REVIEW ARTICLE

Assessment of heart murmurs in childhood

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Abstract

Objective: to discuss clinical and laboratorial evaluation of heart murmurs in children, an important problem faced by pediatricians in their practice.

Sources: this review was based on a critical analysis of the current literature, as well as pediatrics and pediatric cardiology textbooks, which were found to be an important source of information on the subject.

Summary of the findings: it is important for pediatricians to know how to obtain precise information regarding the patient's medical history and to perform extensive physical examination of a child with heart murmur. The diagnosis of innocent heart murmur is essentially clinical and it can help the pediatrician to identify situations which are associated with cardiovascular diseases.

Conclusions: the pediatrician is usually the first physician to see a child with heart murmur and he should be able to distinguish between innocent and pathologic heart murmurs.

J Pediatr (Rio J) 2003;79 Suppl 1:S87-S96: heart defects, congenital, heart auscultation, heart murmurs.

Introduction

The detection of changes in cardiac auscultation, especially the presence of murmurs, is the most frequent reason for patient referral to cardiologists, followed by thoracic pain and syncope.[1] Undoubtedly, an evaluation performed by a cardiologist is the gold standard for the diagnosis of heart diseases; however, as these changes often represent variations within the normal range or innocent murmurs, it is important to carry out a more adequate pediatric evaluation, avoiding unnecessary exams and referrals. It must be emphasized that examination of the cardiovascular system goes beyond cardiac auscultation and that some changes, such as differences in pulse palpation, are suggestive of disorders and must be investigated. On the other hand, it is important to keep in mind that a request for ancillary exams may reveal changes that must be interpreted considering the special characteristics of children. Imaging exams, such as echocardiogram, are becoming increasingly sophisticated, showing findings that, despite being within the normal range, are usually not understood as such by the patient and his/her family.[2-4]

Abnormalities in the cardiovascular system development are responsible for a wide variety of clinical effects. Complex structural malformations can cause fetal death or be diagnosed in the first weeks or months of life, based on the symptomatology of cardiocirculatory decompensation. In the transition to postnatal life,

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predominance of the right heart ceases to exist, as well as placental circulation; additionally, pulmonary arterial resistance is modified and left-heart dependent circulation is installed. At this point, malformations that have been compensated during intrauterine life become hemodynamically unstable, and the clinical status will contribute to diagnosis. However, some congenital cardiovascular malformations remain asymptomatic during this transition, and their clinical status will only be manifested later. These conditions, which include abnormal communication between chambers and hypertrophic cardiomyopathy, have a very heterogeneous genetic inheritance, being only acknowledged in adult life and/or under special circumstances. Small malformations, such as bicuspid aortic valve, will only be revealed if they become the site of infectious processes, such as endocarditis or stenosis.[5] Being aware of these developmental characteristics of cardiovascular disorders, it can be assumed that, when facing certain complaints, signs, symptoms and/or auscultation findings, pediatricians will be concerned with diagnosing a congenital or acquired heart disease that has remained asymptomatic until then.

Therefore, when dealing with certain clinical situations and/or with changes in cardiac auscultation, pediatricians must have a systematized approach that allows them to establish the difference between a change within normal range and a possible cardiovascular disorder that needs to be diagnosed by a cardiologist and, then, to decide on the urgency of this referral.

Initial approach to heart murmurs

The incidence of congenital heart diseases is less than 1%, affecting 8 to 12 out of 1,000 living newborns (excluding premature infants). The most common are acyanotic heart diseases and, therefore, those that are less symptomatic and present a more difficult diagnosis, unless they are associated with other cardiovascular malformations, which are frequent in cases of ventricular septal defect, atrial septal defect, and aortic valve stenosis (Table 1).[5,6] On the other hand, acquired heart diseases have a variable occurrence, depending on the population being studied. Disorders such as rheumatic fever, myocarditis, endocarditis, and others have a greater incidence, as children are more exposed to infectious agents and become ill more frequently due to other systemic pathologies.[7-9] Acquired heart diseases, as well as congenital, can also be identified later in life, especially in the case of patients who perform strenuous physical activities. Kawasaki disease, for instance, can be a cause for stroke, valvulopathy, and coronary artery and systemic aneurysms, after having remained asymptomatic for years.[10]

The most common abnormalities in the cardiac auscultation of children are murmurs, which are sounds generated by turbulent sound waves, originated from the heart and/or vascular system. Murmurs must be differentiated from changes in cardiac rhythm, which require a different laboratory approach. Innocent heart murmur is a change in auscultation observed in the absence of anatomical and/or functional abnormalities of the cardiovascular system. Between 50% and 70% of children will, at some point of their childhood or adolescence, present some change in auscultation that will be characterized as a murmur, especially during school years.

