Operations Performed for Patients with Congenital Heart Disease

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When dealing with congenital heart disease, it is vital to understand the basic terminology regarding postoperative shunts, procedures, and surgeries.

Sano Shunt

A Sano shunt is a vascular conduit that shunts blood from the right ventricle to the main pulmonary artery. The shunt narrows as it traverses the right ventricular wall, which limits regurgitant flow. It may be used in combination with a Blalock-Taussig (BT) shunt in patients with hypoplastic left heart syndrome as part I of the Norwood procedure.

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R.R. Richardson, *Atlas of Pediatric Cardiac CTA*,
DOI 10.1007/978-1-4614-0088-2_11,

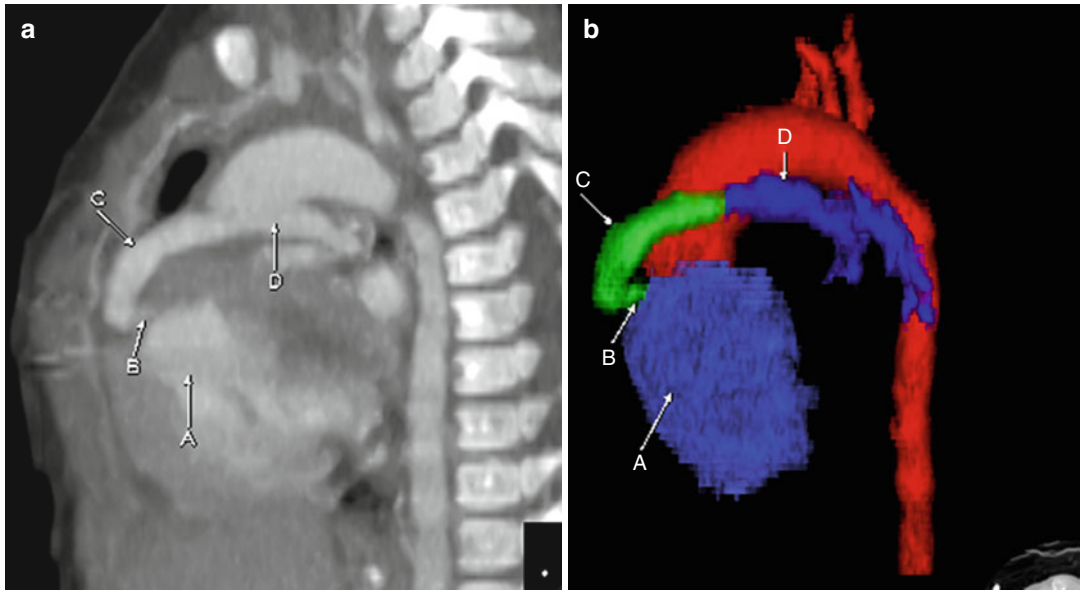

Fig. 11.1 Sagittal maximum-intensity projection (MIP; **a**) and color-coded three-dimensional (3D; **b**) images showing a Sano shunt (*C*) connecting the right ventricle (*A*) to the main pulmonary artery (*D*). Notice the stenotic portion of the proximal shunt (*B*), where the conduit traverses the right ventricular wall

Modified BT Shunt

The modified BT shunt uses a vascular graft (typically Gore-Tex [W. L. Gore and Associates, Elkton, MD]) that shunts blood from the subclavian or brachiocephalic artery to the ipsilateral main pulmonary artery. It often is used as a palliative procedure to increase pulmonary flow and expand the pulmonary arteries before more definitive surgery. The modified shunt differs from the original procedure, which is performed without a graft.

