Heart Murmur in a Child

Case Study and Commentary, David A. Danford, MD

Cardiac murmur is frequently recognized in healthy children, but it can also be the presenting feature in many forms of congenital heart disease, including regurgitation or stenosis of heart valves or left-to-right shunt lesions at the atrial, ventricular, or great arterial levels. Careful examination reveals innocent systolic murmurs in as many as 72% of all school-age children [1]. A high prevalence of innocent murmur also has been documented in infants and neonates [2]. Seven types of innocent heart murmurs are reported in children, including Still’s murmur [3–5], innocent pulmonary flow murmur [6–8], innocent pulmonary branch murmur of infancy [9], supracleavicular bruit [10], venous hum [11], mammary souffle [12], and cardiorespiratory murmur [13]. Generally, the clinical history and physical examination are diagnostic for these murmurs.

Congenital heart disease is much less prevalent than innocent murmur, occurring in only about 0.8% of live births [14], but the natural history of many common congenital cardiac defects can be one of progressive limitation and premature death. The primary care physician, therefore, very frequently faces the challenge of distinguishing between the relatively rare but important pathologic murmur and the ubiquitous innocent murmur. Failure to diagnose heart disease is unacceptable because current treatments can dramatically improve outcomes. On the other hand, the costs of embarking nonselectively on an aggressive laboratory and/or subspecialty evaluation [15] for as much as 70% of the pediatric population would be staggering. This article discusses diagnostic strategies that allow for timely identification of important congenital cardiac defects while at the same time controlling costs.

CASE STUDY
Initial Presentation

A 5-year-old boy is brought to the office by his mother for a health care maintenance visit in advance of the start of kindergarten. The child has been seen in the office regularly since birth for health care maintenance and immunizations, and occasionally for minor acute infectious respiratory ailments. At this visit, the physical examination reveals a systolic murmur for the first time.

Three findings are necessary to make the diagnosis of innocent murmur in a child. First, the examiner must recognize with confidence the classic auscultatory features of a specific innocent murmur. A summary of characteristics that help identify these murmurs is presented in Table 1. Second, a careful, cardiac-specific history must reveal no compelling evidence of heart disease. Third, a careful, cardiac-specific physical examination (beyond simple auscultation of the heart) must reveal no compelling evidence of heart disease. Laboratory testing is not necessary to make the diagnosis of innocent murmur in the vast majority of cases [16,17].

• What elements of the history are important in the evaluation of heart murmur?

Family History and Past Medical History

Clues to heart disease should be sought in the family history and the past medical history. A positive family history for congenital heart defect will raise the level of suspicion of pathologic heart murmur because of the tendency for congenital heart defects to cluster in certain families [18]. Moreover, in the course of taking the history, it is important to assess whether the child’s past medical history includes prior diagnosis of a genetic condition known to be associated with congenital heart disease, such as aneuploidy (eg, Trisomy 21 [19]) or other dysmorphic syndromes (eg, VACTERL [20]). As a matter of routine, physicians evaluating a child with a heart murmur should inquire about any major congenital defects of other organ systems because it is well known that many

