Patient with Persistent Pleural Effusion, Pulmonary Branch Stenosis and Failed Fontan Circulation: “Headache” Resolved With Intervention

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Abstract
Failed Fontan is a serious condition that has high morbidity and mortality. We report the case of a 6-year-old girl with Failing Fontan with persistent pleural effusion for 10 weeks, secondary to anterograde flow and mild stenosis of the left branch pulmonary artery. Covered stent placement was delivered to occlude anterograde flow and uniform pulmonary artery diameter. Effusion resolution was achieved five days after the intervention and discharged ten days later.

Keywords: Pleural Effusion; Pulmonary Branch Stenosis; Fontan Circulation

Introduction
At present, Fontan surgery remains the option that provides the greatest survival to patients with univentricular physiology; its success is based on a thorough evaluation of the hemodynamics of patient prior to surgery. Despite having ideal hemodynamic parameters, the hemodynamic changes involved in total cavopulmonary procedure are not well tolerated in some patients, leading to complications ranging from a slight decrease in oxygen saturation to definitive failure of the Fontan circulation [1]. Pleural effusions continue to present challenges after this operation and account for increased length of stay and time with draining tube thoracostomies [2]. Gupta A et al. [3] in 2004 described significant risk factors for developing this complication such as low preoperative SaO2, prolonged periods of extracorporeal circulation during surgery, diaphragmatic paralysis and the presence of postoperative infections.

Case Presentation
We present the case of a patient with double discordance, left atrioventricular valve atresia, restrictive bulboventricular foramen and hypoplastic right ventricle (HRV). To guarantee systemic flow, an aortopulmonary window with pulmonary artery banding above the aortopulmonary window (APW) was performed. At 4 years of age, Glenn’s surgery was performed without complications and anterograde flow was intentionally maintained through the pulmonary artery. At 6 years, prior to Fontan procedure, the patient’s hemodynamics were evaluated: mean pulmonary artery (PA): 10 mmHg, left ventricle (LV): 80/8 mmHg, pulmonary vascular resistance (PVR) 1.2 Wood units (WU), pulmonary branches with normal Z values; it was thus decided to complete the Fontan procedure. An extracardiac Fontan surgery was performed with fenestration and at the same surgical time, pulmonary anterograde flow was excluded by ligating the pulmonary artery trunk.

The patient presented a satisfactory immediate postoperative evolution and was able to extubate herself in the first 48 hr, but was maintained with high volumes of right pleural effusion. The echocardiogram shows anterograde flow through the PA and mild stenosis in the proximal third of the right pulmonary artery (RPA). Several strategies were implemented with medical management to reduce the pleural drainage. Intermittent improvement was achieved without complete remission of the pleural effusion. To complete the patient’s study, a CT angiography was requested, which showed anatomical distortion of the confluence of the pulmonary branches with RPA stenosis between the confluence and Glenn anastomosis; the pulmonary trunk permeability was also corroborated. It was decided to perform catheterization with the aim of evaluating the hemodynamics of the Fontan system. The catheterization revealed the following: -Anatomic findings: Pulmonary confluence of 10.6 mm (Z = 0.17), reduction of the left...
pulmonary branch (LPA) diameter to 6 mm (Z -4.4) and the RPA of 8.5 mm (Z -2.6) proximal to the anastomosis of both cava, Non- permeable fenestration, preferential flow of both cava to the right lung (Figure 1). Hemodynamic findings: Mean PA pressure 20 mmHg, LPA in its distal portion 15 mmHg, ventricular end-diastolic pressure of 9 mmHg, anterograde flow through the APW to the left branch; this flow was selective towards the left lung and competes with the drainage of the Fontan system (Figure 2A).

Both a non-compliant balloon occlusion test (simulating the stent placement) and an ascending aortic angiogram were simultaneously performed to confirm the absence of passage of contrast to the PA trunk through the APW. Inferior Vena Cava (IVC) and Superior Vena Cava (SVC) angiography was performed to verify the free venous drainage of Fontan circulation (Figure 2 B,C). In view of the stenosis presented and after evaluating both the feasibility and benefit of covered stent placement, we decided to place CP Stent ™ covered 10 Zigs of 39 mm. A 12 × 40 mm POWERFLEX® Pro balloon was used to release the stent and then dilated to 14 mm with a MAXI LD™ balloon without complications, thus resolving pulmonary branch stenosis and total occlusion of the anterograde flow (Figure 3). After 72 hr of the intervention, the pleural tube drainage was minimal; the patient was able to leave the hospital 10 days after the intervention.

Discussion

Glenn circulation associated with accessory flow is generally well tolerated, improves oxygen saturation and prevents the onset of collateral circulation; there are groups opting for this therapy with good results [4,5]. Keeping anterograde or accessory flow after completion of cavopulmonary anastomosis is a totally abnormal condition and can generate, among other complications, persistent pleural effusion as in the presented patient.

Persistent pleural effusion in Fontan patients continues to be a real challenge. Some research groups suggest that fenestration reduces the frequency of pleural effusions [6,7]. However, other authors have not found statistically significant changes with the use of fenestra [3]. New medical management protocols have been developed to reduce pleural effusion with favorable results [8].

Recently, new therapeutic strategies have been chosen in order to resolve or improve the conditions in patients with Fontan failure using mechanical assistance in IVC and cases of percutaneous valve implantation in IVC, even with inconclusive results [9,10]. It is advisable to exhaust all possibilities of medical treatment before considering catheterization or any invasive procedure.

Similar cases have been reported in the literature, where the cause of pleural effusion is resolved through intervention, using
Amplatzer® ASO devices and stents when the problem is accessory flow or pulmonary branch stenosis [11,12].

In the present case, the persistent effusion was generated by the pulmonary branch stenosis and the accessory flow that increased the pressure nonuniformly in the Fontan system. Consequently, through a covered stent, the two problems were solved with a single device; we considered that the occlusion test was useful to evaluate the position and function of the stent without the need to release it and to simulate the final result. It completely solved the problem and a new surgical intervention was avoided, which involves general anesthesia, sternotomy, bleeding, a cardiopulmonary bypass and postoperative intensive therapy. The patient did not require intensive therapy and was discharged from the hospital within 10 days after the procedure. One year after the procedure the patient is asymptomatic and maintains an oxygen saturation of 94%.

Conclusion

There are numerous manifestations that can be presented in a “failing” Fontan circulation: persistent pleural effusion is one of the most frequent. In all the “failing” Fontan patients, an early catheterization should be performed, as discussed in this case. This procedure solved the hemodynamic problem with a percutaneous intervention, thus achieving the discharge of the patient after a long hospital stay.

References


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