

# IMAGES

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## **Cardiac catheter assessment of congenital heart disease prior to total cavopulmonary connection**

JV DeGiovanni<sup>1</sup> and V Grech<sup>2</sup>

<sup>1</sup>Consultant Paediatric Cardiologist, Birmingham Children's Hospital, UK

<sup>2</sup>Consultant Paediatrician (Cardiology), St. Luke's Hospital, Malta

**Contact information:** Dr. Joseph DeGiovanni, Department of Cardiology, The Children's Hospital, Steelhouse Lane, Birmingham B4 6NH, UK Telephone - 0121 3339999 0121 3339999 Fax - 0121 3339441 ; Email: degiovanni@blueyonder.co.uk

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### **Abstract**

This paper summarises the rationale behind cardiac catheter assessment prior to surgical completion of the Fontan circulation in hearts with univentricular pathology.

**MeSH:** Fontan procedure, Heart Catheterization, Heart defects, congenital

Surgery for congenital heart disease (CHD) may be divided into two types: uni- or biventricular. Biventricular repair encompasses situations wherein both ventricles are separately used to support the systemic and pulmonary circuits, such as repair of ventricular septal defect (VSD) or tetralogy of Fallot (figure 1). The term 'biventricular repair' also includes settings wherein a morphologically left ventricle supports the pulmonary circulation and a morphologically right ventricle supports the systemic circulation, such as in congenitally corrected transposition.

Univentricular palliation - also known as 'Fontan' or 'Fontan-type' repair<sup>1</sup> - encompasses all situations wherein, for a variety of reasons, biventricular repair is not possible. The final stage of such repair is usually the culmination of one, or more commonly two previous operations that connect the outflow tracts of one or both ventricles to the aorta with the pulmonary circulation driven without a ventricle (figure 2).

Univentricular palliation may be necessary when the heart cannot be septated, in other words, in the context of a single ventricle (anatomical and/or physiological) such as in tricuspid atresia/pulmonary atresia with intact interventricular septum and hypoplastic RV/hypoplastic left heart syndrome/double inlet ventricle or when biventricular repair is not technically possible such as with straddling atrioventricular valves (AVV) in the setting of a VSD, where closing the VSD would compromise AVV function.

Soon after the initial Fontan experience, Choussat and Fontan listed ten criteria that should ideally be satisfied for optimum low levels of morbidity and mortality with the Fontan procedure.<sup>2</sup> These 10 commandments have been modified and extended over the years and are:

1. Age above four years
2. Normal ventricular function
3. Adequate pulmonary artery size
4. No distortion of pulmonary arteries from prior shunt surgery
5. Low pulmonary artery pressure (below 15 mmHg)
6. Low pulmonary vascular resistance
7. Normal venous drainage
8. No atrioventricular valve leak
9. Normal heart rhythm
10. No right atrial enlargement

Figure 1 Biventricular physiology represented as a circuit diagram

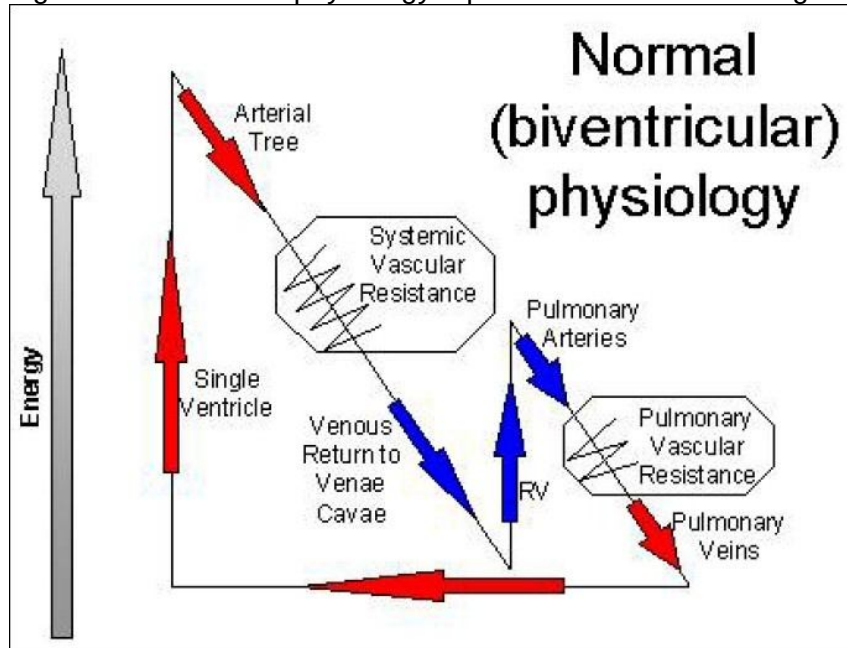
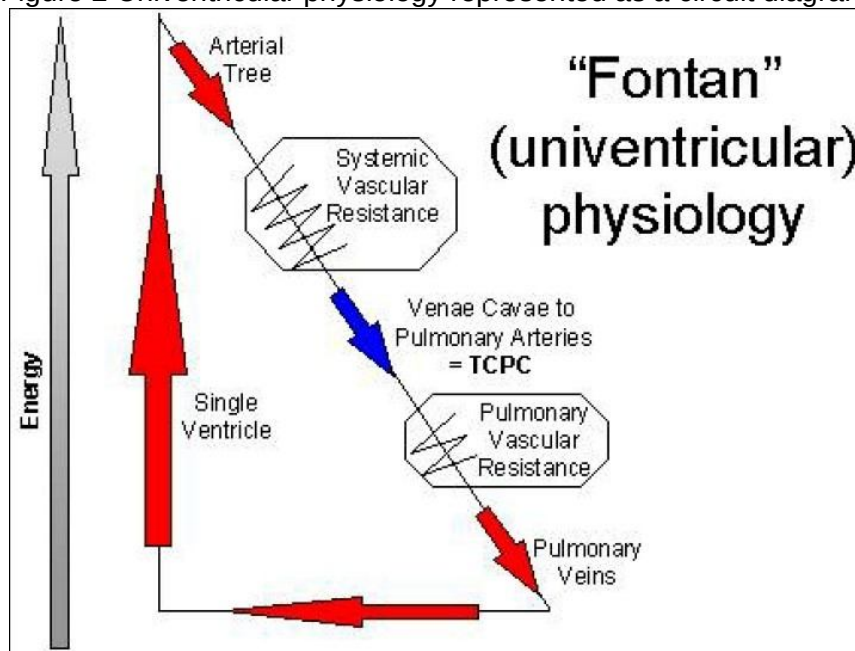


