Pediatric Cardiology Update: Office-Based Practice of Pediatric Cardiology for the Primary Care Provider

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In the following article, selected topics are presented to provide an update on the practice of pediatric cardiology from the perspective of the primary care provider and outpatient-based subspecialist. The approach to the evaluation of very common presenting signs and symptoms are reviewed, including heart murmurs, chest pain, and palpitations. The role of the primary care provider in screening and clearing children for participation in sports activities is discussed. Finally, issues of insurability, employment, and pregnancy in the adolescent and young adult with congenital heart disease are presented.

Heart Murmurs in Children

A murmur in the child is quite common and, consequently, the evaluation of a murmur is the most frequent reason for outpatient referral to most pediatric cardiology practices. It is estimated that up to 72% of children will have a murmur noted at some time during their childhood.1 Given the potential cardiac disease that could cause a murmur, this “diagnosis” is a source of concern for the primary care physician. The primary care physician’s comfort level of diagnosing innocent (or functional) murmurs depends on his or her clinical experience regarding innocent murmurs and any pathologic murmurs that they have detected in their training and practice. It is common to have a new community physician refer more patients for evaluation of a murmur in their first year after residency than it is during their fifth year. Which patient to refer for subspecialty evaluation can be influenced by other factors besides the murmur, including the anxiety level of the family. The family of a child referred to a pediatric cardiologist for murmur evaluation will come with many preconceived ideas and fears regarding their child’s murmur.2 Family history of cardiac disease and patient symptoms will influence how the family approaches this evaluation. In addition, a murmur detected during a routine presports physical can result in significant anxiety for the family as well as the child because of concerns about sudden cardiac death in athletes, a rare occurrence, but one that is prominently reported by the media. However, the current medical community pressures of proper use of resources have to be balanced with the potential medical and legal issues surrounding the evaluation of murmurs and a possible missed diagnosis of congenital or acquired heart disease. Thus, the primary care physician must balance clinical skills, various testing modalities at his or her disposal, and the availability of subspecialty referral options. What follows is a framework for the evaluation of the new murmur. Topics such as diagnostic skills, levels of competency, variety of testing modalities, and a cost analysis will be discussed.

Evaluation

History

The evaluation of the murmur begins at the same point as any other sign or symptom—a thorough history and physical exam. This process is dependent on the age at presentation, which greatly affects the evaluation of the murmur. Murmurs in the first days to weeks of life are more likely to represent cardiac disease as compared to new murmurs in the adolescent.3 The history of the newborn with a heart murmur
Murmurs in the first few weeks to months of life secondary to lesions associated with left-to-right shunting might have associated feeding difficulties, growth problems, and signs of “congestive heart failure” (pulmonary overcirculation). These symptoms can become more apparent or worsen as the pulmonary vascular resistance decreases over this time and there is more shunting of blood into the lungs. On the other hand, murmurs in the preschool and elementary school years are more likely to be innocent. It is common for a previously healthy child to have a “new” murmur when he or she is acutely ill in an urgent care setting. This murmur is most likely created by increased cardiac output and flow velocity in the outflow tracts secondary to fever, anemia, or the underlying illness. Of course, a child with a history of group A streptococcal pharyngitis and a new-onset heart murmur might have acute valvulitis associated with acute rheumatic fever. Other historical factors and symptoms associated with Kawasaki disease or bacterial endocarditis must also be considered in this age group. Finally, a family history of congenital heart disease might focus the physician on possible causes of a murmur at any age. Left ventricular outflow tract obstructive lesions may have a familial predomi-

nance; hypertrophic cardiomyopathy is transmitted in an autosomal dominant pattern with high penetrance, and velocardiofacial or DiGeorge sequence caused by 22q− microdeletion has been noted in many families.4

Physical Exam

General. After a thorough history has been obtained, a comprehensive physical exam should be performed. It is important to not immediately auscultate the child with a murmur, but rather to first observe the child. The general state of the child should be assessed, specifically detailing color, respiratory effort, and any dysmorphic features. Appropriate vital signs should be recorded including length/height and weight, heart and respiratory rate, and properly obtained blood pressures. Next, pulses can be assessed for character, volume, and correlation with the apical impulse. Any discrepancies between brachial and femoral pulses should be noted and suspicion of possible coarctation of the aorta should be entertained if there is a lower intensity in the femoral pulse or a delay between the brachial and femoral pulsation. The precordium should be palpated, noting any thrills or ventricular lifts; both are clues to an underlying cardiac abnormality.

Auscultation. After appropriate observation and palpation, the auscultatory examination is performed. This examination should be undertaken with ritualistic and systematic precision. Each heart sound should be dissected out of the general auscultation and listened to in isolation.5 All medical care providers who auscultate children should generate a personal systematic approach. This might include beginning with evaluating the first heart sound in the appropriate locations, and then isolating the second heart sound. These heart sounds should be evaluated for intensity and appropriate splitting. Any associated valvular clicks should be noted. Additional heart sounds (eg, S3 or S4) might then be assessed.

Next, the period of systole should be evaluated in isolation and any murmurs should be noted. The intensity, frequency, duration, quality, location, and radiation of the murmur should be described. The period of diastole then can be assessed and any distinct murmurs should be recorded using the same parameters as in the evaluation of the systolic murmur. Any continuous murmur should also be noted. The intensity of murmurs has historically been categorized into 6 levels or grades of increasing loudness, with a grade
IV murmur being associated with a thrill. The frequency of the murmur can provide some indirect physiologic information. Higher frequency murmurs are associated with more turbulent flow created by larger pressure changes, as in a small patent ductus arteriosus or ventricular septal defect. Lower-pitch murmurs suggest less turbulence associated with less pressure change, as might be heard in a very large ventricular septal defect.

Probably the most helpful parameter for distinguishing innocent from pathologic murmurs is the quality of the sound. The most common innocent murmur (Still’s murmur) has a typical vibratory and musical quality, whereas most pathologic murmurs have a much harsher, grating sound. In addition, a holosystolic or regurgitant murmur suggests either a ventricular septal defect or atroventricular valve regurgitation, whereas a systolic ejection murmur is usually created by aortic or pulmonary valve stenosis. Finally, the location of maximal intensity of the murmur offers significant insight into the cause of this sound, as does the area of radiation of the murmur. Only after all of the above parameters are noted can a complete description of the murmur be made to suggest a reasonable preliminary diagnosis.

Some murmurs, however, may not be clearly innocent or specific for a cardiac lesion, and a definitive diagnosis remains unclear. To further help qualify abnormal murmurs, McCrindle et al assessed 222 patients with murmurs to determine the qualities of murmurs that suggested cardiac disease. These clinical features included a grade III or louder intensity, pansystolic timing, a harsh quality, a location of maximal intensity at the left upper sternal boarder, and the presence of abnormal second heart sound or an early- to mid-systolic click. These features provided 92% sensitivity and 94% specificity in the diagnosis of pathologic murmurs by this group of experienced pediatric cardiologists.

Auscultation skills. As outlined above, there are many details to the auscultory exam that need to be learned and perfected over a physician’s training and career. There is a great body of literature dealing with various levels of competence in auscultory skills at different degrees of experience, but unfortunately there appears to be difficulty in the diagnosis of innocent murmurs at many levels of expertise. Gaskin et al evaluated the diagnostic accuracy of pediatric residents in the assessment of a wide variety of murmurs generated by a patient simulator. These residents demonstrated only a 33% rate of diagnostic accuracy with only a slight trend for improvement at higher levels of training. Of note, the sound most misinterpreted was the innocent murmur. Similar studies have been performed evaluating the auscultory skills of office-based and academic pediatricians. Haney et al evaluated the ability of 30 office-based pediatricians to correctly identify pathologic murmurs in a group of patients with known cardiac status: 43% of the children had pathologic murmurs, confirmed by echocardiography. These investigators reported a mean sensitivity of 82% and a specificity of 72% in the ability to differentiate pathologic from innocent murmurs. Thus, 18% of patients with pathologic murmurs would be misclassified as having innocent murmurs, and 28% of patients with innocent murmurs would be referred for further testing or evaluation by a subspecialist, a result considered “suboptimal” by the authors. In another study, Rajakumar et al found that academic general pediatricians were able to diagnose murmurs as pathologic with a similar sensitivity as compared with pediatric cardiologists (79% vs 85%). However, there was a marked difference in the accuracy of diagnosing innocent murmurs between the 2 groups, with the pediatric cardiologist more often correctly identifying innocent murmurs and thus having a higher overall diagnostic accuracy (76% vs 92%). Finally, Smythe et al confirmed the high level of accuracy of the pediatric cardiologist’s clinical exam in diagnosing pathologic versus innocent murmurs, with sensitivity values reaching 96% and specificities as high a 95%.

Testing. To increase the diagnostic accuracy of evaluating murmurs, multiple testing modalities, including electrocardiograms, chest radiography, and echocardiograms, have been evaluated. There has been much debate regarding the utility of noninvasive tests in the evaluation of murmurs. Newburger et al performed a prospective study evaluating the electrocardiogram, chest radiography and M-mode echocardiogram in the evaluation of murmurs in children over the age of 1 month. They found these tests to be of very little added value if the pediatric cardiologist believed that the murmur represented “no heart disease” or “definite heart disease.” If “possible heart disease” was suspected, the tests helped to categorize the disease state. These investigators did, however, support obtaining tests with respect to issues such as family reassurance, obtaining baseline values for future comparison, and quantification of severity of heart disease.
disease. Additional studies have found little value in electrocardiograms in the clinical diagnosis of innocent murmurs by experienced pediatric cardiologists, and suggest that electrocardiogram and chest x-ray findings at times lead to revisions of diagnoses that mislead both the primary care provider and the specialist.\textsuperscript{10,11}

However, Swenson et al\textsuperscript{12} report utility for noninvasive testing, with diagnoses such as small atrial septal defects, hypertrophic cardiomyopathy, and Wolff-Parkinson-White syndrome made with the help of chest x-rays and electrocardiograms in patients suspected clinically not to have heart disease. In addition, Danford et al\textsuperscript{13} found that almost 9\% (16 of 187) of children believed to have innocent murmurs by a pediatric cardiologist demonstrated cardiac disease by echocardiogram. However, the majority of these children (88\%, 14 of 16) demonstrated abnormal signs or symptoms, electrocardiograms, or chest x-rays. The authors concluded that there should be a low threshold for the pediatric cardiologist ordering an echocardiogram in very young infants and for select older children with signs or symptoms that suggest cardiac disease.

While echocardiography increases the diagnostic accuracy in the evaluation of murmurs, many studies report unsuspected cardiac disease in a minority of children with innocent murmurs, but at what cost? Yi et al\textsuperscript{14} created multiple models to evaluate the cost-effectiveness for the evaluation of murmurs that involved combinations of obtaining non-invasive studies (chest x-ray, electrocardiogram, echocardiogram) and referral to a pediatric cardiologist. This study concluded that, in general, chest x-ray and electrocardiogram are not cost-effective tools, that referral of only suspected pathologic murmurs to a sub-specialist is the least effective strategy, and that the most effective strategies involve either referral of all murmurs to a pediatric cardiologist or, at a higher cost ($158,000 per additional case detected), performing echocardiograms on all children with murmurs. Unlike Yi and colleagues, who used statistics from the literature as input data in their models, Danford et al\textsuperscript{13} used patient records to model cost-effectiveness and found that referral to a pediatric cardiologist with selective echocardiography to be the most cost-effective model in the evaluation of murmurs.

