114 The Chad With A Heart Murmur

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Detection of a murmur in the heart of a child is a cause of anxiety to the parents, as it is generally considered to be the hallmark of an underlying heart disease. Treatment of heart disease is fraught with difficulties including the possible need of a major cardiac intervention or surgery involving high costs, high morbidity and mortality.

However it should be remembered that all cardiac murmurs in children are not associated with heart disease for as many as 50-60 percent of murmurs heard in young children are either functional or innocent. It is important to distinguish these from pathological murmurs. Also some of the serious heart diseases may occur without any murmur eg, myocarditis, dilated cardiomyopathy, pericardial effusion and cardiac arrhythmias etc. A clinician must know when to refer a child with cardiac murmur to the pediatric cardiologist for further workup and when to reassure the parents regarding the innocent nature of the murmur.

Causes Of Cardiac Murmurs

Murmurs are produced by noisy blood flow through heart or vessels. This may be through normally structured heart and vessels (innocent or functional murmur) or through abnormal heart (pathological murmur).

Innocent or functional murmurs have certain characteristic features described later. The exact reason for these murmurs are not known but it is postulated that there may be due to a small aortic size in children resulting in increased velocity of blood across the aorta during ejection, left ventricular false tendons producing exaggerated vibrations with ventricular contractions or increased cardiac output. Described below are the various innocent murmurs heard in children:

Systolic murmurs

- **Stills murmur** (vibratory systolic murmur) is a mid systolic murmur heard loudest at mid point between left mid sternal border and apex, is a grade 2-3 murmur, has a low frequency (150 Hz) and is best heard supine. It is due to periodic vibrations of semilunar valve, ventricular wall, great artery wall, pulmonary cusps or false tendons in LV.

- **Pulmonary flow murmur** of childhood is an ejection systolic murmur of grade 1-3, heard in thin chest adolescents (8-14 years). It is like a murmur of Pulmonary stenosis (PS) but without a click or thrill and P2 is normal. It is due to vibrations of pulmonary trunk during ventricular systole. *Pulmonary flow murmur of infancy is* ejection systolic murmur heard in

newborns and radiates from lower left sternal border to both axillae and back. It is due to the relatively small pulmonary artery branches as well as the angle of take off from the pulmonary artery. In the fetus, the pulmonary trunk is dilated as it receives its output from the high pressure of the right ventricle. Left and main pulmonary arteries are small branches, as the lungs receive no blood. It disappears by 6months.

- **Supraclavicular murmurs** are grade 3-4 systolic murmurs heard over bracheocephalic artery near aortic origin. They can have thrill and if the shoulder is hyperextended with elbow straight behind the back the murmur decreases. It seldom creates any problem in the differential diagnosis in a child.

Continuous murmurs

- **Venous hum** is the most common innocent murmur with maximum intensity in supraclavicular region, lateral to sternocledomastoid. It may radiate widely. It may be of grade 1-6 and often bilateral. It decreases by compression of internal jugular, lying down or by valsalva maneuver and increases on sitting straight and by deep inspiration. These innocent murmurs are often detected when they increase in intensity during fever, anemia, anxiety or when a child is examined for sports clearance or for any other reason.

Pathological murmurs are produced if there is valvular stenosis {Aortic stenosis (AS), PS, Mitral stenosis (MS)}, valvular leaks {Aortic regurgitation (AR), Mitral regurgitation (MR), Pulmonary regurgitation (PR), Tricuspid regurgitation (TR)}, flow of blood across shunts {Atrial septal defects (ASD), Ventricular septal defects (VSD), Patent ductus arteriosus (PDA)}, narrowing of vessels (Coarctation of aorta) or certain AV fistulas. More than one lesion may coexist and more than one murmur may be present in one patient. Character, timing and site of these murmurs depend on the underlying lesion and these along with other findings on physical and cardiac examination help in the diagnosis of underlying heart disease.

