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Doppler ultrasound in the assessment of severity of coarctation of the aorta and interruption of the aortic arch

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SUMMARY Doppler ultrasound was used to investigate 48 infants and children (age 2 days–16 years, weight 1·0–58 kg) with aortic arch abnormalities. In only 38 of the 42 with an important coarctation was an increased blood flow velocity from the distal arch demonstrated. In three with interruption of the aortic arch an increased velocity recorded from the region of the distal arch was thought to represent ductal flow. There was little difference between the peak to peak and instantaneous maximum gradients in the 20 patients with important coarctation in whom direct pressure measurements both proximal and distal to the obstruction were made at catheterisation. There were poor agreements between Doppler and measured peak to peak and instantaneous gradients in the 17 patients found to have both an increased velocity and important coarctation.

It is concluded that although an increased blood flow velocity in the distal arch is usually demonstrated in coarctation this may not occur with severe obstruction. Furthermore, the maximum velocity is not related to the anatomical severity of the obstruction and the Doppler estimate of pressure drop in coarctation may not even reliably predict that measured at catheterisation.

The diagnosis of coarctation of the aorta is usually based on clinical examination, and further investigations are directed at defining its site and nature to permit the correct surgical approach. Although echocardiography often provides this information and allows surgery to be undertaken safely without catheterisation, another non-invasive diagnostic technique would be useful in patients in whom a clear echocardiographic image of the coarctation site is not obtained.

Reports have suggested that in some instances Doppler ultrasound provides such a technique by demonstrating increased flow velocity in the descending aorta[1–3] and that it can also give accurate information on the gradient across the obstruction.[2] These studies did not measure gradient by invasive means and since our experience has indicated some problems in the application of Doppler ultrasound in this condition, it seemed appropriate to report our findings. Furthermore, previous reports have only briefly considered the different wave form patterns[1] that are obtained from patients with coarctation.

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Patients and methods

We studied 48 patients (14 neonates, nine older infants, and 25 children) aged 2 days–16 years (mean 3 years 6 months) and weighing 1·0 to 58 kg (mean 13·8 kg). In three patients who had undergone coarctation repair the narrowing was minimal on angiocardiography and surgery was not considered necessary. The exact diagnosis was ascertained at surgery in all the others; previous catheterisation had been performed in all but 13 (eight neonates, three older infants, and two children). Of the 23 infants, 18 had preductal coarctation (14 with a ductus arteriosus), two had postductal coarctation, and three had interrupted aortic arch with a ductus arteriosus. Ten patients (including one infant) had already undergone coarctation repair. The table

Table *Summary of the more important clinical and diagnostic features in the different age groups*

Diagnosis and age	*Total No*	*DA present*	*Catheterisation undertaken*	*DA and catheter*	*Previous operation*	*Arch velocity > 2·2 m/s*
Coarctation < 1 month	11	9	4	3	0	11
Coarctation 1–12 months	9	5	6	2	1	8
Coarctation > 1 year	22	1	20	1	6	19
Interruption (all < 1 month)	3	3	2	2	0	3
Insignificant obstruction	3	0	3	0	3	2

DA, ductus arteriosus.

summarises the age groupings, investigation, and findings.

At catheterisation the pressure was measured directly through a fluid filled system both proximal and distal to the obstruction in 23. Pressures on both sides of the obstruction were measured simultaneously in 12; the proximal measurement was made with a catheter in the left ventricle (9) or the aortic arch through the coarctation (3) and the distal one through a needle in the femoral artery or the side arm of a sheath in the iliac artery. In the remaining 11 patients pressures were measured by catheter withdrawal across the coarctation and tracings distal to the coarctation were superimposed on proximal ones to simulate simultaneous pressure measurements; to minimise error, beats with almost equal R–R intervals were chosen. The peak to peak and maximum instantaneous pressure differences were measured from each tracing. Catheterisation was undertaken in seven infants with a ductus arteriosus, with pressures being measured in both the pulmonary artery and descending aorta in five.

