

CHILDHOOD CARDIOVASCULAR LISTINGS

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104.00 Cardiovascular System	104.00 Cardiovascular System
	A. General
	<p>1. <u>What do we mean by a cardiovascular impairment?</u></p> <p style="padding-left: 40px;">a. We mean any disorder that affects the proper functioning of the heart or the circulatory system (that is, arteries, veins, capillaries, and the lymphatic drainage). The disorder can be congenital or acquired.</p> <p style="padding-left: 40px;">b. Cardiovascular impairment results from one or more of four consequences of heart disease:</p> <p style="padding-left: 80px;">(i) Chronic heart failure or ventricular dysfunction.</p> <p style="padding-left: 80px;">(ii) Discomfort or pain due to myocardial ischemia, with or without necrosis of heart muscle.</p> <p style="padding-left: 80px;">(iii) Syncope, or near syncope, due to inadequate cerebral perfusion from any cardiac cause, such as obstruction of flow or disturbance in rhythm or conduction resulting in inadequate cardiac output.</p> <p style="padding-left: 80px;">(iv) Central cyanosis due to right-to-left shunt, reduced oxygen concentration in the arterial blood, or pulmonary vascular disease.</p> <p style="padding-left: 40px;">c. Disorders of the veins or arteries (for example, obstruction, rupture, or aneurysm) may cause impairments of the lower extremities (peripheral vascular disease), the central nervous system, the eyes, the kidneys, and other organs. We will evaluate peripheral vascular disease under 4.11 or 4.12 in part A, and impairments of another body system(s) under the listings for that body system(s).</p>
[104.00A, 1 st ¶] The listings in this section describe childhood impairments resulting from congenital or acquired cardiovascular disease based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of therapy prescribed by a treating source.	<p>2. <u>What do we consider in evaluating cardiovascular impairments?</u> The listings in this section describe cardiovascular impairments based on symptoms, signs, laboratory findings, response to a regimen of prescribed treatment, and functional limitations.</p>

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<p>[104.00A, 3rd ¶] Cardiovascular impairments, especially chronic heart failure and congenital heart disease, may result in impairments in other body systems including, but not limited to, growth, neurological, and mental. Therefore, evaluation should include consideration of the adverse effects of cardiovascular impairment in all relevant body systems, and especially on the child's growth and development, or mental functioning, as described under the Growth impairment (100.00), Neurological (111.00), and Mental retardation (112.05) listings.</p>	
<p>[104.00A, 1st ¶] Reasonable efforts should be made to ensure evaluation by a program physician specializing in childhood cardiovascular impairments or a qualified pediatrician.</p> <p>[104.00E, last ¶] "Appropriate" means that the imaging technique used is the proper one to support the evaluation and diagnosis of the impairment.</p>	<p>3. <u>What do the following terms or phrases mean in these listings?</u></p> <p>a. <u>Medical consultant</u> is an individual defined in §§404.1616(a) and 416.1016(a). This term does not include medical sources who provide consultative examinations for us. We use the abbreviation "MC" throughout this section to designate a medical consultant.</p> <p>b. <u>Persistent</u> means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been present, or is expected to be present, for a continuous period of at least 12 months, such that a pattern of continuing severity is established.</p> <p>c. <u>Recurrent</u> means that the longitudinal clinical record shows that, within a consecutive 12-month period, the finding(s) occurs at least three times, with intervening periods of improvement of sufficient duration that it is clear that separate events are involved.</p> <p>d. <u>Appropriate medically acceptable imaging</u> means that the technique used is the proper one to evaluate and diagnose the impairment and is commonly recognized as accurate for assessing the cited finding.</p> <p>e. <u>A consecutive 12-month period</u> means a period of 12 consecutive months, all or part of which must occur within the period we are considering in</p>

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	<p>connection with an application or continuing disability review.</p> <p>f. <u>Currently present</u> means that the finding is present at the time of adjudication.</p> <p>g. <u>Uncontrolled</u> means the impairment does not respond adequately to standard prescribed medical treatment.</p>
	<p>B. Documenting Cardiovascular Impairment</p>
<p>[104.00B, 1st ¶] Each child's file must include sufficiently detailed reports on history, physical examinations, laboratory studies, and any prescribed therapy and response to allow an independent reviewer to assess the severity and duration of the cardiovascular impairment.</p> <p>[104.00A, 1st ¶] A longitudinal clinical record covering a period of not less than 3 months of observations and therapy is usually necessary for the assessment of severity and expected duration unless the child is a neonate or the claim can be decided favorably on the basis of the current evidence.</p>	<p>1. <u>What basic documentation do we need?</u> We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular impairment. A longitudinal clinical record covering a period of not less than 3 months of observations and treatment is usually necessary, unless we can make a determination or decision based on the current evidence.</p>
<p>[104.00A, 5th ¶] Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the child's functioning, and the frequency, severity, and duration of symptoms.</p> <p>[104.00A, 4th ¶] Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is evidence of such treatment, the longitudinal clinical record must include a description of the therapy prescribed by the treating source and response, in addition to information about the nature and severity of the impairment. It is important to document any prescribed therapy and response because this medical management may have improved the child's functional status. The longitudinal record should provide information regarding functional recovery, if any.</p> <p>[104.00A, 5th ¶] Also, several listings include a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.</p>	<p>2. <u>Why is a longitudinal clinical record important?</u> We will usually need a longitudinal clinical record to assess the severity and expected duration of your impairment(s). If you have a listing-level impairment, you probably will have received medically prescribed treatment. Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your treating or other medical source. It should also include your response to this medical management, as well as information about the nature and severity of your impairment. The record will provide us with information on your functional status over an extended period of time and show whether your ability to function is improving, worsening, or unchanging.</p>
	<p>3. <u>What if you have not received ongoing medical treatment?</u></p>

