Systemic Vein to Pulmonary Artery Shunts: Glenn, Fontan, and Kawashima Procedures

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In a normal biventricular heart, the systemic and pulmonary circulations are in series and each circulation is supported by its respective ventricle. In patients with single ventricular chamber morphology, the two circulations are in parallel and patients only survive by mixing of the systemic and pulmonary venous blood, usually via a septal defect. Systemic venous to pulmonary artery surgical connections have been created to provide venous flow to the pulmonary circulation to allow oxygenation of the blood.

25.1 Glenn Shunt

The original (classic) Glenn shunt is an end-to-end anastomosis of the superior vena cava to the right pulmonary artery, which is divided from the main pulmonary artery. All proximal superior vena cava blood flow is directed to the right pulmonary artery (Figs. 25.1 and 25.2). The azygous vein is also ligated. This shunt was used to palliate congenital heart defects with right-sided hypoplasia or atresia such as tricuspid atresia, Epstein anomaly, and pulmonary atresia with intact ventricular septum.

The modified Glenn shunt, also known as the bidirectional Glenn shunt, consists of an end-to-side anastomosis of the superior vena cava to the right pulmonary artery, which is not divided from the main pulmonary artery (Figs. 25.3 and 25.4). Because the right pulmonary artery maintains continuity with the main pulmonary artery, blood flows from the superior vena cava into both the right and left pulmonary arteries. The complication of both the original and modified Glenn shunts is the development of pulmonary arteriovenous malformations. Currently, the modified Glenn shunt is used as a staging procedure in children with single-ventricle physiology who will later undergo a Fontan procedure.

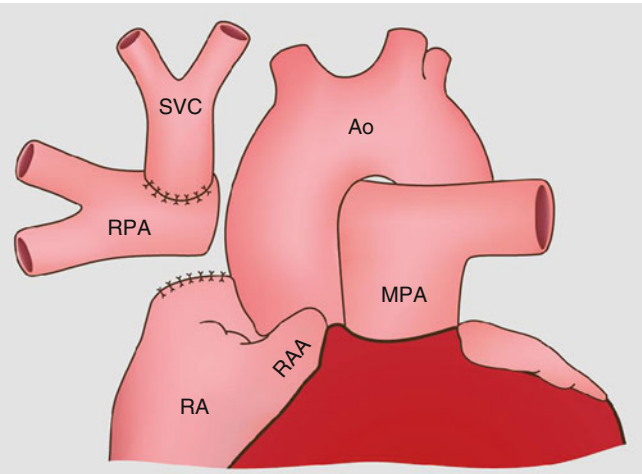

Fig. 25.1 Glenn shunt. This shunt is an end-to-end anastomosis of the right pulmonary artery (*RPA*), which is divided from the main pulmonary artery to the superior vena cava (*SVC*), directing all proximal SVC blood flow to the RPA. The SVC is ligated at its entrance to the right atrium (*RA*). *Ao* aorta, *MPA* main pulmonary artery, *RAA* right atrial appendage, *RPA* right pulmonary artery

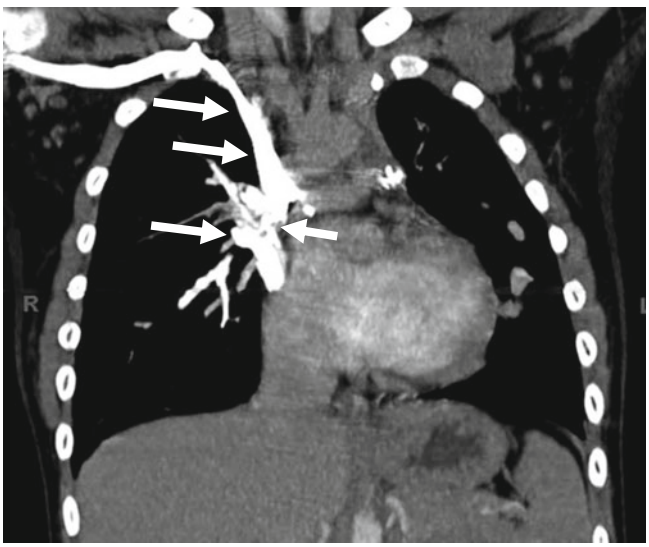

Fig. 25.2 Glenn shunt for tricuspid atresia. This coronal image shows the shunt (*white arrows*), which extends from the superior vena cava to right pulmonary artery

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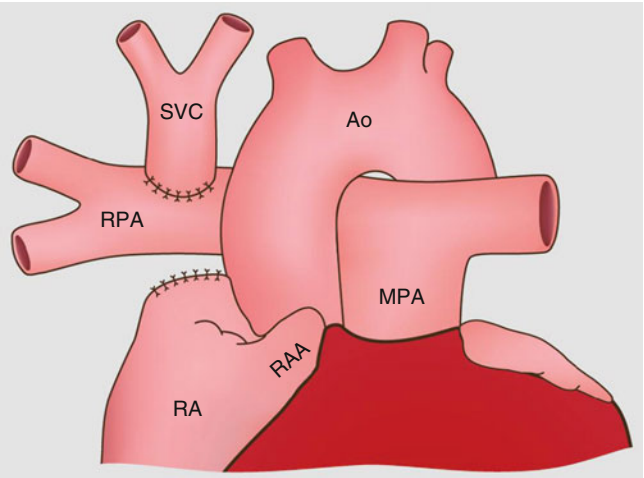

Fig. 25.3 Modified or bidirectional Glenn shunt. This shunt is an end-to-side anastomosis between the superior vena cava (*SVC*) and the right pulmonary artery. The SVC is divided from the right atrium. The right pulmonary artery is not separated from the main pulmonary artery (*MPA*) so venous blood flows into both the right and left pulmonary arteries. *RAA* right atrial appendage, *RPA* right pulmonary artery, *Ao* aorta. This procedure is used in patients with single-ventricle physiology who eventually will undergo a Fontan procedure