Figure 2 Univentricular physiology represented as a circuit diagram



Overall, these criteria are pre-requisites for smooth functioning of the Fontan circuit. Since its inception, the Fontan procedure has undergone many modifications with the most well known being the total cavopulmonary connection that eliminates the right atrium (or indeed any other cardiac chamber) from the pulmonary circuit, with blood driven through the lungs by central venous pressure and the negative intrathoracic pressure generated during inhalation.<sup>3</sup>

At the time of writing, individuals who undergo univentricular palliation generally undergo three procedures:

1. The first procedure optimises pulmonary blood flow; if pulmonary flow is excessive - with potential heart failure and damage to the pulmonary vascular tree, increasing pulmonary vascular resistance, volume overloading the single ventricle and possibly producing AVV regurgitation and jeopardising the above criteria - it is reduced e.g. by pulmonary artery banding. On the other hand, reduced pulmonary blood flow with cyanosis may be alleviated by means of a systemic to pulmonary artery shunt. Additional procedures may also need to be carried out during the first procedure such as aortic arch repair in associated coarctation or hypoplastic aortic arch.
2. The next operation converts the pulmonary supply from a high systemic pressure source to a more passive and continuous low pressure flow driven by the central venous pressure and enhanced by the negative intra-thoracic pressure during inspiration. This is known as a cavo-pulmonary shunt. It has been shown that morbidity and mortality is lower if the cavo-pulmonary connection is done in two stages with the initial procedure joining the superior vena cava to the right pulmonary artery and referred to as a bi-directional Glenn or bi-directional cavo-pulmonary shunt. If the SVC is joined solely to the right pulmonary artery (which is disconnected from the main pulmonary artery), this is referred to as a classical Glenn. This is, nowadays, very rarely performed. If there are two SVCs, the operation is a bilateral bi-directional Glenn i.e. bilateral bi-directional cavo-pulmonary shunts. Occasionally, the original systemic to pulmonary artery shunt is left in-situ and, rarely, antigrade flow through a restricted pulmonary valve is also left partly to enhance blood flow to the lungs but this is likely to reduce the incidence of pulmonary AV malformations which are known to occur with cavo-pulmonary shunts. This procedure of cavo-pulmonary shunt is carried out between 6 and 9 months after the first stage.
3. The final operation is designed to direct the hepatic and IVC blood flow to the pulmonary circulation and is carried out at a later stage, usually at around the age of four although some have been carried out in some as young as one year or much older depending on local departmental policy and clinical situations. There are various modifications to this final stage including the atrio-pulmonary connection, the lateral tunnel and the extra-cardiac conduit.

Cardiac catheterisation is invariably carried out as part of the preoperative assessment before this final stage of palliation. Catheterisation allows accurate measurements of intracardiac pressures and saturations, assessing not only suitability of such palliation, but also identifying problems that may not be readily identified by echocardiography, such as distal pulmonary artery stenoses. Such lesions may be dealt with at the time of the catheter itself (e.g. ballooning of stenosed pulmonary arteries or coil occlusion of veno-venous or veno-atrial collateral vessels) or at the time of surgery. This article describes the methodology of such catheterisation and all stills and animations displayed here were obtained during assessment of individuals with regard to suitability for TCPC.

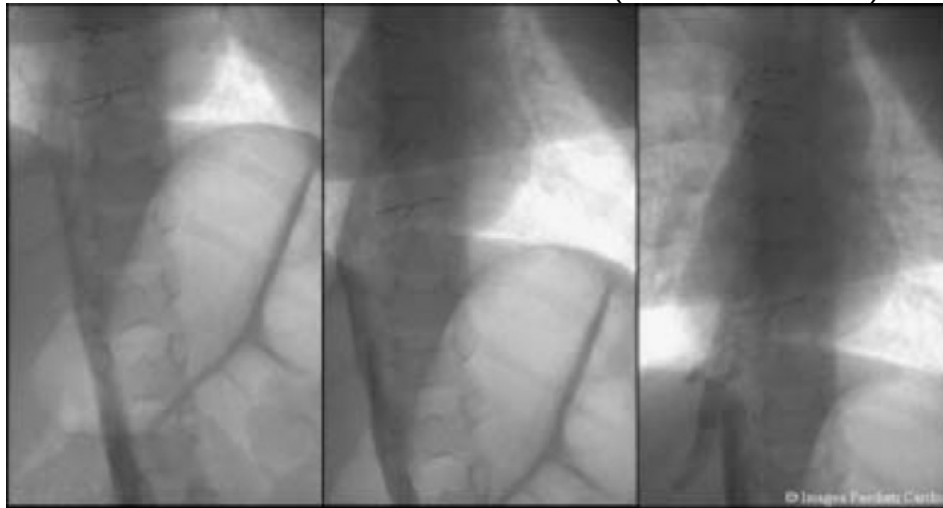
## Methods

Prepare for saturations and pressure run.

Use suitable size multi-hole catheters (e.g. Gensini) for both femoral and internal jugular venous approaches.

In the low IVC check saturation and do a contrast injection to check for collaterals. The IVC is a long structure and one must therefore pan up to the heart in order to follow the contrast during the injection (figure 3).

Figure 3 Hand injection of the inferior vena cava in mesocardia, tricuspid atresia and discordant ventriculo-arterial connections (PA tube orientation).



Enter RA and check saturation and pressure (phasic and mean), cross to the left atrium and make sure that there is no gradient at atrial level at the site of a previous septectomy, then enter the pulmonary veins to ensure patency and no stenosis and, finally, obtain a pulmonary vein wedge as a indirect measure of the pulmonary pressure both on the right and the left.

The ventricular end diastolic pressure is also measured and it is important to make sure that there is no significant AV valve regurgitation and no outflow tract obstruction. In patients who have had a Damus-Kaye Stansil procedure, it is important to make sure that the anastomosis is not stenosed. Saturations should be measured throughout. A left ventriculogram is now performed (figures 4,5).

In the left ventriculogram, it is crucial to distinguish important atrioventricular valve regurgitation that may jeopardize a Fontan procedure from a technical failure (figure 6).

If the patient had had repair of coarctation or hypoplastic arch (as in a Norwood stage 1 procedure), go up the aorta, check saturation and pressure and perform an aortogram in the posteranterior and in the lateral views.

Go down the descending aorta, check pressure and do a pullback to the systemic ventricle and to the atrium. If anatomically possible, cross to the hypoplastic ventricle and check saturation and pressure. In pulmonary atresia with intact septum and hypoplastic RV, look for coronary fistulae.

