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Original Article

Closure of Fontan fenestration with the use of covered stents: short- and mid-term results in a cohort of 50 patients

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Abstract *Objective:* The use of covered stents to close fenestration in total cavopulmonary connection is presented. *Methods:* We retrospectively reviewed data of all patients undergoing the procedure of a covered stent to close fenestration of total cavopulmonary connection between 2005 and 2012. *Results:* A total of 50 patients met the inclusion criteria. Median age and weight were 7.7 years and 20 kg, respectively. Median interval between Fontan completion and fenestration closure was 13 months. The femoral vein was used in 42 patients and the jugular vein in eight patients. Of the patients, seven received two stents. Covered stents were CP stents in 42 patients and Atrium Advanta V12 in eight patients. BIB balloons were used in 24 patients and simple balloons in 18 patients. Simultaneous occlusion of venous collaterals was observed in five patients. Median procedural and fluoroscopy times were 49 and 8 minutes, respectively. Mean central venous pressure rose from 10 to 12 mmHg. Mean oxygen saturation increased from 88% to 96%. Full occlusion was confirmed in 47 patients. The remaining had residual shunts: two patients had intracardiac Fontan, and one patient had a stent that could not be fully opened. Following the procedure, five patients had local bleeding, and three delayed discharge 48 hours after the procedure. There was no thromboembolic event after a mean follow-up of 49 months. *Conclusion:* Covered stent is a good option to close fenestration in extracardiac total cavopulmonary connection. It is safe, easily achievable with low fluoroscopy time, with very low risk of complication or failure. Good results are sustainable when excluding patients with none circular pathway.

Keywords: Total cavopulmonary connection; fenestration; covered stents

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The Fontan procedure has provided good palliation for patients with single ventricular physiology.[1] Surgical techniques have evolved over time. Total cavopulmonary connection with extracardiac conduit has been one of the technical modifications of the Fontan operation with major advantages: prevention of right atrial dilatation and scarring, reducing the risk of arrhythmias.[2] Creation of a fenestration (systemic-to-pulmonary venous atrial communication) during the completion of the Fontan pathway has shown many advantages for the immediate postoperative period: increased cardiac output immediately after surgery, prevention of central venous pressure elevation, reduced pleural effusions, and shorter intensive care unit course.[3,4]

Some fenestrations undergo spontaneous closure but many do not. Although they are useful during the immediate postoperative period, they can become a liability afterwards. They cause chronic desaturation and are a source of paradoxical emboli. If patent, they can be closed in patients with good haemodynamics.

Up to now the benefit of fenestration closure is still unclear.[5] Imielski et al have shown that increased oxygen saturation was the only benefit demonstrated.[6] The timing of closure remains unknown, the same

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study concluded that events could be prevented if closure was carried out before 11 months after Fontan completion, although they concluded that further studies were needed to have guidelines for the timing of closure.

In extracardiac cavopulmonary connections, several devices have been used to close fenestrations: atrial septal occluder – CardioSEAL, Amplatzer septal occluder, or duct occluder (AGA Medical, Golden Valley, Minnesota, United States of America) – coils, or vascular plug.[7,8] All have necessitated introduction of a guidewire and a long sheath into the systemic atrium. This approach can be made difficult by the fenestration's geometry and the risk of systemic embolism.

Covered stents are used in interventional cardiology to treat vascular abnormalities.[9,10]

Our team has described the results of covered CP stent in six patients to close fenestration of extracardiac total cavopulmonary connection.[11] We report here the results of a cohort of 50 patients in whom fenestration was closed with a covered stent focusing on the technical aspects, haemodynamic data, and mid-term results.

Materials and methods

Patients who had undergone Fontan fenestration closure with a covered stent between 2005 and 2012 were reviewed. Patients not undergoing fenestration closure, that is, patients with unfavourable haemodynamics (basal central venous pressure > 15-mmHg), were excluded from this study.

Procedures were performed under general anesthesia. Heparin (50 UI/kg) and antiobiotics (cefaclor 50 mg/kg) was administered to all the patients. Central venous pressure, pulmonary artery pressures, and wedge pressure were recorded. On the basis of these data, the decision was made to close or not the fenestration. An angiogram in the conduit was performed in two planes (lateral view and right atrial oblique view) to visualise the fenestration and the pulmonary arteries. Patients with central venous pressure greater than 15-mm Hg and no anatomical reasons – that is, pulmonary artery stenosis – were considered not suitable for fenestration closure and the procedure was abandoned at this stage. In others, a long sheath was advanced in the conduit over a stiff guide wire; a covered stent was then positioned within the conduit over the stiff guide wire. The dimension of the balloon was choosen according to the diameter of the Gore-Tex conduit used. Post-procedure central venous pressure was recorded and an angiogram was repeated to check residual shunt. In case of residual shunt, balloon inflation was repeated. If fenestration was not totally occluded, a second covered stent was placed afterwards.

