Conclusion

Origin of LSCA from the AA can be explained by Edwards' double arch hypothesis.³ The right arch has fully developed. In addition, there is persistence of a part of the left fourth aortic arch and seventh left ISA in tandem forming the LSCA.

References

1. Hastreiter AR, D'Cruz IA, Cantez T, Namin EP, Licata R. Right-sided aorta. I. Occurrence of right aortic arch in various types of congenital

- heart disease. II. Right aortic arch, right descending aorta, and associated anomalies. *Br Heart J*. 1966;28:722-39.
2. Velasquez G, Nath PH, Castaneda-Zuniga WR, Amplatz K, Formanek A. Aberrant left subclavian artery in tetralogy of Fallot. *Am J Cardiol*. 1980;45:811-8.
3. Edwards JE. Anomalies of the aortic arch system. *Birth Defects*. 1977;13:47-63.
4. Gross R, Ware P. The surgical significance of aortic arch anomalies. *Surg Gynecol Obstet*. 1946;83:435-48.
5. Higashimo SM, Ruttenberg HD. Double aortic arch associated with complete transposition of great vessels. *Br Heart J*. 1968;30:579-81.

Intermediate outcomes of fenestrated Fontan procedures

S. Garekar, MD,^a H. L. Walters, III, MD,^b R. E. Delius, MD,^b R. L. Thomas, PhD,^c and R. D. Ross, MD,^a Detroit, Mich

The fenestrated Fontan (FF) procedure is associated with better hemodynamics postoperatively than the modified Fontan procedure without a fenestration.^{1,2} Although a fenestration improves cardiac output, there is a cost of right-to-left shunting, cyanosis, and the subsequent risk of thromboembolism.

Our goal was to evaluate our experience with FF procedures. Specifically, we sought (1) to assess the outcomes of closed fenestrations, (2) to determine the rate of spontaneously closed fenestrations, and (3), among patients with patent fenestrations, to determine whether a clinical or laboratory variable could be used to predict their outcomes.

After institutional review board approval, 23 FF procedures with no other source of significant right-to-left shunt were identified. The cardiac lesion in the majority of patients (16/23 [70%]) was hypoplastic left heart syndrome.

Pre-Fontan Catheterization Hemodynamics

The average mean pulmonary artery pressure was 11 ± 2.8 mm Hg. The mean pulmonary vascular resistance (PVR) was 2.3 ± 1.5 indexed Wood units. The mean systemic ventricular end-diastolic pressure was 7.8 ± 2.6 mm Hg.

Surgical Results

The mean age at the time of the operation was 24.7 ± 9 months. Twenty-two of the 23 patients received the lateral tunnel Fontan procedure, and the remaining patient received an extracardiac conduit. The fenestration was a single 4-mm punch in 14 patients. Additional procedures performed along with the Fontan procedure were pulmonary artery patch angioplasty ($n = 4$) and atrioventricular (AV) valvuloplasty ($n = 3$). The mean hospital stay after surgical intervention was 13.4 ± 7.4 days. The median follow-up period after the Fontan procedure was 16 months (range, 2-134 months).

Spontaneously Closed Fenestrations

There was spontaneous closure of the fenestration in 5 (22%) patients (Table 1). One of these did not tolerate this closure and has had a transplantation. The other 4 patients are clinically stable. The size of the fenestrations varied from 2 punches of 2.5 mm ($n = 2$), a single 4-mm punch ($n = 2$), and a single 5-mm punch ($n = 1$). There was no correlation between the size of fenestration created and the likelihood of closure.

Catheterization Laboratory Closure of Fenestrations

Data on catheterization laboratory closure of fenestrations are shown in Table 1. Three patients were referred to the catheterization laboratory for device closure of the fenestration, of which 2 underwent device placement after favorable hemodynamics were demonstrated on test occlusion of the fenestration. The patients were referred for device closure because of desaturation in 2 patients and persistent headaches in the third. There were no infarcts seen on brain imaging in the patient with headaches, and the headaches have persisted after device closure.

Patent Fenestrations

There were 20 patients with complete data, of whom 14 had patent fenestrations. The cohort was divided into 3 groups on the basis of fenestration status on the most recent echocardiogram: group 1,

From the Division of Cardiology, Department of Pediatrics,^a the Division of Pediatric Cardiovascular Surgery,^b and the Department of Pediatrics,^c Children's Hospital of Michigan, Detroit, Mich.

Received for publication Aug 1, 2005; revisions accepted Sept 2, 2005; accepted for publication Sept 8, 2005.

Address for reprints: Swati Garekar, MD, Pediatric Cardiology, Children's Hospital of Michigan, 3901 Beaubien Blvd, Detroit, MI 48201 (E-mail: swatigar@gmail.com).