If part of the repair had included an RV-PA connection in addition to the cavopulmonary shunt to simulate a one-and-a-half ventricle repair, it is usually possible to enter the superior vena cava from this ventricle through the RPA.

In this situation, a lateral right ventriculogram should also be performed to demonstrate proximity or otherwise to the anterior chest wall of conduit/outflow tract patch connecting RV-PA and to check for calcification of the conduit/patch. This information is useful for the surgeons because a conduit that is adherent to the

anterior chest wall may be damaged during sternal opening and, in preparation for this eventuality, femoral bypass may be established prior to opening the chest wall. If it proves difficult or impossible to enter the innominate vein, a contrast injection from the left hand may be useful to delineate the anatomy.

Figure 4 Left ventriculogram in the above individual (AP tube orientation) showing mesocardia with apex pointing to the right, and a small rudimentary right ventricle

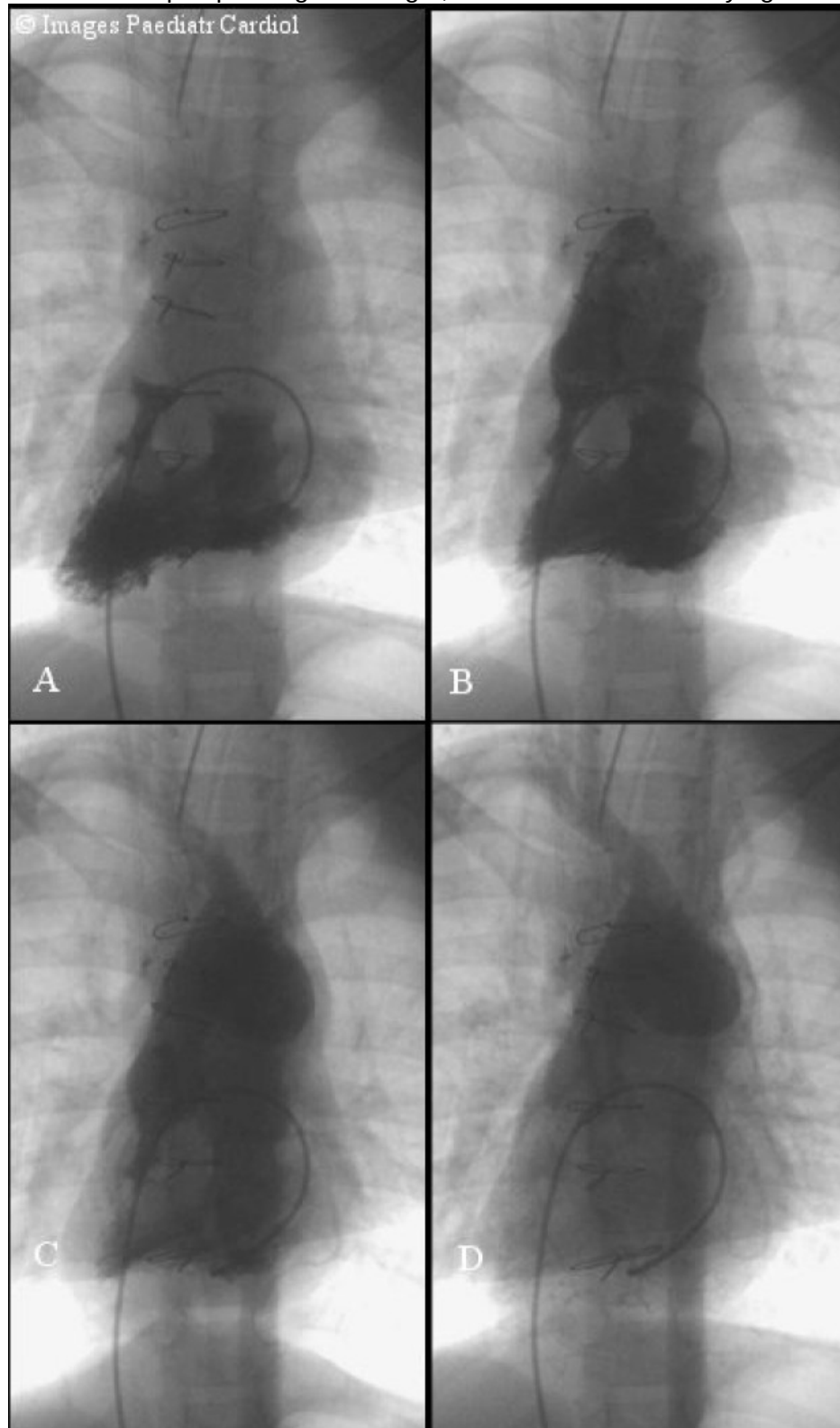


Figure 5 Left ventriculogram in double inlet left ventricle, discordant ventriculoarterial connections after Damus-Kaye-Stensel procedure anastomosing both outflows to the aorta in order to overcome subaortic obstruction or a restrictive outlet foramen (RAO tube orientation). Note that the VSD is restrictive, being much smaller than the aortic valve, and that the RV is just a rudimentary outlet

