Evaluation of ejection murmurs by pulsed Doppler echocardiography*

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SUMMARY A common problem is a soft basal ejection murmur in an asymptomatic child with a normal electrocardiogram and chest x-ray films. If the diagnosis is aortic stenosis, there is a need for prophylaxis for subacute bacterial endocarditis and concern about development of calcific aortic stenosis. In 40 consecutive children referred for this differential diagnosis, aortic stenosis was diagnosed in 30, based on an ejection murmur at the second right interspace (not necessarily louder than at the second left), which transmitted well to the neck, accompanied by a normal second heart sound. Pulsed Doppler echocardiography confirmed turbulence at the aortic valve in 26. In the 10 children diagnosed clinically as having an innocent murmur, three had evidence on pulsed Doppler echocardiography for an abnormal aortic valve. The conventional echocardiographic findings of an eccentric aortic orifice were present in only 21 of 29 patients with aortic turbulence. Though our current clinical criteria are reasonably specific (87%), they are not as sensitive (70%). Pulsed Doppler echocardiography provides powerful non-invasive assistance for this important differential diagnosis.

Clinically significant aortic stenosis is diagnosed by the presence of a grade 4/6, or louder, rough ejection murmur at the base of the heart, prominent at the second right intercostal space, well transmitted to the suprasternal notch and carotids, and towards the cardiac apex.¹ There may be an ejection click,² and audible aortic regurgitation.³ The additional presence of electrocardiographic (left ventricular hypertrophy) or radiological changes (left ventricular enlargement or post-stenotic dilatation of the ascending aorta), and the presence of a thrill are emphasised by others.⁴⁻⁶

A more common—and more difficult—problem is the asymptomatic child with a grade 2-3/6 ejection murmur at the base of the heart with a normal electrocardiogram and chest x-ray film. Since over a half of normal children have innocent murmurs, the majority of which are ejection murmurs, the differential diagnosis between aortic origin and pulmonary origin of this murmur is probably the most common cardiological problem that the family practitioner and paediatrician encounter in their practice. Whether this murmur represents an innocent ejection murmur, mild

pulmonary stenosis, or mild aortic stenosis will determine whether the patient is at risk for infective endocarditis and progression in later life to early calcific aortic stenosis.⁷ Though there are no known preventive measures for calcific aortic stenosis, infective endocarditis is a serious complication with high mortality and morbidity, and prophylaxis is thought to be effective in its prevention. Clearly, the cost and morbidity⁸ for invasive studies argue against their application to this huge group of youngsters with the differential diagnosis of innocent murmur versus mild aortic or pulmonary stenosis. The problem then is an absence of objective information to validate the conclusions based only on physical examination, a situation vulnerable to arbitrary definitions.

Non-invasive approaches of validating the diagnosis of the milder forms of aortic stenosis have not been specific enough. Phonocardiography is an established method of timing auscultatory findings, but it cannot establish the intravascular origin of the murmur. Conventional echocardiography greatly enhanced the detection of bicuspid aortic valves; eccentricity of the coapted aortic leaflets in diastole is well accepted,² and the absence of high frequency systolic flutter of the aortic leaflets has

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been suggested as diagnostic of abnormal aortic valves.⁹

Pulsed Doppler echocardiography is a relatively new technique capable of localising murmurs within the heart and blood vessels.¹⁰ In essence, pulsed Doppler echocardiography is a non-invasive intravascular phonocardiogram, combined with echocardiography. The combination permits the precise localisation of turbulence within the heart and great vessels. Obviously, specific inferences as to the cause of the turbulence require additional clinical information. In the present application, we are concerned with the origin of an ejection murmur, and whether there is abnormal systolic or diastolic turbulence at either semilunar valve. If there was no turbulence for either valve, and the valve orifice was concentric, the ejection murmur could logically be labelled innocent. If turbulence was found at, above, or below the aortic or pulmonary valve, that valve would be established as the site of origin of the murmur. Pulsed Doppler echocardiography was conducted without knowledge of the results of the physical examination. Our purpose was to validate the clinical conclusions based on conventional criteria of the physical diagnosis with the independent data of intracardiac turbulence by pulsed Doppler echocardiography.

