Chapter 27
Fontan Fenestration Closure

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27.1 Introduction

Since Fontan and Baudet first described their technique for univentricular repair in 1971, numerous modifications have been described [1]. The technical approach to the Fontan operation itself has evolved into two major approaches: the lateral tunnel technique and the extracardiac technique. The risk of death is greatest in the immediate postoperative period, often in the setting of a low cardiac output state. Some of the complications that contribute to the early mortality may be transient or reversible, i.e. elevated pulmonary vascular resistance and ventricular dysfunction, or treatable in the case of residual distal pulmonary artery distortion. The concept of a fenestration between the systemic venous and the pulmonary venous pathways was introduced in 1971 [2], when the first patient of

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the atriopulmonary connections series received a 6-mm fenestration to serve as a "pop-off" valve, allowing decompression of the systemic venous pathway into the left atrium. Right-to-left shunt at the atrial level tends to improve cardiac output at the expense of systemic oxygen desaturation. Fenestration has been shown to improve early outcomes, including a decreased duration and quantity of chest tube drainage, a shorter duration of mechanical ventilation and a shorter postoperative hospitalization [3].

27.2 Timing of Closure

Spontaneous fenestration closure is well recognized, but persistent patency may warrant closure to improve arterial oxygen saturations and prevent cerebrovascular accidents due to paradoxical thromboembolism. Whether and when to intentionally close a fenestration remains debatable: depending on institutional protocol, management may vary from active fenestration closure at predetermined intervals to a "hands-off" approach, allowing the fenestration to follow its "natural history". In our institution, the total cavopulmonary connection (TCPC) by interposition of a 16–20-mm extracardiac Gore-Tex graft (WL Gore and Associates, Flagstaff, AZ) is currently the operation of choice, and a fenestration is created virtually in all patients. Despite all controversies surrounding routine fenestration closure, it seems reasonable to close fenestrations in patients with favourable haemodynamic assessment and clinically significant desaturation, based on the secondary effects of cyanosis.

Timing of fenestration closure remains debatable, but current recommendation is to postpone fenestration closure to at least 6 months after completion after the Fontan circulation if O_2 saturations are <90 % and test occlusion is tolerated.

27.3 Patient Selection

Patients with resting oxygen saturation (SaO_2) of less than 92 % or significant desaturation on exertion should be further assessed. Defining the "ideal" patient for fenestration closure is debatable, but factors that can be considered more favourable are the following:

- Uncomplicated postoperative course after bidirectional Glenn shunt and Fontan procedure (absence of prolonged pleural effusions or chylothorax and discontinuation of diuretics within weeks after surgery)
- No clinical evidence of low cardiac output or systemic congestion
- Exclusion of a high-velocity shunt through the fenestration on echocardiography
- Unobstructed Fontan connections and low pulmonary vascular resistance (PVR)
- Good ventricular function and absence of significant valve regurgitation
- Unobstructed systemic outflow and pulmonary venous return
- Normal AV conduction on ECG and absence of significant arrhythmias

27.4 Evaluation Before Catheterization

Most information can be obtained by clinical assessment (including ECG and exercise testing, if appropriate) and transthoracic echocardiography (TTE). However, TTE may fail to detect some thrombi and, therefore, transoesophageal echocardiography (TEE) may be necessary in some patients. In case of suspected pulmonary venous obstruction or pulmonary artery distortion, computed tomography or magnetic resonance imaging should be performed prior to catheterisation. These imaging techniques can also give additional information about the presence of venovenous or aortopulmonary collaterals.