From the Joint Division of Pediatric Cardiology, University of Nebraska Medical Center and Creighton University School of Medicine, Children’s Hospital, Omaha, NE.
<table>
<thead>
<tr>
<th>Type of Innocent Murmur</th>
<th>Timing, Location</th>
<th>Character, Pitch</th>
<th>Helpful Maneuvers</th>
<th>Common Clinical Setting</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Still’s</td>
<td>Systolic ejection; slightly lateral to left lower sternal border</td>
<td>Vibratory, musical, like a low-pitched stringed instrument</td>
<td>Diminishes in intensity with inspiration, sitting up, or standing</td>
<td>Although it can occur at any age, it is particularly common in young school-age children, often detected at the kindergarten physical; it is accentuated by fever or other high cardiac output states</td>
<td>Harmonic vibrations of the left ventricular outflow tract</td>
</tr>
<tr>
<td>Pulmonary flow</td>
<td>Systolic ejection; left upper sternal border</td>
<td>Low-intermediate pitch; non-harmonic, non-harsh</td>
<td>Diminishes in intensity with inspiration, sitting, or standing</td>
<td>Common in all pediatric age groups, particularly infants and preschool children; accentuated by fever or other high cardiac output states</td>
<td>Minor degrees of turbulence in the right ventricular outflow tract and main pulmonary artery in the absence of disease of these structures</td>
</tr>
<tr>
<td>Pulmonary branch murmur of infancy</td>
<td>Systolic ejection; left and right upper sternal border with strong radiation to axillae and back</td>
<td>Intermediate pitch; non-harmonic, non-harsh</td>
<td>None</td>
<td>Often first noted at about 2 weeks of age, it is especially common in premature infants; disappears in the first several months</td>
<td>Physiologic pulmonary branch turbulence due to size disproportion of main pulmonary artery and right and left branches</td>
</tr>
<tr>
<td>Venous hum</td>
<td>Continuous; right infraclavicular, less commonly on left or bilateral</td>
<td>Low-intermediate pitch; “machinery-like”</td>
<td>Disappears with supine position or with pressure on jugular vein</td>
<td>Common throughout childhood</td>
<td>Physiologic turbulence of normal superior vena cava flow</td>
</tr>
<tr>
<td>Supraventricular bruit</td>
<td>Systolic ejection; base of neck, right, left, or bilateral</td>
<td>Intermediate pitch; non-harmonic, non-harsh</td>
<td>Diminishes with neck extension with shoulders thrown back</td>
<td>School-age children</td>
<td>Minor physiologic turbulence in normal brachiocephalic arterial origins</td>
</tr>
<tr>
<td>Cardiorespiratory</td>
<td>Inconsistent timing and location; often at the cardiac apex</td>
<td>Cooing</td>
<td>May disappear when patient holds breath</td>
<td>School-age children</td>
<td>Technically this is a breath sound, arising from variable cardiovascular compression of an airway</td>
</tr>
<tr>
<td>Mammary souffle</td>
<td>Usually continuous, but may be systolic only; right, left or bilateral mid to lower sternal border</td>
<td>Low-intermediate pitch; “machinery-like”</td>
<td>Disappears with firm pressure with stethoscope</td>
<td>Pregnant or lactating women</td>
<td>Turbulence in engorged arteries and veins carrying high volume flow to and from the breasts</td>
</tr>
</tbody>
</table>
patients have cardiac anomalies as part of a constellation of birth defects, even if the constellation does not represent an identified genetic syndrome [21,22]. Because of the known association between connective tissue abnormalities and acquired cardiovascular disease, one should view a cardiac murmur in a child known to have Marfan syndrome [23,24] with a high level of suspicion. Specific inquiry about prior diagnosis of connective tissue disorders is appropriate. Finally, there are myriad inborn errors of metabolism that predispose to cardiomyopathies [25], some of which may produce murmurs of valvular insufficiency or dynamic left ventricular outflow obstruction. Therefore, learning that a patient with a murmur also has an inborn error of metabolism greatly changes the context of the murmur evaluation. The physician should also ask if there has been any prior diagnosis of a cardiovascular condition, such as a past episode of rheumatic fever, a past episode of Kawasaki disease, or known cardiac arrhythmia.

**Cardiac-Specific Issues in the Current History**

Some innocent murmurs have a specific time course over which they appear (eg, the innocent pulmonary branch murmur of infancy [26]), and some become prominent primarily at times of high cardiac output (eg, innocent pulmonary flow murmur, Still’s murmur [27]). Accordingly, inquiry into the time course over which the murmur has been noted and the circumstances under which it has been observed can be very revealing. Growth history is important because many forms of congenital heart disease compromise the patient’s rate of growth [28]. Generally, patients with compromised growth have large left-to-right shunts that produce the dual burdens of increased metabolic demands and poor oral intake. Older children may manifest similar types of heart disease by exercise intolerance, easy fatigue, and diaphoresis either at rest or on exertion. Respiratory complaints may include chronic cough, asthma-like symptoms, dyspnea on exertion, and poor feeding, especially in infants unable to simultaneously breathe heavily and suck and swallow. Inquiry concerning these symptoms of pulmonary edema due to large volumes of pulmonary blood flow and/or left heart failure is a central part of cardiac-specific history taking for pediatric heart murmur. Occasionally, patients with heart disease capable of producing a murmur will have associated cardiac arrhythmias. With this in mind, the physician should ask about dizziness, syncope, palpitations, and chest pain.

**Limited Cardiac History**

This 5-year-old boy has no history of heart murmur. There has been no dyspnea, cough, wheezing, or other chronic or recurrent respiratory symptoms. He has had no trouble keeping up with his peers on the playground. There have been no complaints of chest pain, dizziness, or syncope. He has grown well and met all his developmental milestones appropriately. He has no known disease of other organ systems, and was not born with any known malformation syndrome, connective tissue disorder, inborn error of metabolism, or chromosomal anomaly. The patient’s mother recalls that her brother had a heart murmur as a child, but she believes it resolved. She says there is no family history of specifically diagnosed congenital heart disease.