The Doppler investigations were performed by means of a 2 MHz probe with a continuous and pulsed wave system (Alfred, Vingmed) and a spectrum analyser (Doptek). After echocardiographic examination a routine Doppler study was performed, the suprasternal or left upper parasternal positions being used to obtain the optimal signal of flow through the aortic arch and coarctation site. From the Doppler signals from the ascending aorta and aortic arch the maximum velocity of blood flow in m/s (V) was measured and the modified Bernoulli equation ($\Delta P = 4V^2$)[4] was used to convert this into Doppler estimate of gradient in mm Hg (P). We then compared the Doppler measurement of flow velocity in the ascending and descending aorta and the maximum Doppler gradient in the descending aorta with the measured maximum instantaneous and peak to peak gradients over the coarctation. In those infants with a preductal coarctation and a ductus arteriosus or arch interruption who underwent catheterisation we also compared the Doppler findings with the gradient from the main pulmonary artery to the descending aorta. In one patient the Doppler study was performed while a balloon catheter was inflated within the ductus.

In the normal individual forward flow in both the ascending and descending aorta is of approximately the same velocity,[5] that is < 2·0 m/s and is usually confined to systole. The wave forms from the distal arch in many of those with coarctation, however, demonstrated continuing forward flow in diastole. The Doppler wave forms were reviewed to assess the timing of forward flow paying particular attention to diastolic flow patterns and determining whether there was no diastolic flow or early diastolic flow only or pandiastolic flow.

Results

INFANTS WITH COARCTATION

Maximum velocities in the ascending aorta were < 2·2 m/s (20 mm Hg) in 19 of the 20 infants, the other having an increased velocity related to the presence of associated aortic stenosis. Doppler echocardiography demonstrated a velocity, apparently from the distal arch, indicating flow away from the transducer of > 2·4 m/s (equivalent to a gradient of 23 mm Hg) in 19 of the 20 infants (fig 1); Doppler gradients ranged from 23 to 80 mm Hg with a mean of 38 mm Hg. No increased velocity was found in the remaining 6 month old infant with a narrow isthmus and a web proximal to a 4 mm ductus; catheterisation was not performed but blood pressure measurements showed the leg pressure to be 30 mm less than that in the arm.

In one neonate in whom Doppler echocardiography had recorded increased velocity of flow away from the transducer (2·85 m/s, equivalent to a gradient of 33 mm Hg) catheterisation while prostaglandin E2 was being infused showed a tight coarctation and a ductus arteriosus with a measured drop from main pulmonary artery to aorta of 31 mm Hg. The Doppler signal disappeared when a balloon was inflated in the ductus arteriosus; it reappeared when the balloon was deflated, which suggests that this flow was ductal and not through the coarctation.

The gradient from the main pulmonary artery to the descending aorta was measured at cath-