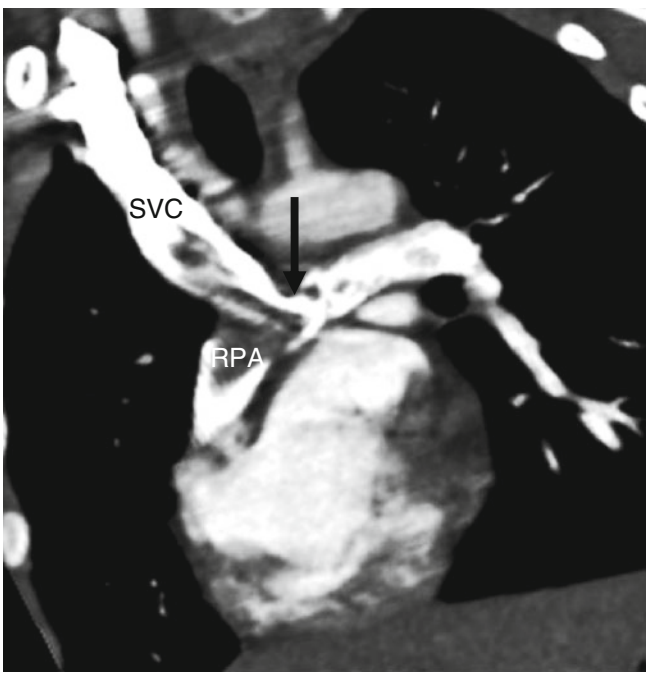

Fig. 25.4 Bidirectional Glenn shunt. This coronal view shows the end-to-side anastomosis between the superior vena cava (*SVC*) and the right pulmonary artery (*RPA*) at the junction with the main pulmonary artery (*arrow*). Note that contrast-enhanced blood flows to both the right and left pulmonary arteries. The non-opacified blood from the superior vena cava is a flow artifact (not thrombus). Artifact can be differentiated from thrombus by reimaging at 2 min which allows enough time for optimal venous opacification

25.2 Fontan Operation

In 1971, Francis Fontan and Eugene Baudet described a procedure that diverted all systemic venous blood into the pulmonary arteries without the interposition of a ventricle as surgical palliation for tricuspid atresia. The Fontan procedure revolutionized the treatment of complex congenital heart defects and remains the palliative treatment of choice for patients born with one functional ventricle. The primary aim of the Fontan procedure is to establish a circulation in which the systemic venous return enters the pulmonary arteries directly.

Creation of the Fontan circulation is considered in all patients with complex congenital heart disease when a biventricular repair is not possible. These include patients with tricuspid atresia, pulmonary atresia with intact ventricular septum, double-inlet left ventricle, hypoplastic left heart syndrome, double-outlet right ventricle, and complete unbalanced atrioventricular septal defects. Candidates for Fontan procedure should be in sinus rhythm and have adequately developed central pulmonary arteries and good ventricular function. The absence of any one of these criteria is a strong predictor of an early poor outcome.

The classic Fontan operation consists of a valved conduit between the right atrium or right atrial appendage and main pulmonary artery (Figs. 25.5 panel a and 25.6). However, this procedure virtually always resulted in severe enlargement and dilatation of the atria which eventually loses contractility and then fails which ultimately further diminishes pulmonary blood flow. Subsequently, there have been many variations of the Fontan procedure, most recently the extracardiac conduit Fontan and lateral tunnel Fontan (Figs. 25.5 panels b, c, 25.7, and 25.8).

The Fontan is usually done early in life as a two-staged repair. Initially a bidirectional Glenn shunt is done to direct superior vena caval blood flow to the lungs, and a tunnel or conduit Fontan is performed with the aim of directing inferior vena cava blood flow to the lungs. In some neonates, a classic or modified Blalock–Taussig shunt may be performed prior to the Fontan procedure in order to establish pulmonary blood flow.

Stage 1 Fontan consists of a bidirectional Glenn shunt or a hemi-Fontan procedure and is usually undertaken as soon as the pulmonary arteries have grown sufficiently to allow a low pulmonary vascular resistance, usually between 2 and 6 months of age. The hemi-Fontan procedure involves formation an atriopulmonary anastomosis between the dome of the right atrium and the underside of the right pulmonary artery with patch in the superior portion of the right atrium to direct blood flow from the superior vena cava atriocaval junction into the atriopulmonary anastomosis (Fig. 25.9). The stage 1 operation provides low-pressure pulmonary blood flow to the lungs and decreases

