

## Functional Assessment of the Fontan Operation: Combined M-Mode, Two-Dimensional and Doppler Echocardiographic Studies

DONALD J. HAGLER, MD, FACC, JAMES B. SEWARD, MD, FACC, A. JAMIL TAJIK, MD, FACC, DONALD G. RITTER, MD, FACC

Rochester, Minnesota

Combined M-mode, two-dimensional and Doppler echocardiographic studies were used to assess the postoperative status of 33 patients who had undergone the modified Fontan procedure. Twenty-four patients had surgical repair with use of a simple direct right atrium to pulmonary artery anastomosis. The remaining patients had repair with use of a prosthesis or associated Glenn shunt. Twenty-seven patients were studied early in the postoperative period (2 months or less) and the remaining patients were studied up to 6 years postoperatively. A total of 36 examinations were performed. Of the 33 patients, 13 had tricuspid atresia, 12 had double inlet left ventricle with hypoplastic right ventricular outlet chamber and 8 had complex lesions with atrioventricular canal, double outlet right ventricle or a hypoplastic ventricle.

Postoperative assessment by M-mode and two-dimensional echocardiography demonstrated normal or

mildly reduced ventricular function (ejection fraction > 40%) in 22 patients. In 24 patients, a "normal" flow pattern was observed in the pulmonary artery by pulsed Doppler echocardiography, with predominant diastolic flow and accentuation by atrial systole somewhat similar to the venous flow pattern observed in the superior vena cava. "Abnormal" flow patterns (disorganized systolic flow, absence of atrial waves and little or no increase with inspiration) were observed in nine patients with reduced ventricular function or residual shunt. Continuous wave Doppler study also demonstrated mild dynamic subaortic obstruction in two patients. Combined pulsed and continuous wave studies showed atrioventricular valve insufficiency in 10 patients. Follow-up studies revealed a satisfactory clinical course in most patients. Three patients died approximately 4 to 8 months after their Fontan operation.

The Fontan operation (1) has been used for the correction of various complex congenital cardiac defects. More than 260 modified Fontan procedures were performed at our institution from October 1976 through May 1983 (2-4). Although the overall results of the operation have been encouraging and most patients have had an improved clinical status, the long-term results have not been adequately assessed. Because of the complexity and variety of congenital heart lesions that have been treated with this operation, the postoperative status has been difficult to differentiate and categorize (4,5).

To provide a noninvasive repeatable form of long-term follow-up evaluation, we analyzed the two-dimensional, M-mode and Doppler echocardiographic examinations of 33 patients who had undergone the Fontan procedure and who

were consecutively studied in our echocardiography laboratory between August 1982 and October 1983.

### Methods

**Study patients.** The patients (13 female and 20 male) ranged in age from 3 to 33 years (median 10). Of these 33 patients, 27 were studied in the first 2 months after operation and the 6 remaining patients were studied up to 6 years after operation. Most patients were referred for study routinely during the early postoperative period. A total of 36 studies were performed (three patients had 2 studies each).

*Preoperatively*, 13 patients had tricuspid atresia with pulmonary stenosis or pulmonary artery band, 12 patients had double inlet left ventricle with hypoplastic right ventricular outlet chamber, 3 patients had hypoplastic right ventricular chamber (dextrotransposition of the great arteries, levotransposition of the great arteries and pulmonary atresia in 1 patient each), 1 patient had hypoplastic left ventricle and atrioventricular (AV) valve, 3 patients had complex forms of complete AV canal defect with common AV valve and

From the Division of Pediatric Cardiology and of Cardiovascular Diseases and Internal Medicine, Mayo Clinic and Mayo Foundation, Rochester, Minnesota. Manuscript received December 19, 1983; revised manuscript received April 23, 1984, accepted May 4, 1984.

Address for reprints: Donald J. Hagler, MD, Mayo Clinic, 200 First Street S.W., Rochester, Minnesota 55905.

1 patient had complex double outlet right ventricle with remote ventricular septal defect.

*Surgical correction with the modified Fontan procedure* was achieved in 24 patients by simple direct anastomosis of the right atrial appendage to the pulmonary artery with a pericardial augmentation. Notable exceptions included three patients with an associated Glenn shunt (right superior vena cava to right pulmonary artery), three patients with a right atrium to pulmonary artery graft (22 mm and 30 mm), one patient with direct anastomosis of the right atrial appendage to the right ventricular outflow tract, one patient with a 30 mm graft from the right atrium to the right ventricular outflow tract, two patients with a 22 mm valved right atrium to right ventricular outflow tract conduit and one patient with an intraatrial 25 mm nonvalved conduit from the inferior vena cava to the pulmonary artery.

*The patients' clinical course* was normal except in seven patients who had clinical evidence of right heart failure or persistent retention of fluid. One patient had clinically suspected conduit obstruction. Two patients died approximately 4 months postoperatively, apparently from pulmonary dysfunction or infection. One patient (Case 6, Table 1) died 8 months postoperatively with chronic congestive heart failure and ventricular failure. Eight patients subsequently had postoperative catheterization hemodynamic studies for comparison (Table 1).

