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Seminars in ULTRASOUND CT and MRI

Pearls and Pitfalls in Pediatric Fontan Operation Imaging

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The Fontan operation or the total cavopulmonary connection is a palliative surgery for single ventricle congenital heart disease where the systemic venous return circumvents a pumping chamber and flows directly into the pulmonary circuit. With surgical and medical advances, there has been improvement in life expectancy of these patients, however, it has also resulted in unique complications from the physiology that requires diligent surveillance. A critical component relies on optimal imaging for diagnosis and treatment of these complications. This article describes the normal anatomy of the Fontan circulation, current imaging modalities and techniques, and frequently encountered complications seen when imaging the patients who have undergone Fontan palliation.
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Introduction

A number of congenital cardiac anomalies interfere with normal cardiac physiology and result in a single functional ventricle. These anomalies include hypoplastic left heart syndrome, tricuspid atresia, pulmonary atresia with intact ventricular septum, and a double-inlet ventricle. In normal cardiac physiology, there are subpulmonary and subaortic pumping chambers to propel blood into the pulmonary and systemic circulations respectively, however, in a single functional ventricle, there is a single pumping chamber for both the systemic and pulmonary circulations.

In 1971, Fontan and Baudet described a surgical procedure which diverted systemic venous flow into the pulmonary circulation by allowing passive flow into the lungs, circumventing the need for a pumping ventricle, and as a result, established the systemic and pulmonary circulations in series powered by a single ventricle.[1] This palliative strategy, referred as the Fontan procedure, placed the single ventricle in the unique position of sustaining the circulations without the benefit of a second ventricle and as such, placed it at risk for possible failure. Complications include not only heart failure but protein loosing enteropathy, plastic bronchitis, poor exercise performance and liver fibrosis to name just a few. Recent advances in cardiac surgery for congenital heart anomalies and improvement in postsurgical care have significantly increased the life expectancy of the patients following Fontan palliation.[2] As a result, long-term complications of Fontan circulation are more frequently encountered. Diagnosis and optimal imaging work-up of Fontan physiology and its complications are essential for radiologists and physicians to guide appropriate treatment options. In this article, we describe the anatomy of the Fontan operation, current imaging modalities and techniques, and frequently encountered complications seen when imaging the patients who have undergone Fontan palliation.

Fontan Palliation

Completed single ventricle palliation ultimately culminates in a total cavopulmonary connection or Fontan palliation.[1] There is a 2-3 staged approach to this palliation. Stage 1 involves establishing a stable source of systemic and pulmonary blood flow in parallel. Pulmonary blood flow is often established via a surgical shunt such as a modified Blalock-Taussig shunt from the right

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subclavian artery or innominate artery to the right pulmonary artery or a Sano shunt from the right ventricle to the pulmonary artery. To establish unobstructed systemic circulation, a Norwood/DKS (Damus-Kaye-Stansel) procedure is often performed during the same operation to establish an adequate systemic outflow and augment the aortic arch as needed. This stage can possibly be skipped if the single ventricle has a balanced source of pulmonary blood flow such as tricuspid atresia with normally related great arteries where the outflow of blood to the pulmonary arteries is enough to not be too cyanotic but not too much as to induce heart failure. Stage 2 involves a superior pulmonary venous connection where the initial surgical shunt is ligated and the superior vena cava flow is directed to the pulmonary arteries by either a bidirectional Glenn (end to side Superior Vena Cava/Right Pulmonary Artery (SVC/RPA) anastomosis) or hemi-Fontan (side to side SVC/RPA anastomosis). This is followed by stage 3, the total cavopulmonary anastomosis or Fontan,[3] where inferior vena caval blood is additionally directed into the pulmonary arteries.

There have been several modifications of the Fontan operation since the originally described intervention. The modified classic Fontan involves a direct anastomosis of the right atrial appendage to the pulmonary artery (atriopulmonary Fontan), but this has been supplanted by creation of a lateral wall tunnel or extra-cardiac Fontan which directs the inferior vena cava flow to the pulmonary artery. A lateral wall tunnel uses both native atrial tissue and a prosthetic patch to create a baffle that courses along the lateral wall of the right atrium whereas in the extra-cardiac Fontan, a conduit is placed joining the inferior vena cava to the pulmonary artery external to the right atrium[4] (Fig. 1). Additionally, this surgery also incorporates (in general) the use of a fenestration in the Fontan, allowing a controlled right-to-left shunt between the systemic venous pathway and the pulmonary venous pathway which increases ventricular preload and decreases venous pressure,[5] maintaining cardiac output at the expense of cyanosis.[6]

Imaging of the Fontan Circulation

Cardiac MRI (CMR)

Advances in imaging in the past few decades have bolstered the position of cardiac MR (CMR) in pediatric and adult patients with Fontan circulation. CMR is a complimentary imaging modality to echocardiography whose inherent limitations become increasing apparent with advancing age of the patient[7]. Additionally, estimation of ventricular function in the single ventricle patient can be difficult and assessment of volumes are more reproducible on CMR than echocardiography.[8,9] CMR can also more accurately measure hemodynamics and blood flow along with tissue characterization such as myocardial perfusion and scarring. The advantages of CMR include lack of invasiveness and ionizing radiation, high spatial and temporal resolution (temporal resolution lower than on echocardiography), and excellent tissue characterization. Additionally, volumetry for functional assessment and the ability to quantify flow within vessels and conduits makes it the gold standard for shunt estimation and aortopulmonary (AP) collateral flow.[10-12] It however has some shortcomings, including lengthy examination techniques often necessitating sedation, particularly in the pediatric population, and in claustrophobic patients and is therefore difficult to use in the acute setting.[11-13] Although MR incompatible devices remain a contraindication, improvement in pacemaker technology and design have resulted in devices safe for CMR.[14-16]

In brief, our CMR imaging protocol involves electrocardiogram (ECG)-gated static and cine steady state-free precession for the initial evaluation of cardiac anatomy and function, phase contrast velocity mapping to map vascular flows including quantification of systemic to pulmonary collateral flow. This is followed by postgadolinium-enhanced MR angiography for further evaluation of anatomy, myocardial delayed enhancement and T1 mapping to assess for myocardial fibrosis.[17] Additionally, a noncontrast T2 MR lymphangiogram is performed for evaluation of the thoracic duct and lymphangiectasia.[18] Advanced techniques such as four-dimensional (4D) flow phase-contrast acquisitions and 4D multiphase steady-state imaging with contrast have also been used to acquire functional and anatomic data in a single acquisition.[19,20]

Table 1 lists the complete protocol for Fontan evaluation at our institution.

