AHA Medical/Scientific Statement
Special Report

Guidelines for Evaluation and Management of Common Congenital Cardiac Problems in Infants, Children, and Adolescents

A Statement for Healthcare Professionals From the Committee on Congenital Cardiac Defects of the Council on Cardiovascular Disease in the Young, American Heart Association

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Appropriate use of diagnostic and therapeutic techniques is important to ensure optimum and efficient treatment of patients in a cost-effective manner. Ideally, guidelines for appropriate, cost-effective care of patients should be based on outcome studies of management strategies. In the absence of reliable outcome studies, the conventional wisdom of experienced pediatric cardiologists can be used to develop guidelines. However, such guidelines must be subjected to review when outcome studies become available. In addition, the guidelines should change as diagnostic and therapeutic technologies change.

Approximately 50 years of experience in evaluation of patients with congenital heart defects has accumulated. Outcome analyses are now available for several conditions and defects, and others are being published. Knowledge of pediatric cardiology is sufficient to allow the development of diagnostic and treatment guidelines for relatively simple but common problems. Recognizing that critical analysis of diagnostic and treatment practices improves patient care and could reduce the cost of medical care, the Committee on Congenital Cardiac Defects of the American Heart Association Council on Cardiovascular Disease in the Young developed the following sets of management and treatment guidelines.

These guidelines are designed for use by board-eligible/certified pediatric cardiologists (hereafter referred to as pediatric cardiologists) and others who have a specialized cognitive knowledge of the clinical and natural history of congenital heart defects. Patients for whom these guidelines have been designed should be screened before referral to a pediatric cardiologist. This statement represents recommended guidelines to supplement practitioners in their clinical judgment and is not intended as a standard of care for all cases.

These guidelines are restricted to relatively simple, uncomplicated problems in infants, children, and adolescents. This is because 1) there are more patients with these problems than those with complex defects, and 2) the diversity of the natural history and diagnostic and therapeutic problems associated with complex defects preclude development of widely applicable useful guidelines.

The following guidelines are the consensus of committee members and consultants to the committee guided by results of published outcome studies or, when not available, by current practice patterns.

Construction of Algorithms

The algorithms were constructed over 3 years during several meetings of the Committee on Congenital Cardiac Defects of the Council on Cardiovascular Disease in the Young. Ten board-certified pediatric cardiologists and Kathryn Taubert, PhD, senior science consultant at the AHA, were members of the committee during the construction of these algorithms. One of the 10 cardiologists, Howard Gutgesell, MD, represented the cardiology subsection of the American Academy of Pediatrics. Another pediatric cardiologist, Arthur Garson, Jr, MD, represented the American College of Cardiology. Input was also obtained from a cardiovascular surgeon, Gordon Danielson, MD, and an internist cardiologist, David Skorton, MD.

The algorithms were reviewed and approved by the appropriate scientific committees of the AHA.
Classification of Testing Procedures

Class I: Conditions for which (or patients for whom) there is general agreement that a specific evaluation or test is useful and reliable. In this report, all evaluations, procedures, and tests are Class I unless stated otherwise.

Class II: Conditions for which (or patients for whom) there is a divergence of opinion with respect to the usefulness of an evaluation or test.

Definition of Defects, Exclusion Criteria, and Annotation of Algorithms

Asymptomatic Child with a Systolic Murmur (“Innocent Murmurs of Childhood”) (Fig 1)

Exclusion Criteria

1. The patient is 2 years old or younger.
2. The patient is symptomatic.
3. The patient is uncooperative during the examination.

Definition of Type of Murmurs

1. Still's murmur: a low-frequency systolic murmur less than or equal to grade 3/6 in intensity; heard best along left sternal border; normal second heart sound; no click; no diastolic murmur.
2. Pulmonic flow murmur: a mid-frequency murmur less than or equal to grade 3/6 in intensity; heard best at upper left sternal border; normal second heart sound; no click; no diastolic murmur.
3. Venous hum: a high-frequency murmur of grade 3/6 or less in intensity; best heard in sitting or standing position; best heard at base of the neck or the infraclavicular or supraclavicular area; normal second heart sound; systolic and/or diastolic in timing; abolished by compression of jugular vein, change of head position, or assumption of supine position.
4. Carotid bruit: grade 3/6 or less in intensity; heard over carotid artery; no associated precordial ejection click; no clinical evidence for aortic stenosis; no diastolic murmur; normal second heart sound.