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<p>[104.00A, 5th ¶] Some children will not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). ...Even though a child who does not receive treatment may not be able to show an impairment that meets the criteria of these listings, the child may have an impairment(s) that medically or functionally equals the listings.</p> <p>[104.00A, 5th ¶] Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the child's functioning, and the frequency, severity, and duration of symptoms.</p>	<p>a. You may not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In this situation, we will base our evaluation on the current objective medical evidence and the other evidence we have. If you do not receive treatment, you cannot show an impairment that meets the criteria of these listings. However, we may find you disabled because you have another impairment(s) that in combination with your cardiovascular impairment medically equals the severity of a listed impairment or that functionally equals the listings.</p> <p>b. Unless we can decide your claim favorably on the basis of the current evidence, a longitudinal record is still important. In rare instances where there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the severity and duration of your impairment.</p>
	<p>4. <u>When will we wait before we ask for more evidence?</u></p> <p>a. We will wait when we have information showing that your impairment is not yet stable and the expected change in your impairment might affect our determination or decision. In these situations, we need to wait to properly evaluate the severity and duration of your impairment during a stable period. Examples of when we might wait are:</p> <ul style="list-style-type: none"> (i) If you have had a recent acute event; for example, acute rheumatic fever. (ii) If you have recently had a corrective cardiac procedure; for example, open-heart surgery. (iii) If you have started new drug therapy and your response to this treatment has not yet been established; for example, beta-blocker therapy for dilated congestive cardiomyopathy. <p>b. In these situations, we will obtain more evidence 3 months following the event before we evaluate your impairment. However, we will not wait if we</p>

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	have enough information to make a determination or decision based on all of the relevant evidence in your case.
[104.00B, 2 nd ¶] Results of additional studies necessary to substantiate the diagnosis or to document the severity of the impairment, including two-dimensional and Doppler echocardiography, and radionuclide ventriculograms, should be obtained as appropriate according to part A, 4.00C3. Ambulatory electrocardiographic monitoring may also be obtained if necessary to document the presence or severity of an arrhythmia.	5. <u>Will we purchase any studies?</u> In appropriate situations, we will purchase studies necessary to substantiate the diagnosis or to document the severity of your impairment, generally after we have evaluated the medical and other evidence we already have. We will not purchase studies involving exercise testing if there is significant risk involved or if there is another medical reason not to perform the test. We will follow sections 4.00C6, 4.00C7, 4.00C8, and 104.00B7 when we decide whether to purchase exercise testing. We will make a reasonable effort to obtain any additional studies from a qualified medical source in an office or center experienced in pediatric cardiac assessment. (See §416.919g.)
[104.00B, 5 th ¶] Cardiac catheterization will not be purchased by the Social Security Administration. If the results of catheterization are otherwise available, they should be obtained.	6. <u>What studies will we not purchase?</u> We will not purchase any studies involving cardiac catheterization, such as coronary angiography, arteriograms, or electrophysiological studies. However, if the results of catheterization are part of the existing evidence we have, we will consider them together with the other relevant evidence. See 4.00C15a in part A.
<p>[104.00B, 3rd ¶] Exercise testing, though increasingly used, is still less frequently indicated in children than in adults, and can rarely be successfully performed in children under 6 years of age. It may be of value in the assessment of some arrhythmias, in the assessment of the severity of chronic heart failure, and in the assessment of recovery of function following cardiac surgery or other therapy. It will only be purchased by the Social Security Administration if the case cannot be decided based on the available evidence and, if purchased, must be performed in a specialty center for pediatric cardiology or other facility qualified to perform exercise testing for children.</p> <p>[104.00B, 4th ¶] Purchased exercise tests should be performed using a generally accepted protocol consistent with the prevailing state of medical knowledge and clinical practice. An exercise test should not be purchased for</p>	<p>7. <u>Will we use exercise tolerance tests (ETTs) for evaluating children with cardiovascular impairment?</u></p> <p>a. ETTs, though increasingly used, are still less frequently indicated in children than in adults, and can rarely be performed successfully by children under 6 years of age. An ETT may be of value in the assessment of some arrhythmias, in the assessment of the severity of chronic heart failure, and in the assessment of recovery of function following cardiac surgery or other treatment.</p> <p>b. We will purchase an ETT in a childhood claim only if we cannot make a determination or decision based on the evidence we have and an MC, preferably one with experience in the care of children with cardiovascular impairments, has determined that an ETT is needed to evaluate your impairment. We will not purchase an ETT if you are less than 6 years of age. If we do purchase an ETT for a child age 12 or younger, it must be performed by a qualified medical source in a specialty center for pediatric cardiology or other facility qualified to perform</p>