Discharge was planned the day after the procedure. Subcutaneous heparin was administered after the procedure for 24 hours. Following our institutional policy, aspirin (100 mg) was added to their antiocoagulation regimen (Coumadin) for 6 months. After 6 months, Coumadin was stopped leaving the patients on aspirin alone until adolescence where aspirin was switched again with Coumadin. Demographic, haemodynamic, procedural data, and outcome were recorded as well as the need for recatheterisation for any reason – recurrent cyanosis, failing Fontan and so on.

Results

A total of 50 consecutive patients met the inclusion criteria. Of them, 17 patients had a follow-up of < 2 years, and 33 patients had their fenestration closed for more than 2 years. Median age and weight were 7.8 years (2.6–20.4; ± 3.8 years) and 20 kg (11.5–67; ± 14.6 kg), respectively. Median interval between Fontan completion and fenestration closure was 13 months (1–171; ± 37 months). Of the patients, five had previous history of thromboembolic events. Patient characteristics are shown in Table 1.

Cardiac catheterisation data are described in Table 2.

Procedures were performed through the femoral vein in 42 patients and the jugular vein in eight patients. Mean basal saturation was 89% (75–97; ± 5.8%) and increased to a mean of 96% (85–100; ± 2.9%)

Table 1. Patient characteristics.

	n = 50
Age at closure (years); (median; minimum–maximum–SD)	7.8 (2.6–20.4; ± 3.8)
Body weight (kg)	20 (11.5–67; ± 14.6)
Interval between surgery and closure (mo) (median; minimum–maximum–SD)	13 (1–171; ± 37)
Heart disease	
Right heart lesion	21
Left heart lesion	13
Undifferentiated	16
Follow-up (months) (median; minimum–maximum–SD)	49 (1–131; ± 29)

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after closure. Basal central venous pressure was 11 mmHg (7–14; ± 2.8 mmHg) and increased to 13 mmHg (9–19; ± 2.8 mmHg) after closure. Mean procedure duration was 45 minutes (20–120; ± 24 minutes), and mean fluoroscopy time was 8 minutes (2–35; ± 8 minutes). Mean radiation was 103 mGy (29–354; ± 90 mGy). These parameters did not change over time.

Table 2. Procedure characteristics.

	n = 50
Access	
Femoral	42
Internal jugular	8
Sheath (Fr)	
10	7
11	17
12	5
14	21
Saturation pre (%)	89 (75–97; ± 5.8)
Saturation post (%)	96 (85–100; ± 2.9)
CVP pre (mmHg)	11 (7–14; ± 2.8)
CVP post (mmHg)	13 (9–19; ± 2.8)
Procedure (minute)	45 (20–120; ± 24)
Fluoroscopy (minute)	8 (2–35; ± 8)
Stents	
Covered CP stent	49
Atrium V12 stent	8
Balloon type	
BIB	26
Tyshak	16
Atlas	10
Balt	6
Number of stent	
1	43
2	7
Residual shunt	
Early	5
Late	2*
Associated procedures	5
Procedural complications	2*
Access complications	5
Late complications	1

CVP = central venous pressure
*One lost to follow-up

A total of 57 stents were used. Two stents were used in seven patients (CP stent group) because of incomplete sealing of the fenestration. The covered stents were CP stents used in 42 patients (Fig 1) and Atrium Advanta V12 in eight patients (Fig 2). For stent insertion, we used BIB balloons in 24 patients or simple balloons (Tyshak or Balt) in 18 patients. In eight cases, two different balloons were used: five in patients receiving two stents, and three in patients receiving an Atrium stent needing post dilatation. Five patients had simultaneous occlusion of venous collaterals.

In seven patients, two stents have been used because of incomplete sealing of the fenestration with one stent. Of these patients, two had an intracardiac Fontan with a composite tube between the vena cava and the pulmonary arteries made of fragments of Gore-Tex and of the atrium; moreover, despite two stents, significant residual shunt across fenestration was seen on angiogram at the end of the procedure. Another patient needed two stents with failure to fully open the stents in the conduit and persistent shunt across fenestration. Unfortunately, this patient was lost to follow-up. The remaining four patients had successful closure of fenestration with the two stents.

The dimension of the sheath needed for the covered stent placement as well as the type of covered stents and balloons evolved over the period. The dimensions of the sheath decreased over time from 14 to 11 Fr ($p = 0.009$ when comparing the patients operated upon > and < of 2 years ago). We moved from using CP covered stent (32/33) mounted on BIB balloons (20/33) to premounted Atrium V12 (9/17) or CP covered stents on simple balloons (8/17).