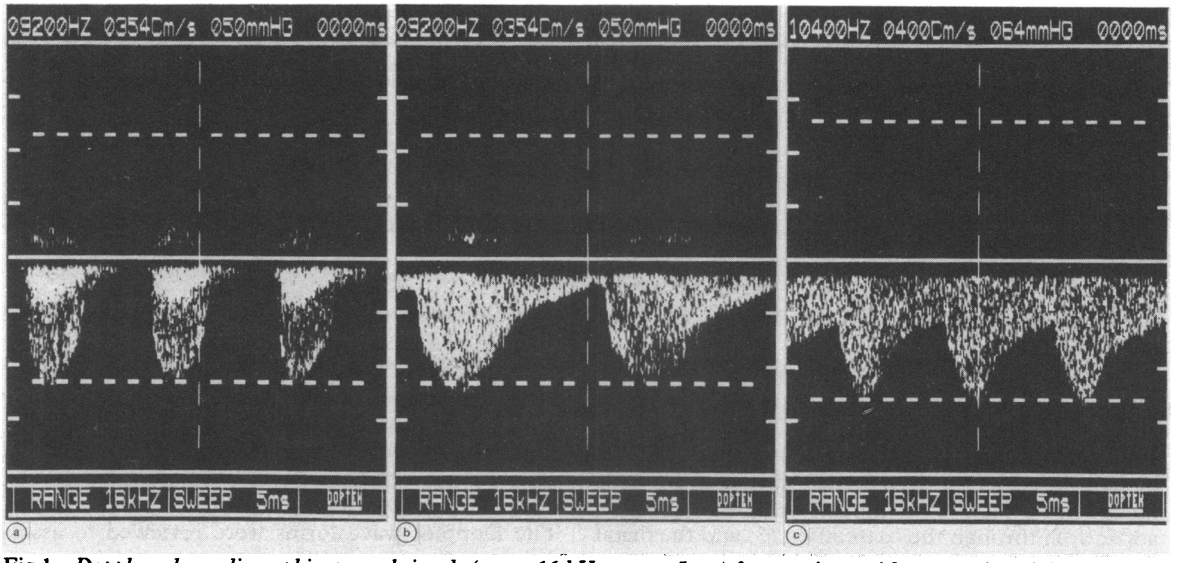

Fig 1 *Doppler echocardiographic spectral signals (range 16 kHz, sweep 5 ms) from patients with coarctation of the aorta. The transverse marker line is positioned at the maximum frequency shift which is displayed on the top left (Hz), followed by the velocity this represents (cm/s) and the gradient (mm Hg) calculated from the simple modified Bernoulli formula. The flow is mainly in systole in (a) and continues throughout diastole in (b) and (c).*

eterisation in five patients with preductal coarctation and ductus arteriosus: the pressure was higher in the main pulmonary artery than in the descending aorta in two (15 and 31 mm Hg difference), approximately equal in the two vessels in two, and 20 mm Hg lower in the main pulmonary artery in the other.

OLDER CHILDREN

Flow velocities from the ascending aorta were normal in all 25. The signal from the distal arch showed velocities of 1·4, 2·4, and 2·9 m/s in the children with recoarctation who were considered not to require further surgery and of at least 2·7 m/s (29 mm Hg) in 19 of the remaining 22. In the other three patients no high velocities away from the transducer were demonstrated. Two had previously undergone coarctation repair; one who had a patch repair when 1·2 kg in weight was shown to have a very long segment with a diameter of only 1–2 mm and the other who had end to end repair had a fibrous membrane. In the third the only demonstration of flow was a continuous signal towards the transducer (fig 2). Careful study failed to demonstrate flow through the coarctation; it proved possible to pass a French gauge 7 catheter through it but angiocardiography showed that the catheter completely occluded the lumen and that there were large collateral vessels.

INTERRUPTED AORTIC ARCH

From the suprasternal notch, with the transducer angled as if to pick up arch flow, Doppler ultrasonography showed flow away from the transducer in excess of 2·2 m/s (4·4, 3·1, and 2·3 m/s) in all three neonates with interrupted arch. Presumably this was ductal flow.

COMPARISON OF PRESSURE GRADIENTS

In all 20 patients with important coarctation in whom pressures were measured on both sides, the instantaneous maximum appeared to be greater than the peak to peak gradient, the difference ranging from 1 to 6 (mean 4) mm Hg in 17 and being 9, 12, and 17 mm Hg respectively in the other three. An increased velocity jet was demonstrated in 17 of these, and comparison of the maximum Doppler gradient with the measured one gave a correlation coefficient r of 0·42 against the peak to peak ($y = 0·46x + 26·4$) and $r = 0·36$ ($y = 0·37x + 27·2$) against the instantaneous maximum gradient. Comparison of the values by the method of Bland and Altman[6] (fig 3) confirms the poor agreement; the mean difference in gradients is −7·5 mm Hg with the differences within 2 SD being from −45·4 to 30·3 mm Hg, which is unacceptable for clinical purposes. There may have been delay in the recordings in the femoral artery but this would have given a greater instantaneous gradient, and the results suggest that in coarctation there is little difference between this and the peak to peak gradient. No relation was observed between maximum velocity (and thus gradient) and the anatomical obstruction;

