

Review Article



Computed Tomography in the Evaluation of Fontan Circulation

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Conflict of Interest

The authors have no financial conflicts of
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ABSTRACT

The Fontan procedure is a well-established surgical technique to improve survival in patients with univentricular heart disease. The procedure reroutes the systemic venous flow to the lungs, bypassing the right ventricle. The originally proposed method involved direct anastomosis of the right atrium to the pulmonary artery. Since then, several modifications have been made in the original technique leading to the modern Fontan, or total cavopulmonary connection. The modern Fontan technique has shown improved surgical outcomes and increased life expectancy in patients with univentricular disease. Due to the increased survival of these patients, long-term complications are becoming more prevalent. Common complications of Fontan procedure include right atrial dilatation and thrombosis; conduit stenosis and thrombosis; right-to-left and left-to-right shunts; hepatic congestion and cirrhosis; and lymphovascular. Computed tomography (CT) can reliably depict the normal Fontan anatomy and various postoperative complications. A fundamental understanding of the techniques of CT, including imaging protocols and common interpretive pitfalls, allows targeted imaging and precise reporting of clinically significant findings. Radiologists should be familiar with the multiple stages of single-ventricle palliation, normal Fontan anatomy, pathophysiology, and imaging features of common Fontan-related complications.

Keywords: Fontan circuit; Fontan circulation; Fontan operation; Fontan palliation; Computed tomography

INTRODUCTION

The Fontan procedure refers to the surgical technique that diverts the systemic venous blood flow to the pulmonary arterial circulation, bypassing the right ventricle (RV). It was initially performed by Fontan in 1971 for tricuspid atresia (TA) repair.¹⁾ It is the primary surgical technique for a wide variety of congenital heart diseases with single-ventricle physiologic characteristics, like TA, mitral atresia, hypoplastic left heart syndrome, double inlet left ventricle (LV), and unbalanced atrioventricular septal defect. Since its original description, several modifications have been made to advance the procedure for better patient outcome. The modern Fontan procedure has shown good results in terms of reduced postoperative mortality and morbidity. Due to the increased longevity of these patients after

Fontan procedure, long-term complications have increasingly been recognised. Computed tomography (CT) is an important imaging modality for the serial evaluation of these patients. CT provides an accurate and reliable assessment of normal Fontan anatomy and various Fontan-related complications. The purpose of this review is to illustrate the normal Fontan anatomy; elucidate the role of CT, including suggested protocols and interpretive pitfalls; and discuss the pathophysiology and imaging features of common Fontan-related complications.

ANATOMY OF THE FONTAN CIRCUIT

Functional single ventricle represents a spectrum of congenital heart disease, with multiple anatomic variations but similar surgical treatment strategies. Anatomically, it is defined as 1) connection of both atria to the same ventricle (2:1 connection) or as 2) connection of both the atria to separate ventricles, one of which is hypoplastic (1:1 connection). The surgical palliation of functional single ventricle began in 1971 with the treatment of TA, described by Fontan and Baudet. The original Fontan procedure involved a direct atriopulmonary connection between the right atrium (RA) and left pulmonary artery (PA) using a homograft. It was postulated that the contractile function of the RA would improve the pulmonary flow. However, it was seen the RA dilates with time after the procedure and loses contractility, resulting in turbulent flow and right atrial thrombosis.²⁾ This surgical palliative technique was further redefined, and many variations were built upon it, leading to the modern Fontan or total cavopulmonary connection. As the name suggests, the procedure establishes a direct connection between both the superior and inferior vena cava and the pulmonary arteries, bypassing the RA and RV. A single-stage total correction results in massively increased pulmonary blood flow, leading to pulmonary congestion and pleural effusions. Therefore, the cavopulmonary connections are made in a staged manner to reduce the postoperative mortality and morbidity; and to facilitate cardiopulmonary adaptation to the new physiology. Currently, the total cavopulmonary Fontan circulation is performed in two stages: the bidirectional cavopulmonary shunt (or bidirectional Glenn) and Fontan completion procedure (intratrial or extracardiac) (**Figure 1**). Before the construction of cavopulmonary connections, a palliative procedure is typically performed in the early postnatal period to augment the pulmonary flow. The commonly performed palliative procedure is a modified Blalock-Taussig (BT) shunt, in which a polytetrafluoroethylene (or Gore-Tex) graft is interposed between the subclavian artery and the pulmonary artery. The purpose of this shunt is to provide regulated blood flow to the lungs, allowing growth of the pulmonary arteries in preparation for the complete future repair.³⁻⁵⁾

The first stage of creating the Fontan circulation involves bidirectional cavopulmonary shunt (or bidirectional Glenn) formation, performed at the age of 4–10 months. It involves directing the superior vena cava (SVC) blood to the pulmonary arteries. The SVC is ligated and anastomosed with the right pulmonary artery in an end-to-side manner. Due to the confluent nature of the pulmonary arteries, blood from the SVC advances into both pulmonary arteries.⁶⁾ An alternative to bidirectional Glenn is the hemi-Fontan procedure, which involves diversion of the SVC blood to the pulmonary arteries by creating a baffle between the SVC-RA confluence and the central pulmonary artery. The antegrade pulmonary flow is interrupted by transecting the central PA close to the cardiac end. In comparison to the bidirectional Glenn procedure, the natural SVC-RA confluence remains intact. This simplifies the lateral tunnel procedure, as there is no competition between the two opposite streams of blood flow from the SVC and inferior vena cava (IVC). Though the outcomes of the bidirectional Glenn