Finally, there are many forms of congenital heart disease that do not manifest themselves in the neonatal period until the pulmonary vascular resistance begins to decrease (small ventricular septal defects), and atrial septal defects might produce quite subtle murmurs (but characteristic fixed split second heart sound) in very early childhood. These murmurs might evolve into the more classic sounds as the child matures, and the primary care provider has the opportunity to repeat evaluations over time to help determine if a possible cardiac defect is present. An encouraging finding with most of the studies previously mentioned involves the severity of the missed diagnoses when murmurs were initially believed to be innocent. The forms of congenital heart disease were usually mild and of no hemodynamic significance (mitral valve prolapse, small atrial septal defects, and ventricular septal defects) and thus time spent in repeat evaluations would probably result in no adverse outcome.

\textbf{Summary}

Murmurs are common in the pediatric population and the role of the primary care provider is to differentiate the common innocent murmur from more pathologic sounds that could be created by structural heart disease. This differentiation begins with a pointed history and a systematic physical examination that dissects different heart sounds from the myriad of “noises” heard during auscultation. Expertise is enhanced with thoughtful analysis and experience, but many murmurs remain ambiguous. Many different testing modalities are available to aid in the assessment of the unclear murmur, but “routine” electrocardiograms and chest x-rays are probably not helpful. Referral to a pediatric cardiologist appears to be the next step for the ambiguous murmur. If the murmur still remains unclear, then an echocardiogram, performed by a technician trained in pediatric echocardiography, using appropriate equipment for the pediatric patient, and interpreted by a pediatric cardiologist, is the next most appropriate step in the evaluation.\textsuperscript{15}

Routine echocardiograms ordered by primary care physicians without pediatric cardiology consultation do not appear to be cost effective, and can be difficult to interpret if performed by laboratories that do not specialize in pediatric echocardiography.\textsuperscript{14,16}

\textbf{Chest Pain in Children: What to Do and When to Refer}

The anxious family sat as the pediatric cardiologist was about to address them. Their 10-year-old son had
just been referred for an echocardiogram because of a recent history of chest discomfort. He seemed to be well in all other regards, was not overweight, and was in no obvious distress as he was putting his shirt back on. The cardiologist had just reviewed the study and was matter-of-factly peering at the family while choosing his words carefully. “His heart is structurally and functionally normal based on the information obtained from this test. A full report will be forwarded to his physician.” As he turned to leave the room, the boy’s bewildered mother rustled in her chair and quickly inquired, “But doctor, then what is causing his chest to hurt?”

The above occurrence is frequently encountered in the circles of pediatric cardiology. Primary care physicians are routinely asked to assess children with complaints of pain, most commonly headaches and abdominal discomfort. However, the third most common “pain” confronted in this age group emanates from the chest.17 Recent statistics suggest 6 in 1000 pediatric emergency room visits are because of chest pain. This rate has more than doubled since the mid-1980s. Slightly more boys than girls are referred,18–20 and all races are proportionally represented.21 The most common age range for presentation is 8–16 years. Adolescents are more likely to have associated obesity and systemic hypertension.19,21

Most pediatric patients with chest pain have chronic or recurrent discomfort.21 Why have referral rates for chest pain increased, given the fact that pediatric chest pain is rarely life-threatening?19 Since the late 1970s and early 1980s the prevalence of coronary artery disease, obesity, and significant systemic hypertension in young adults has increased fairly dramatically.22 In addition, media reports of sudden unexpected death in young athletes have become commonplace in a society where the benefits of physical fitness are widely publicized.18,22 More than 50% of young patients with chest discomfort believe their heart is the cause, yet it is known that “children are not born with the idea that chest pain comes from the heart.”23,24 Coupling these facts with a possible inclination to transfer our own adult fears onto our children has resulted in substantial increases in emergency room visits, calls to 911, and referrals to pediatric cardiologists.18,19 It is common practice for youth to be released from school because of chest pain. Practitioners fear the repercussions, medical and legal, of misrepresenting myocardial infarction or the forerunners of sudden death for benign chest discomfort.17,19 However, heart disease is more likely to be recognized during routine evaluation of children and adolescents than during an evaluation for chest pain, especially if the latter assessment occurs in an office setting.23,25

As previously stated, the overwhelming majority of young patients with chest pain have etiologies that are considered benign.17 In an attempt to uncover the cause in a given individual, it is often useful to distinguish cutaneous or musculoskeletal pain historically from visceral pain. The former tends to be brief, sharp or stabbing, and usually is localized. It immediately follows a stimulus, is maximal at the onset and slowly dissipates, is often positional, and normally is reproducible on examination.26 Benign chest discomfort is apt to be recurrent, and may often be associated or replaced with symptoms such as fatigue, abdominal pain, and headache.27

Most important, it should be emphasized that one of the least likely causes of chest pain in children is the heart.19,26 Assuredly, musculoskeletal, gastrointestinal, pulmonary, and psychiatric etiologies for chest pain are far more commonplace, despite a fervent misconception to the contrary.19,26

Noncardiac Etiology

Many of the noncardiac etiologies of chest discomfort are categorized in Table 1. Discrete musculoskeletal pathology explains approximately 20% of these maladies.19 Traumatic injuries, even weeks after the event, are easily understood sources.26 The keen observer likewise recognizes that overuse afflictions such as sprains and spasms can also occur in the chest wall.26 Costochondritis is common in adolescents, especially in young women. It is usually unilateral, often in the left thorax, and significant discomfort is produced by movement or palpation of the costochondral or chondrosternal articulations.17,26 When this pain is associated with a solitary, spindle-shaped swelling in the chest wall, typically at the right sternoclavicular junction, it is referred to as Tietze’s syndrome. A rare entity in the pediatric world, slipping rib syndrome, involves abnormal mobility of the eighth, ninth, and tenth ribs. The ribs do not directly attach to the sternum but attach to each other by means of dense, fibrous tissue. Activity or unrecognized trauma may result in thoracic pain, abdominal pain, and can...
be elicited by bending over or by pulling anteriorly with the fingers hooked under the lower rib cage. This pain is believed to be generated by weakened fibrous tissue irritating intercostal nerves. While most musculoskeletal ailments benefit from rest and anti-inflammatory medications, the slipping rib syndrome may occasionally require surgical intervention. Pulmonary sources of chest discomfort comprise an additional 15% to 20% of the total cases. Chest pain associated with reactive airway disease, especially when exacerbated by exercise and activity, can be particularly difficult to differentiate from cardiac causes and often requires extensive investigation. Pneumonia, specifically when coupled with pleural inflammation, can be very painful because the pleural membrane contains many pain receptors. Activation leads to searing, sharp discomfort, which may affect breathing. The development of a spontaneous pneumothorax, common in cystic fibrosis and Marfan syndrome, is a well-known cause of chest pain. Another etiology that is difficult to distinguish from cardiac disease is a pulmonary embolism. This appears to be more prevalent in the female, those who are overweight, those taking oral contraceptives, or anyone with a history of leg trauma. Thrombophilic conditions should be investigated in the child with a pulmonary embolism.

While not common as a cause of chest pain, gastrointestinal etiologies are responsible for approximately 5% of the cases. In the pediatric population, gastroesophageal reflux disease is the primary culprit, often leading to severe, erosive esophagitis. This pain often awakens the patient while they are supine, and can be precipitated by the introduction of cold or hot substances. Gastritis and peptic ulcer disease are less common as causes of chest pain, and are typically associated with chronic disease, immune deficiency, or medications. Rarely, unrecognized foreign body ingestion can become a source of chest discomfort.

Often overlooked but extraordinarily important causes of chest pain are psychiatric issues. In at least 10% of all individuals presenting with complaints of chest pain, the primary underlying pathology is psychological. The stress associated with family members dying suddenly or unexpectedly, a myocardial infarction in a parent or grandparent, and the concern that they may experience a similar event can be devastating to a child. Anxiety attacks with subsequent hyperventilation may add to the symptoms. It is not unusual to develop an overt conversion reaction from the overwhelming stress of difficult family events. Chest pain is a response that is most common in adolescent women, while younger children will typically complain of headache or abdominal discomfort. A number of other etiologies for chest pain in young people have been described as well. The most common of the “miscellaneous” causes is the precordial catch syndrome, a frequently seen but poorly understood entity. The pain associated with the precordial catch syndrome is sudden in onset; very sharp in nature, usually described as knife-like, needle-like, or shooting; and the pain does not radiate beyond the precordium. The pain is exacerbated by inspiration in almost all cases, is sporadic with little relationship to exercise, and is generally brief in duration (30 seconds to 5 minutes). While its etiology is unknown, the chest pain associated with precordial catch syndrome is benign. Precordial catch syndrome may be the most common cause of chest pain in children referred for elective outpatient evaluation. The acute chest syndrome, linked specifically to sickle cell disease, is frequently confused with myocardial ischemia and can be extensive in its duration. The presence of a chest tumor may also present with chest discomfort, but this is rare and usually not isolated in it symptoms. Breast tenderness in both adolescent males and females is encountered with relative regu-

TABLE 1. Noncardiac causes of chest pain

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<thead>
<tr>
<th>I. Musculoskeletal (20%)</th>
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<tr>
<td>A. Trauma</td>
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<td>B. Overuse injury</td>
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<td>C. Costochondritis</td>
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<tr>
<td>D. Tietze syndrome</td>
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<td>E. Slipping rib syndrome</td>
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<th>II. Pulmonary (15%-20%)</th>
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<tr>
<td>A. Reactive airway disease</td>
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<td>B. Pneumonia</td>
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<td>C. Pleuritis</td>
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<tr>
<td>D. Pneumothorax</td>
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<td>E. Pulmonary embolism</td>
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<th>III. Gastrointestinal (5%)</th>
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<tbody>
<tr>
<td>A. Esophagitis</td>
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<tr>
<td>B. Gastroesophageal reflux disease</td>
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<tr>
<td>C. Gastritis</td>
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<tr>
<td>D. Peptic ulcer disease</td>
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<td>E. Foreign body ingestion</td>
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| IV. Psychiatric (1%) |

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<th>V. Miscellaneous (15%-20%)</th>
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<tbody>
<tr>
<td>A. Precordial catch syndrome</td>
</tr>
<tr>
<td>B. Sickle cell disease</td>
</tr>
<tr>
<td>C. Chest tumor</td>
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<tr>
<td>D. Breast pain</td>
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| VI. Idiopathic (25%) |
Cardiac Etiology

Despite exhaustive efforts to assess the heart in pediatric patients with chest pain, a cardiac origin is rarely implicated. No more than 3% to 4% of pediatric chest pain patients have a cardiac basis. Cardiac causes of chest pain can be categorized as congenital lesions, acquired processes, and miscellaneous conditions (Table 2).

