CT and MRI Evaluation of the Fontan Pathway: Pearls and Pitfalls

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The Fontan pathway is the result of a palliative surgical procedure achieved by direct anastomosis of systemic veins to the pulmonary arteries, bypassing a ventricle. It is performed in patients with functional univentricular heart physiology in which biventricular repair is not possible. Advances in surgical techniques with modified Fontan procedures have led to improved long-term results and increased life expectancy in such patients. Consequently, late complications of the Fontan procedure are being increasingly encountered, particularly in patients with poor hemodynamics. Accordingly, radiologists are increasingly likely to encounter long-term complications of the Fontan pathway in certain cardiac patients. The purpose of this article is to familiarize radiologists with the surgical techniques of the Fontan procedure, to describe the technical considerations for optimal image acquisition and the expected normal postoperative anatomy, and to illustrate the imaging findings of postoperative complications in these patients.

Key words Heart defects, congenital · Fontan procedure ·
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INTRODUCTION

In functional single ventricle or univentricular hearts, one of the two cardiac ventricles may be underdeveloped or may not function normally due to lack of a normal atrioventricular valve (Fig. 1). An uncorrected single ventricle has a parallel relationship with the right-to-left shunt, causing cyanosis and volume overload and leading to heart failure. The Fontan operation is a palliative surgical procedure performed in patients with single ventricle to divert the venous flow from the superior and inferior venae cavae to the pulmonary arteries without passage through a pumping ventricle. The most common congenital cardiac abnormalities palliated with the Fontan procedure are tricuspid atresia (Fig. 1), hypoplastic left heart syndrome, pulmonary atresia with an intact ventricular septum, and double-inlet ventricle.

In 1971, Francois Fontan and colleagues proposed a surgical technique as a palliative procedure for tricuspid atresia. They initially used a classical Glenn shunt, forming a connection between the superior vena cava (SVC) and the right pulmonary artery with ligation of the SVC-right atrial junction. In addition to this, a connection between the right atrium and the left pul-

monary artery was created with an aortic homograft [1]. The Fontan procedure has undergone diverse modifications in order to improve patient outcomes (Fig. 2). In recent years, the lateral tunnel or the extracardiac Fontan operation have become the most commonly used modified methods to direct blood from the systemic venous system to pulmonary circulation.

Due to increased survival rates with the use of advanced surgical techniques, long-term complications of the Fontan circulation are more commonly observed. Imaging follow-up and diagnosis of these complications are essential for early detection and treatment. In this article, we review the normal anatomy of common variations of the Fontan pathway, various multidetector computed tomography (MDCT) and cardiac magnetic resonance imaging (CMR) techniques for optimal imaging diagnosis of the Fontan pathway, and various spectra of imaging findings regarding potential complications in patients with failing Fontan.

ANATOMY OF THE FONTAN PROCEDURE (Fig. 2)

In the classic Fontan procedure, the right atrium or the right atrial appendage is directly connected to the pulmonary arteries, collectively termed an atriopulmonary connection (Figs. 2A

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Fig. 1. A 24-year-old male patient with tricuspid atresia. (A and B) Two-dimensional reformatted images show tricuspid atresia with absent right atrioventricular connection between the right atrium (RA) and the right ventricle (RV). Note the right coronary artery within the epicardial fat of the deep right atrioventricular sulcus (arrows). Main left ventricle (LV) with hypoplastic RV is a typical morphology of functional single ventricle. Obligatory right-to-left shunt is well demonstrated by contrast media flow (arrowheads) through a large atrial septal defect (ASD), which results in cyanosis. VSD: ventricular septal defect, Ao: aorta, LA: left atrium.

