

Updates in Pediatric Cardiology



East Tennessee Pediatric Cardiology P.C.

Yvonne A. Bremer, MD, FAAP, FACC

Michael R. Liske, MD, FACC, FASE

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Welcome to our Third Issue!

Welcome to the third issue of "Updates in Pediatric Cardiology"! We enjoy using this forum to interact with you as we partner together in taking care of infants, children, teens, and young adults with cardiac concerns.

In our first issue, we began a discussion of the evaluation of cardiac murmurs. We'll wrap up that topic here in Issue 3. We hope you find this

communication helpful, and would ask up front that if you have a topic of interest or an issue of concern, that you notify us.

Going green?

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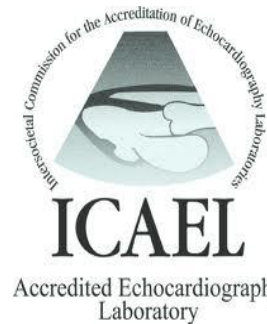
2001 Highland Avenue
Suite B

Knoxville, TN

Phone: 865.971.6897

Fax: 865.971.6817

www.etpc-hearts.com



Grand Rounds at East Tennessee Children's Hospital

On August 16th at noon, Dr. Liske will be presenting, "Acute Acquired Heart Disease: Kawasaki Disease, Myocarditis, and Acute Rheumatic Fever." We hope to see you there!

The Clinical Evaluation of Cardiac Murmurs, Part 2

A. What are the characteristics of innocent murmurs? (Please see the first issue for a complete discussion of these points)

1. Negative cardiac history.
2. These murmurs are only a systolic ejection or continuous type.
3. Innocent murmurs are soft, not harsh.
4. Most (not all) innocent murmurs are louder when the patient is supine
5. The remainder of the cardiac exam is normal.
6. Specifically, the second heart sound is physiologically split.

Please recall the sage advice of the eminent clinical cardiologist, Amnon Rosenthal, "The guilt or innocence of a murmur is more often than not based on other features than the murmur itself."

B. What are some specific innocent murmurs?

1. Still's murmur: This murmur was first described by Dr. George Frederick Still in England in 1909 and it's been around ever since. It may be caused by vibration of the LV chordae tendinae or the LV free wall. It is a very characteristically "musical" systolic ejection murmur loudest between the left lower sternal border and the apex, and it louder when supine. Some describe it as "twangy," or "harmonic," or "vibratory."

2. Venous hum: This is another classic and easily recognizable murmur of childhood that is continuous, present at the left or right infra-clavicular space, is modified by turning the head while sitting, and is substantially diminished in the supine position.

These maneuvers readily distinguish it from the murmur of a patent ductus arteriosus, which does not vary with body position or head turn. The venous hum is caused by a physiologic turbulence of the veins of the upper thorax.

"The younger the patient, the more expeditious should be the evaluation."

3. Innocent pulmonary flow murmur: Vibration of the pulmonary valve and main pulmonary artery is thought to cause this murmur that is present along the left sternal border. Like a Still's murmur, it also can be louder in the supine position but tends to be a bit harsher. This murmur may sound very similar to the systolic murmur of an atrial septal defect or valvar pulmonary stenosis; however, the absence of wide splitting of S2, absence of a diastolic rumble, absence of a systolic click, and absence of an RV lift would be more supportive of an innocent condition.

4. Innocent peripheral pulmonary stenosis: "PPS" is commonly noted in newborn babies. It is caused by a relative narrowing and acute angle take off of the branch pulmonary arteries. The murmur can be quite high pitched and if auscultation is confined to only the precordial area, it may lead the practitioner to believe there is a small ventricular septal defect. To make this diagnosis, one should note that the murmur is as loud over the lung fields as the precordium. Special care should be taken to confirm normal femoral pulses since a coarctation of the aorta gives a similar murmur at the posterior intrascapular space. Also, if this murmur doesn't resolve by 6 months of age, it may suggest pathologic PPS, an ASD, or other conditions.