• What elements of the physical examination are required to evaluate this murmur?

**Cardiac-Specific Issues in the Physical Examination**

Physical examination in this setting must include careful auscultation of the murmur with attention to timing, location, radiation, pitch, intensity, character, and alterations in the murmur with changes in posture. Furthermore, one must evaluate the character of heart sounds, listen closely for accompanying clicks, gallops, and rubs, and palpate the precordial impulses and the pulses in the upper and lower extremities. Vital signs must be measured, including heart rate (and regularity), respiratory rate (and observation for cardinal features of respiratory distress: grunting, flaring, and retractions), blood pressure (upper and lower extremities), and height and weight (plotted on a growth chart). The patient’s general appearance must be observed for features suggestive of dysmorphic syndrome or chromosomal abnormality, distress of any kind, cyanosis, pallor, diaphoresis, and abnormalities of peripheral perfusion. The respiratory examination should include evaluation for chest deformities as well as auscultation for adventitial sounds (rales, wheezes, rhonchi, pleural rubs) and for discrepant breath sounds on the right versus left sides. The gastrointestinal examination should include palpation for the location of the liver (abdominal situs), the size of the liver and spleen, and the presence of ascites.

Certain characteristics of heart murmurs should raise suspicion that they are not innocent. For example, the continuous murmur of patent ductus arteriosus [29] will not disappear in the supine position like the venous hum will. Furthermore, innocent murmurs generally do not have the pansystolic timing observed in cases of ventricular septal defect [30] or mitral regurgitation [31]. While diastolic flow rumble and abnormal second heart sound are frequently recognized in atrial septal defect [32], these are not features of innocent murmur. Ejection clicks, often identified in association with pulmonary valve stenosis [33] and aortic valve stenosis [34] are absent in the examination of the normal...
heart. It is not surprising, therefore, that pansystolic murmur, loud murmurs (grade 3 or higher), harsh quality, left upper sternal border location, and murmurs associated with early- or mid-systolic clicks or an abnormal second heart sound all have been found to have an independent association with the discovery of heart disease [17]. Other authorities suggest that all diastolic murmurs, late systolic murmurs, continuous murmurs (that do not disappear with changes in posture or firm stethoscope pressure), and murmurs accompanied by other abnormal cardiac findings should be considered pathologic until proven otherwise [35].

Limited Cardiac Physical Examination

The patient has a normal appearance of a healthy 5-year-old, with height and weight plotting at the 75th and 80th percentiles, respectively, for age. The heart rate is 95 bpm and regular. Respiration are 20 breaths/min and not labored. Blood pressure is 95/55 mm Hg in the right arm and 102/60 mm Hg in the right leg. The chest is without deformity, and the breath sounds are clear and equal bilaterally. The precordium is without abnormal impulse. The first and second heart sounds (S1 and S2) are probably normal, but the physician is somewhat uncertain if there is abnormal splitting of S2. Also, the physician is uncertain whether there is an early systolic ejection click present or if this is merely a split S2. There are no gallops. A 2/6 low-to-medium pitched nonvibratory systolic ejection murmur is heard well in the third intercostal space at the left sternal edge. However, the murmur is also audible at the right sternal edge and faintly in the back; the physician is uncertain of the precise location of its maximal intensity. The murmur does not change when the patient sits up. There are no diastolic murmurs. Pulses are normal in intensity and equal in the arms and in the legs. Hepatosplenomegaly, ascites, or peripheral edema are not present.

- Can a clinical diagnosis of innocent murmur be made in this patient?

A limited cardiac-specific history has been taken, and this failed to identify any historical features to suggest heart disease in this patient. Most of the cardiac-specific physical examination is also reassuring. However, because of the lingering uncertainties about the location of the murmur, the characteristics of S2, and the presence or absence of an ejection click, the physician cannot claim to unequivocally recognize the features of a particular innocent murmur (based on the list in Table 1). The clinical diagnosis of innocent murmur cannot be made at this point.

- If this is not an innocent murmur, what is the differential diagnosis?