27.5 Catheterization Procedure

- Catheterization is performed with intubation and general anaesthesia in room air. Antibiotic prophylaxis and heparin (100 IU/kg IV, maximum 5,000 IU) should be administered routinely.
- After obtaining femoral venous and arterial access, a complete haemodynamic assessment should be performed, documenting saturations and pressures throughout the Fontan pathway and systemic circulation.
- Angiography should then be performed in the superior and inferior caval veins and pulmonary arteries to visualize the Fontan connections, surgical fenestration (Fig. 27.1), additional interatrial leaks and possible venous collaterals.
- Selective injection in the innominate vein and right hepatic vein is indicated to exclude venovenous connections. An aortogram should be performed to exclude significant aortopulmonary collateral arteries.
- Anatomical abnormalities amendable to interventional treatment should be addressed first: balloon dilation and/or stenting of obstructed Fontan connections or stenosed/hypoplastic pulmonary arteries, occlusion of significant collaterals if appropriate and treatment of systemic obstruction (i.e. recoarctation) by balloon dilation and/or stenting.

27.6 Haemodynamic Assessment and Test Occlusion of Fenestration

- Identifying "favourable" haemodynamics for fenestration closure is ill defined [4].
- Measurement of PVR in Fontan circulation is fraught with difficulties due to inability in accounting for collateral circulation,

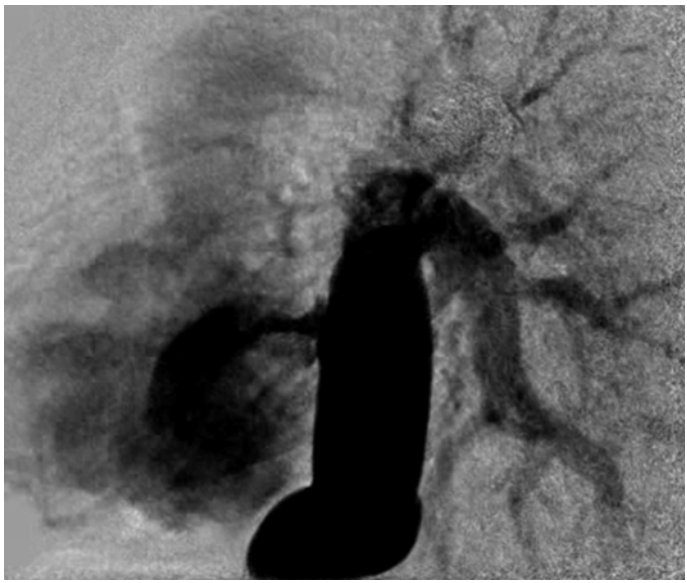

Fig. 27.1 Lateral view of contrast injection in inferior caval vein: a 20 mm conduit is mounted between the inferior caval vein and pulmonary artery; a 4.5 mm fenestration allos righ-to-left shunt into the left atrium

possibility of pulmonary arteriovenous malformation, low cardiac output state, presence of systemic venous obstruction, unequal distribution of lung flow and possibility of pulmonary venous obstruction. All these factors multiply the error in accurate assessment of PVR.

- Test occlusion of the fenestration is used during catheterization to identify patients presumably unsuitable for fenestration closure, by quantifying changes in the systemic or mean venous pressure and systemic saturation. Whether temporary test occlusion in a sedated and intubated patient is a reliable surrogate for predicting physiology in the awake and spontaneously breathing Fontan patient is debatable but is certainly recommended in case of unfavourable baseline haemodynamics and in high-risk patients.

- Test occlusion can be performed using a 7-F balloon-tipped, multi-lumen catheter (Swan-Ganz catheter). The balloon catheter is passed over a wire into the systemic atrium; the balloon is inflated using 1 cc diluted contrast and pulled back against the atrial wall/fenestration to allow for temporary occlusion (at least for 15 min).
- Alternatively, a small compliant balloon (typically a 6–8-mm Tyshak balloon (depending on fenestration size)) can be inflated within the fenestration itself, using the femoral sheath for pressure and saturation measurement.
- Complete occlusion should be confirmed by angiogram (through the proximal port of the balloon-tipped catheter or the femoral sheath) (Fig. 27.2).
- Measurements should be repeated, documenting VCI mean pressure and saturation and aortic pressure and saturation.
- Fenestration occlusion can probably be undertaken safely in patients with a systemic venous pressure of <18 mmHg during test occlusion or in the absence of a significant (>4 mmHg) increase in mean systemic venous pressure or reduction in mixed venous saturation of >10 %.
- We would strongly discourage fenestration closure in patients with systemic venous pressure of ≥20 mmHg.