5. Innocent aortic flow murmur: This murmur is loudest at the left mid sternal border and right upper sternal border and may radiate into the neck. It is caused by physiologic turbulence across the aortic valve or subaortic space. Three lesions that must be distinguished from this innocent murmur are pathologic valvar aortic stenosis (AS), membranous subvalvar aortic stenosis (subAS), and hypertrophic cardiomyopathy (HCM). Importantly, the murmur of HCM tends to be harsher, and becomes louder with standing, while that of AS is also harsher and is usually associated with a systolic click. Membranous subAS also tends to be harsher than an innocent aortic flow murmur, but may have no other distinguishing characteristics.

6. Innocent supraclavicular bruit: This is a murmur that can sound frightful at times, and is caused by turbulence at the origins of the brachiocephalic arteries at the base of the neck. When noted in an adult, it is similar to the sound of a carotid bruit, and can also sound similar to the radiation coming from valvar AS, subAS, or HCM. This murmur is louder when sitting and can be extinguished by simultaneously flexing the elbows then extending the arms backwards.

7. And more! There are a number of other innocent murmurs that may be noted from time to time, such as the mammary soufflé, the cardiorespiratory murmur, and the "nonspecific innocent murmur."

C. Is further testing needed for the evaluation of cardiac murmurs?

Here is the answer: **"It depends!"**

I realize that this is not a very satisfying response, but here are some examples which may provide you with some additional insight:

If a murmur clearly meets the criteria for an innocent venous hum (the six general characteristics of an innocent murmur and the specific findings of the venous hum), then no further testing is needed. There is no pathologic murmur that can reproduce the venous hum.

Likewise, if the examination satisfies the criteria for a Still's murmur, especially in a child 2 years or older when physiologic splitting of the first heart sound should be readily evident, then no further testing is needed.

However, if a patient has features of a Still's murmur, and had a sibling that died of "SIDS," then further evaluation would be indicated.

Likewise, if a patient has a murmur that sounds like innocent PPS at 4 weeks of age, and happens to have an EKG that has an upright T wave in lead V1 (a finding of RV hypertrophy), or has a PPS murmur at 6 months, then additional evaluation would be required to rule out pathologic PPS or another condition.

Likewise, if a patient is less than 14 days old, the practitioner must be VERY CAREFUL that the murmur is not associated with a ductal dependent lesion. For example, a patient with hypoplastic left heart syndrome in the setting of a large ductus arteriosus and elevated pulmonary vascular resistance may demonstrate only a subtle "innocent pulmonary flow murmur," and might be sent home only to present later in cardiogenic shock.

Likewise, a child with a murmur who is at high risk for structural heart disease should undergo further evaluation. Examples include patients with syndromes such as Down, Turner, Williams, and Noonan syndrome. Additionally, those with multiple congenital anomalies, a family history of HCM, and other conditions may deserve further investigation.

Likewise, a patient with a seemingly "innocent murmur" and failure to thrive or chronic respiratory symptoms deserves a further look. Note that technically, these patients don't meet the criteria for an innocent murmur because they have symptoms that potentially have a cardiac origin.

So, putting it all together, "It depends!" A final additional good rule of thumb to consider is, "The younger the patient, the more expeditious should be the evaluation."

D. What is the approach to further evaluate a murmur that may not be innocent?

Should testing first be done, then a cardiology visit if needed, or should the patient first see the cardiologist? If testing is done, which studies are most helpful?

If I had to choose one test that the primary physician can use that is quick and relatively cost effective, it would be the 12 lead EKG. Although certainly this study can't identify many cardiac anomalies, the combination of a normal EKG, good femoral pulses, and a normal lower extremity saturation in the absence of cardiac symptoms is fairly reassuring that the patient doesn't have a condition that is immediately life threatening. It's imperative to note that a normal EKG in a newborn or 2 week old infant is very different from that of an adult, with changes in voltages and T-waves being particularly important.