A prospective study evaluated the characteristics of outpatients referred for echocardiography from a pediatric cardiology outpatient clinic [36–39]; the most common cardiac

<table>
<thead>
<tr>
<th>Cardiac Lesion</th>
<th>No. of Patients</th>
<th>New Patients with Heart Disease Who Have Cardiac Lesion, %*</th>
<th>Mean Age at Diagnosis (25th, 50th, 75th Percentiles), yr</th>
<th>Male Sex, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect—small muscular</td>
<td>239</td>
<td>31.9</td>
<td>1.23 (0.04, 0.17, 0.83)</td>
<td>44.8</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>140</td>
<td>18.7</td>
<td>0.96 (0.08, 0.25, 0.75)</td>
<td>43.6</td>
</tr>
<tr>
<td>Aortic valve disease</td>
<td>120</td>
<td>16.0</td>
<td>5.63 (0.94, 4.00, 10.5)</td>
<td>68.9</td>
</tr>
<tr>
<td>Ventricular septal defect—large and/or not muscular</td>
<td>87</td>
<td>11.6</td>
<td>0.44 (0.05, 0.12, 0.23)</td>
<td>46.0</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>79</td>
<td>10.5</td>
<td>1.66 (0.38, 0.75, 1.33)</td>
<td>31.6</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>58</td>
<td>7.7</td>
<td>1.51 (0.04, 0.23, 1.37)</td>
<td>51.7</td>
</tr>
<tr>
<td>Mitral valve disease</td>
<td>39</td>
<td>5.2</td>
<td>6.90 (1.27, 5.50, 13.0)</td>
<td>38.5</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>26</td>
<td>3.5</td>
<td>3.93 (0.71, 2.25, 5.25)</td>
<td>65.4</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>16</td>
<td>2.1</td>
<td>6.37 (3.00, 4.50, 9.00)</td>
<td>62.5</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>10</td>
<td>1.3</td>
<td>0.03 (0.01, 0.015, 0.06)</td>
<td>60.0</td>
</tr>
<tr>
<td>Atrial ventricular septal defect</td>
<td>9</td>
<td>1.2</td>
<td>3.43 (0.16, 2.25, 6.50)</td>
<td>44.4</td>
</tr>
</tbody>
</table>

Note: Data from the Nebraska Defect Specific Diagnostic Accuracy Study [39], updated November 2001.

*Sum is greater than 100% because patients can have more than 1 lesion.
Heart Murmurs

In this patient, although the physical examination is somewhat ambiguous, an ejection click, if present, would suggest either pulmonary valve stenosis or aortic valve stenosis. The murmur in aortic stenosis would classically be more prominent at the right upper sternal border, and might transmit well to the neck. None of these findings were identified in this case. The description of this murmur’s probable left sternal border location and possible transmission to the back fits better with a main pulmonary arterial origin for the murmur, such as would be present with pulmonary valve stenosis.

If the click is actually absent, aortic or pulmonary valve disease is still possible but distinctly less likely. Innocent pulmonary flow murmur is possible, and subaortic membrane disease is still possible but distinctly less likely. Innocent pulmonary murmur would be present with pulmonary valve stenosis. With a main pulmonary arterial origin for the murmur, such a description of this murmur’s probable left sternal border and a widely split S2 would be unusual but is possible. Atrial septal defect generally produces a systolic ejection murmur of pulmonary origin but is usually associated with a diastolic flow rumble at the left lower sternal border and a widely split S2 that does not vary with respirations. The examination suggested the possibility of a split S2, but did not mention a diastolic flow rumble. Flow rumble can, however, be subtle findings. Whatever this murmur represents, it does not appear to be a significant hemodynamic burden for the heart since the patient is asymptomatic and does not have increased left or right ventricular precordial impulses.

Diagnostic Options

In general, when faced with uncertainty about a diagnosis, a primary care physician needs to gather more data, usually with laboratory testing, or by seeking a specialty or subspecialty opinion. For heart murmur, the laboratory testing considered is usually chest radiography, electrocardiography, or echocardiography. The subspecialty opinion would be available through pediatric cardiology consultation. Table 2 highlights some of the advantages and disadvantages of each of these approaches for the 2 most common examples of heart murmur in children, innocent murmur and ventricular septal defect. Although inexpensive, the chest radiograph and electrocardiogram (EKG) are neither sensitive enough nor specific enough to be helpful to the generalist trying to distinguish ventricular septal defect from innocent murmur [40,41]. The echocardiogram is diagnostically definitive, but this modality would be expensive for general application to large numbers of patients who turn out to have an innocent murmur [15]. Pediatric cardiology consultation to evaluate the not-clearly-innocent murmur screens out most innocent murmurs, and when ventricular septal defect is present, an echocardiogram can be arranged to define its anatomic location and physiologic significance.

