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Obstruction of superior vena caval pathway after Mustard's repair[1]

Reliable diagnosis by transcutaneous Doppler ultrasound

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SUMMARY Superior vena caval pathway obstruction is an important cause of late morbidity and mortality after Mustard's operation. Clinical detection of such obstruction is frequently difficult because of the development of an extensive collateral venous circulation which decompresses the superior vena cava.

As flow is less likely to be normal than pressure, we recorded jugular venous flow profiles transcutaneously in 34 patients after Mustard's operation for simple transposition using a bidirectional Döppler blood velocimeter. There was an independent diagnosis by catheterisation in 14 cases, 4 of whom were restudied after revision of obstruction. Most patients showed a dominant forward flow peak during ventricular diastole and none of these showed any clinical evidence of pathway obstruction. This pattern was considered normal after a Mustard repair, particularly as it was also seen in 4 patients immediately after a revision operation. The jugular flow profile is not the same as in children without heart disease, presumably because of the anatomical differences and the reduction in atrial compliance produced by Mustard's operation. Jugular venous flow in patients even with mild superior vena caval pathway obstruction differed from that in the unobstructed group by exhibiting either non-pulsatility or, more commonly, a dominant forward flow peak in ventricular systole. This quick, simple, and convenient test proved to be diagnostic of the presence or absence of obstruction and, as many patients are asymptomatic, is now the most effective way of screening for this complication. It may thus make catheterisation unnecessary.

Superior vena caval pathway obstruction is an important cause of late morbidity and mortality after Mustard's operation (Mustard, 1964) for the physiological correction of complete transposition of the great arteries (Macartney *et al.*, 1979). The incidence is related to the material and shape of the patch (Stark *et al.*, 1974). The obstruction is not within the superior vena cava itself, but is within the superior vena caval pathway created in forming a new systemic venous atrium during the operation (Fig. 1), and is either upstream or downstream of the right atrial appendage. This has interesting and important effects on the clinical presentation of superior vena caval pathway obstruction, because the haemodynamic effects are different from those of the more familiar superior vena caval obstruction, which occurs within the cava itself. Whatever process occludes the superior vena cava usually occludes the azygos vein as well. By contrast, in caval pathway obstruction, the superior vena cava and azygos vein are wide open. An extensive collateral circulation therefore develops through a dilated azygos and lumbar venous system, allowing superior vena caval blood to return to the heart through the inferior vena cava (Fig. 1) (Stark *et al.*, 1972, 1974; Silove and Taylor, 1976).

Thus complete superior vena caval pathway obstruction can occur without any clinical signs if the collateral venous and inferior vena caval pathways are unobstructed. Though jugular venous engorgement and facial oedema may appear before the development of a collateral venous circulation, they are usually late signs indicating obstruction of

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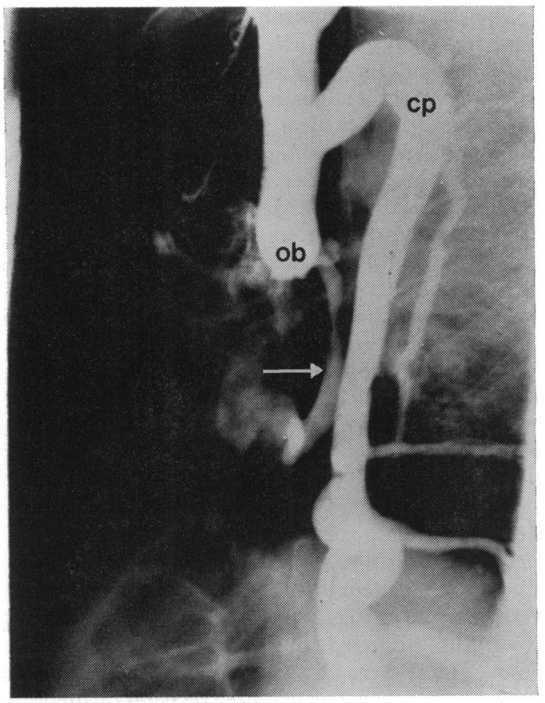

Fig. 1 *Superior vena caval angiogram showing post-Mustard obstruction (ob) with drainage through a collateral pathway (cp), blood eventually returning via the inferior vena cava. The vessel just posterior to the heart* (white arrow) *is the coronary sinus which was opacified from a left superior vena cava.*

both the inferior and superior vena caval pathways. The need for a method of detection of pathway obstruction before the development of clinical signs is obvious. Previous studies have concentrated on patients who are symptomatic by virtue of a raised superior vena caval pressure (Stark *et al.*, 1972, 1974; Venables *et al.*, 1974; Silove and Taylor, 1976). However, when asymptomatic patients are routinely investigated after Mustard's operation, our own experience and that of others (Hagler *et al.*, 1978) shows that severe, even complete, superior vena caval pathway obstruction may be found angiocardiographically, even when there is only a very small pressure gradient between the superior vena cava and systemic venous atrium, presumably because of a large collateral flow. This is consistent with the finding that such patients do not necessarily have a raised or abnormal jugular venous pressure and pulse.