Algorithm for Asymptomatic Child With a Systolic Murmur

This algorithm is intended to address the evaluation of the innocent murmur of childhood. Since “Time 0” is the initial evaluation by a pediatric cardiologist, the murmur will not have been classified as innocent. Hence, the algorithm is directed at the evaluation of a systolic murmur in an asymptomatic child 2 years of age or older. While many pediatric cardiologists are comfortable diagnosing an innocent murmur on the basis of the history and physical examination alone, some pediatric cardiologists obtain a chest radiogram and/or electrocardiogram. Thus, an ECG and a chest radiogram have been designated as Class II tests at the time of the initial evaluation. Because there are cases for which the history and physical findings are atypical for an innocent murmur, the algorithms provides for echocardiography* in selected cases. Once a murmur is determined to be innocent, further cardiac evaluation is unnecessary.

Patent Ductus Arteriosus (Figs 2 and 3)

Exclusion Criteria

1. The patient has associated cardiovascular anomalies.
2. The patient has symptoms or signs of congestive heart failure.
3. The patient was born prematurely.
4. The patient has pulmonary vascular obstructive disease.

Algorithms for Patent Ductus Arteriosus

Two algorithms are provided for the management of patent ductus arteriosus. For the first, “Time 0” is the
initial evaluation by a pediatric cardiologist and the patient is less than 6 months old, and for the second the patient is 6 months old or older at the time of that evaluation. The algorithms provide for an initial history, physical examination, electrocardiogram, and echocardiogram. In patients with a patent ductus arteriosus that has been documented echocardiographically, surgical closure of the defect can be done without additional echocardiographic studies if the physical findings remain typical for an uncomplicated patent ductus arteriosus. If the findings become atypical, one additional echocardiographic study is considered necessary. The algorithm provides for elective closure of the patent ductus arteriosus sometime within the first year of life.

After surgery, successful ligation/division of the ductus arteriosus can be documented by physical examination. The algorithm provides for a posthospital outpatient evaluation, usually within 3 months of surgery. A chest radiogram may be necessary to exclude persistent perioperative problems. Echocardiography to confirm ductal closure is a class II test and is intended to demonstrate complete closure of the patent ductus arteriosus. It is recognized that additional postoperative studies may be needed for treatment of individual patients in the perioperative period. The evaluation provides for detection of additional perioperative problems such as wound infection, pleural effusion, etc. After documentation of successful ligation/division of the ductus arteriosus and resolution of all perioperative problems, further cardiac evaluation is unnecessary.

The algorithm for patent ductus arteriosus in patients older than 6 months differs from that for patients younger than 6 months only in that surgical closure can be performed shortly after diagnosis.

Ostium Secundum Atrial Septal Defect (unoperated) (Figs 4 and 5)

Exclusion Criteria
1. The patient is older than 21 years.
2. The patient has congestive heart failure.

Fig. 3. Algorithm for patent ductus arteriosus in a patient older than 6 months. History, physical examination, chest radiogram, electrocardiogram, and echocardiographic studies will be performed in postoperative period before patient has entered postoperative phase of this protocol. It is assumed that documentation of resolution of all perioperative problems such as pleural and pericardial effusion or postoperative wound infections will be obtained before patient is enrolled in this protocol. ECG indicates electrocardiogram; PDA, patent ductus arteriosus.

Fig. 4. Algorithm for atrial septal defect for a patient 5 years old or younger. ECG indicates electrocardiogram; CNS, central nervous system; SX, symptoms; ASD, atrial septal defect.

Definition of Shunt Size
1. Small shunt: Absence of diastolic flow, normal chest x-ray, echocardiographic evidence of normal right ventricular dimension, and normal interventricular septal motion or a pulmonary to systemic flow ratio (Qp/Qs) <1.5.
2. Large shunt: Presence of diastolic flow rumble or ECG evidence of right ventricular hypertrophy, chest radiographic evidence of cardiomegaly or increased pulmonary vascular markings, echocardiographic evidence of right ventricular enlargement or paradoxical septal motion or a Qp/Qs ≥1.5.

