Current Practice

DISEASES OF THE HEART AND BLOOD-VESSELS

Innocent Systolic Murmurs in Childhood

E. GOLDBLATT,* B.SC., M.R.C.P.ED., D.C.H.

On careful auscultation over 50% of apparently perfectly healthy, normal children will be found to have a systolic murmur. The incidence of congenital heart disease is approximately six cases per thousand live births. Of these many die in early infancy and by the time the child reaches school age only about 2.5 per thousand have definite organic disease. It is apparent, therefore, that the vast majority of children with systolic murmurs have normal hearts. It is not always easy to distinguish the innocent systolic murmur from the murmur due to an organic lesion, particularly if it is a mild one. Murmurs are often heard on routine medical examination in the neonatal period, or later in infancy at maternity and child welfare clinics. Sometimes a murmur is heard on examination during an intercurrent illness. In other children the murmur may be heard for the first time on routine school medical examination.

The public are becoming increasingly aware of the significance of the words "heart murmur." By telling parents that a murmur has been heard in a child, tremendous anxiety may be aroused until the diagnosis has been clarified. It is therefore extremely important to be familiar with the characteristics of the innocent systolic murmur, so that parents may be confidently reassured if they have been told about the presence of the murmur. Unnecessary worry can be prevented by not disclosing the fact that a murmur has been heard, provided the doctor is certain that it is insignificant.

Incidence of Innocent Murmurs

At the Royal Liverpool Children's Hospital approximately a third of the 4,500 patients seen over the past 17 years have been considered to have normal hearts. This is a very conservative estimate of the incidence of insignificant systolic murmurs, as these children have, in the main, been referred by consultant paediatricians who have been in some doubt about the innocence of the murmur. In all probability a large number of children with typical "functional" systolic murmurs are not referred, as the paediatricians have had no doubt about the innocence of the murmurs.

Diagnosis of Systolic Murmurs

Children with cyanotic heart lesions or definite evidence of organic disease are not included in this discussion: these patients need further assessment and investigation and invariably should be referred to a centre equipped to investigate and treat such lesions.

Constantly improving techniques of surgical correction of congenital and acquired heart disease make accurate diagnosis essential. In childhood an accurate diagnosis can be made by

clinical examination alone in many patients with systolic murmurs. In some, however, further specialized investigations may be necessary to establish the diagnosis, though these techniques are usually restricted to patients with organic disease, and are carried out to confirm the clinical assessment and to obtain detailed haemodynamic data.

In those cases in which it is difficult to be certain whether the systolic murmur is insignificant or is due to a mild organic lesion further opinion should be sought. The vast majority of these children are symptom-free and the unsuspecting parent is usually shocked to learn that the child has a murmur. The parents will be perturbed and upset at the thought of a child having a cardiac abnormality. Though the murmur may be insignificant, referral to cardiac clinics may cause unnecessary parental anxiety. Therefore, when referring patients, the parents should be encouraged with cautious optimism, as this may help to alleviate unnecessary anxiety.

Systolic Murmurs in Infants

Systolic murmurs are often heard in newborn and young infants. During the neonatal period there may be a murmur due to the closing ductus arteriosus. A fairly loud, harsh, blowing systolic murmur may be heard, particularly down the left sternal border, in the slightly older infant. The assessment of its significance may be difficult, but the following points may be of assistance in diagnosis. The murmur is likely to be innocent if the infant is symptom-free and progressing normally, the systolic murmur is the only abnormality, the heart sounds are normal and there are no diastolic murmurs, the electrocardiogram is normal, and radiological examination of the cardiac silhouette is normal. If the murmur is innocent it becomes softer at subsequent follow-up visits and disappears entirely within a few months or by the age of two years in most cases.

In paediatric cardiac clinics this group of symptom-free infants is usually not subjected to further specialized cardiac investigation. The children are reviewed at regular intervals.

In a number of cases the murmur may have been due to a small ventricular septal defect, which has subsequently closed spontaneously. The diagnosis is never proved beyond doubt unless the infant is subjected to cardiac catheterization when first seen. It may be misleading to make a diagnosis in the infant based on the localization of maximal intensity of the systolic murmur. A common error occurs, for example, in differentiating between a ventricular septal defect and pulmonary stenosis. Only with growth, associated with better localization of the site of maximal intensity of the murmur, will the diagnosis become clear.