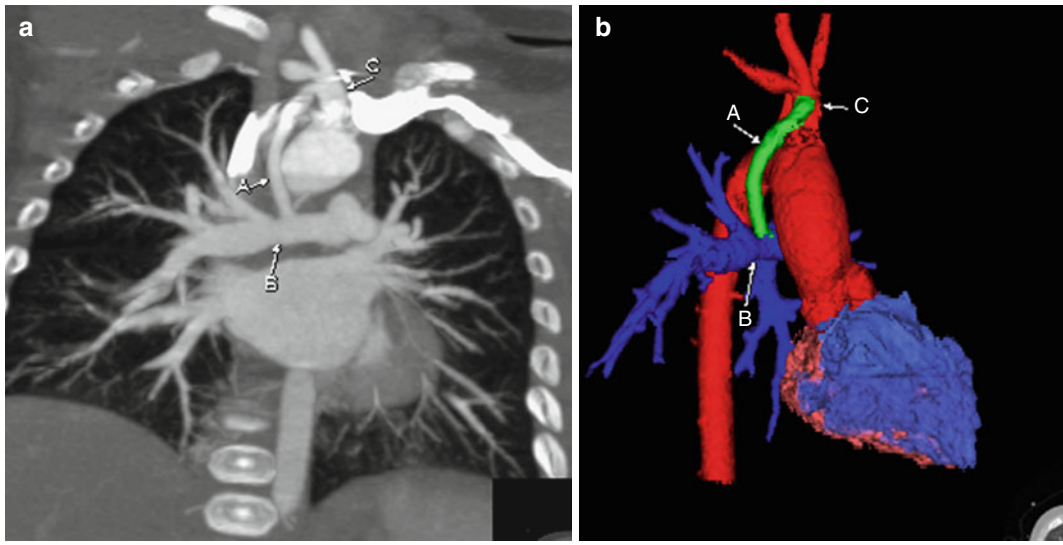

Fig. 11.2 Enhanced coronal (**a**) and color-coded 3D (**b**) images showing a modified BT shunt (*A*) connecting the right brachiocephalic artery (*C*) to the right main pulmonary artery (*B*)

Glenn Shunt

The Glenn shunt is a vascular conduit that shunts blood from the superior vena cava (SVC) to the right pulmonary artery. It is used in a staged palliative procedure in patients with single-ventricle physiology. Eventually, a more definitive Fontan shunt is performed to also supply blood from the inferior vena cava (IVC) to the pulmonary arteries. Glenn shunts typically are performed at 3–9 months of age, when pulmonary vascular resistance has decreased. If a left SVC also is present, bilateral bidirectional Glenn shunts are performed.

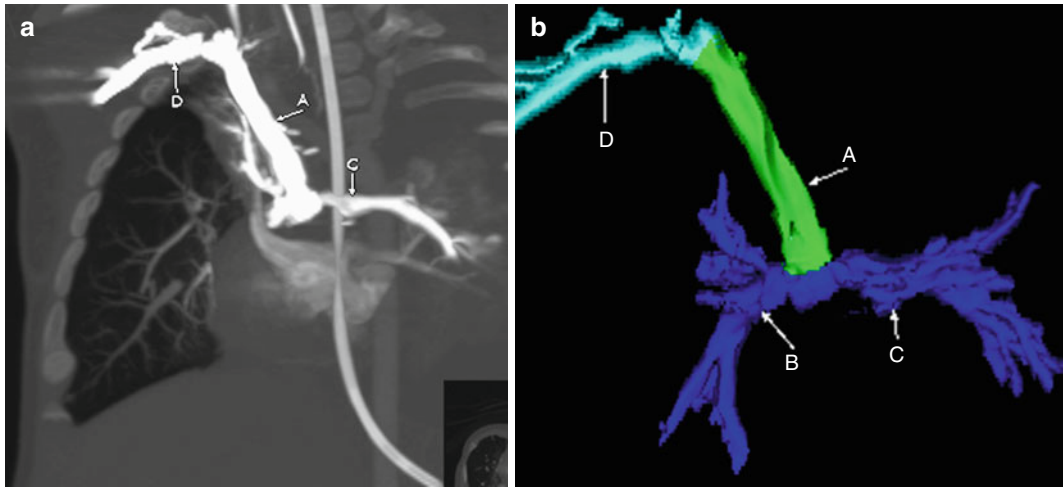

Fig. 11.3 Coronal MIP (**a**) and color-coded 3D (**b**) images showing a Glenn shunt channeling upper-extremity blood flow from the SVC (*A*) to the right pulmonary artery (*B*). The flow is bidirectional, with the left pulmonary artery (*C*) also filling. Contrast was injected from the right upper extremity, with the right subclavian vein (*D*) also opacified

Fontan Procedure

The Fontan procedure involves the placement of a vascular shunt that channels blood from the IVC to the pulmonary artery. It is the final procedure for patients with single-ventricle physiology to provide definitive blood flow to the pulmonary arteries. The surgery may be intra-atrial, in which a tunnel is created to channel blood to the pulmonary arteries, or extra-atrial in which a synthetic graft is placed to connect the IVC to the pulmonary arteries.