Algorithm for Ostium Secundum Atrial Septal Defect (unoperated)

Two algorithms were developed for the management of ostium secundum atrial septal defect. For one, "Time
Fig 6. Algorithm for postoperative ostium secundum atrial septal defect. For this algorithm, "Time 0" is first postoperative, out-of-hospital visit subsequent to resolution of all perioperative problems such as pleural and pericardial effusions, postoperative wound infections, etc. In general, this visit will occur within 3 months postoperatively. ASD indicates atrial septal defect; ECG, electrocardiogram.

0" is the initial evaluation by a pediatric cardiologist and the patient is aged 5 years or younger at the time of the initial evaluation. For the other, "Time 0" is the initial evaluation by a pediatric cardiologist but the patient is older than 5 years at the time of that evaluation.

For patients initially evaluated at the age of 5 years or younger, the algorithm allows for medical treatment or surgical closure (ie, off protocol) for symptomatic patients with a large left-to-right shunt. The algorithm also allows for continued observation of patients with a large left-to-right shunt and symptoms in anticipation of possible spontaneous closure of the atrial septal defect. In asymptomatic patients with a large left-to-right shunt, surgical closure should be performed between 2 and 5 years of age.

For patients initially evaluated when they were older than 5 years, the atrial septal defect can be closed electively, assuming there are no contraindications to surgery.

Ostium Secundum Atrial Septal Defect (operated) (Fig 6)

The algorithm implies that a history, physical examination, chest radiogram, electrocardiogram, and echocardiogram will have been performed in the postoperative period before the patient has entered into this protocol.

Exclusion Criteria

1. The patient is older than 21 years.
2. The patient has evidence of pulmonary hypertension.
3. The patient has a residual atrial septal defect (echocardiographically demonstrated).

Algorithm for Ostium Secundum Atrial Septal Defect (operated)

For this algorithm, "Time 0" is the first postoperative outpatient visit subsequent to resolution of all perioperative problems such as pleural and pericardial effusions or postoperative wound infections. In general, this visit occurs within 3 months after surgery. The algorithm allows one postoperative echocardiogram to document complete atrial septal defect closure. In addition, a second postoperative echocardiogram to assess right ventricular size/function is a class II test.

Small Ventricular Septal Defect (Fig 7)

Exclusion Criteria

1. The patient is symptomatic.
2. The patient has an abnormal second heart sound.
3. The patient has diastolic flow rumble.
4. The patient has a murmur of aortic insufficiency.
5. The patient has an abnormal chest radiogram.
6. The patient has an abnormal electrocardiogram.
7. The patient has a supracristal outlet or inlet ventricular septal defect.
8. The patient has a prolapsed aortic valve cusp.
9. The patient has left ventricular volume enlargement.
10. There is Doppler evidence of significant aortic or mitral insufficiency or evidence of right ventricular or pulmonary arterial hypertension.
11. The patient has associated congenital heart defects.

Algorithm for Small Ventricular Septal Defect (Echocardiographically Demonstrated)

For this algorithm, "Time 0" is the initial evaluation by an experienced pediatric cardiologist. There was
considerable debate among committee members as to whether echocardiographic definition of a small ventricular septal defect was necessary to exclude other conditions that could mimic the physical findings of a small ventricular septal defect or associated problems such as prolapse of an aortic valve cusp. After considerable discussion, it was decided to construct the algorithm for patients with a small ventricular septal defect in whom an echocardiogram had been done. However, the committee recognized that some pediatric cardiologists diagnose a small ventricular septal defect without echocardiographic confirmation of the defect or echocardiographic exclusion of associated problems. The committee agreed that diagnosis and management of patients with a small ventricular septal defect without echocardiographic confirmation was acceptable. Under these circumstances, it is incumbent on the clinician to provide appropriate ongoing evaluation to detect and manage conditions that could mimic the physical findings of a small ventricular septal defect or the problems associated with it.

Postoperative Ventricular Septal Defect (Fig 8)

The algorithm implies that a history, physical examination, chest radiogram, electrocardiogram, and echocardiogram are performed after surgery and before the patient has entered into this protocol.

Exclusion Criteria

1. The patient is asymptomatic.
2. A residual ventricular septal defect of any size is detected echocardiographically.
3. The patient has an abnormal chest radiogram.
4. The patient has an abnormal electrocardiogram (except isolated right bundle branch block).
5. The patient has transient perioperative second- or third-degree atroventricular block.
6. The patient has evidence of persistent postoperative pulmonary arterial hypertension.
7. A left ventriculotomy is used for surgical closure of the ventricular septal defect.
8. The patient has a supracristal outlet or inlet ventricular septal defect.

