

REVIEW ARTICLES

**GUIDELINES FOR ECHOCARDIOGRAPHY IN
SUDANESE CHILDREN**

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INTRODUCTION:

The feeling of helplessness among paediatric registrars and even young paediatricians when confronted with a child with a cardiac problem is not without plausible reasons.

Looking into few thousands of referrals for echocardiography in the last 6 years at 3 centres (Sudan Heart Centre, Ja'ffar Ibn Ouf Hospital and the Academy Medical Centre), one could easily pick a vast number whose diagnosis could have been reached by comprehensive history and clinical examination. The reason behind indiscriminate referrals in my opinion, is partly due to the widespread notion that echocardiography is replacing clinical examination. In this article we will try to explain the continuing importance of clinical examination in cardiovascular diagnosis generally, and especially in Sudan, where investigations are still expensive or not available. We will also try to suggest the best application for echocardiography and offer guidelines for its use in paediatrics in Sudan.

Clinical Examination versus Echocardiography:

- + Is it true that the teaching of the old astounding Physicians who diagnosed all cardiac ailments by looking, touching and listening is vanishing?
- + Are the modern diagnostic technologies going to replace the expert clinical examination?
- + The following views of some Clinicians may answer these questions or reflect their worry about the neglect of the clinical approach.

Rapezzi ⁽¹⁾ wrote: Current trends towards routine mass use of sophisticated diagnostic tools are killing off the science and art of clinical reasoning. Besides burning a lot of public and private money to make diagnostic work rather superficial, doctors also risk the intellectual pleasure that comes from careful diagnostic reasoning.

Rapezzi ⁽¹⁾ also wrote: If one investigative quality marks out the mature clinician, it is the ability to spot possible inconsistencies among the clinical, instrumental and laboratory examinations, considering not only what is present, but also what is missing. For well honoured clinicians, the clinical part of the diagnostic investigation is not just a question of medical history and physical examination, but rather the capacity to establish links among various physical and laboratory or instrumental findings with an eye to both the consistencies and inconsistencies. In this context, simple examinations really do have the same as the more complex and expensive ones.

Markel ⁽²⁾ wrote:

But with all due respect to these and a host of other treasured tokens, I contend that the stethoscope best symbolises the practise of medicine. Whether absentmindedly worn around the neck like an amulet or coiled gunslinger-style in the pocket ever ready for the quick draw, the stethoscope is much more than a

tool that allows us to eavesdrop on the workings of the body. Indeed it embodies the essence of doctoring; using human skills of listening to determine what ails a person.

Outlines of cardiovascular examination in the diagnosis of heart lesions^(3,4).

Inspection may be the single most important part of paediatric examination and it is especially so in cardiology. Many cardiac lesions have constant association with special syndromes. Recognising the dysmorphic features of the syndrome may yield the lesion, e.g. Turner's and coarctation of the aorta, Noonan's and pulmonary stenosis, William's and supraaortic stenosis, etc.

Mild cyanosis can easily be missed by the hurried examiner, while dark lips can be mistaken for cyanosis if the tongue is not inspected.

Many important signs are seen and not felt, e.g. clubbing, tachypnoea, precordial bulge and the characteristic subcostal recession of a large left to right shunt.

Careful palpation of the peripheral pulses can add very useful information like the *small pulses* in aortic stenosis, mitral stenosis, acute myocarditis and Eisenmenger's syndrome, the *collapsing pulse* in PDA, aortic regurgitation and truncus arteriosus, the *weak or absent femoral pulse* in coarctation of the aorta. The *bounding large dorsalis pedis pulse* may be the only (test) to diagnose PDA in a premature baby.

Location of the apex gives a good idea about the left ventricle volume, while the left parasternal heave tells about the right ventricle volume or pressure overload.

We always think that the pathology lies below the thrill,

therefore, careful localisation of the maximal vibration of a thrill may localise the lesion. This may be easier for those who have difficulties with murmurs.