Cardiac CT Angiography

Although CMR is most commonly used for routine assessment of patient's following Fontan palliation, ECG-gated

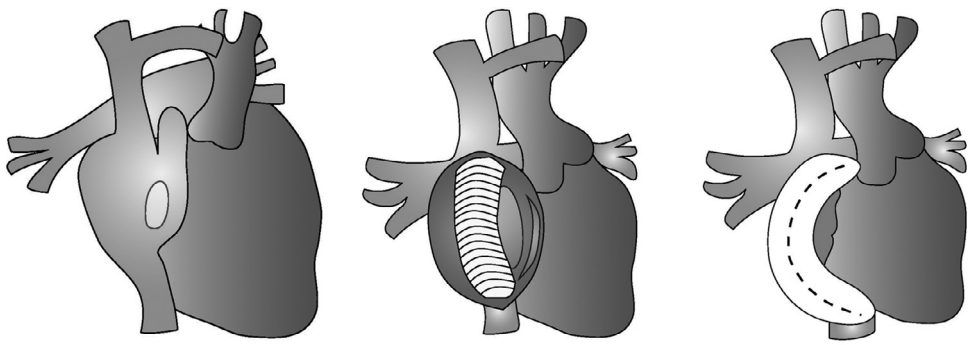

Figure 1 Schematic drawings of Fontan palliation. (a) Atriopulmonary Fontan, (b) lateral tunnel Fontan, and (c) extracardiac Fontan.

Table Cardiac MR Protocol for the Evaluation of Fontan Palliation

Imaging Sequence		Why Perform?
Cine SSFP in	***Short axis***	Quantification of ventricular volumetry and qualitative assessment of wall motion and valvular insufficiency
	Two chamber	
	Four chamber	
	Ventricular outflow tract	
Phase contrast velocity mapping	***Ascending Aorta***	Quantification of AP collateral flow, AV valve insufficiency, fenestration flow
	Right and Left PA	
	Cavae	
	Pulmonary veins	
	Superior Fontan pathway	
	AV valve	
Noncontrast T2 lymphangiogram		Evaluation of thoracic duct and lymphangiectasia
Gadolinium enhanced MR angiography		Anatomic assessment of vascular structures
Myocardial delayed enhancement and T1 mapping		Assessment of myocardial fibrosis

SSFP, steady state free procession; PA, pulmonary artery; AP, aortopulmonary; AV, atrioventricular.

cardiac CT angiography (CCTA) is used when CMR is not feasible. Such circumstances include postsurgical patients with non-MR compatible devices and for assessment of stents. The benefits of CCTA include short scan duration leading to less sedation and high spatial resolution. Additionally, retrospectively gated CCTA can also be used for volumetric and functional assessment when needed with a lower temporal resolution than CMR with an additional cost of increased ionizing radiation.[21] CCTA can detect complications such as thrombosis, stenosis, pulmonary thromboembolism, arteriovenous fistula as well as map arterial and venous collaterals.[22-24] Its drawbacks are exposure to ionizing radiation, poor tissue contrast, poor valvular assessment, lower temporal resolution than CMR and lack of flow data.

ECG-gated synchronization modes that are commonly utilized with CCTA include a high-pitch helical scan mode, prospective gating, and retrospective gating. With high-pitch, images are acquired rapidly during a single heartbeat often eliminating the need for sedation in infants and uncooperative children.[25,26] With prospective ECG gating, imaging is performed during a single point in the cardiac cycle over multiple heartbeats.[25] Finally, retrospective gating imaging is performed throughout the cardiac cycle enabling evaluation of both end-systole and end-diastole allowing volumetric and functional analysis.[27]

Homogeneous opacification of vasculature of interest is paramount when imaging patients post-Fontan palliation. Either bolus tracking or the use of a timing bolus can be performed. With bolus tracking, the CCTA imaging is initiated at a threshold of Hounsfield units in a vessel of interest whereas a timing bolus uses a small amount of contrast to optimally time the CCTA acquisition.[28,29] Regardless, the heterogeneous distribution of blood from the superior vena cava and inferior vena cava can make optimal imaging challenging due to mixing of opacified and unopacified blood, particularly in the pulmonary arteries. Dual injections of the upper and lower extremity and delayed acquisitions may overcome these difficulties. With a dual injection technique, 60% of the contrast is injected in the lower extremity at a rate of 3-4 mL/sec and the remaining contrast is injected in the upper extremity at 2-2.5 mL/sec. Given the rates, the use of two power injectors should be used but in the absence of a second power injector a fast hand injection in the upper extremity can be performed.[3]

Imaging of Complications of Fontan Palliation

Ventricular Dysfunction

It is known that patients after Fontan palliation have low cardiac output and have a high incidence of heart failure.[30,31] Potential etiologies of this include the use of a systemic right ventricle, ventriculotomy during palliation, and chronic hypoxemia.[32,33]

Both retrospective CCTA and CMR can evaluate for global ventricular dysfunction and also wall motion abnormalities. CMR is the preferred imaging modality for this evaluation and has the advantage of assessment of fibrosis via myocardial delayed enhancement (Fig. 2) along with higher temporal resolution. End-diastolic volume measured by CMR is a predictor of death and transplant following Fontan palliation.[34] The presence of fibrosis is associated with nonsustained ventricular tachycardia, decreased function, and regional wall motion abnormalities (Fig. 2).[35]

