ACC/AHA Guidelines for the Clinical Application of Echocardiography

A Report of the American College of Cardiology/American Heart Association Task Force on Assessment of Diagnostic and Therapeutic Cardiovascular Procedures

( Subcommittee to Develop Guidelines for the Clinical Application of Echocardiography)

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Preamble

It is becoming more apparent each day that despite a strong national commitment to excellence in health care, the resources and personnel are finite. It is, therefore, appropriate that the medical profession examine the impact of developing technology on the practice and cost of medical care. Such analysis, carefully conducted, could potentially have an impact on the cost of medical care without diminishing the effectiveness of that care.

To this end, the American College of Cardiology and the American Heart Association in 1980 established a Task Force on Assessment of Diagnostic and Therapeutic Cardiovascular Procedures with the following charge:

The Task Force of the American College of Cardiology and the American Heart Association shall define the role of specific noninvasive and invasive procedures in the diagnosis and management of cardiovascular disease.

The Task Force shall address, when appropriate, the contribution, uniqueness, specificity, indications, contraindications, and cost-effectiveness of such specific procedures.

The Task Force shall include a Chairman and six members, three representatives from the American Heart Association and three representatives from the American College of Cardiology. The Task Force may select ad hoc members as needed upon the approval of the Presidents of both organizations. Recommendations of the Task Force are forwarded to the President of each organization.

The members of the Task Force are: George A. Beller, MD, Roman W. DeSanctis, MD, Harold T. Dodge, MD, J. Ward Kennedy, MD, T. Joseph Reeves, MD, Sylvan Lee Weinberg, MD, and Charles Fisch, MD, Chairman.

This document was reviewed by the officers and other responsible individuals of the two organizations and received final approval in May 1990. It is being published simultaneously in Circulation and the Journal of the American College of Cardiology. The potential impact of this document on the practice of
cardiology and some of its unavoidable shortcomings are clearly set out in the introduction.

Charles Fisch, MD, FACC

General Considerations and Scope

Echocardiography is an examination technique that provides images of cardiac and great vessel anatomy and blood flow by ultrasound. Although ultrasound may be applied in different forms (M-mode, two-dimensional, spectral, and color flow Doppler imaging) and by different techniques (trans-thoracic, transesophageal), all are encompassed in the term echocardiography. When applied by the customary transthoracic approach, the examination involves little, if any, patient discomfort and has not been associated with patient risk. The ability of echocardiography to provide unique information regarding cardiac structure and function, the lack of ionizing radiation, the portability of the instrument, and the potential for repeated studies have led to the widespread utilization of echocardiography for virtually all categories of known and suspected cardiovascular disease.

When the role of echocardiography is discussed, it is assumed that the patients who are given the test have undergone a complete physical examination with medical history and that the usefulness and likely diagnostic yield have been carefully considered. The decision to perform an echocardiogram and interpret the results should not be done without consideration of other factors relevant to the individual patient.

In discussing the optimal use of echocardiography, it is important to consider the relation between the information derived and the ability to establish a definitive diagnosis or make a therapeutic decision. In some cases, echocardiography will definitively establish the etiology of a symptom complex (such as acquired ventricular septal defect after myocardial infarction). Echocardiography may establish the need for interventional therapy (as in patients with left atrial myxoma or cardiac tamponade, for example). Commonly, echocardiography will indicate the need for corrective therapy or specific follow-up (as in mitral or aortic stenosis). Often, it provides data that are confirmatory in nature, that have prognostic significance, or that aid in the overall evaluation of a disease state. The primary reason not to perform echocardiography in many cases is economic. The cost-effectiveness of echocardiography in these settings is related to the individual case. Societal resources must be considered by the physician. In addressing instances where echocardiography provides neither definitive diagnosis nor data on which therapeutic decisions may be based, this report attempts to indicate those situations in which the performance of echocardiography is not generally cost-effective.

The report is based on the assumption that the echocardiogram is performed and interpreted in accordance with the guidelines for optimal training set forth by the American Society of Echocardiography, the American College of Cardiology,1,2 and the Society of Pediatric Echocardiography.3

This report is divided into two parts, the adult and the pediatric guidelines. In classifying the usefulness of echocardiography in clinical cardiovascular practice, each part is divided into two major sections. In the first, the usefulness of echocardiography in specific cardiovascular disorders is considered. In the second, the usefulness of echocardiography in evaluating general symptoms and signs is considered. The latter method of categorizing the appropriateness of echocardiography corresponds to the frequent use of the technique in the evaluation of the significance and mechanism of such common presenting complaints as dyspnea, chest discomfort, and cardiac murmur. It is hoped that this symptom- and sign-based classification will provide guidance to physicians regarding the utility of echocardiography in the evaluation of common clinical problems. The pediatric guidelines include those for fetal echocardiography.

Throughout this report, we have used the following classification system:

Class I: Conditions for which or patients for whom there is general agreement that echocardiography is appropriate

Class II: Conditions for which or patients for whom echocardiography is frequently used but there is a divergence of opinion with respect to its appropriateness

Class III: Conditions for which or patients for whom there is general agreement that echocardiography is not appropriate

Valvular Heart Disease

In evaluating the patient with valvular heart disease, the clinician seeks information about valvular structure and function, as well as about the effect of the valvular abnormality on cardiac anatomy and physiology. Assessment of the cardiac consequences of stenotic valve lesions requires estimation of the transvalvular pressure gradient and valve area and assessment of the cardiovascular consequences. In the case of regurgitant valves, assessment requires determination of not only the degree of regurgitation but also the anatomy of the receiving chamber and ventricular function. Thus, complete evaluation of the patient with acquired valvular heart disease requires attention not only to the valve lesion as such but also to its integrated role in overall cardiac function.

Echocardiography has become the noninvasive diagnostic method of choice for the evaluation of valvular heart disease. Because of the precision and accuracy of modern echocardiographic data, selected patients may undergo definitive surgical therapy without the need for cardiac catheterization.4-6 In general, echocardiographic methods are more precise in defining the severity of valvular stenosis than
of valvular regurgitation; however, this is also true of the alternative methods.

Native Valve Disease

Echocardiography is extremely useful in the diagnosis of stenosis and regurgitation of all four native cardiac valves. The largest clinical experience and most definitive validation studies have been reported in mitral and aortic valve disease, with less experience in the study of tricuspid and pulmonary valve disease.7–9

A particular strength of echocardiography is its ability to help identify the etiology of specific valvular abnormalities. Thus, calcification and fibrosis (due to degenerative and rheumatic diseases), prolapse (either idiopathic or related to specific connective tissue disorders), endocarditis, and other pathophysiologic mechanisms of valve dysfunction may be characterized using ultrasound examination techniques. Special mention should be made of mitral valve prolapse. Although undisputed value in the diagnosis of mitral prolapse, some controversy continues as to the optimal echocardiographic criteria for this diagnosis.10,11

The degree of valvular fibrosis, calcification, and immobility bears an approximate relation to the degree of valvular stenosis. The severity of mitral stenosis may be accurately quantified in most patients by planimetric measurement of the valve area.26 In all other valvular stenoses, hemodynamic measurement with Doppler echocardiography gives more accurate assessment of severity. Doppler echocardiographic techniques can accurately estimate instantaneous and mean transvalvular pressure gradient across stenotic mitral,12 aortic,12–14 and pulmonary6 valves. In the evaluation of semilunar valvular stenosis, peak instantaneous and mean pressure gradient, as well as estimates of aortic valve orifice area based on the “continuity” principle,6,14 have been successfully used to assess severity. In studies of stenotic aortic valves, measurement of mean pressure gradient as well as estimation of orifice area by the continuity principle or by the pressure half-time method15–19 have been employed.

Doppler echocardiographic methods are by far the most sensitive means of identifying the presence of valvular regurgitation. In fact, Doppler techniques are so sensitive that care must be taken not to interpret a physiologic phenomenon as indicating pathologic regurgitation. Studies20,21 have indicated that mild apparent retrograde flow disturbances are frequently detected in Doppler echocardiograms of normal subjects by both pulsed and color flow Doppler methods.

Precise assessment of the severity of regurgitant valvular lesions is difficult using any invasive or noninvasive imaging technique. Pulsed Doppler and Doppler color flow mapping techniques yield only semiquantitative estimates of the severity of mitral and aortic regurgitation.22–25 Due in part to difficulties in defining an appropriate independent standard, studies validating echocardiographic grading of the severity of tricuspid and pulmonary valve regurgitation are few. Echocardiography is the most sensitive noninvasive method of identifying valvular annular calcification.