Our hypothesis was that flow is less likely to be normal than pressure and that detection of superior vena caval pathway obstruction might therefore be possible non-invasively using a transcutaneous Döppler ultrasound bidirectional blood velocity probe. If this were so, use of this method would have the added advantage that it might obviate the need for confirmation of the diagnosis by cardiac catheterisation, which is not only time-consuming, expensive, and unpleasant for the patient, but may also present a real problem of access to veins which tend to become thrombosed either because of previous catheterisations or because of polycythaemia before Mustard's operation.

Methods

Jugular venous flow velocity recordings were made in 34 patients (age range 0·4 to 17·2 years, mean 6·8 years), all of whom had previously had a Mustard's repair for simple complete transposition of the great arteries between 1 week and 12·3 years (mean 5·0 years) before investigation. Four patients were studied before and after revision of obstruction giving a total of 38 recordings. The baffle was fashioned from pericardium in 23 and from Dacron in the remainder. Recordings were made either routinely at an outpatient clinic or when patients were admitted for cardiac catheterisation or revision of Mustard's operation. A transcutaneous bidirectional Döppler ultrasound blood velocimeter[1] was used, which incorporated an electrocardiogram and flow channel, plotting both directly onto an integral paper chart recorder. The machine also emits an audible tone whose frequency is proportional to the velocity signal. The transmitter frequency was 7·52 MHz.

Patients were quiet and supine with no pillow and the head was turned slightly away from the examination. The limb leads of the electrocardiograph were attached to the patient and the instrument checked for an appropriate electrocardiographic deflection (demonstrating the P waves in particular) on the recording paper. The external probe was applied to the skin using Aquasonic 100 ultrasound transmission gel[2]. The probe was placed over the jugular vein and directed toward the inferior border of the contralateral scapula. Its position was then adjusted until the signal reached a maximum as determined both by ear and by looking at the display on a Sonicaid BV 383 memory oscilloscope. When this signal was consistent, reproducible, and pure, free from components of flow from adjacent arteries, the electrocardiogram and jugular flow signals were then recorded simultaneously on the chart recorder at a paper speed of 50 mm/s using a foot-switch control. Recordings were obtained from both left and right jugular veins, after which a carotid

[1]Sonicaid Ltd., England. Model BV381.
[2]Parker Laboratories, New Jersey.

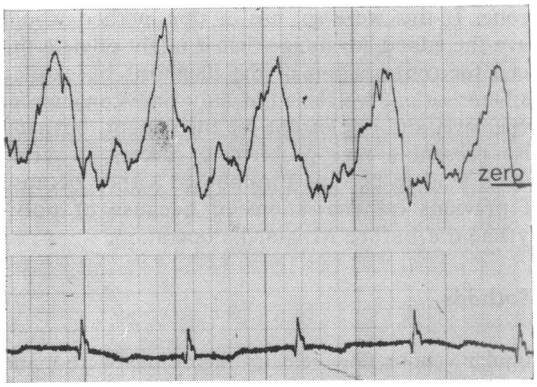

Fig. 2 *Jugular venous flow recording after Mustard's operation with no superior vena caval pathway obstruction. Dominant forward flow occurs during ventricular diastole.*

recording was also made to verify that there was no arterial component in the venous flow records. Zero flow was recorded routinely as a reference level and both venous and arterial forward flows were displayed as positive deflections by the appropriate use of the reversal switch, standard to this instrument.

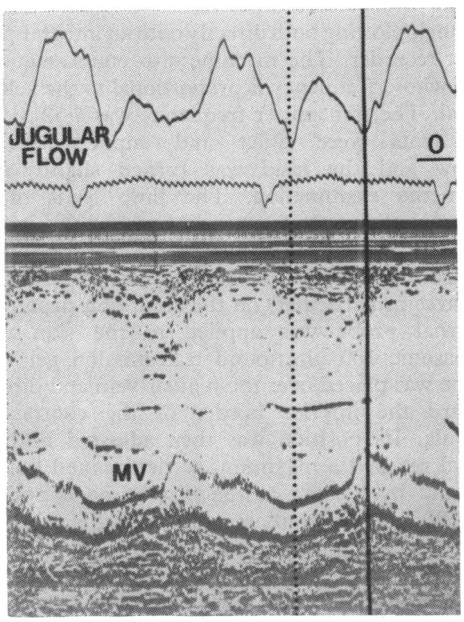

Fig. 3 *Simultaneous echocardiogram* (below), *electrocardiogram* (middle), *and jugular flow recording* (above) *in a patient with no superior vena caval pathway obstruction. Systolic forward flow begins just after the left ventricular posterior wall starts to move forward* (dotted line). *Diastolic forward flow begins just after mitral valve opening* (solid line). *Paper speed 50 mm/s.*

