Catheterization and Cardiovascular Interventions 75:60–65 (2010)

PEDIATRIC AND CONGENITAL HEART DISEASE

Original Studies

Percutaneous Management of A Fontan Fenestration: In Search for the Ideal Restriction—Occlusion Device

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Objective: Most devices devices available for percutaneous closure of Fontan fenestrations tend to be bulky. The aim of this study was to evaluate a low profile custom made device and assess its efficacy and safety. **Patients and Methods:** A 15 mm PFO star was used as the basis. The following modifications were made: removal of the left disc to reduce thrombogenicity in the left atrium, increase the length of the LA legs from 2 by 15 mm to 3 by 20 mm to prevent dislodgement and later adding a pivot between the left and right umbrella. A partial occluder was made by removing two opposite quadrants from the proximal disk. **Results:** Device deployment was possible in 93% (63 of 68) patients. In five patients, the device could not be deployed and an alternative device was used. In 45 patients complete closure of the fenestration was obtained and saturations increased from 84% ± 4% to 95% ± 2% ($P < 001$). In 18 high risk patients with suboptimal Fontan circulation, a modified device was used to effect partial occlusion: saturations increased from 79% ± 7% to 90% ± 4% ($P < 0.001$); a residual shunt persisted in most patients for several months. No thrombotic events were recorded during follow-up. **Conclusions:** The modified PFO star device can safely be deployed in Fontan patients to occlude or restrict flow through a fenestration. It has a low profile with minimal foreign material, is non-obstructive and minimally thrombogenic. © 2009 Wiley-Liss, Inc.

Key words: congenital cardiology; interventional catheterization; univentricular heart; fenestration

INTRODUCTION

Mortality and morbidity can be reduced in selected patients with univentricular heart lesions by creating an interatrial fenestration at the time of Fontan surgery [1–4]. Persistent cyanosis may lead to decreased exercise tolerance and a risk of paradoxical embolic events if spontaneous closure of the fenestration does not occur [5–10]. In addition to the use of surgically placed abdominal snares [11,12], a variety of transcatheter devices have been used for fenestration closure [1,13–22]. However, most of these devices lack desirable criteria such as ease of deployment, low profile with minimal material, nonthrombogenicity and a perfect closure rate. The aim of this study was to assess the efficacy and safety of a custom

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Conflict of interest: Nothing to report.

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Received 12 August 2009; Revision accepted 31 August 2009

DOI 10.1002/ccd.22275
Published online 23 November 2009 in Wiley InterScience (www.interscience.wiley.com)

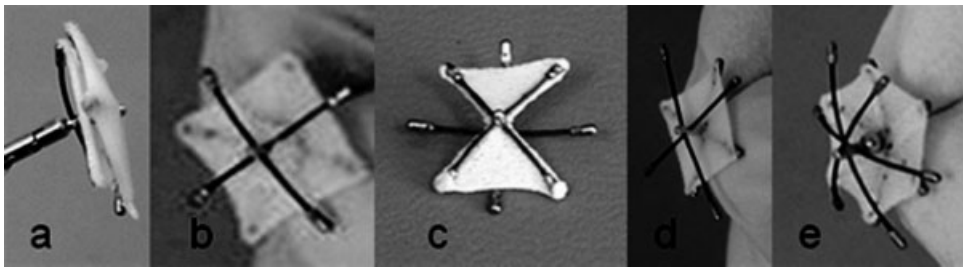

Fig. 1. Device modifications Picture of (a) standard FFD15 with 2 discs, (b) 115S with 2 × 15 mm covered legs at right side, (c) 115S partial occluder where two opposite quadrants were removed, (d) 115S20: 2 × 20 mm bare legs for left side, (e) 320A15 3 × 20 mm bare legs, pivot between left and right.

made device in an attempt to fulfil most of these criteria.

METHODS

Patients

Interventions were performed in two university hospitals: Leuven, Belgium and Birmingham, United Kingdom. Over an eight year period (March 2001–June 2009), fenestration closure using the PFO star type device, was attempted in 68 patients 1.4 ± 2.1 years after Fontan operation at an average age of 6.4 ± 2.9 years. The majority of patients had an extracardiac Fontan conduit ($n = 63$) with only five ($n = 5$) having lateral tunnel intracardiac conduits. The median size of the fenestration was 5.0 mm (range: 4–6) in a 20 mm (range 14–22) conduit. Patients were divided into two groups: The aim in the first group ($n = 50$) was to obtain complete closure of the fenestration. In the other group ($n = 18$), only partial closure was aimed for due to suboptimal Fontan physiology. All procedures were performed according to the regulations of the local ethical committees with knowledge of the modifications. Informed consent was obtained from the patients and their parents.