As a rule, cardiac chest pain is either ischemic, inflammatory, or arrhythmogenic in origin. Ischemic pain is characterized as squeezing, crushing, or pressure-like, often with radiation to the arms, neck, back, or jaw. It tends to be reproducible, is provoked by exercise, and is relieved at rest. Ischemic pain can also be precipitated by cold exposure, eating, and emotions. Inflammatory conditions result in almost constant discomfort, at times improved with positioning, but clearly exhibiting a visceral quality. Arrhythmogenic discomfort usually results from the incessant pounding of the heart, and on occasion is associated with sharp chest pain.

Certain congenital cardiac anomalies may be associated with ischemic chest pain, but most are not. Patients with “mild” congenital heart disease (e.g., atrial septal defects, ventricular septal defect) are no more likely to experience ischemic chest pain than normal children. On the other hand, those with moderate to severe left ventricular outflow tract obstruction (valvar, subvalvar, or supravalvar aortic stenosis) may develop subendocardial ischemia under specific circumstances, such as exercise. Rarely will severe pulmonary stenosis act similarly. The most likely right ventricular lesions to result in exertional chest discomfort are pulmonary hypertension and Eisenmenger’s syndrome.

Coronary artery anomalies comprise a very important set of lesions that must be considered in the context of cardiac chest pain. The most prevalent lesion, origin of the left coronary artery from the pulmonary artery, results in myocardial ischemia due to poor perfusion pressure. This may present as early as 6 weeks of age, when pulmonary vascular resistance falls with resultant perfusion deficits to the left coronary system. When the left coronary artery arises from the right coronary artery, it may course between the aorta and the main pulmonary artery, and has potential for compressive ischemia when the great vessels dilate, as during exercise. The same holds true when the right coronary artery arises from the left coronary artery. Coronary artery fistula can establish a coronary steal phenomenon, especially if the vessel is large.

Hypertrophic cardiomyopathy is a relatively rare but important lesion to recognize. During periods of increased myocardial oxygen demands, such as exercise, which increases both heart rate and blood pressure, diastolic filling time shortens and systolic wall stress increases. This results in substantial subendocardial ischemia and even malignant arrhythmias, all a function of diminished coronary perfusion.

Although mitral valve prolapse is seen with some degree of frequency in the pediatric population, whether there is simply “an association” with chest pain or it is “a cause” of chest pain is not clear. It is correlated with tachyarrhythmias, but it is unknown if mitral valve prolapse actually predisposes the individual to papillary muscle ischemia.
A most unusual congenital cardiac lesion known to cause chest pain is absence or partial absence of the pericardium.\textsuperscript{19,22,34} Usually the pain associated with it is sharp, non-exertional, but often is positional, becoming more intense when the patient is supine or left in a lateral recumbent position. The discomfort itself relates to herniation of the left atrial appendage or other portions of the heart muscle through the deformed pericardium. This surgically amenable lesion is occasionally recognized initially during postmortem examination.\textsuperscript{18}

While inflammatory or acquired cardiac conditions are uncommon in children, the chest pain associated with them is most often encountered when the pericardium is involved, such as in pericarditis, post-pericardiectomy syndrome, and autoimmune disease.\textsuperscript{17,26} Patients with a history of Kawasaki disease should always be taken seriously when complaining of chest pain, because of possibly unrecognized coronary artery stenosis. Patients with significant left ventricular dysfunction (myocarditis and dilated cardiomyopathy) develop chest discomfort with exertion because of coronary insufficiency.\textsuperscript{18,26}

In children, true myocardial infarctions unassociated with congenital or acquired heart disease are quite rare, yet they do occur. More and more young patients with familial hyperlipidemias are being recognized because of identification of these disorders in parents or family members. Unfortunately, illicit drugs remain prevalent in our society, specifically cocaine. Cocaine is a well-known to increase sympathetic activity, increase myocardial oxygen consumption, and cause coronary artery vasospasm—with subsequent myocardial ischemia.\textsuperscript{26} Patients with Marfan syndrome and Turner syndrome are prone to aortic root dissection, a well-described cause of acute chest pain in the adult.\textsuperscript{19,26,32}

**History**

Of course, the initial approach to these patients is a thorough, complete history.\textsuperscript{18,26} Whether a primary practitioner or a pediatric cardiologist, the process is time-consuming and should not be rushed. Even if one’s goal is simply to exclude a cardiac etiology, the effort can be extensive. It is vital that the physician allow the patient to describe the pain, as much as possible, in their own words. This also prolongs the encounter and may confuse the portrayal, but it may represent the most important aspect of the interaction.\textsuperscript{26}

It is important to obtain as much information as possible about the pain before its occurrence, during the episode, and after the event.\textsuperscript{19} Ischemic chest pain is typically severe, mid-sternal, radiating to the shoulder, jaw, or left arm, and is described as diffuse, crushing, pressure-like, and is associated with significant difficulty breathing.\textsuperscript{18} There can be nausea, diaphoresis, and pallor, though the latter is quite nonspecific. Palpitations or a feeling of inappropriate tachycardia before the episode should be a tip to aggressively investigate for an underlying rhythm disturbance. Of utmost importance is association with exercise. Cardiac chest pain is frequently precipitated by activity and dissipated by rest, and it is characteristically reproducible. Most benign chest pain is sporadic, unpredictable, and is not reproducible unless there is palpable tenderness over the sternum.\textsuperscript{18,19,26} It tends to be nonvisceral in quality, usually sharp, and with little radiation.\textsuperscript{18} Because the pain can worsen with deep inspiration, associated shortness of breath and hyperventilation symptomatology is common, even in benign chest pain. Its relationship to exercise is inconsistent at best. Pay particular attention to very young children (<6 to 8 years of age) when they complain of chest pain. They often perceive palpitations as pain and have difficulty explaining and understanding what they experience.\textsuperscript{19}

**Physical Examination**

Most children with chest pain have normal examinations, unless they have underlying congenital heart disease.\textsuperscript{26} Particular attention should be paid to the overall level of distress, the vital signs, and tenderness over the chest or abdomen.\textsuperscript{19} Intensity of the pulses and the cardiac impulse are noted. Lung auscultation should detect the status of air exchange and any unusual adventitious sounds.\textsuperscript{18} Cardiac auscultation is focused on the detection of murmurs, rubs, clicks, and extra heart sounds.\textsuperscript{19}

An abnormal cardiac exam further heightens suspicions of a cardiac cause for the pain. Left or right ventricular outflow tract obstruction murmurs should be readily apparent. A coronary artery fistula is suggested by a soft, continuous murmur over the precordium.\textsuperscript{22} Hypertrophic cardiomyopathy with obstruction usually produces a fairly harsh systolic murmur at the right upper sternal border, with radiation to the apex and carotids. The click and subsequent murmur resulting from mitral valve prolapse may be accentuated with the patient in multiple positions, including...
supine, sitting, standing, and squatting. Friction rubs may be a sign of a pericardial process or exacerbation of an autoimmune disease.18

**Laboratory**

Following completion of the history and physical examination of the patient with chest pain, the focus shifts to laboratory testing. While each patient must be addressed individually, there are many for whom laboratory studies are simply unnecessary.19 Therefore, the need for testing and further evaluation should be tailored to the patient in all cases.18 Generally speaking, the need for extensive studies is rare.31 For those patients where there is a strong suspicion of a pulmonary or a cardiac etiology to the pain, some or all of the following may be indicated: electrocardiography, chest x-ray, echocardiography, exercise testing (with or without radionuclide imaging), ambulatory electrocardiographic monitoring, and pulmonary function tests. Depending on the history and physical examination, laboratory studies such as a complete blood cell count, antinuclear antibody titer, sedimentation rate, creatine phosphokinase concentration, troponin I, thyroid function tests, and drug screening may be warranted.18,22 Clearly, the more impressive the history and physical findings, the more important and the more detailed laboratory testing will be.18

As a primary practitioner initially evaluating a child with chest pain, it can be difficult to know how far to proceed with the evaluation. A complete, thorough history and physical examination is mandatory for all individuals. Both the patient and the parents take the symptomatology complex seriously and expect the physician to do the same. A cursory time commitment and a lack of interest on the part of the physician will result in considerable frustration within the family and efforts to seek appraisal elsewhere.23 As a general rule, the primary care provider should be neither too hasty nor overly exhaustive in the assessment, but seek a comfortable balance.19 Those individuals with obvious musculoskeletal symptoms should be treated with rest, anti-inflammatory agents, and strong reassurance. Those with pulmonary or gastrointestinal symptoms or findings should be managed with the appropriate therapy and followed accordingly. An electrocardiogram with (or without) a chest x-ray may serve as useful "screening" tools here and are cost-effective entities. It is indeed very difficult to place a price on reassurance.35

**Referral**

As noted above, most patients and families seeking medical attention because of chest pain in their child do so because of concerns about the heart. However, only a very small percentage (3% to 4%) is cardiac-based and, thus, it would be inappropriate for all of them to be referred to a pediatric cardiologist. Patients with visceral-type chest discomfort associated with nausea, palpitations, syncope, presyncope, or exercise, as well as those with abnormal cardiac examinations, chest x-rays, or electrocardiograms, deserve a much more thorough cardiovascular assessment. Particular attention should also be given to those who have undergone previous cardiac surgery and those with a family history of sudden unexpected death or early coronary artery disease.26 These patients are best evaluated by a pediatric cardiologist. While most patients do not require immediate referral to an emergency room, those with a history of significant heart disease, chest trauma, severe respiratory distress, palpitations, or syncope are best evaluated in this setting if the discomfort is persistent.18

The most difficult patients to assess with chest pain are those where the etiology is not readily apparent to the practitioner. While most of these individuals have idiopathic or benign chest discomfort, there remains an uneasiness regarding the extensiveness of their testing. The primary caregiver in this situation is concerned about missing a life-threatening cardiac condition, tries to do the right thing, and calm an anxious child and his or her family.22 While many of these children are ultimately referred to a pediatric cardiologist, it is important not to routinely order studies unless they can be justified. It is the role of the pediatric cardiologist to carefully evaluate the heart and confidently relate those findings to the patient and the family.26 When no cardiac condition is found, the ability of the cardiologist to communicate the benign nature of the discomfort to the patient and their family is vital. If the family is left with the impression that the reason for the pain is unknown, they will continue to seek out others to help them and repeated trips to the emergency room will occur.17,23

**Summary**

In summary, chest pain in children and adolescents is quite common, but rarely is it cardiac in origin or life-threatening. A thorough history and examination, followed by appropriate testing is germane for their
initial assessment. In patients with visceral-type discomfort associated with exercise, palpitations, syncope, significant congenital or acquired heart disease, abnormal physical exams, and a positive family history, a more extensive cardiac evaluation is justified. Above all, the practitioner is obligated to effectively communicate the findings to the patient and family, and when appropriate, reassure them in order to allay any unnecessary anxiety or emotional turmoil.