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<p>a child for whom the performance of the test is considered to constitute a significant risk by a program physician. See 4.00C2c.</p>	<p>exercise tests of children.</p> <p style="padding-left: 40px;">c. For full details on ETT requirements and usage, see 4.00C in part A.</p>
	<p>C. Evaluating Chronic Heart Failure</p> <p>1. <u>What is chronic heart failure (CHF)?</u></p> <p style="padding-left: 40px;">a. <u>CHF</u> is the inability of the heart to pump enough oxygenated blood to body tissues. This syndrome is characterized by symptoms and signs of pulmonary or systemic congestion (fluid retention) or limited cardiac output. Certain laboratory findings of cardiac functional and structural abnormality support the diagnosis of CHF.</p> <p style="padding-left: 40px;">b. CHF is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, or rheumatic, congenital, or other heart disease. However, if the CHF is the result of primary pulmonary hypertension secondary to disease of the lung (cor pulmonale), we will evaluate your impairment using 3.09 in the respiratory system listings in part A.</p>
<p>[104.00E, 2nd ¶] Cardiomegaly or ventricular dysfunction must be present and demonstrated by imaging techniques, such as two-dimensional and Doppler echocardiography. (Reference: Feigenbaum, Harvey, "Echocardiography," 4th Edition, Lea and Febiger, 1986, Appendix, pp. 621-639.)</p> <p>[104.00E, 3rd ¶] Findings of cardiomegaly shown by appropriate medically acceptable imaging evidence must be accompanied by other evidence of chronic heart failure or ventricular dysfunction.</p> <p>[104.00E, 2nd ¶] Chest x-ray (6 ft. PA film) will be considered indicative of cardiomegaly if the cardiothoracic ratio is over 60 percent at age 1 year or less, or 55 percent at more than 1 year of age.</p>	<p>2. <u>What evidence of CHF do we need?</u></p> <p style="padding-left: 40px;">a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization.</p> <p style="padding-left: 80px;">(i) Cardiomegaly is present when:</p> <p style="padding-left: 120px;">(A) Left ventricular diastolic dimension or systolic dimension is greater than 2 standard deviations above the mean for the child's body surface area;</p> <p style="padding-left: 120px;">(B) Left ventricular mass is greater than 2 standard deviations above the mean for the child's body surface area; or</p> <p style="padding-left: 120px;">(C) Chest x-ray (6 foot PA film) is indicative of cardiomegaly if the cardiothoracic ratio is over 60 percent at 1 year of age or less, or 55 percent or greater at more than 1 year of age.</p>

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<p>[104.00E, 1st ¶] Chronic heart failure in infants and children may manifest itself by pulmonary or systemic venous congestion, including cardiomegaly, chronic dyspnea, tachypnea, orthopnea, or hepatomegaly; or symptoms of limited cardiac output, such as weakness or fatigue; or a need for cardiotoxic drugs. Fatigue or exercise intolerance in an infant may be manifested by prolonged feeding time associated with signs of cardiac impairment, including excessive respiratory effort and sweating. Other manifestations of chronic heart failure during infancy may include failure to gain weight or involuntary loss of weight and repeated lower respiratory tract infections.</p> <p>[104.00E, 3rd ¶] This evidence may include clinical evidence, such as</p>	<p>(ii) Ventricular dysfunction is present when indices of left ventricular function, such as fractional shortening or ejection fraction (the percentage of the blood in the ventricle actually pumped out with each contraction), are greater than 2 standard deviations below the mean for the child's age. (Fractional shortening, also called shortening fraction, reflects the left ventricular systolic function in the absence of segmental wall motion abnormalities and has a linear correlation with ejection fraction. In children, fractional shortening is more commonly used than ejection fraction.)</p> <p>(iii) However, these measurements alone do not reflect your functional capacity, which we evaluate by considering all of the relevant evidence.</p> <p>(iv) Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema. These findings need not be present on each report, since CHF may be controlled by prescribed treatment.</p> <p>b. To establish that you have <u>chronic</u> heart failure, your medical history and physical examination should describe characteristic symptoms and signs of pulmonary or systemic congestion or of limited cardiac output associated with the abnormal findings on appropriate medically acceptable imaging. When an acute episode of heart failure is triggered by a remediable factor, such as an arrhythmia, dietary sodium overload, or high altitude, cardiac function may be restored and a chronic impairment may not be present.</p> <p>(i) Symptoms of congestion or of limited cardiac output include easy fatigue, weakness, shortness of breath (dyspnea), cough, or chest discomfort at rest or with activity. Children with CHF may also experience shortness of breath on lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). They may also experience cardiac arrhythmias resulting in palpitations, lightheadedness, or fainting. Fatigue or exercise intolerance in an infant may be manifested by prolonged feeding time, often associated with excessive respiratory effort and sweating.</p> <p>(ii) During infancy, other manifestations of chronic heart failure may include failure to gain weight or involuntary loss of weight and repeated lower</p>