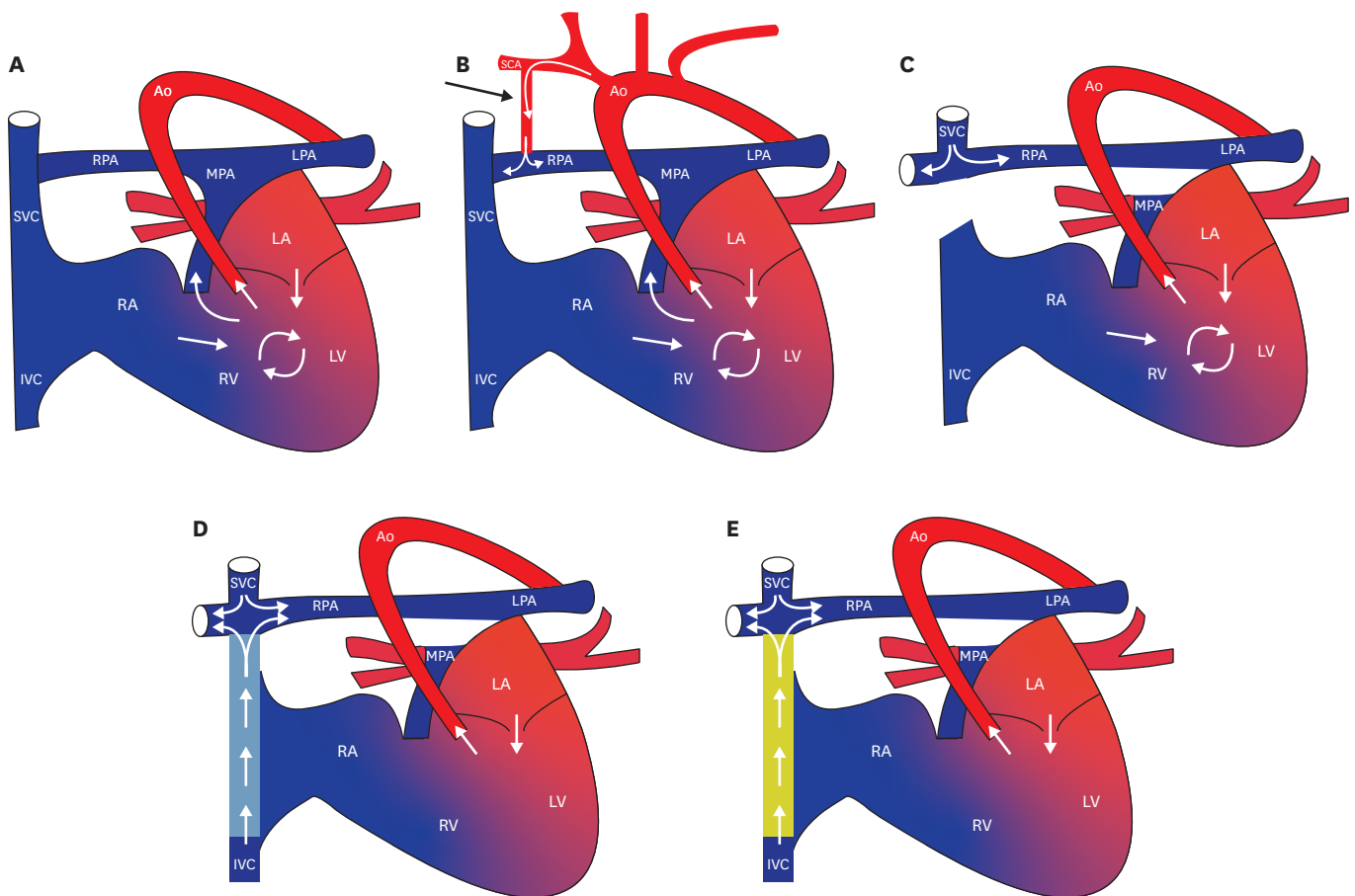


Figure 1. Graphical illustration of univentricular anatomy and modern Fontan circulation. (A) Diagram shows a univentricular heart. Blood from both the atria drains into a functionally single ventricle, and the mixed blood is then pumped into the aorta and pulmonary artery. (B) Diagram shows a modified Blalock-Taussig shunt. A graft (black arrow) is placed between the subclavian artery and the right pulmonary artery. (C) Bidirectional Glenn procedure. The cavoatrial junction is ligated and the superior vena cava is anastomosed to the right pulmonary artery. (D) Lateral tunnel Fontan operation. The inferior vena cava is connected to the pulmonary artery via an intra-atrial lateral tunnel made of the posterior atrial wall and a prosthetic patch. (E) Extracardiac Fontan operation. A tube graft or a conduit is placed entirely outside the atrium, and it connects the transected inferior vena cava and the pulmonary artery, bypassing the right atrium. Ao: aorta, IVC: inferior vena cava, LA: left atrium, LPA: left pulmonary artery, LV: left ventricle, MPA: main pulmonary artery, RA: right atrium, RPA: right pulmonary artery, RV: right ventricle, SCA: subclavian artery, SVC: superior vena cava.