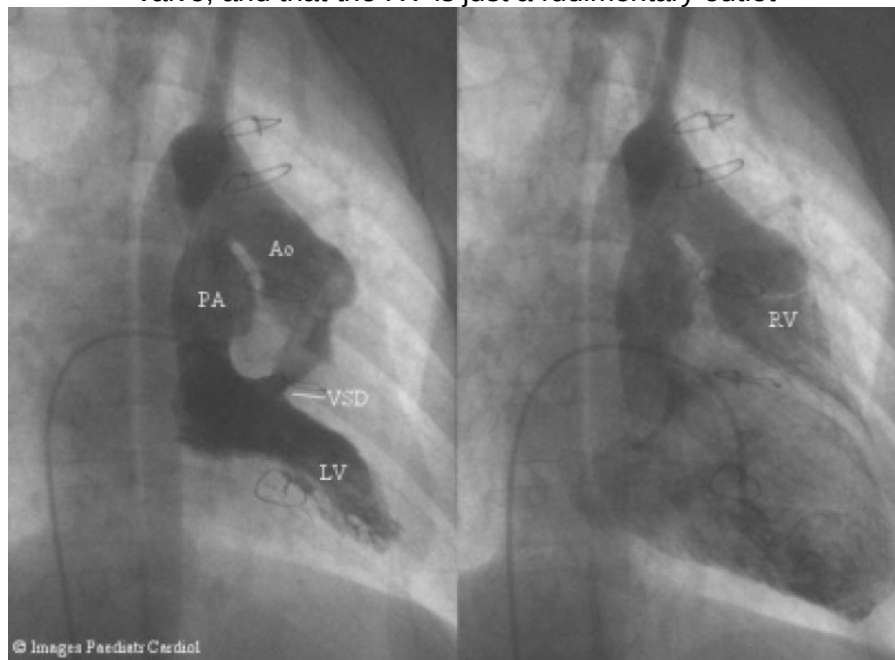
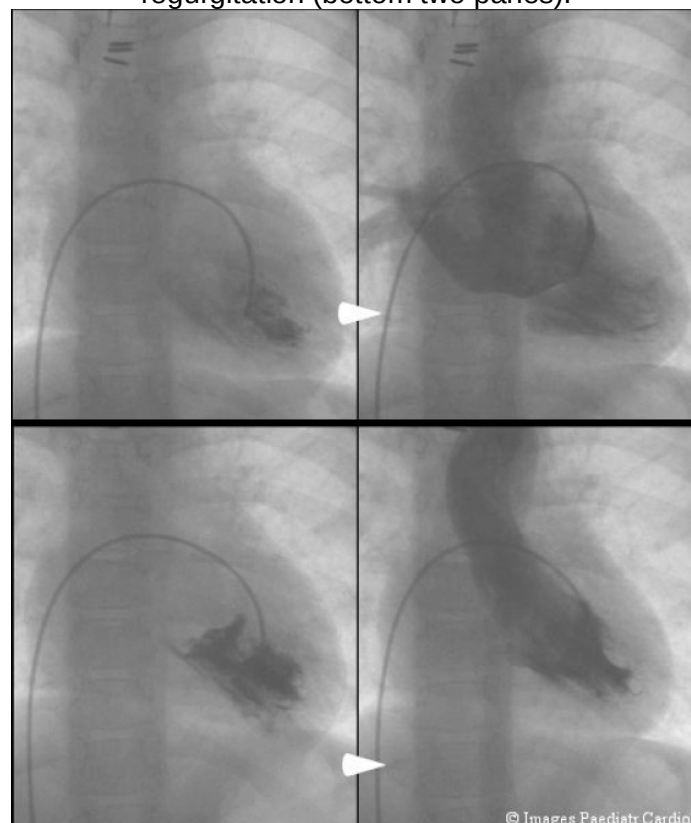


Figure 6 Left ventriculogram in pulmonary atresia with intact interventricular septum shows mitral regurgitation in a patient with pulmonary atresia due to catheter recoil into the left atrium with dye filling pulmonary veins (top two panes). Repeat angiography with the catheter closer to the left ventricular apex showed no mitral regurgitation (bottom two panes).



The cavo-pulmonary shunt is assessed by access through the internal jugular or subclavian vein (usually the right). An angiogram of the SVC will show the anastomosis and also details of the anatomy of the pulmonary arteries as well as the distribution of the pulmonary vasculature within the lung parenchyma. It is also an opportunity to look for acquired or congenital pulmonary arterio-venous malformations and these can take the form of large malformations localised to a segment of the lung or they can be diffuse and involve both lung fields. In addition, it is important to look for veno-venous or veno-atrial shunts especially from the innominate vein and the inferior vena cava. Right subclavian access allows easier coil embolisation of collaterals due to the almost straight approach to the innominate vein. It is also important to follow the angiogram through to the levophase to ascertain normal pulmonary venous drainage.

The pressure is measured in the SVC, RPA and LPA both on phasic and mean; it may be necessary to ask the anaesthetist to stop ventilation in order to abolish the swings of pressure related to the intra-thoracic pressure.

Figure 7 Aortogram in tricuspid atresia and discordant ventriculoarterial connections after Norwood stage II procedure. Note distorted arch (PA tube orientation).



Figure 8 Lateral aortogram in the same individual (lateral orientation).



Figure 9 Right ventriculogram in pulmonary atresia and intact ventricular septum (AP tube orientation).

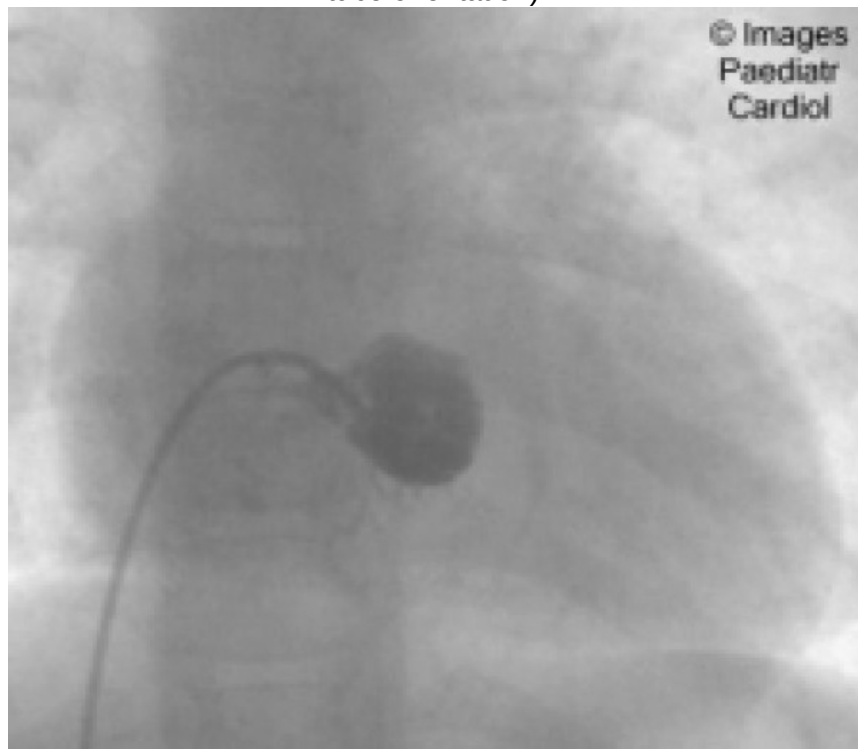


Figure 10 Right ventriculogram in pulmonary atresia after one-and-a-half ventricle repair. Note significant pulmonary regurgitation (AP tube orientation).