**Echocardiography.** All of the patients were studied postoperatively with two-dimensional echocardiography. Patients were examined in the usual manner with a 90°

mechanical sector scanner with 3 and 5 MHz transducers (6). Real time images were recorded on 3/4 inch (1.9 cm) video cassettes, and stop frame images were subsequently photographed with 35 mm film.

During the two-dimensional study, attempts were made to obtain satisfactory short-axis M-mode recordings of the ventricular chamber to allow measurements of ventricular dimension and assessment of ejection fraction (7). If satisfactory M-mode recordings of ventricular short-axis dimensions could not be obtained, a subjective assessment of global ventricular function was used.

**Doppler recording.** All of the patients in this study had echocardiographic examination with a commercially available pulsed Doppler system (American Technology Laboratory). Doppler recordings were obtained with the two-dimensional image to place the sample volume with the specific chamber to be examined. Routine recordings were obtained to assess AV and semilunar valve function, and recordings from the right atrium and main pulmonary artery were used to assess pulmonary flow characteristics. When possible, recordings were obtained from the superior vena cava for analysis and comparison with the pulmonary flow pattern. Continuous wave Doppler studies were available in the more recent examinations. For these studies, a continuous wave Doppler system with two-dimensional echocardiographic direction or an isolated Doppler (Pedof; Irex System III B) probe was used. The continuous wave Doppler system was primarily used to assess ventricular outflow tract obstruction and AV valve function (8).

**Table 1.** Correlation of Echocardiographic and Postoperative Cardiac Catheterization Data in Eight Patients

Case	Pressures (mean) (mm Hg)			CI (liters/min per m <sup>2</sup> )	EF (%)		AV Valve Insufficiency	Shunt	Pulmonary Flow (Doppler)
	RA	PA	Ventricle		By Angio	By Echo			
1	24/21 (22)	26/21 (23)	103/10 to 15	0.9	17	↓↓↓	Mild	None	Abnormal (↓)
2	27/18 (22)	22/13 (18)	108/5 to 12	2.8	56	Normal	Mild	None	Systolic flow*
3	27/22 (24)	30/20 (23)	97/16 to 24	2.6	20	↓↓↓	Mild	None	Abnormal (↓)
4	16/4 (7)	16/4 (8)	93/4	3.2†	N/A	48	None	None	a waves
5	20/16 (18)	19/14 (16)	120/9 to 16	2.1	48	48	None	Tiny residual VSD	Abnormal (↑)
6	28/23 (26)	28/22 (25)	105/12 to 17	1.9	21	21	None	None	Abnormal (↓)
7	22/15 (18)	22/15 (18)	125/6 to 14	3.6	60	Normal	None	None	a waves
8	20/16 (19)	18/14 (17)	111/10 to 19	2.5	40	37	Moderate	None	Abnormal (↓)

\*Patient had a right atrium to right ventricular outflow tract conduit with mild obstruction. †Catheterization data obtained through the courtesy of Donald Girod, MD, Indianapolis, Indiana. Angio = angiocardiology; AV = atrioventricular; CI = cardiac index; Echo = echocardiography; EF = ejection fraction; N/A = not available; PA = pulmonary artery; RA = right atrium; VSD = ventricular septal defect; ↓ ↓ ↓ = severely decreased; ↑ = increased flow; ↓ = decreased flow.

## Results

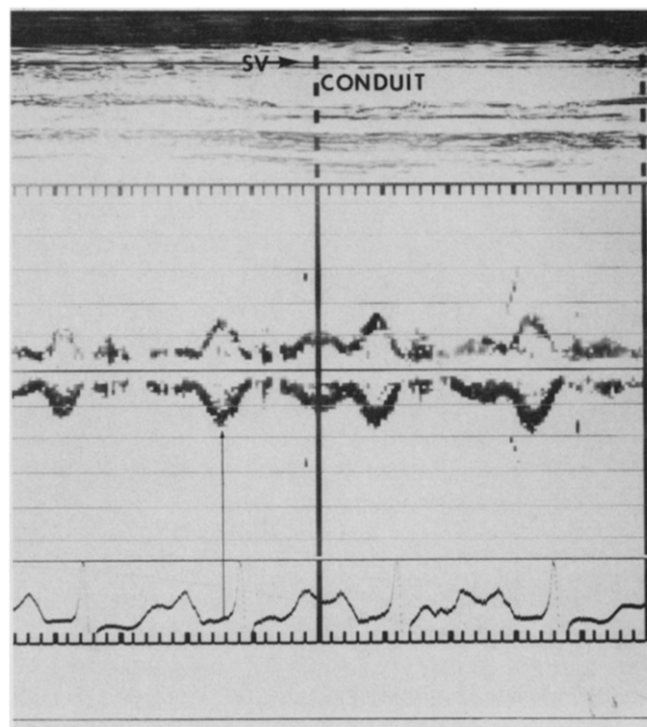
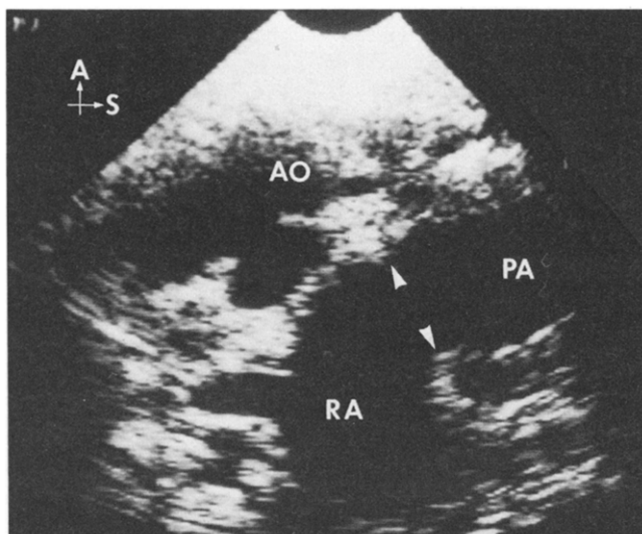
### Two-dimensional and M-mode echocardiography.