* Consultant Paediatrician, Royal Liverpool Children's Hospital; Part-time Lecturer, Department of Child Health, University of Liverpool.

Innocent Systolic Murmurs in Older Children

There are certain characteristics of innocent systolic murmurs in older children which allow the diagnosis to be made with confidence.

The children are entirely symptom-free. The murmur is the only abnormality detected. These murmurs are not accompanied by systolic thrills: they occur at the beginning of systole, immediately after the first heart sound, and they are usually short—in most cases they do not occupy more than half of systole—and of low frequency. They have a buzzing or coarse vibratory quality, and are usually best heard at the left border of the sternum, internal to the apex, and are frequently limited to a localized area. They may, however, also be heard in the pulmonary area.

These murmurs often change in intensity with alteration in posture and can sometimes be almost completely obliterated by change of position, such as from the standing to the supine position. The heart sounds and respiratory variation of the second sound are normal, the electrocardiogram is normal, and the cardiac silhouette on x-ray examination is normal.

Short insignificant murmurs may be heard when there is tachycardia associated with fever, excitement, anaemia, or infection. Such murmurs may not be heard when the heart rate has returned to normal.

Venous Hum

The venous hum is commonly heard and is of no clinical significance. It is a continuous murmur, with diastolic accentuation, heard below either the right or left clavicle. It often extends into the neck. The murmur is thought to be due to blood flow in the great veins or to turbulence produced at the junctions of the large veins. The murmur may be obliterated or diminished by light pressure over the jugular vein. Rotation of the head or change in position and flexion of the head will usually obliterate the murmur entirely.

The heart is usually normal in all other respects, but it should be pointed out that the venous hum may also be heard in children who have murmurs due to organic heart disease.

The importance of the venous hum is that it may be mistaken for a patent ductus arteriosus, but the murmur in this condition cannot be obliterated by head rotation, flexion, or alteration in posture.

Organic Causes of Systolic Murmurs

Where the innocent murmur is fairly loud, it must be differentiated from one of the organic lesions which are characterized by systolic murmurs. Unless the lesion is severe or there are some complications the majority of children with these lesions will also be symptom-free. They are:

- (1) Ventricular septal defect.
- (2) Pulmonary stenosis.
- (3) Aortic stenosis.
- (4) Mitral incompetence.
- (5) Atrial septal defect.
- (6) Coarctation of the aorta.

Each of these murmurs may be differentiated from the insignificant murmur of childhood by careful clinical examination.

Ventricular Septal Defect

The cardiac impulse may be normal or there may be evidence of left or right ventricular dominance. A systolic thrill may be palpable. The systolic murmur is loud, harsh, rough, or

blowing in character, and it occupies the whole of systole. It is loudest at the left sternal border and at the apex, but it may also be heard to the right of the sternum. Sometimes it is loudest in the second and third left intercostal spaces, particularly in infants, when it may be difficult to distinguish from the murmur of pulmonary stenosis. The heart sounds are normal. In patients with larger defects a diastolic murmur, due to increased flow through the mitral valve, may be heard at the apex.

Pulmonary Stenosis

There may be a right ventricular heave. A systolic thrill may be palpable in the pulmonary area. There is a rough ejection-type systolic murmur which is loudest in the pulmonary area. The murmur may be heard down the left sternal border and over the chest posteriorly. In mild cases both components of the second sound will be heard, the pulmonary component being soft. The interval between the aortic and pulmonary components is wider than normal and does not vary with respiration. In the more severe degrees of pulmonary stenosis the pulmonary component of the second sound may not be audible and only the single sound of aortic valve closure may be heard.

In mild cases an early clicking sound following the first sound may be heard in the pulmonary area or at the left sternal border. This is referred to as an ejection click.

Aortic Stenosis

In the more severe cases of aortic stenosis there will be a left ventricular thrust due to left ventricular hypertrophy. A systolic thrill will usually be palpable over the carotid arteries and in the suprasternal notch. In the more severe cases it is also palpable over the aortic area. A rough or harsh ejection systolic murmur is heard over the praecordium. It is loudest over the aortic area and at the apex, but may be well heard along the left sternal border. The murmur radiates up into the neck as a rough ejection systolic murmur of variable length over both carotids.