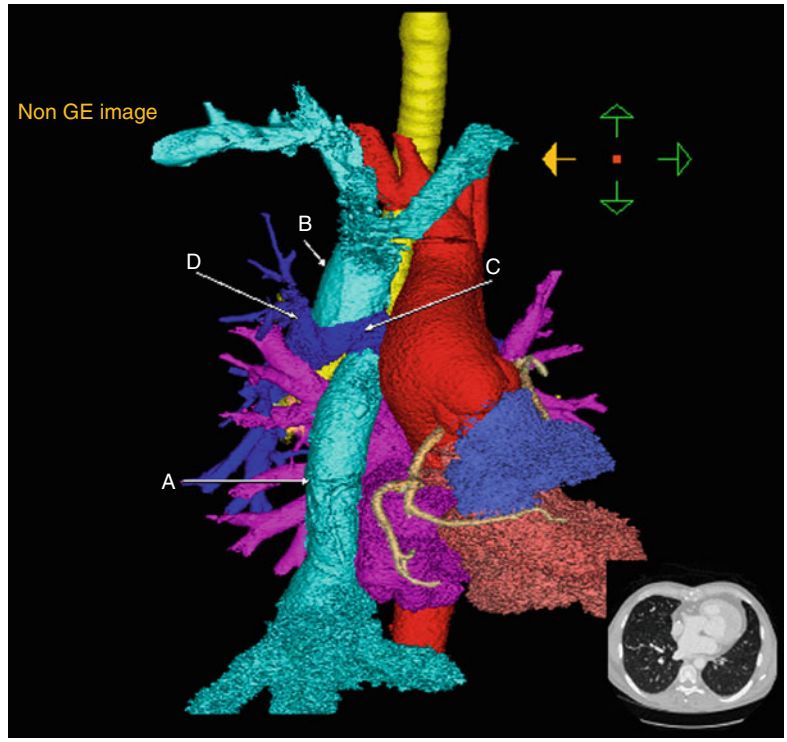

Fig. 11.4 Color-coded 3D image from a CTA scan showing an extracardiac Fontan graft (*A*) extending from the IVC to the main pulmonary artery (*C*). Also notice the presence of a Glenn shunt (*B*) channeling blood from the SVC to the right main pulmonary artery (*D*)

Hybrid Procedure

The hybrid procedure typically is performed for palliation in patients with hypoplastic left heart syndrome who may not be candidates for a stage I Norwood procedure. In the hybrid procedure, which does not require cardiopulmonary bypass, a vascular stent is placed in the patent ductus arteriosus (PDA) while bilateral pulmonary banding is performed to limit pulmonary blood flow.