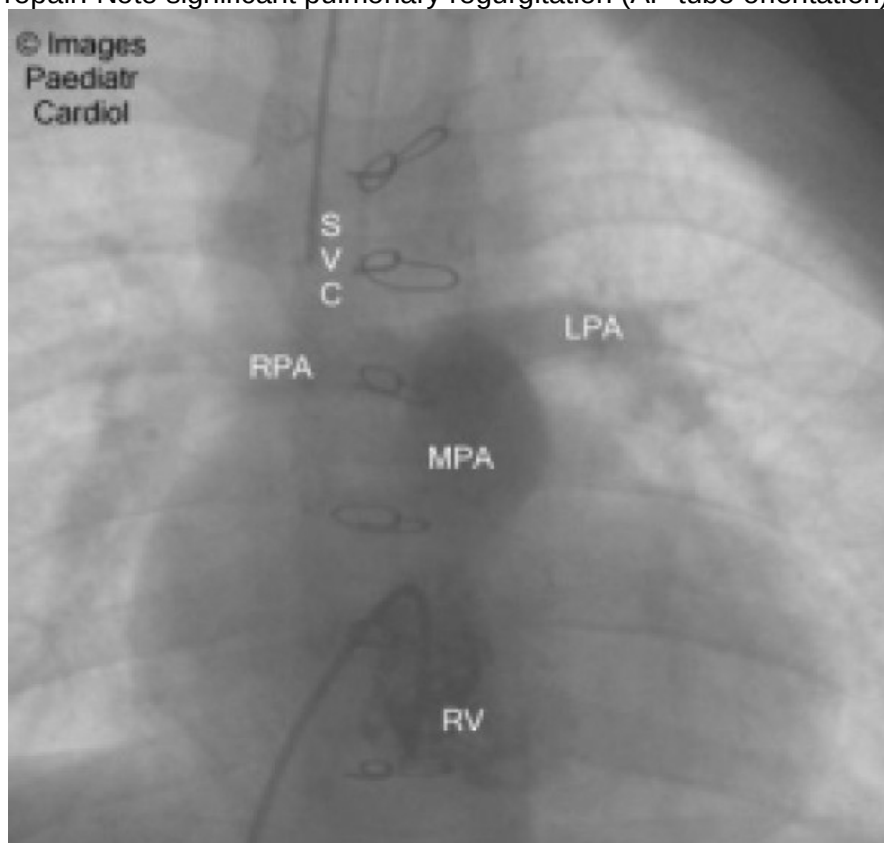


Figure 11 Same individual as in previous figure (lateral tube orientation).

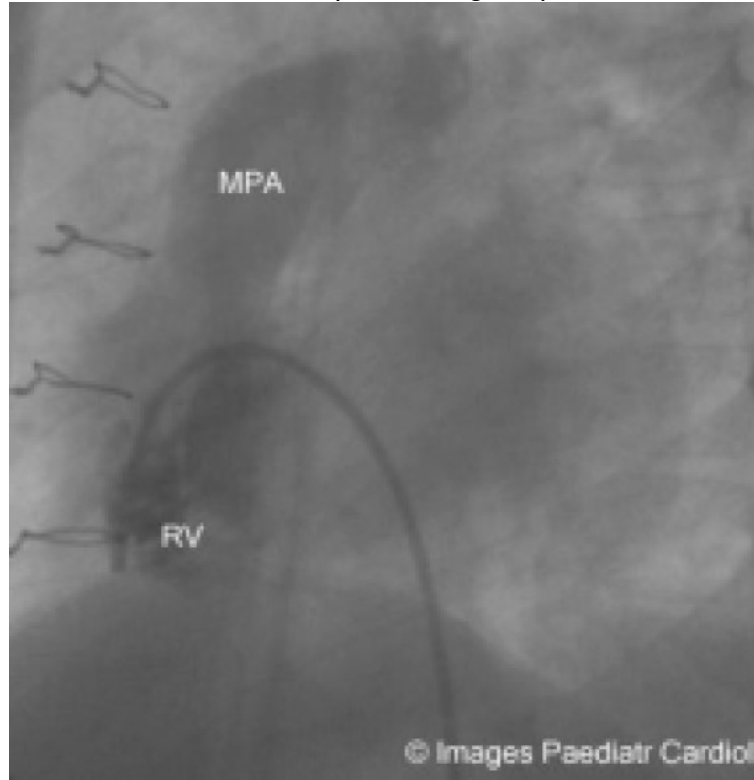


Figure 12 Same individual as in previous figure – catheter passed through IVC, RA, RV, RPA to SVC. Dye passes from SVC to both pulmonary arteries and also to the valveless RV-pulmonary artery connection (PA tube orientation).

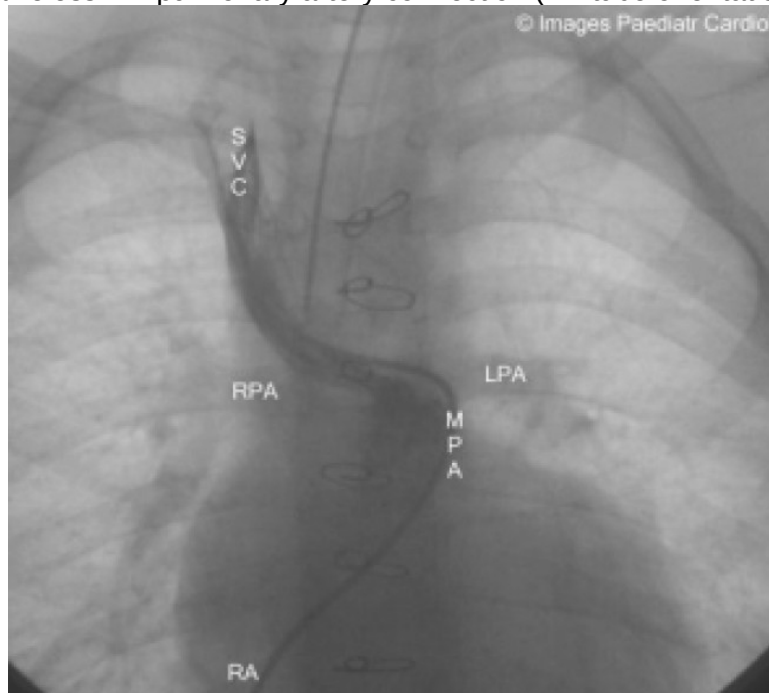


Figure 13 Same individual as in previous figure (PA tube orientation).



Figure 14 Isolated Glenn anastomosis - SVC to pulmonary arteries (PA tube orientation).

