INTRODUCTION

Coronary artery fistula, defined as an abnormal communication between the coronary arteries and a cardiac chamber or a thoracic great vessel, may result in hemodynamically significant problems due to vascular shunting in children. Echocardiography, cardiac catheterization, cardiac MRI, and cardiac CT may be used to evaluate coronary artery fistula in children. Recently, CT has played a pivotal role for the accurate diagnosis of coronary artery fistula in children. Surgical or interventional treatment is performed for hemodynamically significant coronary artery fistulas. In this pictorial review, the detailed imaging findings of coronary artery fistula in children are described.

Keywords: Cardiac catheterization; Children; Cardiac CT; Coronary artery anomalies; Echocardiography
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usually asymptomatic, the majority of pediatric patients are symptomatic, as children commonly develop large fistulous connections [15]. The most common clinical presentation of coronary artery fistula in children is a continuous murmur. With increasing patient age, the fistula gradually enlarges with increased shunt flow via the fistula. In contrast, spontaneous closure of the fistula is rare, accounting for 1.0%–2.0% of cases [7]. Aneurysmal changes are frequently seen in the distal segments of coronary artery fistulas (24.2%–87.1%) [4,6,9,12,13]. The amount of shunt flow depends on the size of the fistula, the presence of stenosis, and the pressure level at the drainage site.

**Fig. 1. Coronary cameral fistula between the right coronary artery and the RV.**
A. Initial frontal volume-rendered CT image showing the diffusely dilated right coronary artery in a 5-month-old boy. B, C. Initial inferior volume-rendered CT images with a full slab (B) and with a thinner slab (C) demonstrating the single drainage site of the fistula into the inferomedial basal portion of the RV. Two narrowings (arrows) are noted at the drainage site. D, E. Follow-up frontal (D) and inferior (E) volume-rendered CT images performed 8 years later showing increased dilatation of the right coronary artery and the development of aneurysmal change (asterisk) at the drainage site. The two narrowings (arrows) showed interval increase in diameter due to increased left-to-right shunt through the fistula. LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle

**Fig. 2. Coronary cameral fistula between the LAD artery and the RV in a 5-year-old boy.**
A. Oblique lateral volume-rendered CT image showing the diffusely dilated LAD artery and a D branch in a 5-year-old boy. B, C. Inferior volume-rendered CT images with a full slab (B) and with a thinner slab (C) demonstrating the single drainage site (arrows) of the fistula into the inferomedial basal portion of the RV. D = diagonal, LA = left atrium, LAD = left anterior descending, LV = left ventricle, RA = right atrium, RV = right ventricle
According to the drainage site, the shunt flow may be left to right or left to left, and may even be right to left in cases with hypertensive right ventricles. When the shunt is large enough to decrease perfusion distal to the fistula, a coronary steal phenomenon may occur. Owing to this hemodynamic effect of coronary artery fistulas, various complications, including myocardial ischemia, heart failure, arrhythmia, and infective endocarditis, have been described in adults [7].

**Diagnostic Imaging Modalities**

Transthoracic echocardiography may show a large coronary artery fistula of typically greater than 3 mm in diameter [16]. However, its diagnostic accuracy for identifying coronary artery fistula is low, and was only 19.1% in a study of 72 adult patients with coronary-to-pulmonary artery fistula [13]. Reports on the diagnostic accuracy of transthoracic echocardiography for identifying coronary artery fistula in children are lacking. Although the acoustic window of transthoracic echocardiography is less limited in children compared to adults, its diagnostic accuracy is expected to remain insufficient to be used as a confirmatory imaging test for coronary artery fistula in children. Therefore, additional imaging methods, including catheter coronary angiography, coronary CT angiography, and coronary MR angiography, are required to make accurate diagnosis of coronary artery fistula.

The reported diagnostic accuracy of catheter coronary angiography is in the range of 35.0%–50.0% [17,18]. Due to procedure-related complications, radiation exposure, and the use of iodinated contrast agents, diagnostic catheter coronary angiography should not be primarily used for coronary artery fistulas, especially in young children.