and hemi-Fontan procedures are similar in the early postoperative period, the future lateral tunnel performs better with the hemi-Fontan procedure.⁷⁾

The second step, also called as, Fontan completion is performed at the age of 1–5 years and involves connecting the IVC to the pulmonary arteries by a conduit. The conduit can be intra-atrial or extracardiac.⁸⁾ In the intra-atrial method, a tunnel is created inside the RA using the right atrial wall and synthetic material (**Figure 2**). This is also called lateral tunnel procedure. In the extracardiac Fontan procedure, a polytetrafluoroethylene tube graft is placed between the IVC and right pulmonary artery (**Figure 3**).

In some high-risk patients, fenestrations are made between the lateral tunnel and the RA. These fenestrations act as a pressure release valve to prevent pulmonary volume overload. Recent studies indicate that the fenestrated approach leads to fewer early postoperative complications, such as pleural effusions and Fontan failure, and shorter lengths of hospitalization; however the long-term outcomes are similar to non-fenestrated method.^{9,11)}

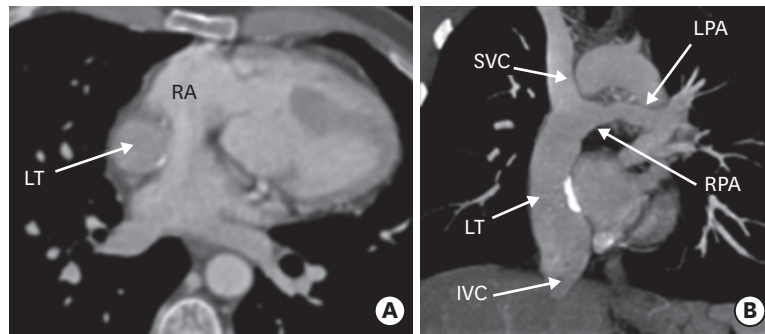


Figure 2. Lateral tunnel Fontan procedure. Axial (A) contrast-enhanced computed tomography image shows an intra-atrial lateral tunnel baffle in the right atrium. Oblique (B) coronal reformatted image show the anastomosis of the superior and inferior ends of the lateral tunnel with the right pulmonary artery and the inferior vena cava, respectively. The SVC is divided and connected to the right pulmonary artery.
IVC: inferior vena cava, LPA: left pulmonary artery, LT: lateral tunnel, RA: right atrium, RPA: right pulmonary artery, SVC: superior vena cava.

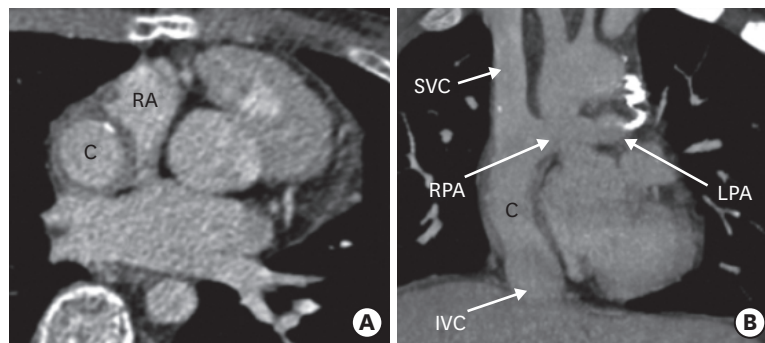


Figure 3. Extra cardiac Fontan procedure. Axial (A) contrast-enhanced computed tomography image shows an extra-atrial conduit placed entirely outside the right atrium. Oblique (B) coronal reformatted image shows the superior and inferior connections of this conduit to the right pulmonary artery and inferior vena cava, respectively. The superior vena cava is connected to the right pulmonary artery.
C: conduit, IVC: inferior vena cava, LPA: left pulmonary artery, RA: right atrium, RPA: right pulmonary artery, SVC: superior vena cava.

Patients with heterotaxy syndrome require more palliative surgeries before definitive correction. One such technique, the Kashiwama procedure, is performed in patients with left isomerism. It includes an end-to-side anastomosis of the SVC and hemiazygos vein to the ipsilateral pulmonary arteries and ligation of the main pulmonary artery (**Figure 4**). This operation diverts the entire systemic venous flow to the pulmonary arteries, except for the hepatic venous return. The selective deficiency of hepatic venous blood in the pulmonary circulation predisposes to pulmonary arteriovenous malformations (PAVM). Diversion of the hepatic venous blood into the pulmonary circulation using the intra- or extracardiac Fontan conduit resolves these PAVMs.⁵⁾ Studies have indicated that early mortality after the Fontan procedure is higher in heterotaxy patients, but the long-term survival is comparable to the overall Fontan population.¹²⁾