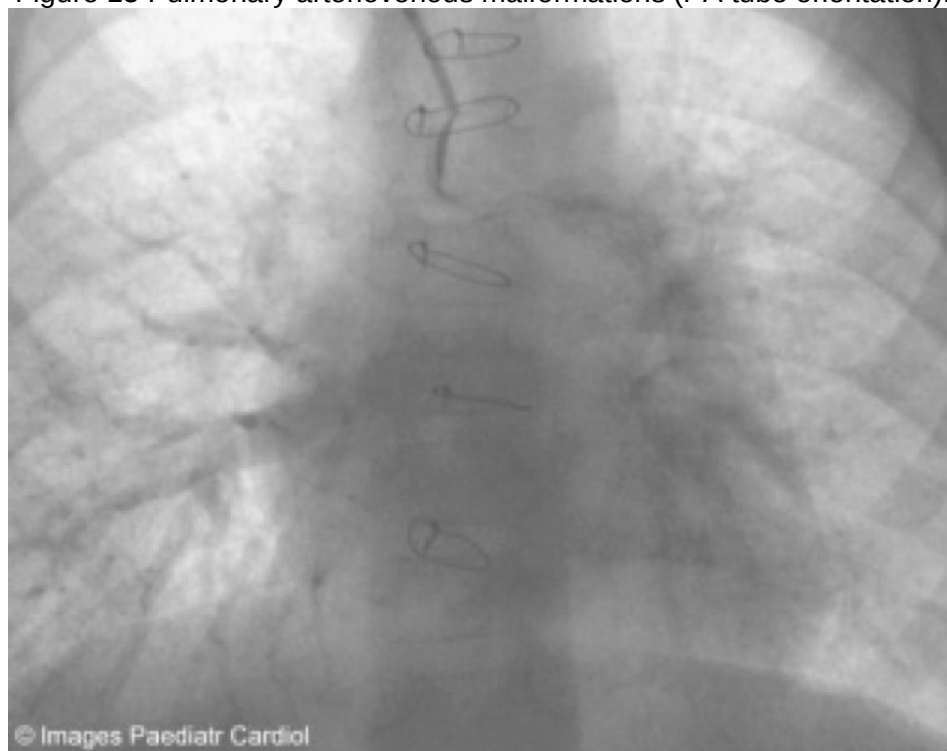


### **Simultaneous LA to PA pressures**

If the mean gradient between the pulmonary arteries and the mean left atrial pressure is less than 6mmHg, the Fontan operation is likely to work whereas, if this is between 6 and 9mmHg there is a high risk of failure and if the gradient is more than 9mmHg the pulmonary vascular resistance is usually too high to attempt any Fontan. If the gradient between the pulmonary arteries and left atrium is borderline, it may be worth considering measuring the RPA and LPA pressures individually the contralateral PA occluded with a balloon as a form of “stress test”.

Pulmonary arteriovenous malformation/s can occur in up to 25% of patients after a Glenn anastomosis.<sup>5</sup> The aetiology of these intrapulmonary right-to-left shunts is uncertain and may be related to low pulsatile blood flow, exclusion of hepatic flow/factors to the lungs, or abnormal distribution of blood flow preferentially to the lower lobes (figure 15). These malformations almost invariably resolve spontaneously after rerouting hepatic flow into the lungs i.e. after TCPC completion.

Figure 15 Pulmonary arteriovenous malformations (PA tube orientation).



From the SVC, enter both branch pulmonary arteries and do pullbacks to SVC. Dye failing to fill the left pulmonary artery implies that it is filled from a left SVC, and on entering this artery, it should be possible to navigate past this artery and up into the left SVC (figure 16).

Figure 16 Bilateral bi-directional Glenns (bilateral bi-directional cavo-pulmonary shunts). Arrow indicates veno-venous malformation.

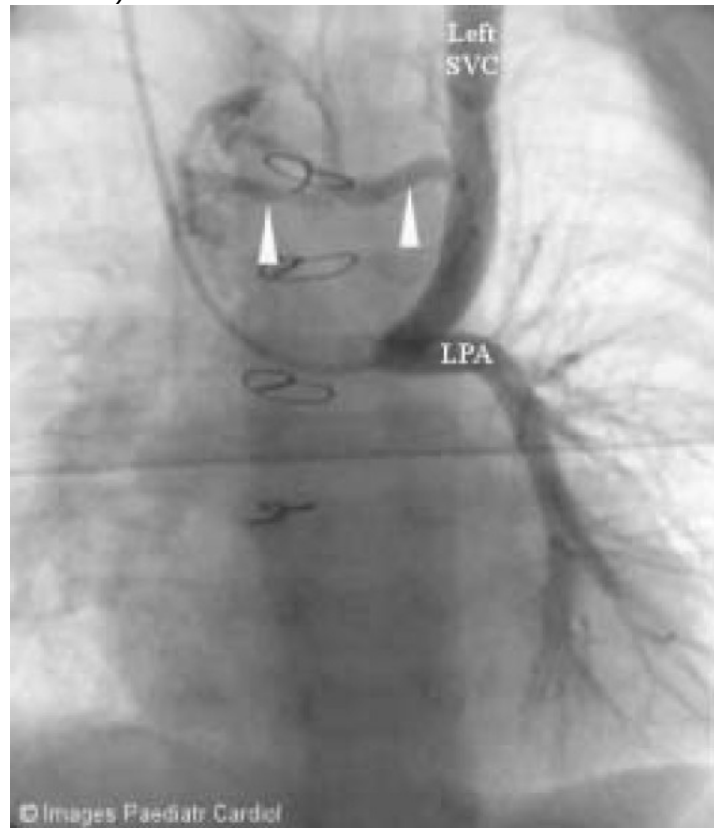


Figure 17 Same patient as in previous figure showing selective angiography of the veno-venous malformation.