Fig. 2. Diagrams showing various anatomies of classic and modified Fontan procedures. (A) Atriopulmonary Fontan operation: the right atrium or the right atrial appendage is directly anastomosed to the pulmonary artery. (B) Lateral tunnel Fontan operation: IVC is connected to the pulmonary artery via the intra-atrial lateral tunnel made of the posterior atrial wall and a prosthetic patch. The SVC is divided and re-anastomosed to the superior and inferior walls of the RPA. (C) Extracardiac Fontan operation: a tube graft or a conduit is placed entirely outside the atrium, and it connects the transected IVC and the pulmonary artery, bypassing the right atrium. The SVC is also transected and anastomosed to the superior wall of the RPA. (D) Kawashima procedure: in patients with a single ventricle along with IVC interruption and azygos continuation, cavopulmonary connection is created by division of the SVC distal to the drainage of the azygos vein and anastomosis of the cranial aspect of the SVC to the pulmonary artery. RPA: right pulmonary artery, IVC: inferior vena cava, SVC: superior vena cava.

and 3). This method was predominantly used up to the late 1980s. However, it is now understood that, as a consequence of this method, significant right atrial dilatation can result in atrial arrhythmias and atrial thrombus formation [2,3]. As a result, the classic Fontan procedure is no longer employed, and it has been replaced by the more energy efficient lateral tunnel (Fig. 2B) or extracardiac Fontan procedure (Fig. 2C). This total cavopulmonary connection comprises a variety of cavopulmonary connections including the bidirectional cavopulmonary shunt (BCPS), the lateral tunnel, and the extracardiac conduit [4,5]. The BCPS is performed to redirect SVC flow to the pulmonary circulation, bypassing the right heart by end-to-side anastomosis of the SVC to the right pulmonary artery after division of the

superior cavo-atrial junction (Fig. 4). The main pulmonary artery is typically divided to completely bypass the right heart. The BCPS is performed as a permanent palliative procedure, an intermediate procedure of a staged Fontan operation (Fig. 4), or a component of the primary Fontan operation (Figs. 5, 6, and 7).

Total cavopulmonary connection is completed by redirecting inferior vena cava (IVC) flow to the pulmonary circulation using an intra-arterial lateral tunnel or an extracardiac conduit. In the lateral tunnel method (Figs. 2B, 5, and 6), a lateral tunnel is formed by an intra-atrial tunnel-like baffle using both the lateral wall of the right atrium and a prosthetic patch. The superior aspect of the lateral tunnel is anastomosed to the inferior wall of the pulmonary artery, and the inferior aspect of the lat-

Fig. 3. A 33-year-old female patient who underwent atriopulmonary Fontan operation for tricuspid atresia. Three-dimensional volume rendered image (A) and oblique coronal and axial reformatted images (B and C) show direct anastomosis (*) of the right atrial appendage (RAA) and the main pulmonary artery (MPA). The right atrium (RA) along with the inferior vena cava (IVC) and the hepatic vein (HV) are markedly dilated. She suffered from severe dyspnea and underwent conversion to the Fontan operation. P: dilated pericardial vein, S: superior vena cava, arrow: endocardial pacemaker.

Fig. 4. A 31-month-old male infant with bidirectional cavopulmonary shunt (BCPS) for double-inlet left ventricle with rudimentary right ventricle. Three-dimensional volume rendered image (A) and oblique coronal reformatted image (B) show end-to-side anastomosis of the superior vena cava (SVC) to the right pulmonary artery (RPA) after division of the superior cavo-atrial junction. In this patient, BCPS is performed as an intermediate procedure of staged Fontan operation. LPA: left pulmonary artery.

eral tunnel is anastomosed to the divided IVC at the IVC-right atrial junction. In the extracardiac Fontan technique (Figs. 2C and 7), a polytetrafluoroethylene conduit or a tube graft is positioned entirely outside the right atrium and connects the transected IVC and the pulmonary artery, bypassing the right atrium.