How The Diagnosis Is Confirmed

A family practitioner should be aware of the features that differentiate an innocent murmur from a pathologic one. The presence of the following features warrants referral to a child specialist and further evaluation:

- Symptoms of chest pain, congestive heart failure (CHF), cyanosis
- Family history of sudden death in a young member
- Malformation syndrome
- Increased precordial activity
- Decreased femoral pulses
- Abnormal intensity or splitting of S2
- Clicks or opening snaps
- More than grade 3 systolic murmurs
- Any diastolic murmur
- Increase in the intensity of murmur on standing. Evaluation of a murmur in a child requires complete history and clinical

examination. History of cyanosis/ cyanotic spells and CHF (excessive sweating, poor feeding, rapid breathing) should be elicited.

Examination includes looking for cyanosis, anemia, all pulses and features of CHF {tachypnea, tachycardia, raised Jugular venous pressure (JVP), edema feet and hepatomegaly}. JVP is difficult to assess in infants and edema feet is a late sign in this age group.

Position and character of apex beat, precordial activity and thrills should be looked for. Intensity of SI and S2 and splitting of S2 should be checked. Various systolic, diastolic and continuous murmurs may be heard.

Classification of intensity of murmurs

- Grade I is the faintest murmur that can be heard (with difficulty)
- Grade II murmur is also a faint murmur but can be identified immediately
- Grade III murmur is moderately loud
- Grade IV murmur is loud and associated with thrill.
- Grade V murmur is very loud but cannot be heard without the stethoscope It may be heard with edge of the stethoscope on the precordium.
- Grade VI murmur is the loudest and can be heard without a stethoscope ie stethoscope being at some distance away from the precordium.

The gradation of intensity is purely subjective. However, it allows recognition of changes in the intensity of the murmur, which has diagnostic relevance.

Table 1: Causes of systolic murmurs

		Pathological				Innocent
Ejection-systolic		**Holosystolic-murmurs**		**Late Systolic murmurs**		
Causes	**Associated diagnostic findings**	**Causes**	**Associated diagnostic findings**	**Causes**	**Associated diagnostic findings**	**Causes** (These have no other associated diagnostic clinical findings)
Obstructed semilunar valves		**Valvular leaks**				
AS	LV heave, thrill in suprasternal notch,,ejection click in valvular AS, Narrow, single or paradoxical split of S2	MR, TR	TR alone is rare and does not need differential diagnosis	MVP	Midsystolic clicks, beat to beat intensity variation	Pulmonary flow murmur
PS	Thrill in suprasternal notch, ejection click best heard in expiration. P2 absent in infundibular PS and soft but present in valvular PS.					Stills murmur
HCMP	Cardiomegaly and heaving apex					Spraclavicular murmur
Increased blooc normal valve flow through		**Shunts**				
ASD,VSD ,TAPVC (flow through pulmona ry valve)	Wide and fixed split of S2 in ASD	VSD	Harsh murmur, mid diastolic murmur at apex or AR murmur may coexist			
AR, PR (Excess flow Through aortic or pulmona ry valve)	But features of AR/PR, ie, diastolic murmur and wide pulse pressure in AR will predominate					

MVP: Mitral valve prolapse
TAPVC: Total anomalous pulmonary venous return
HCMP: Hypertrophic cardiomyopathy

- **Systolic murmurs** occur between S1 and S2 (first and second heart sounds), and therefore are associated with mechanical systolic and ventricular ejection. Mid-systolic murmurs typically have a crescendo-decrescendo character, that is, they start softly and become loudest near mid-systole, followed by a decrease in sound amplitude. This type of murmur is caused by either aortic or pulmonic valve stenosis. A second type of systolic murmur is holosystolic (sometimes called pansystolic) because the amplitude is high throughout systole. This type of murmur is caused by MR, TR, or by VSD.

- **Diastolic murmurs** occur after S2 and are therefore associated with ventricular relaxation and filling. They may be caused by aortic or pulmonic valve regurgitation, or by mitral or tricuspid valve stenosis. They can occur early (eg, AR), mid-diastolic, or late diastolic (eg, MS).

