Pulsed Doppler echocardiographic assessment of the pulmonary venous pathway after the Mustard or Senning procedure for transposition of the great arteries

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ABSTRACT     Pulsed Doppler evaluation of pulmonary venous flow was performed in three groups of patients who had undergone either a Mustard or Senning procedure. Group I consisted of 43 patients in sinus rhythm who had undergone the Mustard procedure 9 months or more before the Doppler examination and 16 who were evaluated immediately after surgery. Group II consisted of 12 patients in sinus rhythm who had undergone the Senning procedure 9 months or more before the Doppler evaluation. Group III consisted of eight patients with mid baffle obstruction of the pulmonary venous atrium and seven with isolated stenosis of the pulmonary vein of which but one had previously had associated mid baffle obstruction. In those without obstruction, the pulmonary venous flow pattern mirrored the left atrial pressure trace, with peak forward flow occurring during the x and y descent. Obstruction produced a specific high-velocity turbulent pattern, whether at the mid baffle or pulmonary venous level. This technique provides reliable noninvasive information about pulmonary venous flow after the Mustard or Senning procedure. *Circulation* 73, No. 4, 765-774, 1986.

INTERATRIAL REROUTING of pulmonary and systemic venous blood by the Mustard or Senning procedure is still the most common operation for patients with simple transposition of the great arteries. In the former the pericardium is used to form the baffle, while in the latter the native atrial tissue is used.1-3 The theoretical advantages of the Senning procedure is that it preserves normal atrial function and growth of the pathways.4-6

Both procedures may be complicated by systemic or pulmonary venous obstruction (PVO), and more rarely by isolated stenosis of the pulmonary veins.7-11 Unless PVO is severe, clinical recognition is difficult and diagnosis necessitates cardiac catheterization.12-17

Recently cross-sectional echocardiography has been used to assess the midportion of the pulmonary venous atrium after the Senning procedure.18,19 However, problems may occur in the older patient in whom a reliable window is absent. Furthermore, this technique does not identify those patients with stenosis of the pulmonary veins.11

The left atrium acts as a reservoir for pulmonary venous return and augments left ventricular filling during atrial contraction. Pulmonary venous flow measured experimentally appears to mirror left atrial pressure events, particularly the slope of the x and y descent.20-22 More recently it has been possible to assess pulmonary venous flow by pulsed Doppler echocardiography.23,24 The purpose of this study was to compare pressure and flow profiles in patients who have undergone the Mustard Senning procedure with normal subjects and those with PVO either at mid baffle or pulmonary venous level.

Materials and methods

Three groups of patients were studied. Group I consisted of 59 patients who had undergone the Mustard procedure and were evaluated at the Hospital for Sick Children. In 43, the surgery had been performed more than 9 months before the study. In
these 43 the age at study ranged from 2 to 14 years (mean 8.3), with a mean age at surgery of 1.5 years. Postoperative hemodynamic data were available on 33, with nine studies being performed at the same time as the Doppler assessment. In these nine the pulmonary venous atrial pressure measurement was made with a No. 5 Millar high-fidelity catheter introduced retrogradely through the tricuspid valve. The pressure and Doppler traces were recorded at a paper speed of 100 mm/sec.

In the other 16 patients the studies were performed in the immediate postoperative period. Their ages ranges from 0.33 to 1.1 years (mean 0.66), with a mean age at surgery of 0.75 years. Simultaneous left atrial pressure measurement (from indwelling pulmonary venous atrial pressure lines) and Doppler interrogation were performed in eight. In nine, left atrial pressure was measured at the preoperative cardiac catheterization.

Group II consisted of 12 patients who had undergone a Senning procedure at either the Montreal or Ottawa Children’s Hospital more than 9 months before the study. Their ages ranged from 0.91 to 6 years (mean of 2.65), with a mean age at surgery of 0.6 years. In group II, postoperative hemodynamic data obtained at a different time were available in eight patients.