Usually, murmurs are detected in routine medical appointments, or during appointments scheduled for ordinary problems, such as infectious febrile illness, anemia and others.[6,11] These situations raise questions

Table 1 - Frequency of congenital heart diseases

Cyanotic		Acyanotic	
Ventricular septal defect- VSD	15 to 20% *	Tetralogy of Fallot - TOF	10%
Atrial septal defect - ASD	5 to 10% *	Transposition of the great arteries - TGA	5%
Patent ductus arteriosus - PDA	5 to 10%		
Coarctation of the aorta - CoA	8 to 10% †		
Pulmonary stenosis - PS	8 to 12%		
Aortic stenosis - AS	8 to 12%		
Atrioventricular septal defect	< 1% ‡		

* VSD and ASD occurrence is associated with other congenital heart diseases in 30% to 50% of the cases.
† Frequent association with Turner's syndrome.
‡ It occurs in 30% of the patients with Down's Syndrome.

as to the possibility of a previously existing murmur and/ or modification of its characteristics; possibility that the current pathology is causing a definite injury in a previously normal cardiovascular system; chance that an innocent murmur is being exacerbated by a hyperkinetic state; chance that a murmur, associated with a heart disease, is being exacerbated by the same condition, among others. Therefore, the approach must consider these various possibilities; some situations can be prioritized in order to be more carefully evaluated, in consequence of the risk of a cardiovascular event.[6]

Initially, changes observed in the auscultation of a newborn or a six to 12 month-old infant require a more detailed investigation. Approximately 50% of newborns weighing less than 1,500g present some heart disease, usually patent ductus arteriosus (PDA): the lower the birthweight the greater the chances of this event. On the other hand, newborns considered big for gestational age – usually children of diabetic mothers – have a greater risk of heart diseases such as transposition of the great arteries (TGA). Low birthweight children can have a cardiovascular disorder associated with the pathology that caused intrauterine compromise (such as a genetic syndrome or a congenital infection).[6,11] In young children with congenital heart diseases (congestive heart failure), the low specificity of the symptomatology of decompensation is another reason for concern. Initial manifestations can include eating difficulty, head sweats while being breastfed, long-lasting jaundice, respiratory discomfort, and others. Also in this age group, changes in auscultation that are related to obstructive injuries can be audible at birth, but those caused by shunts (ventricular/ atrial septal defect) can only be detected later, when changes in pulmonary circulation are completed. In face of these considerations, it is advisable that newborns and infants be evaluated by a cardiologist in their first year of life whenever they present symptomatology or findings on physical examination suggestive of heart disease, regardless of the existence of innocent murmurs in this age group.[12-15]

Secondly, some pregnancy conditions and personal and/or familial background associated with an increased risk of cardiovascular compromise must be investigated, even in the absence of changes in cardiac auscultation.[16,17] Malformations, such as ventricular septal defect (VSD), PDA, coarctation of the aorta, TGA, and hypertrophic cardiomyopathy, are frequently associated with gestational diabetes. Children of mothers with systemic lupus erythematosus or other collagen diseases can present congenital heart block and atrial-ventricular septal defects. If the mother has seizures, medication used during pregnancy can be teratogenic (use of hydantoin and occurrence of pulmonary and aortic stenosis, coarctation of the aorta, PDA). The use of other medication (amphetamines, lithium, progesterone, and estrogen), alcohol or drugs can have a similar effect, being associated especially with VSD, transposition of great vessels, tetralogy of Fallot, and PDA. Arterial hypertension and infections during pregnancy represent an increased risk of prematurity and, therefore, PDA. Infections during the first three months are potentially teratogenic – possibly resulting in complex heart diseases –, while infections in the last three months can cause myocarditis or other cardiac inflammatory processes, mainly in cases of rubella, cytomegalovirus, herpes virus, coxsackie B virus, and human immunodeficiency virus.[8]