27.7 Choice of Device

- When planning transcatheter closure, various factors should be considered, including the size and location of the fenestration, its geometry, the distance between the atrial chamber and the internal edge of the conduit and the possibility of placing a long sheath in the systemic atrium. Patient size and weight should also be taken into consideration.
- The ideal device must not only provide complete occlusion with reliable stability but also have a low profile without

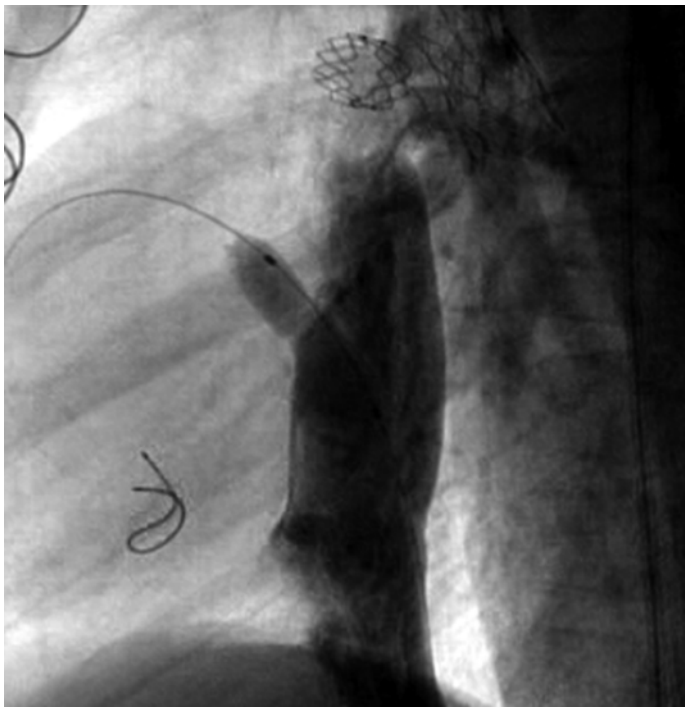

Fig. 27.2 Low-pressure balloon occlusion of 4.5 mm fenestration with a 6 mm Tyshak balloon; contrast injection through venous sheath confirms total occlusion

distorting the anatomy or obstructing flow within the Fontan conduit/baffle.

- Along with the diversity of methods to complete the Fontan operation, a number of techniques have been utilized to create fenestrations, depending on the type of Fontan operation (extracardiac conduit versus lateral intra-atrial tunnel) and institutional preference. For the lateral tunnel type of Fontan, a coronary punch is used to create a fenestration in the Gore-Tex baffle. In case of the extracardiac conduit, a fenestration

can again be created by placing a coronary punch in the Gore-Tex conduit, after which the atriotomy resulting from detaching the inferior vena cava from the right atrium is sewn to the Gore-Tex graft as a circle of about 2.5 cm, with the fenestration in the centre of the circle. This prevents the adjacent atrial wall from impacting the size of the fenestration. In some centres, the fenestration is made using a short (5–7-mm) polytetrafluoroethylene (PTFE) shunt between the extracardiac conduit and the systemic atrium to decrease unexpected spontaneous closure. Kreutzer et al. described a novel method to create a fenestrated extracardiac Fontan conduit by means of a pericardial tube anastomosed end to end with the inferior inlet of the right atrium [5].