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<p>hepatomegaly, edema, or pulmonary venous congestion; or echocardiographic evidence, such as marked ventricular dilatation above established normals for age, or markedly reduced ejection fraction or shortening fraction.</p>	<p>respiratory tract infections.</p> <p>(iii) Signs of congestion may include hepatomegaly, ascites, increased jugular venous distention or pressure, rales, peripheral edema, rapid shallow breathing (tachypnea), or rapid weight gain. However, these signs need not be found on all examinations because fluid retention may be controlled by prescribed treatment.</p>
<p>[104.00A, 2nd ¶] Examples of congenital defects include: abnormalities of cardiac septation, such as ventricular septal defect or atrioventricular (AV) canal; abnormalities resulting in cyanotic heart disease, such as tetralogy of Fallot or transposition of the vessels; valvular defects or obstructions to ventricular outflow, including pulmonary or aortic stenosis and/or coarctation of the aorta; and major abnormalities of ventricular development, including hypoplastic left heart syndrome or pulmonary tricuspid atresia with hypoplastic right ventricle.</p>	<p style="text-align: center;">D. Evaluating Congenital Heart Disease</p> <p>1. <u>What is congenital heart disease?</u> Congenital heart disease is any abnormality of the heart or the major blood vessels that is present at birth. Examples include:</p> <p style="padding-left: 40px;">a. <u>Abnormalities of cardiac septation</u>, including ventricular septal defect or atrioventricular canal;</p> <p style="padding-left: 40px;">b. <u>Abnormalities resulting in cyanotic heart disease</u>, including tetralogy of Fallot or transposition of the great arteries;</p> <p style="padding-left: 40px;">c. <u>Valvular defects or obstructions to ventricular outflow</u>, including pulmonary or aortic stenosis or coarctation of the aorta; and</p> <p style="padding-left: 40px;">d. <u>Major abnormalities of ventricular development</u>, including hypoplastic left heart syndrome or pulmonary tricuspid atresia with hypoplastic right ventricle.</p>
	<p>2. <u>How will we evaluate symptomatic congenital heart disease?</u></p> <p style="padding-left: 40px;">a. Because of improved treatment methods, more children with congenital heart disease are living longer. Although some types of congenital heart disease may be corrected by surgery, many children with treated congenital heart disease continue to have problems throughout their lives (symptomatic congenital heart disease). If you have congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias, we will evaluate your impairment under 104.02 or 104.05. Otherwise, we will evaluate your impairment under 104.06.</p>

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<p>[104.00D, 1st ¶] Some congenital defects usually lead to listing-level impairment in the first year of life and require surgery within the first year as a life-saving measure. Examples of impairments that in most instances will require life-saving surgery before age 1, include, but are not limited to, the following: hypoplastic left heart syndrome; critical aortic stenosis with neonatal heart failure; critical coarctation of the aorta, with or without associated anomalies; complete AV canal defects; transposition of the great arteries; tetralogy of Fallot; and pulmonary atresia with intact ventricular septum.</p> <p>[104.00D, 2nd ¶] In addition, there are rarer defects which may lead to early mortality and that may require multiple surgical interventions or a combination of surgery and other major interventional procedures (e.g., multiple "balloon" catheter procedures). Examples of such defects include single ventricle, tricuspid atresia, and multiple ventricular septal defects.</p>	<p>b. For 104.06A2, we will accept pulse oximetry measurements instead of arterial O₂, but the arterial O₂ values are preferred, if available.</p> <p>c. For 104.06D, examples of impairments that in most instances will require life-saving surgery or a combination of surgery and other major interventional procedures (for example, multiple "balloon" catheter procedures) before age 1 include, but are not limited to, the following:</p> <ul style="list-style-type: none"> (i) Hypoplastic left heart syndrome, (ii) Critical aortic stenosis with neonatal heart failure, (iii) Critical coarctation of the aorta, with or without associated anomalies, (iv) Complete atrioventricular canal defects, (v) Transposition of the great arteries, (vi) Tetralogy of Fallot, (vii) Pulmonary atresia with intact ventricular septum, (viii) Single ventricle, (ix) Tricuspid atresia, and (x) Multiple ventricular septal defects.
	E. Evaluating Arrhythmias
	1. <u>What is an arrhythmia?</u> An <u>arrhythmia</u> is a change in the regular beat of the heart. Your heart may seem to skip a beat or beat irregularly, very quickly (tachycardia), or very slowly (bradycardia).
	2. <u>What are the different types of arrhythmias?</u> <p>a. There are many types of arrhythmias. Arrhythmias are identified by where they occur in the heart (atria or ventricles) and by what happens to the heart's rhythm when they occur.</p> <p>b. Arrhythmias arising in the cardiac atria (upper chambers of the heart) are called atrial or supraventricular arrhythmias. Ventricular arrhythmias begin in the ventricles (lower chambers). In general, ventricular arrhythmias caused by heart disease are the most serious.</p>
	3. <u>How do we evaluate arrhythmias using 104.05?</u>

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	<p>a. We will use 104.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator and you have uncontrolled recurrent episodes of syncope or near syncope. If your arrhythmias are controlled, we will evaluate your underlying heart disease using the appropriate listing. For other considerations when we evaluate arrhythmias in the presence of an implanted cardiac defibrillator, see 104.00E4.</p> <p>b. We consider <u>near syncope</u> to be a period of altered consciousness, since syncope is a loss of consciousness or a faint. It is not merely a feeling of light-headedness, momentary weakness, or dizziness.</p> <p>c. For purposes of 104.05, there must be a documented association between the syncope or near syncope and the recurrent arrhythmia. The recurrent arrhythmia, not some other cardiac or non-cardiac disorder, must be established as the cause of the associated symptom. This documentation of the association between the symptoms and the arrhythmia may come from the usual diagnostic methods, including Holter monitoring (also called ambulatory electrocardiography) and tilt-table testing with a concurrent ECG. Although an arrhythmia may be a coincidental finding on an ETT, we will not purchase an ETT to document the presence of a cardiac arrhythmia.</p>
	<p>4. <u>What will we consider when you have an implanted cardiac defibrillator and you do not have arrhythmias that meet the requirements of 104.05?</u></p> <p>a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in children who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group of children at risk for sudden cardiac death consists of children with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in children with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator is a unique form of treatment; it rescues a child from what may have been cardiac arrest. However, as a consequence of the shock(s), children may experience psychological distress,</p>

