



## Transcatheter Closure of Fenestration after Modified Fontan Operation in Children

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### Abstract

**Background:** Fenestration in the modified Fontan operation allows right-to-left shunting, which reduces the Fontan pathway pressure and improves cardiac output. However, on account of the fact that persistent right-to-left shunting results in cyanosis and paradoxical emboli, fenestration closure is recommended after recovery from the Fontan operation.

**Methods:** This study recruited 3 patients who underwent the transcatheter closure of the Fontan fenestration with the ASD-Amplatzer because of severe cyanosis and significant intracardiac shunts.

**Results:** Fenestration closure was performed at a mean age of 8 yr (6-12 yr) and average of 15 months after the Fontan operation. Aortic O<sub>2</sub> saturation increased by an average of 17.6% (9-26%). During more than a two-year period of follow-up (mean: 27 months), two patients had complete occlusion on echocardiography and the other one had a small residual shunt. One of these patients had atrial flutter during the follow-up.

**Conclusion:** The transcatheter closure of the Fontan fenestration is a safe and feasible technique that is effective in elevating systemic O<sub>2</sub> saturation and well-being and confers acceptable growth and development in children.

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**Keywords:** Fontan procedure • Catheterization • Amplatzer

### Introduction

The Fontan operation, performed for the first time by Francis Fontan in 1968, has undergone many a modification over the years.<sup>1-6</sup> Fenestration in the Fontan pathway allows right-to-left shunting, which lowers the Fontan pathway pressure and improves cardiac output and short-term clinical outcome in the standard risk population.<sup>6-13</sup> Nevertheless, persistent right-to-left shunting between the neo-right atrium

and neo-left atrium begets cyanosis and increases the risk of paradoxical embolism.<sup>6,8</sup>

Fenestration closure is, therefore, recommended after recovery from the Fontan operation.<sup>9,14</sup> Several devices such as the Coil and Amplatzer have thus far been utilized for fenestration closure. We safely used the ASD-Amplatzer for the transcatheter closure of the Fontan fenestration.

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## Methods

A total of three patients have undergone the transcatheter closure of the Fontan fenestration at Rajaei Hospital in Tehran since 2003. These patients had persistent systemic desaturation and no signs of increased systemic venous pressure or low cardiac output such as hepatomegaly, ascites or edema. The patients' mean age at the time of fenestration closure was 8 yr (6-12 yr). The subjects' characteristics are listed in Table 1. All the patients tolerated the intracardiac Fontan operation with pericardial patch tunnel well. One case had a 12-mm fenestration due to technical error in surgery, and the other two had 5-mm and 6-mm punch-hole fenestrations.

The patients' medical records were reviewed to determine their demographic, echocardiographic, hemodynamic and angiographic data.

Table 1. Patients' characteristics

Case	Sex	Type of congenital heart disease	Age at Fontan Procedure	Type of Fontan	Age at fenestration closure	Fenestration size	Amplatzer size	Results
1	Female	Tricuspid Atresia + PS	10 <sup>y</sup>	Intracardiac (pericardial patch)	12 <sup>y</sup>	5 mm	9 mm	No Residue
2	Female	DMGA, DORV, VSD, PS	6 <sup>y</sup>	As above	6 <sup>y</sup> , 2 <sup>m</sup>	12 mm	17 mm	Small Residue
3	male	Single ventricle, LTGA, DILV, PS	4 <sup>y</sup>	As above	6 <sup>y</sup>	6 mm	12 mm	No Residue

PS, Pulmonary stenosis; D-MGA, DMalposition of great arteries; DORV, Double outlet right ventricle; VSD, Ventricular septal defect; LTGA, L-Transposition of great arteries; DILV, Double inlet left ventricle

The presence of residual shunts after fenestration closure was evaluated via angiography and serial echocardiography. After discharge, a clinical follow-up was carried out at a mean of 27 months.

Transthoracic echocardiography was performed before cardiac catheterization to detect possible thrombi and to evaluate patent fenestrations. Arterial and venous collaterals were assessed via angiograms at the aorta, superior vena cava or innominate vein and pulmonary artery. Before fenestration closure with the ASD-Amplatzer, a test occlusion of the fenestration with an ASD-sizing balloon was performed. About 15-20 minutes after the test occlusion, the changes in pressure and O<sub>2</sub> saturation within the Fontan pathway were measured. The criteria for fenestration closure<sup>6,14-17</sup> were:

1. Right atrial pressure ≤ 18 mmHg
2. Difference of the arteriovenous O<sub>2</sub> saturation ≤ 33%
3. Right atrial saturation ≥ 40%
4. Increase in the mean RA pressure < 4 mmHg

If the changes in pressure and cardiac output were acceptable, a suitable ASD-Amplatzer according to the fenestration size was implanted. Heparin was administered during the procedure and warfarin thereafter for thrombus prophylaxis. The differences in O<sub>2</sub> saturation pre- and post-fenestration closure and outpatient clinical follow-up are reported.