In mild cases the second sound will be of normal intensity and will vary normally with respiration. In severe cases, due to delay or aortic valve closure, splitting may be close; the split will apparently become wider on expiration. This is referred to as paradoxical splitting of the second sound. In valvular aortic stenosis (as opposed to subvalvular or supra-valvular stenosis) an ejection click may be heard at the apex or along the left sternal border. This is a similar sound to that referred to under pulmonary stenosis.

Mitral Incompetence

The impulse at the apex may be normal or characteristic of left ventricular hypertrophy. A systolic thrill may be palpable at the apex. The systolic murmur is best heard at the apex and is transmitted to the axilla. It is blowing in quality, beginning at the first sound and extending throughout systole to end at the second sound. The murmur may vary through all grades of intensity.

The first sound may be soft. The second sound is of normal intensity and varies normally with respiration if the condition is not complicated by pulmonary hypertension. A well-marked third sound may be audible at the apex.

Atrial Septal Defect

The impulse may be normal, or suggestive of right ventricular enlargement by a definite parasternal heave. Thrills are usually not palpable unless there is accompanying

pulmonary stenosis. The systolic murmur may be unimpressive. It may be blowing or harsh. It may be short, and best heard at the left sternal border, or, more commonly, there is a louder, rough, harsh ejection systolic murmur best heard over the pulmonary area.

In defects with a large left-to-right shunt a diastolic murmur, which becomes louder on inspiration, due to increased flow through the tricuspid valve, will be heard at the lower left sternal border or over the lower sternum.

The second sound is widely split and of normal intensity. It does not vary with respiration. An early sound following the first sound is frequently heard. This may be due to splitting of the first sound or to an ejection click.

It may be difficult to distinguish clinically between a small atrial septal defect and mild pulmonary stenosis.

Coarctation of the Aorta

The impulse may be normal or left ventricular in character. A systolic thrill may be palpable in the suprasternal notch and carotids. A late systolic murmur, which may not be impressive, is heard at the apex and left sternal border. It may be transmitted into the neck, particularly over the left carotid artery. The murmur may be transmitted to the back, especially over the scapulae.

Sometimes the murmur extends into diastole, particularly below the left clavicle towards the shoulder. A continuous murmur may be audible over the internal mammary arteries and at the medial border of the scapulae. In most cases the heart sounds are normal.

The important diagnostic feature is the inability to palpate the lower limb pulses. Sometimes the femoral pulses are just palpable, but are of small volume and there is delay in timing between the femoral and radial pulses.

Minor Abnormalities

Some "innocent murmurs" may be due to minor abnormalities, such as kinking or mild coarctation in the arch of the aorta. It may be possible to demonstrate these conditions by special radiographic techniques.

As a general rule an organic lesion must be suspected in the presence of cardiac enlargement, palpable thrills, loud systolic murmurs, diastolic murmurs, and abnormalities in the intensity and in the respiratory excursion of the second sound. Difficulty may be experienced in differentiating between innocent murmurs and those produced by mild organic lesions. In the latter group the heart will be of normal size, thrills will usually not be palpable, and the heart sounds will be normal. Electrocardiographic and radiological studies will also be normal.

The diagnosis may not be clear without repeated clinical examinations over a period of years unless the child is subjected to more detailed cardiac investigation at the outset to establish the diagnosis with certainty. There is much to be said for this approach where waiting lists for cardiac investigation permit, as the parent and child can then be reassured with confidence, and any anxiety and feeling of uncertainty allayed.

General Management

When the medical practitioner is certain that a murmur is innocent there is good reason not to mention to the parents that it has been heard. The presence of the murmur, however, should be documented. This may prevent the error of considering the murmur to have been acquired if it is heard at a later date, particularly after an intercurrent illness which may be associated with cardiac complications. When doubt exists about

the significance of the murmur, and a further opinion is sought, the parents should be told the reason for seeking another opinion. At the same time they should be reassured that in all probability if an organic lesion is present it is a mild one.

