Doppler echocardiographic evaluation of pulmonary artery flow after modified Fontan operation: importance of atrial contraction

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Abstract
Doppler echocardiography was used to evaluate blood flow in the pulmonary artery in 14 patients 2 to 42 months (mean (SD) 17 (12) months) after a modified Fontan operation incorporating a direct atrio-pulmonary anastomosis. Preoperatively six patients had tricuspid atresia, six had a double inlet left ventricle, and two had pulmonary atresia with an intact ventricular septum. The postoperative rhythm was sinus in 11 patients, junctional in one, ventricular pacing in one, and atrioventricular sequential pacing in one. In one patient the Doppler trace was unsatisfactory for analysis. In all patients forward flow in the pulmonary artery had biphasic peaks related to both atrial and ventricular contraction. The mean (SD) peak flow velocity that was synchronous with atrial contraction was 80 (30) cm/s and that synchronous with ventricular contraction was 74 (23) cm/s. The atrial contribution to total pulmonary artery flow, assessed by velocity-time integrals, varied between 22% and 73% (mean (SD) 45 (14)%). In patients with tricuspid atresia the mean (SD) peak flow velocity with atrial contraction was 90 (27) cm/s and that with ventricular contraction was mean (SD) 68 (24) cm/s. In patients with double inlet left ventricle the mean (SD) peak flow velocity was 67 (36) cm/s with atrial contraction and 80 (25) cm/s with ventricular contraction. The atrial contribution to total pulmonary blood flow in patients with tricuspid atresia was significantly higher (53 (11)%) than in those with double inlet left ventricle (37 (14)%).

Pulmonary artery flow after modified Fontan operation was biphasic and was related to both atrial and ventricular contraction. The atrial contribution to pulmonary blood flow is greater in patients with tricuspid atresia than in those with a double inlet left ventricle. The mechanism of the second peak related to ventricular contraction is unknown.

Several technical modifications of the original Fontan operation have been used for physiological correction of patients with tricuspid atresia and double inlet left ventricle. These include the incorporation of a subpulmonary ventricular chamber, valved conduits between the right atrium and pulmonary artery, vena cava valves, and direct non-valved atrio-pulmonary anastomosis. There are conflicting reports on the importance of right atrial contraction, and therefore sinus rhythm, to the success of this operation.

We assessed pulmonary artery flow patterns by Doppler ultrasound and evaluated the contribution of atrial contraction to total pulmonary blood flow after a modified Fontan operation with a direct non-valved atrio-pulmonary anastomosis.

Patients and methods
We studied 14 (eight boys and six girls aged 55–175 months (mean (SD) 90 (35) months at operation) randomly selected patients who underwent successful modified Fontan operation between December 1983 and June 1987 at the Royal Liverpool Children's Hospital. The Doppler echocardiographic study was performed between 2 and 42 months (mean (SD) 17 (12) months) postoperatively and the age at the study varied from 60 to 184 months (107 (37) months).

Six patients had tricuspid atresia. Four of these had associated pulmonary or sub-pulmonary stenosis and one had an absent pulmonary valve with intact ventricular septum. All five patients with reduced pulmonary blood flow had undergone palliative systemic-pulmonary artery shunts.

The remaining patient had banding of the pulmonary trunk to reduce excessive pulmonary blood flow in infancy. Six patients had double inlet left ventricle. Two of the three with pulmonary or subpulmonary stenosis had been palliated with systemic-pulmonary artery shunts while three with unrestricted pulmonary blood flow had previous banding of the pulmonary trunk. Two patients had pulmonary atresia with intact ventricular septum. Both of these had undergone previous systemic-pulmonary artery shunts.

A direct posterior non-valved atrio-pulmonary anastomosis was constructed between the undivided superior vena cava, right atrium, and roof of the left atrium and the pulmonary arteries in 12 of the patients. In two patients an anterior anastomosis was constructed between the right atrial appendage and the pulmonary trunk/right pulmonary artery. In seven patients, the pulmonary trunk was divided and passed behind the aorta at the time of the repair, in five it was ligated, and in two

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Doppler echocardiography
Cross sectional and Doppler echocardiography was performed using an Irex Meridian system.

