Surgical correction of functional univentricular heart diseases: spectrum of imaging findings

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Objectives

The learning objectives presented in this study are:

- Review the different surgical options in patients with functionally univentricular heart disease, mainly Glenn and Fontan procedures, and describe the technical aspects of these surgical techniques and their main indications.

- Illustrate the expected normal post-surgical anatomy.

- Describe the potential complications of these procedures, with emphasis on the extracardiac complications, and the spectrum of radiological findings.

Body

DESCRIPTION OF THE SURGICAL TECHNIQUES AND THEIR INDICATIONS

Some congenital heart diseases show impairment of the normal biventricular physiology. In normal circumstances, pulmonary and systemic circulations are driven by their respective ventricular "pump" but there are several congenital heart diseases characterized by having a functional single ventricular chamber. This single ventricle may be real, existing morphological alteration with absence or hypoplasia of one of them (as in the hypoplastic left heart syndrome), or functional, when a biventricular morphology exists but only one ventricle is functional (as in tricuspid atresia or pulmonary atresia with intact ventricular septum).

Physiologically, the single ventricle will receive the flow of both pulmonary and systemic circulations with the subsequent volume overload and a problem in long-term function.

Although definitive treatment of these patients is cardiac transplantation, there are several palliative surgical techniques that have prevented ventricular volume overload diverting part or all of the systemic venous circulation into the pulmonary arteries, and therefore bypassing the single ventricular chamber. The result obtained is a single ventricle that functions both as suction and expelling pump. Low pulmonary and telediastolic resistances are essential for optimum performance of this new hemodynamic system.

One of the first techniques described was **Glenn procedure**. The classic technique is the end-lateral anastomosis of the superior vein cava (SVC) before its entry into the right atrium, to the distal right pulmonary artery (RPA) (Fig. 1 on page 5). The
entire flow of the SVC is directed towards the pulmonary arteries bypassing the right heart, avoiding volume overload and keeping the system functionality. This technique is nowadays normally used as a previous procedure to Fontan correction.

In the 70s Fontan described a new surgical technique with the same aim of reducing ventricular overload redirecting systemic venous flow into the pulmonary circulation and preventing its passage through the ventricle. The original technique was first described for the treatment of tricuspid atresia and consisted on, besides the connection created after a Glenn procedure, a total cavo-pulmonary connection by means of an anastomosis of the right atrium to the pulmonary arteries. (Fig. 2 on page 6).

The significant development of different surgical procedures and the advent of new prosthetic materials in recent years have contributed to the emergence of numerous variations and adaptations of this technique. Currently the original technique has been replaced by other procedures that seek to establish a direct connection between the venous flow of both veins cava superior and inferior (SVC and IVC) and the pulmonary circulation, thus preventing the flow passage through the right atrium and problems associated with it, such as arrhythmias or thrombosis.

Modern Fontan procedure consists in the anastomosis of both SVC and IVC to the RPA. A prosthetic conduit is used to connect the IVC to the RPA. This conduit can be located intraatrial or extracardiac (Fig. 3 on page 8). The total cavo-pulmonary connection should not be done at one time, as increased pulmonary flow occurs abruptly and can cause pulmonary congestion and insidious pleural effusion. To avoid this, a bidirectional Glenn -or hemi-Fontan- (without the section of the RPA as was done in the classic Glenn) must be performed as a first step.

The main indications for these procedures are those congenital heart defects that result in a functional single ventricle. The most important examples are:

- **Tricuspid atresia:** the lack of communication between the right cavities requires a right to left shunt through an atrial septal defect and thus all the volume of the systemic and pulmonary circulation will end into the left ventricle. The patent ductus arteriosus will permit passage into the pulmonary circulation.

- **Pulmonary atresia with intact ventricular septum:** this condition is usually associated with varying degrees of right ventricular hypoplasia and tricuspid atresia. The absence of right ventricular outflow forces again the systemic flow passage through an atrial septal defect to the left cavities. Again, the patent ductus arteriosus will permit passage into the pulmonary circulation. In both cases there is left ventricular overload.