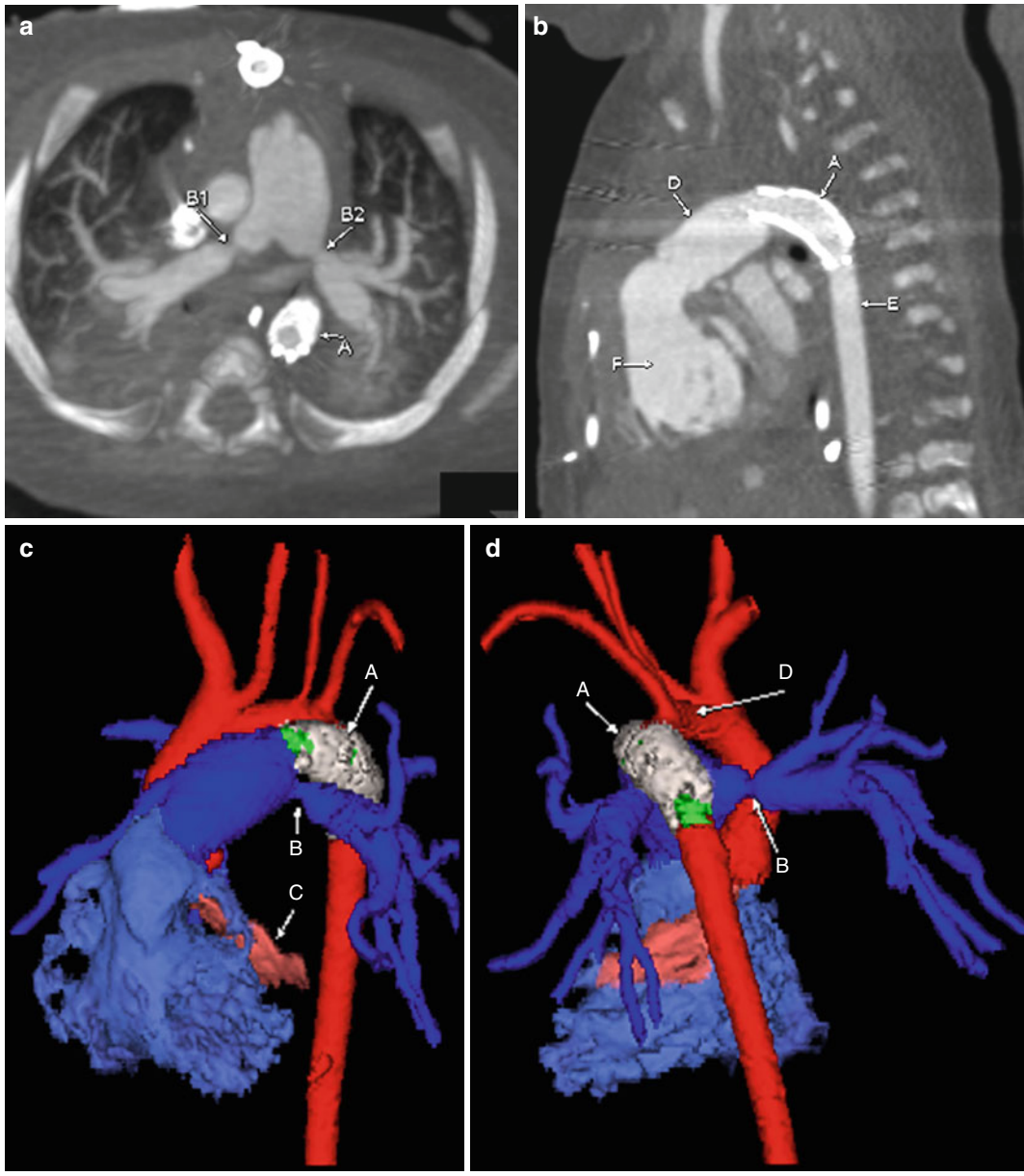

Fig. 11.5 Axial (**a**) and sagittal (**b**) CT images and lateral (**c**) and posterior (**d**) projection color-coded 3D images showing a stent (*A*, *white*) in the PDA (*green*). Notice that the PDA stent (*A*) is across the connection between the transverse aortic arch (*D*) and the descending aorta (*E*), but the holes in the stent allow retrograde flow to the transverse arch. Also note the narrow proximal right and left pulmonary arteries (*B*, *B1*, *B2*) from bilateral pulmonary artery banding. The left ventricle (*C*) is markedly hypoplastic compared with the right ventricle (*F*, *light blue*)

Norwood Procedure

The Norwood procedure is a three-part surgery performed for single-ventricle circulation, such as in patients with hypoplastic left heart syndrome. In part I (Norwood), the main pulmonary artery is disconnected from the right and left pulmonary arteries and connected to the upper portion of the aorta, creating a neoaorta. The aortic arch is widened, if necessary, with homograft tissue. A modified BT and/or Sano shunt is placed, and the atrial septum is resected. In part II (Glenn shunt), the SVC is attached to the pulmonary arteries, the modified BT shunt is taken down, and a patch is placed in the superior aspect of the right atrium. Part III (fenestrated Fontan) usually is performed at 12–24 months of age. During this stage, a graft is placed to channel blood from the IVC to the pulmonary arteries. A fenestration is created between the graft and left atrium to decompress if needed.