Results
In all the patients forward flow in the pulmonary artery had two peaks. Diastolic forward flow started at the second heart sound and was usually of low velocity. This was considered to be due to passive venous return into the pulmonary artery. Augmentation of the forward flow velocities then occurred; this began with the P wave, reached its peak between the P wave and the peak of the R wave, and returned to the baseline at the onset of the first heart sound. A second peak was seen during ventricular systole between the first and second heart sounds. In patients with tricuspid atresia the peak velocity after atrial contraction was higher than that after ventricular contraction; in contrast, in patients with double inlet left ventricle the pattern was reversed (fig 1A and B). In one patient with junctional rhythm the P wave was masked by the QRS complex. Peak forward flow in this patient occurred in early ventricular systole (due to atrial contraction) and a further peak occurred in ventricular diastole with passive venous filling (fig 2). In one patient with ventricular paced rhythm for postoperative complete heart block, there were, in addition to a dominant forward flow peak occurring with ventricular systole, separate peaks unrelated to ventricular systole and caused by atrial contraction (fig 3).

Peak flow velocity in the pulmonary artery synchronous with atrial contraction was
In one patient with pulmonary atresia and intact ventricular septum excision of the tricuspid valve and division of the pulmonary trunk were performed at the time of the modified Fontan operation to incorporate the hypoplastic right ventricle in the pulmonary circulation upstream to the atripulmonary anastomosis. Doppler study showed that the atrial contribution to the total pulmonary blood flow was 48\%, and the flow pattern was similar to that in the patients with tricuspid atresia. Thus in this patient the incorporation of the hypoplastic ventricle did not affect the pulmonary flow pattern.

In two patients a hypoplastic right ventricle remained in continuity with the pulmonary circulation. One of them had pulmonary atresia with intact ventricular septum and a Waterston shunt. A transannular right ventricular outflow patch had been used to decompress the right ventricle. The hypoplastic tricuspid valve was closed at the time of the Fontan operation. When the Doppler study was carried out he was in junctional rhythm. The second patient had tricuspid atresia with an intact ventricular septum and no pulmonary valve. In both patients there was free regurgitation into the hypoplastic right ventricle.

The first patient had forward flow into the pulmonary artery during systole as a result of the atrial contraction, while the remaining forward flow occurred in mid and late diastole due to passive venous filling (fig 2). In the second patient Doppler flow traces were similar to those seen in patients with tricuspid atresia, the atrial contribution to the pulmonary flow being 41\%. Thus the inclusion of a subpulmonary ventricular chamber as a reservoir distal to the atripulmonary anastomosis did not seem to affect the flow pattern in the pulmonary artery.

Two of the patients with double inlet left ventricle had atrial contributions to pulmonary blood flow of 49\% and 58\%. Both these patients had higher preoperative mean right atrial pressures (>7 mm Hg) than the remaining patients with double inlet left ventricle.

**Discussion**

It has generally been accepted that sinus rhythm, and thus atrial contraction, is important for the success of the Fontan operation or its modifications,\(^5\) although controversy continues as to whether the success is due to an improved left ventricular output or to a direct contribution of the right atrium to the pulmonary blood flow. The value of including a subpulmonary ventricular chamber in the repair is also controversial\(^6\) and data from some recent studies suggest that atrial contraction may not be essential.\(^4,10\) Indeed, in some complex defects, such as those in which the left atrioventricular valves are absent or hypoplastic or there are anomalies of venous drainage, total cavopulmonary connection has been proposed as an alternative to Fontan operation.\(^11\) Nevertheless, the original or modified Fontan operation will continue to be used in most patients with tricuspid atresia or double ventricle.
inlet left ventricle. To characterise the type of pulmonary artery blood flow after a direct non-valved atrio-pulmonary anastomosis we studied the Doppler flow patterns in these patients.

It is difficult to evaluate atrial function by a reliable technique. Doppler echocardiography gives a qualitative and possibly semi-quantitative assessment of atrial function, however, by measurement of the ratio of forward flow during atrial contraction to the total flow in the pulmonary artery. Nakazawa et al. suggested that the role of atrial contraction after the Fontan operation may be more important in those patients with a smaller pulmonary artery index, irrespective of whether they have tricuspid atresia or double inlet ventricle.10 Conversely, in patients with a large pulmonary artery index, the right heart output may be less dependent on atrial contraction. These conclusions were based on an invasive study with a catheter mounted Doppler velocity probe. Volume flow is usually calculated from the mean velocity of flow and cross sectional area of the vessel where the flow velocity is measured. After the Fontan operation it is difficult to measure the area of the atrio-pulmonary anastomosis accurately. Because absolute values of volume flow may not be reliable, both we and Nakazawa et al. have assumed this area to be constant throughout the cardiac cycle.10

We have further adapted the intracardiac Doppler methods of flow assessments to the more conventional transthoracic pulsed Doppler techniques.