Auscultation is the fun and art of the situation. If the triad of: the listener, the stethoscope and the patient work in harmony, then the result will be accurate. The confusion comes from the two common mistakes; the hurried auscultation and the neglect of the systematic approach.

I always preach that if one knows how to auscultate the second heart sound, he knows paediatric cardiology. This may look an exaggeration, but it has much truth in it. A normal second heart sound rules out ASD, PS, TGA, Pulmonary hypertension, APVD and severe AS (a good percentage of paediatric cardiology!!). Fixed splitting of S_2 is almost diagnostic of ASD. Ejection clicks at the base point towards stenotic semilunar valves (Registrars raise eyebrows at the mention of clicks). Mitral valve prolapse has a click at the apex.

A useful oversimplification of murmurs suggests that the intensity of the ejection systolic murmur increases when you go up (towards the sternal notch), while the intensity of a pansystolic murmur increases when you go down (towards the xiphisternum).

Ejection Systolic Murmurs (ESM) are due to PS, ASD, Ao.S, PHT, Large unrestrictive VSD and above all, an innocent murmur. Pansystolic murmurs are heard with small or moderate VSD, mitral regurgitation and tricuspid regurgitation. VSDs and TR may share the same room, but the later changes with respiration.

Mitral regurgitation is not commonly heard in children below 5 years of age (Rheumatic).

A continuous murmur below the left clavicle is due to a PDA (with collapsing pulses). The ESM of a COA is better heard over the back between the scapulae (with hypertension and weak femoral pulses). The MVP murmur is a late systolic at the apex. The majority of MVP children are pre-adolescent girls.

The innocent murmur can be heard in 50% of children with careful auscultation. It is a common reason for referrals to echocardiography.

Diastolic murmurs in children are far less common than systolic. The common two are due to rheumatic aortic regurgitation and mitral stenosis. Large VSDs cause diastolic flow murmurs at the mitral area, while a large ASD generates a diastolic flow murmur at the tricuspid valve area.

With a repeated careful cardiovascular system examination, paediatricians can attain a high degree of experience to diagnose and quantify heart disease in children.

This brief account is meant to display the importance of clinical examination and reassure physicians that even without investigations they can do a lot.

Echocardiography:

No doubt, echocardiography made a big leap in cardiology, adult and paediatric alike. Now types of abnormalities can be determined with precision and the haemodynamic aberrations they produce can be measured easily. Still there are limitations to this modality.

Weyman⁽⁵⁾ wrote: Although many people can be taught to interpret high quality echocardiographic study, far fewer can consistently record high quality images.

In Sudan, due to scarcity of paediatric cardiologists at the present

time, we need persons who can produce and interpret high quality images at the same time. Those persons are expected to plan management immediately. Now we have very few individuals who can perform all this at the shortest time. This will make the service rather expensive.

A good number of children with heart disease in Khartoum and the provinces will be denied the best of the service and that will increase the morbidity and mortality.

The alternative is to ask for the help of adults' cardiologists, which is already going on, but with its price, as they have no much experience with complex congenital problems.

For these reasons we think that guidelines for requesting echocardiography are needed.

The recommendations concerning the use of echocardiography follow the indication classification system used in ⁽⁶⁾:

Class I: Conditions for which there is evidence and/or general agreement that a given procedure or treatment is useful and effective.

Class II: Conditions for which there is conflicting evidence and/or divergence of opinion about the usefulness of a procedure or treatment.

Class III: Conditions for which there is evidence and/or general agreement that a procedure or treatment is not useful/or effective, and in some cases, may even be harmful. This classification may help the requesting Physician see the cost-effectiveness of his request.

The following is a humble trial to help the physicians caring for children in this country to understand the usefulness of

echocardiography.