Past history of congenital heart disease in parents or siblings is very important, increasing the risk of equal or similar injuries by three, four or even 10 times, especially if the mother and/or more than one family member is affected. Some forms of heart disease, such as mitral valve prolapse and VSD, have a high risk of recurrence, while tricuspid atresia and truncus arteriosus have low rates of familial recurrence. Rheumatic fever, arterial hypertension and coronary disease also present high rates of familial recurrence.[16,17]

Unexplained sudden death in children or young adults is a background that must be valued. Hypertrophic cardiomyopathy, which has a genetic inheritance in 20% to 60% of the cases, can remain asymptomatic for years and manifest only in special circumstances, being, therefore, a cause of concern for adolescents with such background that engage in competitive sports.[18]

Patients with hereditary diseases, such as mucopolysaccharidosis, muscle dystrophy (Duchenne), neurofibromatosis, osteogenesis imperfecta, tuberous sclerosis, sickle cell anemia, congenital arrhythmias, among others, must have a cardiovascular evaluation not only when the pathology is detected, but throughout life, since structural changes can appear in the course of the disease. Usually, cardiovascular changes occur in 25% of the cases of children with congenital malformations in other systems; in genetic syndromes, such as Marfan's syndrome, there is a possibility that these changes occur either in childhood or adolescence.[11,19-22] (Table 2).

Considering these characteristics of heart diseases in children, the importance of monitoring fetal cardiovascular development through fetal echocardiogram has been discussed.

Such procedure is not done as a routine, but a more detailed study could be indicated in risk situations - fetal (arrhythmias, extracardiac anomalies, fetal hydrops, suspected chromosome anomalies, suspected cardiac disease in pregnancy ultrasound); maternal (mother with congenital heart disease, diabetes, collagen disease, previous history of alcohol, drug or medication abuse); familial (congenital heart disease in first-degree family members, hereditary disorders, genetic syndromes). This approach would allow for a more adequate immediate neonatal care, leading to a greater survival of patients with complex heart diseases.[23]

Table 2 - Genetic syndromes, congenital malformations and frequency of congenital heart diseases

Condition	%CHD	Most common problems
Trisomy 13	90%	VSD, PDA, dextrocardia
Trisomy 18	95%	VSD, PDA, pulmonary stenosis
Trisomy 21 (Down)	50%	VSD, AVSD
Turner's Syndrome (XO)	35%	AS, ASD, COA
Noonan's Syndrome		Pulmonary stenosis
Marfan's Syndrome	–	Aortic aneurysm, aortic insufficiency (adolescents) and mitral insufficiency (childhood)
Kleinefelter's Syndrome	15%	PDA, ASD
Pierre-Robin's Syndrome	30%	VSD, PDA, ASD, COA
VACTERL association	50%	VSD
CHARGE association	65%	TOF, aortic arch anomalies
Diaphragmatic hernia	25%	VSD, TOF
Tracheoesophageal fistula and/or esophageal atresia	20%	VSD, ASD, TOF
Anorectal anomalies	20%	VSD, TOF
Omphalocele/Gastroschisis	20%	Undefined
Renal agenesis	20%	Undefined
Horseshoe kidney	40%	Undefined
Extrahepatic biliary atresia	20%	Undefined

VSD: ventricular septal defect, PDA: patent ductus arteriosus, AVSD: atrioventricular septal defect, AS: aortic stenosis, ASD: atrial septal defect, COA: coarctation of the aorta, TOF: tetralogy of Fallot.

Clinical assessment

Upon detection of heart murmur during a routine medical appointment in an asymptomatic patient, the chance of an undiagnosed heart disease, an innocent murmur or another disorder affecting auscultation (transient or not) must be considered. Therefore, the general assessment of children is just as important as the specific cardiovascular assessment.[17,19,20]