Atrioventricular Valve (AV) Regurgitation

Atrioventricular (AV) valve dysfunction after Fontan palliation has also been shown to be a predictor of poor outcome.[36] Multiple factors cause AV valve dysfunction including prior volume overload, systemic to pulmonary shunting, annular dilatation, abnormal chorda and papillary muscles, and decreased ventricular function.[5] Although echocardiography can evaluate AV valve dysfunction, the use

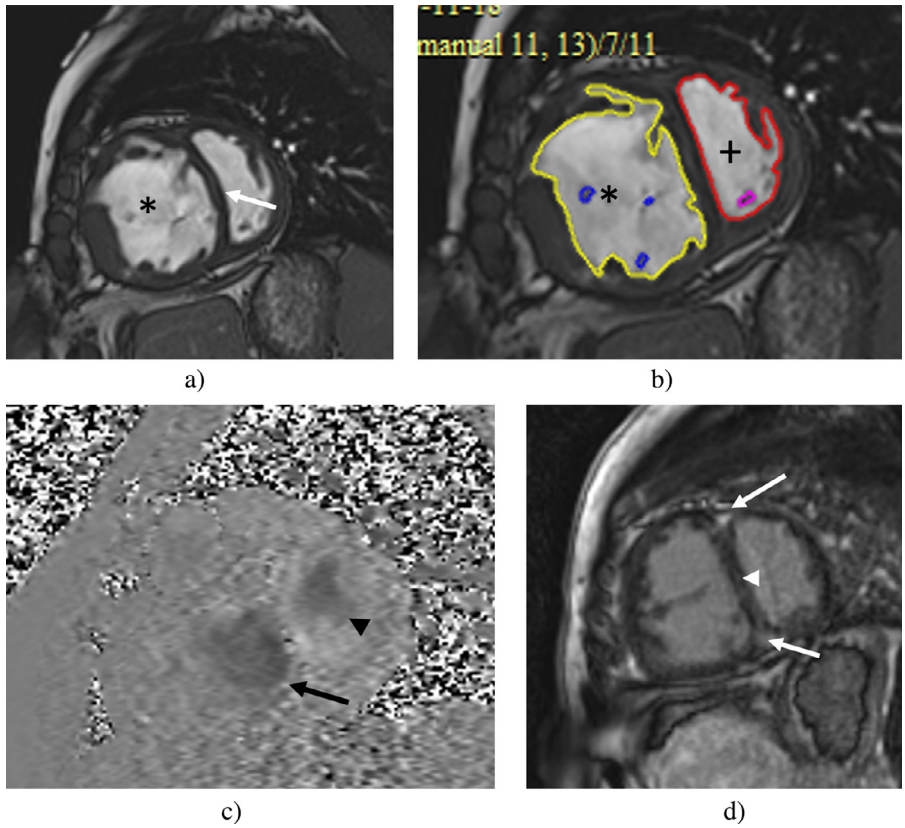

Figure 2 Sixteen-year-old boy with history of hypoplastic left heart syndrome variant, status postextracardiac fenestrated Fontan with decreased ventricular function. (a) Short axis view from a cine image of the heart demonstrates and enlarged right ventricle (*) with bowing of the ventricular septum to the left (arrow). (b) Postprocessed short axis image of the right (*) and the left ventricle (+) revealed decreased biventricular systolic function with a right ventricular ejection fraction measuring 36% and a left ventricular ejection fraction measuring 49%. (c) Two chamber right ventricular view demonstrates a dephasing jet consistent with tricuspid valve insufficiency. (d) Phase contrast images demonstrates reversal of flow within the tricuspid (arrow) and mitral valve (arrowhead) consistent with insufficiency. Calculated tricuspid regurgitant fraction measured greater than 60% and calculated mitral regurgitant fraction measured greater than 30%. (d) Myocardial delayed enhancement demonstrates patchy mid-myocardial fibrosis within the septum along with fibrosis at the superior and inferior hinge points (arrows).

of phase contrast CMR and cine CMR for volumes can quantitatively calculate the regurgitant fraction and flow velocity across the AV valve. There are 2 methods to measure the regurgitant fraction (Fig. 2). One is to compare the ventricular stroke volume to the forward flow within the aorta and the other is to compare the forward flow through the AV valve to the net aortic flow by phase contrast techniques.[37]

Fontan Baffle Leaks/Stenosis and Fenestration

Leak and stenosis of the Fontan pathway can be seen at anastomotic sites and be anatomically evaluated by either CCTA or CMR. In stenosis, phase contrast CMR can estimate the pressure gradient. Baffle leaks can also be quantified by phase contrast CMR.[38]

A fenestration is a modification to the Fontan with connection of the Fontan pathway to the pulmonary venous atrium. Baffle fenestration has been found to improved short-term outcomes such as decreasing pleural drainage, length of hospital stay, and need for additional postoperative procedures.[39,40] When a fenestration is closed following Fontan palliation, central venous pressure and oxygen saturation increases and cardiac index decreases.[41] The simplest way to calculate the flow through the fenestration by phase contrast CMR is by comparing the inferior vena cava flow to the flow within the superior Fontan baffle; another method is the difference between pulmonary venous flow and aortic outflow. Finally, some groups have had success using through plane phase contrast mapping directly across the fenestration.

Systemic to Pulmonary Collaterals

Communication between the systemic circulation and the pulmonary circulation via systemic to pulmonary collaterals is a common finding in Fontan patients. AP collaterals originate most commonly from the descending aorta, subclavian artery, bronchial and intercostal arteries. Long-standing AP collaterals causes left-to-right shunts, increased pulmonary blood flow and pulmonary pressure, which in turn may lead to ventricular volume overload and pleural effusions.