The timing of mitral valve movement was compared with the jugular flow profile in 4 patients by feeding Döppler, electrocardiographic, and echocardiographic[1] signals directly to a Cambridge fibre-optic physiological recorder[2] with echoscan converter, and recording these signals onto the integral ultraviolet paper output.

At cardiac catheterisation, criteria for the diagnosis of superior caval obstruction were a superior vena caval pathway of less than half the diameter of the superior vena cava in the frontal view of the superior vena caval angiogram, the passage of a significant amount of contrast medium from superior to inferior caval territory via the azygos system, and a mean pressure gradient of more than 4 mmHg between superior vena cava and systemic venous atrium (Silove and Taylor, 1976).

Results

Thirty-four patients (89%) had recordings similar to that shown in Fig. 2 in that the dominant forward flow peak occurred during ventricular diastole. Four of these were obtained when the pathway was known to be unobstructed after revision of superior vena caval pathway obstruction. Six more patients in this group had had previous cardiac catheterisation studies in which the superior vena caval pathway was shown to be unobstructed. The jugular venous wave pattern seen in these patients was taken to be that associated with a normal wide unobstructed superior vena caval pathway. This waveform is characterised by a large forward flow in ventricular diastole with less flow in ventricular systole (Fig. 2). This increase in forward flow is shown in Fig. 3 to occur only after the mitral valve has opened, further evidence of an unobstructed superior vena caval pathway.

However, in the 4 cases before revision of pathway obstruction and in a further 4 awaiting revision (after confirmation of diagnosis by cardiac catheterisation), flow patterns were significantly different from those in the unobstructed group. In these patients the waveform was either non-pulsatile (Fig. 4a) or, more commonly, showed dominant forward flow during ventricular systole (Fig. 5a). Whatever the waveform before revision it reverted to the same unobstructed pattern after revision (Fig. 4b and 5b). These results are summarised in the Table.

A number of patients showed flow reversal during atrial systole, but this could not be correlated with the presence, absence, or degree of obstruction,

[1]Ekoline 20, Smith-Kline Instruments Ltd.
[2]Type 01005, Cambridge Medical Ltd., England.

Table *Effect of superior vena caval pathway obstruction (SVCPO) on jugular venous waveform in 18 catheterised or reoperated patients*

		Dominant diastolic wave	*Dominant systolic wave or non-phasic trace*
SVCPO	present	0	8
	absent	10	0

$P < 0.001$.

the placement of the baffle (particularly its relation to the right atrial appendage), or to the cardiac rhythm.

Discussion

Since Kalmanson *et al.* (1968, 1969) showed that jugular venous flow waveforms detected by transcutaneous Döppler ultrasound were an accurate reflection of the total venous return, the technique has so far found few applications in paediatric cardiology apart from the diagnosis of atrial (Kalmanson *et al.*, 1972) and ventricular (Kalmanson *et al.*, 1974) septal defects and estimation of shunt size, for which other equally reliable non-invasive techniques exist. Kalmanson's results suggest that certain jugular venous waveforms are diagnostic of specific lesions and this view has been endorsed by Sivaciyan and Ranganathan (1978), who established that these waveforms reflect

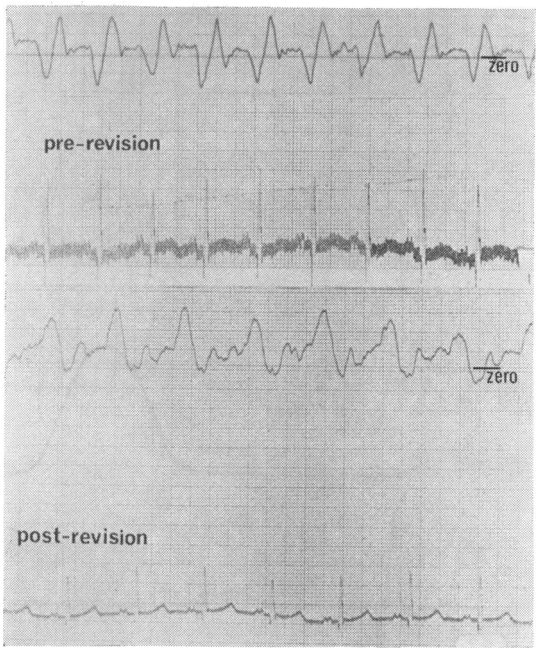

Fig. 5 (a) (above) *Jugular venous flow recording in a patient after Mustard's operation but with severe superior vena caval pathway obstruction. Dominant forward flow occurs in ventricular systole. This was a more common finding than completely non-pulsatile flow.* (b) (below) *After revision of obstruction the dominant wave was in ventricular diastole.*