Changes in growth and/or developmental patterns, although nonspecific, suggest the presence of a severe disorder, either cardiac and/or in other systems, but not an innocent murmur. Weight compromise can occur in ventricular dysfunctions with low output and in the large left-right shunts with pulmonary hypertension. These are also associated with a clinical status of recurrent pneumonia and/or recurrent or persistent wheezing. Weight, structural or developmental compromise can occur in congenital heart diseases with low pulmonary blood flow, which lead to severe hypoxemia and/or thromboembolism. The child's general status can indicate a genetic syndrome or a hereditary disease, as well as reveal a state of discomfort, which would suggest the need for urgent care. Some previous pathological events are relevant, such as vomiting and frequent regurgitation (suggesting a compressive syndrome of vascular malformation); arthritis and/or arthralgia (suggesting an acquired heart disease, such as rheumatic fever or infectious myocarditis); recurrent pneumonia and/or perennial or severe wheezing (suggesting a heart disease with low pulmonary blood flow, such as atrial or ventricular septal defects); and, mainly, anemia, which can be manifested by transitory changes in cardiac auscultation (hyperkinetic state), then develop with cardiovascular compromise (sickle cell anemia). Anemia can also aggravate cardiac insufficiency and hinder the assessment of cyanosis in normal subjects and in those with cyanotic heart diseases.[17,19,20]

On the other hand, the following symptomatology is strongly associated with cardiovascular disorders and should be adequately evaluated in children with heart murmur: cardiac arrhythmia, cyanosis, hypoxemic episodes, syncope, thoracic pain, eating difficulty and/or excessive head sweat, intolerance to physical activity, headache and arterial hypertension (especially in young children), tachycardia and dyspnea, edema and hepatosplenomegaly. Arrhythmias and cyanoses are very indicative of heart disease, while other signs and symptoms are common to several other

disorders. Children with arrhythmia must be referred for investigation, as well as those with perioral cyanosis and cyanosis of the nail bed, if a differential diagnosis has been established with mild vasomotor cyanosis and the levels of oxygenation and hemoglobin are adequate. Hemoglobin levels below 8 mg/dl turn cyanosis into a practically unperceivable disorder, even in patients with right-left shunt.[6,14,15,20]

Infants with congestive heart failure or left-right shunt can present with eating difficulty and excessive head sweat, pallor and irritability; at this age, these can also be manifestations of a cardiac rhythm disorder. Sucking represents an especially great effort for the infant, comparable to exercise intolerance, requiring an adequate evaluation of older children in regard to the type of symptomatology presented and to the level of restriction imposed. The current situation must be compared with previously acquired abilities, lifestyle, sports activities, and performance in relation to same-age children. The degree of intolerance to exercise is directly proportional to the severity of the heart failure in patients with heart diseases.[14,15]

Syncope is a fast and sudden loss of consciousness, which occurs either during exercises or at rest, and can be caused by heart diseases that reduce the flow of arterial supply to the brain (such as severe aortic stenosis), arrhythmias, neurological and metabolic disorders, and others. Syncope can be the initial manifestation of hypertrophic cardiomyopathy during a physical activity, even in the absence of a previous auscultatory change.[24] It must be differentiated from a hypoxemic respiratory failure, which constitutes a sequence of events indicating severe hypoxemia and acidemia, possibly leading to coma and death in patients with cyanotic heart disease, such as tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, and pulmonary stenosis. Hypoxemic episodes can be triggered by exercise, crying, waking, and bowel movement. Older children, when perceiving the episode, usually assume a defensive position, squatting, while infants may assume, as a preferred position, sleeping with their knees folded up against their chest.[25]

Thoracic pain is the second most common reason for referring a child to a cardiologist, although heart diseases are only responsible for 4% to 6% of such pains in children.[1,26] Acute thoracic pain can be caused by pericarditis, traumas, pleurisy, pneumonia, asthma, or foreign body in esophagus. Recurrent or chronic thoracic pain can be associated with muscle strain, costochondritis, esophagitis, mitral valve prolapse, congenital and acquired coronary artery disease, asthma or arrhythmia. In children, angina pectoris can occur in severe left heart obstructive disease, such as aortic stenosis, hypertrophic subaortic stenosis, hypertrophic cardiomyopathy, and in Kawasaki disease. In rare cases of coronary anomaly, angina can occur in the first days of life, manifested by excessive irritability. Mitral valve prolapse and

arrhythmia, palpation and tachycardia can be referred by the child as thoracic pain, and not as changes in heart rate.[6,10,26]

Headache can be a manifestation of systemic arterial hypertension and, when associated with syncope, it can be part of the initial symptomatology of obstructive vascular phenomena.