Algorithm for Postoperative Ventricular Septal Defect

For this algorithm, “Time 0” is the initial evaluation, out-of-hospital visit after resolution of all perioperative problems such as pleural effusions, pericardial effusions, postoperative fevers, or wound infections. In general, this visit will occur within 3 months of hospital discharge. ECG indicates electrocardiogram.

Mild Pulmonary Valve Stenosis (Fig 9)

Exclusion Criteria

1. Clinical findings are atypical.
2. The Doppler peak instantaneous gradient is ≥40 mm Hg measured under resting conditions.
3. The patient has an abnormal electrocardiogram (except only mild right ventricular hypertrophy).
4. The patient has a dysplastic pulmonary valve.

Algorithm for Mild Pulmonary Valve Stenosis

For this algorithm, “Time 0” is the initial evaluation by a pediatric cardiologist. The algorithm provides for
Fig 10. Algorithm for postoperative or post-balloon valvuloplasty for pulmonary valve stenosis. For this algorithm, "Time 0" is first postoperative out-of-hospital visit subsequent to resolution of all perioperative problems such as pleural effusions, pericardial effusions, postoperative fevers, or wound infections. Entry of a patient at "Time 0" assumes that a postoperative echocardiogram has been performed and that the transpulmonary valve gradient is <40 mm Hg. It also assumes that residual subvalvar pulmonary stenosis has resolved. ECG indicates electrocardiogram.

Detection of an increasing transpulmonary gradient. The committee agreed that echocardiographic studies were essential for assessing the severity of pulmonary stenosis. Since progressive severity is highly unlikely in a patient with initially mild (<40 mm Hg) pulmonary stenosis, more than three echocardiographic studies of a patient with a persistent transpulmonary gradient less than 40 mm Hg are probably unnecessary.

Postoperative Pulmonary Valve Stenosis (Fig 10)

The algorithm implies that a history, physical examination, chest radiograph, electrocardiogram, and echocardiographic studies will be performed in the postoperative period before the patient has entered into this protocol.

Exclusion Criteria

1. The residual Doppler peak instantaneous gradient is ≥40 mm Hg.

2. The patient has preoperative critical pulmonary valve stenosis (suprasystemic right ventricular pressure).

3. The patient is younger than 1 year at the time of balloon valvuloplasty or surgical valvotomy.

Algorithm for Postoperative (or Post—Balloon Valvuloplasty) Pulmonary Valve Stenosis

For this algorithm, "Time 0" is the first postoperative, outpatient visit subsequent to resolution of all perioperative problems such as pleural effusions, pericardial effusions, postoperative fevers, or wound infections. Entry of a patient at Time 0 assumes that a postoperative echocardiogram has been performed and that the transpulmonary valve gradient is <40 mm Hg. It also assumes that residual subvalvar pulmonary stenosis has resolved at the time of the initial postoperative, out-of-hospital visit.

Premature Ventricular Contractions (Fig 11)

The algorithm assumes that the patient is referred to a pediatric cardiologist because of an irregular rhythm or symptoms of palpitations.

Exclusion Criteria

1. The patient is aged 21 years or older.

2. The patient has a family history of premature (less than 40 years old) sudden death of presumed or established cardiac origin.

3. The patient has a family history of prolonged QT, interval syndrome.

4. The patient has a family history of cardiomyopathy.

5. The patient has a history suggestive of myocarditis.

6. The patient has a history of syncope, presyncope, or seizures.

7. The patient has a subjective history of sustained tachycardia.

8. The patient has cardiac symptoms in addition to palpitations.

9. The patient has undergone a cardiac examination that was designated "abnormal" by a pediatric cardiologist.

10. The patient has an abnormal electrocardiogram (except for associated premature ventricular contractions.)

11. There is a prolonged QT, interval.

Algorithm for Isolated Premature Ventricular Contractions

For this algorithm, "Time 0" is the initial evaluation by a pediatric cardiologist. The committee debated extensively about the need for 24-hour ambulatory ECG monitoring, echocardiography, and exercise testing. Thus, these were considered class II tests. Use of these studies is influenced by factors such as the patient's age, frequency and intensity of palpitations, and probability of an associated viral illness or other systemic diseases.