Electrocardiogram

Similar to the observations with ventricular septal defect, the electrocardiogram has limited sensitivity for the other common forms of congenital heart disease that present with murmur [42–45]. Because many children with significant heart disease will have a normal EKG, it is hazardous to conclude from a normal EKG that a murmur is innocent. Some authorities point to advantages of the EKG as an integral part of the evaluation of murmur in children [46]. A further argument may be made that the incidental discovery of electrocardiographic abnormalities like Wolff-Parkinson-White syndrome or prolonged QT interval during screening could be very valuable in individual cases. However, large clinical series of children undergoing evaluation for heart murmur in the pediatric cardiology clinic have suggested that the expert clinical examination for evaluation of a murmur is not enhanced by the performance of an EKG [16,17,47]. Despite its low cost, the EKG is unlikely to be of help distinguishing the pathologic murmur from the innocent one in the primary care outpatient setting.

Chest Radiography

Like the EKG, radiography findings in congenital heart disease have poor sensitivity, with many of the classic radiographic features appearing late in the clinical course [42–44,48]. False-positive cardiomegaly is relatively common in young children due to a large thymus or poor inspiratory effort, and this further degrades the value of the test. Although some support the use of chest radiograph as a routine component of the pediatric cardiologist’s evaluation of heart murmur [46], investigators have had difficulty in showing an advantage to this approach [16,17,47]. Radiography should not be routine in the primary care physician’s initial evaluation of heart murmur.

Echocardiography

Echocardiography is an exquisitely accurate means for diagnosis of congenital heart disease when technologists trained and practiced in the pediatric examination perform the test using equipment suitable for children and when the test is interpreted by pediatric echocardiographers [49]. Evidence is accumulating, however, that the accuracy of echocardiography for children is reduced when it is performed in laboratories geared toward adult echocardiography and interpreted by physicians unfamiliar with imaging congenital heart disease [50,51]. Unfortunately, this modality is quite expensive.

**Table 2**

<table>
<thead>
<tr>
<th>Diagnostic Option</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest Radiography</td>
<td>Appears late in clinical course</td>
</tr>
<tr>
<td>Electrocardiogram</td>
<td>Limited sensitivity for other congenital heart disease</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>Definitive for pathologic murmur</td>
</tr>
</tbody>
</table>

**Table 3**

<table>
<thead>
<tr>
<th>Diagnostic Option</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest Radiography</td>
<td>Low cost</td>
<td>Poor sensitivity</td>
</tr>
<tr>
<td>Electrocardiogram</td>
<td>Expert clinical examination</td>
<td>Minimal enhancement</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>Exquisitely accurate</td>
<td>Expensive for general application</td>
</tr>
</tbody>
</table>

**What approaches to diagnosis of heart murmur are available?**

**Which should be used in this patient?**
relative to any of the modalities discussed thus far, and for this reason is poorly suited to serve as a screening tool. The use of echocardiography as a first-line diagnostic test after the primary physician appreciates a heart murmur is an expensive strategy. In 1993, the marginal cost of echocardiography to the patient/payer relative to the cost of pediatric cardiology consultation and subsequent echocardiography if necessary was more than $250/murmur evaluation [15].

**Pediatric Cardiology Consultation**

Probably because of training and practice, the experienced pediatric cardiologist offers greater diagnostic sensitivity and specificity for the diagnosis of congenital heart defect than does the generalist [52,53]. Measured against the anatomic standard of echocardiography, the pediatric cardiology specialty examination carries sensitivity and specificity of approximately 95% for discriminating heart disease from innocent murmur [16,54,55]. Ordinarily, consultation is available in a timely fashion, and the cost is relatively small, enhancing the appeal of using the pediatric cardiologist as a “second screen.” This strategy is less appealing if the pediatric cardiologist is at a great distance, especially if the murmur is in a neonate who must have a medically supervised transport to complete the consultation. The advantages of pediatric cardiology consultation are limited if the cardiologist is not selective in the use of expensive tests, such as echocardiography [15]. In the case patient, the

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**Table 3. Methods to Discriminate Ventricular Septal Defect (VSD) from Innocent Murmur**

