

Occlusion of Interatrial Fenestration with the Amplatzer Septal Occluder Device: First Case Report from Iran

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Abstract

We report successful occlusion of the fenestration after total cavopulmonary connection operation due to cyanosis with the Amplatzer septal occluder device. The procedure was satisfactory; arterial oxygen saturation increased markedly and the general condition of the patient improved remarkably. This procedure was done for the first time in the Islamic Republic of Iran.

A residual communication or fenestration between systemic and pulmonary venous return is often created during surgical construction of the Fontan circulation. This fenestration may prevent excessive increases in venous pressure in the early postoperative phase, especially in high risk patients.^{1,2} Many of these fenestrations close spontaneously;³ however some remain open causing persistent arterial desaturation and are a potential cause of paradoxical embolism.⁴ Subsequent closure of the fenestration using different transcatheter devices, such as double umbrellas⁵⁻⁷ and coils⁸ has been described. We describe our experience with occlusion of a fenestration using the Amplatzer septal occluder device, a procedure done for the first time in the Islamic Republic of Iran.

Key words: fenestrated Fontan operation ■ amplatzer septal occluder device ■ tricuspid valvar atresia

Case Report

A 13-year-old female patient was admitted due to progressive cyanosis and clubbing starting 6 months previously. She had been admitted for the first time two years before. After angiocardiography and echocardiography, she was diagnosed as suffering from tricuspid valvar atresia (TA), large perimembranous ventricular septal defect (VSD), severe infundibular and pulmonary valvar stenosis (PS) and ostium secundum atrial septal defect (ASD), with good-sized pulmonary arterial branches and left aortic arch. A total cavopulmonary connection with fenestration was done. Her

follow-up was unremarkable until 6 months ago, when cyanosis came back again insidiously.

On her latest admission, she had moderate cyanosis and clubbing. Her electrocardiogram was consistent with normal sinus rhythm, right superior axis and prominent left ventricular forces. Chest X-ray showed increased cardiothoracic ratio and normal pulmonary vascular markings. Echocardiographic examination revealed significant right-to-left shunt via the surgical fenestration, in addition to TA and VSD. The catheterization data are shown in Table I.

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Table I: Catheterization data before the procedure

Site	IVC	SVC	LA	LV	RPA	LPA	MPA
Pressure (mmHg)	12	12	12/8 8	100/0- 4	10	10	10
O ₂ saturation	56%	56%	77%	70%	65%	65%	65%

IVC=Inferior Vena Cava, SVC=Superior Vena Cava, LA=Left Atrium, LV=Left Ventricle, RPA=Right Pulmonary Artery, LPA=Left Pulmonary Artery, MPA=Main Pulmonary Artery

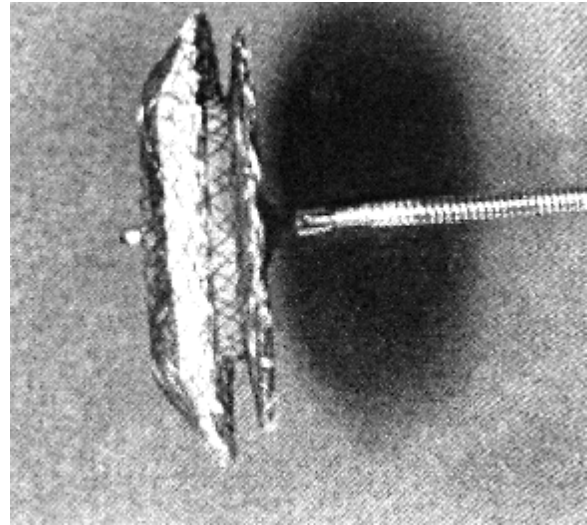
Angiocardiographically, there was no obstruction in the systemic venous to pulmonary arteries connection, but a large shunt through the surgically-created fenestration existed. Arterial blood gas (ABG) analysis showed an arterial oxygen saturation (O₂ sat) of 62% and a partial oxygen pressure (PaO₂) of 45 mmHg.

Due to the risks inherent to right-to-left shunts, such as systemic embolism, we decided to close the fenestration with an Amplatzer septal occluder device (AGA Medical Corp, Golden Valley, Minnesota, USA).

Occlusion Technique

The procedure was performed under light general anesthesia from the right femoral vein using a 7 F sheath. A 6 F end-hole catheter was used to cross the fenestration. Transthoracic echocardiography was used to obtain further measurement of the fenestration, confirm complete balloon occlusion and monitor deployment of the device. Right atrial angiography was performed before and after deployment of the device.

The Amplatzer septal occluder (Fig. 1) is a self-expanding, double-saucer shaped device, with a central stent-like connecting cylinder.^{9,10}

**Fig. 1:** Amplatzer septal occluder device.

Device selection was based on the stretched diameter or known punch diameter used to create the defect. Our patient tolerated test occlusion and device occlusion of the fenestration without significant changes in arterial or venous pressures. After fenestration occlusion with the device, catheterization was performed, data of which are shown in Table II. IVC, SVC, RPA and LPA pressures increased from 12 to 15 mmHg, and O₂ sat increased from 62% to 92%. The fluoroscopy time was 60 minutes. Successful implantation of the occluder device was achieved in the first attempt.