Subjects and methods

Forty consecutive children ranging in age from 2 to 15 years with the clinical question of innocent murmur versus mild aortic stenosis were evaluated by physical examination, electrocardiogram, and chest x-ray film, by conventional M-mode echocardiography, and by pulsed Doppler echocardiography. The patients were examined by two cardiologists, and the consensus remained unknown to the echocardiographers until the data were tabulated.

A conventional M-mode echocardiogram was performed, examining carefully for valvular abnormalities such as redundant leaflet echoes, abnormal motion, asymmetry, and great vessel size, in addition to the usual echocardiographic indices.

At the conclusion of the conventional M-mode examinations pulsed Doppler echocardiography was performed to evaluate blood flow characteristics in heart chambers and great vessels, from standard praecordial and suprasternal approaches. The technique of pulsed Doppler echocardiography has been reviewed recently.¹⁰ The method combines M-mode echocardiography with a simultaneous measurement of Doppler-shift of the frequency of reflected sound; the Doppler signal is obtained at a controlled depth within the cardiac structures. The

sampling site is indicated by a straight line superimposed on the M-mode echocardiogram. The sample volume is relatively small, a tear-drop shaped volume 4 mm in length and 2 mm at its greatest diameter. By moving the sample volume throughout the heart and great vessels, one can sample blood flow characteristics at known sites (Fig. 1). One output from the Doppler instrument* is an audio signal; a smooth audio signal is produced

Fig. 1 Praecordial and suprasternal notch pulsed Doppler echocardiographic examination. (a) Praecordial approach. CW, chest wall; AO, ascending aorta; PA, main pulmonary artery; A, transducer position with sample volume (stippled tear drop) in AO; B, transducer position with sample volume in PA. (b) The suprasternal notch approach. At transducer position A, sample volume is in AO. At transducer position B, sample volume is in right pulmonary artery.

by smooth blood flow, and turbulent blood flow produces a coarse, rough audio signal. Another output of the pulsed Doppler echocardiographic unit is a time-interval histographic spectral record. This graphic method indicates turbulence by an increased spectral bandwidth, presenting as a wide scattering of the dots forming the envelope which outlines the magnitude and direction of blood flow. Fig 2 and 3 show examples of normal and abnormal blood flow records above the aortic valve with pulsed Doppler echocardiography from the suprasternal notch and praecordial approaches, respectively. Fig. 4 shows the normal flow record in the left ventricular outflow tract during systole and diastole.

The audio output is superior to the graphic method of analysis for detecting subtle degrees of abnormal turbulence, in a manner similar to a trained musical ear detecting the sound of an oboe

* Advanced Technology Laboratories, Bellevue, Washington, Model 400A or 600A, 5 or 3 mHz.

Fig. 2 Pulsed Doppler echocardiographic flow record from suprasternal notch examination. These two panels show compressed M-mode echocardiographic tracings at the top, and the time interval histogram at the bottom, as recorded from the suprasternal notch approach. AO, aorta; RPA, right pulmonary artery; SV, sample volume. Left, the walls of the aorta and right pulmonary artery are indicated by the small arrows, and share a common wall echo. The sample volume position is shown by the location of the line on the M-mode tracing. The graphic recording of the flow record is at the bottom of the panel. The small dots comprising the time-interval histographic record are grouped closely together in a wave form in systole (large arrows). This type of flow record is seen when smooth laminar flow is present. Right, from another patient the M-mode echocardiographic tracings are compressed at the top showing the aorta, right pulmonary artery, and the location of the sample volume in the aorta, and the graphic recording of the flow record is at the bottom of the panel. The dots of the time interval histogram are very scattered in systole, representing the rough systolic flow (large arrows). The dots are not scattered in diastole; the diastolic flow is not rough.

Fig. 3 Pulsed Doppler echocardiographic flow record from praecordiol examination. These two panels show compressed M-mode echocardiographic tracings at the top, and the time interval histogram at the bottom as recorded from the praecordial approach. AO, aorta; SV, sample volume; flow, time interval histographic flow record. (a) With SV in AO, shows a record of normal flow above the aortic valve. Systolic flow, above the zero reference line (flow) is comprised of closely grouped dots, indicating normal systolic flow. The diastolic interval is also normal. This record of normal systolic flow would correspond to a smooth audio output. (b) With SV in AO, shows the flow in a patient with aortic stenosis. The systolic portion of the flow record consists of widely scattered dots, indicative of rough systolic flow. This would correspond to a rough audio output.

in an orchestra, whereas spectral analysis might be overwhelmed by the entire orchestra. Though abnormal turbulence is not defined numerically for pulsed Doppler echocardiography, a standardised method has been used in our institution for the past two-and-a-half years on over 1000 children. In this study, objectivity was maintained by performing the examination without knowledge of the clinical impression. The presence or absence of turbulence was determined at, above, or below the aortic valve and at, above, or below the pulmonary valve or in the right pulmonary artery.