## Results

Three patients, who tolerated a modified Fontan operation with fenestration and met the criteria for fenestration closure, were selected. The ASD-Amplatzer was utilized safely, and there were no complications during more than two years of follow-up.

Table 2 depicts the O<sub>2</sub> saturation data at the femoral artery before and after fenestration closure. Aortic O<sub>2</sub> saturation increased by an average of 17.6% (9-26%). During the clinical follow-up, two patients had complete occlusion of the fenestration on serial echocardiography; and the other one, who had a 12-mm fenestration due to technical error in surgery, had a small residual shunt. One of the cases had atrial flutter after the Fontan operation, which was controlled with anti-arrhythmic drugs during the clinical follow-up.

The data on pressure changes in the Fontan pathway before and after the procedure are presented in Table 3. The mean pressure in the Fontan circuit increased by an average of 2.7 mmHg (Range: 2-3.6 mmHg). After fenestration closure, the mean pressure in the Fontan circuit increased in all of the patients.

Table 2. O<sub>2</sub> Saturation before and after fenestration closure (arterial blood gas and pulse oximetry)

Case	Before procedure	After procedure at catheterism lab	The latest follow-up at clinic
1	66%	92%	90%
2	76%	85%	87%
3	72%	90%	89%

Table 3. Pressure changes in Fontan circuit before and after fenestration closure (mmHg)

Case	Before procedure	After procedure	Change
1	9.2	12.8	↑3.6
2	10.1	12.1	↑2
3	8.8	11.3	↑2.5

None of the patients had a significant change in O<sub>2</sub> saturation during more than two years of follow-up after



fenestration closure.

Growth and development of the patients was acceptable during the follow-up according to the growth & development standard charts.

After the Fontan operation, all the patients received warfarin for anti-thrombotic prophylaxis. Warfarin was continued after fenestration closure in these patients. None of the patients had symptomatic thromboembolism within the follow-up period. Serial transthoracic echocardiography during the follow-up showed no thrombosis in the Fontan circuit.

## Discussion

Although fenestration in the Fontan procedure can improve the immediate postoperative course, it may give rise to persistent cyanosis and paradoxical embolism. There has long been debate in the medical community over the advisability of fenestration closure; more recent reports, however, advocate the procedure.<sup>6,17-20</sup>

Goff et al.<sup>21</sup> reported that not only did fenestration closure confer improved oxygenation but it also reduced the need for anti-congestive medication and improved somatic growth. They recommended that patients with the Fontan fenestration procedure undergo late transcatheter closure (>6 months after Fontan) if O<sub>2</sub> saturation rates were less than 90% and test occlusion was tolerated. The good results that we had previously achieved in the ASD occlusion with the Amplatzer in children encouraged us to opt for the transcatheter occlusion of the Fontan fenestration with the ASD-Amplatzer. Our study data demonstrate that Fontan fenestration closure with the Amplatzer in selected patients is a safe and effective procedure that can lead to an increase in O<sub>2</sub> saturation. In addition, more than two years of follow-up was indicative of acceptable growth and development, improved quality of life, no significant change in O<sub>2</sub> saturation and no symptomatic thromboembolic events in our three patients. We did not use Coils in this study because all our cases tolerated intracardiac tunnel-type Fontan. Sung Hye Kim et al.<sup>6</sup> reported that punch-hole fenestration with intracardiac tunnel-type Fontan was not suitable for closure with Coils. In addition, some studies showed that Amplatzer duct occluders and Amplatzer septal occluders such as the ASD-Amplatzer were suitable for fenestration closure.<sup>2,18-20</sup> In our study, two cases had a standard fenestration size and had no residual shunts after closure. However, the other patient, who had a 12-mm fenestration due to technical error in surgery, had a small residual shunt after the procedure. Of great significance is the fact that in all our patients O<sub>2</sub> saturation after fenestration closure increased significantly without any complications. One patient had atrial flutter after the procedure, but it was not a result of fenestration closure.

It is deserving of note that after the Fontan operation atrial arrhythmia is reported frequently.<sup>22</sup> In some studies, the

difference in O<sub>2</sub> saturation between early after fenestration closure and the latest follow-up visit is significant. This difference is believed to be caused by the reopening of the fenestration.<sup>6</sup> In the present study, there was no difference in O<sub>2</sub> saturation between the early and late follow-up visits of our three patients, which was probably due to the complete occlusion of the fenestration with the ASD-Amplatzer and the superior efficacy of the Amplatzer in comparison with the Coil.

## Conclusion

The transcatheter closure of the Fontan fenestration with the ASD-Amplatzer is a safe and feasible technique in selected patients, especially in those that can tolerate transient closure with sizing balloons before permanent closure with the ASD-Amplatzer.

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