from 80.5 to 94.9 percent among pediatric cardiologists, with specificity ranging from 25 to 92 percent.^{32,43} These variations are significant because the lowest specificity corresponds to positive and negative LRs of 1.1 and 0.7, which are uninformative, and the highest specificity corresponds to positive and negative LRs of 10 and 0.21, which are quite accurate. The ability of a pediatric cardiologist to accurately identify pathologic murmurs

SORT: KEY RECOMMENDATIONS FOR PRACTICE

<i>Clinical recommendation</i>	<i>Evidence rating</i>	<i>References</i>
Structural heart disease is more likely when the murmur is holosystolic, diastolic, grade 3 or higher, or associated with a systolic click; when it increases in intensity with standing; or when it has a harsh quality.	C	6, 10, 25
Chest radiography and electrocardiography rarely assist in the diagnosis of heart murmurs in children.	B	5, 6, 29-33
Family physicians should order echocardiography or consider referral to a pediatric cardiologist for newborns with a heart murmur, even if the child is asymptomatic, because of the higher prevalence of structural heart lesions in this population.	B	28, 43

A = consistent, good-quality patient-oriented evidence; B = inconsistent or limited-quality patient-oriented evidence; C = consensus, disease-oriented evidence, usual practice, expert opinion, or case series. For information about the SORT evidence rating system, go to <http://www.aafp.org/afpsort.xml>.

depends on multiple factors, including his or her confidence in the diagnosis. Echocardiography may not be required in newborns with a heart murmur if a pediatric cardiologist has diagnosed an innocent murmur with a high degree of confidence³²; however, it is important to consider the relatively high prevalence of structural heart disease among asymptomatic newborns with a heart murmur.

The evaluation of newborns for CHD may include pulse oximetry after 24 hours of life. Clinical examination of asymptomatic newborns has a sensitivity of 46 percent for detection of CHD; this sensitivity increases to 77 percent when clinical examination is combined with pulse oximetry (with a cutoff of 94 percent).⁴⁴

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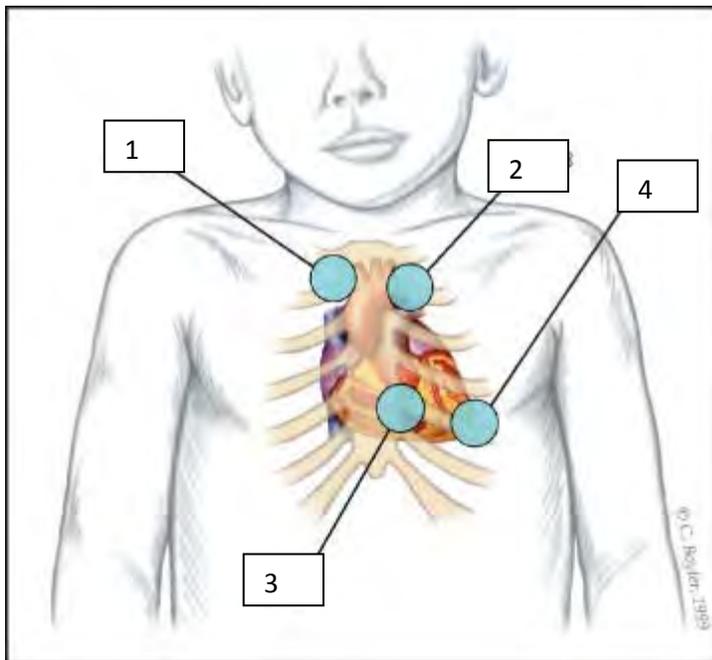
Author disclosure: No relevant financial affiliations to disclose.

Heart Murmurs Quiz

1. What are 6 characteristics of murmurs?

- (a)
- (b)
- (c)
- (d)
- (e)
- (f)

2. Please match the numbered locations with the murmurs auscultated at these sites.



1.
2.
3.
4.

3. Name some key features of Innocent Murmurs:

- (a)
- (b)
- (c)
- (d)
- (e)
- (f)
- (g)

Heart Murmur Cases

You are working in continuity clinic and will be seeing the Thomas family, including David, a 1.5 month-old for early 2mo well and Valerie, a 5 year-old for well visit.

Case 1: David

When asked about any concerns, Mrs. Thomas admits that David has recently had problems breastfeeding and she would like to see a lactation specialist. He seems to now have difficulty latching on and gets “worked up.” Even when he is given expressed breastmilk in a bottle, he seems to have problems with coordination and works so hard that he seems to sweat. Last night, it took him about 1 hour to finish 4oz.

What additional history would you like to know?

Mother’s prenatal history is remarkable for GDMA1. David was born full-term via SVD. “Now that you ask,” Mother reports that she was told David had a murmur on the MICC, but “it disappeared” by his 2-week check. His birth weight was at the 25th percentile. Reviewing his AHLTA growth-chart, you note that his weight has plateaued and is now at the 10th percentile. Mother remarks that he does occasionally have fast-paced breathing “when he gets worked up”. His face also “looks puffy,” but usually after doing tummy-time on his baby-blanket. Mother is unaware of anyone in the family born with heart disease or any unexplained deaths.