Discussion

Innocent Cardiac Murmurs

Three relatively recent studies have focused on the usefulness of diagnostic tests in the initial evaluation of murmurs in children. Newburger et al. assessed results of the initial evaluation of 280 children referred to a
pediatric cardiologist because of a cardiac murmur. After the initial history and physical examination, 142 of the 280 children were deemed to have no heart disease. After further testing and follow-up evaluation, 5 of the 142 patients were thought to have “possible” (2) or “definite” (3) heart disease. The two patients with possible heart disease had ventricular hypertrophy as measured by ECG criteria, and one had a low-normal shortening fraction on M-mode echocardiography. All three patients with definite heart disease had mitral valve prolapse based on a two-dimensional echocardiographic study. Smythe et al. assessed the results of the initial evaluation by pediatric cardiologists of 161 patients with a cardiac murmur. After the initial history and physical examination, 109 of the 161 patients were thought to have an innocent murmur. In two of the 109 patients, a heart defect was detected by echocardiography: one small muscular ventricular septal defect and one small atrial septal defect. There was some question as to whether the patient with an atrial septal defect had in fact a patent foramen ovale. When the results of these were considered, a defect requiring care that differed from the management of an innocent murmur was ultimately discovered in only one patient (0.3% of the group), who had a small ventricular septal defect for which endocarditis prophylaxis would be recommended.

Geva et al. reviewed echocardiograms of 50 children who, following physical examination, electrocardiography, and chest radiography, were thought to have an innocent murmur. The echocardiogram was normal in 48 patients, and the other two patients had a bicuspid aortic valve.

Based on these three studies, examination alone would have failed to identify three patients (1%) for whom endocarditis prophylaxis should be recommended. Given the unestablished efficacy of endocarditis prophylaxis and its very low risk in patients with an atrial septal defect, it may not be cost-effective to perform echocardiographic studies in all patients with an innocent murmur to detect the 1% of patients who might require endocarditis prophylaxis.

**Patent Ductus Arteriosus**

The diagnosis of a patent ductus arteriosus can be made on the basis of a history and physical examination and confirmed by Doppler echocardiography. Cardiac catheterization is necessary only in atypical cases, primarily those in which high pulmonary vascular resistance is suspected.

Complications of a patent ductus arteriosus include infective arteritis, aneurysm of the ductus, congestive heart failure, pulmonary vascular obstructive disease, and calcification of the ductus. Therefore, the presence of a clinically detectable ductus arteriosus is considered an indication for closure. Postoperative sequelae are uncommon, limited primarily to early postoperative complications of thoracotomy, such as but not limited to wound infection, hemothorax, or recurrent laryngeal nerve injury. Other sequelae include incomplete closure or recanalization.

**Ostium Secundum Atrial Septal Defect**

The natural history of secundum atrial septal defect includes development in adulthood of right ventricular failure, atrial arrhythmias, and in rare instances, pulmonary vascular obstructive disease. Investigators have found a greater mortality for patients with unoperated atrial septal defect than for normal subjects or patients who have had surgical closure of the defect. Although spontaneous closure of an atrial septal defect is uncommon in older children, adolescents, and adults, spontaneous closure between 4% and 39.5% has been observed in infancy and early childhood. Most investigators agree that spontaneous closure of an atrial septal defect is unlikely after the age of 4 years and very uncommon after the age of 4 years. Thus, the rationale for conservative treatment of asymptomatic patients with secundum atrial septal defects who are younger than 4 years is justified.

Treatment of symptomatic patients with secundum atrial septal defects in infancy is controversial. Some investigators have demonstrated spontaneous closure of atrial septal defects in infants who had congestive heart failure. Whether it is better to surgically close an atrial septal defect in an infant with congestive heart failure or to treat these patients with anticoagulant therapy is unclear. For this reason, these patients are not considered in this algorithm.