In the case of the symptom-free child it is essential to stress that he should continue leading a perfectly normal unrestricted life until he is seen at the cardiac clinic. Unless the parents are fully reassured they tend to look for symptoms. At cardiac clinics cardiologists are often told that the child with an innocent murmur tires more easily than other children or is breathless on exertion. After detailed questioning it transpires that the symptoms have been noticed only since the parent was made aware of the presence of the murmur.

The vast majority of children with acyanotic heart disease are symptom-free and current practice is to emphasize to the parents that it is essential for the child to continue leading a normal unrestricted life. Even when the lesion is more serious and the child is disabled, it is important to allow him to lead as normal a life as possible within his own exercise tolerance.

If the murmur is functional any doubt about its significance must be eliminated from the mind of the parent. The best way of achieving this is by dispensing with further follow-up.

Schooling

Children with functional murmurs must continue leading normal lives at ordinary schools. The child in whom the systolic murmur is due to a mild lesion should also attend an ordinary school and take part in the full activities of school life.

Even in those children in whom the systolic murmur is due to a more serious lesion, it is desirable that education is pursued at an ordinary school. Where there is some disability a trial at an ordinary school should be made, as in most cases the child will be able to manage quite well. In those cases where there is severe disability there may be a case for education at a school which caters for the physically handicapped if the child has not been able to cope at an ordinary school. It is only under exceptional circumstances that it is necessary to keep a child with severe heart disease out of some sort of school to be educated alone and part-time by a home tutor.

Subacute Bacterial Endocarditis

Subacute bacterial endocarditis is a very serious complication of organic heart disease, no matter how mild the lesion. Once contracted the treatment, even if successful, may be prolonged and most unpleasant.

This condition may complicate any organic heart lesion. The incidence can be reduced by giving an antibiotic cover for all dental treatment. This precaution should be taken in children with systolic murmurs where there is any doubt about their significance. Subacute bacterial endocarditis is to a large extent a preventable disease, and parents should be advised to inform the dental surgeon about the presence of the murmur and the advisability of taking the necessary precautions before proceeding with dental treatment.

Similarly, other infections should be dealt with promptly and adequately.

Advice to Parents

When one is certain that the murmur is innocent the parents must be convincingly reassured that the child's heart is normal. If there is any doubt at all one should inform the parents that in all probability the heart is perfectly normal, but that out of interest one would like to see the child again in a few years

to ensure that the murmur has disappeared. The parents can be told about the high incidence of innocent murmurs in childhood. It should be explained that the reasons for these murmurs are not clear, but that it is known that children with this type of murmur have normally functioning hearts.

If a mild organic lesion is suspected the parents must be reassured that the child will come to no harm and suffer from no disability. If it is felt that further investigation may be necessary at a later date the parents can be informed about this, but told that in all probability surgery will not be necessary. Even if surgery is going to be necessary, the parents must be told confidently that the child will come to no harm by postponing investigation and surgery until the child reaches a suitable age.

When the lesion is more serious the diagnosis should be explained, with the aid of diagrams, in terms that can be

understood by the parents. The proposed timing of the investigations and possible surgery can be outlined. As far as possible the ultimate prognosis should also be discussed with the parents.

Unnecessary restriction of children with systolic murmurs may cause irreparable psychological damage. It may be difficult to get children to readjust themselves to a normal life after treatment if they have been unnecessarily restricted and over-protected pre-operatively. Bearing these simple principles in mind will alleviate unnecessary parental anxiety and contribute largely towards preventing the disabling syndrome of "cardiac neurosis."

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ANY QUESTIONS?

We publish below a selection of questions and answers of general interest.

Propranolol and Heart Disease

Q.—What is the place of β -adrenergic blocking agents in the treatment of heart disease? Which cases might benefit and which cases be adversely affected? What precautions should be taken when using these agents in the treatment of angina?

A.—Propranolol (Inderal) is the currently available β -adrenergic blocking agent, and aspects of its clinical use have recently been summarized.¹ It is now being advertised for the treatment of angina pectoris, cardiac arrhythmias, particularly those associated with digitalis intoxication and anaesthesia, and, in conjunction with α -adrenergic blockade, for the treatment of phaeochromocytoma.

