Chapter 44
Evaluations Before Partial and Total Cavopulmonary Connections

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44.1 Anatomic Description and Physiopathology

Patients with single ventricle physiology have a variety of complex heart diseases that are not suitable to biventricular repair. Fontan circulation can be obtained by performing an atriopulmonary connection or a total cavopulmonary connection (TCPC). Both allow the passive flow of the systemic venous blood into the lungs. In the completed Fontan state, the pressures in the caval and pulmonary circulations must be high enough to ensure flow through the lungs and adequate preload of the left ventricle whilst avoiding high-pressure venous congestion. This goal is achieved when pulmonary arterial pressure (PAP) is between 10 and 14 mmHg. Partial cavopulmonary connection (PCPC), with or without additional pulmonary blood flow (PBF), usually precedes conversion to TCPC. PCPC forces 50 % of cardiac output to bypass the heart and directly enter the lungs, increasing oxygen saturation.

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44.2 Clinical Scenarios

Patients with single ventricle physiology can have duct-dependent pulmonary or systemic circulation and a protected or non-protected pulmonary vascular bed.

(a) Patients with duct-dependent pulmonary circulation or insufficient PBF need either a BT shunt or a ductal stenting.
(b) Patients with duct-dependent systemic circulation need a Norwood operation or a hybrid treatment (Chap. 39).
(c) When obstruction to the systemic outflow exists, a Damus-Kaye anastomosis bypasses the subaortic obstruction, by creating an aortopulmonary anastomosis.
(d) Patients with excessive PBF require pulmonary artery banding.

During follow-up there different scenarios possible:

1. Neonates with adequate PBF can reach the age of 6 months without a neonatal palliation and receive a PCPC as a first intervention.
2. Neonates with pulmonary banding or BT shunt-dependent pulmonary circulation display progressive cyanosis and generally need PCPC at the age of 4–6 months.
   An additional source of PBF such as BT shunt or native antegrade flow can be occluded or left in place at time of PCPC, in order to prepare the child for a TCPC at an older age.
3. TCPC is generally performed after the age of 3 years, depending on the degree of systemic desaturation. At the time of TCPC, any additional source of PBF is removed either surgically or in the cardiac catheterisation laboratory.

44.3 Indications and Patient Selection

PCPC and TCPC are often preceded by cardiac catheterisation, aimed at measuring PAP, assessing pulmonary artery size and treating possible associated anomalies. Catheterisation is performed in either all or selected patients, according to the team policy and depending on the availability of high-quality noninvasive imaging.

Patients in whom catheterisation is commonly performed prior to PCPC include:

- Subject patients with hypoplastic left heart syndrome (having received either Norwood I or hybrid treatment).
- Patients in whom the anatomy of pulmonary arteries needs to be clarified or in whom PAP might be high.

Prior to TCPC a cardiac catheterisation should be considered in:

- Patients with hypoplastic left heart syndrome having had Norwood II (surgical or hybrid treatment).
- Patients with additional PBF.
- Patients in whom the anatomy of pulmonary artery needs to be clarified or in whom PAP might be high.
- Commonly associated anomalies that can be treated percutaneously are aortic recoarctation, aortopulmonary collaterals, restrictive foramen ovale and stenosis of pulmonary arteries or of the superior vena cava.

44.4 Imaging

Noninvasive imaging can provide definitive information on pulmonary artery anatomy (Fig. 44.1).