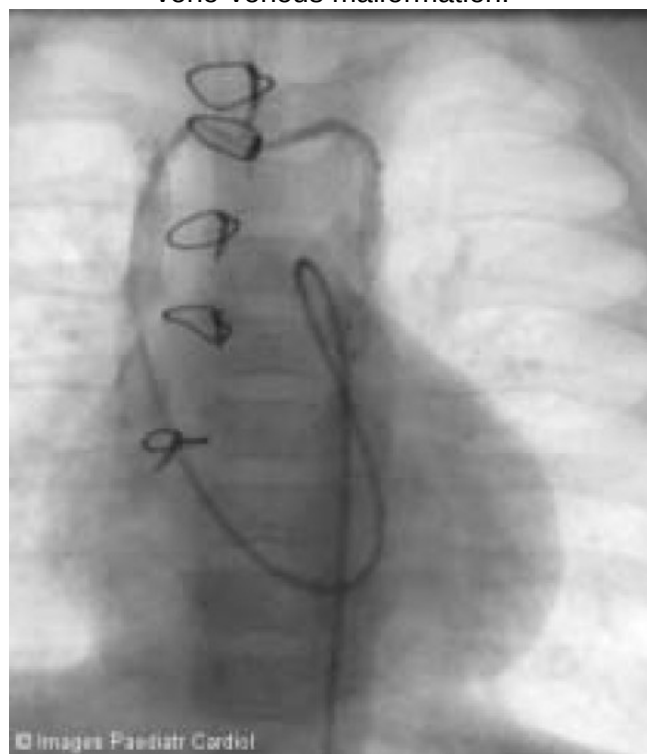
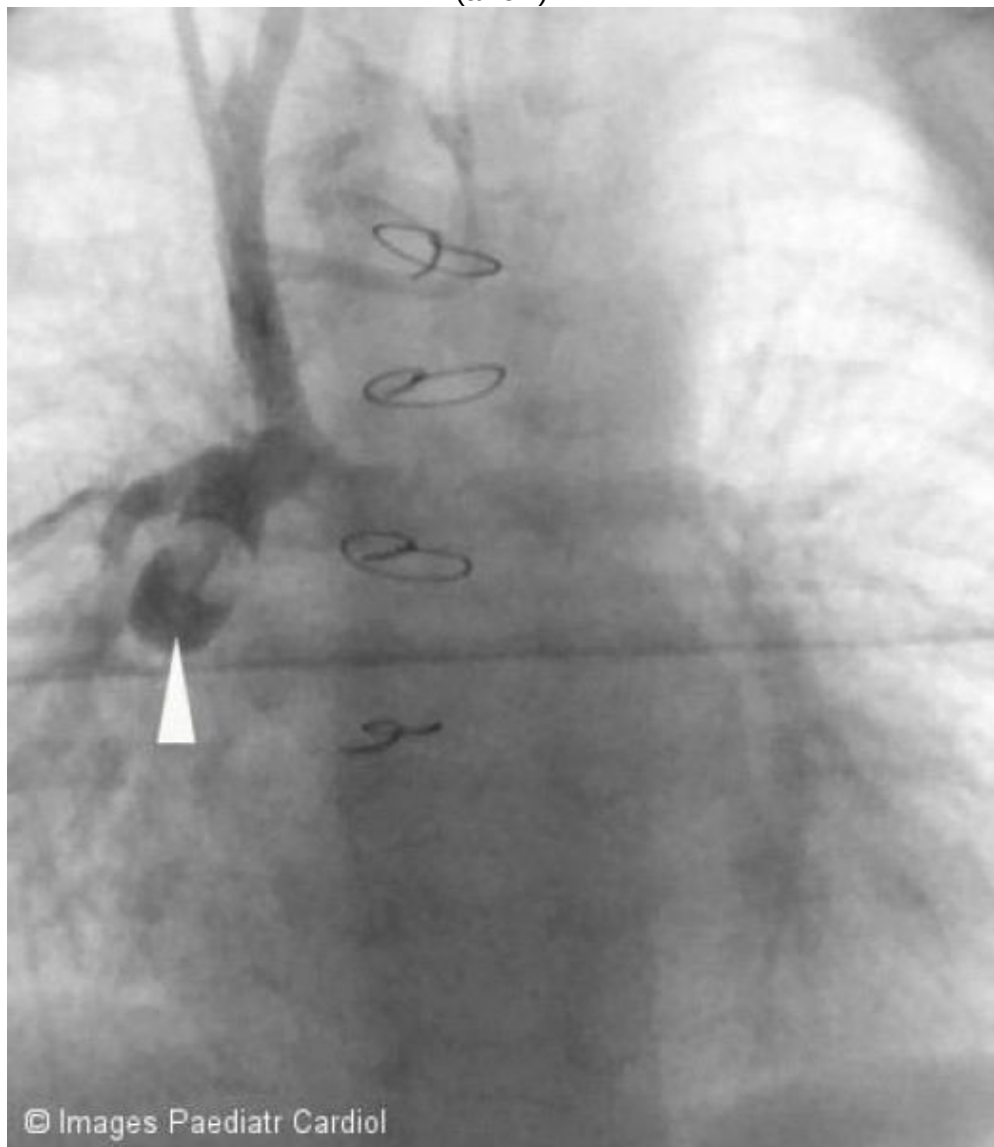


Figure 18 Same patient as in previous two figures. The catheter is balloon tipped (arrow).



Try to enter the innominate vein and do a contrast injection for collaterals and the embryological remnant of the left SVC to the coronary sinus, the vein of Marshall.

In order to enter the innominate vein, it may be necessary to create a loop at the junction of SVC and the pulmonary arteries and pull back with the catheter tip pointing upward and to the left. If innominate vein entry fails with a Gensini, try with a right Judkins catheter.

In azygos or hemiazygos continuation of the inferior vena cava, superior cavopulmonary anastomosis results in total cavopulmonary connection (Kawashima repair).<sup>6</sup> Should reassessment be required after such a surgical procedure, for example, for cyanosis due to acquired pulmonary arteriovenous malformations, the femoral venous route allows access to the pulmonary circulation via the SVC (figures 19-21).

Figure 19 Postoperative left pulmonary artery angiogram in a patient with Kawashima repair – arrows indicate a surgical drain.



Figure 20 Same patient as in the previous figure showing extensive pulmonary arteriovenous malformations of the right lung (lateral view).



Figure 21 Same patient as in the previous figures. Catheter in conduit (arrows) joining hepatic veins to superior cavopulmonary connection.



Cyanosis after TCPC may also be caused by fenestration/s deliberately left during the surgical procedure as runoff to prevent excessively high central venous pressure at the expense of shunting deoxygenated blood from the right side of the circulation to the systemic atrium. Such fenestrations may potentially be closed with a variety of devices (figures 22-26).

Figure 22 Right atrial angiogram showing passage of contrast from right atrium to left atrium.



Figure 23 Same patient as in previous figure after Helex device closure of fenestration. Note minimal residual shunting.