A detailed physical examination can provide data to assist mainly in the differential diagnosis of innocent and pathological murmurs. Such data must be valued, since auscultation in children is often difficult, and pediatricians are often unprepared to diagnose specific changes.[6,11,17] Standard measurements of heart and respiratory rate, arterial blood pressure, weight, and others must be performed repeatedly and interpreted according to age group standards. All these measurements must be taken when the child feels calm. A respiratory rate equal to or above 60 movements per minute, at rest, is abnormal even in newborns; tachypnea, with or without dyspnea, can suggest left heart failure. Measurement of arterial blood pressure in the four limbs is required for the diagnosis of coarctation of the aorta (a difference equal to or above 20mmHg, with hypotension in lower limbs, is considered significant); pulse symmetry must also be assessed. The child's general status can suggest the presence of hereditary or genetic disorders. The presence of pallor, cold head sweats, with or without cyanosis, and without anemia suggest congestive heart failure. Digital clubbing, changes in perfusion, collateral circulation, arthritis, erythema marginatum, and other changes need to be evaluated, as well as the presence of edema, hepatomegaly, and jugular venous distension, which occur in decompensations of the right heart. Hepatosplenomegaly is more common in bacterial endocarditis and severe anemia; abdominal examination must include an auscultation of murmurs suggestive of arteriovenous fistulas or aneurysms. The cervical region must also be screened in search of pulse changes, murmurs, thrills, visible or palpable heartbeats in the sternal furcula (suggestive of hyperkinetic states, disorders of the aorta or aortic valve); the thyroid must also be examined.[19,20]

In thoracic examination of children with murmurs, a precordial bulge may suggest chronic heart hypertrophy, especially in the left ventricle, and the presence of Harrison's groove may point to left-right large shunts and rickets. Pectus carinatum or excavatum may be associated with heart diseases, such as mitral valve prolapse, but rarely with cardiomegaly, altering the cardiac silhouette in chest x-rays. In heart diseases with pulmonary overflow, such as VSD, PDA, mitral or aortic insufficiency, the thorax seems "active": an increase of the anterior-posterior diameter is observed, heart pumps are visible, and the ictus cordis is deviated. However, in skinny children, the ictus and cardiac pumps can be normally visible, though without deviations from the cardiac axis. During palpation of the thorax, the location and extension of the *ictus* and the presence of thrills must be investigated.[20]

In newborns, the horizontal position of the heart makes the *ictus* palpable at the fourth left intercostal space, towards the outside, to the left of the mid-clavicular line; in school-aged children and adults, in the fifth intercostal space, towards the inside of the mid-clavicle line. Deviations occur in cases of hypertrophy of the heart chamber, pulmonary diseases, changes in the position of the diaphragm and the spinal cord.

The identification of thrills is always suggestive of heart disease, especially when associated with murmur. At the upper left sternal border, it suggests pulmonary stenosis and stenosis of pulmonary arteries; in the lower left sternal border, VSD; in the upper right sternal border, aortic stenosis; in the sternal furcula, aortic stenosis, PDA or coarctation of the aorta.

Pulse palpation is also extremely important: high-amplitude pulses in the upper extremities and weak or absent pulses in the lower limbs suggest coarctation of the aorta. High-amplitude pulses occur in hyperkinetic states; PDA with hemodynamic complications; large systemic arteriovenous fistulas; aortic insufficiency (water hammer pulse); and in premature newborns. Low-amplitude pulses are associated with left heart hypoplasia, local trauma, tachyarrhythmia, cardiac insufficiency and shock (low cardiac output). Asymmetric pulse is associated with local vascular malformations and coarctation of the aorta (right radial pulse with higher amplitude than left pulse).[17,19,20]

Cardiac auscultation must be systematized, performed repeatedly and when the child feels calm, evaluating the sounds where they are usually more audible. Therefore, the first sound, S1, (closure of atrioventricular valves at the beginning of the ventricular systole) must be evaluated at the apex of the heart, and the second, S2, at the base of the heart. The detection of the third (S3) and fourth sounds (S4) (gallop sounds) are suggestive of heart disease. In children, the greatest difficulty is the auscultation of S2 (asynchronous closure of the semilunar valves - aortic and pulmonary - in this order), which is usually a split sound, increasing in inspiration and decreasing or becoming only one sound with expiration. The absence of a split (single S2) or a wide split usually indicate abnormality. S1 is rarely split and, when abnormal sounds are heard, it is important to differentiate them from ejection sounds (clicks) of pulmonary stenosis and S4 (atrial gallop). Other equally important changes in cardiac auscultation are sound hyperphonesis and hypophonesis, fixed splits, clicks and snaps.[6,11,17,19,20]