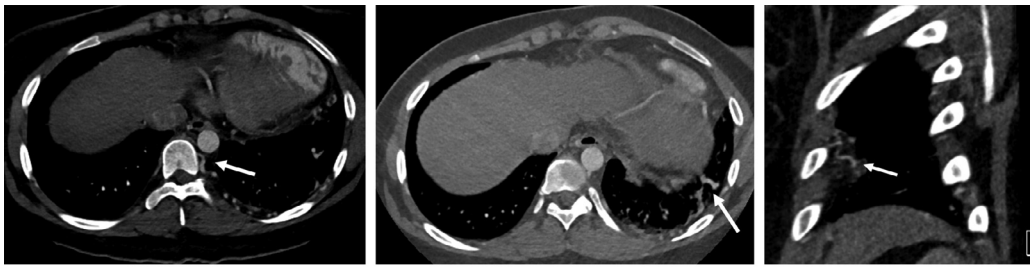

Figure 3 Eighteen-year-old female with a single ventricle in the form of transposition of the great arteries, pulmonary stenosis post fenestrated Fontan. (a) Axial image from a contrast enhanced cardiac CTA demonstrates a prominent vessel posteriorly within the left paravertebral region that originated from the descending aorta and terminated in the left lung consistent with an aortopulmonary collateral (arrow). (b) Axial image from the same cardiac CTA demonstrates prominent vessels within the periphery of the left lower lobe (arrow) consistent with an arteriovenous malformation. (c) Coronal image further demonstrates the prominent vessel in the periphery of the left lower lobe (arrow).

Collaterals can steal blood from other organs, such as the brain.[42] Venovenous collaterals are a result of elevated central venous pressure usually resulting in connections between the superior or inferior vena cava or innominate veins and the pulmonary veins.[38] Although CCTA can depict the location of both AP and venovenous collaterals (Fig. 3), CMR can quantify the collateral burden using phase contrast velocity mapping which has been correlated to postoperative Fontan outcomes such as chest drain volume and duration of hospital stay.[43–45]

Using CMR, calculation of systemic to pulmonary collateral flow is done by subtracting the sum of caval flow from the aortic flow or by subtracting the pulmonary arterial flow from pulmonary venous flow.[46]

Pulmonary Arteriovenous Malformation

In pulmonary arteriovenous malformations (AVMs), systemic venous blood reaches to the pulmonary venous system due to abnormal vascular connections resulting in right to left shunting.[47] Although pulmonary AVM are more commonly described following a bidirectional Glenn, pulmonary AVMs are still seen following Fontan palliation. This occurs due to the asymmetrical distribution of hepatic venous blood to the pulmonary circulation and poor-filling of the pulmonary arteries from the absence of a pumping ventricle.[4] The absence of hepatic vein flow to the lungs, that is, the hepatic factor, has been implicated as a cause of pulmonary AVMs. Data that support this is that in the bidirectional Glenn, where there is only blood from the superior vena cava flowing to the lungs, pulmonary AVMs are more common. Additionally, pulmonary AVMs are more common in patients with single ventricle and interrupted inferior vena cava who undergo the Kawashima procedure. This procedure routes systemic venous return to the pulmonary arteries via the azygous system whereas the hepatic veins empty directly into the right atrium.[48] Finally, pulmonary AVMs have been shown to occur in two-ventricle physiology with an isolated anomalous pulmonary venous connection from the hepatic veins to the left atrium and are seen in patients with advanced hepatic failure.[4,47] When redirection of the hepatic flow to the lungs occurs, such as following the Fontan procedure, pulmonary AVMs have been shown to regress.[47,49]

Although pulmonary AVMs can be see with an MR angiogram, imaging is most commonly performed with CCTA where enlarged pulmonary vessels are seen extending to the periphery of the lung with a tangle of vessels manifesting as the pulmonary AVM[4] (Fig. 3).

Pulmonary Thromboembolism

The causes of pulmonary thromboembolism complications are varied in Fontan patients. Low blood flow, venous stasis, presence of thrombogenic material, along with atrial dilation and stasis are the common precipitating factors to thrombus in the Fontan circulation. Plasma levels of anticoagulant factors like antithrombin, protein C, protein S are low as a result of protein-losing enteropathy (PLE) and this places the patients at risk of thromboembolism.[50]

Thrombus within the Fontan pathway and atrium can lead to pulmonary embolism, which is most commonly diagnosed using CCTA (Fig. 4). Given that flow from the superior vena cava and inferior vena cava into the pulmonary arteries is not uniform, mixing artifact will limit the diagnostic accuracy for this evaluation.[51] Two methods, one with a dual injection and the other with a signal injection, have been described for the assessment of pulmonary embolism in Fontan patients using CCTA. With a dual injection, there is simultaneous injection of CT contrast though an upper and lower extremity intravenous line to achieve homogenous opacification of the pulmonary arteries. With a single injection technique CT, contrast is injected via a single peripheral intravenous line with a set delay ranging from 60 to 100 seconds depending on the institution.[3]

Plastic Bronchitis

Plastic bronchitis is characterized by formation large proteinaceous casts within the airway. Although it can occur with other disorders, plastic bronchitis is most commonly seen following Fontan palliation.[52] This disorder is caused by hypersecretion of airway mucus along with abnormalities of

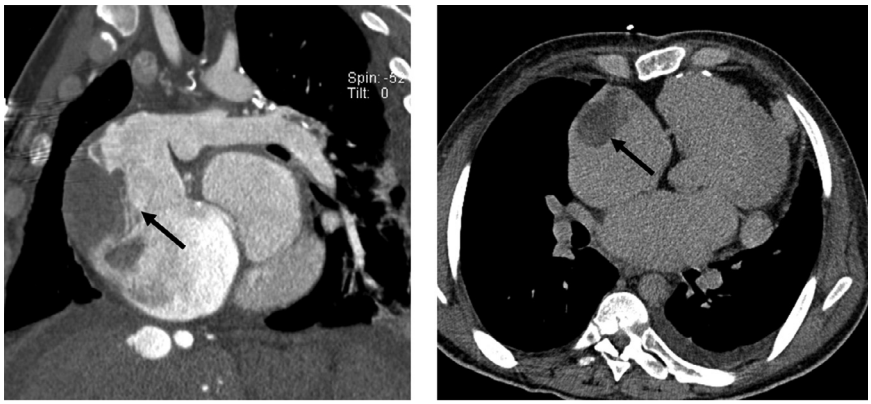

Figure 4 Twenty-year old with history of tricuspid atresia and transposition of the great arteries status postatriopulmonary Fontan. (a) Coronal image from a contrast enhanced CTA demonstrates a large filling defect within the atrium (arrow) consistent with an atrial thrombus. (b) A more delayed axial image from the same examination again shows the atrial thrombus (arrow).