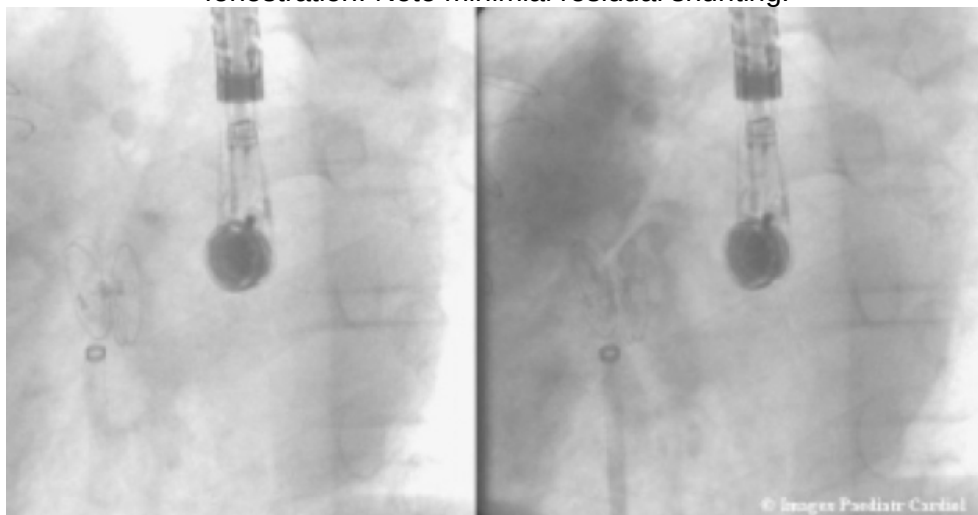


Figure 24 & 25 Innominate vein collaterals indicated by arrows (PA tube orientation). Coiling of a veno-atrial collateral in the same patient shown in figure 24. Upper left pane shows innominate vein angiogram. Upper right pane shows selective angiography of the aberrant vessel. Middle left pane shows one coil deployed in aberrant vessel. Middle right pane shows the deployment of a second coil. Lower left pane shows repeat selective angiography with reduced flow down the vessel. Lower right pane shows the deployment of a third coil.

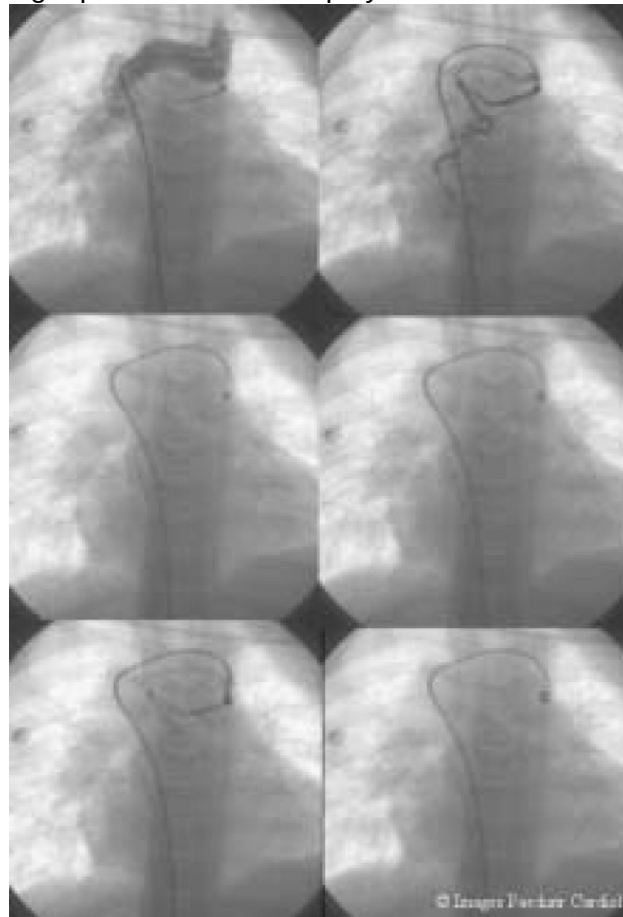
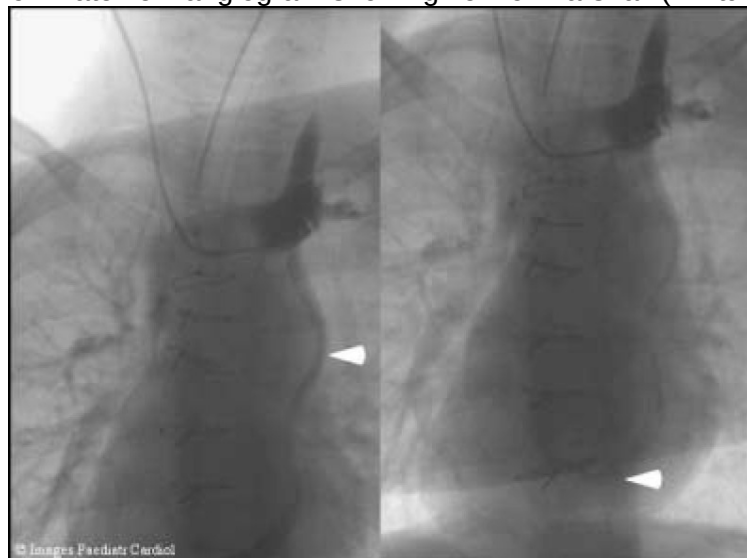
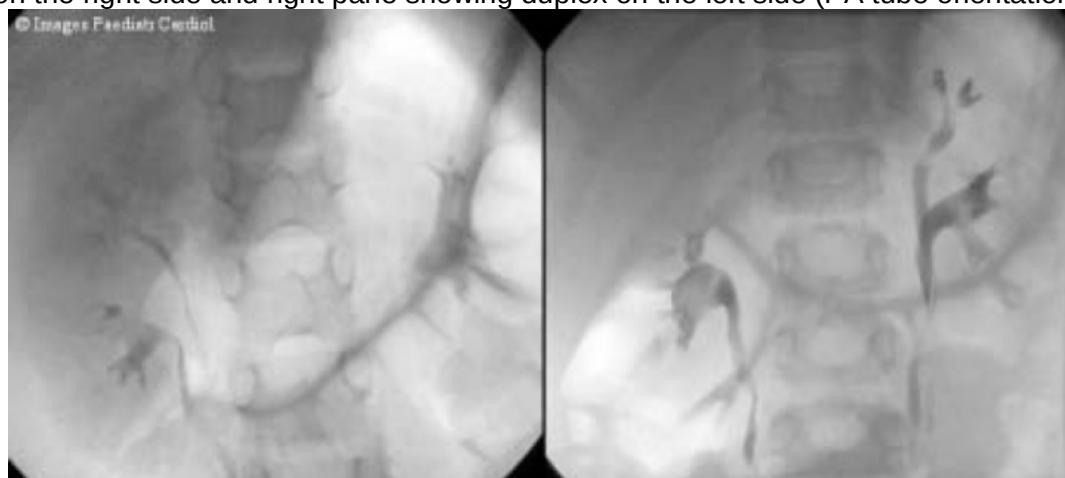


Figure 26 Innominate vein angiogram showing vein of Marshall (PA tube orientation)



For low pressure measurements such as in the atria, pullbacks under apnoea may yield better results. The kidneys should also be screened as usual.

Figure 27 Two of the above patients with duplex kidneys, left pane showing duplex on the right side and right pane showing duplex on the left side (PA tube orientation).



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