In heterotaxy patients with IVC interruption and azygos continuation, the SVC incorporates most (85%) of the systemic venous flow into the heart, with the exception of the venous flow from the coronary sinus and the hepatic vein. In such patients, the cavopulmonary connection of the SVC distal to the drainage of the azygos vein to the pulmonary artery is called the Kawashima operation (Fig. 2D). Exclusion of the hepatic venous

blood from the pulmonary circulation is a major risk factor for pulmonary arteriovenous malformation (PAVM). To prevent or to alleviate PAVM, hepatic veins should be incorporated into pulmonary circulation using the Fontan procedure (Fig. 8), or a graft should be interposed between the hepatic vein and the azygos vein.

In high-risk patients, fenestration can be created between the Fontan pathway and the atrium using a window at the lateral tunnel or a tube graft on the extracardiac conduit. This fenestration can reduce early morbidity by shunting the blood from the Fontan pathway to the atrium when systemic venous pressure is elevated in the early postoperative period (Fig. 9) [6].

Fig. 5. An 18-year-old female patient who underwent lateral tunnel Fontan operation for tricuspid atresia. (A) Transverse CT image shows the lateral tunnel (LT) using an intra-atrial baffle (**) placed on the lateral aspect of the right atrium (RA). Oblique coronal reformatted (B) and volume rendered (C) images show that the superior vena cava (SVC) is divided and connected to the right pulmonary artery (RPA) superiorly (+), and that the superior and inferior ends of the LT are anastomosed to the inferior walls of the RPA and the inferior vena cava (IVC) (*). The main pulmonary artery (MPA) is divided from the ventricle (arrow). Note calcification of the patch in the lateral tunnel.

Fig. 6. A 24-year-old male patient who underwent lateral tunnel Fontan operation. Transverse (A) and oblique coronal (B) reformatted images acquired in the late venous phase show homogeneous enhancement in the lateral tunnel (LT) Fontan pathway and the pulmonary artery. (C) Right pulmonary artery (RPA) stenosis occurs at the site of anastomosis with the LT (arrow). RA: right atrium, LPA: left pulmonary artery, S: superior vena cava.

Fig. 7. A 20-year-old female patient who underwent the extracardiac Fontan procedure using a Gore-Tex tube graft for transposition of the great arteries with a small left ventricle. (A) Transverse CT image shows the Fontan conduit (c) placed entirely outside the right atrium (RA). Late venous opacification of an extracardiac Fontan pathway shows homogeneous enhancement with conduit calcifications. Oblique reformatted (B) and volume rendered images (C) show that the conduit is connected to the transected inferior vena cava (IVC) and the pulmonary artery (PA), bypassing the RA. The superior vena cava (SVC) is connected to the pulmonary artery superiorly.

Fig. 8. An 18-year-old female patient who underwent Kawashima operation and successive Fontan completion for a functional single ventricle with IVC interruption and azygos continuation. (A) Oblique coronal reformatted image shows an extracardiac Fontan conduit (c) connecting the hepatic vein (HV) and the right pulmonary artery (RPA). Note the whitish wall of a thin Gore-Tex tube graft. Anterior (B) and posterior (C) volume rendered images comprehensively show the Fontan pathway and the azygos vein (Az) draining into the left superior vena cava (LSVC). LPA: left pulmonary artery.

Fig. 9. A 20-year-old female patient who underwent the extracardiac Fontan procedure using a 20-mm Gore-Tex tube graft and a 5-mm fenestration graft. Anterior volume rendered (A) and curved reformatted images (B) show a patent fenestrated graft (arrows) connection between the Fontan conduit (c) and the right atrium (RA).

IMAGING CONSIDERATIONS FOR THE FONTAN PROCEDURE

Computed tomography

As in other congenital heart diseases, echocardiography plays a primary and definitive role in imaging of the Fontan procedure. However, echocardiography is often non-diagnostic due to a limited acoustic window, particularly in adult survivors, as well as to shadowing caused by surgical clips, stents, baffles, and conduits. Echocardiography is often insufficient to adequately assess the Fontan pathway and the pulmonary artery. CMR is a useful complementary tool for follow-up in patients who undergo the Fontan procedure in order to demonstrate morphologic abnormalities and to assess functional complications.