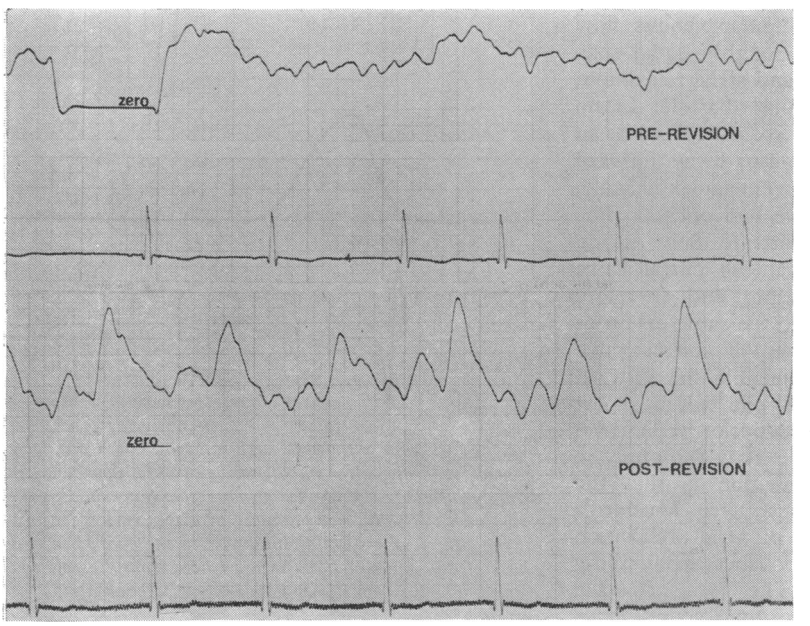

Fig. 4 (a) (above) *Jugular venous flow recording in a patient after Mustard's operation but with severe superior vena caval pathway obstruction. The flow pattern is non-pulsatile.* (b) (below) *The recording in the same patient after surgical revision of obstruction now shows dominant forward flow in ventricular diastole.*

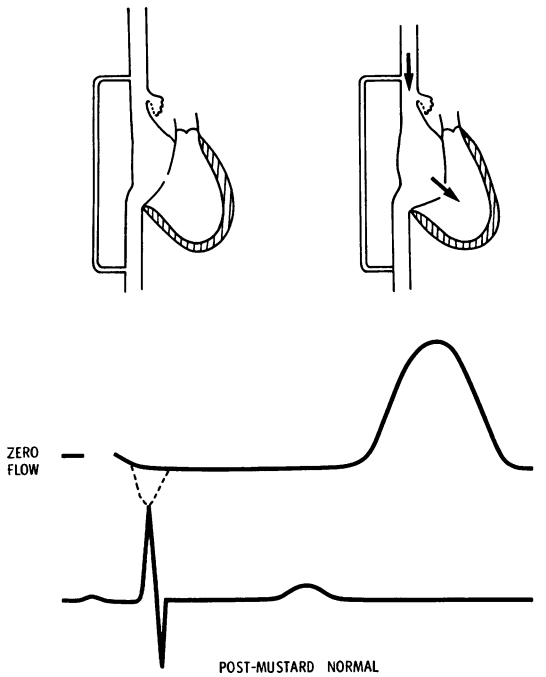

Fig. 6 *With an unobstructed superior vena caval pathway, ventricular filling after mitral valve opening was reflected in a dominant jugular forward flow wave during diastole; there was sometimes flow reversal during atrial systole.*

derangements in right heart haemodynamics irrespective of the underlying aetiology.