Characterization of innocent heart murmurs

Innocent murmurs, which occur in normal cardiovascular systems, have some characteristics in common, such as: being more easily audible in hyperkinetic circulatory states; being systolic or continuous; never occurring in isolation in diastole; having short duration and low intensity (1+/4+); not being associated with thrills or accessory sounds (snaps,

clicks); being located in a small and well defined area; not being associated with sound changes; occurring in the absence of a previous history of murmur or evidence compatible with acquired cardiac disorder; being associated with normal chest x-ray and electrocardiogram (although changes can be observed in echocardiogram, such as the finding of false tendon in the left ventricle).[2,6,28]

The most frequent innocent murmurs in children are vibratory (Still's) murmur, pulmonary ejection murmur, pulmonary branch murmur, supraclavicular murmur, and venous hum. The origin of innocent murmurs is still controversial; most of the time, it is attributed to turbulent outflows, originated from narrow areas of the left (Still's murmur) or right ventricle output (pulmonary ejection murmur), or from areas of arterial branching (supraclavicular murmur). In the case of venous hum, the origin is attributed to the turbulence originated from the venous return in the confluence among innominate, internal jugular and right subclavian veins. These turbulences are probably more audible in children because the outflow tracts are proportionally narrower, and the cardiac structures are closer to the thoracic wall, which is thinner in comparison to adults.[2,6,11,28] Still's murmur has been associated with the presence of false tendons; however, as the prevalence of this finding is high in adults (15% to 20%), it becomes difficult to justify the disappearance of such murmur, common in childhood, during adult life.[28]

Still's murmur is the most common innocent murmur, identified in 75% to 85% of school-aged children, and being rarely found in younger children or adolescents. It is better detected in the medial left sternal border or between the lower sternal border and the xiphoid appendix, in supine position. Still's murmur occurs at the beginning of the systole, with vibratory characteristics, low intensity, and it is never unpleasant or very loud. The murmur can disappear with the pressure exerted by the device over the thorax, and its intensity decreases while standing. Differential diagnosis is made with low diameter VSD, hypertrophic cardiomyopathy, and discrete subaortic stenosis, which present murmurs without melodic patterns, greater intensity, invariable with changes in position, and possibly associated with thrills (Table 3).

Pulmonary ejection murmur is found in children, adolescents and young adults, being most common between the ages of eight and 14. More audible in the upper left sternal border, these murmurs are protosystolic, ejective, with low intensity and non-vibratory, like Still's murmur. Pulmonary ejection murmur is better auscultated in supine position; it is exacerbated in the presence of *pectus excavatum*, flat chest and kyphoscoliosis. Differential diagnosis is made with atrial septal defect (which is usually has a S2 fixed split) and valvar pulmonary stenosis.

Pulmonary branch murmur occurs frequently in newborns, especially in premature and low birthweight infants. It originates from relative hypoplasia of the right and left pulmonary branches. Persistence after six months

Table 3 -   Differential diagnosis of innocent heart murmurs

Type of innocent heart murmur	Differential diagnosis
Still's murmur	Ventricular septal defect (VSD) Hypertrophic cardiomyopathy Mild subaortic stenosis
Pulmonary flow murmur	Atrial septal defect (ASD) Pulmonary valve stenosis
Pulmonary branch murmur	Pulmonary stenosis Pulmonary branch stenosis
Supraclavicular bruit	Aortic stenosis Bicuspid aortic valve Pulmonary stenosis
Venous hum	Patent ductus arteriosus (PDA) Cervical arteriovenous malformation

of age suggests a pathological stenosis of pulmonary branches. Better auscultated in the upper left sternal border, these murmurs are ejective, systolic, with low intensity, irradiating towards the left and right sides of the thorax, axilla, and back. This innocent murmur imposes the greatest difficulty for pediatric evaluation, since it occurs in an age group in which the incidence of heart disease is significantly high. Therefore, its diagnosis is to be made by a cardiologist.

Supraclavicular carotid or systolic murmur can be heard in normal children of any age, being better asculated above the clavicles, in the supraclavicular fossa and/or unilaterally in the neck, above the carotid arteries. With a low tone, it begins suddenly and, in the half or two thirds of the systole, the intensity either decreases or disappears completely when shoulders are hyperextended. Differential diagnosis includes aortic stenosis, bicuspid aortic valve, and pulmonary stenosis, considering that, in these pathologies, murmurs are of greater intensity in the base, with irradiation towards the neck.