Technical Aspects

Procedures were performed under general anaesthesia. Intravenous heparin (100 IU/kg) was given after femoral access was obtained. All the Birmingham patients were on Coumadin before the procedure. Patients who were on Coumadin with an international normalized ratio of more than two received 50 U/kg of heparin during the procedure. The indication and timing of closure was highly dependent on the clinician and condition of the patient. Patients scheduled for complete closure typically would have a short, uneventful postoperative stay in hospital, no chylothorax and discontinuation of diuretics within weeks of surgery. They were in a good clinical condition with a good cardiac output. All other patients were considered high-risk (e.g., risk of protein losing enteropathy). Hemodynamic analysis was performed and eligibility for fenestration closure was decided according to the standard protocol of each unit. Test-occlusion of the fenestration was only performed in patients considered to be at high risk (less favorable hemodynamic parameters). Stenoses in the Fontan circuit and aorto-pulmonary collaterals were usually addressed before fenestration closure. Transesophageal echocardiography was routinely used during device placement in Birmingham, but only in some cases in Leuven. Size 9–11 F sheaths were required for deployment of the device. Patients were discharged on acetylsalicylic acid (1–2 mg/kg/d); Clodipogrel (0, 2 mg/kg/d) was added in cases with partial occlusion and also in some high risk patients for a period of one to six months after device implantation. Patients from Birmingham continued with Coumadin if they were receiving the drug before the procedures. Patients were routinely evaluated (clinically and echocardiographically) 24 hours, 1 month, and 6 months after the intervention with specific attention to residual leaks and clots. Percutaneous saturations were also controlled. Complete closure was defined as improved saturations clinically and the absence of clinically significant shunt on color.

Device

A 15 mm PFO star (FFD15, CARDIA™, Burnsville, MN) was used as the basis because of the flat profile once deployed. Essentially the original CARDIA™ device is a double umbrella unit made of polyvinyl alcohol foam fabric mounted on two curved 15 mm nitinol retention legs (Fig. 1a). In comparison, the Starflex® device (NMT Medical, Boston, MA), uses spring-loaded retention legs made from MP35N and surgical polyester matrix fabric. Modifications were made in a stepwise fashion, trying to overcome some of the technical difficulties that were encountered early in our experience such as difficult deployment in small conduits

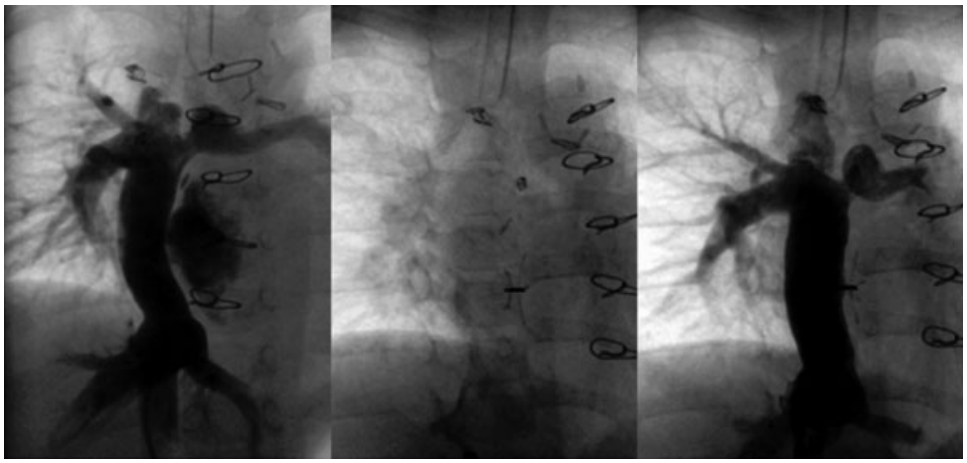

Fig. 2. Angiogram of extracardiac conduit showing shunting through the fenestration into the atrium (left); a 115S20 has been released in the fenestration (middle); angiogram after release shows complete obstruction of the fenestration without any protrusion of the device into the conduit (right).

due to prolapse of the distal legs through the fenestrations in some cases.