OVERVIEW OF THE MODALITIES

Patients with Fontan repair are evaluated by cardiopulmonary exercise testing, transthoracic echocardiography (TTE), CT, cardiovascular magnetic resonance (CMR) imaging, and cardiac catheterization. TTE is the most commonly available imaging tool, but its utility is

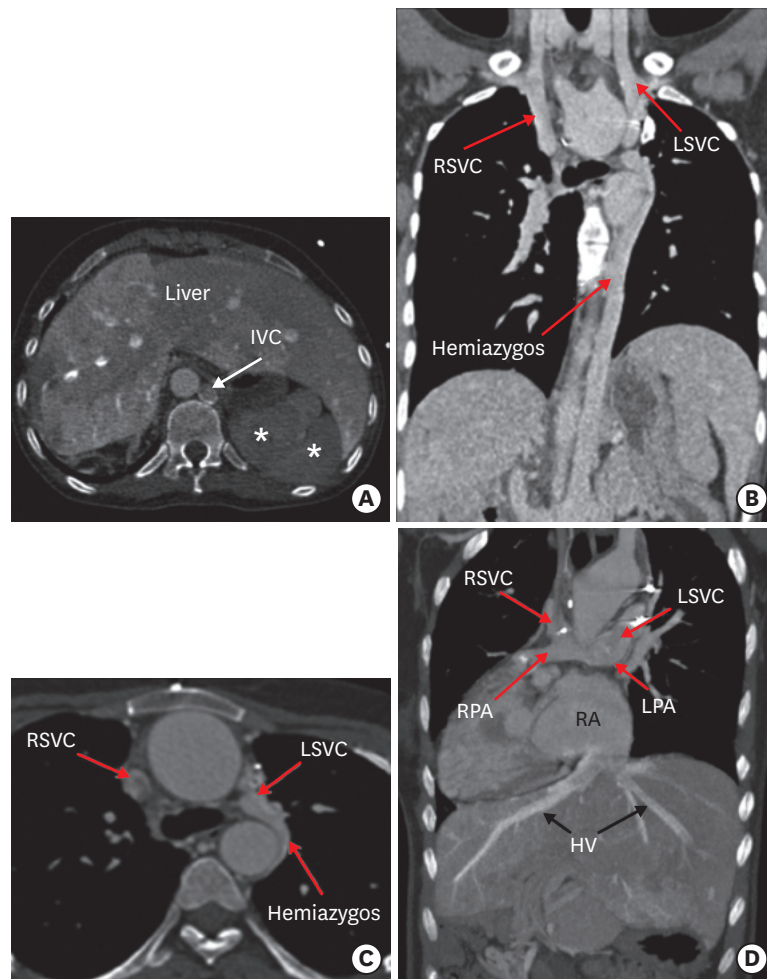


Figure 4. Kawashima procedure in a case of left isomerism. Axial (A) CECT image shows a midline liver, polysplenia (*), and a left-sided IVC. A coronal (B) maximum intensity projection image shows the hemiazygos continuation of the left sided IVC in the thorax, which finally drains into a left-sided SVC. Axial (C) CECT image shows draining of the left hemiazygos vein into the left-sided SVC. Coronal (D) maximum intensity projection image shows dextrocardia and bilateral cavopulmonary connections with both right and left SVCs draining into the right and left pulmonary artery, respectively. The hepatic veins drain directly into the RA. CECT: contrast-enhanced computed tomography, HV: hepatic veins, IVC: inferior vena cava, LSVC: left superior vena cava, RA: right atrium, RPA: right pulmonary artery, RSVC: right superior vena cava, SVC: superior vena cava.

substantially limited in the evaluation of conduits and cavopulmonary anastomosis. CMR provides a comprehensive assessment of conduit anatomy, flow velocities, and ventricular functions; however, it has a limited role in assessing ambiguous morphology and in patients with contradictions to MRI (pacemakers, stents, and unstable patients). The pacemaker is a crucial factor in imaging Fontan circuit. A study by Hoashi et al.¹³⁾ showed that approximately 33% of patients with Fontan procedure required pacemaker implantation later in the life. Recent guidelines recommend that MRI can be performed in patients with implanted devices (MRI conditional or not) as long as safety conditions are met, but the significant artifacts generated due to the device, mainly from the packing box implanted in the chest wall, make the study non-diagnostic.¹⁴⁾

CT is an alternative method to assess the morphology of Fontan conduit and various Fontan related complications. It provides a comprehensive analysis of cardiac and extracardiac

structures together with ventricular functional assessment if needed. The main advantages of CT over MRI are the short acquisition time, high spatial resolution, and ability to image patients with various metallic stents, pacemakers, and defibrillators. Because of the quick acquisition time, there is no need for sedation, making it a modality of choice for pediatric and hemodynamically unstable patients. The advent of third generation dual source CT (DSCT) scanners has made it possible to scan non-cooperative patients without anaesthesia due to higher scan speed and improved temporal resolution (75 ms for second generation DSCT and 66ms for third generation DSCT). In spite of the high heart rates in children (around 120 beats per minute), the high pitch mode provides diagnostic images in most of the scans.¹⁵⁾ ECG-gated scan can calculate both RV and LV volumes and ejection fraction. The construction of three-dimensional (3D) models is another advantage of CT. Sometimes, it is difficult to comprehend the visualization of cardiac structures on 2D images. CT data can be used to create patient-specific 3D heart models which can improve spatial visualization; assist in preoperative planning; and serve as useful tools in medical education and training. Recent studies have validated the role of CT in the functional evaluation of patients with cavopulmonary connections. A study by Piotr et al. indicates a significant positive correlation between the circumference and area of the conduit, and the predicted maximum oxygen uptake. These findings support the more extensive use of cardiac CT in evaluating functional status in adults after total cavopulmonary connection.¹⁶⁾¹⁷⁾

