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An unusual cause of cyanosis after intra-extra cardiac Fontan procedure: anastomotic leakage between conduit and inferior vena cava

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A child aged 7 years was admitted to our hospital nearly 5 years after an intra-extra cardiac Fontan operation due to aggravated cyanosis. He was diagnosed with pulmonary atresia (PA), atrial septal defect, patent ductus arteriosus (PDA) and thickened tricuspid valve with severe regurgitation 1 month after being born, which was considered PA with intact ventricular septum (IVS) with severely hypoplastic right ventricle and was similar to functional single ventricle. All surgical approaches are based on the hemodynamic approximation of a functional single ventricle. His surgical strategy was as follows: first stage, a modified Blalock-Taussig (B-T) shunt was performed at 1 month of age to promote development of dysplastic pulmonary vessels; at 6 months, cardiac catheterization demonstrated well-developed pulmonary arteries after B-T shunt, so we performed a bidirectional Glenn shunt operation, enlargement of atrial septal defect, ligation of B-T shunt and PDA as a transition to Fontan operation; finally, a fenestrated intra-extra cardiac Fontan operation was conducted to achieve a total cavopulmonary anastomosis at 3 years of age. Mild cyanosis appeared during 3-month follow-up after Fontan operation. We considered that it was caused by increased pulmonary vascular resistance, contributing to a growing right to left shunt through the fenestration and regular follow-up visit was required. However, he was lost to follow-up for almost 3 years. His cyanosis became more severe and exercise tolerance was decreased when he visited in October 2022, and SpO_2 was only 75%. However, there was no specific finding other than bidirectional shunt with dominant left to right shunt through fenestration from echocardiography. He was admitted to hospital for further examination. CT angiography demonstrated a high signal at anastomotic between conduit and inferior vena cava and extended to atrium, which implied a possibility of anastomotic leakage (figure 1). Further cardiac catheterization indicated that there was no significantly elevated pressure in pulmonary artery and inferior vena cava. Cardiac catheterization demonstrated that there was a tunnel-like defect at the lower back of conduit with a width of 7.5 mm and catheterization angiography revealed a right to left shunt (figure 2A). During catheterization we tried a closure of fenestration (figure 2B). The SpO_2 increased to 86% from 82% 15 min after closure of conduit fenestration. A PDA occluder was placed for closure of anastomotic leakage (figure 2B). SpO_2 increased to 94% after 15 min. No shunt was found through fenestration and anastomosis with cardiac catheterization angiography and SpO_2 increased to 100% when he was transferred to inpatient ward. He was discharged

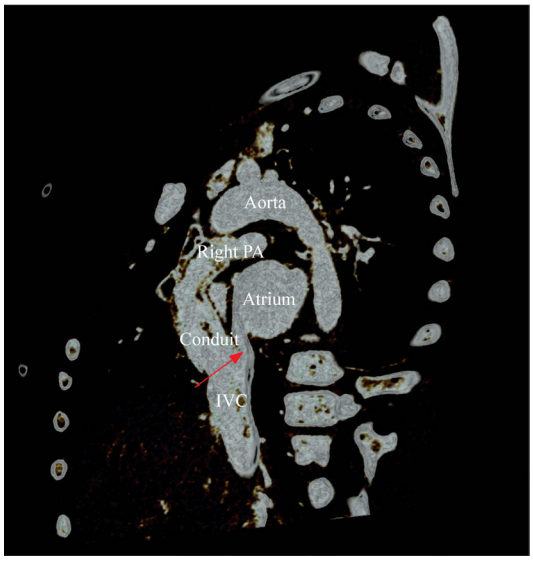

Figure 1 Cardiac CT angiography. Red arrow points to the residual shunt at anastomosis between conduit and inferior vena cava (IVC). PA, pulmonary artery.

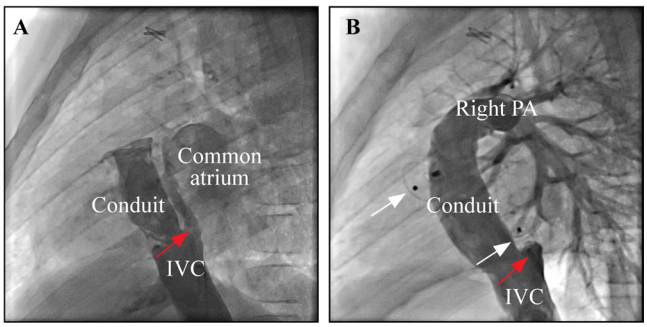

Figure 2 Cardiac catheterization angiography before and after occlusion. (A) Red arrow indicates the residual shunt at anastomosis between conduit and inferior vena cava (IVC). (B) Red arrow indicates that the residual shunt is disappeared. Upper white arrow points to the fenestration occluder and lower white arrow points to the PDA occluder for closure of residual shunt. PA, pulmonary artery.

from hospital on the second day after closure without cyanosis. One month after discharge, he came to our hospital for follow-up without cyanosis and exercise tolerance was improved. Echocardiographic examination revealed occluders were in the right place without residual shunt.