Group III consisted of eight patients with mid-baffle obstruction of the pulmonary venous atrium, two of whom had undergone a Mustard repair and six a Senning procedure. Angiographic and/or surgical confirmation of the site of obstruction was available in seven. Six patients had isolated pulmonary vein stenosis, of which all but one had had previous mid-baffle obstruction. In the remaining patient left-sided venous obstruction was documented by Doppler echocardiography before and was subsequently confirmed at the Mustard procedure.

**Echocardiographic technique.** All patients were studied while in the supine position, and the subcostal and apical windows were used whenever possible. In 45, it was possible to obtain a satisfactory image from the subcostal window, while in every case, the apical position allowed adequate identification of the pulmonary venous pathway. The patients were studied during quiet respiration with an Advanced Technology Laboratory Mechanical Sector Scanner (Mark 600) with the use of a 3, 5, or 7.5 MHz scanhead for direct visualization of the pulmonary venous pathway and ostia of the pulmonary veins (figure 1). The ATL system also incorporates a pulsed Doppler unit with a transmitted frequency of 3 and 5 MHz. The sampling position can be adjusted to interrogate any specific area in question, at depths from 3 to 17 cm, while the axial sample volume size can be varied between 1.5 to 9 mm.

The sample volume size was maintained between 1.5 and 2 mm (axial dimension) to minimize interference from wall movement. The wall filter was set at 200 Hz to avoid filtering of the lower frequency signals.

From the apical four-chamber cut, the pulmonary and sys-

![FIGURE 1. Top left. An apical four-chamber cut from a patient who underwent the Mustard procedure. The sample volume is placed where the right pulmonary veins enter the pulmonary venous atrium. Bottom left. The spectral trace from the same patient. Note the three phases of flow, one reversed peak during atrial systole followed by one forward during the x descent and a second peak during the y descent. Top right. An apical four-chamber view demonstrating the sample volume in a left pulmonary vein. Bottom right. The spectral trace demonstrating a biphasic flow pattern. The velocity of flow is lower than from the right pulmonary veins due to the increased angle between the Doppler sample volume and the direction of pulmonary venous flow. ax = atrial systole; lv = left pulmonary vein; lv = left ventricle; pva = pulmonary venous atrium; rv = right ventricle; sva = systemic venous atrium; vd = flow during y descent; vs = flow during the x descent.](http://circ.ahajournals.org/content/111/6/766/F1)

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temic venous pathways could be individually visualized, requiring superior angulation for the former and inferior angulation for the latter. The left lower lobe pulmonary vein could be seen entering near the left atrial appendage (figure 1), and the right upper lobe vein could be seen entering toward the center of the pulmonary venous atrium (figure 1). Doppler interrogation was performed within the vein, just distal to the vein, and then in the middle of the pulmonary venous atrium and was finally continued inferiorly through the tricuspid valve into the right ventricle. With this latter maneuver associated tricuspid regurgitation could be identified. Tricuspid regurgitation was graded according to the width and depth of the regurgitant jet as detected by flow mapping. The pulmonary venous atrium was arbitrarily divided into three areas. Mild regurgitation was a jet within the proximal one-third from the anulus, moderate regurgitation a jet in the proximal one-third to two-thirds, and severe regurgitation a jet in an area greater than two-thirds.

In the precordial four-chamber cut, the orifice, and the first 2 to 3 mm of the vein could be seen, but not enough of its length was visualized to allow for angle correction and calculation of an absolute peak velocity. For this reason a ratio of the area of flow occurring during the x descent vs that in the y descent, which would be independent of the angle of Doppler interrogation, was calculated. This was achieved by dropping a perpendicular line to the base in the trough between the period of flow during the x and y descent. The areas were planimetered and the heart rate was calculated.

Flow patterns in patients in groups I and II, who were in sinus rhythm, were compared with those in normal subjects examined in our laboratory and with those in patients who had documented PVO. Patients with significant tricuspid regurgitation were excluded because the regurgitant jet interfered with the spectral trace.