Functionally, all but three patients had a single ventricular chamber. In these three, a complete AV canal, AV and ventriculoatrial discordance with hypoplasia of the morphologic right ventricle and a complex double outlet right ventricle were found. In all patients, the right atrial patch appeared to be well attached without abnormal motion. Direct anastomosis of the right atrium to the pulmonary artery could be clearly defined from upper parasternal long- and short-axis scans (Fig. 1). In all of the patients, a widely patent anastomosis was noted. In patients with a right atrium to pulmonary artery conduit, frequently only the distal conduit and pulmonary artery anastomosis could be clearly visualized. In addition, in patients with a right atrium to right ventricular conduit, although the right ventricular outflow tract could be demonstrated, the proximal conduit and valve could not be demonstrated echocardiographically because of their retrosternal course.

Normal or mildly reduced ventricular function or left ventricular ejection fraction ( $> 40\%$ ) was demonstrated by M-mode echocardiography in 22 patients. In eight patients, the ventricular function was moderately or severely reduced. Three patients had a small posterior pericardial effusion.

**Doppler echocardiography.** Doppler echocardiographic examinations were primarily obtained for pulsed Doppler assessment of the venous flow patterns within the pulmonary

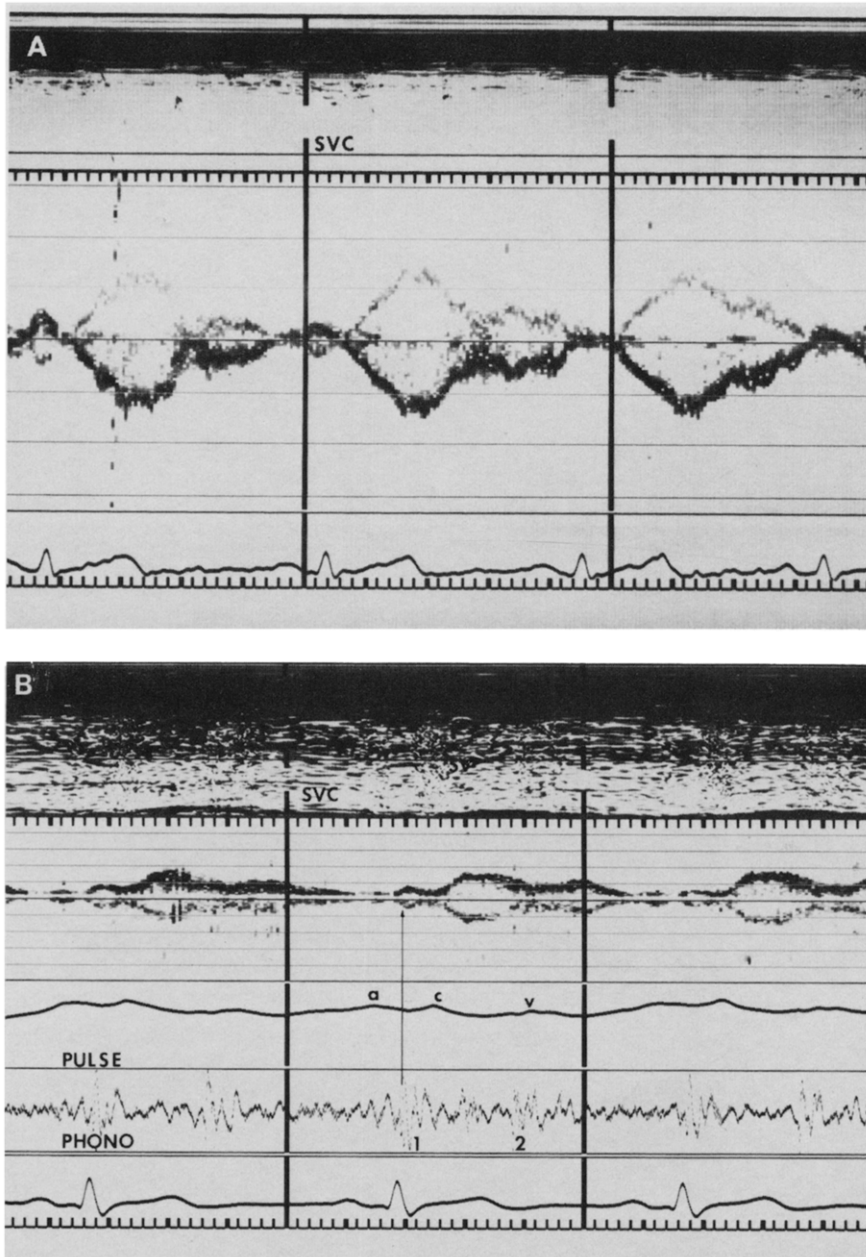
**Figure 1.** Two-dimensional echocardiogram in a 13 year old boy after a Fontan procedure with a direct right atrium (RA) to pulmonary artery (PA) anastomosis. The upper parasternal long-axis scan demonstrates a widely patent right atrium to pulmonary artery anastomosis (arrowheads). Rightward transducer tilt was occasionally necessary for demonstration. A = anterior; AO = aorta; S = superior.