- Fenestration size is usually between 3.5 and 5 mm, depending on the type of fenestration (punch hole vs. short PTFE shunt) and patient characteristics (risk stratification).
- In addition to intentional fenestrations, significant Fontan baffle leaks exist in up to 15 % of patients with a lateral tunnel-type Fontan. The baffle leaks are mostly located at the base of the right atrial appendage (RAA) at the suture line excluding the superior vena cava flow from the RAA. This suture line seems particularly susceptible to tiny leaks being left postoperatively due to the difficulty in tightly joining a smooth patch material to the corrugated surface created by the pectinate muscles. The increased venous pressure within the baffle can enlarge these channels creating a clinically significant shunt over time. While the origin of the leaks may be similar, the anatomy of the fistulous tract may vary.
- Due to the varying location, size and type of fenestrations, several catheterization methods have been described for fenestration closure by multiple authors, including Gianturco coils, detachable coils, clamshell devices, CardioSEAL devices, Amplatzer septal occluders, Amplatzer duct occluders, Amplatzer vascular plugs, Helex septal occluder, Angel Wings devices, Gianturco-Grifka vascular occlusion devices and CARDIA™ PFO star device.

- The placement of clips at the time of surgery to mark the location of the fenestration or to narrow the mid-portion of a tube graft for better anchoring of coils or devices has facilitated closure at the time of catheterization.
- Over the past two decades, the following devices have been used in our unit for closure of fenestrations and baffle leaks: Rashkind device, CardioSEAL, Amplatzer ASD occluder, Amplatzer VSD occluder and PFO star type device. In search of an ideal device, we modified a 15-mm PFO star (FFD15, CARDIA™, Burnsville, MN) by removal of the left disc to reduce thrombogenicity in the left atrium, increase the amount and 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 [6]. We considered this device "ideal" because of its low profile, minimal fabric and metal, good closure rate and non-thrombogenicity. However, introducer sheaths are much larger than needed with the newer devices and although the loading mechanism has been simplified, there remains a learning curve. Currently, the Amplatzer duct occluder type II has become our device of choice for closing the typical punch-hole-type fenestration performed in our extracardiac Fontan conduits. This device has a high conformability and its dual articulating discs makes placement in the fenestration relatively easy. The fabric-free technology allows for delivery through a low-profile 4-F catheter while maintaining a high rate of occlusion without being bulky and potentially obstructive.

27.8 Crossing and Outlining the Fenestration/ Baffle Leak

- The position of the fenestration and/or baffle leak should be delineated using angiography in different views.
- TEE can give additional information in patients with a baffle leak, or in cases where the size of the atrial chamber is small or a residual atrial septum may be problematic.

- Depending on the location and shape of the fenestration, it can be crossed by the aid of various preshaped or custom heat-shaped catheters such as the right Judkins catheter and a floppy exchange wire [i.e. 0.035-in Terumo guide wire or Woolley Hi-torque Floppy wire (Mallinckrodt, St. Louis, MO)].
- Once the wire is advanced, it can be exchanged for a straight catheter if necessary, facilitating placement of stiffer wires.
- Depending on the method of test occlusion, a 7-F balloon wedge catheter is passed over an exchange wire, or a small compliant balloon (i.e. Tyshak balloon) is passed over the appropriate wire (depending on balloon). Test occlusion is performed, and if necessary, balloon sizing can be performed if the punch-hole size of the fenestration is not known or in case of a baffle leak or fistulous connection.
- After selecting the appropriate device, a long sheath with dilator or delivery system (depending on device) is passed across the defect over the exchange guide wire.
- The dilator and wire should be removed slowly, allowing for spontaneous backflow of blood through the sheath, followed by careful flushing to avoid air embolism.
- Loading and deployment of the device are performed in the usual way as described for the specific device. Prior and after release, the device position should be checked angiographically (Figs. 27.3a, b) and on TEE if necessary. Haemodynamic measurements and saturations should be repeated.