- 115X: The polyvinyl alcohol foam on the left disc was removed to further reduce thrombogenicity in the left atrium (Fig. 1b).
- 115S: A partial occluder was made by removing two opposite quadrants from the right atrial disc (Fig. 1c).
- 115S20 and 215S20: The length and number of the left atrial legs were increased from two 15 mm legs to three 20 mm legs, to reduce the risk of prolapse of the distal legs when deploying the proximal disc. The device with 2 × 20 mm legs are currently most commonly used (Fig. 1d).
- 320A15: A pivot was placed between the left and right umbrella to further reduce the risk of prolapse of the distal legs due to the sharp angulation of the introducer sheath in the Fontan conduit when deploying the device (Fig. 1e). Note that the articulated version is not currently available due to patent issues.

Where possible, before the operation, the surgeon was asked to make the fenestration in such a manner (as lateral as possible and suture the atrial wall a few millimeters around the fenestration) to facilitate safe and easy deployment of a device.

Statistical Methods

Data are described as means or medians with standard deviations and ranges (where appropriate). Before and after datasets were analyzed using paired *t* tests. Data was analyzed using GraphPad Prism version 5.00 for Windows, GraphPad Software, San Diego, CA. Statistical significance was defined as a *P*-value <0.05.

RESULTS

The device could be successfully deployed in 63 of the 68 attempted cases (91%). In five patients the device could not be implanted due to technical difficulties: repetitive prolapse of the small 15 mm distal legs when deploying the proximal disc ($n = 2$), insufficient space to deploy the left atrial legs due to anterior orientation of the fenestration ($n = 2$) and failure to advance the introducer sheath through the fenestration ($n = 1$; 4.5 mm fenestration in an 18mm conduit). Most of these were closed using a different device. Complete closure (Fig. 2) was possible in 45 patients and partial closure in 18. One patient had a significant residual shunt, which was closed with a second device during the same session. Devices used to achieve complete closure were: FFD15 ($n = 9$), 115× ($n = 15$), 115S20 ($n = 16$), 320A15 ($n = 6$). For partial closure the 115S device was used in 15 patients. Due to prolapse of the distal legs in one of these patients, the device was replaced with a manually tailored 320A15 device. In three other patients manually tailored devices were also used (115S20 in 2; 320A15 in 1).

Saturations improved significantly in both groups. Results can be viewed in Table I. Venous pressures were measured in 16 of the patients undergoing partial closure and changed from a median of 12 mm Hg (25th–75th percentile: 9.2; 13.8) to 12.5 mm Hg (25th–75th percentile: 10.0; 14.8) [$P = 0.005$; 95% CI: 0.3–1.2].

TABLE I. Saturations Before and After Implantation of Devices

	Saturations (mean ± SD)			
	Before procedure	After procedure	*P* value	95% CI
Complete closure	84% ± 4%	95% ± 2%	<0.001	9–12
Partial closure	79% ± 7%	90% ± 4%	<0.001	8–13

SD, standard deviation; CI,confidence interval.

Follow-Up

In the complete closure group, a minimal shunt was observed during follow-up in three patients after 1 month, which still persisted in one after 6 months as a mild shunt. In the partial closure group mild to moderate shunting residual shunting remained in all but two after one month. In one patient the device was percutaneously removed 51 days after implantation. Six months after device implantation, residual shunting was still documented by echocardiography in 12 patients with partial closure (saturations 90% ± 3%).

Complications

Complications observed during the procedure were: removal and reloading of device prerelease ($n = 3$), inadvertent deployment in the interatrial septum ($n = 1$), minor air embolism in the left atrium and left ventricular apex respectively ($n = 2$), an insignificant clot within the conduit ($n = 1$), a short run of ventricular tachycardia during device deployment ($n = 1$) and transient bradycardia during manipulation of the long sheath in the left atrium ($n = 1$). No thrombotic events were recorded during 4.9 ± 2.4 years follow-up.

DISCUSSION

The ideal closure device should have a low profile, consist of minimal fabric and metal, be easy to load and deploy, have a perfect closure rate (or the possibility of predictable partial closure) and be non-thrombogenic. In many institutions the extracardiac Fontan operation is now routinely performed in selected patients. In these extracardiac Fontan conduits, the profile of the device used for fenestration closure becomes even more important, since bulky devices may cause partial obstruction, especially in smaller conduits. Location, size, and type of fenestrations vary widely. Consequently, numerous devices have been used for fenestration closure, including Gianturco coils [14], detachable coils [20], Raskind duct occluders [13], Clamshell devices [4], CardioSeal devices [17], Amplatzer septal occluder [15,16,22], Amplatzer duct occluder [18], AngelWings devices [19], and covered stents [21]. Choice of device to close a fenestration is guided by several factors, including the type of Fontan circulation (intracardiac versus extracardiac), size and location, geometry of the fenestration, likelihood of placing a long sheath in the systemic atrium, ease of availability and atrial size [21]. Most of these devices produce satisfactory results, but are bulky and protrude significantly into the conduit. Although the incidence of thrombus formation is low with current medical strategies, a well profiled device may have additional benefits considering the increased risk for thrombosis as a result of ageing.