Infective Endocarditis

Transesophageal two-dimensional echocardiography has been utilized for the diagnosis and characterization of these valvular masses. However, the sensitivity of transthoracic examination in visualization of valvular vegetations (≥4–5 mm in size) has ranged from 50% with M-mode echocardiography to 70% with two-dimensional echocardiography. Transesophageal echocardiography promises to improve the sensitivity of visualization of vegetations to nearly 90%.26 Because of the possibility of false-negative examination results, echocardiography should not be used as the sole means of excluding the presence of endocarditis. Echocardiography may be of use in identifying vegetations in patients with culture-negative endocarditis.27

In addition to identifying the presence of valvular vegetations, echocardiography offers important information about the complication of endocarditis, including not only the valvular sequelae (such as ruptured chordae tendineae), abscesses, and shunt lesions, but also the hemodynamic and pathologic consequences.

Prosthetic Valve Dysfunction

Echocardiographic imaging can be used to define abnormalities of prosthetic disk or ball motion; however, this usefulness is limited. Thrombus on struts, stents, or disks is difficult to appreciate because of reverberations and shadowing from the prosthetic valve structure. This is especially true of mitral prosthetic valves. In this situation the addition of transesophageal echocardiography is helpful since the combination of techniques allows a view of the valves from several angles.

Evaluation of the severity of prosthetic valvular dysfunction is best performed by Doppler techniques. The pressure gradient across aortic and mitral prostheses may be estimated accurately utilizing Doppler techniques in the majority of prosthetic valves.28,29 Special care must be taken in the identification of the highest velocity jets, particularly in valves that produce eccentric flow, such as tilting disk and ball valve prostheses. The transvalvular pressure gradient expected across normally functioning prosthetic valves will vary with valve type and size. However, sufficient data are available concerning in vivo and in vitro flow behavior of these valves to interpret the velocities measured in the clinical setting.30 Assessment of prosthetic valve effective orifice areas, particularly AV valve areas by pressure halftime methods, may be less accurate than the assessment of mean and peak pressure gradients.31 Diagnosis of mild prosthetic obstruction may be difficult because of the variable transvalvular pressure gradients expected in valves of differing designs and sizes.

Echocardiography can identify bioprosthetic valve regurgitation. When employing Doppler tech-
niques to identify abnormal mechanical prosthetic valve insufficiency, the normal occurrence of small amounts of regurgitation through these devices should be kept in mind.\textsuperscript{32} Doppler color flow mapping appears capable of discriminating between central regurgitant flow jets and eccentric jets due to paravalvular leaks, although this differentiation is not perfect.\textsuperscript{33,34}

The strengths and limitations of assessing severity of prosthetic regurgitation are in general similar to those noted for native valves. However, reverberations and attenuation (shadowing) associated with prostheses complicate the process of mapping regurgitant tricuspid and mitral jets. Transesophageal imaging methods avoid some of these problems,\textsuperscript{35} especially when imaging the prosthetic mitral valve. Insufficient data are available to classify the usefulness of echocardiographic assessment of the severity of dysfunction of regurgitant pulmonary valve prostheses.

**Prosthetic Valve Infective Endocarditis**

Diagnosis of prosthetic valve endocarditis by the transthoracic technique is more difficult than the diagnosis of endocarditis of native valves because of the reverberations, attenuation, and other image artifacts related to both mechanical valves and bioprostheses. Particularly in the case of mechanical valves, echocardiography is probably helpful only when there is a large or mobile vegetation. Thus, the technique cannot be used to exclude the presence of vegetations. These limitations are diminished with the use of transesophageal recording techniques because of the superior imaging quality. Transesophageal techniques have enhanced echocardiographic assessment of prosthetic valve infective endocarditis, especially of the mitral valve and of both mitral and aortic annular areas for abscesses.

Doppler techniques offer important information about the functional consequences of endocarditis of prosthetic valves, such as the existence of paravalvular leaks. It should be noted, however, that paravalvular leaks are not specific for endocarditis. Importantly, echocardiography may identify vegetations on native valves in patients with suspected prosthetic endocarditis.

**Indications for Echocardiography**

**Class I**
1. Native cardiac valve disease
2. Prosthetic cardiac valve disease
3. Suspected or proved infective endocarditis

**Class II**
None

**Class III**
None

**Ischemic Heart Disease**

Because atherosclerosis still represents the major health threat to adults in Western societies, there has been tremendous effort expended on the development of noninvasive techniques for the diagnosis of coronary artery disease and its consequences. As a result, echocardiography has evolved to become a powerful tool in the assessment of patients at all stages of the ischemic disease process. It is useful to consider its application in the following circumstances:

**Diagnosis**

**Acute myocardial infarction.** Segmental ventricular wall motion abnormalities are characteristic of myocardial infarction in all but the smallest of infarctions and correlate with specific coronary artery distribution and pathology.\textsuperscript{36-45} However, the loss of systolic contraction, the loss of wall thickening during systole, and even frank systolic bulging of a ventricular segment may also occur in acute and chronic ischemia or myocardial scar and therefore are not pathognomonic for infarction.\textsuperscript{37-45} Segmental wall motion abnormalities may also occur in myocarditis and other conditions not associated with coronary occlusion, but the lack of systolic wall thickening is more specific for ischemia.\textsuperscript{46} In patients with acute myocardial infarction, the ventricular wall motion abnormalities include not only the acutely infarcted segment but also previously infarcted segments as well as ischemic, “stunned,” and “hibernating” myocardium in adjacent zones.\textsuperscript{39,41,42,45,47} Together these represent the functional infarct size, which may be an overestimation of the true anatomic infarct size in some patients. Nevertheless, echocardiography-derived infarct size\textsuperscript{42} correlates with thallium-201 perfusion defects,\textsuperscript{40} peak creatine kinase levels,\textsuperscript{41} hemodynamic changes, \textsuperscript{42} results of catheterization ventriculography,\textsuperscript{43} and coronary angiography,\textsuperscript{44} early\textsuperscript{48} and late\textsuperscript{49} complications, mortality,\textsuperscript{42,50} and pathologic findings.\textsuperscript{45}

In acute myocardial infarction, clinical status and ventricular function may not be static, especially after reperfusion therapy. Serial echocardiography is a noninvasive method for monitoring the changes that occur.

Evaluation of myocardial segments uninvolved by the acute infarction is also important. The lack of the expected compensatory hyperkinesia can indicate multivessel disease,\textsuperscript{47} and, therefore, the presence of asynergy in a remote segment carries an increased risk of postinfarction angina, progression in Killip classification, shock, and death.\textsuperscript{41}

The right ventricle is involved to some degree in one third of inferior myocardial infarctions,\textsuperscript{51} and an associated right ventricular infarction can have significant hemodynamic implications for patient management. Although somewhat more difficult to image, echocardiography is useful in the diagnosis of biventricular infarction.\textsuperscript{52}

**Complications of Acute Myocardial Infarction**

In addition to revealing and allowing quantitation of wall motion abnormalities, the echocardiogram can be used to detect five major early complications of acute myocardial infarction:
1. **Thrombus.** Echocardiography is the definitive test for intracardiac thrombi.\(^{53-58}\) The true frequency of postinfarction thrombus was not appreciated until echocardiographic studies were done. Thrombi are more common in anterior and apical than in inferior infarctions.\(^{56-58}\) Patients developing a mural thrombus are at increased risk of mortality.\(^{54}\) The therapeutic implications of left ventricular thrombi are evolving and therefore the need for serial imaging is controversial.

2. **Acute mitral regurgitation.** The importance of new onset of mitral regurgitation is demonstrated by its correlation with prognosis.\(^{59}\) Acute mitral regurgitation can occur from several mechanisms. One of the heads of the papillary muscle may acutely rupture.\(^{60}\) Alternatively, the papillary muscle and associated free wall may be acutely ischemic or may develop late fibrosis and foreshorten, rendering the valve regurgitant.\(^{61}\) Both are evident echocardiographically. Doppler ultrasonography can be used to locate the regurgitant jet, and color flow mapping can help in its quantitation.\(^{62}\)

3. **Ventricular septal rupture.** Two-dimensional echocardiography can be used to visualize the typical interventricular septal defect\(^{63}\) and Doppler ultrasonography can be used to locate the left to right shunt\(^{64}\) and differentiate it from mitral valve regurgitation\(^{65}\) or from tricuspid valve regurgitation resulting from right ventricular infarction. This information is essential when surgical intervention is being considered.

4. **Free wall rupture.** Patients who survive free wall rupture develop a pseudoaneurysm that has a characteristic echocardiographic appearance.\(^{66}\) Echocardiography also provides information regarding the presence or absence of associated cardiac tamponade\(^{67}\) and can prove helpful in determining the timing of surgical intervention.

5. **Infarct expansion.** Infarct expansion may occur after myocardial infarction. When this occurs, the patient’s prognosis has been shown to be worse.\(^{68}\) Echocardiography has been shown to be excellent at making this diagnosis\(^{69}\) and at differentiating expansion from infarct extension.