Norwood Part I

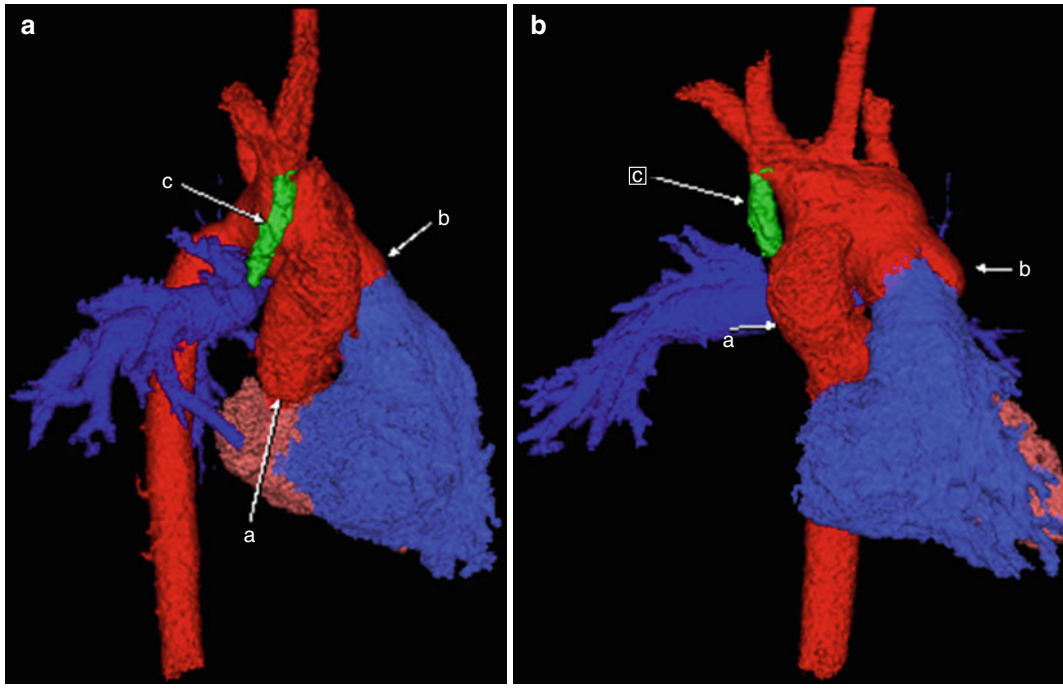

Fig. 11.6 (**a**, **b**) Color-coded 3D images showing a neoaorta (*red*) created by joining the main pulmonary artery (*b*) with the ascending aorta (*a*). A vascular conduit (*c*) connects the brachiocephalic artery to the right pulmonary artery, compatible with the modified BT shunt (*c*)

Norwood Part II

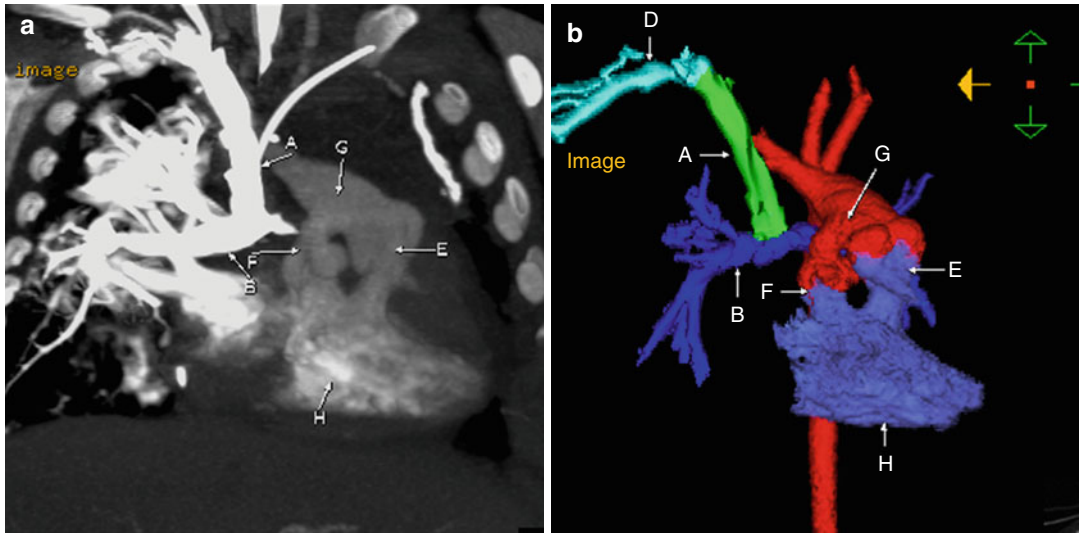

Fig. 11.7 Coronal MIP (**a**) and color-coded 3D (**b**) images from a cardiac CTA scan showing a Glenn shunt connecting the SVC (*A*) to the right pulmonary artery (*B*). Notice the neoaorta (*G*) created by joining the main pulmonary artery with the ascending aorta. Also notice that the right (*E*) and left (*F*) ventricular outflow tracts come off the right ventricle (*H*), which is the only functional ventricle