However, CMR is still contraindicated in patients with pacemakers and defibrillators and is not able to provide suitable image quality in patients with susceptibility artifacts due to surgical materials such as hemostatic clips, stents, and embolization coils.

MDCT has been increasingly used in the morphologic evaluation of extracardiac vasculature in congenital heart disease with the development of a faster scanner to improve temporal resolution with a decrease in cardiac motion artifacts, higher spatial resolution, isotropic reformatted images in any plane, and reduction of the radiation dose. When echocardiography and CMR provide insufficient information or when CMR is contraindicated in patients with the Fontan pathway, MDCT angiography is utilized as an alternative imaging modality to detect

complications such as thrombosis, stenosis, pulmonary embolism, pulmonary arteriovenous fistula, arterial collaterals, and venous collaterals. Using electrocardiogram (ECG) triggering or ECG gating, MDCT scans can also be utilized to evaluate intracardiac morphology and systolic function.

Diagnostic pitfall in MDCT scans: “Streaming artifact”

In patients who undergo the Fontan procedure, successful computerized tomography (CT) scanning requires optimal and uniform simultaneous contrast enhancement of the Fontan pathway and pulmonary arteries. Differential timing of opacification of the superior and inferior venae cavae, incomplete mixing in the Fontan circuit, and differential streaming of contrast into pulmonary arteries result in inhomogeneous opacification of the Fontan pathway. Therefore, proper selection of injection sites, timing of contrast administration, and initiation of scanning are critically important. Because the Fontan circuit drains two different systemic venous sources, and because Fontan circulation flow is characterized as passive laminar flow, homogeneous enhancement of the Fontan pathway cannot be obtained until the venous phase. If the acquisition of CT scans is routinely initiated before the venous phase, then the incompletely opacified blood can be either non-diagnostic or misdiagnosed as thromboembolism (Fig. 10) [7].

CT techniques for Fontan circulation

To mitigate the streaming artifact of the Fontan pathway, various enhancement protocols have been established, including dual injection techniques, delayed imaging, and bolus tracking methods.

Dual injection protocol is a method involving simultaneous injection of iodinated contrast through both upper and lower extremities, which allows denser opacification of the entire Fontan circuit. Greenberg and Bhutta [8] successfully used the dual injection technique via simultaneous intravenous (IV) injections into a dorsal foot vein and an upper extremity vein. Sandler et al. [9] performed simultaneous injections into a central lower extremity vein and an upper extremity vein, with a catheter placed in the central femoral vein under sonographic guidance, in addition to placing an IV catheter in an antecubital vein. The American College of Radiology also suggest simultaneous injection via catheters placed in both upper extremity and lower extremity veins, preferably with two power injectors. Disadvantages of the dual injection technique are invasiveness and difficulty in cases of poor IV access. Also, some patients will still have a swirling artifact, unopacified hepatic venous inflow, or incomplete mixing, all of which require a second delayed scan in the venous phase.

Another option is delayed scanning when the venous blood returns to the Fontan pathway following systemic circulation. A one-minute-delayed scan usually provides adequate contrast opacification of the intrathoracic vasculature with only minor inhomogeneity. In patients with an atriopulmonary Fontan connection, significant ventricular dysfunction, or severe atrioventricular valve regurgitation, scans should be acquired even after one minute due to slow circulation. The three-minute-delayed scan provides the most homogeneous contrast opacification for the detection of a thrombus in the Fontan pathway. However, overall reduction of contrast density can make image interpretation difficult, particularly if low-radiation dose protocols are

Fig. 10. A 14-year-old female who underwent extracardiac Fontan operation for complete AVSD. Transverse (A) and oblique sagittal (B) reformatted images acquired in the early venous phase show a streaming artifact mimicking thrombosis. The venous delayed phase was scanned too early, at about a 40 second delay, and the streaming artifact is still seen in the Fontan conduit. Note thick circumferential calcification of the conduit (arrows).