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	<p>which we may evaluate under the mental disorders listings in 112.00ff.</p> <p>b. Most implantable cardiac defibrillators have rhythm-correcting and pacemaker capabilities. In some children, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. (The shock is like being kicked in the chest.) Implanted cardiac defibrillators may deliver inappropriate shocks, often repeatedly, in response to benign arrhythmias or electrical malfunction. Also, exposure to strong electrical or magnetic fields, such as from MRI (magnetic resonance imaging), can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. We must consider the frequency of, and the reason(s) for, the shocks when evaluating the severity and duration of your impairment.</p> <p>c. In general, the exercise limitations imposed on children with an implanted cardiac defibrillator are those dictated by the underlying heart impairment. However, the exercise limitations may be greater when the implanted cardiac defibrillator delivers an inappropriate shock in response to the increase in heart rate with exercise, or when there is exercise-induced ventricular arrhythmia.</p>
	<p>F. Evaluating other cardiovascular impairments</p>
	<p>1. <u>What is ischemic heart disease (IHD) and how will we evaluate it in children?</u> <u>IHD</u> results when one or more of your coronary arteries is narrowed or obstructed or, in rare situations, constricted due to vasospasm, interfering with the normal flow of blood to your heart muscle (ischemia). The obstruction may be the result of an embolus, a thrombus, or plaque. When heart muscle tissue dies as a result of the reduced blood supply, it is called a myocardial infarction (heart attack). Ischemia is rare in children, but when it occurs, its effects on children are the same as on adults. If you have IHD, we will evaluate it under 4.00E and 4.04 in part A.</p>
	<p>2. <u>How will we evaluate hypertension?</u> Because <u>hypertension</u> (high blood pressure) generally causes disability through its effects on other body systems, we will evaluate it by reference to the specific body system(s) affected (heart, brain, kidneys, or eyes) when we consider its effects under the listings. We will also consider any limitations imposed by your hypertension when we consider whether</p>

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	<p>you have an impairment that functionally equals the listings.</p> <p>3. <u>What is cardiomyopathy and how will we evaluate it?</u> Cardiomyopathy is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy). There are various types of cardiomyopathy, which fall into two major categories: <i>Ischemic</i> and <i>nonischemic</i> cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: Dilated, hypertrophic, and restrictive. We will evaluate cardiomyopathy under 4.04 in part A, 104.02, 104.05, or 111.06, depending on its effects on you.</p>
<p><i>F. Valvular Heart Disease</i></p> <p>Valvular heart disease requires documentation by appropriate imaging techniques, including Doppler echocardiogram studies or cardiac catheterization if catheterization results are available from a treating source or other source of record. Listing-level impairment is usually associated with critical aortic stenosis in a newborn child, persistent heart failure, arrhythmias, or valve replacement and ongoing anticoagulant therapy. The usual time after valvular surgery for adequate assessment of the results of treatment is considered to be 3 months.</p>	<p>4. <u>How will we evaluate valvular heart disease?</u> We will evaluate valvular heart disease under the listing appropriate for its effect on you. Thus, we may use 4.04 in part A, 104.02, 104.05, 104.06, or an appropriate neurological listing in 111.00ff.</p>
	<p>5. <u>What do we consider when we evaluate heart transplant recipients?</u></p> <p>a. After your heart transplant, we will consider you disabled for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year.</p> <p>b. However, heart transplant patients generally meet our definition of disability before they undergo transplantation. We will determine the onset of your disability based on the facts in your case.</p> <p>c. We will not assume that you became disabled when your name was</p>

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	<p>placed on a transplant waiting list. This is because you may be placed on a waiting list soon after diagnosis of the cardiac disorder that may eventually require a transplant. Physicians recognize that candidates for transplantation often have to wait months or even years before a suitable donor heart is found, so they place their patients on the list as soon as permitted.</p> <p>d. When we do a continuing disability review to determine whether you are still disabled, we will evaluate your residual impairment(s), as shown by symptoms, signs, and laboratory findings, including any side effects of medication. We will consider any remaining symptoms, signs, and laboratory findings indicative of cardiac dysfunction in deciding whether medical improvement (as defined in §416.994a) has occurred.</p>
[104.00G] The diagnosis should be made in accordance with the current revised Jones criteria for guidance in the diagnosis of rheumatic fever.	<p>6. <u>How will we evaluate chronic rheumatic fever or rheumatic heart disease?</u> The diagnosis should be made in accordance with the current revised Jones criteria for guidance in the diagnosis of rheumatic fever. We will evaluate persistence of rheumatic fever activity under 104.13. If you have evidence of chronic heart failure or recurrent arrhythmias associated with rheumatic heart disease, we will use 104.02 or 104.05.</p>
	<p>7. <u>What is hyperlipidemia and how will we evaluate it?</u> <u>Hyperlipidemia</u> is the general term for an elevation of any or all of the lipids (fats or cholesterol) in the blood; for example, hypertriglyceridemia, hypercholesterolemia, and hyperlipoproteinemia. These disorders of lipoprotein metabolism and transport can cause defects throughout the body. The effects most likely to interfere with function are those produced by atherosclerosis (narrowing of the arteries) and coronary artery disease. We will evaluate your lipoprotein disorder by considering its effects on you.</p>
	<p>8. <u>How will we evaluate Kawasaki disease?</u> We will evaluate Kawasaki disease under the listing appropriate to its effects on you, which may include major coronary artery aneurysm or heart failure. A major coronary artery aneurysm may cause ischemia or arrhythmia, which we will evaluate under 4.04 in part A or 104.05. We will evaluate chronic heart failure under 104.02.</p>
	<p>9. <u>What is lymphedema and how will we evaluate it?</u></p>