- **Continuous murmurs** are heard in systole as well as they continue in diastole.

Tables 1-5 shows causes and characteristics of a murmur in a child without cyanosis and Table 6 shows the same in a child with cyanosis.

Almost 50 percent of patients with congenital heart diseases will not have a murmur at birth and so the absence of a murmur at birth does not rule it out. Thus, currently there is no known method of spotting these neonates for further evaluation at this time.

Investigations

X-ray and ECG: Most of the times there is no need for an x-ray and ECG for suspected innocent murmurs. They are unlikely to reveal clinically unsuspected heart disease.

The role of x-ray and ECG is questionable even in cases where cardiac pathology is strongly suspected. Various studies have shown that x-ray and ECG can be normal with heart disease and sometimes can be abnormal without it. These (x-ray and ECG) are however helpful in follow up and in reducing the frequency of serial Echo's. Also, there may be other findings in the x-ray eg, pneumonia or rib notching in Tetralogy of Fallot cases with bronchial collaterals, which may have their own implications in patient management.

Table 2: Listening areas for various pathological systolic heart murmurs

RUSB	Valvular AS
Apex	MR, (radiates to left axilla and back),MVP
LLSB	VSD, TR, HCMP (subvalvular AS)
LUSB	PS, ASD, TAPVC

LUSB: Left upper sternal border
LLSB: Left lower sternal border
RUSB: Right upper sternal border

Table 3: Causes of diastolic murmurs

Early diastolic murmurs	Valvular leaks of major vessels ieAR (soft systolic murmur, difficult to detect. Listen with patient sitting up, leaning forward, in full expiration), also wide pulse pressure and other peripheral signs of AR). PR (congenital rare. Mostly after surgery of TOF or balloon pulmonary valvotomy)
Mid diastolic murmurs	Narrowing of atrioventricular valves ie, MS, LV filling murmurs (VSD, PDA, MR) Decreased LV compliance (AS, Coarctation, LVCMP)
Late diastolic murmurs or presystolic murmurs	Late atrial contraction forcing blood through narrow atrioventricular valves ie, severe MS

LVCMP: Left ventricular cardiomyopathy

In cyanotic patients, an x-ray may be helpful in differentiating cardiac or respiratory cause of cyanosis. They are generally ordered in all pathological murmurs. Useful findings on x-ray are cardiomegaly, pulmonary vascularity etc, and on ECG are right ventricular hypertrophy, left ventricular hypertrophy, right bundle branch block or arrhythmias. Table 7 shows some common E C G findings and associated heart diseases.

Table 4: Listening areas for various diastolic murmurs

Site	**Lesion**
RUSB	AR
LUSBorLMSB	ARPR
Apex	MS, Increased LV filling (VSD, PDA, MR), decreased LV compliance (AS, Coarctation, LVCMP)

Table 5: Causes and Listening areas for various continuous murmurs

Site	Lesion
Pathological	
LUSB	PDA (most common cause of continuous murmurs), AP window of aorta
Back	Coarctation
Axilla	Peripheral PS
Site of fistula /shunt	AV fistulas and surgical shunts
Innocent	
Left and right upper chest	Venous hum

Table 6: Flow chart for the diagnosis of heart disease presenting with a heart murmur and cyanosis

Increased pulmonary blood flow (Increased vascular markings on x-ray)		**Decreased pulmonary blood flow Decreased vascular markings on x-ray)**	
Malformation	**Clinical findings other than systolic murmur**	**Malformation**	**Clinical findings other than systolic murmur**
Transposition of great arteries, single ventricle Tricuspid atresia	None specific	**With pulmonary artery hypertension**	
Total anomalous Pulmonary venous return	Increased precordial activity and bulge, Wide and fixed split S2 may be present but not a must.	Eisenmenger complex	Loud p2
Truncus arteriosus	Wide pulse pressure, Aortic ejection click after SI. Loud SI and single S2	**Without pulmonary artery hypertension**	
		ASD/VSD with PS TOF physiology (TOF, DORV, TGA with PS, Pulmonary atresia, single	Single S2

		ventricle ie, SV with PS (TA) with PS	
		ECD	Wide split S2
		Ebstein's anomaly	Cardiomegaly