- **Mitral atresia:** due to the absence of communication between the left chambers, a left to right shunt is established through an atrial septal defect. The ductus arteriosus must be permeable to allow the blood passage to the aorta and systemic circulation.
- Hypoplastic left heart syndrome: in these patients there is no functional left chambers. The passage of blood into the systemic circulation also occurs through the ductus arteriosus, from the pulmonary artery to the aorta. Blood from the pulmonary circulation to the right atrium passes through an atrial septal defect, establishing a left to right shunt. In the last two cases the right ventricle is the one overloaded.

**COMPLICATIONS**

Late complications arising from the new hemodynamic and physiological conditions created after the performance of these procedures are observed with increasing frequency due to increased survival of these patients. Currently, survival rates are around 85% at 20 years. There are risk factors that predict a worse outcome and more frequent occurrence of complications such as high pulmonary pressure, anatomical alterations in the pulmonary arteries or ventricular dysfunction. Imaging tests (CT and MRI) are essential in monitoring these patients, allowing functional assessment and an early detection of complications.

Complications that can occur in these patients are very diverse, and can be cardiac and extracardiac. In the heart, the most common complication after the classic Fontan technique is atrial enlargement (Fig. 2 on page 6), that can lead with high frequency to arrhythmias and thromboembolic problems.

There is a wide range of extracardiac complications occurring in these patients and imaging plays an important role in the early diagnosis. The radiologist must therefore recognize the spectrum of imaging findings which include:

1. **Hemodynamic complications.**
   The low pressures achieved after the Fontan repair paradoxically condition an increase in the pulmonary vascular resistance. This complication is attributed to the absence of pulsatility of the pulmonary arteries and the inability to achieve complete filling of the pulmonary vasculature. Pulmonary vascular resistance is a determinant factor in the cardiac output of this new hemodynamic condition.

   Pulmonary vascular resistance may also be affected by morphological alterations in the pulmonary arteries, mainly stenosis (Fig. 4 on page 8), that sometimes occur at the site of anastomosis. Its detection and early treatment can improve system functionality and can determine a better prognosis.

2. **Right to left shunts.**
   Right to left shunts may occur due to the development of pulmonary arterio-venous fistulae and the formation of systemic-pulmonary venous collaterals.

   Although the etiology is not completely established, the absence of arterial pulsatility and hepatoenteric inflow in the lungs appear to be associated with an increased risk
of developing pulmonary arterio-venous fistulas (Fig. 5 on page 9, Fig. 6 on page 9).

Veno-venous collateral development between the systemic and pulmonary circulation is a consequence of the existence of an elevated central venous pressure (Fig. 5 on page 9, Fig. 6 on page 9, Fig. 7 on page 10, Fig. 8 on page 10, Fig. 9 on page 12B).

These conditions entail a right to left shunt and may cause desaturation and cyanosis.

3. Liver complications.
Increased systemic venous pressure causes a passive venous congestion in the liver that has negative effects on the parenchyma. The increase in venous pressure causes an increase in retrograde pressure in the hepatic sinusoids which can develop centrilobular necrosis, fibrosis, cirrhosis and portal hypertension (Fig. 9 on page 12, Fig. 10 on page 12). Liver dysfunction, splenomegaly, ascites and porto-systemic shunts and collateral veins may occur.

These patients may develop cirrhosis of cardiac origin and, as in all other causes of cirrhosis, dysplastic nodules and hepatocarcinomas.

4. Lymphatic complications.
The increased venous pressure may also affect the lymphatic circulation.

In the lungs, the increased pressure in the lymphatic circulation can cause interstitial edema, and pleural and pericardial effusion. Plastic bronchitis is another potential complication of lymphatic dysfunction that, although rare, can be severe. Hypersecretion of mucin influenced by alterations in lymphatic drainage and a low cardiac output are factors related to the formation of large and dense secretions that impact into the airway and may produce atelectasis. Clinically, patients present with respiratory symptoms, such as cough and expectoration.