Results

Flow was detected in the right upper lobe pulmonary vein in every patient. The left lower lobe pulmonary vein was sampled in 75 patients, with flow being detected in 74. In one patient who had previously undergone surgery for mid baffle obstruction associated with left PVO, absence of flow in the left-sided veins corresponded with absence of flow on a lung perfusion scan. In the remaining patients, who were assessed early in the study, documentation of left-sided pulmonary venous flow had not been obtained.

Group I. Of the 43 patients who had undergone surgery more than 9 months before the study, 31 were in sinus rhythm and did not have significant tricuspid regurgitation. In 29 patients forward flow was biphasic. These forward peaks mirrored the pressure waveform in the nine patients with a simultaneous left atrial pressure trace so that there was forward flow during the a to x and v to y descent, with minimal flow at the peak of the v and a waves (figure 2). In two patients there were two peaks during the x descent, one corresponding with the a to c and the second with the c to x descent of the pressure trace. When the sample volume was moved just beyond the orifice of the pulmonary vein, the flow changed from being smooth and laminar to an irregular pattern with the component during the x descent being affected more than the y descent. As the sample volume was moved closer to the tricuspid valve, the y component again became laminar. When the sample volume was moved through the tricuspid valve, the atrial systolic component could be identified in each case.

Sampling in the left pulmonary vein from the precordial four-chamber cut demonstrated a similar pattern, but it was of lower velocity due to the increased angle, with loss in many of the atrial systolic components (figure 1).

In the remaining patients (n = 16), who were studied immediately after the Mustard procedure, a change in the velocity profile was noted. Before surgery dominant flow had occurred during the x descent, which is seen in normal subjects (figure 3). After the Mustard procedure, in those in sinus rhythm the velocity profile changed from that of dominant flow during the x descent to dominant flow during the y descent, despite a similar heart rate before and after surgery (figure 3). Baffle movement, observed during respiration in each case, had an effect on the velocity profile, resulting in variations in the flow during the x and y descent of the left atrial pressure trace.

Group II. This group consisted of patients who had undergone the Senning procedure. Twelve were in sinus rhythm and had no evidence of tricuspid regurgitation. As in the patients who underwent the Mustard procedure, the precordial four-chamber cut provided the best assessment of both right- and left-sided pulmonary veins. In 11 patients forward flow in the right pulmonary vein was biphasic, while in one triphasic forward flow was documented. In each case reversed flow during atrial systole was detected in the pulmonary vein, and forward flow was detected through the tricuspid valve during the same phase of the cardiac cycle. The velocity profile in the left pulmonary veins from the precordium was similar to the right-sided pattern, but was of lower velocity, as in the patients who underwent Mustard repair. In eight patients, as the sample volume was moved distal to the vein orifice there was no change in the velocity profile, unlike in the Mustard group. In each, the central portion of the pulmonary venous atrium was narrower than the proximal or distal segments, although there was no pressure drop across this region. With further movement toward the tricuspid valve the pattern changed to one of dominant flow during the y descent. In four patients in whom the midportion of the pulmonary venous atrium was wider, there was decreased flow during the x descent that was similar to that in the patients who underwent the Mustard procedure.
Pulmonary venous atrial waveforms: Senning vs Mustard procedure. Analysis of covariance, performed to remove the effect of the heart rate on the ratio of flow during the x and y descent, demonstrated a significant difference between the patient groups and the normal population. The mean heart rate–adjusted ratio for the normal population was 1.33 ± 0.01, that for the Senning group was 1.02 ± 0.04, and that for the Mustard group was 0.85 ± 0.01. The level of significance for the differences between the Senning and control groups, the Mustard and control groups, and the Senning and Mustard groups was p < .001 (figure 4).

Rhythm disturbances. Major changes associated with junctional rhythm were reduced flow during the x descent with a corresponding increase during the y descent (figure 5). This was observed in eight patients, while in one, the two components were equal. In each there was an absent a wave through the tricuspid valve and in one reversed flow in the pulmonary vein during atrial contraction against a closed tricuspid valve could be detected. In those patients who varied between junctional and sinus rhythm, flow during the x descent increased in sinus rhythm and decreased on return to junctional rhythm. This was also observed in a patient who was atrially paced (figure 5).