SCAN PROTOCOL

The main diagnostic challenge in evaluating Fontan circuit is the uniform and optimal opacification of the Fontan circuit and pulmonary arteries. Due to the differential timing of the opacification of the SVC and IVC, there is incomplete contrast mixing in the Fontan conduit and pulmonary arteries, producing streaming artifacts, which can mimic thrombosis (**Figure 5**). Various contrast injection protocols such as dual-injection protocol and single-injection single-phase technique have been proposed to avoid this.

The dual-injection protocol involves simultaneous contrast injection in the upper and lower extremity veins, allowing dense uniform opacification of the Fontan circuit and pulmonary arteries. Because many of these patients have right-to-left shunts due to fenestrations, attention should be paid to clear the air bubbles from the tubing to avoid the risk of embolic stroke.¹⁸⁾¹⁹⁾ The arterial diagnostic scan should be initiated once the contrast is visualized

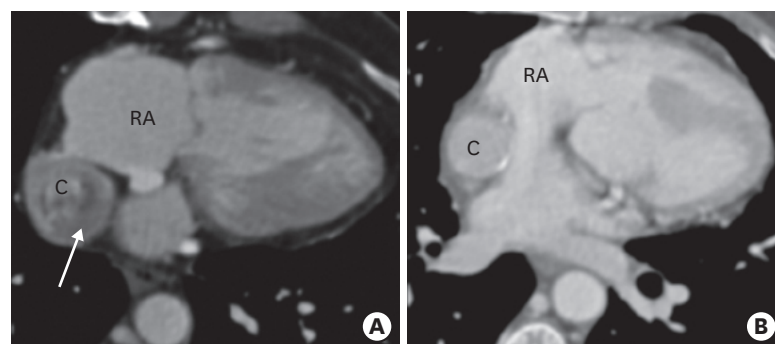


Figure 5. Streaming artifact in a lateral tunnel Fontan conduit. Axial (A) CECT image acquired in the early venous phase shows streaming artifact mimicking an intra-tunnel thrombosis (arrow). Axial (B) CECT image shows uniform opacification in the delayed venous phase. C: conduit, CECT: contrast-enhanced computed tomography, RA: right atrium.

in the left branch pulmonary artery. No further imaging is needed if the answers to clinical questions are obtained. Opacification of the conduit is usually delayed by 60–90 seconds after the first scan. Hence, a delayed venous phase is acquired, which provides appropriate opacification of the Fontan circuit and pulmonary arteries. It is not possible to enhance both the aorta and Fontan conduit in the same phase settings. Therefore, an early aortic phase scan is followed by a delayed phase for the complete evaluation of aortic pathologies, pulmonary arteries, and the cavopulmonary circuit.

Single-injection single-phase is an alternative technique. A total of 3 mL/kg of contrast material is injected with a constant rate of 2 mL/sec, followed by a saline chaser. The scan is initiated 70 seconds after contrast administration. If the optimum opacification of the aorta is also needed, the injection rate is increased to 3–5 mL/sec a few seconds before scan initiation. The timing of the increase in the contrast rate is determined by calculating the time to the peak enhancement in the aorta from the test bolus injection. The time to peak enhancement is subtracted from the total duration of the injection. For example, if the total duration of the injection is 70 seconds and the time to the peak enhancement is 15 seconds, then the injection rate should be increased at 55 seconds (70 sec – 15 sec = 55 sec). Contrast administration should be stopped at least 10 seconds before the scan initiation to avoid streak artifacts in the pulmonary arteries and SVC.²⁰⁾

COMPLICATIONS AND THEIR IMAGING FEATURES

A number of complications are seen after Fontan procedure. These complications can be divided into early or late and cardiac or extracardiac categories. The early complications (≤ 6 months after surgery) include Fontan obstruction, persistent pleural effusion, chylothorax, ventricular dysfunction, and atrioventricular valve regurgitation. Lymphatic circulation plays a significant role in the development of early Fontan complications. A study by Ghosh et al.²¹⁾ demonstrated that patients with high-grade perfusion abnormalities on preoperative T2 imaging are more prone to early postoperative complications. Delayed complications can be cardiac (progressive exercise intolerance, ventricular failure, arrhythmias, sudden cardiac death), conduit-related (conduit stenosis, thrombosis, wall calcification), vascular (PAVM, aortopulmonary collaterals [APC], pulmonary thromboembolism), hepatic (hepatic congestion, cardiac cirrhosis, regenerative liver nodules), and lymphovascular (**Table 1**).²²⁾²³⁾

Fontan failure is a complex clinical syndrome in which the Fontan circuit can no longer meet the demands of the body or drain the systemic return and lymphatic circulation. It results from various causes, such as structural issues (obstruction in the Fontan pathway), rhythm disturbances, elevated pulmonary vascular resistance, and single-ventricle dysfunction.²⁴⁾ CT is an effective modality to diagnose most of these complications. The imaging features of common post-operative complications are discussed below.