Therefore, in patients with acute myocardial infarction, echocardiography can be used to make a rapid diagnosis, to stratify patients into high- or low-risk categories, to monitor serial changes, to look for associated injury such as right ventricular infarction, and to diagnose the complications of infarction.

**Chronic Ischemic Heart Disease**

Echocardiography is frequently useful in assessing patients with chronic ischemic heart disease for both prognostic and therapeutic reasons. Quantitative global and regional systolic function indexes include fractional shortening, fractional area change, and ejection fraction.

A left ventricular aneurysm is a finding of chronic ischemic heart disease, and echocardiography is a helpful test for this condition.\(^{70,71}\) Assessment of function of residual cardiac segments is one of the key factors in selecting medical or surgical therapy in these patients. Doppler echocardiographic evaluation of the tricuspid valve usually detects sufficient tricuspid regurgitation to allow an estimation of right ventricular and pulmonary artery systolic pressure.\(^{72}\) Doppler echocardiography is also helpful in assessing diastolic dysfunction (see below).

**Detection of Myocardial Ischemia**

Echocardiography in coronary artery disease does not image the primary lesion but only its consequences. Segmental wall motion abnormalities with lack of normal systolic thickening are therefore highly specific but not sensitive for latent underlying coronary artery disease.

Stress echocardiography, including exercise and pharmacologic stress, is more informative than studies at rest for the detection of myocardial ischemia.\(^{73-75}\) Stress echocardiography is discussed in more detail in the section on chest pain. Exercise examination may be warranted in patients with clinical evidence of coronary artery disease in those circumstances in which the standard graded exercise tests may prove nondiagnostic. Examples include conditions that produce abnormal rest echocardiographic results and others likely to produce false-positive stress echocardiographic results.

**Indications for Echocardiography**

**Rest Echocardiography**

**Class I**

1. Myocardial infarction (acute and chronic) when there is a specific question that can be resolved by echocardiography

**Class II**

1. Clinical evidence of coronary artery disease

**Class III**

1. Screening test for coronary disease in the general population

**Stress Echocardiography**

**Class I**

None

**Class II**

1. Whenever there is a high pretest probability that an indicated standard exercise stress test would be inadequate, nondiagnostic, or produce false-positive results

**Class III**

1. Routine screening of the general population without significant coronary risk factors
Disease of the Heart Muscle
Cardiomyopathies are heart muscle disorders of unknown etiology and are classified into three broad categories: dilated, hypertrophic, and restrictive forms. Echocardiography provides comprehensive morphologic assessment as well as characterization of the hemodynamics.

Dilated Cardiomyopathy
Echocardiography demonstrates dilation of ventricles, usually with normal wall thickness and reduced systolic function. Intracardiac thrombi may be detected. Doppler echocardiography is used to determine valvular regurgitation, pulmonary pressures, and diastolic dysfunction. Echocardiography is useful for serial follow-up as well as for assessment of the effectiveness of therapeutic interventions. It is useful in assessing and monitoring patients at risk of developing toxic cardiomyopathy, such as adriamycin cardiomyopathy.

Hypertrophic Cardiomyopathy
Echocardiography not only establishes the diagnosis of hypertrophic cardiomyopathy by revealing diffuse or localized areas of ventricular hypertrophy but also permits comprehensive morphologic assessment, specifically in patients with unusual areas of ventricular hypertrophy. Doppler techniques are utilized to assess severity of intraventricular obstruction at rest and with provocative maneuvers and to assess associated mitral regurgitation and diastolic filling abnormalities. In many patients, cardiac catheterization is no longer necessary to establish the diagnosis or for hemodynamic assessment. Echocardiography is useful for serial follow-up and for assessment of the effectiveness of therapeutic interventions.

Restrictive Cardiomyopathy
The two-dimensional echocardiographic features of restrictive cardiomyopathy are distinctive in that the ventricular chambers are usually normal in dimension and wall thickness and that frequently there is normal systolic function. However, the atria are markedly dilated, reflecting abnormal diastolic compliance of the ventricles. Doppler studies have shown characteristic ventricular inflow velocity profiles consisting of increased peak early flow velocity, reduced peak late flow velocity, and shortened deceleration time. Combined two-dimensional and Doppler echocardiographic examination may be helpful in patients with restrictive cardiomyopathy and may allow differentiation from constrictive pericarditis.

Indications for Echocardiography
Class I
1. Establishment of the morphologic diagnosis and assessment of hemodynamic status of patients with cardiomyopathies
2. Systemic illness associated with cardiac involvement, with clinical symptoms
3. Exposure to cardiotoxic agents

Class II
1. Systemic illness with high incidence of cardiac involvement but no clinical evidence of cardiac involvement
2. Clinical evidence suggesting cardiomyopathy
3. Family history of genetically transmitted cardiac disease

Class III
1. Systemic illness with low incidence of cardiac involvement and no clinical evidence of cardiac involvement

Pericardial Disease
One of the earliest applications of echocardiography was in the detection of pericardial effusion, and it remains the procedure of choice for evaluating this clinical problem. The pericardium usually responds to disease or injury by inflammation that may result in pericardial thickening, the formation of an exudate, or both, which in turn is manifested in the clinical picture of pericardial effusion, with or without tamponade or constriction. The anatomic evidence of pericardial disease and its effects on cardiovascular physiology can often be seen on M-mode, two-dimensional, and Doppler echocardiograms.

Pericardial Effusion
Pericardial effusions as small as 15 ml in volume can be detected by echocardiography, their location and configuration determined, and their size estimated in a semiquantitative fashion. Differentiation among types of pericardial fluid (blood, exudate, transudate, and others) cannot be made, but fibrous strands, tumor masses, and blood clots can often be distinguished. It should be remembered that all “echo-free” spaces adjacent to the heart are not the result of pericardial effusion.

Most pericardial effusions that require pericardiocentesis are located both anteriorly and posteriorly, but loculated effusions may occur, particularly after cardiac surgery. In such cases, echocardiography can define the distribution of the fluid so that the safest and most effective approach (subcostal, apical, or parasternal) can be planned for the pericardiocentesis.

Cardiac Tamponade
Enlarging pericardial effusions may cause cardiac tamponade. Although the diagnosis of cardiac tamponade can usually be made on the basis of the clinical evidence, when two-dimensional or Doppler echocardiography, alone or in combination, is combined with those clinical findings, the diagnosis is more certain, even in difficult cases. The elevated intrapericardial pressure in tamponade decreases the transmural pressure gradient between the pericardium and the right atrium and ventricle and increases the distending force necessary for ventricular filling.
Echocardiographic evidence of right atrial invagination (collapse) at end-diastole and right ventricular collapse in early diastole are signs of hemodynamic compromise.85–87 Right atrial collapse is a sensitive sign of tamponade but is not specific; diastolic right ventricular compression is more specific. Distension of the inferior vena cava that does not diminish on deep inspiration may also be seen and indicates an elevation of central venous pressure.88 Doppler flow studies have shown marked respiratory variation in transvalvular flow velocities, left ventricular ejection, and left ventricular isovolumetric times in patients with pericardial tamponade.89,90

**Increased Pericardial Thickness**

Increased echo density behind the posterior wall suggests pericardial thickening, but echocardiographic measurement of the precise pericardial thickness may be inaccurate.91 The causes of such thickening include fibrosis, calcification, and neoplasms, and it is usually not possible to differentiate the specific cause by echocardiography.

**Pericardial Tumors and Cysts**

Tumor in the pericardium is usually metastatic from the breast or lung, but other types occasionally occur.92 The clinical findings are typically a sizable pericardial effusion, at times leading to tamponade, but tumor may also present as single or multiple epicardial tumor nodules, as effusive-constrictive pericarditis, or even as constrictive pericarditis. The effects of radiation therapy on the tumor may further affect the pericardium, resulting in inflammation, effusion, or fibrosis.

Pericardial cysts are rare and are usually located at the right costophrenic angle. They are readily visualized by echocardiography and their cystic nature can be differentiated from that of a solid mass.93

**Constrictive Pericarditis**

In constrictive pericarditis there are such prominent pathologic and physiologic changes that echocardiographic abnormalities are always present, and in most cases there are multiple abnormalities. However, there is no single echocardiographic sign, or combination of signs, that is absolutely diagnostic of constrictive pericarditis. Some frequently seen findings are pericardial thickening, mild atrial enlargement with a normal-sized left ventricle, dilation of the vena cava, flattening of left ventricular endocardial motion in mid and late diastole, various abnormalities of septal motion, and premature opening of the pulmonary valve. Although these findings are nonspecific,94–98 when they are considered in the clinical context they can usually confirm the diagnosis of constrictive pericarditis. Doppler flow studies show marked changes in early mitral and tricuspid flow velocity at the onset of inspiration and expiration, and they provide useful information about physiology that can be combined with anatomic information from two-dimensional echocardiography.99,100 These Doppler findings are helpful in differentiating restrictive cardiomyopathy from constrictive pericarditis.