**Palpitations**

Palpitations are defined as “forcible pulsations of the heart perceptible to the patient usually associated with an increase in frequency, with or without an irregularity in rhythm.” Palpitations in children have been shown to be neither a sensitive nor a specific symptom of heart disease. Children and adolescents may use a variety of terms to describe their feelings of palpitations. Young children may have a difficult time describing the sensations they feel. Further complicating this is the fact that the sensation of palpitations can often occur in the presence of normal sinus rhythm. In contrast, patients with incessant tachycardias may not report feelings of palpitations and some life-threatening arrhythmias may not present with palpitations. How, then, can one best evaluate a patient presenting with a complaint of palpitations?

**History**

The key to an accurate diagnosis and effective treatment begins with a thorough history of the patient’s symptoms. The report of palpitations should be taken seriously and the child should be asked to describe the symptoms in their own words. It may be difficult for young children to describe the sensations they experience and the report from parents of what the child has sensed or words that the child has used with them to describe the symptoms should be obtained. Historical points recommended for inclusion in a thorough evaluation include:

- How long has the patient had these symptoms? When did the symptoms first begin? Did the symptoms begin after some specific event? How many episodes have actually occurred?

These are questions that help a physician understand what the patient is experiencing. For example, if the patient had been riding a bicycle and the parents noted the heart beating rapidly, and this was the only event, it was likely related to the patient’s recent exercise rather than an arrhythmia. Of course, if the parents report that the child participated in only quiet activities and presented with a rapid heart rate associated with chest pain, dizziness, or other such symptoms, then this was more likely an arrhythmic event.

- How often do the episodes occur?

Patients with an arrhythmogenic etiology to their symptoms rarely have symptoms on a daily basis. Their symptoms are typically separated by weeks or even months. Sometimes their symptoms can occur in clusters consisting of several episodes occurring over a several day period, and then interspersed with periods of quiescence. However, patients without an arrhythmogenic etiology to their symptoms may complain of symptoms occurring several times each day. This question also allows selection of testing that is most likely to lead to a diagnosis. For example, patients who have symptoms on a daily basis should undergo 24-hour to 48-hour ambulatory electrocardiographic monitoring (Holter monitoring) in order to capture a recording of the event. On the other hand, a patient with an episode once a month will need other types of monitoring.

- How long do the episodes last?

Duration of symptoms is important to determine the likelihood of an arrhythmia. Patients who report palpitations of a few seconds duration are less likely to have a clinically significant arrhythmia than those that report palpitations lasting several minutes or more. Patients that notice irregular heartbeats or “skipped heart beats” lasting only seconds in duration require different types of monitoring than those with sustained episodes.

- Are there any particular events that make the episodes occur?

Some patients have specific triggers for their episodes. Exercise may be an important precursor, or the events may occur at random times. This information allows development of an appropriate strategy to monitor such events.
What do you typically do when you have an episode?

This is useful to determine any specific actions that produce exacerbations or remissions. Children may have learned to self-perform a Valsalva maneuver, including gagging, yawning, drinking cold water, or other vagal stimulant activities that terminate supraventricular tachycardia. Other patients may report that the episode gradually resolves. Episodes that have a gradual onset and gradual resolution are less likely to be arrhythmogenic than those with sudden onset and sudden termination.

Are there any associated symptoms with your palpitations?

It is more likely to be an arrhythmogenic event should the patient report dizziness, loss of consciousness, or anginal type chest pain during their symptoms. Patients with arrhythmias will often complain of feeling the palpitations “in the back of their throat.”

How does the patient appear during an episode?

Again, it is more worrisome and more likely to be an arrhythmogenic event if the patient appears pale, has dizziness, or appears quite frightened during the episode. It is important to inquire about diaphoresis and nausea, as these are more typically associated with an arrhythmia.

Finally, inquiring about the patient’s general lifestyle is useful.

Does the patient ingest an excessive amount of caffeine? Are they athletic? Is there a family history of arrhythmia or sudden death?

Investigate anxiety as a cause of (or created by) the patient’s symptoms, especially if a relative has recently been diagnosed with an arrhythmia or even died because of an arrhythmia. Such anxiety may exacerbate a benign problem or create the illusion of an arrhythmia when none is present.

Physical Examination

Coupled with a thorough history is a complete physical examination, which should emphasize the cardiovascular system. Structural heart disease should be actively investigated. Abnormalities in the cardiovascular examination should be evaluated with additional laboratory testing because the prognosis of an arrhythmia depends on the presence of underlying cardiac disease. For example, premature ventricular contractions in a structurally normal heart are likely benign. However, the same rhythms in a patient with structural heart disease, for example, dilated cardiomyopathy or postoperative tetralogy of Fallot, may be a risk factor for sudden death. One should also be aware that children with rhythm problems might have a completely normal cardiac examination. Abnormalities of the electrical system may not produce abnormal physical findings, such as murmurs, cardiomegaly, or abnormal pulses, unless associated with structural heart disease.

In addition, searching for other disease processes is important. Disease processes such as fever and anemia may secondarily cause murmurs related to a high-output cardiac state and may also lead to sinus tachycardia. Hyperthyroidism can lead to sinus tachycardia and the perception of palpitations.

Differential Diagnoses

Following the history and physical examination, one should have an idea of the patient’s diagnosis. The differential diagnoses commonly include patient anxiety, systemic illness, premature beats resulting from atrial or ventricular ectopy, or a more complex arrhythmia of some type.

Anxiety. Anxiety in children can often trigger the sensation of palpitations. The anxiety is usually related to either school or family stressors. From the answers to the historical questions outlined previously, the child with anxiety will typically relate that the episodes occur multiple times each day and are short-lived in nature. The child will not have associated symptoms although occasionally they may have shortness of breath associated with panic attacks. The episodes resolve gradually. Specific inquiries about the family situation or the child’s situation at school are extremely helpful. Developing a rhythm monitoring strategy that allows these events to be recorded is important.

Systemic illness. Systemic illnesses that result in a high-output cardiac state can also induce the symptoms of palpitations. Usually the patient will have other symptoms associated with the palpitations. Any illness that results in a fever will increase the heart rate and cause sinus tachycardia. Systemic illnesses that cause anemia may also result in palpitations. If anemia is suspected clinically from the physical examination due to patient pallor, a complete blood count will confirm it. Hyperthyroidism can also result in symptoms of palpitations due to intermittent sinus tachycardia. Patients may have symptoms associated with
their hyperthyroidism, such as emotional lability, tremors, weight loss, or exophthalmos. Thyroid function testing provides confirmation. These patients may require a monitoring strategy to evaluate their rhythm, which will reveal sinus tachycardia or, rarely, atrial fibrillation.

**Ectopic beats.** Some patients are able to sense and report premature beats. These can be due to premature atrial contractions or premature ventricular contractions (Figures 1 and 2). Although isolated premature atrial or premature ventricular contractions are benign with a structurally normal heart, a monitoring strategy is necessary to capture these events and make a definitive diagnosis. The patients can then be counseled regarding the benign nature of these events. Occasionally, should the events persist with continued patient symptomatology, medications may rarely be necessary to decrease the frequency of the premature beats and therefore the frequency of symptoms. In a structurally abnormal heart, further evaluation of both the underlying cardiac disease and the arrhythmia is necessary to determine a prognosis and select effective treatment.

**Arrhythmia.** The most common arrhythmogenic etiology for palpitations in children is supraventricular tachycardia (Figure 3). In younger children this is typically orthodromic atrioventricular reentry tachycardia utilizing a concealed accessory pathway. This means that the tachycardia has antegrade conduction through the atrioventricular node and retrograde conduction through an accessory pathway with reactivation of the atrial tissue. This mechanism is called reentry because the impulse “reenters” the atrium to initiate another beat of tachycardia. This is the most common type of supraventricular tachycardia in young children. In older children, adolescents, and young adults supraventricular tachycardia is more likely the result of atrioventricular nodal reentry. Atrioventricular nodal reentry tachycardia is the result of reentry within the atrioventricular node itself.

Ventricular tachycardia is an uncommon presentation for the complaint of palpitations, but may be noted occasionally in children (Figure 4). If the episodes are nonsustained, with a structurally normal heart, then the episodes may be benign. If the episodes are sustained, however, or should the child have a
structurally abnormal heart, then further evaluation and therapy are necessary. Ventricular tachycardia can be the result of electrolyte abnormalities, myocarditis, arrhythmogenic right ventricular dysplasia, hamartomas, Purkinje cell tumors, other ventricular tumors, central nervous system disorders, postoperative cardiac disease, mitral valve prolapse, drug-induced ventricular tachycardia, long QT syndrome, coronary artery disease, or any systemic illness that results in anoxia or hypoxia.46

Finally, atrial fibrillation or atrial flutter may also present in childhood (Figure 5). Investigation for structural heart disease is required, focusing on those conditions associated with dilation or stretch of the atria, such as atrial septal defects, or following extensive atrial surgery, such as the older Mustard repair of transposition.

**Adjunctive Testing**

**Electrocardiogram.** All patients with palpitations should have an electrocardiogram performed to look for substrates for arrhythmia such as long QT syndrome or the presence of pre-excitation (Figures 6 and 7). The electrocardiogram can also identify hypertrophy of chambers or abnormal conduction intervals. It is important that pediatric electrocardiograms be performed properly and interpreted by those experienced with pediatric standards. The electrocardiograms of a 5-year-old, a 10-year-old, and an adult are dramatically different.

**Chest X-ray.** A chest x-ray is rarely of benefit in the evaluation of palpitations unless one is using it to identify those with known or suspected structural heart disease. Patients with cardiomyopathy may demonstrate an enlarged cardiac silhouette on the chest radiograph. However, chest x-rays have been shown to be quite inaccurate for the diagnosis of specific types of heart disease. Patients with palpitations, and particularly those without evidence of structural heart disease by the physical examination, will typically demonstrate a normal chest x-ray.

**Echocardiogram.** An electrocardiogram is useful when structural heart disease is suspected. Echocardiog-
raphy is a highly accurate tool for the diagnosis of cardiomyopathy in children, both dilated cardiomyopathy and hypertrophic cardiomyopathy. Patients with incessant tachyarrhythmias deserve an echocardiogram to evaluate myocardial performance, because incessant tachyarrhythmias can sometimes lead to dilated cardiomyopathy.

**Holter monitor.** Ambulatory electrocardiographic monitoring (Holter monitor) is useful for patients experiencing symptoms on a daily basis. In the absence of daily symptoms, a Holter monitor can document normal heart rate variation and identify clues to arrhythmia risk, such as abnormal conduction. Again, the test needs to be performed and interpreted by those experienced with pediatric norms. Children have a wide variety of normal heart rates. Heart rates of 50 beats per minute when sleeping and 180 or 190 beats per minute when exercising may be normal. This normal variation should not be misinterpreted as abnormal bradycardia or tachycardia.

**Event monitor.** Event monitoring is also beneficial, and there are of 3 basic types. The first is self-applied and a recording performed at the time of patient symptoms. It is useful for patients who are old enough to perform this test themselves and have symptoms that last at least several minutes in duration. After recording the event, the patient typically transtelephonically transmits the data to a receiving station for interpretation.