**Figure 2.** Pulsed Doppler recording from the distal conduit in a 15 year old patient approximately 5 days after the Fontan procedure. Note the diastolic flow with prominent atrial waves (thin arrow). SV = sample volume.

artery, right atrium and superior vena cava. The results described reflect our attempt to qualitatively assess flow patterns. The pulsed Doppler recordings were not used to determine actual flow velocities. For pulmonary artery flow, maximal gain and low filter settings were used to maximize Doppler recordings. These settings were particularly necessary when very low flow was present and they allowed interpatient comparison of pulmonary flow patterns. Our initial pulsed Doppler studies demonstrated a venous flow pattern somewhat similar to that normally observed in the superior vena cava (Fig. 2 and 3). The most commonly observed flow pattern in the pulmonary artery was a late systolic and diastolic flow with a prominent *a* wave or accentuation of flow related to atrial systole. For better timing of these events, we obtained simultaneous jugular venous pulse tracings and phonocardiograms. These recordings confirmed the predominant diastolic flow pattern with accentuated flow related to atrial systole (Fig. 4). Similar venous flow patterns were recorded in the superior vena cava; however, systolic flow was more prominent than that observed in the pulmonary artery.

The Doppler flow observed within the systemic venous atrium was generally reduced in amplitude. A larger chamber size and lack of parallel flow orientation could explain the observed flow patterns within the right atrium.



**Figure 3.** A, Pulsed Doppler recording from the superior vena cava (SVC) in normal male subject. Note the triphasic appearance with late systolic, early diastolic and small atrial waves. B, Simultaneous jugular venous pulse tracing (PULSE) and phonocardiogram (PHONO) confirm timing of triphasic flow pattern. a = a wave; c = c wave; v = v wave; 1 and 2 = first and second sounds in the phonocardiogram.

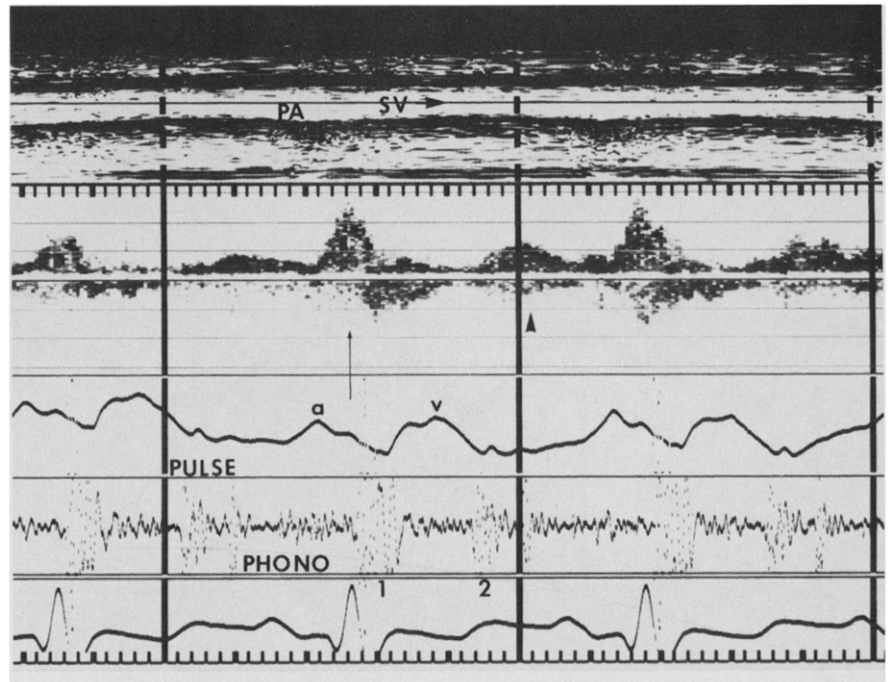
In none of the patients was turbulent flow present to suggest residual atrial shunt. In one patient, satisfactory Doppler flow recordings could not be obtained from the right atrium or the pulmonary artery, and markedly reduced ventricular function was demonstrated by two-dimensional echocardiography.