**Congenital Absence of the Pericardium**

In both total and partial absence of the pericardium, there are echocardiographic findings that are helpful in establishing the diagnosis.99,100

**Indications for Echocardiography**

**Class I**

Patients with clinical manifestations of or suspected pericardial disease.

**Class II**

Follow-up studies. The precise timing of follow-up studies is highly individualized, but they are usually done when there is other clinical evidence that the clinical status of the patient has changed or when further information is needed for guiding treatment.

**Class III**

None

**Cardiac Masses**

Echocardiography is a well-established technique for diagnosis of various types of intracardiac masses.53,54,58,101 Cardiac masses include tumors, thrombi, and vegetations. The most common primary cardiac tumor is a myxoma, and echocardiography has proved to be the diagnostic technique of choice for characterization of this tumor (location, attachment, size, appearance, and mobility). Echocardiography similarly is highly sensitive and specific for diagnosis of rhabdomyoma in patients with tuberous sclerosis. It also is useful for diagnosis of suspected cardiac metastases.

Intracardiac thrombi can be present in any of the cardiac chambers, and echocardiography is also the diagnostic technique of choice for localization and characterization of various intracardiac thrombi. With transesophageal echocardiography, thrombi in the left atrial appendage can be readily visualized. Echocardiography not only detects thrombus but also provides information pertaining to its size and shape and on whether it is sessile, pedunculated, or free-floating. Such characterization has important clinical and therapeutic implications.

**Indications for Echocardiography**

**Class I**

1. Evaluation of patients with suspected cardiac masses

**Class II**

None

**Class III**

None

**Diseases of the Great Vessels**

Echocardiography can be effectively utilized to visualize the entire thoracic aorta in most adults. Complete aortic visualization by combined transtho-
Aortic Aneurysm

Aneurysms of the ascending aorta can be characterized by transthoracic echocardiography. The aneurysm may be localized to one of the sinuses of Valsalva. With Doppler color flow imaging, rupture of an aneurysm in the sinus of Valsalva can be diagnosed and its communication with the receiving cardiac chamber can be documented. Annuloaortic ectasia as well as localized atherosclerotic aneurysms of the ascending aorta can be well visualized with use of the left as well as the right parasternal windows. Echocardiography is particularly well suited for serial follow-up of patients with ascending aortic aneurysms (especially in patients with Marfan’s syndrome) for determination of increase in the size of the aneurysm in a serial manner. Descending thoracic aortic aneurysms are difficult to visualize with the transthoracic approach. Transesophageal echocardiography is particularly suited for complete characterization of these aneurysms.

Aortic Dissection

Acute aortic dissection is a life-threatening emergency, and an early and prompt diagnosis is mandatory for appropriate patient care. The feasibility of using transthoracic echocardiography to visualize the intimal flap has been demonstrated by various investigators. Transesophageal echocardiography is currently regarded as a sensitive diagnostic procedure. In cooperative European studies the sensitivity and predictive accuracy of transesophageal echocardiography in aortic dissection were 98% and 99%, respectively. Limitations of single plane transesophageal echocardiography include the “blind spot” of the ascending aorta because of the interposition of the trachea and the difficulty of detecting branch vessel involvement. In addition to establishing the diagnosis and the extent of aortic dissection, echocardiography is extremely useful in delineating any associated complications, such as pericardial effusion, with or without tamponade, and aortic regurgitation, as well as in evaluating left ventricular size and function. Transesophageal echocardiography is also suited for serial follow-up of postoperative patients with aortic dissection.

The Great Veins

Echocardiography is a useful technique for visualizing the superior vena cava and for diagnosing various congenital and acquired abnormalities. A persistent left superior vena cava often can be imaged directly from the left supraclavicular fossa. Its connection, which is frequently to the coronary sinus, can be seen from a parasternal window as dilation. In some cases the connection to the coronary sinus can be better delineated with the use of contrast echocardiography with injection into the left arm. Other abnormalities, such as vena caval thrombosis, can also be diagnosed with combined use of echocardiographic and Doppler techniques. The proximal inferior vena cava can be readily visualized in nearly all patients and vena caval dilation and thrombosis or extension of tumors from the inferior vena cava to the right heart chambers have been diagnosed. The hepatic veins, their size, connection, and flow dynamics can be characterized with combined use of two-dimensional and Doppler echocardiography. Although visualization of all four pulmonary veins is not feasible in the majority of adult patients with use of the transthoracic approach, transesophageal echocardiography permits clear visualization of the pulmonary vein connections.

Indications for Echocardiography

Class I
1. Acute aortic root dilation or clinical suspicion of aortic dissection
2. First-degree relatives of patients with genetically transmitted connective tissue disorders

Class II
1. Chronic aortic root dilation
2. Suspected connective tissue disorder in athletes
3. All other suspected disease of the great vessel

Class III
None

Pulmonary Disease

As a general rule, patients who have primary pulmonary disease are not ideal subjects for echocardiographic examinations because the hyperinflated lung is a poor conductor of ultrasound. Despite these technical limitations, transthoracic echocardiography can still be very informative in some patients with primary lung disease. The usual precordial or parasternal windows are frequently unavailable in patients with hyperinflated lungs. However, in these same patients the diaphragms are frequently lower than normal. Thus, the subcostal or subxyphoid transducer position can offer an ideal window for echocardiographic examinations. For those few patients in whom transthoracic and subcostal echocardiographic windows are totally unavailable, there is now the transesophageal approach, which provides an unobstructed view of the heart in patients with lung disease. As a result, with use of one examining technique or another, almost all
Indications for Echocardiography

Class I
1. Unexplained pulmonary hypertension
2. Pulmonary emboli and suspected clots in the right atrium or ventricle

Class II
1. Lung disease with clinical suspicion of cardiac involvement
2. Pulmonary emboli

Class III
None

Hypertension

In adults, hypertension is the most common cause of concentric left ventricular hypertrophy and congestive heart failure. Data from the Framingham Study have shown that both the risk of cardiac failure and mortality are increased in patients with electrocardiographic (ECG) criteria of left ventricular hypertrophy compared with patients who have hypertension and normal findings on electrocardiography. More recently, in the same population, an association of echocardiographic left ventricular mass with coronary heart disease events has been demonstrated that is independent of traditional coronary risk factors.

Echocardiography must now be considered the noninvasive procedure of choice in evaluating the cardiac effects of systemic hypertension. M-mode and two-dimensional echocardiographic estimates of left ventricular mass are more sensitive and specific than either the ECG or chest roentgenogram in diagnosing the presence of left ventricular hypertrophy, and these estimates have been shown to correlate accurately with left ventricular mass at necropsy. These techniques have been used to evaluate the relation of left ventricular mass to rest and exercise blood pressure as well as to multiple other physiologic variables. Echocardiography can also be used to evaluate systolic and diastolic properties of the left ventricle, such as speed and extent of contraction, end-systolic wall stress, and the rate of ventricular filling throughout diastole.

A decrease in left ventricular mass in hypertensive patients through control of blood pressure or weight loss has been demonstrated in several studies. The need for serial quantitation of left ventricular mass in assessing drug therapy for hypertension is underscored by the poor association of blood pressure control with regression of left ventricular hypertrophy. However, although preliminary data are encouraging, more study will be required to prove that regression of left ventricular hypertrophy alters cardiac morbidity and mortality and that echocardiography is a cost-effective method for both the detection and follow-up evaluation of the large number of patients with hypertension.

**Indications for Echocardiography**

**Class I**
1. Hypertension with clinical evidence of heart disease

**Class II**
1. Hypertension without signs or symptoms of heart disease

**Class III**
1. Borderline hypertension without signs or symptoms of heart disease

**Dyspnea**

Dyspnea is a common cardiac symptom in patients with lung or cardiac disease. The role of echocardiography in patients with lung disease is addressed in the section on pulmonary disease. Impairment of left ventricular systolic function is the most common cause of heart failure. Recently, it has been recognized that up to one third of patients who have dyspnea of cardiac origin have abnormalities of left ventricular diastolic function as the cause of their symptoms. Diastolic abnormalities severe
enough to cause symptoms without systolic dysfunction are most commonly seen in elderly patients who have left ventricular hypertrophy and a history of hypertension. Evidence suggests that diastolic abnormalities precede detectable left ventricular systolic dysfunction and are associated with increased cardiovascular morbidity.15 Since treatment to improve systolic cardiac performance may not benefit or may even be detrimental to diastolic function, a noninvasive method to help determine the cause of cardiac dyspnea should ideally be able to evaluate both systolic and diastolic cardiac performance.