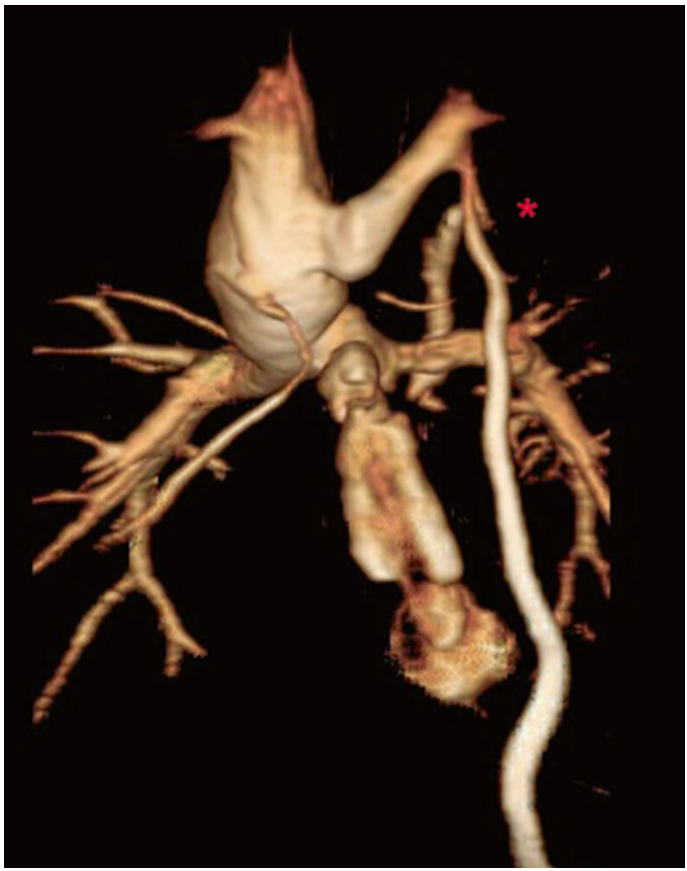

Fig. 44.1 3D MRI in AP view shows a dilated superior vena cava, the presence of antegrade flow and a venovenous collateral (*)

In addition, it can offer information on intracardiac and extracardiac structures, display unsuspected anomalies and offer accurate data on ventricular function.

Radiologic imaging, in order to be valuable, has to be of high quality.

Operators should be skilled at obtaining and interpreting the images of the hearts with congenital anomalies.

44.5 Pre-PCPC Catheterisation

Vascular access, technique and materials can vary in accordance with the anatomy.

The aim of the exam is to measure PAP, assess the anatomy of pulmonary arteries and rule out or treat associated anomalies.

Intravenous heparin at a dose of 50–100 UI/kg should be administered.

Arterial access is generally needed to visualise pulmonary arteries in patients with pulmonary atresia and a BT shunt.

Angiography rules out the presence of aortic coarctation and aortopulmonary collaterals.

If the shunt cannot be entered, PAP can be measured via pulmonary veins [2]. We penetrate a pulmonary vein until the catheter is wedged and the shape of the pressure curve changes and a transpulmonary gradient appears.

The same approach is used in patients with inadequate antegrade PBF and a BT shunt.

In our view, it is easier and safer to measure PAP via pulmonary veins rather than via a small BT shunt or a small native pulmonary outflow.

Patients with hypoplastic left heart syndrome are complex patients who need a complete and precise evaluation.

In subjects after a Norwood I operation, venous and arterial access should be obtained. In some cases it is possible to obtain

a complete evaluation via venous access alone. The catheter enters the right atrium, and then the left atrial pressure and pulmonary vein wedge pressure are obtained. Then ventricular pressure is measured.

The catheter enters the neoaorta and is guided into the descending aorta. If difficulty is encountered when trying to enter the neoaorta or the child shows instability, this approach should be abandoned and the arterial approach should be adopted.

An angiography should be performed into the BT shunt to visualise pulmonary arteries. It is often difficult to enter the brachiocephalic artery using venous access. Arterial access should be used in those cases. Possible coexisting problems are recoarctation and aortopulmonary collaterals. Both conditions can be treated, if necessary.

Patients with hypoplastic left heart syndrome having received a hybrid treatment are potentially fragile and can display several associated anomalies such as restrictive atrial septal defect, proximal or distal displacement of the ductal stent, stent thrombosis and preductal or postductal coarctation. The treatment of these lesions, including stenting of the atrial septum and of the native aortic arch, will be discussed in the dedicated chapter.

In these patients, the neoaorta provides pulmonary, systemic and coronary circulations. High-volume angiography (2–3 ml/kg) and appropriate views are necessary to obtain an adequate visualisation of the banded pulmonary arteries, the stented arterial duct and the native aortic arch.

I rarely manage to enter a banded pulmonary artery and generally prefer to measure pulmonary pressure via pulmonary vein wedge pressure [2]. If PAP is high, we need to know if this is due to excessive flow, restrictive atrial septal defect, obstructed pulmonary venous return, recoarctation or ventricular dysfunction. The pressure measurement should be repeated once the associated lesions are treated.

44.6 Pre-TCPC Catheterisation

Catheterisation is performed, according to the presence of forward flow to the lungs, via the femoral vein or the internal jugular vein [1].

Arterial catheterisation can be needed to rule out or treat aortic recoarctation and aortopulmonary collaterals.

If a pulmonary banding or a BT shunt is left in place, the pulmonary artery can be entered via femoral venous access or femoral arterial access.

However, it is generally easier and rapid to reach pulmonary arteries via the internal jugular vein. PAP and either wedge or left atrial pressure are measured.

Pulmonary angiography is performed in a four-chamber view.