In the normal heart, the jugular venous flow record shows two maxima of roughly equal size, one during ventricular systole and atrial relaxation, and the other during ventricular diastole. In no post-Mustard case was this so. Even with an unobstructed superior vena caval pathway, forward flow was much greater during ventricular diastole, occurring once the mitral valve had opened. The explanation of the reduced forward flow during ventricular systole may be that the mitral valve descends less than the tricuspid and therefore cannot increase atrial volume to the same extent as in the normal heart. However, if mitral valve movement is to be held responsible for forward flow during ventricular systole, one would expect less flow in the presence of superior vena caval pathway obstruction, whereas exactly the opposite occurs. A more likely explanation is that the reduction of atrial compliance after Mustard's operation negates the effects of atrial relaxation and reduces or completely abolishes atrial filling during ventricular systole. The loss of atrial compliance is reflected in the systemic venous atrial pressure pulse by a steep y-descent (Silove and Taylor, 1976). Such a reduction in atrial compliance might also explain why a few of the jugular venous recordings from post-Mustard patients showed flow reversal during atrial systole. The origin of the diastolic flow peak is depicted diagrammatically in Fig. 6.

How pathway obstruction creates an abnormal jugular venous waveform is depicted in Fig. 7. Because of the obstructed pathway the forward flow peak corresponding to mitral valve opening is lost. Pulsatility may be lost completely when collateral channels are poorly developed or inferior vena caval pathway obstruction also present. However, some patients with severe obstruction of both caval pathways had pulsatile flow nevertheless. Why, in patients with superior vena caval pathway obstruction, the peak of jugular venous forward flow occurs during ventricular systole remains unclear at present. One explanation would be that the jugular flow pattern is out of phase with forward flow through the mitral valve during ventricular diastole, because of the length, volume, and compliance of the collateral venous system. How-

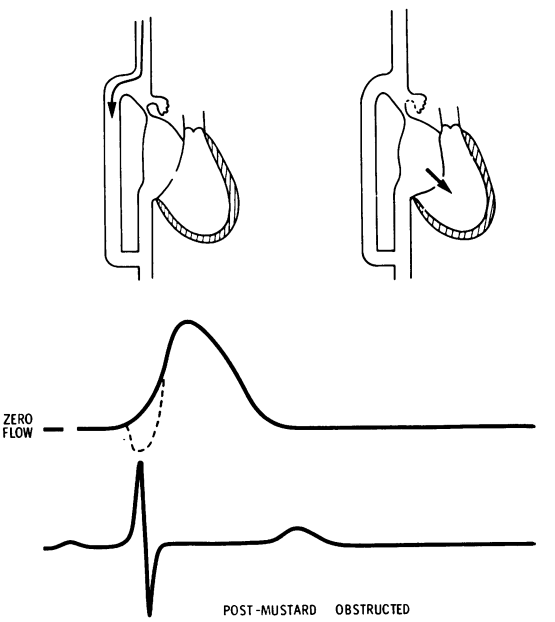

Fig. 7 *With an obstructed superior vena caval pathway, ventricular filling after mitral valve opening does not directly influence the jugular venous flow pattern. Blood flow down the collateral pathway probably occurs mainly during ventricular systole giving rise to the change in phase of dominant jugular forward flow from diastole to systole. There is sometimes flow reversal during atrial systole.*

ever, if this were the explanation, it is surprising that the timing with ventricular systole is so exact. An alternative explanation is that motion of the heart during systole exerts traction on the superior vena cava in such a way as to encourage forward flow. Nevertheless, whatever the mechanism involved, the resultant pattern of jugular venous flow is so obviously different from that in unobstructed patients as to make the technique reliably diagnostic of superior vena caval pathway obstruction. Further, it has been shown to detect even mild and asymptomatic total obstruction which was hitherto impossible without cardiac catheterisation, and has done so with no false positive or negative results. The patient population was deliberately biased towards patients with obstruction, so the overall incidence of obstruction is not representative.

The use of Dacron baffles, which were associated with rather rapid progression of obstruction, has been discontinued. Our present policy in patients with pericardial baffles is not to operate unless both superior and inferior vena caval pathways are obstructed, since experience has shown that even complete obstruction of one pathway is well tolerated for many years provided the other pathway is clear, because of the collateral circulation. This means that though an asymptomatic postoperative patient with a normal jugular flow trace does not require cardiac catheterisation to exclude caval pathway obstruction, an asymptomatic patient with an abnormal jugular venous flow trace does require catheterisation not to detect superior vena caval pathway obstruction, but to assess its severity and ensure that the inferior vena caval pathway is clear. In a patient with facial oedema and jugular venous engorgement, a jugular venous flow recording denoting obstruction has already been used as an indication for revision of the Mustard operation without further cardiac catheterisation. As the technique also provides an early diagnosis of impending superior vena caval obstruction, the information thus obtained can be used to assess the effects of modifications of surgical technique which attempt to reduce the incidence of caval pathway obstruction. The study is now being extended to patients with complex transposition, and also to assessment of the inferior vena caval pathway.

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