Group III: PVO in patients who underwent Mustard repair. In two patients with mid baffle obstruction, a high-velocity (2.7 and 2.1 m/sec) turbulent systolic jet with diastolic spillover was identified just distal to the obstruction (figure 6). More distal in the pulmonary venous atrium, systolic and diastolic turbulence could be detected resembling that resulting from a patent ductus arteriosus (figure 6). Above the obstruction, the flow velocity decreased (30 cm/sec). Doppler interrogation in the pulmonary veins demonstrated loss of phasic flow (figure 6). Revision of the baffle was performed in both patients. In one, the orifices of the pulmonary veins were small and they were dilated at the time of surgery. Repeat Doppler assessment immediately after surgery demonstrated forward biphasic

FIGURE 2. A, The upper picture is a pulmonary venous atrial pressure trace from a patient who underwent the Mustard procedure. Note the deep y descent in comparison with the a to x descent. The lower picture is the spectral trace and likewise mirrors the pressure events. Flow occurs predominantly during the y descent, with lack of flow during the a to c phase and a later peak coinciding with the c to x descent. vs = flow during the x descent; vd = flow during the y descent. B, The upper picture is a pulmonary venous atrial pressure trace obtained after the Mustard procedure. The lower picture is the simultaneous venous Doppler spectral trace. Note that the flow mirrors the pressure events, with peak flow occurring during the x and y descent and minimal flow during the a and v waves. Flow occurs during both the a to c and c to x descents.
flow, with an adequate mid baffle area. Several weeks later the child developed pulmonary venous congestion and repeated Doppler assessment demonstrated a high-velocity turbulent pattern in the vicinity of the pulmonary veins.

In the other patient, reassessment immediately after surgery revealed significant stenosis, with turbulence detected throughout the pulmonary venous atrium.

Both of these children subsequently died and autopsy confirmed the presence of bilateral stenosis of the pulmonary vein. In one other patient who had previously undergone revision of mid baffle obstruction and received a patch to the left pulmonary veins a similar turbulent pattern was identified. The flow in the right veins in this patient consisted of low-velocity biphasic forward flow (figure 7). There were three other

**FIGURE 3.** Top, A spectral trace of pulmonary venous flow in an infant before Mustard procedure. The dominant peak occurs during the phase corresponding with the x descent of a left atrial pressure trace. Bottom, From the same patient immediately after Mustard procedure. Despite the similarity in heart rates, the profile has changed to that of dominant flow during the y descent. There is a variation in waveform from one cycle to another due to baffle movement secondary to respiration. vs = flow during the x descent; vd = flow during the y descent.
patients with documented abnormalities of the left-sided pulmonary veins. In one patient, flow could not be detected in the left-sided pulmonary veins, which corresponded with the finding of complete absence of perfusion of the left lung by radionuclide techniques. In the other two patients, both of whom had surgical procedures performed on the left-sided pulmonary veins at the time of revision for mid baffle obstruction, a perfusion scan demonstrated grossly reduced flow to that side. No evidence of turbulence was detected in the pulmonary venous atrium, but the individual pulmonary veins were not sampled during this examination.

At surgery the presence of stenosis of the left-sided veins was observed in the patient who was recognized before surgery as having abnormal left venous flow. The right-sided flow pattern demonstrated no evidence of turbulence, but in the left-sided veins there was increased flow velocity with turbulence. A lung perfusion scan showed that 83% of flow was to the right lung and only 17% was to the left lung.