Coronary MR angiography does not involve ionizing radiation, which may be considered a merit in pediatric imaging. However, its diagnostic accuracy in children is relatively low, mainly due to its low spatial resolution [19]. Lengthy examination time, commonly requiring deep sedation or general anesthesia, is another limitation of pediatric coronary MR angiography [20]. Consequently, cardiac MRI has been rarely used for the diagnosis of coronary artery fistula [21].

Coronary CT angiography is currently considered the imaging modality of choice for evaluating coronary artery fistula in adults [4,6-9,12-14]. Coronary artery visibility on electrocardiography-synchronized CT is also significantly improved in children [22-24]; therefore, cardiac CT has been useful for evaluating coronary artery anomalies such as coronary artery fistula in children [5,15,20,25-27]. Three-dimensional CT imaging with a thinner slab over the area of interest is especially useful for the treatment planning of...
coronary artery fistulas. However, coronary artery fistulas with mildly dilated distal coronary artery segments and small drainage sites, particularly seen in young children, may be missed given the significantly low visibility of distal coronary artery segments on cardiac CT [28]. In this case, ascending aortography or selective catheter coronary angiography may be helpful in making the correct diagnosis.

**Coronary Cameral Fistula**

Coronary cameral fistula is the most common type of coronary artery fistula in children, accounting for 75.0%–100.0% of cases [5,15]. Coronary cameral fistula in children most commonly originates from the right coronary artery, followed by the left coronary artery (Figs. 1-5) [5,15]. In this type of fistula, the coronary arterial origin is usually single and rarely multiple [5]. When the fistula occurs in association with pulmonary atresia and an intact ventricular septum, the origin is frequently multiple. The most common draining site of coronary cameral fistula in children is the right ventricle, followed by the left ventricle [15] (Figs. 1-5), and less frequently, the right or left atrium (Fig. 6). Coronary cameral fistula can be easily detected on imaging studies because the affected coronary artery is usually large.

**Coronary-to-Pulmonary Artery Fistula**

As previously mentioned, coronary-to-pulmonary artery...
Fig. 5. Coronary cameral fistula between the right coronary artery and the LV in an 8-month-old girl.
A. Superior volume-rendered CT image showing the diffusely dilated right coronary artery (asterisks) in an 8-month-old girl. B. Inferior volume-rendered CT image revealing the abnormal connection (arrow) between the right coronary artery and the LV. C. Curved planar reformatted CT image demonstrating the entire course (arrows) of the fistula. AA = ascending aorta, CS = coronary sinus, LV = left ventricle, RA = right atrium, RV = right ventricle.

Fig. 6. Coronary cameral fistula between the right coronary artery and the RA in an 11-day-old boy.
A. Oblique coronal CT image showing the severely dilated origin and proximal portion of the right coronary artery (asterisk) in an 11-day-old boy. The left coronary artery (arrows) was normal in size. B. Oblique axial CT image revealing the fistulous connection (arrow) between the dilated proximal right coronary artery (asterisks) and the RA. AA = ascending aorta, DA = descending aorta, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle.
fistula is predominantly found in adults and rarely in children. In contrast to coronary cameral fistula, this fistula type most commonly arises from the proximal left anterior descending artery (45.3%–93.1%) and the proximal right coronary artery (24.5%–76.4%), and multiple origins are fairly common (30.2%–69.4%) [5,8,12,13,29]. Coronary-to-pulmonary-artery fistulas typically course anteriorly to the main pulmonary artery and drain into the anterolateral aspect, and often form a vascular network with multiple origins. Coronary arteries supplying the central pulmonary artery, as one of the major aortopulmonary collateral arteries, have been identified with ascending aortography or selective catheter coronary angiography in 10.0% of pediatric patients with pulmonary atresia and ventricular septal defect [30]. However, the tiny connection between the coronary artery and the central pulmonary artery may be easily missed on non-selective catheter angiography and other imaging modalities, including echocardiography, CT, and MRI [13].

**Coronary Artery-to-Coronary Sinus (or Any Other Cardiac Vein) Fistula**

Coronary artery fistulas terminating in the coronary venous system are relatively rare, accounting for 7.0% of cases [7]. The fistula may terminate in the coronary sinus (Fig. 7) or in its tributary cardiac veins (Fig. 8). Coronary artery-to-coronary sinus fistulas frequently present with congestive heart failure [31].