*'Normal' pulmonary artery flow pattern.* In 24 patients, pulsed Doppler examination of the pulmonary artery demonstrated a venous type flow pattern with late systolic and early diastolic flow and atrial waves concurrent with

atrial systole. In addition, inspiration resulted in a significant increase in the early diastolic and atrial waves (Fig. 5).

*Abnormal pulmonary artery flow pattern.* An abnormal pulmonary artery flow pattern was most frequently associated with reduced ventricular function. In five patients, the pulmonary artery Doppler flow pattern was reduced and occurred in systole, but small atrial waves were present. Two of these patients later had further deterioration in ventricular function, and the atrial waves were then absent. In four patients with markedly reduced ventricular function, a

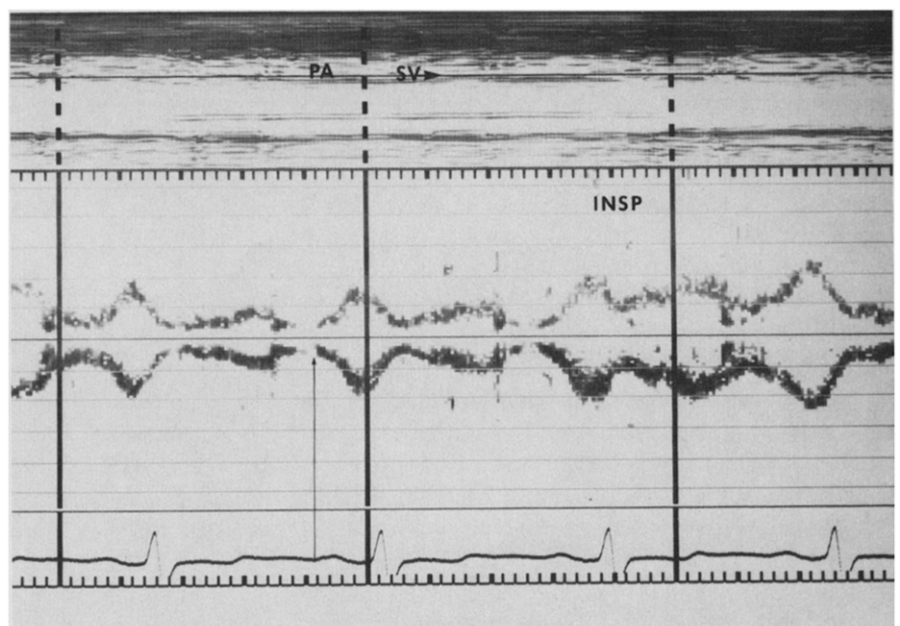
**Figure 4.** Simultaneous pulsed Doppler recording from the pulmonary artery (PA), jugular venous pulse tracing (PULSE) and phonocardiogram (PHONO) in a 20 year old patient approximately 3 weeks after a Fontan procedure and mitral valve replacement (4 mm Starr-Edwards prosthesis). Note the diastolic flow (**arrowhead**) with atrial (*a*) waves (**thin arrow**) preceding the first heart sound (1) on the phonocardiogram; diastolic flow (**arrowhead**) begins before the mitral valve opening sound. Abbreviations as before.

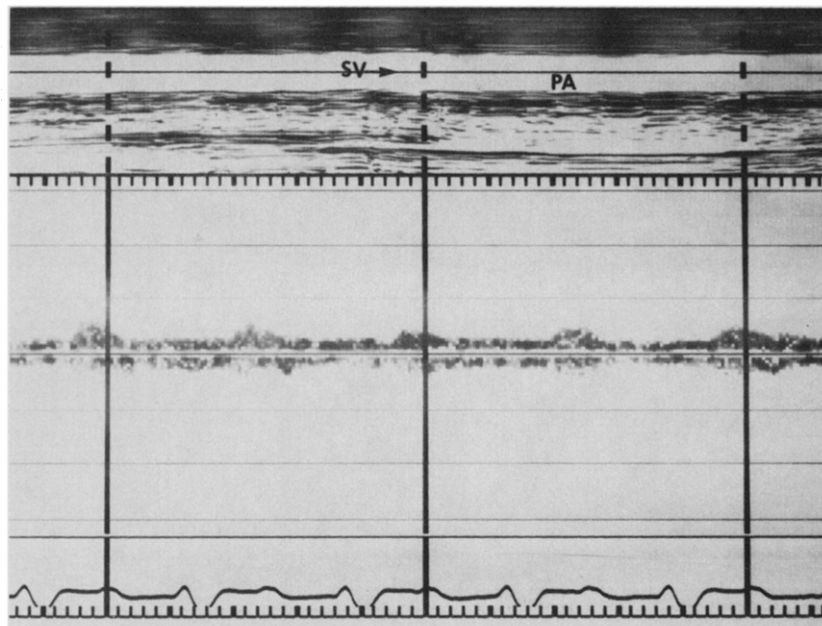


disorganized (predominantly systolic) flow with reduced amplitude and absent atrial waves was observed (Fig. 6). Also, the normal respiratory increase in flow velocity was absent or reduced in these patients (Fig. 7). On follow-up examination, two patients were noted to have further deterioration of ventricular function (ejection fraction was 21 and 39%, respectively) in association with disorganized (systolic) pulmonary artery Doppler flow patterns with absent *a* waves.