Fig. 11. (A) This arterial acquisition shows a streaming artifact in the extracardiac Fontan conduit and bilateral branch pulmonary arteries. (B) The one-minute-delayed scan shows adequate contrast opacification in the Fontan pathway. (C) The three-minute-delayed scan shows the most homogeneous contrast opacification at the expense of overall reduction of contrast density.

used (Fig. 11) [10].

An early arterial phase scan is needed for detection of aorto-pulmonary collateral (APC), which can be a source of life-threatening hemoptysis (Fig. 12). The bolus tracking method is considered the most effective method to initiate arterial phase CT scanning because of the unpredictable degree of contrast enhancement secondary to variable blood flow velocity in the Fontan pathway and the pulmonary artery [11].

Magnetic resonance imaging

When echocardiography is not feasible and is non-diagnostic, CMR can play a complementary role in obtaining comprehensive anatomical and functional information, particularly in older patients who have undergone the Fontan procedure. CMR can evaluate morphologic abnormalities, including the Fontan conduit, systemic veins, pulmonary arteries and veins, and collaterals. To evaluate any structural abnormality following the Fontan procedure, black blood spin-echo imaging and contrast-enhanced magnetic resonance angiography (MRA) are used. CMR readily provides functional parameters using flowmetry and volumetry to quantify valvular regurgitation, pulmonary and systemic blood flow, and APCs [6,12], which cannot be obtained by MDCT. Magnetic resonance imaging (MRI) evaluation is limited in patients with surgical or interventional ferromagnetic materials, which cause large susceptibility artifacts.

To obtain functional information, cine steady-state free-precession (SSFP) imaging and phase-contrast velocity-encoded cine imaging are typically used (Fig. 13) [13].

Cine SSFP imaging is used to obtain functional parameters, such as ventricular volume and ejection fraction, using volumetry. These MR parameters are thought to be the reference standard for the assessment of ventricular function, and they are clinically important for follow-up in patients who have received

the Fontan procedure.

Phase-contrast velocity-encoded imaging allows accurate flowmetry for quantitative evaluation of valvular regurgitation, pulmonary to systemic blood flow ratio (Qp/Qs), and burden of collateral flow. The Qp/Qs ratio is usually calculated across the main pulmonary artery and the ascending aorta by phase-contrast imaging, which provides important information about the presence and degree of right-to-left shunts, systemic to pulmonary venous shunts, or baffle leak. CMR also allows calculation of APC blood flow [14].

Late gadolinium enhancement (LGE) CMR is utilized to detect myocardial fibrosis and infarction. An increased extent of LGE was associated with a lower ejection fraction, increased CMR-derived ventricular end-diastolic volume index and mass index, and non-sustained ventricular tachycardia [15]. Contrast-enhanced MRA is used for the identification of collateral vessels and extracardiac vascular anatomy.

ABNORMAL IMAGING FEATURES OF THE FONTAN PATHWAY

Many patients who undergo the Fontan procedure have substantially prolonged survival and improved quality of life in comparison to those who undergo only shunt operation. Due to the prolonged survival of these patients with abnormal palliative physiology, however, late complications are being increasingly observed in children and young adults. Commonly encountered cardiac and extracardiac complications include Fontan conduit stenosis and thrombosis, SVC stenosis, peripheral pulmonary artery stenosis, right atrium dilatation and arrhythmia, pulmonary embolism, systemic venous collateralization, PAVMs, hepatic problems, and lymphatic dysfunction [3,13,16].