27.9 Alternatives to Device Closure

- Device closure necessitates introduction of a guide wire and a long sheath into the pulmonary atrium. Technical difficulties in closing fenestrations by different devices have been shown in TCPC patients with residual native atrial septum, forming an intermediate chamber on the pulmonary venous side of the fenestration and additionally carrying the risk of

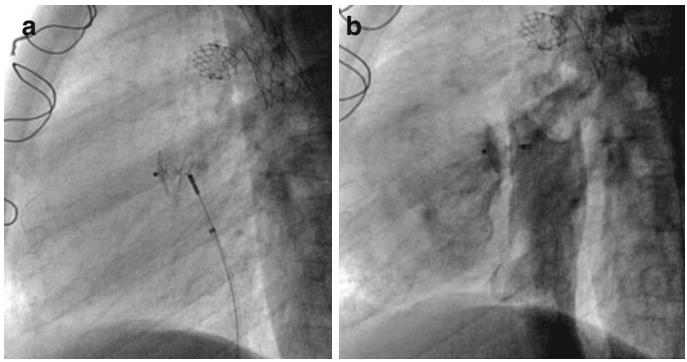

Fig. 27.3 (**a**) deployment of Amplatzer duct occluder type II in fenestration, device still attached to delivery cable. (**b**) cavogram after release of device: both disks are clearly at appropriate end of the fenestration; there is still some contrast like "smoke" through the device which will disappear within minutes after release

systemic embolism in the case of difficult manipulations with wires and sheaths.

- In these situations the use of a covered Cheatham Platinum (CP) stent (NuMED, Hopkinton) could be a valuable option, at least in patients weighing more than 15 kg. A 12-F long sheath is advanced over a stiff guide wire across the TCPC conduit, positioning the tip of the wire in the superior caval vein. The stent is hand crimped onto a BIB balloon catheter (NuMED, Hopkinton), with a diameter equal to or 1–2 mm larger than the angiographic conduit diameter. Short procedural and fluoroscopy times required by this procedure are attractive, as well as the complete immediate fenestration closure. The technique also avoids protrusion of prosthetic material in the pulmonary atrium that could prompt apposition of thrombotic material and systemic embolism. Disadvantage of this technique is the relatively large sheath size needed for covered stent delivery.
- The combination of Fontan baffle stenosis "downstream" from the fenestration or baffle leak may significantly worsen right-to-left shunting especially during exercise. Device

occlusion of fenestrations or leaks may additionally narrow the pathway in these patients and is therefore undesirable. Balloon expandable covered stents may be less desirable in this setting as there is often a significant size discrepancy between the stenotic area and the largest baffle diameter, which can potentially result in either incomplete closure of the baffle leak or an inadvertent baffle tear. Madan et al. recently described two patients with the combination of Fontan baffle stenosis and patent fenestration successfully treated with a Zenith abdominal aortic aneurysm endograft (Cook Medical) [7]. The Cook Zenith endograft is constructed using full-thickness woven polyester fabric sewn to a self-expanding stainless steel endoskeleton. This framework with fabric on the outside provides good graft to vessel wall apposition. The delivery system of the Cook Zenith stent offers an advantage over balloon expandable stents by enabling precise positioning and readjustment of the graft before final deployment. In addition, post-deployment, the self-expanding stent conforms to the vessel wall and selective dilation of specific areas using different balloons can then be performed. This is advantageous in the Fontan patient where the baffle is not of uniform calibre in order to minimize residual leak. Due to the large delivery sheath size (16Fr), this technique should be reserved for older children or adults.

27.10 Closing the Stented Fenestration

- Spontaneous closure of a fenestration during the early postoperative period may lead to haemodynamic deterioration associated with elevated systemic venous pressures, low cardiac output, progressive oedema and effusions. The use of intravascular stents to reopen or create a fenestration in these unstable patients can be life-saving.