Norwood Part III

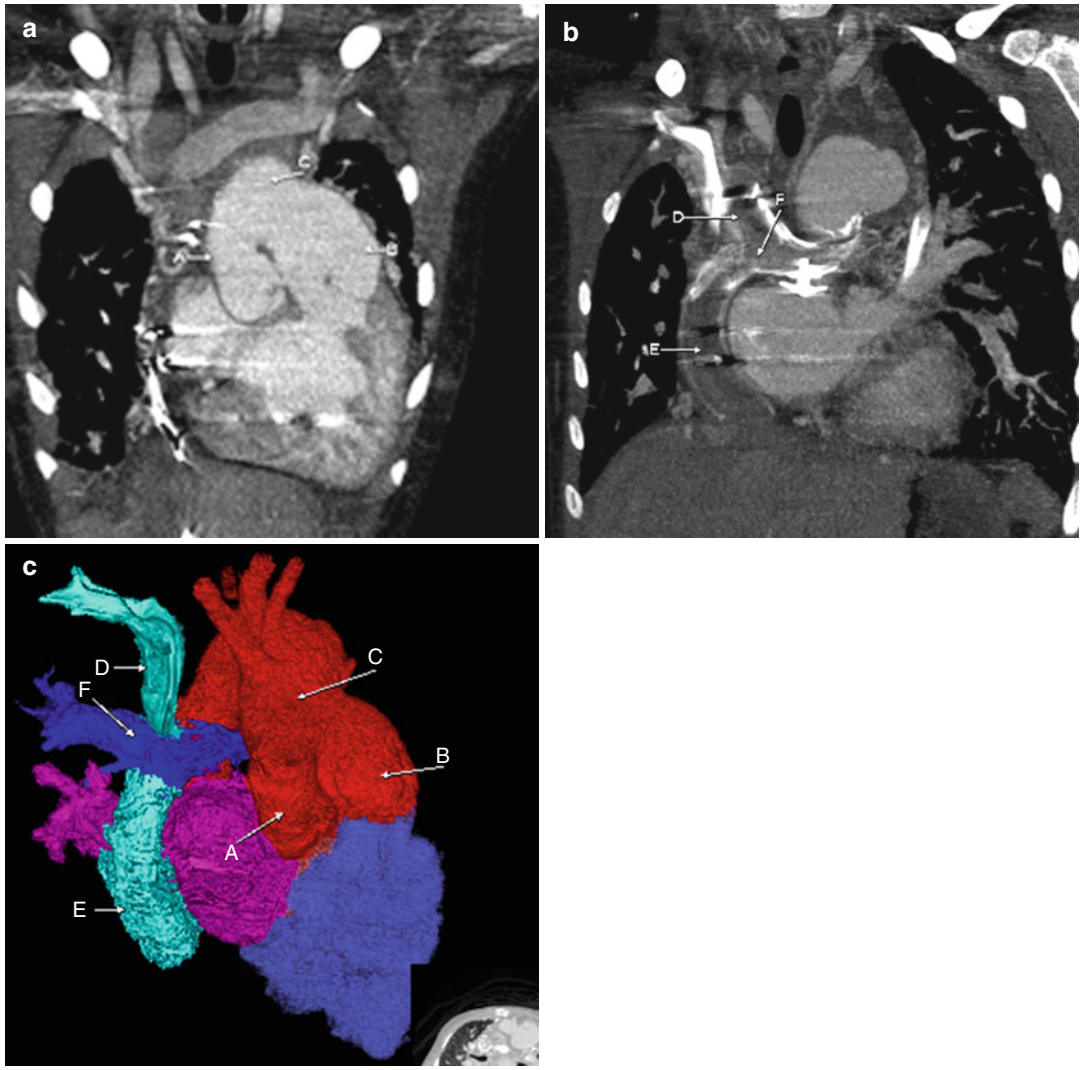

Fig. 11.8 Coronal CT (**a, b**) and color-coded 3D (**c**) images showing the result of a Norwood part III operation, with an extra-atrial Fontan shunt (*E*) channeling blood around the atrium (*pink*) to the pulmonary artery (*F*). A Glenn shunt also is present, with the SVC (*D*) shunting blood to the main pulmonary artery (*F*). Notice the neoaorta (*C*) created by joining the main pulmonary artery (*B*) with the ascending aorta (*A*)