This study shows that the modified Cardia TM PFO device is effective and safe for both complete and partial closure of Fontan fenestrations. This device can be considered ideal due to the low profile, minimal metal and fabric as well as good closure rates. In cases where complete closure was desired, a 98% ($n = 44$ of 45) closure rate was observed after 6 months.

To our knowledge, this is the first transcatheter device used to effect partial closure of a Fontan fenestration. Partial closure resulted in significant improvement of almost 10% in oxygen saturations without clinically important changes in venous pressures. Although the precise amount and duration of residual shunting was less predictable, early complete closure occurred in only one patient, requiring elective removal of the device due to persistent venous hypertension and chylothorax. The 115S partial occluder was designed by removing two opposite quadrants from the right atrial disc (Fig. 1c). The devices can also be manually tailored in the catheterization laboratory by removing one or more quadrants of the polyvinyl alcohol foam on the proximal disc (depending on the magnitude of residual shunt required). None of the remaining 17 patients with partially closed fenestrations has so far been considered for percutaneous closure of residual shunts. Closure of these shunts should be technically feasible using coils or a covered stent.

The loading mechanism has been simplified and ease of deployment has certainly improved with the modified devices (115S20 and 320A15). Gaining experience, the currently used loading technique evolved: after flushing the system properly, the delivery cable is passed through a standard short 11 F introducer sheath as well as the short plastic folding sheath included in the unit. The device is then attached using the bioptome and gently pulled into the folding sheath with the legs of both discs pointing distally. The device is now slowly pulled back until the proximal (right-sided) legs protrude and is subsequently pushed forward into the folding sheath. This ensures appropriate folding direction of both the proximal and distal legs. The device is now retrieved into the 11 F short sheath and is ready for delivery after flushing. Experience proved

that using a short 11F introducer sheath facilitates smooth loading of the device and also allows introduction of the device in Mullins® (Cook, Bloomington, IN) sheaths ranging from 9F and larger. Further suggested technical improvements should include reducing the size of the introducer sheath and adding a 0–90° pivot at the release mechanism.

The device seems to be sufficiently nonthrombogenic. In the 115S20 device, the polyvinyl alcohol foam of the left disc was removed to even further reduce thrombogenicity in the left atrium. No specific and reliable risk factors for thrombus formation after Fontan circulation have been identified and no guidelines regarding anticoagulation based on controlled trials have been published [6]. All our patients were discharged on some form of oral antithrombotic therapy for a period of one to six months after device implantation. No thrombotic events were recorded during follow-up.

Complications mostly occurred early in our experience, were not severe and could be easily managed. Deployment of the device in the interatrial septum could have been avoided if transesophageal echocardiography was used, since remnants of the interatrial septum often interfere with procedures after Fontan operation.

Study Limitations

This study has only been limited to patients considered for fenestration closure and therefore represents a select group. The aim of this study was to evaluate the efficacy and safety of a modified occlusion device. The indications for fenestration closure were not evaluated in this study. Therefore also, no comparisons were made to other modalities of treatment. In patients with suboptimal Fontan physiology, ongoing clinical experience is needed to define the optimal "device-tailoring-technique" to obtain a more predictable partial closure in the individual patient. Transthoracic echocardiography may fail to detect some thrombi and although routine transesophageal echocardiography is more invasive, it should probably be considered in certain patients.

CONCLUSIONS

The Cardia™ PFO device and its modifications can safely be used to achieve complete as well as partial closure of Fontan fenestrations. Complete elimination of right-to-left shunting can easily be accomplished, but when aiming for partial closure the size and duration of residual shunting is less predictable. Because of a minimal amount of fabric and metal, the device is negligibly thrombogenic, has a perfectly low profile and as a result non-obstructive.

ACKNOWLEDGEMENTS

The authors acknowledge the design modifications as suggested by doctor Joseph DeGiovanni and implemented by the manufacturer. They also appreciate the contributions of Paul Miller and Rami Dhillon from Birmingham.

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