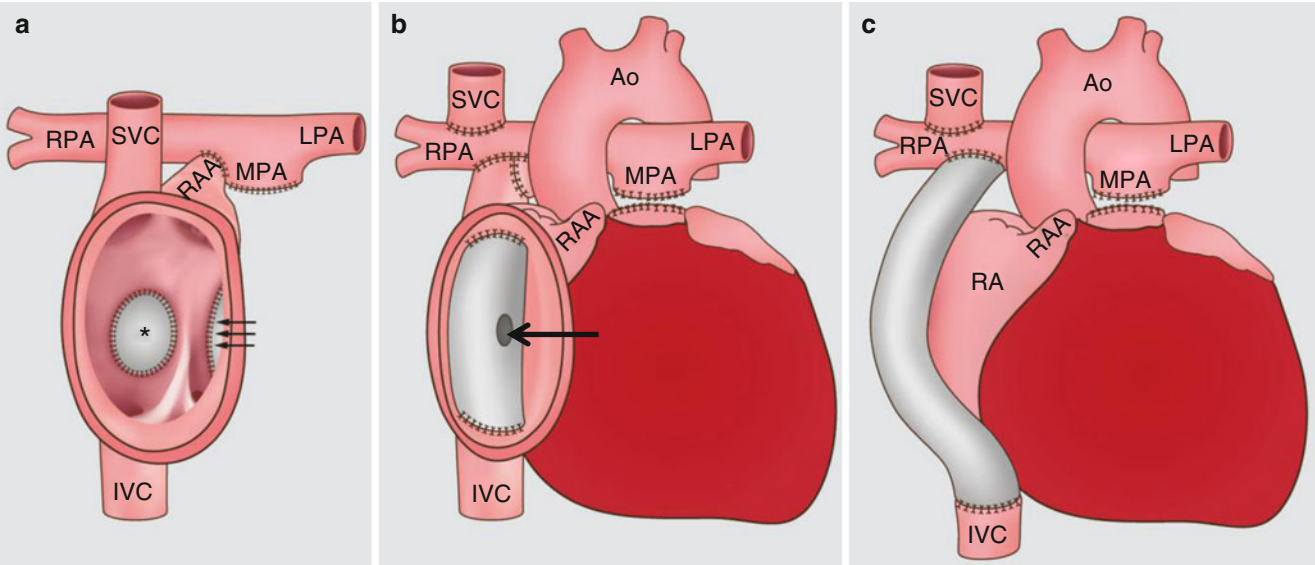

Fig. 25.5 Fontan procedures. Panel (**a**) demonstrates the classic Fontan procedure. The right atrial appendage (*RAA*) is connected to the pulmonary artery. The asterisk (*) marks the closed atrial septal defect. The three *arrows* point to the over sewn tricuspid valve. Panel (**b**) shows the lateral tunnel procedure. Systemic blood from the inferior vena cava (*IVC*) is redirected via an intra-atrial tunnel to the pulmonary arteries. The main pulmonary artery is ligated. The superior vena cava (*SVC*) is connected to the right pulmonary artery as a bidirectional cavopulmonary anastomosis (bidirectional Glenn shunt). Note the presence of fenestration (*arrow*) in the interatrial tunnel. Panel (**c**) illustrates an extracardiac conduit procedure. Systemic blood flow from the IVC is redirected via an extracardiac conduit to the pulmonary arteries. Similar to the intra-atrial tunnel, the SVC is connected to the right pulmonary artery as a bidirectional cavopulmonary anastomosis. Currently, the Fontan circulation is achieved by using a bidirectional Glenn shunt and a lateral tunnel or extracardiac conduit Fontan. *Ao* aorta, *MPA* main pulmonary artery, *LPA* left pulmonary artery, *RPA* right pulmonary artery

the volume load on the single ventricle. The atria continue to receive venous blood directly from the inferior vena cava and oxygenated blood returning via the pulmonary veins. There is mixing of oxygenated and deoxygenated blood via an atrial septal defect, resulting in peripheral oxygen saturation in the range of 80–85 %. Because patients have residual mixing of oxygenated and deoxygenated blood and hypoxia, a second-stage operation (described below) is required after the pulmonary arteries grow adequately and are less fragile.

Stage 2 Fontan, also called Fontan completion, is usually performed at 2–5 years of age when pulmonary arteries are of sufficient size to allow high pulmonary flow. It involves redirecting blood from the inferior vena cava or hepatic veins into the pulmonary circuit either via a lateral tunnel (Figs. 25.5 panel b and 25.7) or an extracardiac conduit Fontan (Figs. 25.5 panel c and 25.8). With the lateral tunnel Fontan, a tunnel is created in the right atrium using prosthetic material. The tunnel is anastomosed inferiorly to the inferior vena cava and superiorly to the right or main pulmonary artery. With an extracardiac conduit Fontan, the inferior vena cava is separated from the right atrium and a synthetic (polytetrafluoroethylene) tube graft is created adjacent to the right atrium (rather than within it) to connect the inferior vena cava with the right or main pulmonary arteries. A small fenestration or opening may be created between the venous conduit or tunnel and the atrium to prevent volume overload to the pulmonary circulation. The fenestration limits caval pressure and venous congestion. It also results in increased systemic ventricular preload and cardiac output at the expense of mild desaturation. The stage 2 Fontan corrects the hypoxia and leaves the single ventricle responsible only for supplying blood to the body.