Venous hum is the only continuous innocent murmur, affecting children between the age of three and six. It is heard in the anterior lower part of the neck, supraclavicular region, possibly extending to the infraclavicular area of the anterior thoracic wall, bilaterally. Venous hum can either change or disappear when the patient turns the head to the opposite side of the murmur, or when finger pressure is applied to the jugular vein. Differential diagnosis is made with cervical arteriovenous fistulas and PDA, considering that, in this pathology, murmurs have clearly audible systolic and diastolic components, and that the sound is more intense in the left infraclavicular region, or in the upper left

sternal border, associated with palpation of high-amplitude peripheral pulses.

Characteristics of pathological murmurs

Pathological murmurs also have common characteristics that suggest the presence of a cardiovascular system disorder: isolated occurrence in diastole or continuous murmur; greater intensity (2+/4+ or over) or unpleasant tone; well defined and fixed irradiation to other areas; association with normal cardiac sounds (sound hyperphonesis, clicks and snaps) and/or thrills; association with symptomatology suggestive of heart disease, especially cyanosis and changes in rhythm and pulse palpation; abnormal lab tests, with the presence of changes in heart size and/or cardiac silhouette, or pulmonary vascular abnormalities in chest x-ray, changes in electrocardiogram and/or in echocardiogram.[2,6,11,14,15]

When identifying pathological murmurs, it is important to establish a relation between the time when murmurs occur and the cardiac rhythm, as well as to observe murmur transmission, abnormal S2 splits, accessory sounds, noises, and thrills.

Regurgitant or holosystolic murmurs are never innocent murmurs, being associated with VSD, mitral or tricuspid stenosis. Ejection murmurs can be innocent, such as Still's, but those with increasing-decreasing patterns can be associated with stenosis or deformities in semilunar valves (aortic and pulmonary). Diastolic murmurs are always pathological and occur due to aortic or pulmonary insufficiency, stenosis, and changes in outflows through mitral or tricuspid valves. The same pattern is identified in

continuous murmurs, which are associated with PDA, coarctation of the aorta, and pulmonary artery stenosis (except in venous hum).

Murmur transmission is also indicative of heart disease; therefore, for example, a systolic ejection murmur, heard at the base of the heart and transmitted to the neck, suggests an aortic murmur. On the other hand, murmurs that are transmitted towards the dorsum suggest pulmonary murmur.

In regard to S2 splits, wide split S2, single S2 or paradoxical split S2 are always considered abnormal, showing changes that extend ejection time for the right ventricle (or shorten for the left one), or disorders of electrical conduction in the heart. Hypophonesis in S2 suggests the absence of pulmonary and aortic components, and, therefore, stenosis of these valves. Fixed split S2, not varying while breathing, suggests atrial septal defect; when no audible splits are found in the tricuspid area, it suggests bad position of the great vessels at the base of the heart. Hyperphonesis of an aortic component of S2 suggests systemic arterial hypertension or dilatation of the aortic root. Hyperphonesis in a pulmonary component suggests pulmonary hypertension, and hypophonesis suggests pulmonary stenosis, tetralogy of Fallot or tricuspid stenosis.

Laboratory investigation

The definition of innocent murmur includes normal results on chest x-rays, electrocardiogram and echocardiogram, although several authors do not point the need of these exams, if the initial clinical approach and follow-up are carefully done and reveal the characteristics of normality previously discussed here. When a more detailed diagnosis is needed, the best option, with the lowest cost, is to refer the patient to a cardiologist, who will request more adequate ancillary exams. The occasional request for exams must be delayed when a murmur with innocent characteristics is detected in the presence of hyperkinetic states.[2,6]

In practice, pediatricians can evaluate previously taken tests, such as, for example, in cases of children with frequent wheezing or recurrent pneumonias, who have already taken several chest x-rays in different situations. These radiographs can be useful in the diagnosis of heart diseases, in the evaluation of pulmonary complications secondary to these diseases, and in the differential diagnosis. In the evaluation of the cardiac area, the adequate positions are posterior-anterior and lateral, during inspiration and while standing. In children, a cardiothoracic index (the relation between the greatest transversal diameter of the heart and the greatest internal diameter of the thorax) equal to or below 0.6 is considered normal. Changes in cardiac silhouette can suggest pathologies in the following situations: boot-shaped heart, with low pulmonary flow (tetralogy of Fallot); "snowman" appearance (supracardiac anomalous drainage of pulmonary veins); and ovoid shape, narrow pedicle, pulmonary overflow (transposition of the great arteries). Coarctation of the aorta

can be considered if the ribs present notching in the lower border.