**Ventriculo-Coronary Arterial Connections in Pulmonary Atresia with an Intact Ventricular Septum**

Ventriculo-coronary arterial communications in pulmonary atresia with an intact ventricular septum are defined as abnormal vascular connections between the hypoplastic right ventricle and the coronary artery. Ventriculo-coronary arterial communications are present in approximately 40.0%
of cases [32]. Such communications typically occur in cases with severe hypoplasia and hypertrophy of the right ventricle [32]. Right ventricle-dependent coronary arterial circulations may cause right ventricular steal phenomenon if the affected coronary arteries are patent, and may cause myocardial ischemia or infarction when coronary artery obstruction coexists. For the imaging diagnosis of a right ventricle-dependent coronary arterial circulation, there should be angiographic evidence of myocardial perfusion through the fistulous communication. Right ventricle-dependent coronary arterial circulation occurs in slightly over half of the cases with ventriculo-coronary arterial connections, and is considered a risk factor for poor outcomes in patients undergoing an attempted biventricular repair, in which the right ventricle is decompressed and the coronary arterial pressure is subsequently dropped [32]. Therefore, Fontan operation without cardiopulmonary bypass is now preferred to avoid adverse coronary arterial events, as right ventricular unloading during cardiopulmonary bypass reduces coronary perfusion pressure in right ventricle-dependent areas, and results in myocardial ischemia [32]. Ventriculo-coronary arterial connections have been traditionally observed with cardiac catheter angiography [32], but recently, such abnormal fistulous connections can be delineated with coronary CT angiography (Figs. 9, 10) [5].

**Treatment**

Coronary artery fistulas should be treated if the fistula is large with substantial shunt flow that may lead to myocardial ischemia, ventricular dysfunction, and congestive heart failure [7,15,33]. Surgical ligation was solely performed before the development of transcatheter closure techniques, and is now used for fistulas not amenable to interventional treatment (Fig. 7). Since its introduction in the early 1980s, transcatheter closure of the fistula has been widely used as an effective and safe treatment for coronary artery fistulas (Fig. 10) [33].

Fig. 8. Coronary artery-to-middle cardiac vein fistula.
A. Axial coronary MR angiographic image showing a fusiform enlarged vascular space (asterisks) at the basal portion of the RV in a 2-year-old boy. B, C. Cardiac volume-rendered CT images performed 3 years later showing the mildly dilated left anterior descending artery (arrows). However, the enlarged vascular space (asterisks) detected on MRI was barely seen. D, E. Frontal selective catheter left coronary arteriographic images performed 4 years later demonstrating the abnormal connection (arrow) between the left anterior descending artery and the middle cardiac vein, delayed opacification of the enlarged vascular space (asterisks), and the cardiac veins. CS = coronary sinus, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle
Contraindication to percutaneous transcatheter closure include fistulas draining close to the atroventricular annulus, fistulas with origins at normal coronary artery branches in proximity to the occlusion site, extreme tortuosity or very small patient size making the procedure difficult, a wide draining site increasing the risk of coil migration, as well as multiple communications and drainage sites [7,15]. Coronary artery-to-coronary sinus fistula increases the risk for post-treatment adverse events such as coronary artery thrombosis and myocardial infarction [34].

**CONCLUSION**

In this article, the imaging findings of coronary artery fistula in children have been described. Coronary cameral fistula is the most common type of coronary artery fistula in children, and can be accurately diagnosed with CT. However, small or distal parts of other coronary artery fistula types in children may not be clearly depicted on CT, and selective catheter angiography may be necessary to characterize the whole angioarchitecture of such coronary artery fistulas. The detailed angioarchitecture delineated on coronary CT
angiography is not only useful for differentiating coronary artery fistulas from other abnormalities manifested as dilated coronary arteries, including anomalous coronary artery origin from the pulmonary artery and Kawasaki disease, but is also crucial for treatment planning and post-treatment evaluation.