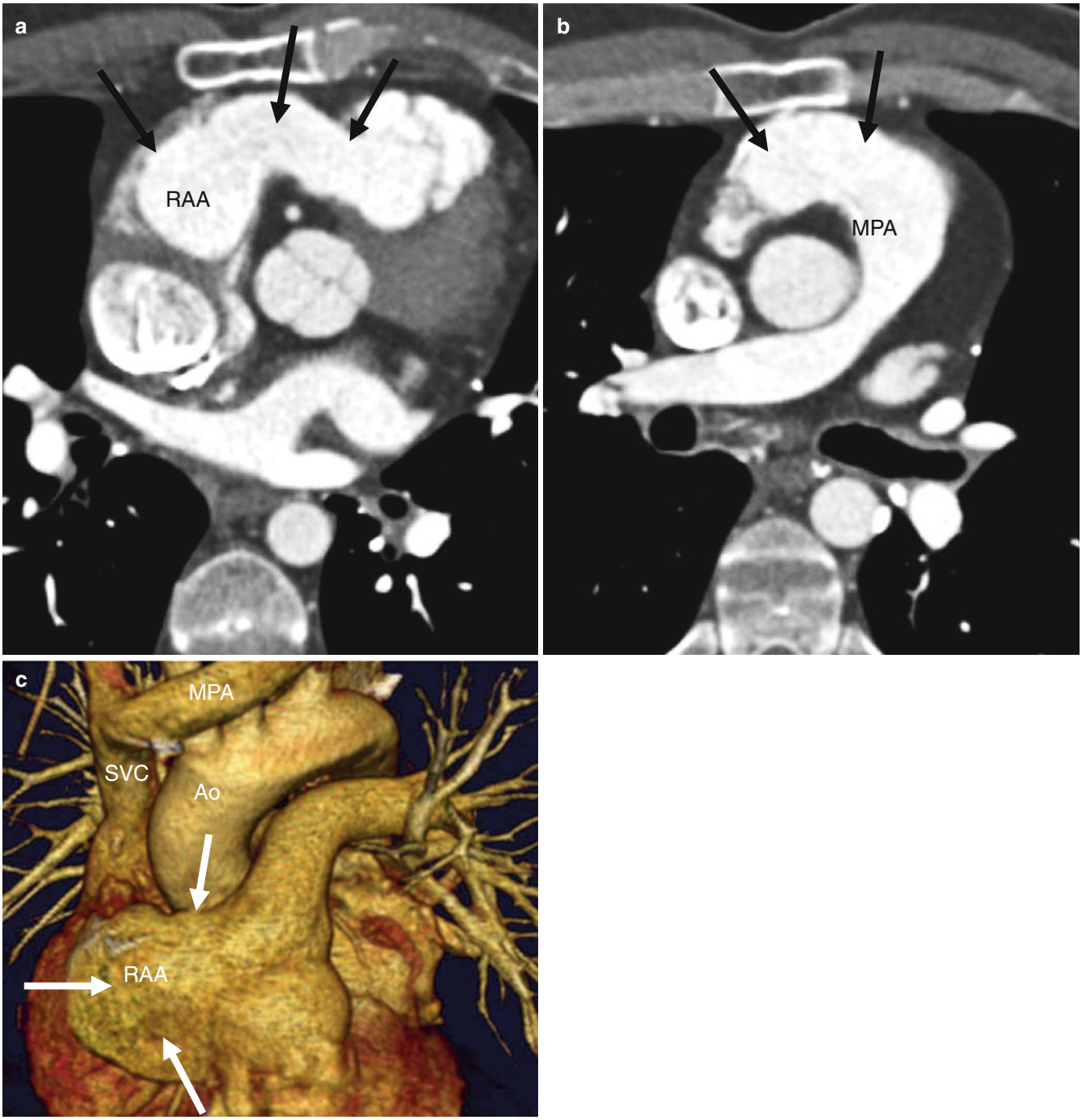

Fig. 25.6 Classic Fontan shunt for tricuspid atresia. Panels (**a**) and (**b**) are transaxial views. Panel (**c**) is a 3D image. The classic Fontan (*arrows*) from the right atrial appendage (*RAA*) to the main pulmonary artery (*MPA*) is depicted. In panel (c), note the superior vena cava (*SVC*) which is surgically connected to the right pulmonary artery (the anastomosis is posterior and thus not visualized in this image)

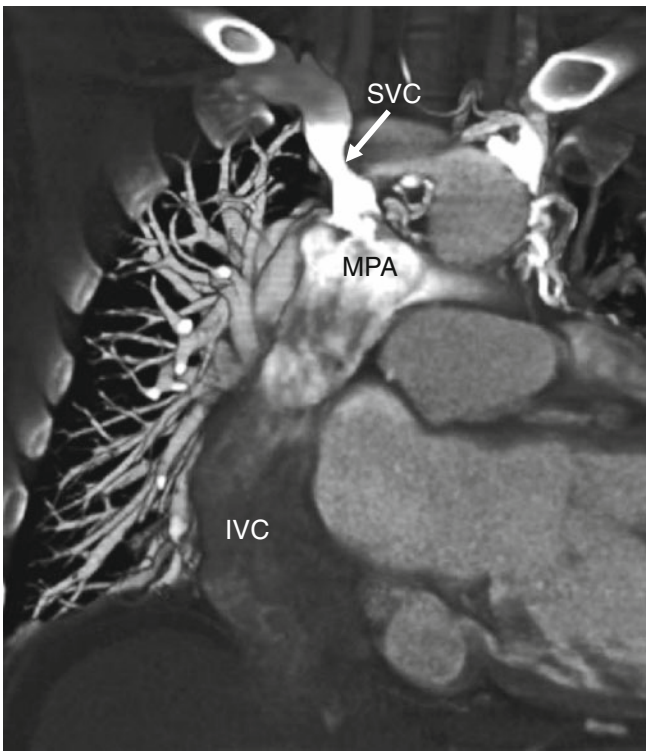

Fig. 25.7 Lateral tunnel Fontan for tricuspid atresia. A coronal 3D image shows the opacified superior vena cava (*SVC*) and unopacified inferior vena cava (*IVC*), both of which are connected to the main pulmonary artery (*MPA*). The tunnel is created within the right atrium using prosthetic material. *RA* right atrium