The second type of event monitor has a loop memory. The patient wears the monitor continuously, and  

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**FIG 5.** Rhythm strip demonstrates atrial flutter. Initially, typical “saw-tooth” flutter waves are difficult to identify. During transient higher grade atrioventricular nodal block, flutter waves are clearly identified.

**FIG 6.** Rhythm strip shows sinus bradycardia with prolonged QT interval.
when symptoms occur presses a recorder button. The monitor retains the data from a predetermined time period prior to the occurrence, during the occurrence of symptoms, and a predetermined period after the button has been pressed. These monitors are typically the size of a pager and are useful for patients with fleeting symptoms or those who have neurologic impairment of some type during the symptoms, which would prevent self-application of a monitor at the time of symptoms.

The third type of monitor is implanted. These monitors are surgically placed subcutaneously, typically in the pectoral area. Once an event occurs, the patient reports to his or her cardiologist to have the information transmitted. This is used for patients with significant neurologic symptomatology, rarely occurring events, or when the patient has been noncompliant with previous attempts at documentation.

**Stress test.** Graded exercise testing is another adjunctive test that is useful for patients whose symptoms are provoked by exercise.\(^{46,53}\) Using this approach, the patient’s cardiac rhythm is monitored in a controlled environment during strenuous exercise to document the type of rhythm that occurs.\(^{54}\)

**Therapy**

Once the correct diagnosis is determined, therapy can be initiated. It is important to withhold therapy until a definitive diagnosis is reached. Many patients reporting palpitations have normal chronotropic or inotropic variation in their heart rate. These patients sense this variation and feel anxious. Reassurance can be given to these families after the normal variation is demonstrated. Other patients may have arrhythmias of a benign nature. Those with structurally normal hearts and isolated premature atrial or premature ventricular contractions can also be given reassurance. Patients with arrhythmias secondary to other conditions should have those problems treated. For example, hyperthyroidism, anemia, and fever can result in sinus tachycardia but the tachycardia is not harmful to the patient and the underlying disease process must be treated. Finally, some patients will have an arrhythmogenic etiology to their palpitations. These patients require medical, surgical, or catheter-based radiofrequency therapy. Some patients benefit from a combination of therapies. A discussion of specific therapies for each rhythm problem is beyond the scope of this article but consultation with a pediatric cardiologist is essential for effective management.

**Summary**

The evaluation of palpitations begins with a complete history and physical examination. Important general principles have been discussed and are summarized:\(^{43}\):

- The description of the symptoms of palpitations varies depending on the patient’s age.
Documenting the event is essential before advancing a diagnosis or initiating treatment.

The urgency of an evaluation for palpitations depends on the symptoms experienced and the likelihood of structural heart disease.

Physical examination may be entirely normal, even in those with serious rhythm problems.

Adjunctive testing should be performed and interpreted by those familiar with pediatric norms.

Therapy is guided by the specific type of arrhythmia, associated symptomatology, presence of structural disease, and each patient’s unique clinical circumstances.

Athletic activities invariably place an increased demand on the cardiovascular system. To be able to participate safely in an athletic activity, one must be able to accommodate the hemodynamic demands imposed by the activity. The most useful classification of athletic activities with respect to the demands placed on the cardiovascular system divides exercise into 2 main categories, dynamic exercise and static exercise (Table 3). Dynamic exercise is characterized by large changes in muscle size and repetitive joint movements with little developed intramuscular force, whereas static exercise is characterized by increased intramuscular force with little change in muscle length. Many athletic activities share features of both dynamic and static exercise. This classification is believed to be more useful in the formation of participation guidelines than is the metabolic-based classification of sports as aerobic or anaerobic.

The cardiovascular adaptations to dynamic and static exercise are well characterized. In response to dynamic exercise one observes significant increases in oxygen consumption and heart rate, and a substantial increase in cardiac output. During dynamic exercise systolic blood pressure rises while diastolic blood pressure typically falls, reflecting a decrease in peripheral resistance. The heart of the competitive athlete who engages in a high dynamic exercise sport is characterized by an increase in chamber size and left ventricular mass, reflecting the volume load imposed by dynamic exercise. In contrast, static exercise results in little change in cardiac output and heart rate, and elevations of both systolic and diastolic blood pressures occur. The heart of the athlete who engages in primarily static exercise is characterized by an increase in left ventricular mass without increases in chamber size, reflecting the predominant pressure load of static exercise. Finally, it should be recognized that even the most “sedentary” of athletic activities, such as rillelry or billiards, might result in significant, transient elevations in heart rate or blood pressure, as a reflection of competition-related stress with resultant activation of the sympathetic nervous system.

Children Without Known Cardiovascular Disease

For the primary care physician, providing the child with known heart disease appropriate counseling for participation in athletic activities requires matching a known diagnosis with a set of reference guidelines.
and at times referral to a pediatric cardiologist. The apparently normal child without known heart disease presents a somewhat different challenge. While precise figures are difficult to come by, it is estimated that sudden unexpected cardiac death occurs in children (excluding SIDS) at a rate of 1.5 to 8 per 100,000 per year, and may be somewhat higher (1 in 50,000 to 1 in 100,000) in competitive athletes. A lower rate of sudden death, 1 per 200,000, was reported over a 12 years among high school athletes in Minnesota. It is estimated that 20 to 50 sudden cardiac deaths occur annually among high school and college athletes. Though still rare, when such a tragedy occurs it often results in widespread media coverage, probing questions of school officials and health care providers, and long-lasting anxiety and emotional trauma within the community. By examining the known causes of sudden unexpected cardiac death in children, and considering the accuracy and costs associated with various screening approaches for occult heart disease, a rational approach for the identification of “at risk” children and adolescents can be proposed.

In one of the most comprehensive series addressing sudden unexpected cardiac death in young competitive athletes, Maron et al identified the following causes in 158 cases (Table 4): hypertrophic cardiomyopathy and possible hypertrophic cardiomyopathy (46%), coronary artery anomalies (19%), rupture of the aorta as part of Marfan’s syndrome (5%), aortic stenosis (4%) and other causes, including myocarditis, mitral prolapse, arrhythmogenic right ventricular dysplasia, asthma, heat stroke, and arrhythmias. With the excep-

### TABLE 3. Classification of sports according to peak dynamic and static components during competition

<table>
<thead>
<tr>
<th></th>
<th>A. Low dynamic</th>
<th>B. Moderate dynamic</th>
<th>C. High dynamic</th>
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<tbody>
<tr>
<td>I. Low static</td>
<td>Billiards</td>
<td>Baseball</td>
<td>Badminton</td>
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<td></td>
<td>Bowling</td>
<td>Softball</td>
<td>Cross-country skiing (classic)</td>
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<td></td>
<td>Cricket</td>
<td>Table tennis</td>
<td>Field hockey</td>
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<td></td>
<td>Curling</td>
<td>Tennis (doubles)</td>
<td>Orienteering</td>
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<td>Golf</td>
<td>Volleyball</td>
<td>Race walking</td>
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<td>Rifery</td>
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<td>Racquetball</td>
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<td>Running (long distance)</td>
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<td></td>
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<td></td>
<td>Soccer†</td>
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<td></td>
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<td>Squash</td>
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<td></td>
<td></td>
<td></td>
<td>Tennis (singles)</td>
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<tr>
<td>II. Moderate static</td>
<td>Archery</td>
<td>Fencing</td>
<td>Basketball†</td>
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<td>Auto racing††</td>
<td>Field events (jumping)</td>
<td>Ice hockey</td>
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<tr>
<td></td>
<td>Diving††</td>
<td>Figure skating†</td>
<td>Cross-country skiing (skating)</td>
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<td></td>
<td>Equestrian††</td>
<td>Football (American)†</td>
<td>Football (Australian rules)†</td>
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<td></td>
<td>Motorcycling††</td>
<td>Rodeoing††</td>
<td>Lacrosse†</td>
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<td>Rugby†</td>
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<td>Running (sprint)</td>
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<td></td>
<td>Surfing†</td>
<td>Team handball</td>
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<td></td>
<td></td>
<td>Synchronized swimming†</td>
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<td>III. High static</td>
<td>Bobsledding††</td>
<td>Body building††</td>
<td>Boxing†</td>
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<td></td>
<td>Field events (throwing)</td>
<td>Downhill skiing††</td>
<td>Canoeing or kayaking</td>
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<td>Gymnastics††</td>
<td>Wrestling†</td>
<td>Cycling†</td>
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<td></td>
<td>Karate or judo*</td>
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<td>Decathlon</td>
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<td>Luge††</td>
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<td>Water skiing††</td>
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<td>Weight lifting††</td>
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<td></td>
<td>Windsurfing††</td>
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*Danger of bodily collision.
†Increased risk if syncope occurs.

### TABLE 4. Causes of sudden cardiac death in young athlete

<table>
<thead>
<tr>
<th>Disorder</th>
<th>%</th>
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<tr>
<td>Hypertrophic cardiomyopathy</td>
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<tr>
<td>(and possible hypertrophic cardiomyopathy)</td>
<td></td>
</tr>
<tr>
<td>Coronary artery anomalies</td>
<td>19</td>
</tr>
<tr>
<td>Aortic rupture (Marfan syndrome)</td>
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</tr>
<tr>
<td>Aortic stenosis</td>
<td>4</td>
</tr>
<tr>
<td>Other causes (myocarditis, mitral valve prolapse, arrhythmogenic)</td>
<td>&lt;3</td>
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</tbody>
</table>

tion of aortic rupture, heat stroke and possibly asthma, the final common pathway to sudden cardiac death in each condition is a lethal arrhythmia. The category of “arrhythmia” may include conditions such as Wolff-Parkinson-White syndrome and long QT syndrome, which represent primary electrical abnormalities in which electrophysiologic substrates for a lethal arrhythmia may exist in the absence of an underlying structural heart defect. Others such as coronary artery anomalies, and arrhythmogenic right ventricular dysplasia are conditions associated with structural defects that can be quite subtle and difficult to detect. Finally, geographic differences in the etiology of sudden cardiac death in young athletes have been reported. For example, in Italy arrhythmogenic right ventricular dysplasia, not hypertrophic cardiomyopathy, is the leading cause of sudden cardiac death in young athletes. Genetic differences, as well as the inclusion of an electrocardiogram as part of the routine screening of all student athletes in Italy, which may detect and thus exclude from athletics children with hypertrophic cardiomyopathy, may account for this geographic difference.