Fig. 4 Pulsed Doppler echocardiographic record of normal flow in the left ventricular outflow tract. M, mitral valve; SV, sample volume; flow, time interval histographic flow record. The SV has been placed in the left ventricular outflow tract, just anterior to the mitral valve leaflet (M). The flow record is comprised of closely grouped dots, and would correspond to a smooth audio output. As the mitral valve opens, flow proceeds towards the praecordial transducer, corresponding to valve motion. During systole, flow proceeds from the outflow tract toward the aorta, indicated by negative deflection of the flow record during systole. Throughout the cardiac cycle, there is close grouping of dots in the flow record. Such a record of normal flow in the left ventricle could be seen in a patient with normal aortic flow, or in a patient with aortic stenosis. In the normal patient, the aortic flow record would be similar to that shown in Fig. 3a. In a patient with valvar aortic stenosis, the normal left ventricular flow record would change to one similar to Fig. 3b, as SV is moved to aortic valve. The dot scatter, noted as SV is placed at and distal to the aortic valve is indicative of turbulent blood flow arising at the aortic valve.

Results

On the basis of our clinical findings, we diagnosed aortic stenosis in 30 of the 40 patients (Fig. 5). In all of these children, an ejection murmur was

Fig. 5 The patient distribution based upon clinical evaluation and the diagnoses based upon pulsed Doppler echocardiography evaluation are shown.

present at the base, prominent in the aortic area, though not necessarily louder than at the pulmonary area, and the murmur was judged to transmit well to the neck. An additional finding that was regarded as evidence of an aortic valve abnormality was a soft diastolic murmur presumed to be of aortic origin. An ejection click was frequently present, but not consistently. The second sound in the pulmonary area was judged to be normal in all.

Of the 30 patients diagnosed clinically as having mild aortic stenosis, 26 had one or more M-mode or Doppler criteria of an abnormal aortic valve. Eccentricity of the valve leaflets was diagnosed in 20. Turbulence was detected by pulsed Doppler echocardiography at the aortic valve in all 26, and turbulence in the ascending aorta was found in 24. Mild aortic regurgitation (diastolic turbulence below the aortic valve) was present in 12 subjects. Of the four subjects diagnosed clinically as aortic stenosis but without aortic abnormalities on pulsed Doppler echocardiographic examination, one had Doppler evidence of pulmonary regurgitation (the regurgitant murmur on physical examination had incorrectly been attributed to the aortic valve), one had systolic turbulence only in the main pulmonary artery, and two children had no detectable abnormalities by conventional echocardiogram or pulsed Doppler echocardiography.

In the 10 youngsters who were diagnosed clinically as having an innocent murmur, three had turbulence at the aortic valve and ascending aorta, and one of those had eccentricity of the aortic valve orifice (Fig. 5).

Discussion

Pulsed Doppler echocardiography is of particular benefit in those subjects with mild disease, for whom invasive studies are scarcely justified. Without some objective, independent analyses, clinical diagnoses are in danger of being unchallenged and somewhat arbitrary. Though we,^{11 12} and others¹³ have validated the results of pulsed Doppler echocardiography by comparison with catheterisation and angiocardiography, a logical case can be made for the superiority of a technique for detecting turbulence, the pulsed Doppler echocardiogram, over these invasive techniques, when the purpose of the study is to evaluate the anatomical origin of a murmur.¹⁴ The pulsed Doppler echocardiogram has the same usefulness—and limitations—as intracardiac phonocardiography, with the major advantage that cardiac catheterisation is not required. Study of the left heart requires arterial cannulation, and the possibility of compromising arterial flow to the leg includes a significant risk of retarded limb growth.⁸