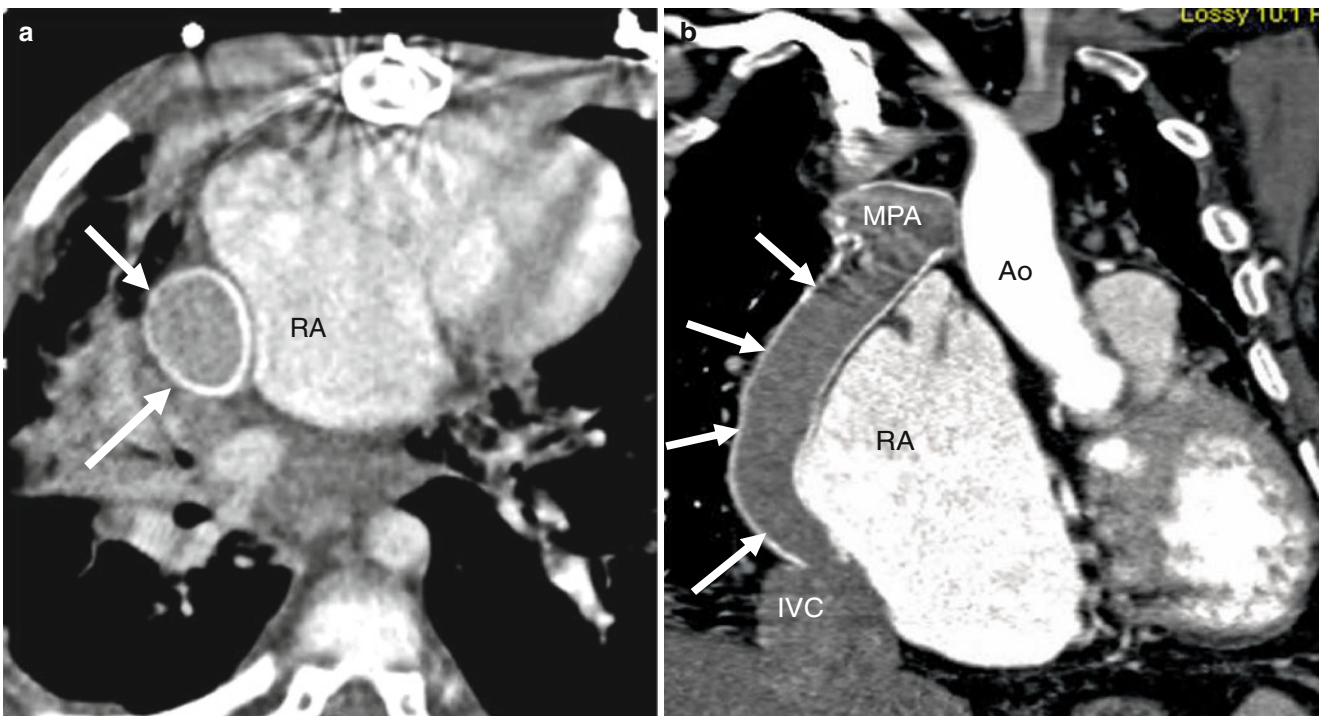

Fig. 25.8 Extracardiac conduit Fontan. Panel (**a**) is an axial image and panel (**b**) is a coronal reformatted image, showing a synthetic tube graft (*arrows*) adjacent to the enlarged right atrium (*RA*). The shunt connects the inferior vena cava with the main pulmonary artery (*MPA*). *Ao* aorta, *IVC* inferior vena cava

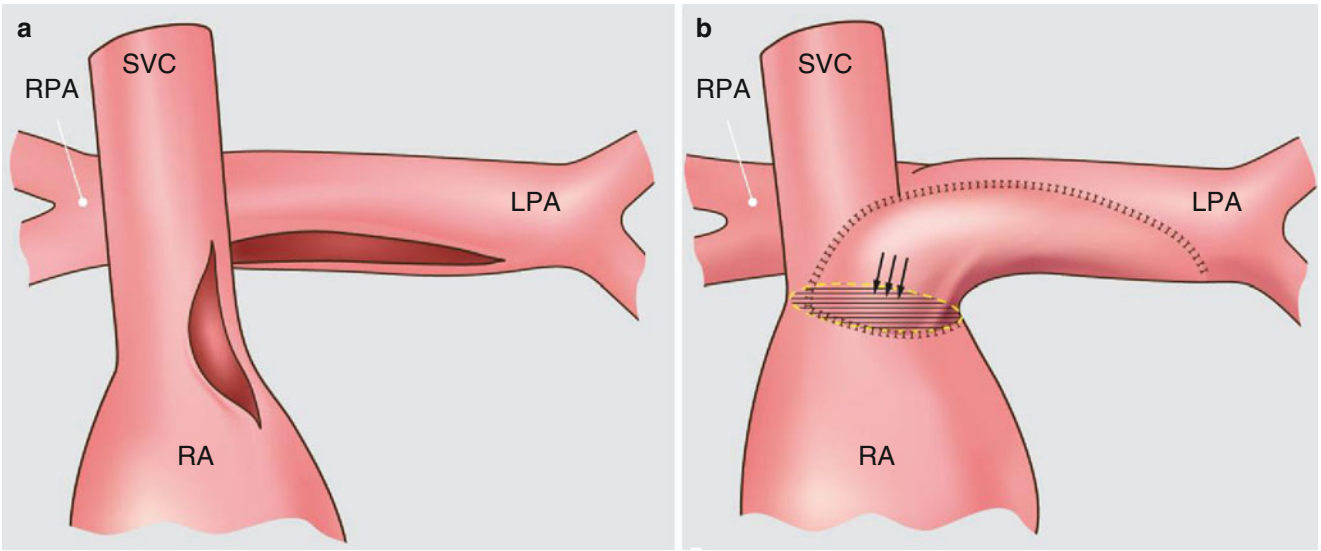

Fig. 25.9 The hemi-Fontan procedure. This is an alternative first stage procedure to the Glenn shunt in the Fontan operation. Panel **a**. The procedure involves an atriopulmonary anastomosis between the dome of the right atrium (*RA*) and the underside of the right pulmonary artery (*RPA*). A patch is placed in the superior aspect of the *RA* (shown in Panel **b**, *arrows*) to direct blood flow from the superior vena cava (*SVC*) atriocaval junction into the atriopulmonary anastomosis. The patch typically extends into the left pulmonary artery (*LPA*) augmenting the pulmonary artery area

25.3 Cardiovascular Complications of Fontan Circulation

Despite the advances in surgical procedures, there are several complications associated with the Fontan procedure. A well-known complication of both the Glenn and original Fontan circulation (right atrial appendage to pulmonary artery) is the development of pulmonary arteriovenous malformations (Fig. 25.10). It is thought that lack of admixture with hepatic venous blood and the absence of pulsatile flow in the pulmonary bed resulting from the absence of a pumping ventricle between the systemic venous return and the pulmonary bed play a role the development of arteriovenous malformations. These have been reported to resolve with reestablishment of exposure to inferior vena cava blood flow. Arteriovenous malformations can result in ventricular volume overload and may lead to irreversible pulmonary hypertension and ultimately failure of the Fontan circulation.