pulmonary lymphatic drainage that are related to the chronically elevated central venous pressure.[53,54]

Diagnosing plastic bronchitis with CCTA or CMR can be challenging. CT of the chest may demonstrate bronchial casts causing partial or complete obstruction of airway in the absence of bronchiectasis.[55] Although heavily T2 weighted imaging can demonstrate dilatation of the thoracic duct and pulmonary lymphangiectasia, dynamic contrast enhanced MR lymphangiography is the preferred technique to further characterize this disorder demonstrating abnormal lymphatic flow to the lung parenchyma (Fig. 5).[53,54]

Fontan Associated Liver Disease

Fontan associated liver disease (FALD) is a group of diagnoses resulting from congestive hepatopathy including liver cirrhosis, focal nodular hyperplasia, and hepatocellular carcinoma.[56] With Fontan palliation, there is chronically elevated hepatic vein pressures, which leads to passive congestion, dilatation of hepatic sinusoids and arterialization of hepatic flow.[4,57] Serum markers are not helpful in diagnosing early stages of congestive hepatopathy since they remain normal until end stage disease and do not correlate with fibrosis.[57,58]

Although conventional CT and ultrasound is insensitive in evaluating fibrosis, congestive hepatopathy manifests as hepatomegaly with nodularity along with dilatation of the hepatic veins and inferior vena cava.[57] Congestive hepatopathy will also have a similar appearance as chronic venous obstruction, such as Budd-Chiari syndrome where there is typically reticular enhancement in the periphery of the liver.[4] Later regenerative nodules may form which must be distinguished from hepatocellular carcinoma most often on a contrast enhanced MRI.[4]

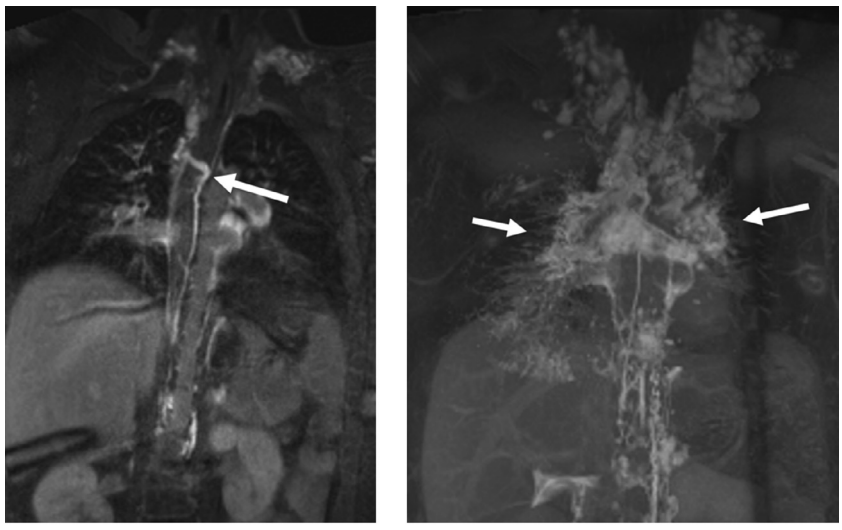

Figure 5 Fourteen-year-old female with history of double inlet left ventricle and interrupted aortic arch, extracardiac fenestrated Fontan who presents with plastic bronchitis. (a) Respiratory navigated gradient echo MR lymphangiogram reveals a dilated and tortuous thoracic duct with a prominent channel (arrow) crossing the midline. (b) Maximum intensity projection of an MR lymphangiogram demonstrates bilateral pulmonary lymphatic perfusion (arrows).

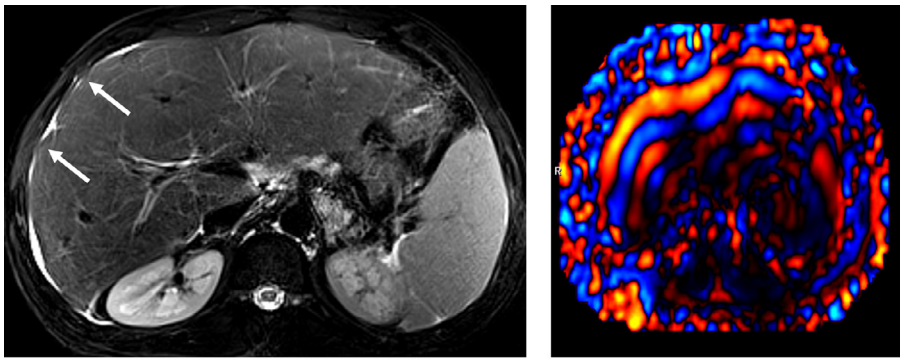

Figure 6 Twenty-one-year old with hypoplastic left heart syndrome status post Fontan with increased liver stiffness. (a) Axial T2-weighted image demonstrates mild surface nodularity (arrows). Spleen is also enlarged in size. (b) MR elastogram revealed increased liver stiffness measuring up to 5 kPa (normal <2.9 kPa).

There is growing evidence that elastography, by either ultrasound or MR, is a valuable tool in determining liver stiffness, which is representative of liver fibrosis and congestion that is seen with FALD.[5] With ultrasound elastography, shear wave elastography is the most commonly used technique where the velocity of shear waves, produced through mechanical vibration of acoustic radiation force impulses, through a tissue correlates with fibrosis. With MR elastography, mechanical vibration produces waves through the liver which are converted into liver tissue stiffness maps (Fig. 6).[59]

