The Innocence of Innocent Heart Murmurs

Family physicians and parents alike need clarification on what innocent heart murmurs are and how they can be diagnosed.

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Innocent murmurs in childhood remain a significant source of morbidity and cost to the healthcare system. Of 600 new patients assessed in our Pediatric Cardiology Outpatient Clinic last year, over 25% (almost 200) new patients were seen because of innocent murmurs.

Parental understanding is important

We attempted to assess parental understanding of innocent murmurs, both prior to and following cardiology referral. Parents were given a questionnaire regarding their knowledge and attitude toward their child’s murmur before seeing one of four cardiologists.

Prior to cardiology referral, 3% of parents felt their child could potentially have a serious heart problem, 19% felt their child had a non-serious heart problem, 76% felt their child had a normal heart, and 2% admitted to being confused.

Following cardiology consultation, and despite one-half of the patients receiving an information pamphlet, the results shifted in an unexpected manner. Fortunately, none of the parents left with the impression that their child had a serious heart problem. However, those with the perception that their child had a non-serious problem actually increased to 30%, and the number who left with the impression that their child had a normal heart fell to 68%. The number of parents who were confused increased from 2% to 3%.

These results may be an interesting and perhaps amusing commentary on the effectiveness of physician consultation and information pamphlets. But the results also point to the significant morbidity associated with the diagnosis of innocent heart murmur.

At present, the cost of a full cardiac ultrasound exceeds the cost of specialist consultation, and the combined cost of referrals and ultrasounds to the health-care system for thousands of patients each year is truly significant.

Clearly it would be of great benefit if we could develop a system to dispense with the majority of innocent heart murmurs in childhood with a minimum of referral and diagnostic testing.
**Frequently Asked Questions:**

**Innocent Heart Murmurs**

<table>
<thead>
<tr>
<th>Questions:</th>
<th>Answers:</th>
</tr>
</thead>
<tbody>
<tr>
<td>What are the five common innocent heart murmurs?</td>
<td>There are two types of innocent heart murmurs: systolic and continuous. There are four systolic murmurs and one continuous. <strong>Systolic:</strong> 1. Still's murmur 2. Peripheral pulmonary stenosis of the newborn 3. Pulmonary systolic flow murmur 4. Supraclavicular systolic bruit <strong>Continuous:</strong> 1. Continuous venous hum</td>
</tr>
<tr>
<td>What steps should be taken when diagnosing an innocent heart murmur?</td>
<td>• It is important to confirm the presence of a quiet precordium, as no murmur associated with a thrill can be seen as innocent. • Confirm the normal heart sounds. • Verifying the absence of extra sounds, especially clicks at the apex, must always be considered important because of the relative high prevalence of bicuspid aortic valve and mitral valve prolapse. • Timing the murmur, no diastolic murmur is innocent. • Positively identify the specific innocent murmur.</td>
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</tbody>
</table>

**Basic approach**

Auscultation is a learned skill and there is no substitute for experience. Fortunately this experience can now be gained online through the Internet and through several widely available CD-ROMs.\(^1^,\(^2\) When asked, many practitioners will define an innocent murmur negatively, *i.e.*, as a murmur present in the absence of pathology. This in turn, naturally leads to a search for pathology, just to be sure. In fact, the diagnosis of an innocent murmur
should be a positive diagnosis, and patients should leave with a specific diagnosis and description of what the murmur is.

This, perhaps dogmatic, approach means that once identified, most of these murmurs do not require other diagnostic testing, such as electrocardiogram (EKG), chest X-ray, or ultrasound.

**Where to begin?**

It is important to confirm the presence of a quiet precordium as no murmur associated with a thrill can be taken as innocent. Secondly, confirmation of the normal heart sounds, including normal splitting of the second heart sound, is also important. The presence of extra sounds, especially clicks at the apex, must always be considered important because of the relative high prevalence of bicuspid aortic valve and mitral valve prolapse. Timing the murmur is important, as no diastolic murmur is innocent.

Given the above normal findings, what follows is a brief description of the innocent murmurs of childhood, which can be readily diagnosed by auscultation. In childhood, there are five common innocent heart murmurs (four which are systolic and one which is continuous).

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**What are the innocent childhood murmurs?**

1. **Still’s murmur**

Still’s murmur is felt to be related to vibrations caused by the effect of ejection on trabeculae/tendons in the left ventricle.\(^4\) The peak incidence is at three to five years of age with a range from infancy to adolescence. The murmur has a unique relationship to puberty, and always disappears at that time.

The murmur is best described as a Grade I-II/VI, vibratory, ejection systolic murmur. There are two important keys to the diagnosis. One is the unique sound of the murmur, which can be learned from practice, and is likely best described as twanging. Therefore, it is often best heard with a light application of the bell. Secondly the murmur is best localized inside the apex along the sternal border, *i.e.*, it is not maximal at the apex.

The murmur does increase with increased cardiac output, and so is best heard with the patient lying or at times of high cardiac output, such as with fever. There is no significant differential diagnosis.