The chest X-ray is helpful if it is abnormal, but there are certainly many cardiac conditions, even severe ones, that are seen in the setting of a normal CXR. Additionally, cardiomegaly may be difficult to determine in an infant. For these reasons, I find the CXR less helpful than the EKG.

The echocardiogram is of tremendous value, however, in my opinion, for most situations, this test should be performed after clinical cardiac evaluation for these reasons:

1. Some lesions may be missed by echo unless the person performing the scan knows to look for them specifically. A cardiologist's history and physical examination prior to the scan will significantly diminish the frequency of these "misses."
2. Often, the cardiologist will feel comfortable diagnosing an innocent murmur based on history, physical examination, and 12 lead EKG alone, and will forgo the echocardiogram. This cost benefit was detailed in the 1993 study by Danford.
3. Finally, in situations where a pathologic murmur is confirmed, an explanation of the lesion, anticipated clinical course, and need for intervention can be provided immediately in our office. This tends to lessen the family's anxiety considerably.

F. Summary

Innocent cardiac murmurs are frequently noted, with prevalence rates higher than 50% during the childhood years. Many can be identified by clinical evaluation alone without the need for expensive testing. One other factor important in the evaluation of murmurs is YOU. Your expertise and experience are invaluable. Granted, some of you are more experienced with cardiac auscultation than others. Murmur identification is a learned skill. Many practitioners, by making a concerted effort to identify specific types of innocent murmurs can become very accurate in their assessment. The venous hum and Still's murmurs are both common and peculiar enough that they can be confidently identified in your office. Beginning with those murmurs is a great way to build your confidence. Happy examination!

Michael Liske, MD, FAAC, FASE

Rosenthal A, Ped Clin N Amer, 31: 1229-1240, 1984
Danford A, Nasir A, Gumbiner C, Pediatrics, 91:365-368, 1993

E. Counseling for the family of a child with an innocent murmur

If an innocent murmur is diagnosed, then the following points should be emphasized:

1. Your child's heart is normal.
2. This sound is due to normal blood flowing through a normal heart causing innocent vibration.
3. Your child's heart is normal.
4. This murmur may become louder with fevers, and that is OK.
5. Your child's heart is normal.
6. This murmur will probably get softer as your child ages, but may not completely resolve, and that is OK. Some adults have innocent murmurs.
7. Your child's heart is normal.
8. Antibiotics are not needed prior to dental visits, but it is still good for your child to brush her teeth.
9. Your child's heart is normal.
10. There is no need for exercise or sports restrictions.



**East Tennessee
Pediatric Cardiology, PC**

2001 Highland Avenue
Suite B
Knoxville, TN 37916
Phone: 865.971.6897
Fax: 865.971.6817



We're on the Web!
www.etpc-hearts.com

About East Tennessee Pediatric Cardiology

Some of you have asked about our practice and how to most easily access the services we provide.

We are located 2 blocks north of East Tennessee Children's Hospital (not on the Children's campus itself.) We are at the corner of Highland Ave and 20th Street. Please see our web-site or Facebook page for phone contact, address, and map: www.etpc-hearts.com.

For patients needing testing, we offer EKG, echocardiography, and other diagnostic capabilities on site. Ours is the only pediatric and fetal echo lab in the region that is certified by the Intersocietal Commission on the

Accreditation of Echocardiogram Laboratories (ICAEL), and our doctors perform the scanning themselves in the office.

We maintain inpatient privileges at East Tennessee Children's Hospital, University of Tennessee Medical Center, St. Mary's Medical Center, and Ft. Sanders Regional Medical Center. If your patient is immediately ill and needs transportation to one of these centers, we'd love to hear from you as he is en-route to further facilitate his care. Our direct physician phone line is 865-771-9070.



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