Ebstein's anomaly ECD: Endocardial cushion defects, DORV Double outlet left ventricle, TGA Transposition of great arteries, TA: Tricuspid Atresia

Table 7: Main x-ray and EGG findings in heart disease X-ray findings Condition

X-Ray Findings	**Condition**
Cardiomegaly	Cardiomyopathy, large shunt (VSD, ASD, PDA, single ventricle), severe AR, MR, ebstein's anomaly, pericardial effusion, truncus arteriosus
Increased pulmonary blood flow	Shunts (VSD, ASD, PDA, single ventricle, ECD) transposition of great arteries, tricuspid atresia, truncus arteriosus.
Decreased pulmonary blood flow	TOF physiology (TOF, DORV, TGA with PS, pulmonary atresia, single ventricle with PS) tricuspid atresia with PS
ECG findings	**Condition**
LVH	AS, HCMP, VSD, MR, AR, SV
RVH	TOF, TGA, MS, ASD, ECD, SV
BVH	Large VSD, SV, truncus arteriosus
PAH	MS, eisenmenger complex, primary PAH
LAD	TA, AS, AR
RAD	TOF physiology

Note: It is important to know that these are some of the common salient findings and there may be more specific and non-specific findings. The E C G may be normal also. There are many complex congenital malformations other than the ones mentioned above and Echo is the only reliable diagnostic tool especially in cyanotic congenital heart disease.

Echo: The Echo would be the most appropriate and key investigation required while evaluating pathological murmurs. However, in suspected innocent murmurs there is no role of Echo. However any murmur in neonates should be investigated by Echo as cardiac murmurs are rare in newborns (<1%) but nearly half of those with a murmur will have heart disease.

Cardiac catheterization is required only when a definite intervention or cardiac surgery is being considered and not for initial diagnosis. Detailed analysis of anatomy, quantitation of shunts and pressures can be done with this.

How Is It Managed

Innocent murmurs require reassurance and infrequent follow up (may be once or twice in a year during childhood or till murmur disappears). Pathological murmurs require further investigations and after complete diagnosis of the underlying heart disease is made, it is managed accordingly. Medical management includes managing congestive heart failure, infective endocarditis, rheumatic prophylaxis, pharmacological closure of PDA in neonates etc. Cardiac intervention or surgery is done at the appropriate age.

Key Points

- Majority of murmurs in children are innocent.
- Some serious heart diseases can occur without murmurs.
- X-ray, ECG and Echo are not required in the evaluation of suspected innocent murmurs.
- X-ray and ECG have limited role in the evaluation of heart disease in pathological murmurs but Echo is the key investigation in such children.
- All cardiac murmurs in new born should be investigated by Echo.
- Cardiac catheterization is required only when intervention is being considered.
- For Innocent murmurs only reassurance is required and heart disease associated with pathological murmurs should be treated medically or surgically as appropriate.

Reading List

1. Allen HD, Phillips JR, Chan DP. History and physical examination. In: Allen HD, Gutgesell Hp, Clack EB, Driscoll DJ (eds) Moss and Adams Heart disease in infants, children and adolescents. Philadelphia: Lippincot Wiliams and Wilkins; 2000; 143.
2. Friedman W Congenital heart disease in infancy and childhood. In: Heart disease. I Braunwald E (ed). Philadelphia: WB Saunders. 1997; 877.
3. Gessner IH. Evaluation of the infant and child with a heart murmur. In: Pediatric cardiology - A problem oriented approach. Gessner IH and Victoria BE (eds) Philadelphia: WB Saunders Co;1993; 131.
4. Perloff JK. Normal or innocent murmurs. In: The recognition of heart disease Perloff JK. (ed). Philadelphia: WB Saunders; 1987:8.