Postoperative results in patients in the Senning group. Six patients in this group had mid baffle stenosis. Two were asymptomatic but had a definite pressure drop between the upper and lower portions of the pulmonary venous atrium. Both forward flow components of the Doppler trace were increased, with peak velocities between 1.4 and 1.6 m/sec and spectral broadening (figure 8). In the pulmonary veins, however, the velocity profile was biphasic (figure 8). One of these patients has undergone postoperative catheterization and the pulmonary angiogram demonstrated hold up of dye at the mid baffle level of the levophase. In two other patients with definite pulmonary hypertension, the narrowing was more severe. The flow distal to the obstruction peaked at 1.96 and 1.6 m/sec, with equal forward flow components and associated spectral

\[ \text{FIGURE 4. Graph showing the differences between the flow during the x and y descent, expressed as a ratio and corrected for heart rate, between normal subjects, patients who underwent Senning repair, and those who underwent Mustard procedure. The mean and two standard errors of the mean are shown.} \]

\[ \text{FIGURE 5. Top, A pulmonary venous atrial pressure trace obtained after the Mustard procedure while the patient was in junctional rhythm. Note the retrograde P wave. Middle, The Doppler spectral trace, which mirrors the pressure trace. The majority of forward flow occurs during the y descent. Bottom, From the same patient after atrial pacing. The pattern has changed to one of biphasic flow. vs = flow during the x descent; vd = flow during the y descent.} \]
broadening. In the fifth patient there was a dominant jet during the y descent that peaked at 2.25 m/sec. Distal to this the pattern was similar to that of the other patients. Catheterization 2 days after the Doppler assessment in this patient (under sedation) revealed a gradient of 15 mm Hg between the upper and lower limb of the pulmonary venous atrium. Sampling in the right and left pulmonary veins from the suprasternal position demonstrated loss of phasic flow with a continuous pattern and some respiratory variation. All

FIGURE 6. Top left, The subcostal cut obtained after Mustard repair with middle baffle obstruction of the pulmonary venous atrium. This is indicated by the two white arrows. The sample volume is just distal to the obstruction. Middle left, The Doppler spectral trace, demonstrating a high-velocity turbulent jet. Bottom left, When the sample volume is moved more distally into the pulmonary atrium, a high-velocity turbulent pattern not unlike that seen in patients with patent ductus arteriosus is demonstrated. Top right, The sample volume placed just proximal to the obstruction. Middle right, The Doppler spectral trace. Note that the velocity is reduced. Bottom right, The lack of phasic flow when sampling within the pulmonary vein. dias = diastole; pva = pulmonary venous atrium; sva = systemic venous atrium; sys = systole.
the ultrasound findings. In the sixth patient, the mid baffle narrowing was due to severe mitral regurgitation and subsequent dilatation of the systemic venous atrium. The flow in the pulmonary veins in this patient appeared normal, but when the sample volume was placed through the mid baffle area, an increased peak during the y descent with associated spectral broadening was evident.

Discussion

Interatrial repair by the Mustard or Senning procedure is still the most common operation performed in patients with transposition of the great arteries. Complications include systemic and pulmonary venous obstruction,7-10 arrhythmias, and right ventricular dysfunction.25 Noninvasive studies are frequently employed to evaluate arrhythmias, right ventricular function, and more recently, systemic venous obstruc-
Subcostal cross-sectional echocardiography has been used after the Senning procedure to evaluate the pulmonary venous pathway, with significant mid baffle obstruction having a distinct appearance from the pathway without obstruction or significant tricuspid regurgitation. This is of value in the younger patient in whom the whole limb of the baffle can be visualized, but in older patients it is not possible to use this approach because anatomical detail is less precise and no information about the flow characteristics is gathered.

Animal experiments and studies in humans suggest that pulmonary venous flow directly mirrors left atrial pressure events. Reversed flow, which is due to atrial contraction, is followed by forward flow coincident with passive atrial relaxation. This is succeeded by and often blends with a second peak of forward flow that results from active atrial relaxation as the atrioventricular valves descend. This corresponds with the a to c and c to x descent of the left atrial pressure curve. The third peak of forward flow occurs during rapid diastolic filling as the atrioventricular valve opens and corresponds with the y descent of the left atrial pressure curve.