Fig. 12. An 18-year-old male with hemoptysis who underwent extracardiac Fontan operation for complete AVSD. Transverse (A and C) and volume rendered (B) images acquired in the arterial phase show numerous aortopulmonary collateral arteries in the mediastinum and around the bronchus. Note the metallic artifact due to embolization coils (A and B, arrows) in the intercostal and internal mammary arteries. Lung window image (C) shows ill-defined patchy consolidations and ground-glass densities in the right lower lobe, suggestive of pulmonary hemorrhage.

Fig. 13. An 18-year-old male who underwent atriopulmonary Fontan operation for an right ventricle (RV)-type functional single ventricle and hypoplastic left ventricle (LV). (A) Gadolinium-enhanced MR angiogram (MRA) shows a dilated right atrium (RA) and a patent connection between the superior aspect of the right atrium and the main pulmonary artery (MPA). (B) Gadolinium contrast material-enhanced MRA also shows the extracardiac vascular anatomy. (C) Cine b-steady-state free-precession (SSFP) sequence demonstrates RV hypertrophy and a small LV cavity with biventricular EF= 61.6% and biventricular mass=81.9 g/m². (D) Oblique coronal cine b-SSFP sequence demonstrates turbulent slow flow (arrow) in the atriopulmonary Fontan circuit. SVC: superior vena cava.

Conduit stenosis

Stenosis of the conduit usually occurs at the site of anastomosis with the pulmonary artery, and conduit problems include pseudointimal peel, thrombosis, calcification, or a small conduit relative to the physical growth of the patient. Such stenosis is a potential complication of the Fontan procedure, which causes severe symptoms of systemic venous obstruction and requires stenting or surgical replacement. MDCT can provide excellent information about the presence of conduit stenosis, along with its cause and degree (Figs. 14 and 15). Using three-dimensional volume rendering and multiplanar reformatted MDCT images, the diameters of the Fontan conduit and branch pulmonary arteries should be analyzed.

Thrombosis

Pulmonary embolism is a life-threatening thromboembolic

complication of Fontan circulation due to stasis and slow flow. In this situation, Fontan circulation has an imbalance between procoagulant and anticoagulant factors [17]. High mortality from thromboembolic events is also related to arrhythmia as a result of increased atrial pressure and distention, particularly in atriopulmonary Fontan procedures. The reported incidence of postoperative thromboembolic disease varies from 3% to 19% [9,18-20]. Moreover, a recent retrospective study of asymptomatic patients with Fontan circulation reported that 13% had a mural thrombus within the extracardiac conduit [21].

On MDCT, low-density thickening within the Fontan conduit suggests conduit thrombosis and a central filling defect surrounded by homogeneous IV contrast material, which suggests pulmonary thromboembolism (Fig. 16). CMR also provides excellent anatomic information on atrial dilatation and the presence of a thrombus. Differentiating the thrombus from any "swirl-

Fig. 14. A 15-year-old female who underwent extracardiac Fontan operation for a functional single ventricle, coarctation of the aorta, and supra-aortic (arrow) stenosis. (A) Non-opacified contrast is still seen in the Fontan conduit, which resulted in incomplete evaluation of conduit thrombosis. (B) Significant stenosis is noted in the mid portion of the conduit due to folding of the graft and thick wall calcification (arrow). She suffered from pleural effusion due to significant obstruction in the Fontan conduit.

Fig. 15. A 20-year-old female who underwent extracardiac Fontan operation with an 18-mm Hemashield vascular graft for a functional single ventricle with double-outlet right ventricle. Oblique coronal reformatted (A) and axial images (B) obtained with a late venous phase CT scan show severe conduit (c) stenosis caused by concentric wall calcifications (arrows). (C) Mild left branch pulmonary artery stenosis is noted on the MPR image (arrows). SVC: superior vena cava, IVC: inferior vena cava, RPA: right pulmonary artery, LPA: left pulmonary artery. MPR: multiplanar reformatted.

ing artifacts” of a Fontan conduit is potentially difficult on both MDCT and CMR. A thrombus is most reliably identified using delayed MDCT scanning in the venous phase and contrast-enhanced MRA [16].