J Thorac Cardiovasc Surg 2006;131:247-9

0022-5223/\$32.00

Copyright © 2006 by The American Association for Thoracic Surgery

doi:10.1016/j.jtcvs.2005.09.004

TABLE 1. Patients with spontaneous or device closure of fenestrations

Patient no.	Lesion	Pre-Fontan MAP/PVR	Time to closure from surgical date	Clinical course after closure
Spontaneous closure of fenestrations (n = 5)				
8	HLHS	13 mm Hg/4.8 WU	12 mo	Experienced protein-losing enteropathy. A fenestration was recreated in the catheterization laboratory. Worsened clinically. Listed and received a heart transplant.
10	Pulmonary atresia/IVS	14 mm Hg/3.3 WU	9 mo	NYHA class I
16	HLHS	16 mm Hg/5.4 WU	83 mo	NYHA class I
19	HLHS	9 mm Hg/3 WU	11 mo	NYHA class I
23	Complex single ventricle	9 mm Hg/3.4 WU	2 mo	NYHA class I
Device closure of fenestrations (n = 2)				
18	HLHS	10 mm Hg/5.6 WU	15 mo	Experienced frank congestive heart failure within 2 wk of closure. Managed with diuretics; currently stable NYHA class II.
21	DORV with mitral atresia	9 mm Hg/1.4 WU	11 mo	Currently stable NYHA class I

MAP, Mean pulmonary artery pressure; PVR, pulmonary vascular resistance; HLHS, hypoplastic left heart syndrome; WU, indexed Woods units; IVS, intact ventricular septum; NYHA, New York Heart Association; DORV, double-outlet right ventricle.

TABLE 2. Comparison of the 3 groups (data missing on 3 patients)

	Group 1: closed fenestration (n = 7)	Group 2: patent fenestration and saturation <94% (n = 9)	Group 3: patent fenestration and saturation ≥94% (n = 4)
Age (mo) at FF (mean ± SD)	25.6 ± 8.3	20.4 ± 2.5	33.7 ± 17.7
Pre-FF: pulmonary pressure (mm Hg)	11.4 ± 2.9	11.2 ± 2.7	9 ± 3.5
Pre-FF: pulmonary vascular resistance (indexed Woods units)	3.8 ± 1.6	1.7 ± 0.8	1.0 ± 0.2
Type of fenestration			
Single punch			
4 mm	n = 3	n = 7	n = 3
5 mm	n = 2	n = 1	n = 0
Two punches	2.5 mm × 2: n = 2	3.5 mm × 2: n = 1	3.5 mm × 2: n = 1
Pre-FF: end-diastolic pressure (mm Hg)	7.1 ± 2.5	8.2 ± 2.2	8 ± 4.4
Post-FF hospital stay	17.7 ± 9.2	10.3 ± 4	10.2 ± 6.6
≥Moderate AV valve regurgitation at recent visit, n	1 (14%)	4 (44%)	0 (0%)
≥Mild decreased ventricular function at recent visit, n	2 (29%)	4 (44%)	0 (0%)
NYHA class ≥2 at recent visit, n	3 (43%)	3 (33%)	0 (0%)
Hepatomegaly at recent visit, n	5 (71%)	5 (56%)	1 (25%)
Transplantation, n	1 (14%)	1 (11%)	0 (0%)

FF, Fenestrated Fontan procedure; SD, standard deviation; AV, atrioventricular; NYHA, New York Heart Association.

closed fenestration; group 2, patent fenestration and low resting saturation (<94%); and group 3, patent fenestration but normal resting saturation (≥94%).

Comparison Between Groups

Data on comparison between groups are shown in Table 2. There was no difference between the groups with respect to age at the time of the Fontan procedure, the pre-Fontan mean pulmonary artery pressure, or the systemic ventricular end-diastolic pressure. The preoperative PVR was significantly lower in group 3 ($P < .05$). No group 3 patients had significant AV valve regurgitation or ventricular dysfunction at the most recent clinic visit. Patients in groups 1 and 2 had a higher incidence of moderate or severe

decrease of ventricular function, New York Heart Association class 2 or greater, and hepatomegaly at the most recent clinic visit when compared with group 3.

Discussion

Historically, the interval between the FF procedure and device closure of the fenestration has ranged from less than a week to 3 years postoperatively.^{3,4} The trend has been to wait at least 6 months before contemplating closure. Waiting for a longer period exposes the patient to potential adverse effects of cyanosis and the risk of thromboembolism. No patient in this study had clinically detectable thromboembolism. Our data show that spontaneous

closure can occur as early as 2 months and up to 83 months after the operation. Closure of fenestration was not necessarily associated with a good outcome.

There are some proponents of the view that fenestrations should not be closed because they preserve cardiac output and prevent central venous pressure from increasing. This seems like a good option for the patients in our study with patent fenestrations whose resting peripheral arterial saturations were normal. The patients in this group had no significant AV leakage or ventricular dysfunction and were doing well clinically at the most recent clinic visit. These patients might have done well in part because of a lower pre-Fontan PVR. Whether patients with patent fenestrations but low resting saturations would benefit from device closure requires further study; however, those within this group who have preserved ventricular function

and minimal AV regurgitation might be good candidates for this procedure.

References

1. Lemler MS, Scott WA, Leonard SR, Stromberg D, Ramaciotti C. Fenestration improves clinical outcome of the Fontan procedure: a prospective, randomized study. *Circulation*. 2002;105:207-12.
2. Bridges ND, Mayer JE Jr, Lock JE, Jonas RA, Hanley FL, Keane JF, et al. Effect of baffle fenestration on outcome of the modified Fontan operation. *Circulation*. 1992;86:1762-9.
3. Goff DA, Blume ED, Gauvreau K, Mayer JE, Lock JE, Jenkins KJ. Clinical outcome of fenestrated Fontan patients after closure: the first 10 years. *Circulation*. 2000;102:2094-9.
4. Lloyd TR, Rydberg A, Ludomirsky A, Teien DE, Shim D, Beekman RH 3rd, Mosca RS, et al. Late fenestration closure in the hypoplastic left heart syndrome: comparison of hemodynamic changes. *Am Heart J*. 1998;136:302-6.