The conventional echocardiogram performed relatively well in confirming the presence of abnormal anatomy of the aortic valve; a positive finding of a dilated aortic root and an eccentric orifice is useful to corroborate the clinical diagnosis of aortic valve disease. However, the absence of these echocardiographic abnormalities does not rule out the aortic valve as the origin of abnormal turbulence, since eight of the 29 patients with aortic turbulence had no echo abnormalities. The possibility exists that these eight patients had no heart disease, and that the murmurs were "innocent". Stein and Sabbah,¹⁵ indeed, assert that most innocent murmurs originate in the aorta, rather than at the pulmonary valve. They used invasive intravascular phonocardiography in 10 adults; only four of their patients had *any* murmur audible, and only two had a grade 2/6 murmur. These two subjects were 50 and 69 years old, which puts them at 37 and 47 per cent risk of having aortic murmurs according to Bruns and Van der Hauwaert.¹⁶ An unrecognised bicuspid aortic valve would have an even higher probability of calcification at that age.⁷ In fact, the 69-year-old *did* have mild aortic stenosis, and not a "functional" murmur: the authors' Fig. 3 shows a withdrawal pressure curve with a gradient across the valve of 9 mmHg. Furthermore, none of the 10 patients in their series had an energy level even one-half the 3 ergs/s per cm² which they claimed earlier¹⁷ would make the turbulence audible at the chest wall. In short, we do not believe that their data support their contention that the ordinary innocent murmur is aortic in origin.

The pulsed Doppler diagnosis of abnormal turbulence in the eight patients with no M-mode echocardiographic abnormalities we believe reflects mild, obstructive dysfunction of the aortic valve, and not innocent murmurs. In the past two years, with the present personnel and equipment, we have analysed the aortic flow in 1039 children suspected of heart disease because of praecordial murmurs in all. We found 97 records free of any turbulence, 772 with turbulence in various chambers and vessels other than the aorta, and 170 with turbulence in the aorta. Thus, we do not believe that aortic turbulence is normal, or compatible with an innocent murmur. It is also well to remember that only three-quarters of patients with known bicuspid aortic valve have significant asymmetry or eccentricity on echocardiography,¹⁸ and very few, if any, bicuspid aortic valves could be completely unobstructive, because the cusps are tethered, stretching across the valve ring.¹⁹

Though we assert that pulsed Doppler echocardiographic turbulence is not compatible with the diagnosis of an innocent murmur, the absence of such turbulence leaves unanswered the question of origin of the soft innocent murmur. We believe that an innocent ejection murmur may be produced by right ventricular ejection, but with insufficient disturbance of flow to produce turbulence detectable by the pulsed Doppler echocardiogram, considering the proximity of the outflow tract of the right ventricle to the chest wall, in relation to the usual site of maximal intensity of the murmur. Another possible source of the innocent murmur may be retrograde transmission from a carotid bruit.²⁰

Recently, absence of high frequency flutter of the aortic leaflets during systole has been suggested as "distinguishing a functional murmur from one resulting from organic valve stenosis".⁹ We retrospectively examined for this phenomenon in a subset of 24 of our patients. This set included six normal subjects and 18 with aortic stenosis determined by pulsed Doppler echocardiography; we could positively identify flutter in five of the six normals, but also in 10 of the 18 patients with aortic stenosis. It is then obvious that the presence of flutter in no way rules out aortic stenosis.

The criteria for diagnosis of aortic stenosis based on auscultation performed well, in terms of specificity, 26 of 30, or 87 per cent. However, these clinical criteria were not as successful for sensitivity, missing three out of the 10. There were two of the criteria that were misleading in a few instances. Transmission of a basal murmur to the carotids was regarded as favouring aortic stenosis, but we found that pulmonary stenosis could transmit to

the neck, and several normal children had a combination of an innocent ejection murmur and a coincidental carotid bruit.²⁰ The second fallible assumption was that a brief diastolic bruit at the left sternal border, in a patient with no pulmonary hypertension, was a result of aortic regurgitation. We have unequivocal documentation of the occurrence of pulmonary regurgitation in children with no other cardiopulmonary disease.

Considering the presence of a bicuspid aortic valve in approximately 1 per cent of the normal population,²¹ many of these patients are probably not diagnosed at present, and do not receive appropriate protection against infective endocarditis. Since the complications are severe, though infrequent,²²⁻²³ improved non-invasive means of accurate diagnosis and follow-up are needed.

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