On exam, you appreciate a regular heart rhythm with a II/VI blowing holosystolic murmur which is heard best along the LLSB without radiation; there is normal physiological splitting of S2.

What other physical exam findings would be instructive? What additional testing (if any) would you like to perform during this visit?

What is your presumed diagnosis and plan?

Case 2: Valerie

Valerie proudly tells you that she's here for her Girl Scout Daisy camp physical. On review of PMHx, Mother reports that "she's my healthy one"; however, when you check your Autocites (which you haven't figured out how to shut off), you see "**murmur**" listed. Unfortunately, AHLTA crashes before you are able to further investigate her past encounters.

You proceed with her PE. Examining Valerie in a sitting position, you appreciate a Grade II/VI continuous murmur, mildly louder in diastole with a roaring quality, best heard in the right supraclavicular area. **Is your exam done?**

When you ask Valerie to lay down, her initial murmur is no longer audible, but you are now able to appreciate a Grade II/VI early systolic vibratory murmur best heard at the LLSB. **What do you think is going on? How do you counsel this family-- Can Valerie be cleared for sports?**

Heart Murmurs Audio-Clip Game

- You will hear clips of 10 different pediatric murmurs. Identify (a) **innocent vs. not innocent**; and (b) the **specific type of murmur or cardiac lesion**.
- You may work individually or as a group. Faculty may review the answers one-by-one, or after all 10 clips are completed.
- Please be sure to sit with your backs *turned away from* the overhead computer screen, so that you do NOT see the answer key. ***Turn the volume of the speakers up to 100% to hear all of the clips clearly.***

	Innocent or Not That Innocent	Specific Type of Murmur/Cardiac Lesion
1.		
2.		
3.		
4.		
5.		
6.		
7.		
8.		
9.		
10.		

Heart Murmurs Board Review

1. Included in your rounds today is a 36-hour-old boy who was born at term by normal, spontaneous vaginal delivery. His respiratory rate is 80 breaths/min and heart rate is 168 beats/min. He has easily palpable, bounding pulses in all four extremities, and his blood pressure is 72/30 mm Hg. Precordial examination reveals a lift and a 3/6 systolic ejection murmur at the upper left sternal border. You also note a murmur over the anterior fontanelle.

Of the following, the MOST likely diagnosis is

- A. aortic coarctation with congestive heart failure
- B. aortic insufficiency
- C. large ventricular septal defect with congestive heart failure
- D. left-to-right extracardiac shunting with congestive heart failure
- E. right-to-left extracardiac shunting with right heart failure

2. You are evaluating a 2-month-old girl as part of a routine health maintenance visit. Her mother tells you that she has no trouble feeding and is gaining weight like her previous children. Her precordial examination demonstrates a mild lift. The first and second heart sounds are normal. There is a systolic click at the upper left sternal border as well as a 3/6 systolic ejection murmur at the upper left sternal border with radiation to the axillae. Diastole is clear, and her pulses are normal in all extremities.

Of the following, the MOST likely cause of this patient's signs and symptoms is

- A. aortic stenosis
- B. atrial septal defect
- C. patent ductus arteriosus
- D. pulmonary stenosis
- E. ventricular septal defect

3. You are evaluating a 12-year-old girl as part of a sports screening program at the local school. She tells you that she has trouble keeping up with her friends during gym class and on the soccer field. On physical examination, she appears well and is in no distress. Her precordial examination demonstrates a mild lift. The first heart sound is normal, and the second heart sound is prominently split. There is a 3/6 systolic ejection murmur at the upper left sternal border. Diastole is clear, and her pulses are normal in all extremities.

Of the following, the MOST likely cause of this patient's signs and symptoms is

- A. aortic stenosis
- B. atrial septal defect
- C. patent ductus arteriosus
- D. pulmonary stenosis
- E. ventricular septal defect

4. You are conducting a preparticipation evaluation of a 14-year-old girl who is trying out for her school volleyball program. She is athletic and has never had any health problems. On physical examination, her height is at the 90th percentile and weight is at the 50th percentile for her age. Her lungs are clear, and cardiac examination reveals a systolic click and an apical systolic murmur that is late systolic and graded at 2/6 with radiation to the left axilla.

Of the following, the MOST likely diagnosis is

- A. anemia with high-output state
- B. aortic valve stenosis
- C. atrial septal defect
- D. mitral valve prolapse
- E. small midmuscular ventricular septal defect

5. You are evaluating a 15-year-old boy who will be attending sports camp in the summer. He tells you that he is very athletic, has no trouble keeping up with his peers during physical activities, and, in fact, has less fatigue with activities than most of his friends. On physical examination, he is well-developed and comfortable. The first and second heart sounds are normal. There is a systolic click at the upper right sternal border as well as a 3/6 systolic ejection murmur at the upper right sternal border. There is a thrill in his suprasternal notch. Diastole is clear, and his pulses are normal in all extremities.

Of the following, the MOST likely cause of this patient's signs and symptoms is

- A. aortic stenosis
- B. atrial septal defect
- C. patent ductus arteriosus
- D. pulmonary stenosis
- E. ventricular septal defect