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	<p>a. <u>Lymphedema</u> is edema of the extremities due to a disorder of the lymphatic circulation; at its worst, it is called elephantiasis. Primary lymphedema is caused by abnormal development of lymph vessels and may be present at birth (congenital lymphedema), but more often develops during the teens (lymphedema praecox). Secondary lymphedema is due to obstruction or destruction of normal lymphatic channels due to tumor, surgery, repeated infections, or parasitic infection such as filariasis. Lymphedema most commonly affects one extremity.</p> <p>b. Lymphedema does not meet the requirements of 4.11 in part A, although it may medically equal the severity of that listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular listing, such as 4.11, or a musculoskeletal listing, such as 101.02A or 101.03. If no listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we consider whether you have an impairment that functionally equals the listings.</p>
	<p>10. <u>What is Marfan syndrome and how will we evaluate it?</u></p> <p>a. Marfan syndrome is a genetic connective tissue disorder that affects multiple body systems, including the skeleton, eyes, heart, blood vessels, nervous system, skin, and lungs. There is no specific laboratory test to diagnose Marfan syndrome. The diagnosis is generally made by medical history, including family history, physical examination, including an evaluation of the ratio of arm/leg size to trunk size, a slit lamp eye examination, and a heart test(s), such as an echocardiogram. In some cases, a genetic analysis may be useful, but such analyses may not provide any additional helpful information.</p> <p>b. The effects of Marfan syndrome can range from mild to severe. In most cases, the disorder progresses as you age. Most individuals with Marfan syndrome have abnormalities associated with the heart and blood vessels. Your heart's mitral valve may leak, causing a heart murmur. Small leaks may not cause symptoms, but larger ones may cause shortness of breath, fatigue, and palpitations. Another effect is that the wall of the aorta may be weakened and</p>

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	stretch (aortic dilation). This aortic dilation may tear, dissect, or rupture, causing serious heart problems or sometimes sudden death. We will evaluate the manifestations of your Marfan syndrome under the appropriate body system criteria, such as 4.10 in part A, or if necessary consider the functional limitations imposed by your impairment.
	G. Other Evaluation Issues
	<p>1. <u>What effect does obesity have on the cardiovascular system and how will we evaluate it?</u> Obesity is a medically determinable impairment that is often associated with disorders of the cardiovascular system. Disturbance of this system can be a major cause of disability in children with obesity. Obesity may affect the cardiovascular system because of the increased workload the additional body mass places on the heart. Obesity may make it harder for the chest and lungs to expand. This can mean that the respiratory system must work harder to provide needed oxygen. This in turn would make the heart work harder to pump blood to carry oxygen to the body. Because the body would be working harder at rest, its ability to perform additional work would be less than would otherwise be expected. Thus, the combined effects of obesity with cardiovascular impairments can be greater than the effects of each of the impairments considered separately. We must consider any additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular impairment or a listing-level cardiovascular impairment (or a combination of impairments that medically equals a listing), and when we determine whether your impairment(s) functionally equals the listings.</p>
[104.00C, 1 st ¶] In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The overall clinical and laboratory evidence, including the treatment plan(s) or results, should be persuasive that a listing-level impairment exists. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, impairment evaluation may need to be deferred for a period of up to 3 months from the	<p>2. <u>How do we relate treatment to functional status?</u> In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, we may need to defer evaluation of the impairment for a period of up to 3 months from the date treatment began to permit consideration of treatment effects, unless we can make a determination or decision using the evidence we have. See</p>

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<p>date of treatment to permit consideration of treatment effects. Evaluation should not be deferred if the claim can be favorably decided based upon the available evidence.</p>	<p>104.00B4.</p>
<p>[from 104.00A, last ¶] Indeed, it must be remembered that these listings are only examples of common cardiovascular disorders that are severe enough to find a child disabled. When you have a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will consider a determination whether your impairment(s) medically equals or, as appropriate, functionally equals the listings. (See §§ 404.1526, 416.926, and 416.926a.)</p>	<p style="text-align: center;">3. <u>How do we evaluate impairments that do not meet one of the cardiovascular listings?</u></p> <p style="padding-left: 40px;">a. These listings are only examples of common cardiovascular disorders that we consider severe enough to result in marked and severe functional limitations. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.</p> <p style="padding-left: 40px;">b. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926.) If you have a severe impairment(s) that does not meet or medically equal the criteria of a listing, we will consider whether it functionally equals the listings. (See §416.926a.) When we decide whether you continue to be disabled, we use the rules in §416.994a.</p>
<p>104.01 <i>Category of Impairments, Cardiovascular System</i></p>	<p>104.01 <u>Category of Impairments, Cardiovascular System</u></p>
<p>104.02 <i>Chronic heart failure.</i> Documented by clinical and laboratory findings as described in 104.00E, and with one of the following: A. Persistent tachycardia at rest (see table I); OR B. Persistent tachypnea at rest (see table II), or markedly decreased exercise tolerance (see 104.00E); OR C. Recurrent arrhythmias, as described in 104.05; OR D. Growth disturbance, with: 1. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from established growth curve (on standard growth charts) which persists for 2 months or longer; or 2. An involuntary weight loss (or failure to gain weight at an appropriate rate for</p>	<p>104.02. <u>Chronic heart failure</u> while on a regimen of prescribed treatment with symptoms and signs described in 104.00C2 and with one of the following: A. Persistent tachycardia at rest (see Table I); OR B. Persistent tachypnea at rest (see Table II) or markedly decreased exercise tolerance (see 104.00C2b); OR C. Growth disturbance with: 1. An involuntary weight loss or failure to gain weight at an appropriate rate for age, resulting in a fall of 15 percentiles from an established growth curve (on current NCHS/CDC growth chart) which is currently present (see 104.00A3f) and has persisted for 2 months or longer; or 2. An involuntary weight loss or failure to gain weight at an appropriate rate for age, resulting in a fall to below the third percentile from an established growth</p>