Pulmonary arteriovenous malformation

Although the etiology of PAVM remains unclear, the absence of pulsatile blood flow, underfilling of the pulmonary arteries, and the relative lack or asymmetrical distribution of hepatic venous blood to the pulmonary circulation appear to be possible factors (Fig. 17). Also, the Fontan conduit is thrombogenic because of venous stasis and low passive flow. It has been pos-

tulated that a hepatic factor exists, and that it prevents the opening of arteriovenous communications. Bernstein et al. [22] reported that 60% of patients with a cavopulmonary shunt developed PAVM. Heterotaxy patients with left isomerism, interrupted IVC, and azygos continuation who underwent the Kawashima operation showed an increased incidence of PAVM relative to those who underwent the Fontan operation (Fig. 18). In patients who undergo the Kawashima operation, the IVC drains through an azygos vein into the SVC. Accordingly, only hepatic veins drain into systemic circulation, thereby bypassing pulmonary vasculature. On MDCT, abnormally enlarged pulmonary vessels, which form a small tangle of vessels extending

to the periphery of the lung, suggest PAVM [23]. Both MDCT and contrast-enhanced MRA can accurately demonstrate PAVM [24].

Systemic-pulmonary venovenous shunts (venous collaterals)

Venovenous collaterals from the systemic vein to the pulmo-

nary vein are frequent in patients who undergo the Fontan operation as a consequence of elevated central venous pressure. Desaturation by right-to-left shunts through venovenous collaterals may cause cyanosis. When cyanosis is significant, venovenous collaterals are embolized using an embolization coil or a vascular plug. MDCT is able to demonstrate venovenous collaterals, especially in the early arterial phase (Fig. 19) [23].

Fig. 16. A 15-year-old male who underwent extracardiac Fontan operation for functional single ventricle with crisscross heart. Transverse (A) and oblique coronal (B) reformatted images show multifocal intraluminal filling defects in bilateral jugular veins and low-density thickening within the extracardiac Fontan conduit (arrows), which is suggestive of venous and conduit thrombosis.

Fig. 17. A 16-year-old male who underwent extracardiac Fontan operation for tricuspid atresia. (A) Oblique coronal reformatted image in the arterial phase shows preferential flow with dense contrast from the inferior vena cava (IVC) with hepatic vein blood to the right pulmonary artery (RPA). Note that the unopacified superior vena cava (SVC) blood is directed into the left pulmonary artery (LPA). (B) A small tangle of vessels is formed, connecting with the upper pulmonary artery and the upper pulmonary vein in the LUL lingular segment, suggestive of pulmonary arteriovenous malformation.

Aortopulmonary collaterals (arterial collaterals)

In patients who undergo the palliative Fontan procedure, development of APCs is frequently observed due to arterial hypoxemia. Eventually, APCs result in left-to-right shunts (Fig. 12). APCs usually arise from the descending aorta, subclavian artery branches, and bronchial and intercostal arteries. With the passage of time, APCs result in left-to-right shunts and increased pulmonary blood flow and pressure. APCs have many physio-

logic implications, such as ventricular volume overload and pleural effusion. In addition, APCs can be a source of life-threatening hemoptysis in close association with bronchial tree dilatation, airway erosion, and rupture. MDCT depicts the locations of APCs, and CMR allows estimation of APC blood flow. Grosse-Wortmann et al. [14] reported two methods for calculating APC blood flow. Method A involved summation of the individual pulmonary vein flows. Subsequently, the sum of the

Fig. 18. An 18-year-old female who underwent Kawashima operation and Fontan completion for heterotaxy, IVC interruption, and azygos continuation. (A and B) CT scan with a lung window and a volume rendered image show a prominent LPA and a pulmonary vein, and a communication is noted in the LUL and LLL (arrows), suggestive of PAVM. She underwent connection of the hepatic vein to the Az with a Gore-Tex tube graft to relieve severe cyanosis due to PAVM. RPA: right pulmonary artery, LPA: left pulmonary artery, Az: azygos vein, IVC: inferior vena cava, PAVM: pulmonary arteriovenous malformation.