After the procedure, five patients had bleeding: three at the venous puncture site and two in

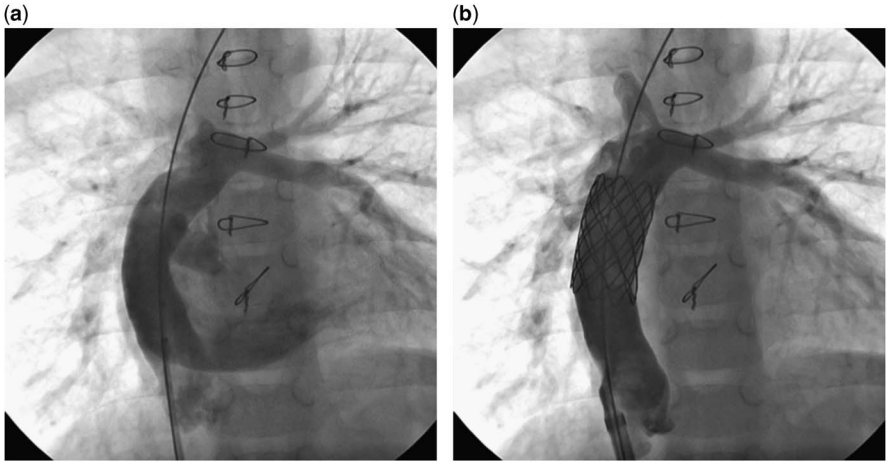

Figure 1.
*(**a** and **b**) Closure of fenestration with a covered CP stent.*

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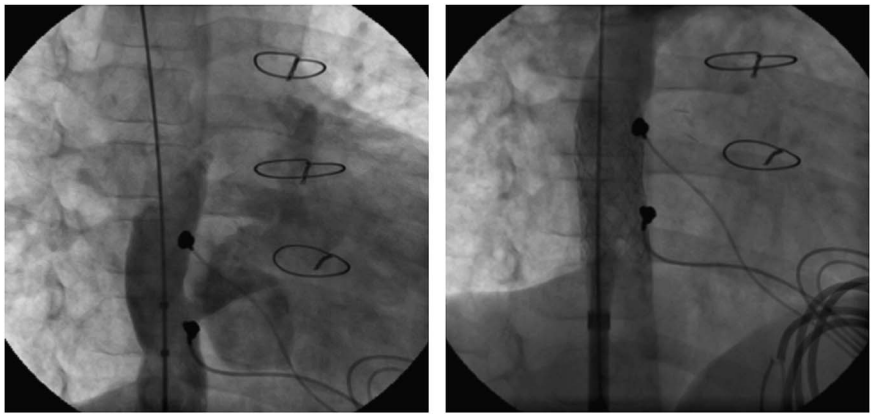

Figure 2.
Fenestration closure with a premounted stent Atrium V12 16 × 41 mm.

retroperitoneal position. Patients with retroperitoneal bleeding had complicated procedures: one had a hepatic puncture because of difficulties to access fenestration, and one patient underwent a long procedure because of the non-circular conduit used for the completion of Fontan circuit. Patients with local bleedings were 4.7, 9.5, and 10.6 years old. Of the patients one required blood transfusion and the other patients remained asymptomatic, of whom three were discharged on day 2 after the procedure.

One patient presented with pleural effusions. This patient underwent closure of the fenestration 15 months after Fontan completion. Central venous pressure was 10 mmHg before closure and 13 mmHg after closure. Two venous collaterals were occluded with coils at the same time. Pleural effusion (exudative fluid) occurred 1 month after fenestration closure, and despite diuretics the patient required recurrent chest drainage. Central venous pressure was 15 mmHg during second cardiac catheterisation with good placement of the stent. The patient was on high doses of diuretics but not on any pulmonary vasodilators. Creation of a new fenestration was considered, but she underwent Fontan take-down; however, she died afterwards from congestive heart failure.

Of the two patients with composite tube made of Gore-Tex and right atrium, one underwent a second cardiac catheterisation because of persistent cyanosis 7 years after the stent placement. Fenestration was occluded with no residual shunt and cyanosis was due to veno-venous collaterals.

After a mean follow-up of 49 months, there was no thromboembolic event reported. No stent fracture or displacement was reported.