In the acute treatment of arrhythmias it is probably wisest to start with a very small dose of propranolol intravenously, say 0.5–1 mg., to repeat it after two to three minutes and to continue likewise as required until a total of 10 mg. has been given, all under E.C.G. control. Propranolol buys time in digitalis intoxication while excess is excreted. In patients with a severe cardiac lesion particularly, special care must be taken to avoid causing acute heart failure and hypotension. Propranolol may also be of value in paroxysmal atrial and ventricular tachycardia, and in helping to maintain normal rhythm after electric shock in ventricular fibrillation.

Before the advent of propranolol no drug had convincingly been shown to reduce the frequency of anginal attacks throughout the day. It therefore represents a useful advance, taking its place alongside reduction in weight (where indicated) and trinitrin in the treatment of angina. One double blind trial has reached high levels of significance in favour of the drug ($P=0.00012$).² There is no good evidence that it is less effective in one type of anginal patient than in another.

The dosage of propranolol is important. Present evidence suggests that it is reasonable to adjust the dose so that the resting pulse rate is about 60 beats a minute. This is sometimes achieved, particularly in elderly patients with ischaemic heart disease and occasionally in younger patients, by giving 5 to 10 mg. four times a day orally. The

initial dosage should not be more than 10 mg. four times a day. However, other patients require far more. Each dose should be increased by, say, 10 mg. as indicated, and when a dosage of 60 mg. four times a day is reached increments of 20 mg. per dose are reasonable. The lowest dose in the trial referred to³ was 30 mg. four times a day. Six patients (out of a total of 14) received 100 mg. four times a day. The average might be 60–80 mg. four times a day, but it must again be emphasized that this dose is arrived at by steady increments from 10 mg. four times a day. Propranolol is valuable in angina, but for optimum effect the dose must be adjusted to suit the individual.

Propranolol should not be given to asthmatics, since it increases obstruction of the airway in such patients. Propranolol may precipitate heart failure in susceptible subjects, and this may occur with a small dose. When there is a history of left ventricular insufficiency and shortness of breath on exertion digitalis and a diuretic should be given before prescribing propranolol. If a patient is already receiving digitalis and diuretics for heart failure, propranolol should not be used unless the indication is strong and the patient is well compensated with therapy. Even then propranolol should be given only with extra caution. As an example, a 71-year-old woman with ischaemia developed heart failure after being given propranolol 10 mg. for two days. After she had been given digitalis and chlorothiazide she tolerated propranolol 50 mg. four times a day.⁴ Recent myocardial infarction as such is not a contraindication to propranolol. Other side-effects are not troublesome.

In conclusion, propranolol is valuable in angina pectoris and is of use in certain arrhythmias and phaeochromocytoma. There is some suggestion that it may be of use in numerous other conditions such as hypertension, myocardial infarction, anxiety state, hypertrophic obstructive cardiomyopathy, and Parkinsonian tremor. Further evidence is awaited with interest.

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Insulin by Jet Injection

Q.—Is high-pressure injection with an injector of the Porton type suitable for giving insulin, particularly in children?

A.—Jet injection has the obvious advantage over syringe and needle that it is less frightening and should therefore be useful for children.

Machines of the Porton type,¹ once assembled, do not require sterilizing, and they are relatively easy to load and prime. The injector must be correctly positioned at right angles to the skin to avoid bruising and bleeding. Tests with the Hypospray injector have been carried out in a limited number of diabetic children,^{2,4} and once the technique is mastered it appears to be as reliable as conventional injections, and in general more acceptable.

There are, however, some possible snags. Dosage calibration may not be sufficiently accurate with the higher strength insulins and mixtures may present problems.³ The children studied were of necessity under supervision, and there is a need for greater experience with the method before it is recommended for use by an individual.

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Dupuytren's Contracture

Q.—What are the current views on the treatment of Dupuytren's contracture?

A.—Many patients with palpable thickening of the palmar fascia need no treatment at all. When finger movements are not affected, or are a little restricted and the condition is showing no sign of worsening, observation alone is sufficient. Passive stretching is almost certainly harmful and should be avoided.

When the contracture is progressive excision of the involved part of the palmar fascia is advised while the deformity is still only mild. Skilled and occasionally prolonged post-operative care is needed. Radical or total fasciectomy is often followed by exces-