(1) The Neonate:

There is no age when echocardiography is so important as in this age group, when the haemodynamics are rapidly changing. It is our belief that neonatologists should be taught neonatal echocardiography and the machine should be part of the nursery equipments. This will allow reducing the morbidity due to the delay in performing the procedure, and the risk of transferring tiny unstable babies long distances under unfavourable conditions.

Cyanosis is so far the commonest indication for echocardiography, but one has to be sure that it is central cyanosis (and not only the lips). It is useful to differentiate ductus dependant lesions early (TGA, COA, Hypoplastic left ventricle, severe PS).

The commonest cause of heart failure in this age group is COA, which can be diagnosed clinically, but nobody will be happy to discover that prostaglandin was given to myocarditis!!

Transient murmurs are common and echocardiography may reassure doctors and parents alike. Primary Pulmonary Hypertension is not uncommon, and echocardiography is essential in the diagnosis and follow up.

Respiratory distress by itself is not an indication for echocardiography if a clear non-cardiac underlying cause is incriminated.

An increased cardiac silhouette in chest x-ray not supported by cardiac findings for heart disease is not an urgent indication. Thymus shadow is a common cause for this appearance in neonates and infants.

Transient atrial extrasystoles are benign and usually the heart is normal. Complete heart block needs an echocardiography to exclude an associated CHD. Syndromes known to be associated with CHD need early echocardiography for screening.

(2) Infants:

The most common cause of referral in this age group is large ventricular septal defects. This may be due to systolic murmur with recurrent pulmonary infections, failure to thrive and tachypnoea. The X-Ray invariably shows cardiomegaly with plethoric lungs.

This makes the clinical diagnosis easy. Medical therapy can be started after a clinical diagnosis.

Early referral to echocardiography is indicated when associated cyanotic cardiac lesions are suspected. These are mainly a TGA when a degree of cyanosis is also observed, coarctation when the lower limbs pulses are weak or absent. Failure to respond to diuretics and vasodilators is an indication for early referrals. Evidence of pulmonary hypertension in any age group is an urgent reason for referral. Generally those infants need to be seen by a paediatric cardiologist when 8-12 months old. The decision for surgery is probably made when they cross their first birthday.

Infants with cyanosis may be referred for echocardiography at an early age. A precise diagnosis is needed early. The majority will need some sort of surgery, so parents counselling should be done as early as possible, and a date and place of surgery may be suggested.

(3) Children at all ages:

MURMURS:

Cardiac auscultation remains the most widely used method of screening for heart disease ⁽⁶⁾; Innocent murmurs are common in

children. These are asymptomatic children with short ejection systolic murmur maximally grade 3 (does not cause a thrill) heard along the left sternal border, and the intensity changes with change of the posture of the children and S₂ is normal.

Aubrey Letham, the father of phonocardiography claimed that he could hear innocent murmurs in 100% of children⁽⁷⁾. A typical innocent murmur does not need echocardiography, but in ambiguous situations echocardiography alone can clarify the truth.

Rheumatic heart disease (RHD) needs echocardiography at intervals. A time will come to decide valve replacement if cardiac deterioration is documented.

Infective endocarditis is a situation where echocardiography is essential for diagnosis and follow up.

DYSRRHYTHMIAS:

Most of rhythm abnormalities are associated with normal heart structure (except RHD). The most common irregular rhythm in children is respiratory sinus arrhythmia. This is defined as changes of heart rate with the respiratory cycle. This is benign and does not require echocardiography. Paroxysmal supraventricular tachycardia needs echocardiography to rule out structural diseases, especially ASDs and tricuspid valve diseases. Complete heart block is usually found with normal hearts, but an echo is usually preferred.

If a dysrhythmia is suspected as cause of syncope with a clinically normal heart, probably 24 hours Holter monitor is more appropriate and economic.

HEART FAILURE:

It is unlikely for heart failure to present without evidence of cardiomegaly, but echocardiography will prove a condition like

Meanwhile and ever, the prompt clinical approach should be the primary stage of diagnosis and management of heart disease in children.

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