With its ability to assess cardiac anatomy, chamber sizes, and myocardial and valvular function, echocardiography is a powerful noninvasive tool to investigate possible causes of dyspnea of cardiac origin. Echocardiography is a good method for detecting and quantitating left ventricular hypertrophy111 and has been shown to be comparable to cardiac catheterization for assessing the severity of valvular and congenital heart disease. In most patients who have pulmonary and tricuspid valve regurgitation, estimates of pulmonary artery systolic and diastolic pressures can be made with use of the modified Bernoulli equation.116 Abnormalities of left ventricular diastolic filling can be assessed by either M-mode or Doppler echocardiography; the results of both have been compared favorably with those of angiographic17 and radionuclide118 techniques. Therefore, echocardiography can be recommended for the evaluation of dyspnea in patients with clinical findings suggestive of significant coronary, valvular, or hypertensive heart disease. However, because dyspnea is a common symptom in patients without organic heart disease, echocardiography cannot be recommended as an initial diagnostic study in patients with normal blood pressure and physical examination.

Indications for Echocardiography

Class I
None

Class II
1. Dyspnea with clinical evidence or suspicion of heart disease
2. Unexplained dyspnea

Class III
1. Dyspnea without clinical evidence of heart disease, pulmonary hypertension, or significant lung disease
2. Hyperventilation syndrome

Chest Pain

There are many cardiac causes of chest pain. The most common clinical entity that presents as chest pain is coronary artery disease (see section on ischemic heart disease). Hypertrophic cardiomyopathy, aortic stenosis, aortic dissection, mitral valve prolapse, and even acute pulmonary embolism can present with fairly distinctive and diagnostic findings on echocardiography.

The role of echocardiography in patients whose chest pain raises suspicion of coronary artery disease is being defined. In patients with ischemic heart disease, the role of echocardiography can be similar to that of electrocardiography. A person can have extensive coronary artery disease; however, if the coronary artery obstructions have not induced any malfunction of the ventricle, then the echocardiographic findings can be completely normal at rest. If the patient has had a previous known or unknown myocardial infarction, then the rest echocardiogram will help to confirm or evaluate that clinical event. Silent ischemia, even to the point of myocardial infarction, is not uncommon. A rest echocardiogram can detect a previously unknown myocardial infarction.

A patient may have coronary artery disease that is not producing angina but is interfering with blood flow so that myocardial function is impaired even in the resting state, as in hibernating myocardium. An echocardiogram can detect regional wall dysfunction and, theoretically, can be diagnostically useful in patients who are suspected of having coronary artery disease. The frequency with which a rest echocardiogram will be informative in such patients is still unknown.

The majority of patients with ischemic heart disease will have normal findings on rest echocardiogram. If there is clinical evidence to suggest a prior myocardial infarction and confirmation or evaluation of the patient's global or regional ventricular function is desired, then the need for a rest echocardiogram can be more easily justified.

As with the ECG, a patient with coronary artery disease may exhibit an echocardiographic abnormality only when ischemia can be induced with some form of stress testing. Exercise is a commonly used form of stress to bring out ECG abnormalities, ventricular wall motion abnormalities, or perfusion defect abnormalities that are not present at rest. A similar strategy is possible with echocardiography. By inducing ischemia, either with exercise,119-121 pharmacologic stress,122,123 or pacing,124 myocardial dysfunction can be produced that can be recognized on the echocardiogram as a wall motion abnormality. The feasibility of using echocardiography to detect ischemia-induced wall motion abnormalities has been well demonstrated.73,125,126

A major issue regarding the use of stress echocardiography is its practicality. However, with advances and improvements in echocardiographic techniques, echocardiographic equipment, and digital recording methods, the majority of patients can be successfully examined with stress echocardiography.127-129

Stress echocardiography is combined with routine exercise testing and offers supplemental information.130 The sensitivity and specificity of the combined tests are improved, especially in certain subsets of patients.131,132 The addition of echocardiography, however, substantially increases the cost of a routine stress test. For any patient whose physician believes that ECG stress testing with clinical eval-
uption alone needs to be supplemented with a test of ventricular function, stress echocardiography is a reasonable option.

**Indications for Echocardiography**

**Class I**
1. Chest pain with clinical evidence of valvular, pericardial, or primary myocardial disease

**Class II**
1. Known or suspected coronary artery disease

**Class III**
1. Noncardiac chest pain

See previous section on chest pain for discussion of stress echocardiography.

**Murmurs**

Cardiac auscultation remains the most widely used method of screening for heart disease. In valvular and congenital forms of heart disease a murmur is usually the major evidence of the abnormality. Heart murmurs are produced by turbulent blood flow and are often signs of stenotic or regurgitant valve disease or of acquired or congenital defects. However, many murmurs are “innocent” and of no functional significance. When the characteristic findings of an individual murmur are considered together with other clinical data from the physical examination, the chest roentgenogram, and the ECG, the correct diagnosis can usually be established. However, echocardiography provides complementary information about cardiac structure and function as well as about blood flow. In some patients the echocardiogram is the only noninvasive method capable of identifying the cause of a heart murmur.

In the evaluation of heart murmurs the purposes of performing an echocardiogram are to:

- Define the primary lesion and judge its severity
- Detect coexisting abnormalities
- Detect lesions secondary to the primary lesion
- Evaluate cardiac function
- Establish a reference point for future observations
- Reevaluate the patient after an intervention

Echocardiography has replaced cardiac catheterization as the definitive study for many types of valvular and congenital heart disease and has become the method of choice for serial observation of patients with these conditions because it is accurate and painless. Furthermore, in many patients surgery can be performed without cardiac catheterization as long as the status of the coronary arteries is not a concern.

As valuable as echocardiography may be, the basic cardiovascular evaluation is still the most appropriate method to screen for cardiac disease and will establish many clinical diagnoses. Accordingly, echocardiography should not be used to replace the cardiovascular examination. Echocardiography can be helpful to determine the etiology and judge the severity of lesions.

**Indications for Echocardiography**

**Class I**
1. An organic murmur in a patient with cardiopulmonary symptoms
2. A murmur in an asymptomatic patient if the clinical features indicate at least a moderate probability that the murmur is organic

**Class II**
1. A murmur in an asymptomatic patient in whom there is low probability of heart disease but in whom the diagnosis of heart disease cannot be reasonably excluded by the standard cardiovascular clinical evaluation

**Class III**
1. A typically innocent murmur in an asymptomatic patient without any other reason to suspect heart disease

**Ischemic Syndromes**

By clinical criteria, cerebral embolism originating from the heart is believed to account for approximately 15% of all ischemic strokes. Using aggregate clinical data, the main conditions believed to be associated with the formation of intracardiac thrombus by percentage are nonhemorrhagic atrial fibrillation (45%), acute myocardial infarction or left ventricular aneurysm (15%), rheumatic mitral stenosis (10%), and prosthetic aortic or mitral valves (10%). Mitral valve prolapse, idiopathic dilated cardiomyopathy, valvular vegetations, calcific aortic stenosis, patent foramen ovale, and left atrial myxoma are other possible sources of cerebral embolism.

Two-dimensional echocardiography is recognized as a sensitive and specific noninvasive method for the diagnosis of intracardiac thrombi. Although it can confirm the presence of abnormalities associated with cerebral emboli, an abnormal result cannot rule out the heart as an embolic source. Two-dimensional echocardiography has a sensitivity of 75–95% and a specificity of approximately 85% for detecting left ventricular thrombi that are >4 mm in diameter. Two-dimensional echocardiography has a sensitivity and specificity of approximately 70–90% for identifying thrombi in the body of the left atrium, but the sensitivity for clots located in the left atrial appendage is <15%. More recently, transesophageal echocardiography has been reported to have an increased sensitivity for diagnosing clots in the left atrium and left atrial appendage that approaches 95%. This is notable since the appendage is the most frequent site for clot formation.

Previous studies have established that at least one third of patients with ischemic strokes have evidence of cardiac disease by history, physical examination, chest x-ray film, or ECG. Because this incidence is higher than that estimated for cerebral embolism from a cardiac source, a causal relation between the two cannot be assumed. Therefore, echocardiography appears warranted in patients with stroke who...
have clinical evidence for heart disease. Echocardiography is frequently performed in stroke patients <45 years of age, regardless of clinical findings, to rule out in-atrial communications or mitral valve prolapse as a possible source of embolism.\(^{137}\) If Doppler echocardiography and color flow imaging do not detect a shunt, a two-dimensional contrast bubble study is frequently performed in these patients because it is the most sensitive method for detecting small in-atrial shunts. Routine echocardiography does not appear warranted as an initial diagnostic study in the majority of stroke patients >45 years old who do not have clinical evidence for cardiac disease because these patients rarely have echocardiographic findings associated with peripheral emboli.\(^{138}\)

**Indications for Echocardiography**

**Class I**
1. Patients with cerebral embolism and clinical evidence of heart disease
2. Patients <45 years of age with a cerebrovascular event

**Class II**
1. Patients >45 years of age with suspicion of cardiogenic brain embolism but without clinical evidence of heart disease

**Class III**
1. Patients with known noncardiac causes of the neurologic disorder

**Syncope**

Determination of the etiology of a syncopal episode can be a difficult clinical problem. Despite a careful history and physical examination, it is often not possible to distinguish syncope of cardiac origin from syncope due to other causes.\(^{139-141}\) Furthermore, because numerous cardiac-related mechanisms can cause a sudden decrease in cerebral perfusion, a diagnostic approach for ordering cardiac studies that is cost-effective should be based both on the features of the individual case and on an awareness of the most common pathophysiologic mechanisms that cause syncope.\(^{139,141}\)

Syncope of cardiac origin is most commonly related to vasodepressor reflexes or cardiac bradycardia or tachycardias.\(^{140,142}\) Uncommon causes of cardiac syncope include severe aortic stenosis, hypertrophic obstructive cardiomyopathy, or atrial myxoma. Therefore, echocardiography should not be performed as an initial diagnostic step in patients with syncope unless the physical examination suggests the presence of a pathologic murmur or valvular heart disease. The decision on performing echocardiography in patients who still have unexplained syncope after evaluation for cardiac arrhythmias should be individualized with the knowledge that the yield of the test is expected to be low.