PA is a rare congenital heart disease, which is divided into PA with ventricular septal defect (VSD) and PA/IVS, depending on whether VSD exists. PA/IVS is also defined as hypoplastic right heart syndrome. The operation method of PA/IVS is selected in consideration of cardiac malformation and the developmental condition of right heart.[1 2] The right heart of this child was severely hypoplastic, which could be regarded as functional single ventricle, so we decided to finally achieve a Fontan operation in this situation.[3 4] Based on previous total cavopulmonary connection operation, an intra-extra cardiac Fontan operation was performed with fenestration between conduit and atrium to balance systemic pulmonary circulation, stabilizing the circulation and reducing hydrothorax and ascites. However, fenestration would lead to increased right to left shunt and risk of low systemic arterial oxygen saturation.[5] To detect the cause of his cyanosis, CT angiography detected a tunnel-like leakage at anastomosis between inferior vena cava and conduit (figure 1). The lower end of the conduit was wrapped by atrium which led to the possibility of anastomotic leakage and would cause cyanosis if the suturing was loosened (figure 3). To confirm this anastomotic leakage, cardiac catheterization angiography was performed and pressure of conduit and pulmonary artery was not high. Angiography illustrated two right to left shunts through fenestration and anastomosis (figure 2A). Fontan fenestration may persist over years and cause systemic desaturation and exertional dyspnea due to a persistent right to left shunt, increasing the risk of thromboembolic events and brain abscess. Therefore, transcatheter device closure of the fenestration is often considered after Fontan operation. Due to satisfactory pressure and resistance of pulmonary artery, we initially tried a closure of fenestration because this shunt was relatively small. After the closure of fenestration, the pressure of conduit changed little and we could close the residual shunt safely to alleviate his cyanosis.[6 7] Finally, he was discharged from hospital with good outcome and informed follow-up.

In this child, an intra-extra cardiac Fontan operation was applied. The strengths of this surgical method are: there is no need to transect inferior vena cava, the conduit cannot easily twist, the direction of blood flow is appropriate, operation is convenient and the risk of arrhythmia is relatively low. However, with lower half of conduit wrapped by atrium, anastomotic leakage would happen if the suture was not perfect or loosened. Therefore, if cyanosis was found after intra-extra cardiac Fontan operation with fenestration, besides right to left shunt through fenestration due to increased pulmonary circulation pressure and resistance, a right to left shunt caused by anastomotic leakage should be considered as well. The leakage was not detected by echocardiography due to the undetectable location of leakage in this child, and it was easy to have missed diagnosis. However, it was detected by CT angiography and confirmed by catheterization.

In conclusion, residual shunt at anastomosis between conduit and inferior vena cava is a rare cause of cyanosis in intra-extra cardiac Fontan operation. If cyanosis appeared postoperatively, anastomotic leakage should be considered. An echocardiography examination should be conducted carefully. Meanwhile, for cardiac surgeon, anastomosis between conduit and inferior vena cava should be enhanced to reduce the risk of leakage.

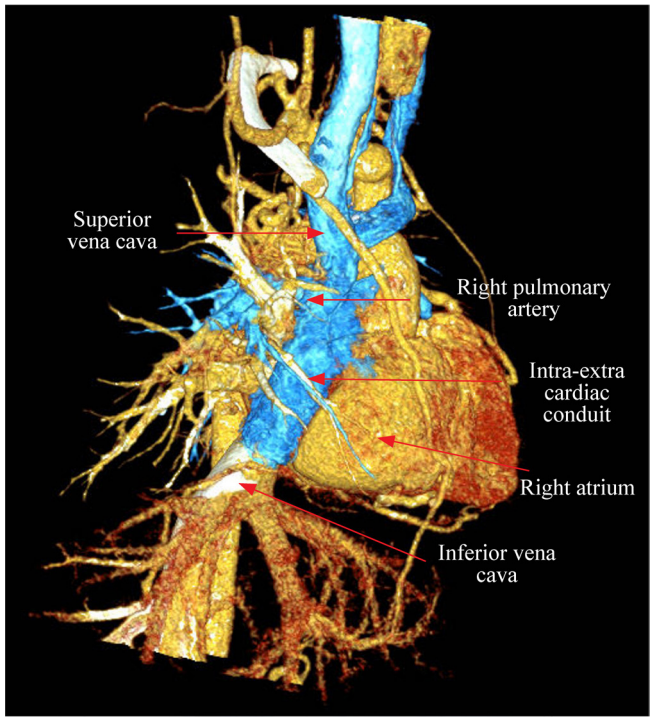

Figure 3 Cardiac CT angiography of the child 3 years after Fontan procedure.

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