*Flow in the conduit.* In four patients with a right atrium to right ventricular outflow tract conduit, prominent systolic flow associated with ventricular contraction was observed (Fig. 8). In two patients, a high velocity jet was recorded, consistent with a residual ventricular septal defect. One patient with a 22 mm right atrium to right ventricular outflow tract valved conduit was studied approximately 6 years after operation to evaluate possible conduit obstruction. At this examination, prominent atrial waves were observed within

**Figure 5.** Pulsed Doppler recording from the pulmonary artery in 17 year old patient 10 days after a Fontan procedure, showing typical flow pattern with prominent increase in flow with inspiration (INSP). **Arrow** correlates the *a* wave after the *P* wave of electrocardiogram. Abbreviations as before.





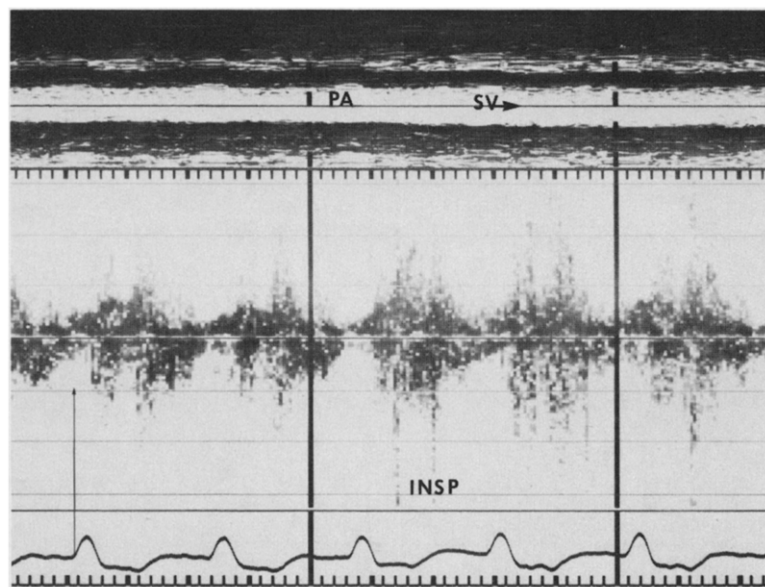
**Figure 6.** Pulsed Doppler recording from the pulmonary artery in a 13 year old patient approximately 3 weeks after a Fontan procedure. Note the markedly reduced flow pattern with systolic flow. Echocardiographic and angiographic evidence of severe decrease in left ventricular function and moderate AV valve insufficiency was also found. Abbreviations as before.

the conduit and reversed atrial waves in the inferior vena cava, suggesting conduit obstruction (Fig. 9A and B). However, further hemodynamic documentation is not available.

*Atrioventricular valve insufficiency.* Occasionally, AV valve insufficiency is observed in patients with tricuspid atresia or univentricular heart. It is even more frequent in patients with common AV valve. In 10 patients, AV valve insufficiency was demonstrated postoperatively with pulsed or continuous wave Doppler studies. In these patients, prominent systolic flow away from the sample volume and transducer was observed. Continuous wave Doppler recordings

confirmed the high velocity and direction of the jet resulting from AV valve insufficiency (Fig. 10). One patient had had replacement of the left AV valve; in this patient, the Doppler flow pattern within the left atrium was normal.

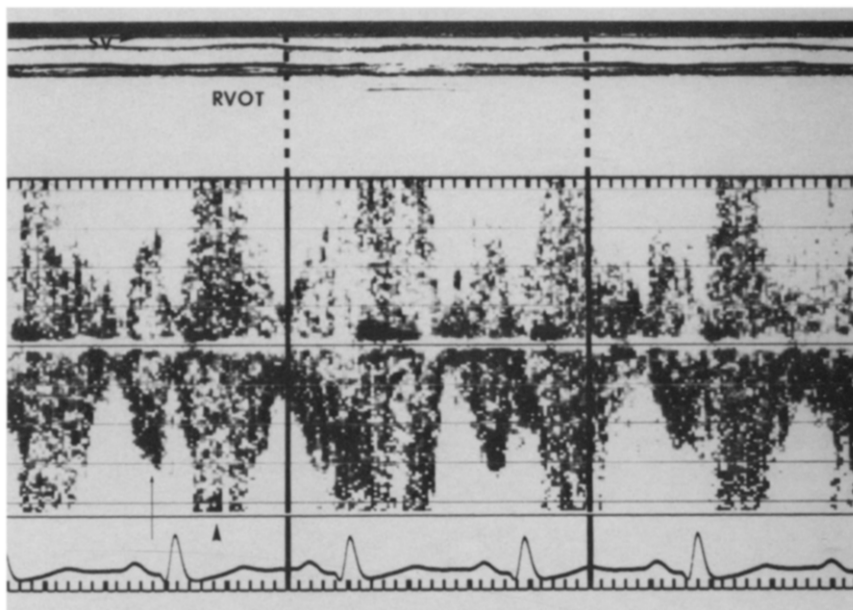
*Subaortic obstruction.* Continuous wave Doppler and two-dimensional echocardiography also suggested mild dynamic subaortic obstruction in two patients with univentricular heart and subvalvular pulmonary stenosis. Velocities of 2.0 and 2.3 m/s, respectively, were recorded in the ascending aorta, consistent with mild (approximately 16 to 20 mm Hg) pressure gradients. A grade 3/6 systolic ejection murmur was