Shemin and associates, in an experimental study, reported that the right atrium did not function as a pump in the maintenance of pulmonary blood flow after a right atrium to pulmonary artery conduit.11 Matsuda et al., in a study on dogs that had right atrium to pulmonary artery conduits with the right ventricle excluded from the pulmonary circulation, induced atrial fibrillation and found that Doppler echocardiography could be used to measure the systemic arterial pressure and pulmonary blood flow were maintained.6 With atrial fibrillation, however, pulsatile flow in the atrio-pulmonary conduit was lost. Nakazawa et al. evaluated seven patients after a Fontan operation for tricuspid atresia. They used pulsed Doppler echocardiography, cardiac catheterisation, and intracardiac Doppler with a catheter tipped velocity transducer.7 They found that the flow patterns obtained by pulsed Doppler echocardiography were biphasic and identical to those obtained with a catheter tipped velocity transducer and that atrial contraction contributed to the forward flow of blood in the pulmonary artery. In a subsequent report, patients with a double inlet left ventricle were studied for comparison and similar flow patterns were obtained.10 DiSesa et al. also confirmed that the pulmonary artery flow was biphasic after the Fontan operation both in patients with tricuspid atresia and in those with double inlet left ventricle.8 The flow, however, was not evaluated quantitatively or semi-quantitatively. Alboliras et al. suggested that synchronised atrial contraction was not necessary for adequate cardiac output, despite finding acceleration in pulmonary artery flow during atrial systole by Doppler.13 Biphasic flow patterns in the pulmonary artery were reported by Hagler et al.,14 but, interestingly, in patients with reduced ventricular function the predominant flow occurred during ventricular systole.

Our measurements showed important differences between the patients with tricuspid or pulmonary artery atresia and double inlet left ventricle. The previous reports showed that the pulmonary artery flow was biphasic and these phases were related to atrial contraction and passive venous filling. In our study the second phase was consistently related to ventricular systole, which was followed by non-pulsatile low velocity flow caused by passive venous filling. These differences have not been emphasised before. We found that right atrial contraction, while contributing significantly to the total pulmonary blood flow in both groups of our patients, made a greater contribution to the pulmonary flow in patients with tricuspid atresia than in those with double inlet left ventricle. This may have been because higher atrial pressures cause greater right atrial hyper trophy. However, the mechanism of the second phase of flow that was synchronous with ventricular systole is not clear. DiSessa et al. suggested that the second phase may have been the result either of right atrial relaxation and vena caval blood flowing into the pulmonary artery or filling of the left atrium from the pulmonary venous bed, which was in turn filled by flow from the pulmonary artery. However, our study showed that this flow occurred only during ventricular systole and was also pulsatile. This suggests "active" rather than "passive" flow into the pulmonary artery. In one of our patients with double inlet left ventricle, in whom ventricular function was impaired, the atrial contribution (58%) to total pulmonary blood flow was higher than the rest of this group, suggesting that Doppler echocardiography was useful in this study, it probably failed to detect slow and transient pulmonary regurgitation into the right atrium because of its very low velocity.

In conclusion, pulmonary artery flow after modified Fontan operation is biphasic. The atrial contribution to total pulmonary flow is greater in patients with tricuspid atresia than in those with a double inlet left ventricle. However, the importance of phasic pulmonary artery flow in the various subsets of patients and its long term effects on the pulmonary vasculature remain important questions for further investigation in a larger group of patients.

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VIEWS FROM THE PAST Louis N Katz

Louis N Katz (1897-1973) had a distinguished career as a physiologist in Carl Wiggers's department in Cleveland, Ohio. In 1924-25 he held a US National Research Council Fellowship at University College, London, in E H Starling's department, where he studied myocardial metabolism with A V Hill (the future Nobel laureate). The two months he spent with Thomas Lewis had a key influence, for after he became the founding director of the Michael Reese Cardiovascular Institute in Chicago in 1930 he made it into a renowned centre of electrocardiography. (Photograph provided by Dr Arnold M Katz.)

DENNIS M KRIKLER