Fig. 19. A 33-year-old female patient who underwent atriopulmonary Fontan operation for an RV-type single ventricle. (A) Volume rendered image shows prominent venous collaterals from the inferior vena cava (IVC) adjacent to the hepatic vein to the left atrium (LA) via the left pulmonary vein (arrow). (B) Note a contrast jet to the LA (arrow). LPA: left pulmonary artery, RV: right ventricle.

Fig. 20. A 20-year-old male patient who underwent lateral tunnel Fontan operation for a double-outlet right ventricle. Arterial (A), portal (B), and delayed phases (C) in liver dynamic CT scan show a hyperdense nodule in the arterial phase, a slightly hyperdense nodule in the portal phase, and an isodense/slightly hyperdense nodule in the delayed phase (arrows), which is suggestive of a focal nodular hyperplasia-like nodule. Inhomogeneous reticular enhancement of the liver is seen in the portal venous phase (arrowheads).

right and left pulmonary arterial flows was subtracted from the sum of the individual pulmonary vein flows. With method B, APC flow was calculated by subtracting the sum of the SVC flow and the descending aorta flow at the diaphragm from the ascending aorta flow.

Cardiac cirrhosis and hepatic nodules

Chronically elevated systemic venous pressure associated with Fontan circulation causes increased retrograde pressure in the hepatic sinusoids. This may lead to passive hepatic congestion, hepatic cirrhosis, and portal hypertension, which can be complicated by dysplastic nodules and hepatocellular carcinoma. Because children are often asymptomatic, congestive hepatopathy is usually first detected on MDCT and CMR imaging. Congestive hepatopathy manifests in inhomogeneous reticular enhancement patterns, most prominent in the periphery of the liver, which are best observed in the portal venous phase. Chronic passive hepatic venous congestion can also lead to the formation of venovenous collaterals.

A chronic increase in hepatic venous pressure results in arterialization of hepatic flow, which can lead to the development of hypervascular dysplastic nodules. These benign regenerative or focal nodular hyperplasia-like nodules are typically isodense to liver on precontrast images, show avid enhancement in the arterial phase, and are slightly hyperdense/isodense to liver parenchyma in the portal and equilibrium phases of MDCT (Fig. 20). Also, they show intense enhancement in the arterial phase and are slightly hyperintense/isointense to liver parenchyma in the portal and equilibrium phases of MRI [23].

Protein-losing enteropathy

Elevated lymphatic pressure may result in lymphedema, pulmonary edema, and pleural and pericardial effusion. Ascites and protein-losing enteropathy are additional late but serious abdominal complications of Fontan circulation.

Protein-losing enteropathy is a rare manifestation of failing Fontan circulation. Although its etiology is not clearly established, enteric protein loss may be due to systemic venous hypertension that is transmitted to the hepatic circulation. Even though protein-losing enteropathy does not manifest specific CT and MRI imaging findings, it should be suspected in patients with abdominal pain, diarrhea, recurrent pleural effusion and ascites, hypoproteinemia, hypocalcemia, and coagulopathy [25].

CONCLUSION

In patients who undergo the Fontan procedure, postoperative imaging follow-up with CMR and MDCT is essential for early detection of cardiac and extracardiac complications. Special modifications to the imaging protocols for these patients are required to optimally evaluate the Fontan pathway. Radiologists should be familiar with the varying types of Fontan pathways, the imaging techniques, and the diverse imaging features of abnormal postsurgical complications, including thromboembolism, stenosis of the conduit, pulmonary artery stenosis, arterial and venous collaterals, PAVM, hepatic congestion, and cardiac cirrhosis.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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