PLE

PLE is a result of excessive protein loss into the intestinal lumen leading to hypoalbuminemia. The incidence in children with Fontan circulation is about 13%.[60] A number of theories have been proposed as to the etiology of PLE, but it is still unclear. In general, it is thought that lack of or failure of pumping action between the systemic venous circulation and pulmonary arteries that leads to chronically elevated systemic venous pressures has a 2-fold effect on the lymphatic system, by both, increasing lymphatic pressure (afterload) and increasing formation of lymphatic fluid from the peripheral organs, particularly the liver. Cumulatively, this stress can cause lymphatic congestion, abnormal flow patters or lymphangiogenesis which can lead to lymphatic disease such as PLE, where lymphatic leakage occurs in the hepato-duodenal lymphatic system.[61,62] T2-weighted unenhanced lymphangiography is fast noninvasive method to visualize lymphatic system demonstrating thoracic duct dilation and tortuosity.[63] The use of intrahepatic MR lymphangiography has demonstrated abnormal lymphatic perfusion to the duodenum with intraluminal leak of contrast in this disorder (Fig. 7).[64] Vascular imaging has also been used to guide

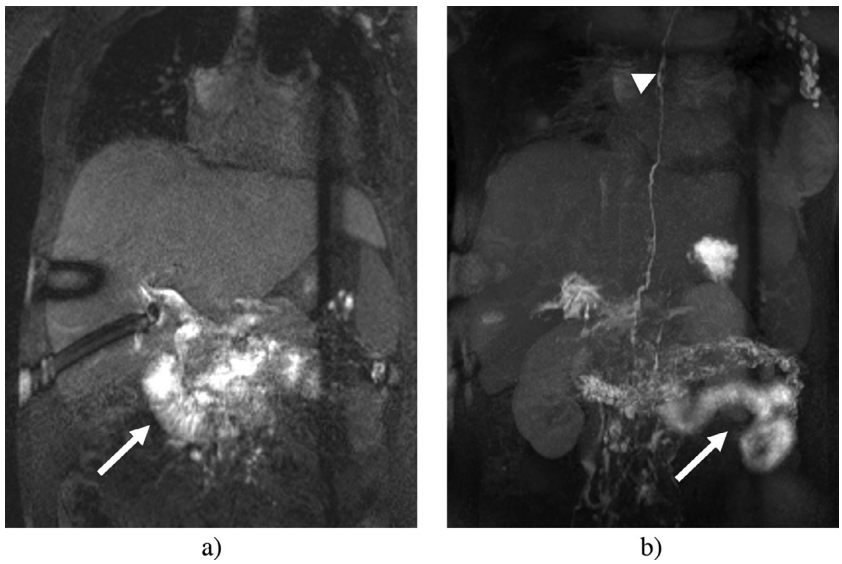

Figure 7 Twenty-four-year-old female with history of double inlet left ventricle post extracardiac fenestrated Fontan who presents with protein losing enteropathy. (a) Respiratory navigated gradient echo image following an intrahepatic MR lymphangiogram demonstrates flow of lymphatic contrast into the duodenum lumen (arrow). (b) Maximum intensity projection from after a subsequent bilateral inguinal node MR lymphangiogram demonstrates contrast continuing to progress through the bowel (arrow) with the thoracic duct extending to the left venous angle (arrowhead).

treatment including percutaneous decompression of the thoracic duct by venous connection to a lower pressure chamber such as the left atrial appendage.[65]

Conclusion

Imaging following the Fontan operation can be complex due to the physiology of a single pumping chamber. Monitoring and complications of this palliation can be performed with CMR and CCTA. CMR particularly is advantageous given its ability to perform both functional and anatomical assessment along with tissue characterization without ionizing radiation, but CCTA has a complementary role due to its speed decreasing the need for sedation in the pediatric population.