**Indications for Echocardiography**

**Class I**
1. Patients with a murmur suggestive of significant valvular heart disease or obstructive cardiomyopathy

**Class II**
1. Patients without clinical evidence of heart disease and normal findings on evaluation for noncardiac causes of syncope

**Class III**
1. Patients with known noncardiac causes of syncope

**Peripheral Emboli**

Patients with documented peripheral emboli involving major arteries should undergo echocardiographic study regardless of clinical findings because the heart is the only likely source for such large emboli. Transesophageal echocardiography improves the ability to detect left atrial thrombi, especially in the appendage, and to detect venous thrombi that have entered the central or peripheral circulation through a patent foramen ovale. Echocardiography should also be performed to look for evidence of endocarditis in patients with fever who have peripheral arterial emboli or embolic findings in the extremities or fundi. Transesophageal echocardiography improves the detection of ascending or descending aortic abnormalities, such as aneurysms, dissecting hematomas, or ulcerations.

**Indications for Echocardiography**

**Class I**
None

**Class II**
1. Patients with peripheral emboli involving major arteries
2. Patients with evidence of infection and peripheral emboli

**Class III**
None

**Arrhythmias and Palpitation**

Echocardiography is useful in defining the cardiac milieu in which arrhythmias occur and therefore is a useful adjunct in the management of cardiac arrhythmias.

Arrhythmias associated with palpitation can be divided into several types. Minor arrhythmias, such as isolated premature contractions, can occur without structural heart disease and further evaluation is not required. Although an echocardiogram may reveal a minor abnormality such as mitral valve prolapse in such patients, the diagnostic yield is low and unlikely to change management.

Some arrhythmias are frequently associated with underlying organic heart disease or may predispose the patient to hemodynamic deterioration. Atrial fibrillation and flutter are examples of arrhythmias in which the echocardiogram frequently is appropriate to assess such an underlying disorder. A specific common use of the ultrasound examination is to
quantitate left atrial size in patients with atrial fibrillation before considering cardioversion.

Certain arrhythmias are more prone to deteriorate into unstable or life-threatening forms. In these patients, there is an important relation between the underlying substrate to sustain the arrhythmia and the need for special treatment. Increasing degrees of cardiac abnormality or dysfunction are associated with greater need to treat specific arrhythmias. Echocardiography is an excellent tool for assessing the presence and degree of cardiac dysfunction and therefore provides essential information for the management of these patients. Furthermore, the assessment of ventricular function may also influence the choice of antiarrhythmic agent, as some have significant negative inotropic effects.

In patients with arrhythmias capable of hemodynamic compromise or life-threatening potential, an echocardiogram can serve as an integral part of the cardiac evaluation.

**Indications for Echocardiography**

Class I

1. Arrhythmias with evidence of heart disease
2. Family history of genetic disorder associated with arrhythmias
3. Arrhythmias commonly associated with, but without evidence of, heart disease
4. Atrial fibrillation or flutter

Class III

1. Palpitation without evidence of arrhythmias
2. Minor arrhythmias without evidence of heart disease

**Edema**

The causes of peripheral edema are numerous and include both cardiogenic and noncardiogenic etiologies. Echocardiographic study could be recommended in any patient who has evidence for an elevated central venous pressure, significant valvular or coronary artery disease, cor pulmonale, or pulsus paradoxus. Uncommon cardiac disorders that might be detected by echocardiography in patients with abnormal findings on physical examination include constrictive pericarditis, restrictive cardiomyopathy, and amyloid heart disease. Echocardiography cannot be routinely recommended in patients with mild peripheral edema who have no evidence for an increase in central venous pressure or clinical findings of heart disease because the diagnostic yield in such patients is expected to be low. In patients in whom central venous pressure cannot be estimated with certainty on physical examination, echocardiographic evaluation of respiratory collapse of the inferior vena cava diameter can determine if the central venous pressure is elevated.

**Indications for Echocardiography**

Class I

1. Edema with other evidence of cardiac disease

Class II

1. Edema without evidence of cardiac disease

Class III

1. Edema of noncardiac origin

**Evaluation of Ventricular Function**

**Global Systolic Function**

Echocardiographic methods can be used to define several indexes of global left ventricular systolic function, including M-mode measurements (fractional minor axis shortening, mitral-septal separation), two-dimensional measurements (fractional area change, ejection fraction), Doppler measurements (peak aortic flow velocity and acceleration), and combined indexes (cardiac output, stroke volume). The M-mode indexes are prone to significant errors in patients with inadequate acoustic access, abnormally shaped ventricles, extreme dilation, and segmental wall motion abnormalities. Two-dimensional data, including linear (fractional shortening), area-based (fractional area change), and volume-based (ejection fraction) measurements, correlate well with independent standards such as chest roentgenogram findings and radionuclide ventriculography and are, therefore, useful in patients in whom studies of adequate quality can be obtained. Doppler indexes may be useful in the serial evaluation of a given patient and less useful for comparisons among patients.

Global right ventricular systolic function in adults is difficult to quantitate by echocardiography because of the frequent difficulty in obtaining accurate geometric information concerning the unusually shaped right ventricular chamber. Useful qualitative assessment of right ventricular size and function may be obtained with echocardiography. In children, useful quantitative measures of right ventricular function may be made. Doppler-derived data may be of more use, particularly in serial studies of a patient or in the determination of pulmonary to systemic flow ratios in patients with shunt lesions.

**Regional Left Ventricular Function**

Echocardiography is an excellent technique for determining regional contractile function of the left ventricle. The attributes of high spatial and temporal resolution and the ability to define regional wall thickening, as well as endocardial excursion, make echocardiography extremely useful in defining regional dysfunction due to ischemic disease, cardiomyopathy, contusion, and other disorders. Considerable controversy still surrounds the optimal method of analyzing echocardiographic data to extract information on regional left ventricular function, but virtually all carefully tested methods have yielded data useful in the clinical examination of regional function.

Right ventricular regional function may also be assessed by echocardiography, but the difficulties mentioned, coupled with the different mechanisms of contraction of the right and left ventricles, combine
to make echocardiographic data on regional right ventricular contraction abnormalities less quantitatively accurate than data for the left ventricle. Nevertheless, clinically useful evidence of regional right ventricular dysfunction, such as that due to infarction, can be garnered from echocardiograms.

**Diastolic Left Ventricular Function**

Recent interest in the noninvasive evaluation of diastolic function has produced a large number of indexes based on information from M-mode, two-dimensional, or Doppler echocardiographic studies. In addition, echocardiography has been used experimentally to obtain complex measures of regional diastolic stress-strain characteristics. Unfortunately, virtually all clinically available indexes of diastolic function (including echocardiography) are of somewhat limited usefulness. This is due to several factors, including 1) the complex nature of diastolic function, which differs in its mechanism in early diastole (when active cellular relaxation occurs) from that in late diastole (when passive muscle material properties are important); 2) the load dependence of the commonly described echocardiographic variables; and 3) the lack of careful validation of most of the variables by comparison to appropriate independent standards. For these reasons, echocardiographic indexes of diastolic function find their greatest usefulness in the serial examination of a given patient but less usefulness in comparing patients or in identifying the degree of diastolic dysfunction.

**Indications for Echocardiography**

**Class I**
1. To evaluate global left ventricular function
2. To evaluate regional left ventricular function
3. Qualitative right ventricular function

**Class II**
1. Diastolic left ventricular function

**Class III**
1. Quantitative right ventricular function (except in children)

**Screening**

Screening tests for cardiac disease can be valuable but may not be very cost-effective. The intent of screening tests is to find those persons who have a serious, potentially treatable abnormality but are unaware of the problem. Although waiting for the patient to have some sort of complaint before investigating the possibility of a treatable illness is common, significant abnormalities can occur in the asymptomatic patient that at times lead to severe organ damage.