If some washout is found, aortopulmonary collaterals should be suspected and aortography should be performed. If a rapid opacification of pulmonary veins is noticed, pulmonary fistulae should be ruled out. The presence of microfistulae can be confirmed by injecting microbubbles obtained by rapidly mixing 80 % blood with 20 % air in both distal pulmonary arteries. Simultaneously, transthoracic or transoesophageal echocardiography is performed. If pulmonary fistulae are present, generally massive opacification of the left atrium is seen.

Post-PCPC patients can have right and left superior vena cava and as such a bilateral bidirectional PCPC may have been performed.

In these patients, the relative size of the superior vena cava should be established. Competitive flow can sometimes prompt the thrombosis of the smaller vena cava that can, if necessary, be reopened using balloon angioplasty and stenting.

Disconnection of pulmonary arteries can also be observed in patients with additional competitive flow (Fig. 44.2).

Any stenosis in the PCPC system must be treated, and collateral vessels connected with the inferior vena cava may need occlusion if they are large before measuring a reliable pressure.

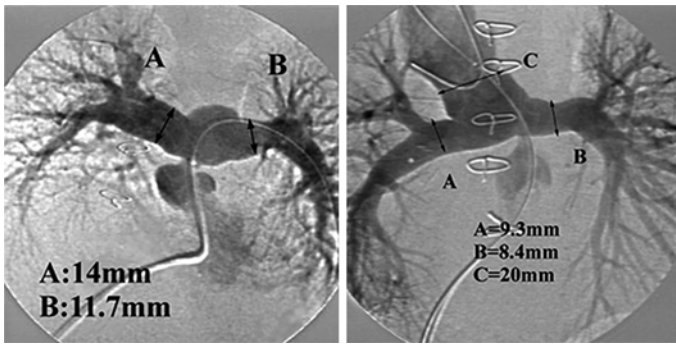

Fig. 44.2 Measurement of pulmonary artery (*double-headed arrows*) size in two different patients before (*left*) and after (*right*) PCPC. In both cases, the catheter enters the pulmonary artery in an antegrade way. In the patient with PCPC, a multi-track catheter (notice the guidewire parallel to the catheter) attains the superior vena cava

When stenosis of the superior vena cava involves the origin of one or both pulmonary arteries, the lesion can be treated percutaneously, using open-cell stents, but can also be treated surgically, at the time of TCPC.

Before TCPC, however, the measurement of pulmonary pressure has to be reliable; therefore, confounding factors that either lower pulmonary pressure (presence of venovenous collaterals and pulmonary fistulae) or increase pulmonary pressure (obstruction to pulmonary venous return, restrictive atrial septal defect, ventricular dysfunction, aortic coarctation, additional flow to the lungs, stenosis/hypoplasia of pulmonary arteries) should be looked for and, if possible, treated.

Depending on the quality of the available noninvasive imaging, the right and left pulmonary artery diameters can be measured immediately before their first branches and are used to calculate cross-sectional areas (Fig. 44.2).

The most useful index to measure pulmonary artery size is the Nakata index.

Accepted values for patients scheduled for TCPC are >200 mm^2/m^2 [3].

44.7 Materials

Diagnostic catheterisation in pre-PCPC and pre-TCPC patients is performed using standard catheters and guidewires, in accordance with the experience of the operators. Open-tip catheters can be more easily manipulated when the anatomy is unusual. However, flow-directed catheters can provide safe manipulation and offer high-quality imaging.

If interventional catheterisation is necessary, various materials are needed.

To occlude the venovenous collaterals, pulmonary fistulae and aortopulmonary collaterals, we use coils, particles, plugs or various devices, according to the anatomy and size of the vessels.

To treat a restrictive atrial septal defect or an intact atrial septum, radiofrequency, balloons, cutting balloons, blade and stents are required.

To treat aortic coarctation and stenosis of pulmonary arteries, we use balloons, cutting balloons and stents.

44.8 Expected Results

The ideal pre-PCPC patient has low PAP and normal pulmonary artery size.

He has non-obstructed pulmonary venous return, non-restrictive atrial septal defect, normal ventricular function and non-obstructed ventricular outflow and does not have aortopulmonary collaterals.

The ideal pre-TCPC patient has mean PAP <14 mmHg, normal pulmonary artery size, normal ventricular function and competent atrioventricular valve(s).

He has no venovenous collaterals or pulmonary fistulae, non-obstructed pulmonary venous return, non-restrictive atrial septal defect and non-obstructed ventricular outflow and does not have aortopulmonary collaterals.

