A 20-year-old man was admitted to our hospital for evaluation of his coronary arteries. His daily life activities were well established without any symptoms indicating myocardial ischemia. He had a history of repeat cardiac surgeries as a result of cyanosis consisting of a right Blalock-Taussig shunt 3 months after birth and a Fontan procedure at 2 years of age for pulmonary atresia with an intact ventricular septum (PA-IVS). Then, dilation of the middle left anterior descending (LAD) coronary artery without collaterals, an elevation of the right ventricular (RV) systolic pressure (86/8 mm Hg), and a decrease in the aortic pressure (81/51 mm Hg) were detected. Because there were no findings of the formation of coronary aneurysms of the dilated LAD coronary artery, the physicians carefully followed them up. Moreover, because of the RV-dependent coronary circulation, no surgical or catheter interventional repair of the PA to reduce the RV pressure, which could lead to myocardial ischemia in the LAD territory, was performed. He also experienced a myocardial infarction at 4 years of age that probably was attributable to the RV-dependent coronary circulation. Thus, he began to undergo medical treatment with anticoagulation therapy. The last coronary angiography at 10 years of age revealed no findings of any coronary aneurysm formation of the dilated LAD coronary artery or collaterals to the LAD territory. A recent multidetector computed tomography (MDCT) demonstrated the presence of a 23- and a 12-mm fusiform aneurysm in the LAD coronary artery draining into the RV (Figure 1A and 1B). The MDCT cine mode interestingly showed that the aneurysms expanded during the systolic phase (Figures 2A) and contracted during the diastolic phase (Figure 2B) in accordance with the cardiac pulsations (Movie I in the online-only Data Supplement) owing to an elevation in the RV systolic pressure. Moreover, 4-dimensional magnetic resonance imaging also visualized the retrograde coronary perfusion flow from the RV to the LAD coronary artery in accordance with the cardiac contractions (Figure 3 and Movie II in the online-only Data Supplement). Because he had no symptoms and the coronary aneurysms did not grow, we continued with careful medical treatment, including anticoagulation therapy and antihypertensive drugs, with close follow-up by MDCT.

PA-IVS and an RV-dependent coronary circulation are a rare congenital cardiac anomaly. Although very few infants can live beyond the first year without surgical therapy including a Fontan procedure, survival at 15 years after the Fontan procedure is 81.3%.1 The survivors are now reaching adulthood. Nearly half of the patients with PA-IVS have coronary arterial abnormalities,2 but giant congenital coronary artery aneurysms are very rare.3 Although aneurysmal dilation is also rare, it can be complicated by ruptures, distal thrombi, and myocardial ischemia.4 Moreover, myocardial ischemia and infarctions associated with PA-IVS and an RV-dependent coronary circulation are a well-established complication.1 On the other hand, in our present case, MDCT and magnetic resonance imaging interestingly helped to demonstrate the mechanism of the formation of the coronary aneurysm. Because of the elevated RV systolic pressure after the PA-IVS and tricuspid valve hypoplasia leading to RV volume overload, the RV to coronary arterial connections (fistulas) may have finally induced dilatation of the coronary aneurysms. To the best of our knowledge, there have been no reports of images demonstrating aneurysms that expanded during the systolic phase (Figures 2A) and contracted during the diastolic phase (Figure 2B) in accordance with the cardiac pulsations (Movie I in the online-only Data Supplement) and the retrograde coronary perfusion flow from the RV to the LAD coronary artery in accordance with the cardiac contractions (Figure 3 and Movie II in the online-only Data Supplement). In view of these findings, even after the abolishment of cyanosis leading to coronary dilation, coronary aneurysms may occur as a result of the RV-dependent coronary circulation of the PA-IVS. Furthermore, because the natural history and adequate treatment for coronary aneurysms are currently still unknown, physicians should follow them up closely.
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Disclosures
None.

References

Figure 1. The 3-dimensional (A) and transverse (B) imaging of multidetector computed tomography demonstrates the presence of a 23-mm fusiform aneurysm (arrows) on the left anterior descending (LAD) coronary artery draining into the right ventricle (RV).

Figure 2. The multidetector computed tomography cine mode interestingly shows that this aneurysm (arrows) on the left anterior descending (LAD) coronary artery draining into the right ventricle (RV) expands in the systolic phase (A) and contracts in the diastolic phase (B) in accordance with the cardiac pulsations.
Figure 3. Four-dimensional magnetic resonance imaging also visualizes the retrograde coronary perfusion flow from the right ventricle (RV) to the aneurysm (arrows) on the left anterior descending (LAD) coronary artery in accordance with the cardiac contractions.
Giant Pulsatile Coronary Aneurysms Associated With Pulmonary Atresia With an Intact Ventricular Septum After a Fontan Procedure
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