The criteria for an ideal screening test include being accurate, harmless, rapid, painless, and inexpensive. Echocardiography meets some of these criteria. The examination is painless and accurate, and as best can be determined, it is harmless. The test is relatively rapid, depending on the information desired. Unfortunately, echocardiography is not inexpensive. The examination is less costly than some other sophisticated procedures, such as invasive testing, or other noninvasive tests, such as nuclear stress testing, computed tomography scanning, or nuclear magnetic resonance imaging. On the other hand, it is considerably more expensive than an ECG or a chest roentgenogram. There are certain groups of persons who might benefit from a routine echocardiogram because of a relatively high risk of cardiac disease; an example might be patients with a family history of inheritable cardiovascular disease.

In general, echocardiography is too costly to be considered as a routine screening test for the general population. There may be certain subgroups of persons for whom the cost of this procedure may be warranted, provided that there is a reasonable likelihood that the results of the test will influence an individual patient’s management or prognosis.

**Indications for Echocardiography**

**Class I**
1. Patients with a family history of cardiovascular disease that is clearly inheritable

**Class II**
1. Competitive athletes

**Class III**
1. General population

**Use of Echocardiography in the Pediatric Patient**

Echocardiography has become the definitive diagnostic method for the recognition and assessment of congenital and acquired heart disease in children. Its use has eliminated the need for invasive or other noninvasive studies in some and decreased the frequency and improved the timing and performance of invasive studies in other patients. Serial evaluations in some conditions improve medical or surgical management. Echocardiographic evaluation reduces trauma to the child with insignificant cardiac abnormality and provides reassurance to the family. The outcome is improved for those patients with significant cardiac abnormality by guiding management decisions and providing early education and support for the family.

Although congenital heart disease is the most common type of cardiovascular disease recognized in the pediatric population, the appearance of Kawasaki disease and human immunodeficiency virus–related myocarditis and the recrudescence of rheumatic heart disease have increased the prevalence of inflammatory diseases in this age group. Cardiomyopathy, whether familial, acquired, or idiopathic, is also commonly seen. Additionally, a variety of serious cardiopulmonary diseases occur in neonates.

Serial follow-up studies are frequently utilized to follow the late cardiovascular adaptation to surgical repair or palliation, to demonstrate the recurrence of abnormalities, and to provide new knowledge about the relative benefits of new surgical techniques, such
as the arterial switch procedure for transposition of the great arteries. such information serves retrospectively to enlighten the clinician in the selection of the correct interventional approach and its timing. For these reasons, echocardiography provides improved outcome and lowered health care costs by the streamlined use of medical resources.

Congenital Heart Disease

Two-dimensional echocardiography provides essential structural information in all forms of cardiac and great vessel disease in pediatric patients. Doppler echocardiography provides important physiologic information that, when combined with anatomic data, is sufficient to guide therapeutic management in some diagnostic categories. Serial examinations allow tracking of hemodynamic changes such as those occurring during the transition phase from fetal to newborn and infancy periods. Echocardiography provides clinical information for the initial evaluation, before medical or surgical intervention, during medical or surgical intervention, and in postoperative patients.

Perinatal physiologic changes often mask or obscure the presence of hemodynamically important cardiovascular lesions. Echocardiography allows early recognition of lesions in which either the pulmonary or the systemic circulation is dependent on the patency of the ductus arteriosus. Definitive diagnosis in these lesions before ductal closure may prevent death or severe morbidity. Infants with a loud murmur, signs of congestive heart failure, cyanosis, or failure to thrive have a high probability of significant heart disease and should undergo echocardiographic evaluation.

Congenital heart disease in the child or adolescent commonly presents as an asymptomatic heart murmur; nevertheless, the cardiac murmurs of this age group are more commonly functional than pathologic. History and physical examination by a skilled observer are usually sufficient to distinguish functional from pathologic murmurs. In the presence of ambiguous clinical findings, echocardiography can demonstrate the presence or absence of abnormalities such as bicuspid aortic valve, mildly obstructive subaortic stenosis, mitral valve prolapse, or cardiomyopathy. Such determination directs a need for further follow-up or endocarditis prophylaxis, or both. For patients with clinical findings of hemodynamically significant heart disease, anatomic and physiologic data provided by serial two-dimensional and Doppler echocardiography may provide a definitive diagnosis and allow the most efficient timing of invasive or interventional procedures.

Echocardiography may be employed in concert with cardiac catheterization to limit the quantity of radiographic contrast material and to direct interventional maneuvers.

Intraoperative echocardiography has been utilized to provide timely information about the success of septal defect closure and valve palliation. In some lesions, the ability to scan the heart by direct transducer placement on the heart surface or by transesophageal echocardiography allows the patient to undergo surgical repair without prior cardiac catheterization.

Indications for Echocardiography

Class I
1. Cyanosis, respiratory distress, abnormal arterial pulses, or cardiac murmur in a neonate
2. Loud or abnormal murmur or other abnormal cardiac finding in an infant or older child
3. Failure to thrive in the presence of an abnormal or unusual cardiac finding
4. Presence of a syndrome associated with cardiovascular disease and dominant inheritance or multiple affected family members
5. Presence of a syndrome associated with heart disease, with or without abnormal cardiac findings, for which an urgent management decision is needed
6. Cardiomegaly on chest radiograph
7. Dextrocardia, abnormal pulmonary, or visceral situs
8. Most ECG abnormalities
9. Postoperative congenital or acquired heart disease
10. Postcardiac or cardiopulmonary transplant

Class II
1. Murmur of uncertain etiology
2. Failure to thrive in the absence of definite abnormal clinical findings
3. Clinical findings of small ventricular septal defect after the neonatal period
4. Presence of a syndrome associated with a high incidence of congenital heart disease for which there are no abnormal cardiac findings and no urgency of management decisions

Class III
1. An asymptomatic heart murmur in a child or adolescent that is positively identified by an experienced observer as functional or innocent

The common categories of congenital heart disease are summarized as follows:

<table>
<thead>
<tr>
<th>Diagnostic Groups</th>
<th>Information Provided</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Left to right shunts</td>
<td>Presence, position, configuration, and size of defect, direction of flow and gradient across defect, pulmonary/systemic flow, ventricular compensation, associated lesions</td>
</tr>
<tr>
<td>2. Obstructive lesions</td>
<td>Location, configuration, and severity of obstruction, ventricular compensation, associated lesions</td>
</tr>
<tr>
<td>3. Regurgitant lesions</td>
<td>Valve configuration, assessment of severity, chamber dilation, ventricular compensation, associated lesions</td>
</tr>
</tbody>
</table>
4. Venous connections
Location and connections of all systemic and pulmonary veins, assessment of left-to-right and right-to-left shunts, associated lesions
194,196

5. Conotruncal abnormalities
Position of great arteries, location of ventricles and ventricular septal defect, nature of subaortic obstruction, great artery anatomy, associated lesions, ventricular compensation

6. Coronary anomalies
Origin, size, and flow in coronary arteries, ventricular compensation

7. Complex lesions
Cardiac segmental analysis of situs and connections, size and location of all cardiac chambers, atrioventricular valve morphology, subarterial and arterial obstruction, interatrial and interventricular communications, venous and great artery anatomy, ventricular compensation

Inflammatory Diseases
Kawasaki disease may result in abnormalities in the proximal or distal coronary circulation, myocarditis, myocardial infarction, and pericardial effusion. Baseline and serial evaluations by echocardiography are recommended in all patients with clinical stigmata of this disease because echocardiographic findings influence management decisions. Since long-term abnormalities of the coronary arteries have been noted after resolution of initial aneurysms, these patients may require lifelong follow-up.

Children with human immunodeficiency virus infection acquired during the fetal or newborn period have an aggressive form of the disease with early and prominent myocardial involvement and therefore should have a baseline study and serial follow-up studies as indicated by the appearance of tachycardia, congestive heart failure, and respiratory distress.

There is a resurgence of acute rheumatic fever in the United States. Newer diagnostic criteria include echocardiographic assessment of mitral valve function, ventricular function, and pericarditis. Echocardiography is an important component of the diagnostic evaluation of children with fever, new cardiac murmur, migratory polyarthritis, and chorea.

See previous sections for discussion of pericardial disease and infective endocarditis.