Real patients are often very different from ideal patients. Some degree of ventricular dysfunction, incompetence of atrioventricular valve and small aortopulmonary collaterals can be tolerated. TCPC can be performed also in patients with occluded inferior vena cava. Stenoses in the PCPC anastomosis must however always be treated, either in the catheter laboratory or at the time of surgery.

44.9 Tips and Tricks

Do not forget that PAP can be measured via pulmonary veins. Always obtain pressure measurement before performing a pulmonary angiography to avoid increasing PAP.

Be aware that PAP changes in accordance with aortic pressure.

The simultaneous measurement of pulmonary vein wedge pressure and end-diastolic ventricular pressure can rule out stenosis of pulmonary veins.

Simultaneous angiography in disconnected pulmonary arteries allows the measurement of the distance between disconnected segments (Fig. 44.3).

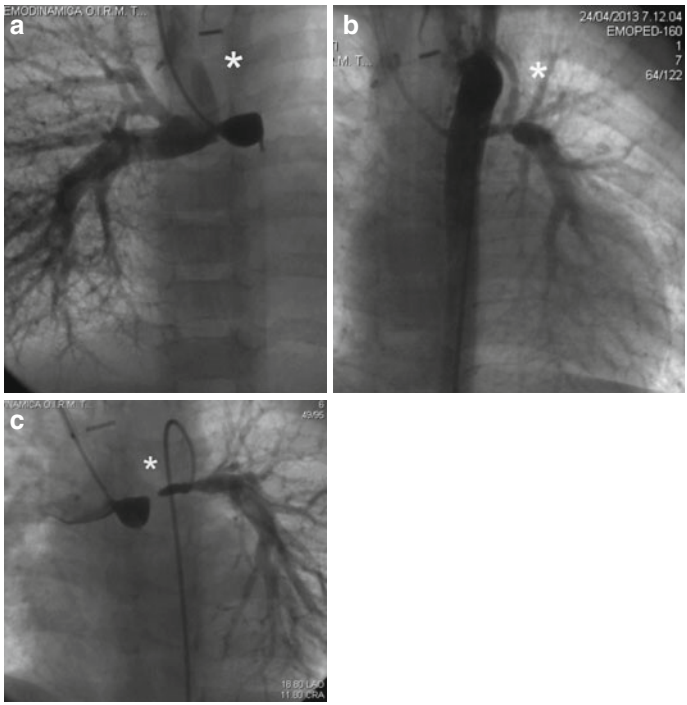

Fig. 44.3 Disconnected pulmonary arteries in a patient with competitive flow. The right pulmonary artery is fed via the superior vena cava (**a***) and the left pulmonary artery via a BT shunt (**b***). Simultaneous injection allows to appreciate the distance between the pulmonary arteries (**c***)

Patients with a single ventricle who had a prolonged stay in intensive care unit can lose femoral venous access.

In patients with antegrade pulmonary flow, an arterial retrograde cardiac catheterisation can be performed from the venous approach.

In patients in whom a persistent left superior vena cava is suspected, a hand injection in a vein of the left arm will easily demonstrate this condition.

To obtain a reliable measure of PAP, we can perform a balloon test occlusion of any additional source of PBF. We have to be aware that we need two vascular accesses or two catheters to measure pressure during test occlusion, unless we use Berman and reversed Berman catheters. However, these catheters have a small balloon and rarely provide a stable occlusion.

44.10 Pitfalls

PAP can be unusually low even when pulmonary arteries are small. In this case always look for anomalies able to lower PAP such as pulmonary fistulae and venovenous collaterals.

44.11 Complications

Patients in whom a diagnostic catheterisation was performed have the general risks of any cardiac catheterisation.

Jugular catheterisation is burdened by the risk of arterial puncture and bleeding.

Rarely, transient atrioventricular block occurs when the catheter is manipulated from the ventricle into the aorta. When this occurs the catheter should be removed from the vein and an arterial approach used. Ventricular pacing is rarely necessary.

Patients in whom an interventional catheterisation was performed are subject to the risks of the respective interventions. There are generic rules and specific rules to manage complica-

tions. Generic rules are be quiet, be logical and avoid useless manoeuvres. Follow the sequence A-B-C-D (A, airway; B, breathing; C, circulation; D, drugs). Specific rules are read in the chapters referring to the respective interventions.

44.12 Post-procedural Care and Follow-Up

Patients having had a diagnostic catheterisation should undergo standard follow-up. In anti-aggregated patients having had a jugular catheterisation, particular attention should be paid to the risk of bleeding. After interventional catheterisation post-procedural care varies in accordance with the intervention performed.

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