The most relevant abdominal complications derived from the increased lymphatic pressure are ascites and protein losing enteropathy (Fig. 11 on page 12). The latter, although it is rare and its etiology is not defined, is associated with venous and lymphatic congestion that occurs in the splanchnic circuit and causing a loss of protein into the intestinal lumen with hypoalbuminemia and immunodeficiencies. The hypoproteinemia may also be the cause of pleural effusion and ascites. Patients presenting with protein losing enteropathy have a poor prognosis, and it is a potentially lethal disease.

Images for this section:
Fig. 1: 15-year-old male with Glenn correction. Coronal MRA image shows the connection performed between superior vein cava and right pulmonary artery (arrow).
**Fig. 2:** 27-year-old female with classic Fontan procedure for tricuspid atresia correction. Connection between right atrium and right pulmonary artery (arrow) and moderate dilatation of the right atrium are seen on this angio-MR image.

**Fig. 3:** Total cavo-pulmonary connection with extracardiac Fontan conduit (arrow).
**Fig. 4:** Left pulmonary artery stenosis (arrow) in a patient after been performed a Fontan procedure. This image also shows a significant dilatation of the aortic root (arrowhead).

**Fig. 5:** 20-year-old woman with multiple PAVMs and VVS. Contrast-enhanced thoracic CT image shows countless small PAVMs in the RUL (arrows) (A). Strong and early contrast opacification of the right upper pulmonary vein is seen (B) due to the presence of PAVMs. Abundant collateral circulation is also seen within the mediastinum representing several systemic-pulmonary veno-venous shunts which connect the left subclavian vein and SVC (arrowheads) with right and left superior pulmonary veins (arrows) (B). Arteriography confirms the presence of PAVMs (C). The patient presented with cyanosis and oxygen saturation of 82%.
Fig. 6: 27-year-old woman with classic Fontan procedure for tricuspid atresia correction (same patient as Fig 2). Contrast enhanced thoracic CT images show several peripheral PAVM in the LUL (yellow arrows) and VVS (white arrow).

Fig. 7: 20-year-old female with multiple mediastinal venous collaterals secondary to veno-venous shunts connecting left subclavian vein and SVC (yellow arrows) to both superior pulmonary veins (white arrows).
Fig. 8: 19-year-old male with tricuspid atresia corrected with a Fontan circuit. Contrast-enhanced MRA (MIP reconstruction) shows veno-venous shunts between left subclavian vein (yellow arrow) and left superior pulmonary vein (white arrow).

Fig. 9: 16-year-old male who developed several extracardiac complications after Fontan procedure. CT images show signs of chronic hepatopathy due to liver congestion, cirrhosis and portal hypertension with ascites and splenomegaly. Prominent venous collaterals (arrow) and connection between left hepatic vein and lower left pulmonary vein can be also seen.

Fig. 10: Congestive hepatopathy after Fontan correction in two different patients. Heterogeneity and peripheral perfusion alterations are seen on A and B. Extracardiac Fontan conduit can also be seen on B (arrow).
**Fig. 11:** Protein-losing enteropathy in a patient with liver cirrhosis and anasarca (same patient as in figure 10). CT shows mural thickening of small bowel loops, free fluid and edema in the subcutaneous tissue.
Conclusions

Fontan procedure is the main palliative surgical procedure in congenital heart diseases that result in a functional univentricular heart. Its main objective is to redirect the venous flow from the systemic circulation into the pulmonary circulation, bypassing the ventricular chamber and, consequently, avoiding the volume overload that this flow would suppose.

The importance of imaging in the detection of the potential complications that can happen after surgical correction of univentricular heart diseases requires the radiologist to be familiar with these circumstances.