Normal pulmonary venous flow patterns as assessed recently by pulsed Doppler echocardiography correspond with the above experimental data. Since these flow profiles clearly reflect left atrial events they can be used to assess alterations after baffle procedures. Normal flow is defined as an atrial contraction detected by forward flow through the tricuspid valve or reversed flow in the pulmonary veins followed by two forward peaks of virtually equal magnitude. The first of these represents atrial relaxation patterns a to c and c to x on the left atrial pressure trace, and the second the period of rapid diastolic filling.

The Doppler data from this study correspond fairly closely with hemodynamic information on patients who have undergone Mustard and Senning repairs. Apart from Silove and Taylor, few authors have examined the pulmonary venous waveform in detail. We showed that even when patients in whom flow profiles may have been compromised were not considered, pressure-flow characteristics in both patient groups differed from normals and those in the Mustard group differed from those in the Senning group. This suggests that in patients undergoing Mustard repair, the baffle prevents normal relaxation of the pulmonary venous atrium after atrial systole. Patients who undergo Senning repair appear to have a greater degree of relaxation of the atrium, but it is still abnormal.

Similarly, when a patient is reoperated on after the Senning procedure for mid baffle obstruction with enlargement of the pulmonary venous atrium by a patch, the velocity profile is similar to that of a patient in the Mustard group, with loss of dominant flow during the x descent.

These pulmonary venous flow patterns correspond with Doppler assessments of systemic venous flow after Mustard repair, which demonstrates dominant flow during the y descent and a smaller or absent peak during the x descent. In patients who undergo Senning repair the x descent component is greater but still abnormal. This likewise suggests reduced compliance of the systemic venous channel after both operations, which is more pronounced in the Mustard group.

With current surgical techniques the incidence of mid baffle obstruction after Mustard or Senning procedure has decreased. When obstruction is severe, pulmonary hypertension occurs, with a corresponding increase in left ventricular dimension. Doppler interrogation just distal to the obstruction is very specific, with a high-velocity systolic and/or diastolic jet and associated spectral broadening. Even if the sample volume is not placed at the site of obstruction, the turbulent pattern can be identified as far distal as the tricuspid valve. Proximal to the obstruction the pattern changes, with a loss of phasic flow that can also be seen in the pulmonary veins. What is apparent is that the upper limb of the baffle becomes tense with increased pressure and no longer appears to be under the influence of left atrial pressure events. The pattern of the pulmonary venous flow under these conditions resembles that documented by early experimental data, from which it was postulated that pulmonary venous flow was under the control of right ventricular pressure events.

In those patients who had undergone a Senning procedure and had less severe narrowing, the pulmonary vein velocity profile was normal. This indicates that although there is an element of narrowing, the upper limb of the baffle is still compliant and remains under the influence of left atrial pressure events. What is not yet known is whether this narrowing progresses to cause pulmonary venous hypertension or whether with time growth will occur.

Isolated stenosis of the pulmonary vein associated with transposition of the great arteries is a rare but serious disorder. It may be present before the Mustard procedure, or result from suture lines placed too close to the venous orifices. The left veins are more commonly affected, and the outcome is uniformly dismal if the stenosis is bilateral, even with surgical intervention. Postoperative pulsed Doppler is invaluable in
detecting this problem, especially when mid baffle obstruction masks venous stenosis before surgery.

Except in neonates and young infants, it is impossible to sample all four veins independently. What can be achieved is sampling of both sides of the pulmonary venous atrium to determine whether the stenosis is unilateral or bilateral. Stenosis of the pulmonary vein should not be diagnosed by increased peak velocity alone, particularly in patients with junctional rhythm or significant tricuspid regurgitation in whom the velocity of flow during the y descent may be increased. Spectral broadening, as in all of our patients, should also be present.

One potential limitation illustrated in three of our patients is that obstruction may not be recognized if the flow in the veins is absent or greatly reduced.

Pulsed Doppler echocardiography provides a reliable noninvasive technique for the assessment of pulmonary venous atrial pressure flow characteristics after a Mustard or Senning procedure. This technique can be used to detect the presence of mid baffle obstruction and isolated stenosis of the pulmonary vein and to monitor changes after surgery.

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