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<p>age) resulting in a fall to below the third percentile from established growth curve (on standard growth charts) which persists for 2 months or longer; or 3. Growth impairment as described under the criteria in 100.00.</p> <p style="text-align: center;">TABLE I. – TACHYCARDIA AT REST</p> <table border="1" style="margin-left: auto; margin-right: auto; border-collapse: collapse;"> <thead> <tr> <th style="text-align: center;">Age</th> <th style="text-align: center;">Apical heart rate (beats per minute)</th> </tr> </thead> <tbody> <tr> <td>Under 1 year</td> <td style="text-align: center;">..... 150</td> </tr> <tr> <td>1 through 3 years</td> <td style="text-align: center;">..... 130</td> </tr> <tr> <td>4 through 9 years</td> <td style="text-align: center;">..... 120</td> </tr> <tr> <td>10 through 15 years</td> <td style="text-align: center;">..... 110</td> </tr> <tr> <td>Over 15 years</td> <td style="text-align: center;">..... 100</td> </tr> </tbody> </table> <p style="text-align: center;">TABLE II – TACHYPNEA AT REST</p> <table border="1" style="margin-left: auto; margin-right: auto; border-collapse: collapse;"> <thead> <tr> <th style="text-align: center;">Age</th> <th style="text-align: center;">Respiratory rate over (per minute)</th> </tr> </thead> <tbody> <tr> <td>Under 1 year</td> <td style="text-align: center;">..... 40</td> </tr> <tr> <td>1 through 5 years</td> <td style="text-align: center;">..... 35</td> </tr> <tr> <td>6 through 9 years</td> <td style="text-align: center;">..... 30</td> </tr> <tr> <td>Over 9 years</td> <td style="text-align: center;">..... 25</td> </tr> </tbody> </table>	Age	Apical heart rate (beats per minute)	Under 1 year 150	1 through 3 years 130	4 through 9 years 120	10 through 15 years 110	Over 15 years 100	Age	Respiratory rate over (per minute)	Under 1 year 40	1 through 5 years 35	6 through 9 years 30	Over 9 years 25	<p>curve (on current NCHS/CDC growth chart) which is currently present (see 104.00A3f) and has persisted for 2 months or longer.</p> <p style="text-align: center;">TABLE I. – TACHYCARDIA AT REST</p> <table border="1" style="margin-left: auto; margin-right: auto; border-collapse: collapse;"> <thead> <tr> <th style="text-align: center;">Age</th> <th style="text-align: center;">Apical heart rate (beats per minute)</th> </tr> </thead> <tbody> <tr> <td>Under 1 year</td> <td style="text-align: center;">..... 150</td> </tr> <tr> <td>1 through 3 years</td> <td style="text-align: center;">..... 130</td> </tr> <tr> <td>4 through 9 years</td> <td style="text-align: center;">..... 120</td> </tr> <tr> <td>10 through 15 years</td> <td style="text-align: center;">..... 110</td> </tr> <tr> <td>Over 15 years</td> <td style="text-align: center;">..... 100</td> </tr> </tbody> </table> <p style="text-align: center;">TABLE II – TACHYPNEA AT REST</p> <table border="1" style="margin-left: auto; margin-right: auto; border-collapse: collapse;"> <thead> <tr> <th style="text-align: center;">Age</th> <th style="text-align: center;">Respiratory rate over (per minute)</th> </tr> </thead> <tbody> <tr> <td>Under 1 year</td> <td style="text-align: center;">..... 40</td> </tr> <tr> <td>1 through 5 years</td> <td style="text-align: center;">..... 35</td> </tr> <tr> <td>6 through 9 years</td> <td style="text-align: center;">..... 30</td> </tr> <tr> <td>Over 9 years</td> <td style="text-align: center;">..... 25</td> </tr> </tbody> </table>	Age	Apical heart rate (beats per minute)	Under 1 year 150	1 through 3 years 130	4 through 9 years 120	10 through 15 years 110	Over 15 years 100	Age	Respiratory rate over (per minute)	Under 1 year 40	1 through 5 years 35	6 through 9 years 30	Over 9 years 25
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<p>104.03 <i>Hypertensive cardiovascular disease</i>. With persistently elevated blood pressure equal to or greater than the 95th percentile for age (see table III), and one of the following: A. Impaired renal function, as described in 106.02; OR B. Cerebrovascular damage, as described in 111.06; OR</p>	<p>(Removed)</p>																																												