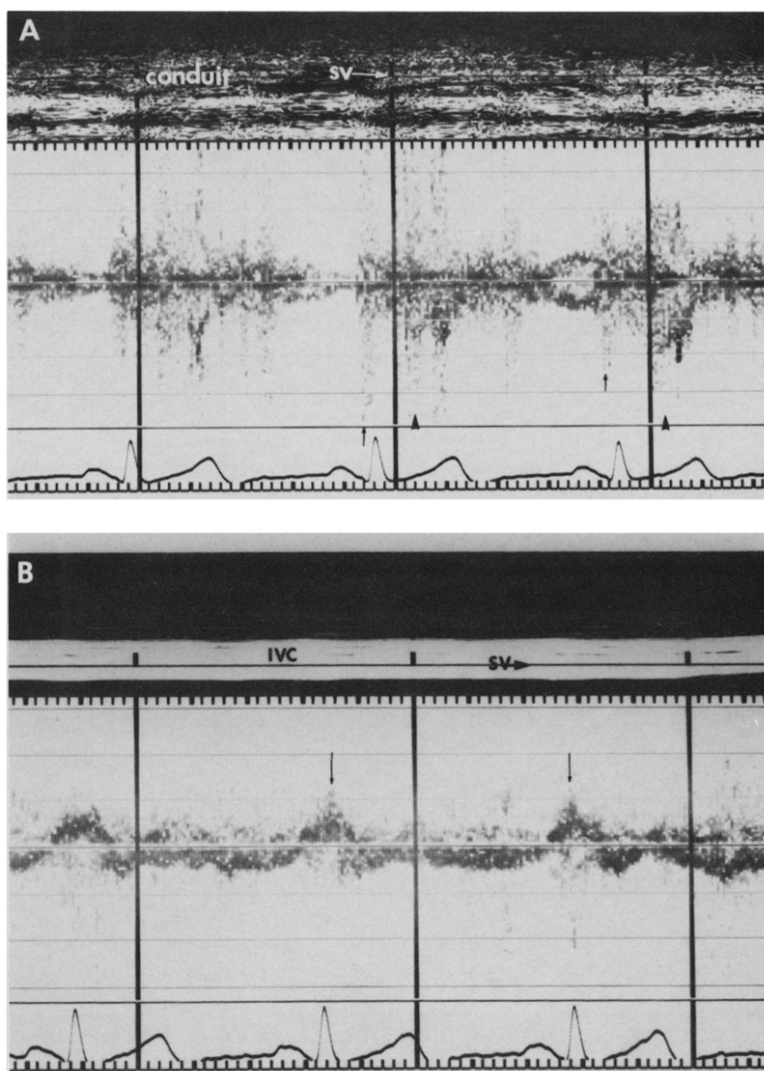
**Figure 7.** Pulsed Doppler recording from the pulmonary artery in a 14 year old patient with common atrium and common AV valve 8 days after a Fontan procedure. Poor ventricular function and AV valve insufficiency were noted at catheterization. Note the systolic flow, loss of *a* waves and little increase in flow with inspiration. Abbreviations as before.