**Conflicts of Interest**
The author has no potential conflicts of interest to disclose.

**REFERENCES**

1. Krause W. Über den Ursprung einer akzessorischen A. coronaria aus der A. pulmonalis. Z Klin Med 1865;24:225-227
2. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc
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17. Shabestari AA, Akhlaghpoor S, Fatehi M. Findings of bilateral coronary to pulmonary artery fistula in 64-multislice computed tomographic angiography: correlation with catheter angiography. *J Comput Assist Tomogr* 2008;32:271-273

18. Erol C, Seker M. Coronary artery anomalies: the prevalence of origination, course, and termination anomalies of coronary arteries detected by 64-detector computed tomography coronary angiography. *J Comput Assist Tomogr* 2011;35:618-624

19. Tangcharoen T, Bell A, Hegde S, Hussain T, Beerbaum P, Schaeffer T, et al. Detection of coronary artery anomalies in infants and young children with congenital heart disease by using MR imaging. *Radiology* 2011;259:240-247

20. Goo HW. Coronary artery imaging in children. *Korean J Radiol* 2015;16:239-250

21. Detorakis EE, Foukarakis E, Karavolias G, Dermitzakis A. Cardiovascular magnetic resonance and computed tomography in the evaluation of aneurysmal coronary-cameral fistula. *J Radiol Case Rep* 2015;9:10-21

22. Tsai IC, Lee T, Chen MC, Fu YC, Jan SL, Wang CC, et al. Visualization of neonatal coronary arteries on multidetector row CT: ECG-gated versus non-ECG-gated technique. *Pediatr Radiol* 2007;37:818-825

23. Ben Saad M, Rohanean A, Sigal-Cinquabre A, Adler G, Paul JF. Evaluation of image quality and radiation dose of thoracic and coronary dual-source CT in 110 infants with congenital heart disease. *Pediatr Radiol* 2009;39:668-676

24. Goo HW, Yang DH. Coronary artery visibility in free-breathing young children with congenital heart disease on cardiac 64-slice CT: dual-source ECG-triggered sequential scan vs. single-source non-ECG-synchronized spiral scan. *Pediatr Radiol* 2010;40:1670-1680

25. Goo HW, Seo DM, Yun TJ, Park JJ, Park IS, Ko JK, et al. Coronary artery anomalies and clinically important anatomy in patients with congenital heart disease: multislice CT findings. *Pediatr Radiol* 2009;39:265-273

26. Goo HW. Identification of coronary artery anomaly on dual-source cardiac computed tomography before arterial switch operation in newborns and young infants: comparison with transthoracic echocardiography. *Pediatr Radiol* 2018;48:176-185

27. Goo HW. Coronary artery anomalies on preoperative cardiac CT in children with tetralogy of Fallot or Fallot type of double outlet right ventricle: comparison with surgical findings. *Int J Cardiovasc Imaging* 2018;34:1997-2009

28. Goo HW. Quantitative evaluation of coronary artery visibility on CT angiography in Kawasaki disease: young vs. old children. *Int J Cardiovasc Imaging* 2021;37:1085-1092

29. Verdini D, Vargas D, Kuo A, Ghoshhajra B, Kim P, Murillo H, et al. Coronary-pulmonary artery fistulas: a systematic review. *J Thorac Imaging* 2016;31:380-390

30. Amin Z, McElhinney DB, Reddy VM, Moore P, Hanley FL, Teitel DF. Coronary to pulmonary artery collaterals in patients with pulmonary atresia and ventricular septal defect. *Ann Thorac Surg* 2000;70:119-123

31. Ogden JA, Stansel HC Jr. Coronary arterial fistulas terminating in the coronary venous system. *J Thorac Cardiovasc Surg* 1972;63:172-182
32. Freedom RM, Anderson RH, Perrin D. The significance of ventriculo-coronary arterial connections in the setting of pulmonary atresia with an intact ventricular septum. *Cardiol Young* 2005;15:447-468

33. Latson LA. Coronary artery fistulas: how to manage them.

34. Valente AM, Lock JE, Gauvreau K, Rodriguez-Huertas E, Joyce C, Armsby L, et al. Predictors of long-term adverse outcomes in patients with congenital coronary artery fistulae. *Circ Cardiovasc Interv* 2010;3:134-139