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<p>C. Chronic heart failure as described in 104.02. (Note: Table III omitted due to space considerations.)</p>	
<p>104.05 <i>Recurrent arrhythmias</i>, such as persistent or recurrent heart block (A-V dissociation), repeated symptomatic tachyarrhythmias or bradyarrhythmias or long QT syndrome arrhythmias, not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled repeated episodes of cardiac syncope or near syncope and arrhythmia despite prescribed treatment, including electronic pacemaker (see 104.00A if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography coincident with the occurrence of syncope or near syncope.</p>	<p>104.05 <u>Recurrent arrhythmias</u>, not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled (see 104.00A3g), recurrent (see 104.00A3c) episodes of cardiac syncope or near syncope (see 104.00E3b), despite prescribed treatment (see 104.00B3 if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography, or by other appropriate medically acceptable testing, coincident with the occurrence of syncope or near syncope (see 104.00E3c).</p>
<p>104.06 <i>Congenital heart disease</i>. With one of the following: A. Cyanotic heart disease, with persistent, chronic hypoxemia as manifested by: 1. Hematocrit of 55 percent or greater on two or more evaluations within a 3-month period; or 2. Arterial O₂ saturation of less than 90 percent in room air, or resting PO₂ of 60 Torr or less; or 3. Hypercyanotic spells, syncope, characteristic squatting, or other incapacitating symptoms directly related to documented cyanotic heart disease; or 4. Exercise intolerance with increased hypoxemia on exertion; OR B. Chronic heart failure with evidence of ventricular dysfunction, as described in 104.02; OR C. Recurrent arrhythmias as described in 104.05; OR D. Secondary pulmonary vascular obstructive disease with a mean pulmonary arterial pressure elevated to at least 70 percent of the mean systemic arterial pressure; OR E. Congenital valvular or other stenotic defects, or valvular regurgitation, as</p>	<p>104.06 <u>Congenital heart disease</u>, documented by appropriate medically acceptable imaging (see 104.00A3d) or cardiac catheterization, with one of the following: A. Cyanotic heart disease, with persistent, chronic hypoxemia as manifested by: 1. Hematocrit of 55 percent or greater on two evaluations 3 months or more apart within a consecutive 12-month period (see 104.00A3e); or 2. Arterial O₂ saturation of less than 90 percent in room air, or resting arterial PO₂ of 60 Torr or less; or 3. Hypercyanotic spells, syncope, characteristic squatting, or other incapacitating symptoms directly related to documented cyanotic heart disease; or 4. Exercise intolerance with increased hypoxemia on exertion. OR B. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure. OR C. Symptomatic acyanotic heart disease, with ventricular dysfunction interfering very seriously with the ability to independently initiate, sustain, or complete activities. OR</p>

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<p>described in 104.00F and 104.07; OR F. Symptomatic acyanotic heart disease, with ventricular dysfunction resulting in significant restriction of age-appropriate activities or inability to complete age-appropriate tasks (see 104.00A); OR G. Growth failure, as described in 100.00; OR H. For infants under 12 months of age at the time of filing, with life-threatening congenital heart impairment that will or has required surgical treatment in the first year of life, consider the infant to be under a disability until the attainment of age 1 or for 12 months after surgery, whichever is the later event; thereafter, evaluate impairment severity with reference to 104.02 to 104.08.</p>	<p>D. For infants under 12 months of age at the time of filing, with life-threatening congenital heart impairment that will require or already has required surgical treatment in the first year of life, and the impairment is expected to be disabling (because of residual impairment following surgery, or the recovery time required, or both) until the attainment of at least 1 year of age, consider the infant to be under disability until the attainment of at least age 1; thereafter, evaluate impairment severity with reference to the appropriate listing.</p>
<p>104.07 <i>Valvular heart disease or other stenotic defects, or valvular regurgitation</i>, documented by appropriate imaging techniques or cardiac catheterization. A. Evaluate according to criteria in 104.02, 104.05, 111.06, or 11.04; OR B. Critical aortic stenosis in newborn.</p>	(Removed)
<p>104.08 <i>Cardiomyopathies</i>, documented by appropriate imaging techniques, including echocardiography or cardiac catheterization, if catheterization results are available from a treating source. Impairment must be associated with an ejection fraction of 50 percent or less and significant left ventricular dilatation using standardized age-appropriate echocardiographic ventricular cavity measurements. Evaluate under the criteria in 104.02, 104.05, or 111.06.</p>	(Removed)
<p>104.09 <i>Cardiac transplantation</i>. Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under 104.02 to 104.08.</p>	<p>104.09 <u>Heart transplant</u>. Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under the appropriate listing.</p>
<p>104.13 <i>Chronic rheumatic fever or rheumatic heart disease</i>. Consider under a disability for 18 months from the established onset of impairment with one of the following: A. Persistence of rheumatic fever activity for 6 months or more which is manifested by significant murmur(s), cardiac enlargement (see 104.00E) or ventricular dysfunction, and other abnormal laboratory findings, as for</p>	<p>104.13 <u>Rheumatic heart disease</u>, with persistence of rheumatic fever activity manifested by significant murmurs(s), cardiac enlargement or ventricular dysfunction (see 104.00C2a), and other associated abnormal laboratory findings; for example, an elevated sedimentation rate or ECG findings, for 6 months or more in a consecutive 12-month period (see 104.00A3e). Consider under a disability for 18 months from the established onset of impairment, then evaluate</p>

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<p>example, an elevated sedimentation rate or ECG findings; OR B. Evidence of chronic heart failure, as described under 104.02; OR C. Recurrent arrhythmias, as described under 104.05.</p>	any residual impairment(s).
<p>104.14 <i>Hyperlipidemia</i>. Documented Type II homozygous hyperlipidemia with repeated plasma cholesterol levels of 500 mg/ml or greater, with one of the following: A. Myocardial ischemia, as described in 4.04B or 4.04C; OR B. Significant aortic stenosis documented by Doppler echocardiographic techniques or cardiac catheterization; OR C. Major disruption of normal life activities by repeated hospitalizations for plasmapheresis or other prescribed therapies, including liver transplant; OR D. Recurrent pancreatitis complicating hyperlipidemia.</p>	(Removed)
<p>104.15 <i>Kawasaki syndrome</i>. With one of the following: A. Major coronary artery aneurysm; OR B. Chronic heart failure, as described in 104.02.</p>	(Removed)