Conduit thrombosis

Thromboembolic complications frequently occur after the Fontan procedure and are a cause of significant morbidity. Several risk factors have been proposed, including low flow rates, blood stasis, associated cardiac arrhythmias, pulmonary hypertension, and the type of conduit material. Flow in the Fontan conduit is driven by passive force. Pulmonary hypertension leads to slow venous flow, which predisposes to right atrial thrombosis.²⁵⁾ A higher risk of thrombosis is seen with polyethylene terephthalate grafts compared to

Table 1. Table showing Fontan-related complications

Conduit-related
Conduit thrombosis
Conduit stenosis
Conduit calcification
Cardiac issues
Ventricular failure
Arrhythmias
Sudden cardiac death
Vascular
Pulmonary arteriovenous malformations
Aortopulmonary collaterals
Pulmonary thromboembolism
Pulmonary hypertension
Pulmonary venous stenosis
Fontan-associated liver disease
Chronic hepatic congestion
Cardiac cirrhosis
Regenerative hepatic nodule
Hepatocellular carcinoma
Lymphovascular
Chronic pleural effusion
Pericardial effusion
Chylothorax
Protein losing enteropathy
Plastic bronchitis
Renal
Chronic renal failure

polytetrafluoroethylene grafts.²⁶⁾ On CT angiography, the thrombus usually appears as eccentric hypodensity which persists in the delayed phase (**Figure 6**). Care should be taken to differentiate swirling artifacts from thrombosis.

Conduit stenosis

Stenosis of the conduit and its venous connections is a well-known late complication of the Fontan procedure. Stenosis can be proximal, distal, or diffuse along the entire conduit length (**Figure 7**). The various etiological factors for conduit stenosis include conduit calcification, thrombosis, neo-intimal hyperplasia, and stretching at the anastomotic site due to physical growth. The stenosis can obstruct the systemic venous return and may require angioplasty or

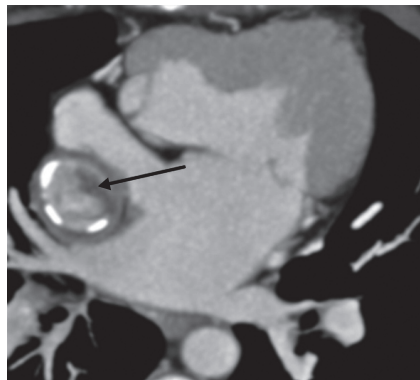


Figure 6. Thrombosis in an extra-atrial Fontan conduit. Axial maximum intensity projection image shows an eccentric hypodensity in the extra-atrial Fontan conduit which persists in the delayed venous phase, suggesting thrombosis (arrow).
C: conduit, RA: right atrium.



Figure 7. Calcification in an extra-atrial Fontan conduit. Coronal contrast-enhanced computed tomography image shows calcification in the conduit wall leading to mild luminal narrowing.

surgical correction. CT provides plentiful information regarding conduit stenosis, including the site, severity, and cause. Multiplanar reformations allow the detailed analysis of the conduit and calculations of various diameters before angioplasty.²⁷⁾

APCs

The development of APCs is frequently seen after the Fontan procedure. They develop in response to arterial hypoxemia and lead to left-to-right shunting. The shunting of arterial blood into the pulmonary circulation increases the pulmonary volume overload and pulmonary resistance. Additionally, these collaterals may lead to life-threatening hemoptysis. The APCs commonly arise from the descending thoracic aorta, subclavian artery, internal mammary artery, thyrocervical trunk, intercostal arteries, and the upper abdominal aorta and its branches (**Figures 8 and 9**). In contrast to bronchial arteries, they supply blood flow to terminal respiratory units and do not travel in close relation to the bronchial tree. Maximum intensity projection images can identify the origin and course of APCs. CT can be used for the evaluation of APCs after coiling or embolization also, when CMR is contraindicated.²⁸⁾

Systemic-to-pulmonary venovenous shunts

Venovenous collaterals are formed between systemic veins and pulmonary veins as a result of elevated systemic venous pressure. They usually arise from the brachiocephalic vein, azygous and hemiazygos veins, epidiaphragmatic veins, thebesian veins, pericardial veins, hepatic



Figure 8. Aorto-pulmonary collaterals in a post-operative case of the Fontan procedure. Coronal maximum intensity projection image shows numerous aorto pulmonary collaterals (arrow) arising from the brachiocephalic artery (white arrow) and supplying the right pulmonary circulation.