When the size of heart chambers is assessed, an increase in the left atrium size can be detected through double cardiac contour, elevation of the left main bronchus and protrusion of the left atrium. An increase in the left ventricle makes the tip of the heart deviate towards the left and downwards (into the diaphragm). Laterally, the lower cardiac border is dislocated towards the spinal cord. An increase in the right atrium size creates a protuberance in the right upper region of the cardiac silhouette, while the right ventricle can only be identified if the tip of the heart is elevated; laterally, retrosternal space is lost. Pathologies of the great vessels modify the pedicle. In evaluation of pulmonary lobes, data suggesting overflow are increase of pulmonary hilum (pulmonary trunk and pulmonary arteries) and presence of visible vascular patterns on the top and lateral third of pulmonary lobes; data suggestive of low flow are reduced images of the pulmonary hilum, darkened pulmonary lobes and thin vessels. Images of diffuse opacification in pulmonary lobes suggest venous congestion.[6,29,30]

When electrocardiogram (ECG) and echocardiogram are requested, they must be interpreted considering their peculiarities in children. In regard to ECG, since birth, physiological overload is the standard in the right ventricle, although this continuously changes to a left ventricle overload.[29]

In younger children, echocardiogram is usually performed under sedation and, due to the low calcification of the breastbone, the echocardiographic window is larger, allowing for a better visualization of the heart and great vessels. A study mapping the flow in colors can better detail small defects, such as apical ventricular septal defect, small arterial vessels, and complex heart diseases.[31]

There are other questions regarding cardiovascular lab tests in children, such as the performance of preventive exams for adult cardiovascular disorders and exams in children who practice sports regularly and extenuatingly.[31-33] In principle, surveillance of obesity, arterial hypertension, adequate physical activity and use of tobacco and drugs is part of the overall pediatric evaluation and guidance. Preventive exams for dyslipidemia must be restricted to children belonging to risk groups, such as those with metabolic disorders and/or first-degree family members with atherosclerotic disoders.[29-31]

In regard to children or adolescents who practice strenuous physical activities, there is some concern with congenital or acquired heart diseases not yet diagnosed, especially septal defect, hypertrophic cardiomyopathy, Kawasaki disease and others, which can remain asymptomatic for long periods, and only manifest during such practices, imposing a life threat to these patients. Here, also, familial and personal backgrounds are important,

especially when there is a family history of cardiomyopathy and complaints of respiratory disorders, hypertensive episodes, thoracic pain, syncope, hypotensive response, and exercise intolerance. In these situations, a request for exams is more complex, and the evaluation by a cardiologist is the most adequate procedure.[35,36]

Referral to a specialist

It can be suggested that, in certain situations, some urgent and others not, children should be referred to a cardiologist for diagnosis and treatment.[6,7,9,36,37] These situations include: when heart murmur or any other auscultatory change presents characteristics that have been described here as pathological; when heart murmur has characteristics considered innocent but accompanied by symptomatology and/or changes on physical examination suggestive of cardiovascular disorder; when children belong to risk groups for cardiovascular disorder, especially in families in which the incidence of congenital heart disease and sudden and/or early death is high, even if they do not present changes on physical examination; when changes in cardiac auscultation and/or symptomatology suggestive of cardiovascular disorder are detected in children less than one year old, especially during the neonatal period; when patients who had initial diagnosis of innocent murmur show, during pediatric follow-up, changes in auscultation and/or clinical history that can suggest cardiovascular disorder; and when requested lab tests suggest cardiovascular changes. Additionally, children with signs and symptoms that can suggest cardiac insufficiency must be urgently referred, even in the absence of auscultatory changes. Finally, those cases in which diagnosis was made during a hyperkinetic state, but clinical and/or auscultatory changes remain after the underlying disease has been resolved, especially in the case of infectious diseases, must also be referred to a cardiologist.

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