Little is known about the fate of asymptomatic patients with a small atrial septal defect. The incidence of small atrial septal defects is unknown, and it is difficult to distinguish a “small” atrial septal defect from a “large” patent foramen ovale. In one study of small atrial septal defect, progression of the left-to-right shunt occurred in 15% of patients, and spontaneous closure occurred in 11.5%. An additional risk of a small atrial septal defect (or a patent foramen ovale) is paradoxical embolization and stroke. Certainly it seems reasonable to close atrial septal defects if the left-to-right shunt becomes progressively larger. For these reasons the algorithm provides for continued follow-up evaluation of these patients. The efficacy of closing an atrial septal defect (or patent foramen ovale) in a patient who has had an unexplained stroke is unknown. However, the algorithm allows the clinician to elect that course of treatment.

Continued long-term follow-up of patients who have had operative closure of an atrial septal defect seems reasonable to detect the onset of new arrhythmias. In one study, 2% of patients with an atrial septal defect had arrhythmias before surgery, but 23% had them 0.1 to 8.0 years after surgery.

**Ventricular Septal Defect**

There have been several outcome studies of the treatment of ventricular septal defect.

**Small Ventricular Septal Defect**

The Second Joint Study on Natural History of Congenital Heart Defects provided comprehensive data on 242 young adults known to have a small ventricular septal defect who were followed up medically from early childhood. Spontaneous closure occurred in 13.7%. Five patients (2.1%) developed clinical signs of congestive heart failure, and 10 (4.1%) were taking digitalis, diuretics, or antiarrhythmic agents. Bacterial endocarditis occurred in five patients (7.1 per 10 000 person-years). In the entire cohort of 570 with ventricular septal defects of all sizes, 12 (2%) developed aortic regurgitation.

Thus, although in most patients a small ventricular septal defect is a relatively benign condition, ongoing...
medical evaluation of these patients is necessary to detect and manage the relatively rare but important complications of endocarditis, arrhythmias, aortic regurgitation, and subvalvar pulmonary stenosis and discrete subaortic stenosis.

**Postoperative Ventricular Septal Defect**

Surgical closure of ventricular septal defects has been done successfully for nearly 40 years. Surgical techniques and timing and the mortality and morbidity of operation have changed over time. Preoperative or postoperative high pulmonary vascular resistance and surgically acquired complete atroventricular block are risk factors for long-term morbidity and mortality. For the purposes of these guidelines, only children with isolated ventricular septal defect and normal pulmonary vascular resistance, intact atroventricular conduction, and complete closure or, at most, a small residual ventricular septal defect were considered.

In the Second Natural History Study 226 young adults had undergone ventricular septal defect closure in childhood. Subsequent operations were performed in 31 of 33 patients with a pulmonary artery band and 7 of 193 patients with primary closure. Episodes of congestive heart failure were noted in 72 patients, and 6 patients had cardiac pacemakers. Bacterial endocarditis occurred in 10 patients.

**Pulmonary Stenosis**

The algorithm for the management of patients with mild pulmonary stenosis is based on the premise that patients 1) with a transpulmonary peak Doppler gradient <40 mm Hg do not require operation or balloon dilation; 2) with a transpulmonary gradient >40 mm Hg may require surgery or balloon dilation; 3) with transpulmonary gradients <40 mm Hg may progress to gradients >40 mm Hg; 4) with gradients <25 mm Hg are less likely to progress to gradients >40 mm Hg than patients with gradients between 26 and 39 mm Hg.

In the First Natural History Study 14% of all patients with valvular pulmonary stenosis had a significant increase of transpulmonary gradient at the conclusion of the study.22 For patients with an initial transpulmonary gradient <40 mm Hg, progression to a gradient >60 mm Hg was rare. Gradients were unlikely to increase in patients older than 12 years and were most likely to increase significantly in patients younger than 4 years, particularly if the initial gradient was >40 mm Hg. There were 94 patients with pulmonary stenosis and a transpulmonary gradient <40 mm Hg on admission to the First Natural History Study. Of these, 5 progressed to a gradient >40 mm Hg at the termination of that study and an additional two patients progressed to a gradient >40 mm Hg at the time of the Second Natural History Study. None of the seven patients underwent surgery. An additional eight patients had an admission gradient <40 mm Hg but had a pulmonary valvotomy. One of the eight progressed to a gradient >40 mm Hg at the end of the First Natural History Study. Although the admission gradient for the remaining seven was <40 mm Hg, the immediate postoperative gradient was unknown. In the Second Natural History Study, no cardiac-related deaths occurred among the medically managed patients.23 For patients who had initial transpulmonary gradients <25 mm Hg, 96.1% were free of cardiac operation over a 25-year period.