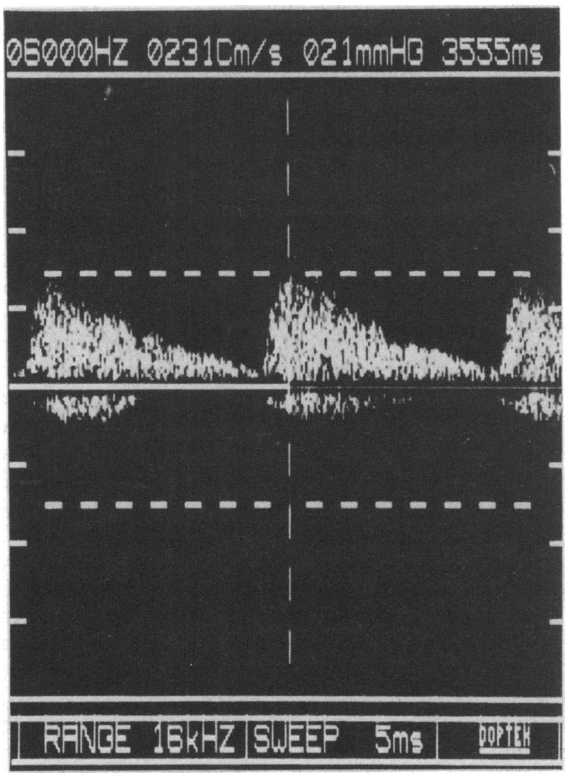

Fig 2 *Doppler echocardiographic spectral signal (range 16 kHz, sweep 5 ms) obtained from the suprasternal notch in the patient in whom the only increased velocity flow was towards the transducer, presumably through collaterals. The marker line indicates a maximum frequency shift of 6000 Hz representing a velocity of 2·31 m/s.*

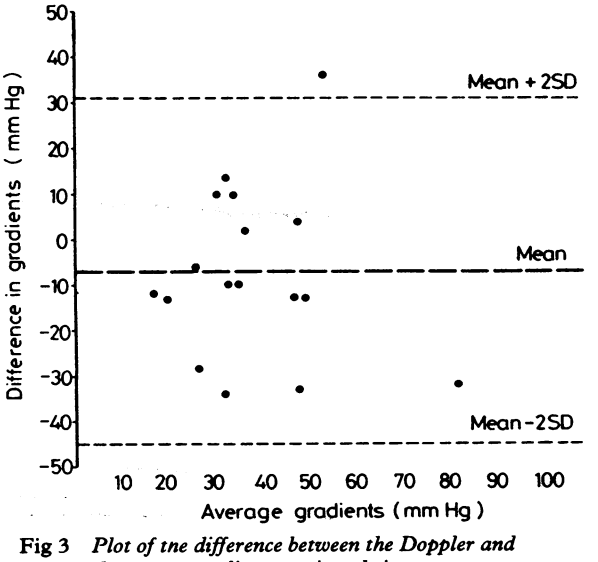

Fig 3 *Plot of tne difference between the Doppler and measured pressure gradients against their mean.*

in two patients in whom the catheter would not pass through the narrowing, Doppler estimate of gradient was 26 mm Hg in one and no flow was demonstrated in the other.

WAVE FORMS

Study of the flow patterns showed that in diastole flow was generally of low velocity and its recognition was related to the level of the high pass filter. With this set at 800 Hz (a velocity of just over 0·3 m/s) continuous flow in diastole was demonstrated in 21 of the 38 with important coarctation and increased velocity in the distal arch. In a further 10 flow apparent in early diastole did not continue throughout diastole and in seven flow was confined entirely to systole. The duration of flow was not related to the maximum velocity or gradient. The end diastolic pressure difference across the coarctation could be measured in 10 patients in whom an increased velocity was recorded; flow throughout diastole occurred in the five with a measured end diastolic gradient of $\geqslant 6$ mm Hg but was not found in those with a lower gradient.

Discussion

In coarctation of the aorta Doppler ultrasound techniques might be expected to be able to confirm the presence and site of the obstruction and measure the pressure difference across it.

CONFIRMATION OF COARCTATION

Hatle and Anglesen reported that with continuous wave Doppler echocardiography a jet from beyond the coarctation could usually be obtained in patients with a moderate coarctation but in only half of those with severe obstruction[1] but Wyse *et al* found a jet in all 30 patients studied.[2] In the latter investigation, performed mainly in patients who had previously undergone coarctation repair, a good correlation was reported between the Doppler estimate of maximum velocity and the difference between blood pressures measured in the arm and leg with a sphygmomanometer, though no invasive pressure measurements were made. Robinson *et al* also published results in six infants; in three with critical aortic stenosis high velocity flow was measured in the ascending aorta whereas in three with coarctation a high velocity flow was measured in the descending aorta; this suggested that a clear distinction could be made on the basis of Doppler results.[3]

For practical purposes it is appropriate to consider the two different patient populations with coarctation, the infant with preductal coarctation and a ductus arteriosus and the others, often older patients, who usually have a postductal coarctation.