2. **Peripheral pulmonary stenosis of the newborn**

This is an extremely common murmur if carefully listened for in newborns. The murmur is caused by the relatively large size of the main pulmonary artery compared to branch pulmonary arteries in the newborn. Due to the large liver and supine position of the newborn the heart is positioned higher in the chest. This position of the heart leads to some kinking, particularly at the left, but also the right branch pulmonary artery. In most cases by catheterization or echo-Doppler, a small but true gradient can be measured at the bifurcation of the pulmonary arteries. The fact that the murmur is even more common in the premature infant is further evidence of its relationship to cardiac positioning. The cause of the murmur will disappear when the infant assumes an upright position.

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**Take-home message**

- With some practice innocent heart murmurs can be readily identified and we would, of course, promote the acquisition of this skill by most generalists.
- The fact that the murmurs are innocent and associated with a completely normal circulation should be made very clear to parents.
- An actual description of the nature and causes of these benign noises may reassure the parents.
- It is those murmurs which are not clearly identified as being typical innocent murmurs that are the ones which suggest the need for further investigation.

“The diagnosis of an innocent murmur should be a positive one, and patients should leave with a specific diagnosis and description of what it is.”
upright posture and is, therefore, rarely heard after a few weeks to one year of age.

The murmur is described as Grade I-II/VI, high-pitched and impure ejection systolic murmurs. The keys to diagnosis are that the murmur is best heard with the diaphragm, and is often very difficult to localize with radiation to the axilla and back. In the rapidly breathing newborn, the murmur is most often confused with respirations, but will persist when there is a brief pause in respiration.

The differential diagnosis would include true peripheral pulmonary stenosis. True peripheral pulmonary stenosis is unusual, and very often associated with several well known syndromes including Turner’s syndrome, Williams syndrome, Noonan’s syndrome, rubella syndrome, and Alagille’s syndrome. These syndromes are not described here in detail, but will be familiar to the pediatrician and generalist. Thus, true peripheral pulmonary stenosis is extremely rare in the absence of dysmorphism, and can usually be dismissed.

3. The pulmonary systolic flow murmur

The pulmonary systolic flow murmur is related to ejection vibrations in the main pulmonary artery. The murmur is also related to the very anterior position of the main pulmonary artery, and in this regard one should be cautious of the so-called “straight back syndrome,” which today may justify an echocardiogram. The pulmonary systolic flow murmur is also greatly affected by cardiac output and may be heard in settings, such as anemia or hyperthyroidism. Such murmurs are most frequently heard from childhood through early adulthood, but are unusual in the fourth decade.

The murmur is a Grade II-III/VI, high-pitched impure ejection systolic murmur. It is
best heard with the diaphragm and well localized to the pulmonary area.

The differential diagnosis would include mild pulmonary valve stenosis. The natural history study demonstrates that valvar pulmonary stenosis is a highly predictable disease. Mild pulmonary valve stenosis is uniformly associated with an ejection click, also best heard in the pulmonary area. The absence of an ejection click allows the differentiation of the benign pulmonary systolic flow murmur from pulmonary valve stenosis.

4. Supraclavicular systolic bruit

The supraclavicular systolic murmur may be related to the subject of innocent cranial bruits in children. In early childhood the relative cardiac output to the head is increased, and may reach 25% compared to the adults of approximately 10%. This high output to the head and neck may lead to systolic vibrations along the path of the subclavian and carotid arteries.

These I-II/VI ejection bruits are always best heard above the clavicles or in the suprasternal notch. In the young infant the bruit can often be traced along the branches of the internal or external carotid arteries. This ability to trace benign cranial bruits will often differentiate them from pathologic cranial bruits related to arterio-venous malformations. The most frequent arterio-venous malformation is the vein of Galen aneurysm, which is often best heard at the occiput or over the anterior fontanelle.

The supraclavicular systolic bruit in children is not to be confused with carotid bruits in adults, which are not considered benign.

5. Continuous venous hum

While still unclear, the continuous venous hum may also be related to the relatively high cardiac output to the head and neck. The high flow venous return to the great veins of the head and neck coupled with compression or deviation by the clavicle at the thoracic inlet likely leads to this murmur. It is most frequently heard from ages one to seven with its peak at three to five years of age.

The murmur is described as a Grade I-III/VI, low-pitched and impure murmur. The murmur is best heard just above or below either clavicle. It is often described as having a whining or musical quality, and is best heard with the bell. The key to diagnosis is, of course, its relation to positioning. The venous hum can be elicited in many children by lifting the chin. The murmur will diminish or disappear with com-
pression of the internal jugular vein or with the supine position.

The differential diagnosis is the patent ductus arteriosus (PDA). This congenital heart malformation also causes a continuous murmur heard below the left clavicle. This murmur is, however, consistent with an arteriovenous malformation, and, while also continuous, it is generally described as harsh, impure, and machinery-like. The PDA will not vary with positioning.

**Murmurs in post-adolescence**

The mammary souffle is a continuous or systolic murmur which is not heard in childhood, but is most frequently heard during late pregnancy or early post-partum. The murmur is caused by increased arterial flow to the breast related to lactation. The murmur is often continuous, and is not related either to changes in cardiac output or positioning.

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