The Child With Known Cardiovascular Disease

The most recent, comprehensive and widely used guidelines to address the eligibility for participation in sports for children and adolescents with known cardiovascular disease are derived from the 26th Bethesda Conference. While death or disability directly resulting from participation in sports is rare, even in the child with a significant congenital or acquired heart defect, certain heart defects are recognized as being more highly associated with a risk for sudden death. These include various forms of left ventricular outflow obstruction, such as aortic valve stenosis, cardiomyopathies, congenital coronary artery anomalies, and Marfan’s syndrome with aortic root dilatation. An elevated risk for sudden death is also present postoperatively in children following repair of aortic stenosis, coarctation of the aorta, and complex congenital heart defects such as tetralogy of Fallot and transposition of the great arteries. With early repair of congenital heart defects becoming the standard, most children of school age will have had repair or palliation of their heart defect already performed. The risk of sudden death after repair of tetralogy of Fallot has been estimated to be approximately 1.5 children per 1000 patient-years, or 1 in 700 operations per year. While elevated right ventricular pressure, significant pulmonary insufficiency, older age of repair and QRS complex greater than 180 ms on electrocardiogram have been identified as potential risk factors for death, predicting level of risk in individual patients is exceptionally difficult. For children who have undergone the older atrial repairs of transposition of the great arteries (Mustard or Senning operations), the risk of sudden death may be as high as 4.9 children per 1000 patient-years, or 1 in 180 operations per year. With the arterial switch now the standard method of repair of transposition of the great arteries, it is expected that the incidence of postoperative sudden death in these patients will be significantly reduced. For both postoperative tetralogy of Fallot and (the atrial repair of) transposition of the great arteries, arrhythmia is the presumed mechanism for sudden cardiac death. The risk of sudden death for other complex heart defects such as double-outlet ventricle and single ventricle, may be of similar or greater magnitude, even after repair. Children with congenital heart disease associated with elevated pulmonary vascular resistance (pulmonary hypertension) are also at significantly elevated risk.

For the child with a known congenital heart defect, the 26th Bethesda Conference offers excellent guidance for basing decisions about eligibility for participation in athletic activities. In general, in the mild forms of the most common congenital heart defects (ventricular septal defect, atrial septal defects, patent ductus arteriosus, valvar pulmonic stenosis, aortic stenosis) most sports listed in Table 3 are permitted. In patients with intermediate forms of the common defects, specific evaluations are recommended to guide final decisions concerning allowable sports. In the most severe forms of these congenital heart defects strenuous athletic activity is usually prohibited. Thus, children with small atrial, or small to moderate ventricular septal defects or small patent ductus arteriosus may participate in all sports. However, in children with atrial septal defects or PDAs with significant pulmonary hypertension, or large ventricular septal defects, participation in sports may be limited to those in class IA in Table 3. For asymptomatic children with valvar pulmonic stenosis, those with peak systolic gradients of less than 50 mm Hg may participate in all competitive sports, while those with peak systolic gradients greater than 50 mm Hg are restricted to low intensity sports (Class IA). Because aortic valve ste-
nosis, particularly when severe, is strongly associated with a risk for sudden death, the recommendations with regards to sports participation for this lesion are more conservative. Any patient with aortic stenosis and exertional symptoms such as chest pain or syncope should be immediately excluded from athletic participation until a complete evaluation can be performed. The asymptomatic child with mild aortic stenosis (defined by the 26th Bethesda Conference as aortic stenosis with less than a 20 mm Hg peak gradient) may participate in all sports as long as the resting electrocardiogram is normal. Those with moderate aortic stenosis, defined as a peak gradient of 21 to 49 mm Hg, who are asymptomatic, have normal exercise stress test, and no greater than mild LVH on electrocardiogram, may participate in low static/low to moderate dynamic, and moderate static/low dynamic sports (classes IA and IB and IIA). Children with coarctation of the aorta with less than a 20 mm Hg gradient between upper and lower extremities, and whose systolic blood pressure with exercise testing remains less than 230 mm Hg at peak exercise are allowed to participate in all athletic activities, although the avoidance of high-intensity chest impact has been recommended by some to reduce the risk of aortic dissection.

Recommendations for athletic participation in postoperative patients are also addressed in the 26th Bethesda Conference report. In general, 6 months after repair of atrial septal defects or ventricular septal defect and 3 months after repair of PDA, if the child is asymptomatic and if there is no evidence of pulmonary hypertension or arrhythmia, and no more than a small residual shunt, participation in all sports is permitted. In the case of children who are post-repair of tetralogy of Fallot, decisions concerning athletic participation generally require a careful evaluation that includes a physical examination, chest x-ray, electrocardiogram, 24-hour electrocardiogram monitoring, and an exercise stress test. An echocardiogram and at times a postoperative cardiac catheterization are often included in the evaluation. Patients who are found to have an excellent repair, with normal or near normal right-heart pressures, only mild RV volume overload (from pulmonary or tricuspid insufficiency), no significant residual shunt and no rhythm abnormality on 24-hour electrocardiogram monitoring or exercise stress testing may be allowed to participate in all competitive sport activities. Similarly, in asymptomatic children who are postatrial repair for transposition of the great arteries, unrestricted athletic participation is permitted in those with no cardiac enlargement on chest x-ray, no history of atrial flutter or ventricular arrhythmia, no history of syncope, and a normal exercise stress test. All other patients require careful evaluation and individualized consideration prior to formulating a recommendation for participation in athletics. For the increasing number of patients who have undergone the Fontan operation for single ventricle complex, in general only class IA athletics are permitted. Specific recommendations for children with other congenital heart defects, including unoperated cyanotic heart disease, Marfan’s syndrome, and coronary artery anomalies (discussed later in this article), are also contained in the 26th Bethesda Conference report.

Children with significant systemic hypertension are believed potentially to be at increased risk for complications including cerebral vascular accidents with exercise. To address this concern the 26th Bethesda Conference set forth recommendations concerning the eligibility for sports participation of children and adolescents with mild, moderate, severe and very severe degrees of systemic hypertension. These guidelines were subsequently revised by the Committee on Sports Medicine and Fitness of the American Academy of Pediatrics in 1997 to more closely link the definition of severe hypertension in children greater than 12 years of age with blood pressure values reported in the Second Task Force on Blood Pressure Control in Children. According to the 1997 AAP recommendations, children and adolescents with no more than ‘significant’ hypertension (136-143/86-91 in a 13-15 year old), in the absence of evidence of target organ damage, may be allowed full participation in athletics without restriction. Children whose hypertension meets the criteria of “severe” hypertension (greater than 143/91 in a 13-15 year old) should be restricted from exercises that are highly static (classes IIIA, IIIB, and IIIC) until their hypertension is controlled, and there is no evidence of end organ damage. Children with common, benign rhythm disturbances may participate in athletic activities without restriction. These include asymptomatic children with isolated premature atrial premature beats and children with premature ventricular contractions who by history and physical examination have a normal heart. For children with premature ventricular contractions, some recommend either a formal exercise stress test, or an informal office-based “exercise test,” to docu-
ment that the premature ventricular contractions do not increase with exercise, and ideally are suppressed at high heart rates. Similarly, asymptomatic children (without structural heart defects) with sinus bradycardia, junctional escape beats, or sinus pauses of less than 3 seconds, or mild first degree atrioventricular block are allowed full athletic participation. For athletes with a history of supraventricular tachycardia, asymptomatic patients with exercise-induced supraventricular tachycardia that is well controlled by medical therapy, or who have sporadic episodes of tachycardia that appear suppressed by therapy, may participate in all athletic activities.

There are cardiac conditions associated with hemo-
dynamically significant and potentially lethal arrhyth-
mias for which clearance for participation in athletics should be withheld by the primary care physician, pending evaluation by a pediatric cardiologist. These include children with ventricular pre-excitation (Wolff-Parkinson-White Syndrome), congenital heart block, congenital prolonged QT syndrome, hypertro-
phic cardiomyopathy, dilated cardiomyopathy, myo-
carditis, and those with documented episodes of sus-
tained, or nonsustained ventricular tachycardia. Evaluation by the pediatric cardiologist may include 24-hour electrocardiogram monitoring, exercise stress testing, echocardiography, cardiac catheterization, MRI imaging and, in some instances, intracardiac electrophysiologic testing. Final recommendations may then be formulated. Complete guidelines for athletic participation for children with cardiac rhythm disturbances are contained in the 26th Bethesda Con-
ference. 62

Conditions Associated With Structural Heart Disease

The conditions described below (as well as aortic stenosis, dilated cardiomyopathy, and myocarditis) share in common the presence of an anatomic or functional abnormality of the heart. Hypertrophic car-
diomyopathy is the leading cause of sudden cardiac death in young athletes in the United States. This disorder is characterized by left ventricular hypertrophy that is the result of dominantly inherited gene mutations of elements of the cardiac sarcomere, that is, the contractile proteins. Mutations in the myosin heavy chain, troponin T, and myosin binding protein account for almost two thirds of all cases. 69 Cardiac hypertrophy in this disorder typically involves the interventricular septum, but may be more concentric or even confined to apical portions of the left ventricle. Hypertrophy of the interventricular septum may be associated with varying degrees of left ventricular outflow tract obstruction, which may be manifest on physical examination as a harsh systolic murmur along the left sternal border. In other patients, minimal outflow tract obstruction occurs and the disorder may be difficult to detect simply by auscultation. The prevalence of hypertrophic cardiomyopathy may be as high as 1 in 500 adults. The true prevalence in the pediatric age group is not known, but it has been identified even in young infants. It is now recognized that, in addition to patients who have marked, life-
threatening symptoms due to outflow tract obstruction, ischemia, and arrhythmias, a significant number of patients are asymptomatic and have a relatively benign clinical course. A family history of hypertrophic cardiomyopathy or unexplained sudden cardiac death in a young family member should alert the clinician to this diagnosis. Of equal importance, the child should be questioned concerning exertional chest pain or syn-
cope, and exercise intolerance. As mentioned earlier, a murmur may or may not be appreciated, depending on the degree of left ventricular outflow obstruction. The electrocardiogram is abnormal in over 90% of infants and children with hypertrophic cardiomyopathy, with left ventricular hypertrophy and abnormal ST-T waves being evident in most. The definitive diagnosis is made by echocardiography, demonstrating either segmental or global left ventricular hypertrophy (Figure 8). Mortality rates reported for hypertrophic cardio-
myopathy range from 0% to 50%.

In a recent report of 99 pediatric patients with hypertrophic cardiomyopathy, sudden death was noted in 18 patients over a follow-up period of 5 years, for an overall death rate of 2.7% after age 8. 70 In this series, predictors of sudden death included ventricular tachycardia on 24-hour electrocardiogram recording, myocardial bridging of the left anterior descending coronary artery, and increased QT dispersion on the 12-lead electrocardiogram. In contrast to other reports in the pediatric age group, age at diagnosis, family history, and symptoms were not predictive of sudden death in this series. For any child with known or suspected hypertrophic cardiomyopathy, participation in competitive or strenuous athletic activities is for-
bidden. Treatments available include medical therapy with β-blockers and, for those who remain symptom-
atic from outflow obstruction, surgical excision of the
obstructing hypertrophied septum. Other therapies include implantation of an automatic internal cardioverter-defibrillator for those with refractory ventricular arrhythmias, and cardiac transplantation.