Arterial Switch or Jatene Procedure

The arterial switch or Jatene procedure is a surgery performed to correct transposition of the great arteries. The aorta and pulmonary artery are switched, resulting in the typical "straddled" appearance of the pulmonary arteries draped around the ascending aorta. The coronary arteries are transected and reimplanted into the neoaortic root. The procedure typically is performed during the first few weeks of life.

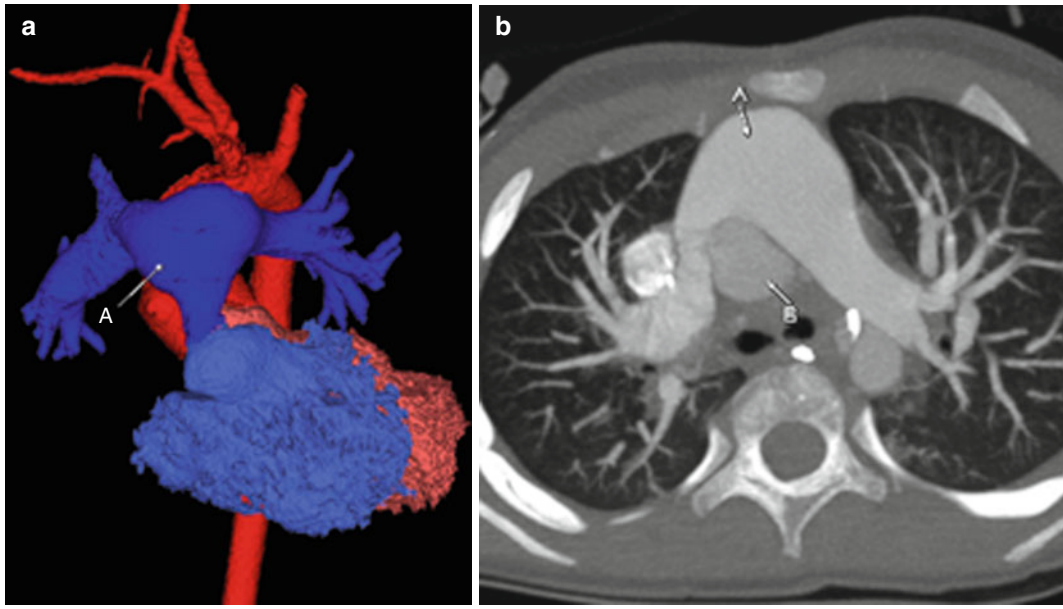

Fig. 11.9 Color-coded 3D (**a**) and axial MIP (**b**) images from a cardiac CTA scan showing the main pulmonary artery (*A*) anterior to the aorta (*B*) status post arterial switch procedure. Notice how the right and left main pulmonary arteries are draped around the ascending aorta

Fig. 11.10 Axial (**a**) and coronal (**b**) MIP images and color-coded 3D images (**c** and **d**) from a cardiac CTA scan showing the SVC (*A*) and IVC (*B*) draining into the left atrium (*C*). Deoxygenated blood then pumps from the left ventricle to the pulmonary arteries (*blue*). Oxygenated blood then returns blood to the right atrium (*F*) via the pulmonary veins (*E*). The right ventricle (*purple*) pumps blood to the aorta (*red*) in this patient with dextrotransposition of the great arteries status post Mustard procedure. Notice the typical appearance of the atrial appendage of the left atrium (*D*)

Mustard Procedure

Historically, the Mustard procedure has been performed in patients with transposition of the great arteries. It currently is not the surgery of choice for transposition repair, except in patients with L-type transposition of the great arteries, in which both the venous and arterial systems must be switched. During the Mustard procedure, the pulmonary veins are baffled to the right atrium and the SVC and IVC are baffled to the left atrium. The mustard procedure results in diverted oxygenated blood to the right ventricle, which pumps it to the body, and deoxygenated blood to the left ventricle, which pumps it to the lungs in patients with transposition.