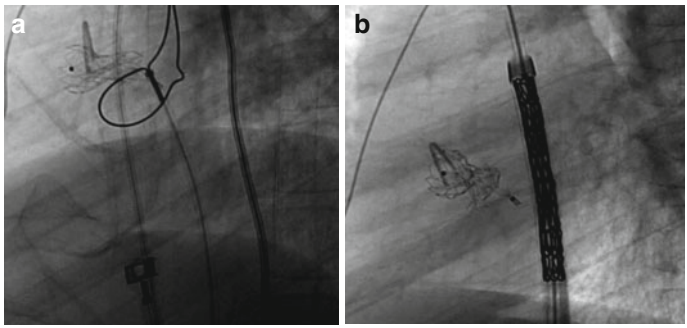

Fig. 27.4 Stented fenestration with previous attempt of closure with an Amplatzer duct occluder type II; now complete closure with a covered CP stent (see text)

- Future reclosing of these stented fenestrations after patients have improved haemodynamically might pose some challenges. Figure 27.4a depicts such a stented fenestration that had to be created postoperatively in a patient due to prolonged chylothorax after early spontaneous closure of the fenestration. Ten months later, an Amplatzer duct occluder II was implanted within the stent, but 3 years later, saturations persisted below 88 % due to residual right-to-left shunting. Clinical and haemodynamic evaluations were favourable for complete fenestration closure, but technically the procedure proved to be challenging. The distal (conduit) part of the stent was snared from the femoral side and gradually pulled caudally against the conduit wall, to prevent sharp edges sticking into the conduit (Fig. 27.4a). The stent was then forced even more against the conduit from cranially to caudally by inflating a 20-mm Atlas balloon, also testing for the risk of balloon perforation due to residual sharp edges. Finally, a 45-mm covered CP stent was implanted using a 22-mm BIB (Fig. 27.4b), obtaining complete closure of the fenestration and a non-obstructive conduit.

27.11 Devices for Partial Occlusion

- In some patients with suboptimal Fontan physiology, the fenestration may be too large in the early postoperative period in a patient not yet stable enough for complete closure of the fenestration. The possibility to partially close such a fenestration could be an attractive option in this setting.
- A customized fenestrated atrial septal occluder device has been used in few patients; however, the incidence of spontaneous closure of the fenestration in the immediate follow-up period was high. We described a partial occluder, the 115S PFO star (CARDIA ™), designed by removing two opposite quadrants from the right atrial disc. These 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). In the 18 patients in which the partial occluder was implanted, mild to moderate residual shunting remained in all but two after 1 month. Six months after device implantation, residual shunting was still documented by echocardiography in 12 of these patients (saturations 90 % ± 3 %). Closure of these shunts should be technically feasible using coils or a covered stent when indicated.

27.12 Follow-Up After Fenestration Closure

- Patients should be routinely evaluated (clinically and echocardiographically) 24 h, 1 month and 6 months after the intervention with specific attention to clinical signs of venous congestion or low output and evidence of thrombus or residual shunt on TTE. Complete closure is defined as improved saturations clinically and the absence of clinically significant shunt on colour.

- Transthoracic echocardiography may fail to detect some thrombi, and although routine transoesophageal echocardiography is more invasive, it should probably be considered in certain high-risk patients.
- The optimal anticoagulation regimen after Fontan completion is unknown. Previous reports have shown an incidence of 20–23 % of thrombus formation in the extracardiac conduit if anticoagulants were not given. Patients with a persistent right-to-left shunt and a tendency to form venous thrombi may be at increased risk for paradoxical embolic events and device occlusion of the fenestration may decrease the risks of systemic thromboembolisation. Treatment protocol before and after fenestration closure differs depending on institutional protocol and risk stratification in individual patients.
- In our institution all patients with a fenestrated Fontan circulation are treated with acetylsalicylic acid 1–2 mg/kg/day orally in combination with clopidogrel 0.2 mg/kg/day orally; the clopidogrel is usually stopped 6 months after fenestration closure, except in patients with "unfavourable" haemodynamics. In the event of previous thrombosis or high-risk patients, lifelong treatment with Coumadin is used aiming for a target prothrombin time of 1.5–2.

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