Discussion

The Fontan pathway offers a good palliation for patients with univentricular physiology. Early and late complications still exist and the technique has been modified over the years to decrease mortality and morbidity. Extracardiac conduit and lateral atrial tunnel are the two modificated techniques mostly used currently.[2] In the 1990s, the Fontan pathway was fenestrated after benefical effects of the systemic to pulmonary venous shunt were demonstrated in the early postoperative course. Indeed, fenestration has been shown to improve cardiac output immediately after surgery, to reduce pleural effusions and to shorten the intensive care unit course.[3,4] After the initial period some fenestrations spontaneously close, but some remain open and they cause chronic desaturation, reduce exercise tolerance, and may be a source of paradoxical embolism.

Extracardiac conduit avoids dilatation of the right atrium and scars in the atrium, which might decrease the risk of arrhythmias. In our unit, extracardiac conduit is the surgical technique used to complete the Fontan pathway. The circuit is always fenestrated at the time of surgery to ease early postoperative period. The risk of thromboembolic event exists. In our series, five patients had thromboembolic events: two before bidirectional Glenn shunt, one during the early postoperative period after the Fontan completion, one after the Fontan completion (non-observance of the anticoagulation), and one after an attempt to close the fenestration with a CardioSEAL device (residual shunt on echocardiography).

The benefical effect of fenestration closure and the timing of closure remain under debate.[5,6,7] We routinely close fenestrations if they are patent. In our institution, the timing of the closure depends on the medical history of the patient, intensity of desaturation, and of the cardiologist in charge, as no guidelines currently exist. In our series, the closure of fenestration was performed at a median after surgery of 13 months.

Many devices have been described to close fenestration: device used to close intracardiac shunts or

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arterio-venous connections: atrial septal occluder (CardioSEAL, Amplatzer setpal occluder) or duct occluder (Amplatzer, AGA Medical), coils, or vascular plug. The choice of the device is related to the size, the location, and the geometry of the fenestration.[8,9,11]

We described the use of covered CP stents mounted on BIB balloons for the closure of extracardiac conduit fenestration.[10] This technique does not require to enter in the systemic atrium, which can be a challenge because of the location of the fenestration (acute angle) and can lead to embolic event (thrombo or air events). In addition, this technique completely obviates the need for TEE and theoretically the need of general anesthesia. We, however, chose to perform the procedure under general anesthesia because the use of large sheaths in small children is painful.

We herein describe 50 cases of Fontan fenestration closure with covered stents and describe the mid-term follow-up for 33 patients who had the fenestration closed for more than 2 years. The rate of success in extracardiac conduit was 100% with low fluoroscopy time and low procedure time. In comparison, we demonstrated that this technique was not valid for composite -total cavopulmonary connection conduit with early failure in two patients. Technically, the procedure was straightforward and usually short. No guidewire or sheath had to be inserted into the pulmonary venous atrium, reducing the risk of systemic gas embolisation or arrhythmias. Of the patients, five had venous collaterals closed with coils during the procedure. In one patient lost to follow-up, the stents could not be fully opened and remained partially opened in the TCPC conduit with a residual shunt through the fenestration. The only significant post-procedural complication was local bleeding, as two patients experienced retroperitoneal bleeding treated medically and three patients experienced minor bleeding at the venous puncture side. To reduce this type of complication, we decided to reduce the size of the long sheath by using different covered stents and balloons for fenestration exclusion. Over time, we used Tyshak balloon instead of BIB Ballon to deliver covered CP stents and premounted stents (Atrium V12, Advanta) when possible. As a result, sheath size decreased from 14 to 11 Fr over time. Our policy is now to use Atrium V12 stents when surgical conduit is 16-mm or smaller and CP-covered stents mounted on Tyshak balloons when conduits are >16-mm. Vessel trauma is as a result reduced.

The closure of fenestration was very well tolerated in all cases, except in one patient who presented with recurrent pleural effusions. Her case was reviewed in our joint cardiac conference and various managements were considered including increasing medical treatment – corticosteroid, pulmonary vasodilators, long-term subcutaneous heparin and so on – transcatheter or surgical creation of a new fenestration, and heart transplantation. The decision was made to take the Fontan down. She unfortunately died after this surgery.

There was no history of conduit thrombosis or thrombus formation in the atrium. Patients were administered a combination of Coumadin and aspirin for 6 months and remained on aspirin afterwards. All patients are alive with no thromboembolic event or no cardiac catheterisation history. There is no report of recurrence of cyanosis, stent fracture, or stent dislodgement so far.

Conclusions

Covered stent is a good option to close fenestration in extracardiac total cavopulmonary connection. It is safe, easily achievable with low fluoroscopy time, and has very low complication rate and failure. The good results are sustainable when excluding patients with no circular pathway. There is no report of late recurrence of cyanosis, of thromboembolic event, and of stent fracture or dislodgement.

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Conflicts of Interest

None.

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