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REFERENCES

1. Fontan F, Baudet E: Surgical repair of tricuspid atresia. Thorax 26:240-248, 1971
2. Gilboa SM, Devine OJ, Kucik JE, et al: Congenital heart defects in the United States: Estimating the magnitude of the affected population in 2010. Circulation 134:101-109, 2016
3. Ghadimi Mahani M, Agarwal PP, Rigsby CK, et al: CT for assessment of thrombosis and pulmonary embolism in multiple stages of single-ventricle palliation: Challenges and suggested protocols. Radiographics 36:1273-1284, 2016
4. Khanna G, Bhalla S, Krishnamurthy R, et al: Extracardiac complications of the Fontan circuit. Pediatr Radiol 42:233-241, 2012
5. Rychik J, Atz AM, Celermajer DS, et al: Evaluation and management of the child and adult with Fontan Circulation: A scientific statement from the American Heart Association. Circulation 140:234-284, 2019
6. Fredenburg TB, Johnson TR, Cohen MD: The Fontan procedure: Anatomy, complications, and manifestations of failure. Radiographics 31:453-463, 2011
7. Prakash A, Powell AJ, Geva T: Multimodality noninvasive imaging for assessment of congenital heart disease. Circulation 3:112-125, 2010
8. Han BK, Rigsby CK, Leipsic J, et al: Computed tomography imaging in patients with congenital heart disease, part 2: Technical recommendations. An expert consensus document of the Society of Cardiovascular Computed Tomography (SCCT). Endorsed by the Society of Pediatric Radiology (SPR) and th. J Cardiovasc Comput Tomogr 9:493-513, 2015
9. Margossian R, Schwartz ML, Prakash A, et al: Comparison of echocardiographic and cardiac magnetic resonance imaging measurements of functional single ventricular volumes, mass, and ejection fraction (From the Pediatric Heart Network Multicenter Fontan Cross-Sectional Study). Am J Cardiol 104:419-428, 2009
10. Lewis G, Thorne S, Clift P, et al: Cross-sectional imaging of the Fontan circuit in adult congenital heart disease. Clin Radiol 70:667-675, 2015
11. Grewal J, Al Hussein M, Feldstein J, et al: Evaluation of Silent thrombus after the Fontan operation. Congenit Heart Dis 8:40-47, 2013
12. Nordmeyer J, Gaudin R, Tann OR, et al: MRI may be sufficient for non-invasive assessment of great vessel stents: An in vitro comparison of MRI, CT, and conventional angiography. Am J Roentgenol 195:865-871, 2010
13. Fratz S, Chung T, Greil GF, et al: Guidelines and protocols for cardiovascular magnetic resonance in children and adults with congenital heart disease: SCMR expert consensus group on congenital heart disease. J Cardiovasc Magn Reson 15:1-26, 2013
14. Martin ET, Coman JA, Shellock FG, et al: Magnetic resonance imaging and cardiac pacemaker safety at 1.5-Tesla. J Am Coll Cardiol 43:1315-1324, 2004
15. Naehle CP, Kreuz J, Strach K, et al: Safety, feasibility, and diagnostic value of cardiac magnetic resonance imaging in patients with cardiac pacemakers and implantable cardioverters/ defibrillators at 1.5 T. Am Heart J 161:1096-1105, 2011
16. Wollmann CG, Thudt K, Kaiser B, et al: Safe performance of magnetic resonance of the heart in patients with magnetic resonance conditional pacemaker systems: The safety issue of the ESTIMATE study. J Cardiovasc Magn Reson 16:1-8, 2014
17. Kutty S, Rathod RH, Danford DA, et al: Role of imaging in the evaluation of single ventricle with the Fontan palliation. Heart 102:174-183, 2016
18. Biko DM, DeWitt AG, Pinto EM, et al: MRI evaluation of lymphatic abnormalities in the neck and thorax after Fontan surgery: Relationship with outcome. Radiology 291:774-780, 2019
19. Vasanawala SS, Hanneman K, Alley MT, et al: Congenital heart disease assessment with 4D flow MRI. J Magn Reson Imaging 42:870-886, 2015
20. Nguyen KL, Han F, Zhou Z, et al: 4D MUSIC CMR: value-based imaging of neonates and infants with congenital heart disease. J Cardiovasc Magn Reson 19:1-15, 2017
21. Lin E, Alessio A: What are the basic concepts of temporal, contrast, and spatial resolution in cardiac CT? J Cardiovasc Comput Tomogr 3:403-408, 2009
22. Hauser JA, Taylor AM, Pandya B: How to image the adult patient with Fontan circulation. Circ Cardiovasc Imaging 10:1-7, 2017
23. Rigsby CK, Mckenney SE, Hill KD, et al: Radiation Dose Management for Pediatric Cardiac Computed Tomography: A Report from the Image Gently "Have-A-Heart" Campaign Pediatr Radiol 48:5-20, 2018
24. Han BK, Rigsby CK, Hlavacek A, et al: Computed tomography imaging in patients with congenital heart disease part I: Rationale and utility. An expert consensus document of the Society of Cardiovascular Computed Tomography (SCCT). Endorsed by the Society of Pediatric Radiology (SPR) and the Nor. J Cardiovasc Comput Tomogr 9:475-492, 2015.. Available from: http://dx.doi.org/10.1016/j.jcct.2015.07.004
25. Han BK, Rigsby CK, Hlavacek A, et al: Computed tomography imaging in patients with congenital heart disease part I: Rationale and utility. An Expert Consensus Document of the Society of Cardiovascular Computed Tomography (SCCT). Endorsed by the Society of Pediatric Radiology (SPR) and the Nor. J Cardiovasc Comput Tomogr. 9:475-492, 2015
26. Lell MM, May M, Deak P, et al: High-pitch spiral computed tomography: Effect on image quality and radiation dose in pediatric chest computed tomography. Invest Radiol 46:116-123, 2011
27. Qin J, yun Liu L, chun Meng X, et al: Prospective versus retrospective ECG gating for 320-detector CT of the coronary arteries: Comparison of image quality and patient radiation dose. Clin Imaging 35:193-197, 2011
28. Scholtz JE, Ghoshhajra B: Advances in cardiac CT contrast injection and acquisition protocols. Cardiovasc Diagn Ther 7:439-451, 2017
29. Bae KT: Optimization of contrast enhancement in thoracic MDCT. Radiol Clin North Am 48:9-29, 2010
30. Fogel MA, Weinberg PM, Chin AJ, et al: Late ventricular geometry and performance changes of functional single ventricle throughout staged Fontan reconstruction assessed by magnetic resonance imaging. J Am Coll Cardiol 28:212-221, 1996.. Available from: http://dx.doi.org/10.1016/0735-1097(96)00111-8
31. Khairy P, Fernandes SM, Mayer JE, et al: Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation 117:85-92, 2008
32. Goldberg DJ, Shaddy RE, Ravishankar C, et al: The failing Fontan: Etiology, diagnosis and management. Expert Rev Cardiovasc Ther 9:785-793, 2011
33. Sundareswaran KS, Kanter KR, Kitajima HD, et al: Impaired power output and cardiac index with hypoplastic left heart syndrome: A magnetic resonance imaging study. Ann Thorac Surg 82:1267-1277, 2006
34. Rathod RH, Prakash A, Kim YY, et al: Cardiac magnetic resonance parameters predict transplantation-free survival in patients with Fontan circulation. Circ Cardiovasc Imaging 7:502-509, 2014
35. Rathod RH, Prakash A, Powell AJ, et al: Myocardial fibrosis identified by cardiac magnetic resonance late gadolinium enhancement is associated with adverse ventricular mechanics and ventricular tachycardia late after