<table>
<thead>
<tr>
<th>Diagnostic Modality</th>
<th>Possible Findings</th>
<th>Advantages</th>
<th>Disadvantages</th>
<th>Usual Findings</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>EKG</td>
<td>RVH</td>
<td>Inexpensive</td>
<td>Insensitivity: will likely be normal in small VSD</td>
<td>Normal</td>
<td>Inexpensive</td>
<td>False-positive findings will provoke anxiety and require further evaluation</td>
</tr>
<tr>
<td></td>
<td>LVH, BVH</td>
<td></td>
<td>Nonspecificity: abnormal findings are not unique to VSD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chest radiograph</td>
<td>Cardiomegaly</td>
<td>Inexpensive</td>
<td>Insensitivity: will likely be normal in small VSD</td>
<td>Normal</td>
<td>Inexpensive</td>
<td>False-positive findings will provoke anxiety and require further evaluation</td>
</tr>
<tr>
<td></td>
<td>Increased pulmonary vascular markings</td>
<td></td>
<td>Nonspecificity: abnormal findings are not unique to VSD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>VSD</td>
<td>Definitive for anatomic location of VSD</td>
<td>Expensive</td>
<td>Normal</td>
<td>Usually definitive and immediately available</td>
<td>Expensive</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Good indication of VSD size and pulmonary arterial pressure</td>
<td></td>
<td></td>
<td></td>
<td>Can be misleading if done in a lab unaccustomed to examining children</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Opportunity to reassess family</td>
<td></td>
<td></td>
<td>Can promote cardiac nondisease if results are not carefully presented</td>
</tr>
<tr>
<td>Pediatric cardiology consultation</td>
<td>Clinical findings</td>
<td>Inexpensive</td>
<td>Insensitivity: occasional failures to recognize small VSD</td>
<td>Clinical findings</td>
<td>Inexpensive</td>
<td>Sometimes not immediately available</td>
</tr>
<tr>
<td></td>
<td>highly suggestive of VSD</td>
<td></td>
<td>Nonspecificity: occasionally results in echocardiography when the heart is actually normal</td>
<td>highly suggestive of innocent murmur</td>
<td></td>
<td>Will, on rare occasions, fail to diagnose minor heart disease</td>
</tr>
</tbody>
</table>

BVH = biventricular hypertrophy; EKG = electrocardiogram; LVH = left ventricular hypertrophy; RVH = right ventricular hypertrophy.
approach of choice is a consultation with a pediatric cardiologist, who will then decide if echocardiography is indicated.

**Referral to Pediatric Cardiologist**

An outpatient evaluation by a pediatric cardiologist is arranged. Reexamination by this physician confirms a 2/6 low-to-medium pitched systolic ejection murmur maximal at the left upper sternal border with faint transmission to the back. The $S_2$ splits prominently enough so that the physician can easily discern the 2 components, but the splitting varies physiologically with respiration. An ejection click is indeed present at the left midsternal border. The physician makes a provisional diagnosis of mild pulmonary valve stenosis and arranges for echocardiography examination.

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**Table 4. Natural History and Risks with Common Shunt Lesions**

<table>
<thead>
<tr>
<th>Defect</th>
<th>Symptoms or Impairment</th>
<th>Potential for Spontaneous Improvement or Resolution</th>
<th>Endocarditis Risk</th>
<th>PVOD Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD, small</td>
<td>Generally none</td>
<td>May close spontaneously (muscular defects more likely to close than others), especially before age 2 years</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>VSD, moderate</td>
<td>May have none, but may have failure to thrive and chronic respiratory symptoms</td>
<td>Can diminish in size, and spontaneous closure is possible</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>VSD, large</td>
<td>Chronic respiratory symptoms and failure to thrive are common</td>
<td>Spontaneous decrease in size or even closure is possible, but severity of symptoms and risk of pulmonary vascular disease often dictates an intervention before this can happen</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>ASD</td>
<td>Usually none in early childhood, but more and more report fatigue, dyspnea, and palpitations with advancing age</td>
<td>Spontaneous closure of secundum ASD is possible, but uncommon, especially after age 2 years ASD in other locations is not reported to close spontaneously</td>
<td>No</td>
<td>Yes, but less likely, and usually at an older age than with large VSD</td>
</tr>
<tr>
<td>PDA</td>
<td>Usually none with small PDA, but chronic respiratory symptoms and failure to thrive are common in infancy with large PDA</td>
<td>Spontaneous closure after 6 months of age would not be expected</td>
<td>Yes</td>
<td>Yes with large PDA and pulmonary hypertension. No with small PDA.</td>
</tr>
<tr>
<td>AVSD, complete</td>
<td>Chronic respiratory symptoms and failure to thrive are common</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>AVSD, partial</td>
<td>As with larger secundum ASD (above)</td>
<td>No</td>
<td>Yes</td>
<td>As with larger secundum ASD (above)</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Can have respiratory symptoms early, but these diminish with time Cyanosis may be present early, tends to be progressive</td>
<td>No</td>
<td>Yes</td>
<td>No*</td>
</tr>
</tbody>
</table>