Coronary artery anomalies are the second leading cause of sudden cardiac death in the young athlete. (Table 4) These anomalies include origin of either the left or right coronary artery from the opposing aortic sinus, with a course that “wedges” the aberrant coronary artery between the aorta and pulmonary artery (Figure 9).71 These are the most common coronary anomalies encountered in the context of sudden death in the competitive athlete. Other anomalies, such as coronary ostial stenosis and aberrant origin of the left anterior descending coronary artery from the pulmonary artery are less common, or present earlier in life. The prevalence of significant coronary artery anomalies ranges from 0.17% in autopsy series, to as high as 1.2% by angiographic series.72 In a recent report, 4 cases of anomalous origin of the coronary arteries were detected out of 2,388 children (0.17%) undergoing echocardiographic study for various indications.72 The mechanism of sudden death in children and adolescents with coronary artery anomalies is presumed to be exercised induced ischemia with or without regional infarction, and resultant ventricular arrhythmia. The physical examination of the child athlete with an aberrant origin of a coronary artery will typically be normal, however, upon close questioning as many as 40% may report exertional chest pain or syncope, which should serve to alert the clinician to the possibility of a coronary artery anomaly.71 The utility of the routine electrocardiogram, exercise stress testing, and echocardiographic screening for wall motion abnormalities in the evaluation of patients with suspected anomalous origin of the coronary arteries has been questioned, because in 1 series these tests were normal in all patients with anomalous coronary origins in whom the tests were performed.71 When anomalous origin of a coronary artery is suspected because of a history of recurrent or exertional syncope or angina, definitive diagnosis should be aggressively pursued. This requires imaging of the coronary arteries either by echocardiography, MRI, or, if necessary, coronary artery angiography. Surgical correction of the anomaly represents the definitive treatment.

Marfan’s syndrome with aortic rupture represents the third leading cause of sudden death in the competitive athlete. Phenotype can vary greatly in Marfan’s syndrome, with some children and adolescents being easily diagnosed by physical examination (pectus, scoliosis, typical arm length to height ratio), whereas in others the diagnosis may be more difficult. A careful family history may be useful when considering the diagnosis of Marfan’s syndrome in the “tall” athlete without obvious stigmata. Evaluation by a geneticist, cardiologist, and ophthalmologist can be helpful in establishing the diagnosis. Most children...
and adolescents with Marfan’s syndrome (30%-60%) have accompanying cardiac abnormalities, including aortic root dilatation, aortic insufficiency, aortic root aneurysms, mitral valve prolapse, and mitral insufficiency (Figure 10). While echocardiographic screening of “tall” athletes, for example, basketball players, for aortic root dilation has been advocated by some, this approach has not been adopted in the United States. Sports participation for patients with Marfan’s syndrome is guided by the extent of cardiac disease, though activities associated with chest collision should probably be avoided.

As described earlier, arrhythmogenic right ventricular dysplasia is the leading cause of sudden cardiac death in competitive athletes in Italy. This entity is characterized by fatty and fibrous infiltration of the right ventricular myocardium that provides an anatomic and electrical substrate for the development of ventricular arrhythmias. Ventricular tachycardia in these patients is often exercise-induced, and has a typical left bundle branch morphology on electrocardiogram. Therefore, a history of exercise-induced syncope, lightheadedness, or palpitations may be elicited. A history of syncope may be elicited in 23.5% of children with arrhythmogenic right ventricular dysplasia. The resting electrocardiogram typically shows abnormal T wave inversions in the precordial leads. The entity may be dominantly inherited, and has been linked to multiple chromosomal loci, including chromosomes 1, 2, 3, 10 and 14. A careful family history looking for young persons with ventricular arrhythmia or sudden death, therefore, may alert one to the diagnosis. In severely affected patients, the diagnosis can be based on demonstration of abnormalities of right ventricular wall motion by echocardiography or by cineangiography. In many cases, however, the degree of fibro-fatty infiltration is subter and MRI or biopsy may be required to establish the diagnosis. Arrhythmogenic right ventricular dysplasia may be the underlying etiology in over 30% of cases of ventricular arrhythmia and a seemingly “normal” heart, as assessed by echocardiography. Treatment may consist of medical therapy (sotalol preferred), and radiofrequency catheter or surgical ablation of the arrhythmogenic substrate. Implantation of an automatic internal cardioverter-defibrillator is also an option.

**Conditions Without Structural Heart Disease**

Causes of sudden cardiac death in the young athlete where no structural heart defect or functional abnormality is present include children with arrhythmias associated with entities such as Wolff-Parkinson-White syndrome and long QT syndrome, as well as lethal arrhythmias induced on the athletic field in association with chest trauma, so-called commotio cordis.

The Wolff-Parkinson-White syndrome (presence of an accessory atrioventricular connection) is a very common substrate for supraventricular tachycardia in infants and children. The classical electrocardiogram pattern of the Wolff-Parkinson-White syndrome (short PR interval, wide QRS complex, delta wave) may be present in up to 0.1% of the population (Figure 7). The spectrum of symptoms varies greatly with many individuals remaining symptom-free, some experiencing palpitations or well-tolerated episodes of supraventricular tachycardia, and a few experiencing life-threatening arrhythmias. Sudden death in association with the Wolff-Parkinson-White syndrome is uncommon, estimated to occur in 1.5 per 1000 patient-years. Death is thought to occur secondary to ventricular fibrillation, initiated in response to rapid conduction of rapid atrial rhythms, such as atrial fibrillation, to the ventricles over the accessory connection. While rare, sudden death or near sudden death episodes have been described in children with the Wolff-Parkinson-White syndrome, including sudden death as the initial clinical presentation. Treatment of children with symptomatic Wolff-Parkinson-White syndrome includes antiarrhythmic therapy and, increasingly common, radiofrequency catheter ablation of the accessory connection, which can be performed in children and
adolescents with a very high rate of success and high degree of safety. 78

Long QT syndrome should be considered in any child or adolescent with a history of unexplained, recurrent, seizures or syncope, particularly when these symptoms are related to activity. A family history of long QT syndrome or sudden unexplained cardiac death in young family members should alert the pediatrician to this diagnosis. It may be as common as 1 in 7000 children. 64 The disorder is the result of cardiac ion channel gene mutations (potassium or sodium ion channel mutations 75) that cause prolongation of the action potential plateau, a delay in the repolarization process, and a propensity to develop life-threatening, polymorphic ventricular tachycardia. Sudden death associated with long QT syndrome occurred in 8% of children followed for 5 years in 1 series. 79 The diagnosis is readily made from the electrocardiogram (Figure 6) when the calculated corrected QT interval (QT interval divided by the square root of the RR interval) exceeds the upper limit of normal of 0.45 seconds. Genetic testing may become more widely available to assist in the diagnosis of suspected patients or family members when QT interval prolongation on electrocardiogram is borderline. Treatment consists of β-blockade, and, in patients in whom symptoms persist, implantation of an automatic internal cardioverter defibrillator.

Sudden cardiac death following a blow to the chest wall has become known as commotio cordis. Although rare in children, it is most commonly observed during baseball or hockey games. 80 Collapse occurs immediately following impact. Resuscitation efforts are often unsuccessful. It has been recently shown that blunt chest trauma, delivered at a critical phase of the cardiac cycle, can induce ventricular tachycardia and ventricular fibrillation. 81

Screening Children and Adolescents for Potentially Life-Threatening Cardiovascular Abnormalities

Many of the conditions associated with sudden death in the young athlete can be detected by electrocardiogram (hypertrophic cardiomyopathy, long QT syndrome, Wolff-Parkinson-White syndrome), or by echocardiography (hypertrophic or dilated cardiomyopathy, aortic root dilatation). As discussed earlier, mass electrocardiogram screening of student athletes in the Veneto region of Italy has resulted in an apparent reduction in the incidence of sudden cardiac death from hypertrophic cardiomyopathy among Italian athletes. 61 While a few US-based studies do report cost-effective detection of “at risk” athletes through universal electrocardiogram or limited-view echocardiographic screening programs, 82–84 other studies do not support this approach. 85 In spite of this, in the United States the routine screening of all athletes by electrocardiogram or echocardiography is currently considered impractical, expensive, and has not been proven to prevent sudden death. The cost of detecting a single case of hypertrophic cardiomyopathy by routine echocardiographic screening has been estimated to be between $200,000 and $250,000, and screening for life-threatening conditions by electrocardiogram, $44,000 per detected case. (57,82) Furthermore, consideration of the costs and burdens associated with false-positive results, and the sheer magnitude of resources required to provide nationwide screening, make a universal screening approach based on noninvasive testing impractical.

In 1996 a consensus statement was issued by the American Heart Association, and endorsed by the American Academy of Pediatrics Section on Cardiology, detailing the critical components of cardiovascular screening of young competitive athletes. 86 The currently recommended and most cost-effective approach for screening children and adolescents for potentially life-threatening cardiovascular abnormalities is through a careful personal history, a focused family history, and a physical examination. Cardiovascular screening is recommended for all athletes, and should be performed prior to participation in high school and college sports. For high school students, screening is recommended every 2 years, with an interim history obtained each year. A careful history is of paramount importance. Of particular concern to the pediatrician should be children who report exertional angina, exertional syncope, near syncope, excessive fatigue, or shortness of breath. These exertional symptoms should not be confused with the very common and quite benign sharp intermittent chest pain that occurs primarily at rest, or simple vasovagal syncope, both of which occur in many children and adolescents. The history should also include a past diagnosis of either hypertension or a heart murmur. The family history should include questions of premature death or disability from cardiovascular disease in close relatives younger than 50 years of age, or known family members with any of the conditions described above,
such as long QT syndrome, hypertrophic cardiomyopathy, or Marfan’s syndrome. The focus of the physical examination should include checking for stigmata of Marfan’s syndrome, measurement of blood pressure, palpation of femoral pulses (to exclude coarctation of the aorta), and auscultation of the heart for a murmur. A positive finding during the cardiovascular screen should prompt a referral to a pediatric cardiologist for further evaluation. If such a referral is not practical, obtaining an electrocardiogram to screen for left ventricular hypertrophy, Wolff-Parkinson-White syndrome or long QT syndrome, and an echocardiogram may provide some guidance. It should again be emphasized that the echocardiographic diagnosis of entities such as anomalous origin of the coronary arteries or arrhythmogenic right ventricular dysplasia is quite specialized and technically challenging, and, therefore, whenever possible, evaluation by a pediatric cardiologist is recommended.

Finally, community-based approaches to aid in the prevention of sudden cardiac death on the athletic field have been proposed. These include education of the public concerning sudden cardiac death, the more widespread teaching of CPR, and an increase in the public awareness and access to automatic external defibrillators during high school and college athletic events. The use of chest protectors to prevent commotio cordis has also been proposed.

Insurance and Employment in Adolescents and Young Adults With Congenital Heart Disease

It is a testament to the benign course of the most common congenital heart defects and the improving survival of those with even the more complex defects that the problems of insurability of these adolescents and young adults have come to the forefront. There are at least half a million adults living with congenital heart disease in the United States, a number that is growing by more than 20,000 per year. The fact that most of these adults are not severely disabled and are capable of working does not appear to improve their access to health insurance, which is frequently denied, limited, or set with prohibitive premiums that may exceed standard premiums by as much as 50%. For those with insurance, lifetime caps on coverage, underinsurance, and disapproval of an ever-broadening list of certain services by managed care organiza-

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of 40 years compared to 85% for the general population of women.