Venovenous connections may also develop between the deoxygenated venous drainage of the upper extremities and the oxygenated pulmonary veins or left atrium. They likely develop secondary to elevated central venous pressures and can result in right-to-left shunting and exacerbation of cyanosis.

Thrombus can develop within conduits or cardiac chambers due to low-velocity pulmonary flow, atrial fibrillation, hypercoagulability, or exposure to thrombogenic suture material (Fig. 25.11) and can result in systemic or pulmonary thromboembolic events. Thromboembolic complications have been reported rarely of in patients within 10 years of the procedure. These include cerebrovascular accidents, pulmonary embolism, and systemic venous clots. Varma et al. reported

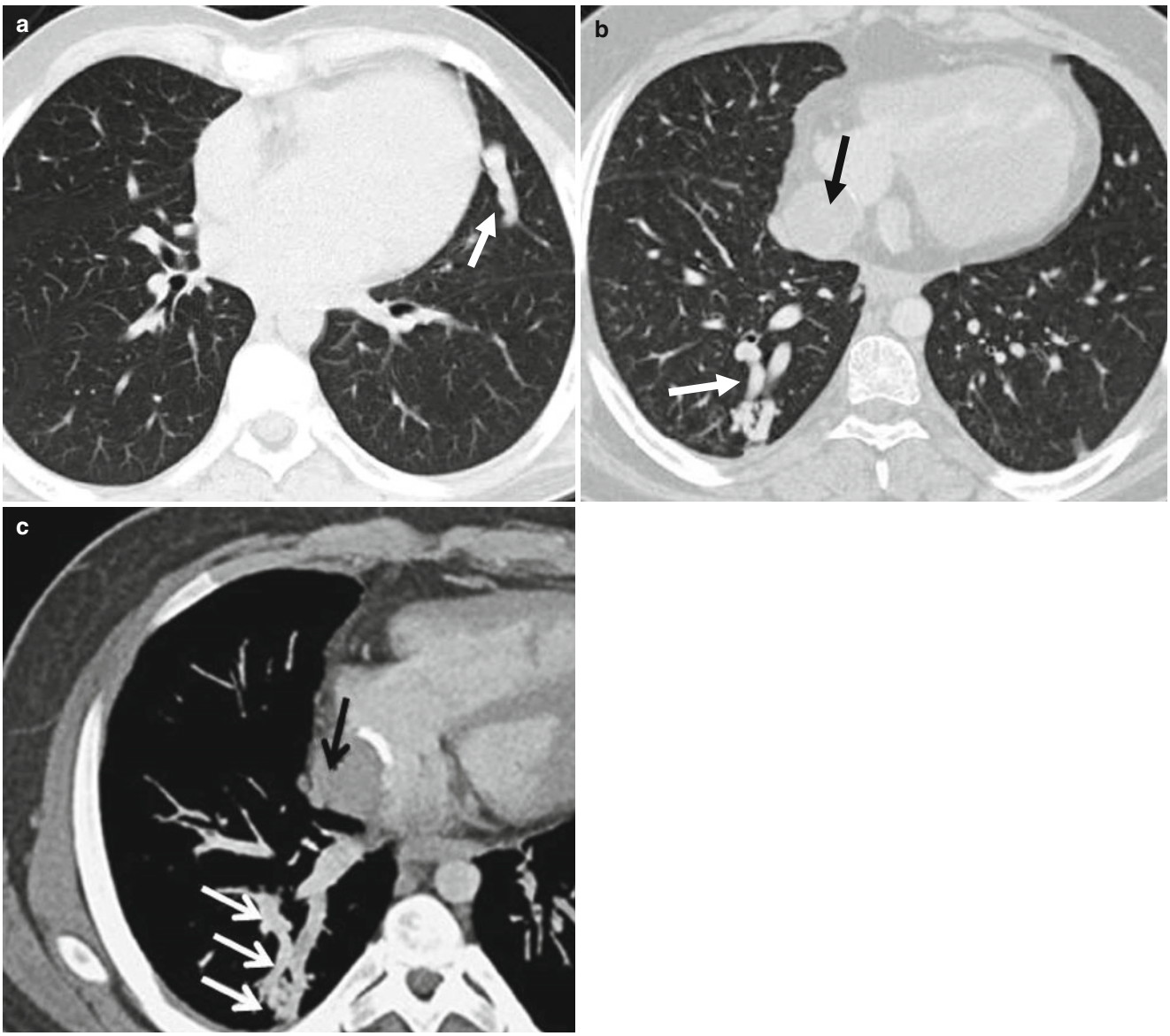

Fig. 25.10 Arteriovenous malformation (AVM) with a Glenn shunt. Panel (**a**) is an axial image showing a pulmonary AVM (*arrow*) in the lingula. Panel (**b**) is an axial slice and panel (**c**) is a maximum intensity projection (MIP). Both panels (**b**) and (**c**) show an AVM in the right lower lobe (*white arrows*) as well as the extracardiac Fontan (*black arrow*). Note that the use of thick-slab MIPs improves delineation of the feeding arteries and draining veins of the AVM