AR = aortic regurgitation; AS = aortic stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defect; LV = left ventricle; MR = mitral regurgitation; PDA = patent ductus arteriosus; PS = pulmonary stenosis; PVOD = pulmonary vascular obstructive disease; RA = right atrium; TR = tricuspid regurgitation; VSD = ventricular septal defect.

*Except in the unusual circumstances of associated large aorticopulmonary collateral arteries or after the surgical placement of a large systemic-to-pulmonary arterial shunt.
What is the rationale for echocardiography in this patient?

The sensitivity and specificity of the pediatric cardiologist's clinical diagnosis of pulmonary valve stenosis are good, but not perfect—73% and 82% in a recent report [37]. Therefore, echocardiography serves 2 purposes: to confirm that pulmonary valve stenosis is in fact the true diagnosis, and to assess the severity of obstruction.

- What are the risks for adverse outcomes in children with a heart murmur?
- What implications do these risks have for ongoing management?

Common Shunt Lesions

Ventricular septal defect, atrial septal defect, and patent ductus arteriosus are associated with well-defined risks for adverse outcome, including chronic respiratory symptoms and failure to thrive attributable to pulmonary overcirculation [28,40,41,56,57]. Some patients are also at risk for bacterial endocarditis, irreversible pulmonary vascular obstructive disease (Eisenmenger’s syndrome), and emergence of other hemodynamically important cardiac abnormalities [58–60]. The natural history and risks of these conditions are summarized in Table 4.

Management

Because all of the common left-to-right shunt lesions (eg, ventricular septal defect, atrial septal defect, patent ductus arteriosus) except for isolated secundum type atrial septal defect are believed to carry risk for bacterial endocarditis [61], the patient and family should be educated regarding use of antibiotics for dental and surgical procedures when any of these conditions is diagnosed. Although diuretics, afterload-reducing agents, and digoxin are commonly used in the initial management of the symptomatic child with a large left-to-right shunt [62,63], definitive management is generally surgical or transcatheter obliteration of the shunt.

Although concurrently controlled studies to demonstrate the superiority of surgical or transcatheter closure over the natural history or pharmacologically modified natural history will likely never be accomplished, the low surgical risks and benign postoperative course in the modern era [64–67] do appear to compare favorably with the natural history of common shunt lesions [60,68,69]. Early intervention is key for conditions likely to produce pulmonary vascular obstructive disease. With minor left-to-right shunts through small defects, the case for surgical or transcatheter intervention is far less compelling. Patients with small defects in the muscular ventricular septum, for example, are far better served by observation, reassurance, and antimicrobial prophylaxis against bacterial endocarditis during times of risk [69].
Common Valvular and Obstructive Lesions

A well-known set of risks is also associated with the commonly encountered valvaral and obstructive lesions such as pulmonary stenosis, aortic stenosis, and coarctation of the aorta. Discovery of such lesions is important because, even if mild, they represent indications for prophylaxis against bacterial endocarditis [59,61]. In addition, some of these patients are at risk for ventricular failure, valvaral regurgitation, arrhythmias, and sudden death [44,70]. Although most cardiac valve disease in children deteriorates hemodynamically with time or is static at best, mitral valve prolapse appears to be an exception. Auscultatory features of prolapse can, for reasons not well understood, disappear in a significant proportion of patients over a period of years [71,72]. The natural history and risks associated with valvaral and obstructive conditions are listed in Table 5.

Management

Relief of coarctation of the aorta is believed to reduce the incidence of early congestive heart failure and the potentially devastating complications of chronic upper body hypertension [73]. Surgical repair is therefore usually recommended upon diagnosis; however, there has been recent enthusiasm for balloon and/or stent aortoplasty [74].