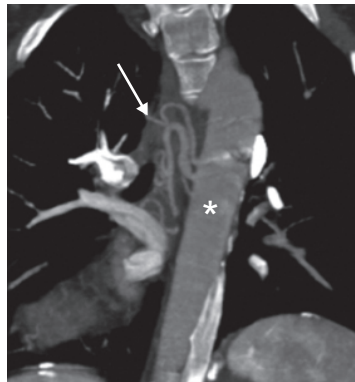


Figure 9. Aorto-pulmonary collaterals in a post-operative case of the Fontan procedure. Coronal maximum intensity projection image shows numerous aorto pulmonary collaterals (arrow) arising from the descending thoracic aorta (*) and supplying the right pulmonary circulation.

veins and drain into pulmonary veins and left atrium (**Figure 10**). The resultant right-to-left shunting may cause cyanosis. When there is significant cyanosis, they are embolized or ligated. CT can reliably assess the anatomy of systemic-to-pulmonary venovenous shunts and is helpful in guiding preprocedural planning. CT can also be used to follow up imaging of these shunts after ligation and coiling, if required.²⁹⁾

PAVMs

Patients with Fontan repairs develop multiple thin-walled PAVMs. These may range from microscopic telangiectasias to large vessels measuring 1–5 cm in diameter. The loss of pulmonary blood flow pulsatility, poor filling of the pulmonary vascular bed, and lack of or uneven delivery of hepatic venous blood to the lungs are the proposed theories for their development. There is a higher incidence of PAVMs in patients undergoing Kashiwama procedure.³⁰⁾ This occurs due to selective deficiency of hepatic venous blood; and redirecting the hepatic venous blood by Fontan completion resolves the PAVMs.³¹⁾ On CT, PAVM appear as small tangle of vessels with arterial phase filling and early opacification of the nearby pulmonary vein (**Figure 11**). PAVMs are usually located in the peripheral parenchyma. If these are large in size or symptomatic, embolization is performed. CT is also helpful for follow up imaging after the embolization procedure.

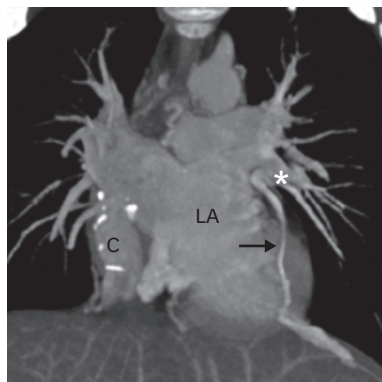


Figure 10. Systemic-pulmonary venovenous shunt in a post-operative case of the Fontan procedure. Coronal maximum intensity projection image shows a prominent systemic-to-pulmonary venous collateral (arrow) draining a sub diaphragmatic vein into the left inferior pulmonary vein (*). C: conduit, LA: left atrium.

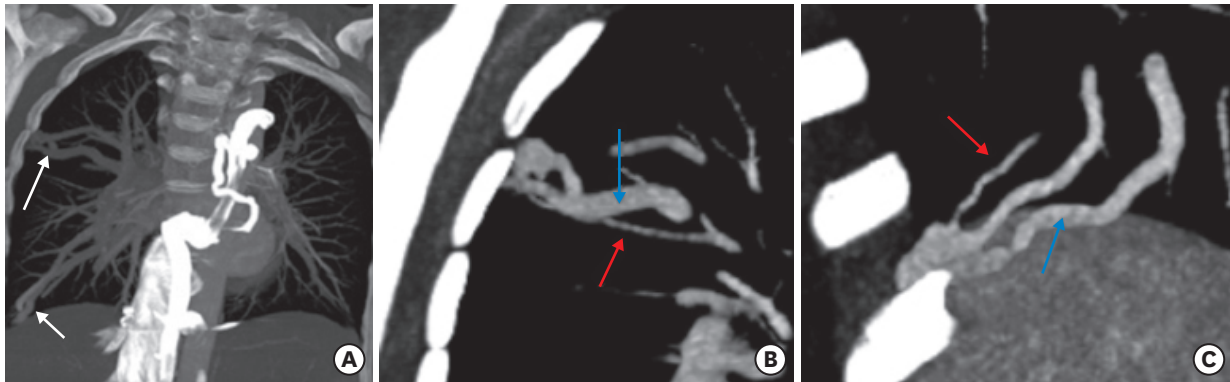


Figure 11. Pulmonary arteriovenous malformation in a patient who underwent the Kashiwama operation and a successful Fontan completion. Coronal (A) MIP image shows tangle of vessels in the right upper and lower lobes (white arrow). The corresponding magnified coronal (B, C) MIP images show an arterial feeder (red arrow) and a draining vein (blue arrow) within the tangles suggesting pulmonary arteriovenous malformations. MIP: maximum intensity projection.