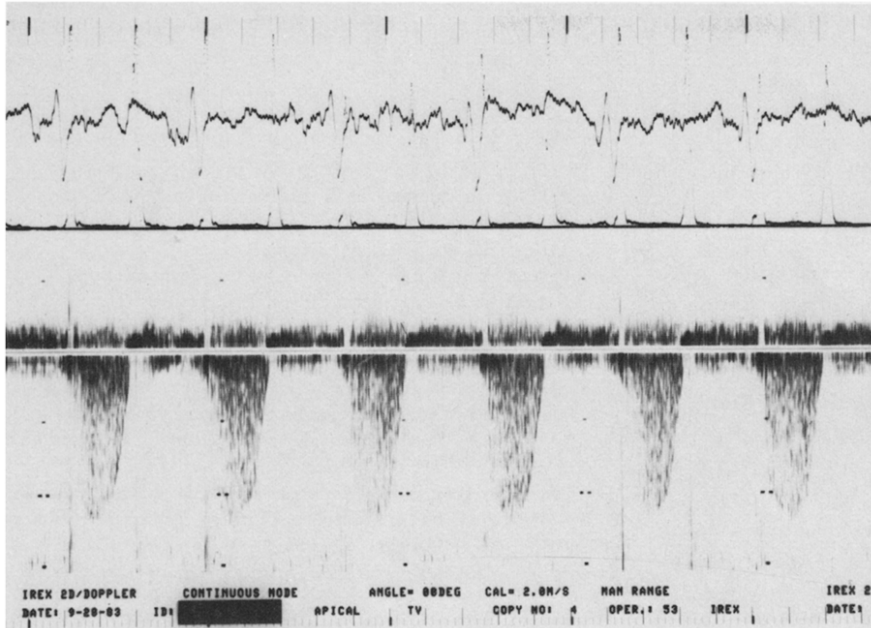


**Figure 8.** Pulsed Doppler recording from the right ventricular outflow tract in a 3 year old patient 2 months after a Fontan procedure with right atrium to right ventricular outflow tract anastomosis. Note the prominent systolic flow (**arrowhead**) due to residual ventricular septal defect. Also note prominent atrial waves (**thin arrow**). RVOT = right ventricular outflow tract; SV = sample volume.



**Figure 9. A,** Pulsed Doppler recording 6 years post-operatively in a 13 year old patient with a 22 mm right atrium to right ventricular outflow tract valved conduit. Note prominent atrial waves with aliasing (**thin arrows**) and systolic conduit flow (**arrowheads**). **B,** Pulsed Doppler recording from the inferior vena cava (IVC) in the same patient, demonstrating retrograde atrial flow into the inferior vena cava (**thin arrows**). SV = sample volume.





**Figure 10.** Continuous wave Doppler recording in a patient with double inlet left ventricle and tricuspid valve insufficiency, demonstrating high velocity regurgitant jet consistent with AV valve insufficiency.

detected clinically in the patients, and pulsed Doppler recordings localized the high velocity jet to the ascending aorta.

**Follow-up studies.** Clinical follow-up in most patients demonstrated a satisfactory course with clinical improvement. An initial clinical incidence of right heart failure or fluid retention did not always correlate with abnormal echocardiographic observations. The condition of most patients slowly improved with medical management. Three patients died late postoperatively. One patient with Down's syndrome had improved cardiac function and normal echocardiographic Doppler findings postoperatively. He subsequently had recurrent pulmonary infections that necessitated tracheostomy and he died approximately 4 months postoperatively. Another patient died approximately 4 months after operation, reportedly of pulmonary infection. His postoperative Doppler studies demonstrated reduced flow with small atrial waves and predominant systolic flow. In one patient (Case 6), reduced pulmonary artery Doppler flow and moderate to severe reduction in ventricular function were noted late postoperatively. Cardiac catheterization demonstrated severely decreased ventricular function. He subsequently died 8 months postoperatively despite medical management.

*Postoperative cardiac catheterization (Table 1).* Eight patients had postoperative cardiac catheterization. Two patients (Cases 1 and 3) with common AV valve were observed to have severe ventricular dysfunction and AV valve insufficiency by angiography. One other patient (Case 6) had markedly reduced ventricular function by angiography. The

findings in these studies were predicted by the two-dimensional and Doppler studies. One patient (Case 2) had mild (4 to 6 mm Hg pressure gradient at rest) conduit obstruction in a 22 mm right atrium to right ventricular outflow tract valved conduit. One patient (Case 5) with a right atrium to right ventricular outflow tract anastomosis had a small residual ventricular septal defect that had been predicted by pulsed Doppler echocardiographic demonstration of a high velocity jet in the right ventricular outflow tract. Two patients (Cases 4 and 7) had normal results of hemodynamic and angiographic studies. A "normal" pulmonary artery Doppler flow pattern had been observed with predominant diastolic flow and atrial accentuation.

## Discussion

In this study, combined M-mode, two-dimensional and Doppler echocardiographic studies provided valuable information for the postoperative noninvasive assessment of patients who had undergone the Fontan procedure (right atrium to pulmonary artery or right ventricular shunt or graft). Predominant diastolic flow with atrial contraction waves was observed in patients with a satisfactory clinical course. Notably different observations were made in three patients with angiographic evidence of reduced ventricular function. In these patients, a disorganized flow pattern with markedly reduced flow, systolic flow and absence of atrial waves was observed with pulsed Doppler recordings of the pulmonary artery.



Combined two-dimensional and pulsed Doppler studies were used in this study because they permit accurate localization of flow patterns (range resolution) and have greater sensitivity in the detection of low velocity flow with atrial contraction. The added use of continuous wave Doppler assessment can permit accurate detection of atrioventricular (AV) valve insufficiency and ventricular outflow tract obstruction. As used in this study, M-mode and two-dimensional echocardiography provided anatomic correlation by demonstration of right atrium to pulmonary artery anastomosis and the pulmonary artery size. Ventricular function may be more accurately assessed with computerized analysis of two-dimensional echocardiographic studies. In this study, a subjective evaluation and M-mode correlation provided a coarse assessment of ventricular function. However, a reasonable correlation with clinical and angiographic observations was noted. Further postoperative studies with hemodynamic and angiographic correlations of late postoperative results will be important for assessment of the results of modified Fontan correction.

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