The natural history of severe pulmonary valve disease is, in all likelihood, favorably impacted by relief of obstruction by balloon pulmonary valvuloplasty. Because transcatheter relief of pulmonary stenosis is quite effective and long-lasting in most instances, surgical valvotomy or valvectomy has fallen out of favor as a first-line approach for treatment of important pulmonary stenosis [75]. Mild pulmonary valve stenosis has such a favorable natural history that surgical or transcatheter intervention to relieve it is generally not believed to be worth the admittedly small risks [76]. Evidence is not strong to support either aggressive intervention or watchful waiting with moderate pulmonary valve stenosis, but current practice is to intervene for patients with a transvalve peak pressure gradient in excess of 40 mm Hg [77].
With a greater tendency for mild disease to progress to severe [78] and a greater risk for life-threatening complications among those with severe valve disease, ongoing regular follow-up of children with mild aortic valve disease is recommended [69]. Balloon aortic valvuloplasty for moderate or severe aortic stenosis should be considered palliative rather than curative, as there is a high likelihood that the aortic valve will eventually restenose or develop important degrees of regurgitation [79]. Nonetheless, it is widely believed that relief of moderate-to-severe aortic stenosis provides advantages over the natural history [70], and balloon valvuloplasty offers comparable hemodynamic results to surgical valvotomy without the disadvantages of sternotomy [79]. Surgical valvotomy or valve replacement is generally considered when balloon valvuloplasty is ineffective in relieving severe aortic stenosis, or when enough aortic regurgitation develops to raise concerns for left ventricular failure in the long term. Like balloon aortic valvuloplasty, surgical valvotomy is generally only palliative [79]. All surgical aortic valve replacement procedures carry significant risks for long-term complications and the all-too-frequent need for further intervention. Mechanical valves require anticoagulation (with the attendant risks) for the prevention of thrombosis and embolism [80]. Homograft valves implanted in the aortic position tend to calcify and deteriorate rapidly in children and adolescents [81]. Currently very popular, the Ross procedure uses the patient’s own pulmonary valve as an autograft in the aortic position and employs a homograft replacement in the pulmonary position, where homograft durability is better compared to the aortic position. Even the Ross procedure, unfortunately, is associated with some incidence of late deterioration of the autograft, or the homograft, or both [82]. Therefore, aortic valve replacement, by whatever means, is reserved for those patients in whom other effective means of treatment have been exhausted and whose valve disease is severe enough that the natural history would likely compare poorly with the postsurgical history.

Nondisease

Although not generally considered a risky condition in the classic sense, innocent murmur carries the peculiar risk of cardiac nondisease. The specter of possible heart disease in a child can be extremely burdensome for the child and the family. The child can be inappropriately “protected” from the normal activities of childhood by the family when the possibility of congenital heart disease remains a concern. Such observations prompted Bergman and Stamm [83] to coin the term “cardiac nondisease” in 1967 to describe the phenomenon. Cardiac nondisease remains a risk for children with innocent murmur in the current era, as noted by McCrindle et al. [84] and Young [85], who discovered that there was persistent concern about heart disease among the families of 10% to 17% of patients after the diagnosis of innocent murmur was made in the pediatric cardiology clinic.

Management

As discussed, the discovery of an innocent heart murmur in a child is not always free from adverse consequences; however, the physician can take steps to reduce the consequences. The admonition of Friedman [86] published a quarter century ago remains true today: “The physician must make clear to the patient with an innocent murmur that there is no need for systematic long-term cardiac supervision, antistreptococcal prophylaxis against rheumatic fever, or antibiotic prophylaxis against bacterial endocarditis. The patient and family must understand that the murmur will play no role in the prognosis of the patient even in the remote future, so there is no need for further cardiac evaluation or for restriction from any specific physical activities because of the murmur.” Physicians must convey these messages effectively to patient and family so that the child’s quality of life does not suffer due to inappropriate lifestyle restrictions imposed for cardiac nondisease.

Echocardiography Evaluation and Diagnosis

Echocardiography in the patient shows a minimally thickened pulmonary valve with minor restriction of systolic excursion at the tips, and accelerated main pulmonary arterial systolic flow velocity to 2.5 m/sec, consistent with a peak systolic pressure gradient of 25 mm Hg. The right ventricle is neither hypertrophied nor dilated. The remainder of the examination is normal. The physician informs the patient and family of the diagnosis of mild pulmonary valve stenosis and its favorable natural history. The physician reassures them that no restrictions are necessary for participation in athletics. Instructions are given for the appropriate use of antibiotic prophylaxis against bacterial endocarditis for dental or surgical work. Follow-up in the cardiology clinic in 3 years is scheduled.

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References