Fontan Operation. J Am Coll Cardiol 55:1721-1728, 2010.. Available from: http://dx.doi.org/10.1016/j.jacc.2009.12.036
36. Mavroudis C, Stewart RD, Backer CL, et al: Atrioventricular valve procedures with repeat Fontan operations: Influence of valve pathology, ventricular function, and arrhythmias on outcome. Ann Thorac Surg 80:29-36, 2005
37. Uretsky S, Argulian E, Narula J, et al: Use of cardiac magnetic resonance imaging in assessing mitral regurgitation: Current evidence. J Am Coll Cardiol 71:547-563, 2018
38. Navarro-Aguilar V, Flors L, Calvillo P, et al: Fontan procedure: Imaging of normal post-surgical anatomy and the spectrum of cardiac and extracardiac complications. Clin Radiol 70:295-303, 2015.. Available from: http://dx.doi.org/10.1016/j.crad.2014.10.005
39. Lemler MS, Scott WA, Leonard SR, et al: Fenestration improves clinical outcome of the Fontan procedure: A prospective, randomized study. Circulation 105:207-212, 2002
40. Bridges ND, Mayer JE, Lock JE, et al: Effect of baffle fenestration on outcome of the modified Fontan operation. Circulation 86:1762-1769, 1992
41. Bradley SM: Use of a fenestration should be routine during the Fontan procedure: PRO. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 13:55-59, 2010. Available from: http://dx.doi.org/10.1053/j.pcsu.2010.01.004
42. Fogel MA, Li C, Wilson F, et al: Relationship of cerebral blood flow to aortic-to pulmonary collateral/shunt flow in single ventricles. Heart 101:1325-1331, 2015
43. Glatz AC, Rome JJ, Small AJ, et al: Systemic-to-pulmonary collateral flow, as measured by cardiac magnetic resonance imaging, is associated with acute post-Fontan clinical outcomes. Circ Cardiovasc Imaging 5:218-225, 2012
44. Odenwald T, Quail MA, Giardini A, et al: Systemic to pulmonary collateral blood flow influences early outcomes following the total cavo pulmonary connection. Heart 98:934-940, 2012
45. Grosse-Wortmann L, Drolet C, Dragulescu A, et al: Aortopulmonary collateral flow volume affects early postoperative outcome after Fontan completion: A multimodality study. J Thorac Cardiovasc Surg 144:1329-1336, 2012
46. Whitehead KK, Harris MA, Glatz AC, et al: Status of systemic to pulmonary arterial collateral flow after the Fontan procedure. Am J Cardiol 115:1739-1745, 2015. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=4450112&tool=pmcentrez&rendertype=abstract%5Cnhttp://dx.doi.org/10.1016/j.amjcard.2015.03.022%5Cnhttp://dx.doi.org/10.1016/j.amjcard.2011.11.040%5Cnhttp://dx.doi.org/10.1016/j.ahj.2011.03.021%5Cnht
47. Duncan BW, Desai S: Pulmonary Arteriovenous Malformations after Cavopulmonary Anastomosis. Ann Thorac Surg 76:1759-1766, 2003
48. Nakamura Y, Yagihara T, Kagisaki K, et al: Pulmonary arteriovenous malformations after a Fontan operation in the left isomerism and absent inferior vena cava. Eur J Cardio-thoracic Surg 36:69-76, 2009
49. Sundareswaran KS, de Zélicourt D, Sharma S, et al: Correction of pulmonary arteriovenous malformation using image-based surgical planning. JACC Cardiovasc Imaging 2:1024-1030, 2009.. Available from: http://dx.doi.org/10.1016/j.jcmg.2009.03.019
50. Tomkiewicz-Pajak L, Hoffman P, Trojnarska O, et al: Abnormalities in blood coagulation, fibrinolysis, and platelet activation in adult patients after the Fontan procedure. J Thorac Cardiovasc Surg 147:1284-1290, 2014
51. Goo HW, Yang DH, Park IS, et al: Time-resolved three-dimensional contrast-enhanced magnetic resonance angiography in patients who have undergone a Fontan operation or bidirectional cavopulmonary connection: Initial experience. J Magn Reson Imaging 25:727-736, 2007
52. Avitabile CM, Goldberg DJ, Dodds K, et al: A multifaceted approach to the management of plastic bronchitis after cavopulmonary palliation. Ann Thorac Surg 98:634-640, 2014. Available from: http://dx.doi.org/10.1016/j.athoracsur.2014.04.015
53. Dori Y, Keller MS, Rychik J, et al: Successful treatment of plastic bronchitis by selective lymphatic embolization in a Fontan patient. Pediatrics 134:e590-e595, 2014. Available from: http://www.ncbi.nlm.nih.gov/pubmed/25002668
54. Dori Y, Keller MS, Rome JJ, et al: Percutaneous lymphatic embolization of abnormal pulmonary lymphatic flow as treatment of plastic bronchitis in patients with congenital heart disease. Circulation 133:1160-1170, 2016
55. Goo HW, Jhang WK, Kim YH, et al: CT findings of plastic bronchitis in children after a Fontan operation. Pediatr Radiol 38:989-993, 2008
56. Egbe A, Miranda WR, Connolly HM, et al: Temporal changes in liver stiffness after Fontan operation: Results of serial magnetic resonance elastography. Int J Cardiol 258:299-304, 2018.. Available from: https://doi.org/10.1016/j.ijcard.2018.01.108
57. Lemmer A, VanWagner L, Ganger D: Congestive hepatopathy: Differentiating congestion from fibrosis. Clin Liver Dis 10:139-143, 2017
58. Wu FM, Earing MG, Aboulhosn JA, et al: Predictive value of biomarkers of hepatic fibrosis in adult Fontan patients. J Hear Lung Transplant 36:211-219, 2017. Available from: http://dx.doi.org/10.1016/j.healun.2016.07.011
59. Agbim U, Asrani SK: Non-invasive assessment of liver fibrosis and prognosis: An update on serum and elastography markers. Expert Rev Gastroenterol Hepatol 13:361-374, 2019. Available from: https://doi.org/10.1080/17474124.2019.1579641
60. Driscoll DJ, Offord KP, Danielson GK, et al. Protein-losing enteropathy after the Fontan operation. 1987;
61. Rychik J: Protein-losing enteropathy after Fontan operation. Congenit Heart Dis 2:288-300, 2007
62. Rychik J, Dodds KM, Goldberg D, et al: Protein losing enteropathy after Fontan operation : Glimpses of clarity through the lifting fog. World J Pediatr Congenit Hear Surg 11:92-96, 2020
63. Dori Y, Keller MS, Fogel MA, et al: MRI of lymphatic abnormalities after functional single-ventricle palliation surgery. Am J Roentgenol 203:426-431, 2014
64. Biko DM, Smith CL, Otero HJ, et al: Intrahepatic dynamic contrast MR lymphangiography: Initial experience with a new technique for the assessment of liver lymphatics. Eur Radiol 29:5190-5196, 2019
65. Smith CL, Hoffman TM, Dori Y, et al: Decompression of the thoracic duct: A novel transcatheter approach. Catheter Cardiovasc Interv 2019