Table II: Catheterization data after the procedure.

Site	IVC	SVC	RPA	LPA
Pressure (mmHg)	15	15	15	15

The immediate post-implantation angiogram revealed only a small residual shunt (Fig.2).

Subsequent follow-up echocardiographies showed complete occlusion 3 months later; oxygen saturation and partial pressure by arterial blood gas analysis were 89% and 64 mmHg, respectively. At the same time, the patient's hemoglobin concentration and hematocrit were 14 mg/dL and 45%, respectively. The position and integrity of the device was satisfactory, and there were no complications over the 2-year follow-up period.

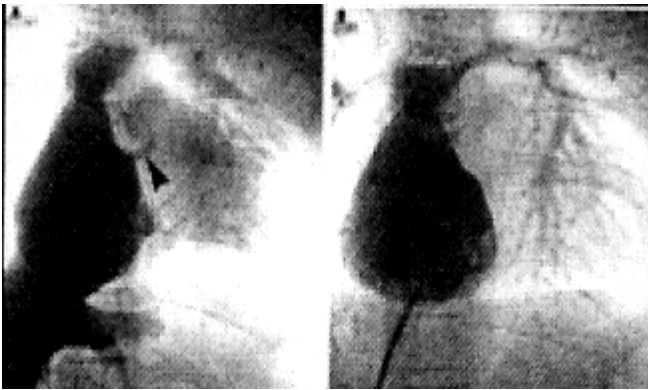


Fig. 2. Immediate post-implantation angiogram

Discussion

Transcatheter occlusion of the fenestration following the Fontan procedure can be done safely and effectively using the Amplatzer device. The complete occlusion on follow-up echocardiograms in our patient is very encouraging. There was no embolization or misplacement of the device.

The Amplatzer septal occluder was designed for the occlusion of left-to-right shunting fossa ovalis defects.^{9,10} The device consists of two saucer-shaped disks with the left-sided disc overlapping the right-sided disc. This design appears quite suitable for fenestration closure as the fenestrations are punched in a semicircular shaped Goretex patch. The saucer-shaped right-sided occlusion disc aligns well with the Goretex patch,

minimizing the amount of protrusion into the stagnant Fontan circulation.

Another advantage of this system is that the attachment mechanism consists of a simple screw design, and the device is easily delivered and retrieved through a 6 F or 7 F sheath. The use of echocardiography was helpful, although not essential, for ensuring that both discs were correctly placed before release.

In our hospital, we do not routinely undertake fenestration closure in the early postoperative phase, but rather reserve it for patients with persistent desaturation from right-to-left shunting through the fenestration for more than one to two years after the Fontan operation. In the patient in question, transcatheter occlusion of Fontan fenestrations was performed because of desaturation one-year-and-a-half after the surgical construction of the circulation. Occlusion of the fenestrations appeared to cause no adverse hemodynamic effects; the central venous pressure was not significantly increased, and there was no undue systemic venous desaturation. However, all these measurements were taken under shallow general anesthesia and do not necessarily reflect the capacity to increase cardiac output during exercise. So far our patient and her parents have reported symptomatic improvement.

References

1. Kopf GS, Kleinman CS, Hijazi ZM, Fahey JT, Dewar ML, Hellenbrand WE. Fenestrated Fontan operation with delayed transcatheter closure of atrial septal defect. Improved results in high-risk patients. *J Thorac Cardiovasc Surg* 1992; 103: 1039-1047.
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: 1762-1769.

3. Pearl JM, Laks H, Barthell S, Drinkwater DC Jr, Capouya ER, Chang PA. Spontaneous closure of fenestrations in an interatrial Goretex patch: application to the Fontan procedure. *Ann Thorac Surg* 1994; 57: 611-614.
4. Wilson DG, Wisheart JD, Stuart AG. Systemic thromboembolism leading to myocardial infarction and stroke after fenestrated total cavopulmonary connection. *Br Heart J* 1995; 73: 483-485.
5. Bridges ND, Lock JE, Castaneda AR. Baffle fenestration with subsequent transcatheter closure. Modification of the Fontan operation for patients at increased risk. *Circulation* 1990; 82: 1681-1689.
6. Redington AN, Rigby ML. Novel uses of the Rashkind ductal umbrella in adults and children with congenital heart disease. *Br Heart J* 1993; 69: 47-51.
7. Sommer RJ, Recto M, Golinko RJ, Griep RB. Transcatheter coil occlusion of surgical fenestration after Fontan operation. *Circulation* 1996; 94: 249-252.
8. Sharafuddin MJ, Gu X, Titus JL, Urness M, Cervera-Ceballos JJ, Amplatz K. Transvenous closure of secundum atrial septal defects: preliminary results with a new self-expanding nitinol prosthesis in a swine model. *Circulation* 1997; 95: 2162-2167.
9. Berger F, Ewert P, Bjornstad PG, Dahnert I, Kings G, Brilla Austenat I, et al. Transcatheter closure as standard treatment for most interatrial defects: experience in 200 patients treated with Amplatzer Septal Occluder. *Cardiol Young* 1999; 9: 468-473.