In the group with postductal coarctation our findings are more in accord with those of Hatle and Angelsen[1]; in most cases Doppler demonstrates high velocity flow but where the obstruction is very severe this may not be demonstrable. In the three children in whom Doppler did not demonstrate the high velocity there was no clinical doubt as to the presence of coarctation, however, and a site near the arch was likely in one because continuous flow towards the transducer suggested collateral flow.

Diagnosis of coarctation of the aorta in the newborn can be difficult; echocardiography is not always diagnostic and there is also the possibility of confusion between coarctation and interruption of the arch. Robinson *et al* suggested that in the neonate increased velocity flow away from the transducer indicates coarctation[3]; in one of their patients pressures on both sides of the obstruction were equal and a high velocity should not have been recorded. We speculate that the increased flow velocity in those with arch interruption is a result of ductal flow and may be wrongly interpreted as representing coarctation. It seems that ductal rather than coarctation flow may also be recorded on occasions in neonates with severe coarctation, as in the newborn infant in whom the obstruction of the ductus abolished the signal. Thus we propose that an increased velocity signal should be interpreted as indicating arch obstruction, not necessarily coarctation, because it may be found with interruption and a restrictive ductus.

MEASUREMENT OF PRESSURE DROP

Since simple measurement of arm and leg blood pressures with a sphygomomanometer will give a good estimate of the systolic pressure drop, the ability of Doppler to measure the gradient could be regarded as being of little practical clinical value. Measurement of leg blood pressure, however, can be difficult and in some patients who have undergone coarctation repair the use of arteries for the repair, pressure monitoring, or cardiac catheterisation can occasionally make measurement of arm blood pressure unreliable.

Although we have found a good correlation between catheter and Doppler derived gradients for other obstructive lesions,[7-9] the present study has shown that in our laboratory the simple Bernoulli formula does not give a good correlation with direct pressure as we recorded it. Nor can the poor agreement be attributed to the difference between instantaneous maximum and peak to peak gradients, which in our experience is negligible.

Doppler echocardiography might have been expected to underestimate the pressure gradient in some patients with coarctation either because of misalignment with the direction of flow or because it is known to give low values where the orifice is small,[10] especially if the narrowed segment is long.[11] It is less easy, however, to explain the overestimation of gradient by more than 20 mm Hg in three children; the study was performed at catheterisation in all three and in only one was the distal pressure measured while a catheter was through the coarctation. It may be that this was collateral flow but the direction and velocity were different from that in the one child where this was considered to have been clearly demonstrated (fig 2).

Since the measurement of pressure gradient with continuous wave Doppler has proved reliable in other situations with discrete obstruction it is difficult to explain why it should overestimate the gradient in some patients with coarctation. This may be due in part to the use of the modified Bernoulli formula, $\Delta P = 4V^2$, which does not take the proximal velocity into account, thus potentially overestimating the pressure drop. The discrepancy may also be related to the measurement technique, in all three the distal pressure having been recorded from the femoral artery. At catheterisation the pressure in the femoral artery can be higher than in the ascending aorta[12] because of the progressive distortion of the pressure wave as it is transmitted through the arterial system.[13] It is not certain whether this applies in coarctation, but the measured gradient may have underestimated the true one and Doppler estimation may well be more accurate than the results presented in this study suggest.

This study has shown that although in most patients Doppler echocardiography allows an accurate demonstration and localisation of coarctation of the aorta, the findings should be interpreted with full knowledge of the weaknesses of the technique and within the context of the whole clinical and noninvasive investigation. The results suggest that in the newborn infant extreme caution must be exercised; the presence of an increased velocity is likely to indicate coarctation but can also be found with a restrictive ductus arteriosus with interruption (or very severe coarctation). Since good images of the aortic arch are usually obtained from these babies it may be that Doppler echocardiography has little to add to clinical examination and conventional cross sectional echocardiography in this situation. In older children it can be difficult to demonstrate the coarctation by cross sectional echocardiography and Doppler echocardiography will often indicate its presence and site. In very severe obstruction, however, the failure of Doppler echocardiography to show an increased velocity in the presence of clinical coarctation requires reassessment of the noninvasive information and might even indicate the

need for angiography. When a signal suggesting the presence of coarctation is obtained the maximum velocity (and thus Doppler gradient) does not correlate with the anatomical severity of the narrowing. Furthermore, our findings indicate that Doppler echocardiography may be less accurate in measuring pressure gradients in coarctation than in other obstructive lesions and we believe that further correlative studies are needed.

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