Fontan-associated liver disease

There is a growing concern about the deleterious effects of chronic systemic venous hypertension after Fontan repair. The elevated right atrial pressure is transmitted to the hepatic parenchyma and causes chronic passive hepatic venous congestion. The elevated sinusoidal pressure ultimately leads to sinusoidal fibrosis, cirrhosis, portal hypertension, intra- or extrahepatic venovenous shunts, regenerative nodules, focal nodular hyperplasia (FNH), and, rarely, hepatocellular carcinoma. The classical appearance of these lesions on CT is helpful in early diagnosis and treatment planning. Chronic hepatic congestion is the earliest event in the sequelae of these lesions. On CT, it shows a characteristic peripheral reticular enhancement pattern on portal venous phase (**Figure 12**). It is essential to diagnose the hepatic changes at this stage because advanced hepatic fibrosis may develop insidiously. Once hepatic fibrosis sets in, the typical imaging features of cirrhosis such as surface nodularity, hypertrophy of the caudate and left lobe, dilatation of the portal vein, splenomegaly, and collateral veins become evident. Over time, multiple regenerative hepatic nodules also called FNH-like nodules appear in hepatic parenchyma. These nodules share imaging and histopathological features with FNH nodules, showing arterial phase hyperenhancement and iso-enhancement in the delayed phase (**Figure 13**). Atypical enhancement patterns may raise a diagnostic dilemma in distinguishing them from hepatocellular carcinoma.³²⁾



Figure 12. Extra-cardiac complication after the Fontan procedure. Upper abdominal contrast-enhanced computed tomography image show signs of early cardiac cirrhosis evident as a nonhomogeneous lacy reticular pattern in the hepatic parenchyma.

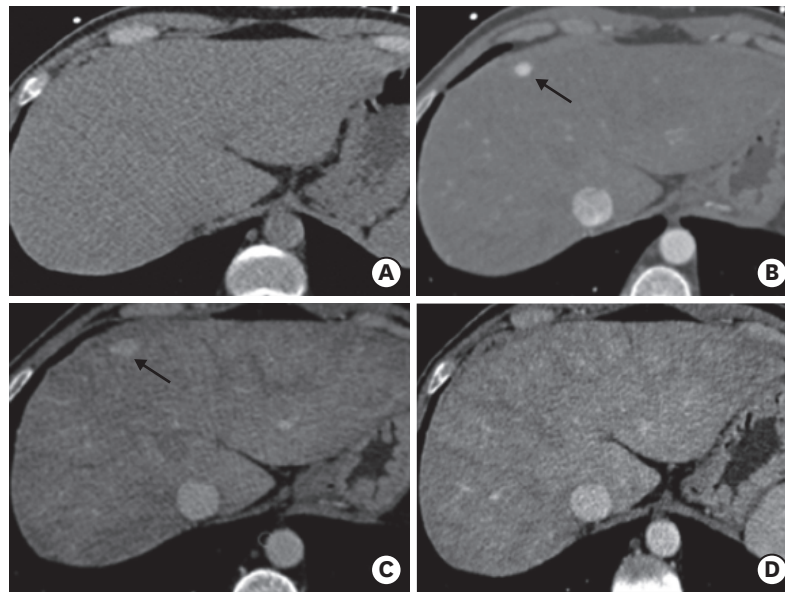


Figure 13. Extra-cardiac complication after the Fontan procedure. Plain (A), arterial (B), porto-venous (C) and delayed phase (D) upper abdominal dynamic CT scan images show a nodule in the right hepatic lobe demonstrating enhancement in the arterial phase which persists in the portal venous phase. The nodule is isodense to liver parenchyma in the delayed phase. Imaging findings are consistent with focal nodular hyperplasia-like nodules.

Lymphatic complications

Elevated central venous pressure results in increased fluid filtration and decreased lymphatic drainage. The resultant fluid accumulation leads to peripheral edema, pleural effusions, plastic bronchitis, and protein-losing enteropathy. Plastic bronchitis is a rare condition characterized by the formation of gelatinous plugs (bronchial casts) in large bronchioles. It has been proposed that elevated venous pressure predisposes to the disruption of bronchial mucosal integrity, leading to the leakage of proteinaceous material in the airways.³³⁾ CT shows branching intraluminal filling defects in the tracheobronchial tree with a variable degree of lung atelectasis or hyperinflation. Protein-losing enteropathy is another rare lymphovascular complication of Fontan surgery. Two theories have been proposed regarding its pathophysiology. One theory suggests that the elevated systemic venous pressure leads to dilation of lymphatics in the gastrointestinal tract, while another suggests that low cardiac output leads to decreased bowel perfusion and increased endothelial permeability. Protein-losing enteropathy commonly presents with hypoalbuminemia, ascites, diarrhea, and abdominal pain. There are no specific findings on CT; however, most often, it presents as submucosal edema in the small-bowel loops (**Figure 14**).³⁴⁾

CONCLUSION

The Fontan procedure is the surgery of choice in functional single-ventricle defects. The increased survival the patients after Fontan procedure necessitates meticulous longitudinal monitoring to identify various cardiac and extracardiac complications. CT can reliably depict the normal Fontan anatomy and various Fontan-related complications. Due to improved temporal resolution, fast scanning, wide field of view, multiplanar reconstruction techniques, and dose reduction algorithms, CT has emerged as an excellent modality to assess the anatomy, hemodynamics, and prognosis of patients after Fontan procedure. Radiologists



Figure 14. Protein losing enteropathy after the Fontan procedure in a patient with edema, hypoalbumenia, and chronic diarrhea. Coronal contrast-enhanced computed tomography image shows abdominal ascites and diffuse circumferential thickening of small bowel loops (arrow).

must be familiar with the normal postoperative anatomy and diverse imaging features of various Fontan-related complications. This can aid early and confident diagnoses guiding therapeutic interventions, leading to better patient outcomes.

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