Indications for Echocardiography
Class I
1. Baseline and follow-up studies on all pediatric patients with suspected or documented Kawasaki disease, human immunodeficiency virus infection, or rheumatic fever

Class II
1. Follow-up examinations after occurrence of acute rheumatic fever in patients with normal cardiac findings

2. Long-term follow-up studies in patients with Kawasaki disease who have no coronary abnormalities during the acute phase of the disease process

Class III
None

Myocardial Disease
Echocardiography provides diagnostic information in patients with hypertrophic, congestive, and infiltrative cardiomyopathy, viral myocarditis, toxic cardiomyopathy, and idiopathic cardiomyopathy. Patients receiving anthracycline or other cardiotoxic agents should have baseline and serial follow-up studies. Echocardiographic assessment of patients with renal disease provides guidance in management of hemodialysis and hypertensive medications. Echocardiography is useful in detecting hypertrophic cardiomyopathy and in determining the presence of subaortic and subpulmonary obstruction, mitral insufficiency, and diastolic compliance abnormalities. Echocardiography is useful in screening family members in all types of cardiomyopathy associated with a dominant or recessive pattern of inheritance.

Indications for Echocardiography
Class I
1. Patients with a family history of genetically transmitted myocardial disease, with or without abnormal cardiac findings

2. Patients with clinical evidence of myocardial disease

3. Baseline and serial examinations of patients receiving cardiotoxic therapeutic agents

4. Patients with severe renal disease and an abnormal cardiac finding

5. Recipients and donors undergoing evaluation for cardiac transplantation

Class II
None

Class III
None

Arrhythmia
Rhythm abnormalities in children may be associated with Ebstein's anomaly of the tricuspid valve, cardiac tumor, cardiomyopathy, mitral valve prolapse, glycogen storage disease, or stimulation from migrated central catheters.

Echocardiography is an important component in evaluation of these patients. Mild rhythm disturbances, such as sinus arrhythmias and low grade supraventricular ectopic beats, or brief and infrequent runs of supraventricular tachycardia, are rarely associated with cardiac pathology. In addition, atrial contraction can sometimes be characterized by echocardiography when findings are obscured on the surface ECG.

Indications for Echocardiography
Class I
1. Arrhythmia requiring treatment
2. Arrhythmia in the presence of an abnormal cardiac finding
3. Arrhythmia in a patient with a family history of a genetically transmitted cardiac lesion associated with arrhythmia, such as tuberous sclerosis or rhabdomyoma
4. Appearance of arrhythmia in a patient with a central venous catheter

Class II
1. Recurring arrhythmia not requiring treatment in the presence of normal findings on cardiac examination

Class III
1. Sinus arrhythmia or isolated extrasystoles in a child with otherwise normal findings on cardiac examination and no family history of a genetically transmitted abnormality associated with arrhythmia

Cardiopulmonary Diseases

A variety of cardiopulmonary diseases are seen in the neonates. Premature infants may have respiratory failure based on a combination of processes: lung immaturity, hyaline membrane disease, persistence of the ductus arteriosus, inflammatory disease, or congenital heart disease. Echocardiography determines the patency of the ductus arteriosus, direction and degree of shunting at the ducal level, and ventilricular compensation for left ventricular volume overload. An indirect assessment of pulmonary artery hypertension can also be made. Echocardiographic study also rules out a ductal-dependent lesion when pharmacologic or surgical ductal closure is planned.

Term newborn infants with primary pulmonary hypertension of the newborn (persistent fetal circulation) may present with or without associated meconium staining or aspiration. Differentiation of this entity from cyanotic heart disease often requires echocardiography. In addition to excluding structural abnormalities, Doppler echocardiography provides additional information about atrial and ductal shunting, pulmonary artery pressure, and ventricular function. Serial studies are useful in monitoring the therapeutic response. In patients with severe disease progressing to extracorporeal membrane oxygenation, this information is useful in assessing the contribution of extracorporeal circulation to ventricular output, the appearance and course of “stunned myocardium,” and flow across the ductus arteriosus. Children with upper airway disease, cystic fibrosis, human immunodeficiency virus infection, and other chronic immunologic disorders may have clinical or ECG evidence of pulmonary artery hypertension. As discussed in a previous section, echocardiography provides indirect documentation of pulmonary artery hypertension and estimation of severity. Follow-up studies reflect response to therapy and are useful in guiding management.

Indications for Echocardiography

Class I
1. Respiratory distress or cyanosis in the newborn

Class II
1. Baseline study of patients with cystic fibrosis and no findings of cor pulmonale
2. Newborn, premature, or term infants with respiratory distress who respond rapidly to initial pulmonary management

Class III
None

Thromboembolic Disorders

Stroke and other manifestations of thromboembolism that occur in childhood may result from intracardiac thrombus, tumor, or vegetation. In some groups of patients, long-term indwelling catheters in the central veins or atria may predispose to thrombus formation or infection. Because children have a lower incidence of distal cardiovascular disease as a cause of stroke or loss of pulse, the yield of echocardiography to rule out an intracardiac cause may be somewhat higher than for adults.

Indications for Echocardiography

Class I
1. Thromboembolic event in an infant, child, or adolescent

Class II
None

Class III
None

Fetal Echocardiography

Widespread use of general fetal ultrasound examinations among women receiving prenatal care has resulted in increased referrals for specific cardiac analysis. Definition of fetal cardiac structures is currently possible at 10–12 weeks’ gestation, with the use of vaginal probes with high resolution transducers. By 16–18 weeks’ gestation, accurate segmental analysis of cardiac structure is possible with a conventional transabdominal approach. Doppler examination provides important information about blood flow across the cardiac valves, great arteries, ductus arteriosus, and umbilical arteries. A general fetal ultrasound examination usually includes a four-chamber or inflow view of the fetal heart. This view is sensitive to abnormalities of the inflow portions of the heart but is insensitive to some septal defects, outflow lesions, and conotruncal abnormalities. Patients are referred for specific fetal echocardiographic examination because of an abnormality of structure or rhythm noted on ultrasound examination or because the patient is in a high-risk group for fetal heart disease. Early recognition of fetal heart disease allows the opportunity for transplacental therapy, as in the case of arrhythmias. When a potentially life-threatening cardiac anomaly is found, the delivery can be planned at a tertiary care center where supportive measures can be instituted before severe hypoxia, shock, or acidosis ensues. Education of the
parents can be initiated early so that complex therapeutic choices can be reviewed and informed consent obtained. When the fetal heart appears normal, the family may be reassured.

Diagnostic difficulties may arise because of modulation of the anatomic and physiologic presentation of certain lesions by the fetal circulation and dramatic changes in the heart and great vessels that may occur throughout gestation. As an example, the severity of pulmonary stenosis cannot be assessed by quantification of valve gradient because of the variability in right ventricular output and the condition of the ductus arteriosus. The outcome of fetal heart disease is often suggested only after serial studies to determine growth of cardiac chambers and vascular structures and changes in blood flow patterns. The spectrum of congenital cardiac lesions is broader than that seen in the neonates and infants because of the presence of noniviable subcategories of disease. The maternal history of a given congenital heart lesion recognized prenatally is not always the same as that of one diagnosed postnatally. A knowledge of prenatal maternal history is as necessary as good imaging in providing proper care for these patients. An additional degree of difficulty is imposed by the inability to see the fetus for orientation reference and the inability to examine the fetus for clinical findings that might guide the performance and interpretation of the echocardiogram.

In skilled hands, the diagnostic accuracy of fetal echocardiography may reach the high sensitivity and specificity of echocardiography in the neonate; however, not all pediatric cardiology centers have specially trained fetal echocardiographers. Such experts may be pediatric cardiologists, obstetricians, or radiologists with special training or experience in fetal ultrasound imaging and a comprehensive knowledge of congenital heart disease, fetal cardiac anatomy and physiology, and arrhythmias. Where specific expertise in fetal echocardiography does not exist, close collaboration between a pediatric cardiologist/echocardiographer and a fetal ultrasonographer may produce similar results once a learning curve has been completed. The collaboration of a multidisciplinary perinatal team provides support for diagnostic and therapeutic decisions.

**Indications for Echocardiography**

**Class I**

1. Abnormal-appearing heart on general fetal ultrasound examination
2. Fetal tachycardia, bradycardia, or irregular rhythm on clinical or screening ultrasound examination
3. Maternal/family risk factors for cardiovascular disease, such as a parent, sibling, or first-degree relative with congenital heart disease
4. Maternal diabetes
5. Maternal systemic lupus erythematosus
6. Teratogen exposure during a vulnerable period
7. Other fetal system abnormalities (including chromosomal) if pregnancy management decisions are required
8. Performance of transplacental therapy or the presence of a history of significant, but intermittent, arrhythmia or a family history of left or right heart obstructive lesions. Serial examinations are required in these conditions.

**Class II**

1. Fetal distress or dysfunction of unclear etiology
2. Previous history of multiple fetal losses
3. Presence of other system abnormality and an unclear prognosis for fetal outcome

**Class III**

1. Multiple gestations
2. Low-risk pregnancies with normal anatomic findings on ultrasound examination
3. Occasional premature contractions without sustained tachycardia or signs of dysfunction or distress
4. Presence of a noncardiovascular system abnormality when evaluation of the cardiovascular system will not alter either management decisions or fetal outcome

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