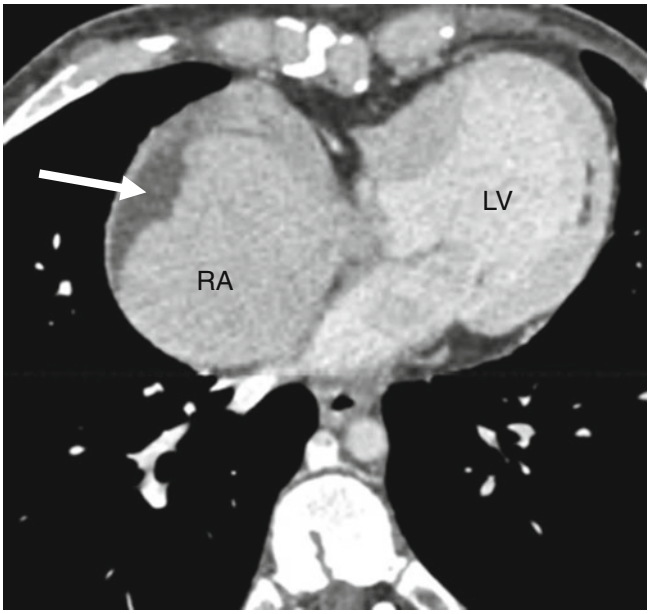

Fig. 25.11 Fontan shunt with thrombus. Axial image showing thrombus formation (*arrow*) in the enlarged right atrium (*RA*) in a patient with tricuspid atresia. *LV* morphologic left ventricle

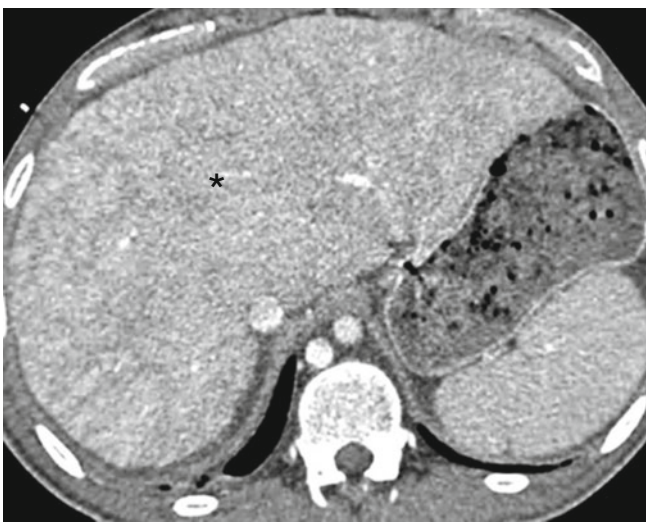

Fig. 25.12 Hepatic congestion secondary to Fontan circulation. This axial computed tomogram shows a heterogeneous enhancement pattern of the liver (*asterisk*), most prominent in the periphery

clinically silent pulmonary emboli in 17 % of patients with Fontan circulation [1]. The univentricular heart after the Fontan operation occasionally exhibits a blind pouch formed by the pulmonary stump or rudimentary ventricle. Almost 2 % of patents have thrombosis in the blind pouch [2].

Systemic ventricular dysfunction can develop as a result of complications mentioned above. Systolic dysfunction of the single ventricle is seen frequently in patients 10 years after the procedure. Ventricular dilatation, global hypokinesis, hypertrophy, and atrioventricular valve regurgitation can occur. Mild desaturation is common secondary to right-to-left shunting via surgical fenestrations and pulmonary arteriovenous malformations. Additionally, anastomotic stenosis can occur resulting in increased venous pressure and decreased cardiac output. Long-term extracardiac complications include hepatic dysfunction (congestion, fibrosis, and cirrhosis) (Fig. 25.12) and protein-losing enteropathy which often lead to the patient's demise [3].

25.4 Cardiac Computed Tomographic Angiography (CT) in the Evaluation of Glenn Shunts and Fontan Circulation

Computed tomography (CT) has a role in evaluating the patency of Glenn shunts and the Fontan circulation as well as the potential complications of these procedures. A noncontrast sequence should be performed in order to identify calcified conduits followed by an arterial phase timed to the aorta. Care must be taken in imaging patients with Fontan anatomy so as not to confuse an unenhanced conduit with thrombus. Hence, a delayed venous-phase CT scan may be useful to allow for contrast enhancement of the conduit or inferior vena cava since more time is required for contrast to opacify these structures (Fig. 25.13) [4]. CT is also helpful in defining complications of shunting at the atrial-conduit level, arteriovenous malformations (Fig. 25.10), and pulmonary embolus/thrombus [5–11]. Arteriovenous malformations are best seen on thick-slab (4–20 mm) maximum intensity projections. CT can also be used to evaluate the percutaneous coil embolizations performed to treat these malformations.