The results of operation (and presumably balloon dilation) of pulmonary valve stenosis are excellent. In the Second Natural History Study, 349 patients opted for relief of pulmonary valve stenosis.23 There were a total of 19 deaths. Only 3.1% of patients required a second operation within 10 years of the initial operation. These data support an algorithm designed to detect the recurrence of significant pulmonary stenosis and detection of the rare patient who experiences late complications.

**Isolated Uniform Premature Ventricular Contractions**

Based on experience with older patients with ischemic heart disease, the notion that premature ventricular contractions lead to ventricular tachycardia that in turn leads to ventricular fibrillation has evolved. This perception has continued to evolve in the ischemic heart disease model, but extrapolation of the adult experience to the pediatric patient is difficult, in part because ischemic heart disease is a rare cause of premature ventricular contractions in the pediatric patient. Nevertheless, the possible association with occult heart disease makes evaluation of premature ventricular contractions an important clinical problem.

Isolated premature ventricular contractions with uniform ECG features are commonly found in older children and adolescents but are infrequently detected in infants. For example, Southall and coworkers24 evaluated 134 healthy full-term infants using 24-hour ambulatory ECG monitoring, and no premature ventricular contractions were recorded in these newborns. In a previous study Southall et al25 reported a prevalence of premature ventricular contractions in newborns of 0.9%. In children between 7 and 11 years of age, 1 of 92 patients had premature ventricular contractions recorded with 24-hour ambulatory ECG monitoring.26 Scott et al27 evaluated 131 presumably healthy boys aged 10 to 13 years and found that 26% had premature ventricular contractions. In a study of boys aged 14 to 16 years, 41% had premature ventricular contractions and 3% had nonsustained ventricular tachycardia.28 Brodsky et al29 found that 50% of 50 male medical students had premature ventricular contractions. Based on these studies, it appears that the prevalence of premature ventricular contractions in otherwise healthy children is uncommon in infancy but increases with age. This suggests that the evaluation of a patient with premature ventricular contractions must be tempered by the patient's age.

Although it has been suggested24,30 that isolated premature ventricular contractions in patients who seem otherwise normal is a benign problem, the confounding effects of age, number of premature ventricular contractions, or variation in ECG features on the natural history of these patients has not been thoroughly addressed. Certainly there are conditions associated with ventricular ectopic activity that are related to increased risk of sudden death, such as myocarditis, myocardial tumors, prolonged QT interval syndromes, and arrhythmogenic right ventricular dysplasia. These are relatively rare and usually associated with other physical or ECG abnormalities. The necessity of extensively testing all patients with premature ventricular...
Bacterial Endocarditis

## Cardiac Conditions

- Endocarditis prophylaxis is recommended for most unoperated congenital cardiac malformations.
- Endocarditis prophylaxis is not recommended for isolated secundum atrial septal defect.
- Endocarditis prophylaxis is not recommended for 6 months after repair (without residual) of secundum atrial septal defect, ventricular septal defect, or patent ductus arteriosus.
- Endocarditis prophylaxis is recommended for patients with a history of previous endocarditis, regardless of current cardiac status.

Specific surgical and dental procedures needing coverage and the antibiotic regimens recommended by the AHA are given in reference 31.

contractions to exclude these relatively rare problems is unclear. For these reasons, echocardiography and 24-hour ambulatory ECG monitoring are Class II tests. A thorough past medical history and family history are essential in the evaluation of these patients to alert the clinician to potentially important conditions.

### Other Considerations

Many patients with congenital cardiac defects require prophylaxis for bacterial endocarditis when undergoing certain surgical and dental procedures (the Table). The AHA has made specific recommendations concerning antibiotics.

Few patients with congenital heart defects are ever truly “cured.” Recommendations for endocarditis prophylaxis, sports participation, and genetic counseling change as new information is acquired. Ongoing contact with patients is necessary to communicate these changes to them. Maintaining contact with many patients over a long period of time is difficult. It is logistically impossible for health providers alone to ensure that this contact is maintained. Many patients will eventually be followed up by an internist cardiologist. It is important that internist cardiologists have a sufficient level of expertise and experience to follow these patients into their adult years.

### References

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