- The frequency of marriage and rates of divorce and separation for the male cohort was similar to the general population of men.

This NHS is especially pertinent in this discussion because the vast majority of young adults with the label of “congenital heart disease” will have (or have had) an atrial, ventricular septal defect, or stenosis of the pulmonary or aortic valve, lesions which are the commonest. The other end of the spectrum, a minority, albeit a growing one, will include patients with varying degrees of palliated complex heart disease, such as tetralogy of Fallot, transposition of the great arteries, and various anatomic substrates with a single ventricle physiology. These are patients whose need for continued consumption of medical and surgical services should be expected to be higher than those of the general population and will have, therefore, higher medical and health insurance expenses.

Fewer than 10% of adults with congenital heart disease are considered disabled,91 yet their ability to secure gainful employment is often markedly hindered by their diagnosis even in the absence of any residual cardiac defects. Many employers balk at hiring such adults because of often ill-informed concerns about frequent absenteeism due to illness, an exaggerated sense of the potential employee’s frailty and subsequent ability to perform, and the real “dollar and sense” worry about increasing health care costs borne by the employer. Even a minor disability may have a negative impact on employability.92 Although the Americans with Disabilities Act of 1990 prohibits discrimination in employment and makes it unlawful to question applicants about their disabilities unless it is prudent to their ability to perform the job, the fact remains that the full protection of the law is something the young adult with congenital heart disease can find extremely difficult to achieve.

There is an ongoing crisis in American health care, which is heightened, in the vulnerable populations such as adults with congenital heart disease. Inflationary pressures created by an ever-increasing demand for services and rapidly evolving but costly technological advances will remain with us in the foreseeable future. In the interim, health insurance companies continue to adopt more restrictive measures and impose higher premiums to offset expenses. Employers in turn avoid hiring those with preexisting conditions in an attempt to keep their health care costs manageable.

Life insurance is another matter altogether. In 1 study, which polled insurance companies to determine which cardiac conditions were considered insurable, there was a marked disparity from company to company for no discernable reason. The factors considered by the companies in making their decisions were often not provided. Some conditions such as transposition of the great arteries and ventricular septal defects with pulmonary hypertension were considered uninsurable.93

### Pregnancy, Delivery, and Contraception in the Young Adult With Congenital Heart Disease

As in other aspects of the medical care of the patient with congenital heart disease, a multidisciplinary approach involving an appropriately trained pediatric or adult cardiologist, obstetrician, perinatologist, geneticist, and anesthesiologist is necessary to provide optimal care to these patients. Ideally, a pre-pregnancy evaluation should be performed to assess the severity of the patient’s heart disease and allow discussion between the medical team and the patient concerning the anticipated impact of pregnancy induced hemodynamic changes on the patient’s condition. The maternal risk depends on the type and severity of the heart lesion, ventricular function, presence of pulmonary hypertension, and the patient’s exercise capacity. The same factors that determine pregnancy outcome in heart disease in general apply to this group of patients with congenital heart disease. Severe systemic ventricular dysfunction, significant oxygen desaturation, and pulmonary hypertension are associated with high morbidity and mortality for the mother and fetus.94,95 A thorough prepregnancy evaluation of all patients with complex congenital heart disease (preoperatively or postoperatively) should therefore include cardiac catheterization. A review of the patient’s medications for potential teratogens, such as angiotensin converting enzyme inhibitors, which may cause renal dysplasia in the fetus, or other possible toxicities should be carried out prior to a planned pregnancy. Finally, genetic counseling with the patient and the infant’s father is often appreciated by the patient. Both men and women are concerned about the risk of congenital heart disease in their offspring.
Genetics

In the general population, the risk of congenital heart disease is 0.4% to 0.6%. If the mother is affected, the average risk is 6.7% (range 2.5% to 18%). If the father is affected, the risk is 2.1% (range 1.5% to 3.0%). If 1 offspring is affected, the risk of having a second child with congenital heart disease is 2.3% (range 1.5% to 5%). If 2 offspring are affected, the risk of having a third child with congenital heart disease rises to 7.3% (range 5% to 10%).

The risks are higher when there is a chromosomal anomaly; and some conditions such as velocardiofacial syndrome show an autosomal dominant inheritance.

Physiology

Plasma volume begins to rise in the first trimester, leveling off in the second trimester. The red blood cell count also rises (20%), but to a lesser extent than the plasma volume which almost doubles. The entire blood volume increases by 40% to 50% reaching a maximum by 30 to 32 weeks of gestation. In addition, the heart rate increases by 10% to 20%, contributing to an increase in the cardiac output, which rises by approximately 30%. Concomitantly, there is a significant fall in peripheral vascular resistance and the venous pressure in the lower extremities, which is why up to 80% of healthy pregnant women develop pedal edema. The pregnant woman therefore has a hyperdynamic cardiovascular physiology. Because of the physiologic decrease in peripheral vascular resistance in pregnancy, obstructive cardiac lesions, such as aortic stenosis, are not well-tolerated as their gradients increase, whereas regurgitant lesions, such as mitral regurgitation, are well-tolerated because of the decrease in afterload.

The majority of women with congenital heart disease who become pregnant have, or have had, simple lesions, are acyanotic, and have normal systemic ventricular function and no pulmonary hypertension. The fetal and maternal outcome in these patients is favorable with no significant difference in labor outcome, or fetal or maternal morbidity and mortality, although the fetuses tended to be smaller. In the pregnant woman with high-risk heart disease, maternal and fetal outcomes are significantly worse. Women with cyanotic heart disease have high fetal loss, with up to a 45% chance of spontaneous abortion when the oxygen saturation is below 82%. As maternal cyanosis worsens, as reflected by an increasing hemoglobin concentration, there is a near linear increase in the incidence of spontaneous abortion, and fetal growth retardation is worsened. When the hemoglobin is higher than 18g/dl, the chance of infant survival becomes remote. Certain cardiac lesions are associated with such a high risk of maternal and fetal morbidity and mortality that pregnancy termination is recommended. Marfan’s syndrome with a dilated aortic root greater than 40 mm is at increased risk for dissection and rupture during pregnancy. Patients with pulmonary vascular obstructive disease (Eisenmenger physiology) have a 50% mortality with pregnancy, and termination of pregnancy, especially in the first trimester, is a safer option. Women with symptomatic obstructive lesions, such as aortic and pulmonary stenosis, fare poorly in pregnancy, because the afterload reduction exaggerates their obstructive gradient. Patients with symptomatic ventricular dysfunction tend to deteriorate during pregnancy as the volume exacts a hemodynamic burden.

Labor and Delivery

There are rapid changes in hemodynamics during labor. With each uterine contraction, 300 to 500 mL of blood is pumped into the circulation, with progressively increasing cardiac output. Following delivery, with the relief of IVC obstruction by the fetus, there is an increase in venous return to the heart. The immediate postpartum period is especially fraught with danger for patients with Eisenmenger syndrome and pulmonary hypertension, and those with hereditary long QT syndrome. Recent studies show a high maternal mortality rate primarily in the postpartum period for patients with Eisenmenger syndrome and exclusively in the postpartum period in those with hereditary long QT syndrome. Hemodynamics return to baseline approximately 4 to 6 weeks after delivery. In the vast majority of patients with congenital heart disease, vaginal delivery is safer unless there is an obstetrical complication; blood loss is significantly greater with a caesarian section. The management of labor and delivery in the patient with a moderate to high risk cardiac lesion is very complex, and requires a multidisciplinary approach to minimize maternal stress, shorten the second stage of labor, and provide intensive monitoring with judicious use of anesthesia. Routine antibiotic prophylaxis is not usually recommended for labor and delivery unless infection is suspected or in high-risk patients with pros-
thetic valves, previous history of endocarditis, complex congenital heart disease, or surgically created systemic to pulmonary conduits.

Contraception

The prevention of pregnancy is desired by patients with congenital heart disease for diverse reasons that are similar to the general population and is the best strategy for those with high-risk cardiac disease. Low estrogen contraceptives are effective and safe except in patients with right-to-left shunting, polycythemia, or a prior thromboembolic event where there is an increased risk of thromboembolism. Estrogen may also exacerbate pulmonary hypertension. In those patients with significant left ventricular dysfunction (ejection fraction < 30%), there is an increased risk of thrombus formation that may be aggravated by use of estrogen containing contraceptive. Medroxyprogesterone acetate (Depo-Provera) and levonorgestrel implant (Norplant) are the preferred methods of contraception, because they are long-lasting and have a higher compliance. Progesterone-only contraceptive pills are less effective and do not prevent ovulation. Intrauterine devices are usually avoided because of the risk of infection. Sterilization (tubal ligation) is usually recommended for the very high risk patient where pregnancy would be life-threatening, but is not recommended for the patient under the age of 21 years.

Summary of Key Points

● Innocent heart murmurs are quite common in children. Most physicians caring for children are “comfortable” diagnosing innocent murmurs. If there is any doubt about the innocent nature of a heart murmur, or associated symptoms of concern, referral to a pediatric cardiologist is the preferred route for further evaluation.

● Chest pain is quite common in children, and rarely signifies a serious underlying cardiac condition. Intermittent sharp, stabbing chest pain is particularly common (precordial catch syndrome), is benign, and requires only reassurance. Laboratory evaluation of chest pain in children should be guided by clinical picture and examination. Thus, a routine chest x-ray is not necessary for every child, but would be indicated in the child with chest pain, fever, and cough. Exertional chest pain should be evaluated by a cardiologist.

● Palpitations in children are most often associated with non-life-threatening events, such as sinus tachycardia or isolated extrasystoles. Children who report lightheadedness, syncope or near syncope, or exertional symptoms, should be evaluated by a cardiologist and monitored to document the nature of the underlying cardiac rhythm.

● Children with known congenital heart disease, either repaired or un repaired, may participate in athletic activities as guided by the recommendations of the 26th Bethesda Conference. Children with mild forms of congenital heart disease, or those with few hemodynamic residua following surgical repair may participate in most athletic activities.

● Children without known cardiovascular disease often request of their primary care physician “clearance” to participate in athletic activities. The pre-participatory screening for these children includes a careful history and family history, looking for exertional chest pain, syncope, or fatigue, or a family history of sudden cardiac death in persons younger than 50 years, or a known inheritable syndrome such as Marfan syndrome, and a focused cardiovascular examination. Routine laboratory testing (electrocardiogram, echocardiogram) is not recommended for pre-participatory screening of all children.

● Most children with congenital heart disease grow to become highly productive and functional adults. Although at times they may encounter difficulty in obtaining insurance, the majority are, in fact, in excellent health. Most young women with congenital heart disease are able to become pregnant and deliver normal newborns. A small number, such as those with persistent cyanosis, or pulmonary hypertension, are considered at high risk and require careful multidisciplinary approaches to issues such as employability, pregnancy, and contraception.

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