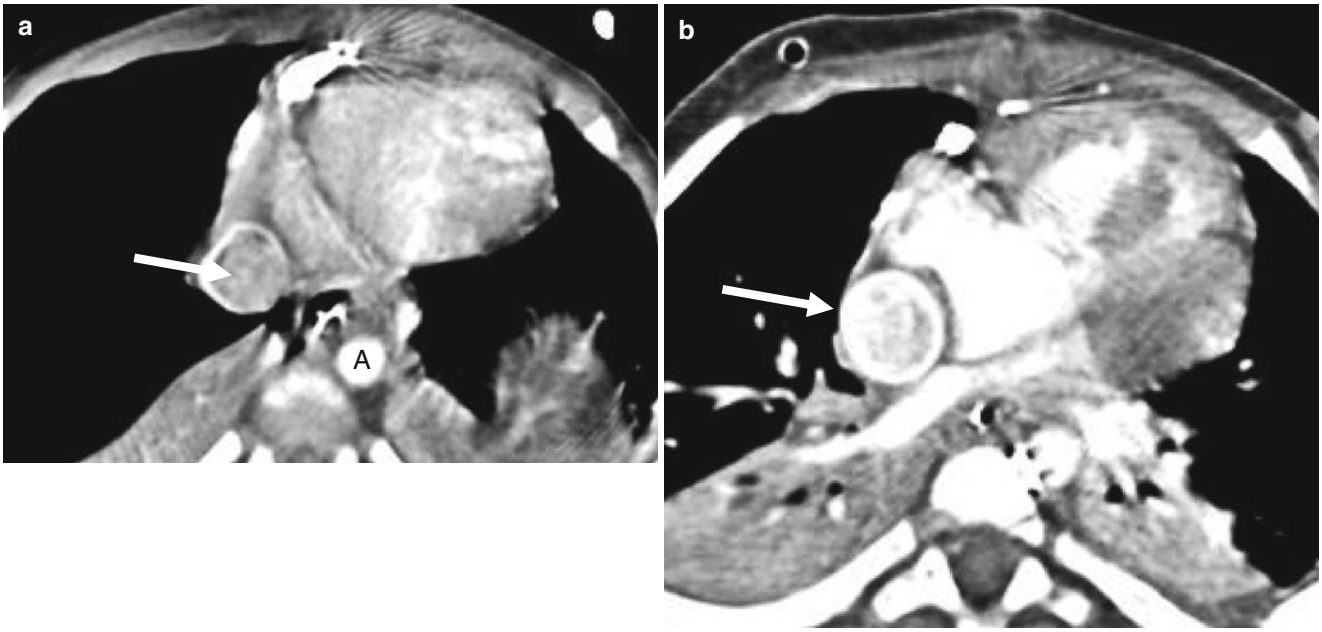

Fig. 25.13 Fontan shunt enhancement techniques. Panel (**a**) an arterial phase image 15 s after injection of contrast medium. The Fontan conduit (*arrow*) is not opacified, while the aorta (*A*) shows intense enhancement. Panel (**b**) a delayed image at 30 s shows enhancement of the conduit (*arrow*)

25.5 Kawashima Procedure

The Kawashima procedure is performed in patients with heterotaxy syndrome and refers to creation of a bidirectional Glenn shunt in the setting of interruption of the inferior vena cava with azygous continuation to the superior vena cava. The azygous continuation of the inferior vena is then anastomosed to the pulmonary artery (Fig. 25.14). This results in redirection of systemic venous blood to the lungs with the exception of the hepatic and coronary venous return. Mild cyanosis is present postoperatively because desaturated hepatic venous blood mixes with pulmonary venous blood. Hepatic venous blood flow is often not redirected because of the complexity of the surgery. However, the complication of not redirecting hepatic venous flow is a high prevalence of the development of pulmonary arteriovenous malformations [12]. If arteriovenous malformations develop, a conduit may be created between the hepatic veins and the innominate vein to redirect hepatic blood flow into the pulmonary circuit, reducing the hemodynamic impact of the malformations [13]. Complications of hepatic conduits include thrombosis or stenosis which also may be evaluated reliably with CT.

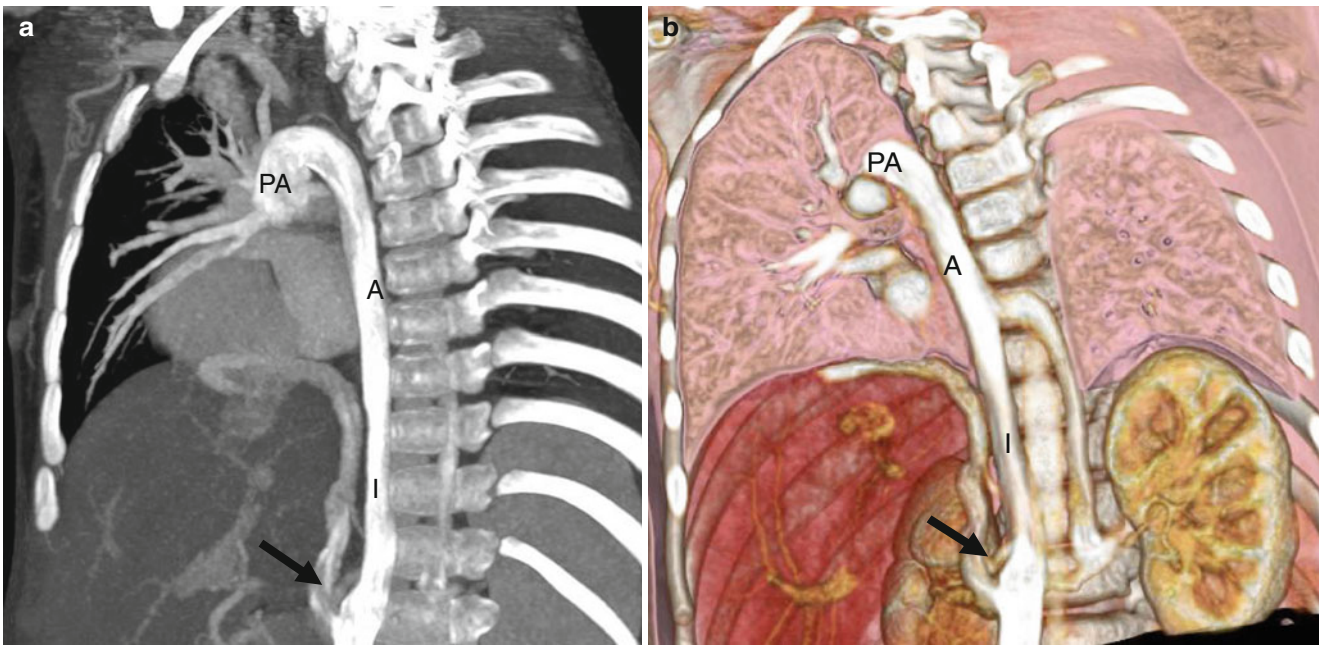

Fig. 25.14 A Kawashima procedure in a patient with interrupted inferior vena cava with azygous continuation, atrioventricular canal defect, and hypoplastic left heart. Panels (**a**) (sagittal image) and (**b**) (3D reconstructions) show the interruption of the inferior vena cava (*I* and *black arrow*) with azygous (*A*) continuation to the pulmonary artery (*PA*) (Images provided kindly by Frandics Chan, MD. Stanford University)

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