Complications after a Bidirectional Cavopulmonary Anastomosis with Accessory Sources of Pulmonary Blood Flow

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Abstract

Keywords

- fistula
- superior vena cava syndrome
- ulceration

Creating an accessory source of pulsatile pulmonary blood flow in a patient with a bidirectional cavopulmonary anastomosis may have advantages and disadvantages. In relation to the latter, we report the complications seen in a cyanotic congenital heart disease patient who developed a superior vena cava syndrome plus severe swelling of his right hand that evolved satisfactorily after percutaneous and medical treatment.

The bidirectional cavopulmonary anastomosis, or the bidirectional Glenn shunt as it is sometimes named, is a surgical procedure performed in cyanotic patients with left or right heart hypoplasia to whom a complete two ventricle circulations cannot be done. However, it generally should not be the sole source of pulmonary blood flow and additional pulmonary blood flow via a native pulmonary stenosis, a pulmonary artery band, a concomitant systemic-to-pulmonary artery shunt or a staged procedure toward the Fontan completion may be necessary if an increase in systemic oxygenation is required.

Case Report

A 28-year-old male patient with cyanotic congenital heart disease with a pulmonary atresia and an intact ventricular septum, who was operated with a left modified Blalock Taussig shunt when he was 10 days of age. Four years later, the patient underwent a bidirectional cavopulmonary anastomosis and at the age of 8 years, a Potts shunt was performed to treat progressive cyanosis and exertional dyspnea. Fontan completion was not achieved due to pulmonary hypertension. Within this context of progressive cyanosis and dyspnea, at the age of 9 years, a Waterston shunt was performed and later on, at the age of 24 years, an axillary arteriovenous

fistula between the right subclavian artery and the right subclavian vein was created (basal oxygen saturation of 81%).

Three years later of this last procedure, the patient developed right upper limb edema secondary to right subclavian vein stenosis which was treated by angioplasty and stent implantation. However, 1 year later, due to previous stent restenosis, a new bare-metal stent implantation was needed in association with angioplasty at the anastomosis between the superior vena cava and the right pulmonary artery. After percutaneous treatment, right brachiocephalic vein pressure dropped from 55 to 27 mm Hg and the mean pressures registered in the superior vena cava and the right pulmonary artery were 22 and 18 mm Hg, respectively.

Treatment with aspirin (325 mg/24 hours) plus warfarin was initiated after stent implantation while tadalafil (20 mg twice daily), which was later changed to bosentan (125 mg every 12 hours), was prescribed for the treatment of pulmonary hypertension. Also, oral levofloxacin (500 mg/24 hours) and oral linezolid (600 mg/12 hours) during 15 days was scheduled due to the ulceration of the second finger of his right hand.

Four months after the percutaneous procedure, the patient has improved, very significantly, his symptoms of facial edema and right upper limb venous congestion while the ulceration of his right index finger has healed (► **Fig. 1A, B**).

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Fig. 1 (A) Right upper limb edema with ulceration and cellulitis of his right index finger before the percutaneous and medical treatment. (B) Four months after the percutaneous procedure, showing significant improvement of the venous congestion of his right hand and the ulceration of his right index finger.

Physical examination shows bilateral machine-like systolic-diastolic murmur at the supraclavicular levels while blood analysis evidences a hemoglobin of 20 g/dL, a hematocrit level of 60.5%, and a basal hemoglobin oxygen saturation of 84%. Echocardiography exhibits a pulmonary valve atresia, a hypoplastic right ventricle with a preserved left ventricular ejection fraction (50%), a functioning Glenn shunt, and the patency of the right and left aortopulmonary fistulas. Meanwhile, chest magnetic resonance angiography shows patency plus multiple aneurysmal dilatations of the right subclavian, axillary, and brachial veins (► **Fig. 2**).

Discussion

Bidirectional cavopulmonary anastomosis is typically performed as one step of the staging procedure in preparation for a Fontan surgery or as a long-term palliation for high-risk Fontan candidates. However, bidirectional cavopulmonary anastomosis can also provide by itself adequate pulmonary

blood flow in most patients with single ventricular heart. For this reason, some authors promote the idea that patients with functionally single ventricles should remain palliated for an extended period of time with a bidirectional Glenn procedure alone¹ due to the increased risk of stroke, protein losing enteropathy, and arrhythmias, without an improvement in survival, seen in Fontan patients. Also, and because many of these patients develop cyanosis, other authors propose leaving or creating an accessory source of pulsatile pulmonary blood flow after performing a classic Glenn or a bidirectional cavopulmonary anastomosis, especially when the patient is a suboptimal candidate for a Fontan procedure, one wants to obtain a definitive palliation or one wishes to offer an effective bridge to transplantation.²

The advantages of leaving an accessory source of pulsatile pulmonary blood flow include the improvement in postoperative arterial oxygen saturation, the promotion of pulmonary arteries' growth, and prevention of pulmonary arteriovenous malformations development.³ Some authors

Fig. 2 (A) Coronal magnetic resonance angiography of the thoracic vasculature showing patency of the right subclavian vein, the right internal jugular vein, the SVC, and the RPA. Multiple aneurysmal dilatations of the right subclavian, axillary, and brachial veins (arrowhead) are also seen. The asterisk shows where the stent is implanted in the right subclavian vein. (B) Coronal magnetic resonance angiography of the thoracic vasculature showing an enlarged and dilated azygos vein (double arrowhead) resulting from a SVC obstruction distal to the azygos entry. Ao, aorta; RPA, right pulmonary artery; SVC, superior vena cava.

advocate performing Glenn shunts with associated pulsatile pulmonary blood flow to improve the clinical status, reduce hospital mortality rates, and diminish the incidence of early reoperations due to hypoxemia compared with Glenn anastomosis.⁴ However, this therapeutic approach may favor pulmonary hypertension, superior vena cava syndrome, volume overload on the single ventricle, persistent pleural effusions, longer hospital stays, higher morbidity and mortality, and the completion of a second stage of Fontan procedure.⁵⁻⁷

Creation of an axillary arteriovenous fistula to augment pulmonary blood flow after a cavopulmonary shunt may also provide useful palliation for complex cyanotic congenital heart diseases when other options are limited or previous fistulae are insufficient to maintain a suitable arterial oxygen saturation.⁸ However, axillary arteriovenous fistulae may also lead to potential disadvantages such as volume load to the heart, pulmonary hypertension, shunt restenosis or thrombosis, endocarditis, and facial or arm swelling due to a malfunction of the anastomosis. This last drawback seems to be related to a perianastomotic mismatch in the elastic properties and the physical trauma associated with cutting and suturing veins and arteries that favor progressive stenoses at the venous anastomotic side.

In relation to cyanosis improvement after axillary arteriovenous fistulae, the data are controversial. On the one hand, Magee et al⁹ found, in 10 patients unsuitable for definitive repair, that the creation of an ipsilateral axillary arteriovenous fistula favored a rise in mean oxygen saturations from 80% before operation to 84% at a mean follow-up interval of 7.4 years. On the other hand, Quiñonez et al¹⁰ showed, in 11 patients with axillary arteriovenous fistula, that this type of shunt did not improve cyanosis, functional class, or polycythemia.

Although percutaneous and pharmacological treatments may improve complications after axillary arteriovenous fistulae, we should only perform them in those rare patients

with intractable cyanosis for whom Fontan completion or transplantation are unappealing.

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