THREE CASES OF NEWLY APPEARED PULMONARY ARTERIOVENOUS FISTULA AFTER FONTAN OPERATION

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History and physical:

Pulmonary arteriovenous fistula (P-AVFs) appearing after bilateral cavopulmonary shunt (BCPS) is well-known phenomenon. However, newly appearing P-AVFs after Fontan operation are reported to be rare. Absence of hepatic factor in pulmonary blood flow had been suspected to influence development of P-AVFs. Here, we present three cases of newly appeared P-AVFs after Fontan operation

Case 1

The patient was diagnosed with right isomerism with functional single ventricle. He underwent staged procedures including balloon pulmonary valvuloplasty and BCPS. There was no P-AVFs in pre-Fontan evaluation. Extracardiac Fontan without fenestration was done at 33 months of age. At 13 years of age, the patient's oxygen saturation at room air (SpO2) was 90-91%. Because of Fontan pathway narrowing, balloon dilation and stent insertion at extracardiac conduit was done. The patient had received growth hormone therapy (GHT) from 15 years of age. He was admitted for progressive desaturation two months after the initiation of GHT. There was no significant Fontan pathway stenosis. Pulmonary angiography revealed diffuse P-AVFs. The patient's condition was tolerable, and thus, he was followed-up without intervention. Two years later, the patient died of severe pneumonia.

Case 2

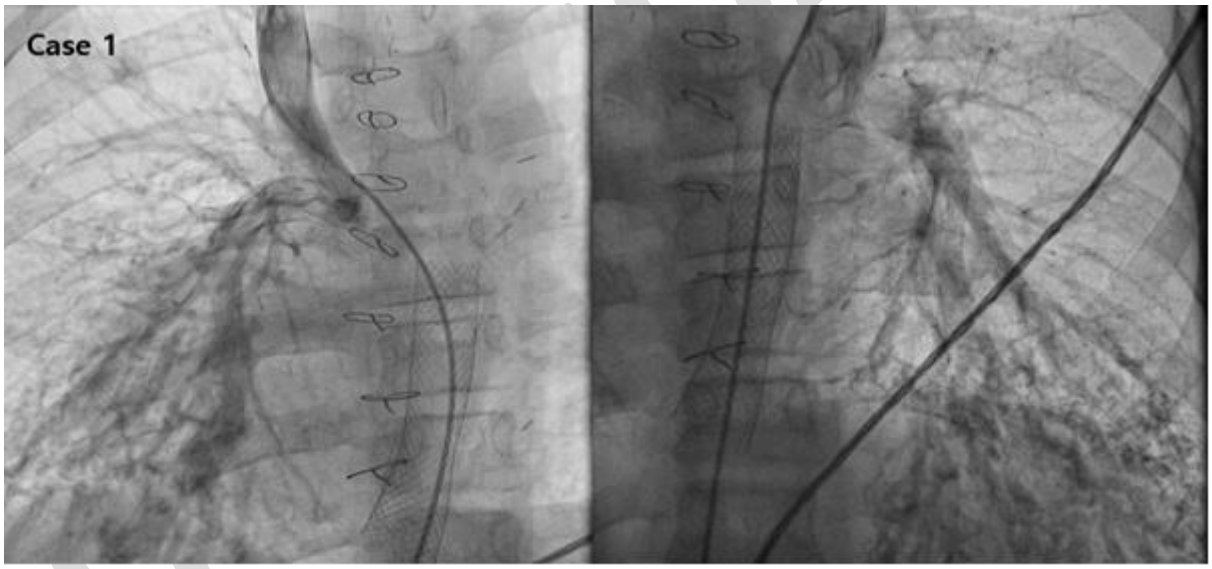
The patient was diagnosed with pulmonary atresia with intact ventricular septum, severe right ventricle hypoplasia. He underwent right modified Blalock-Taussig shunt and right ventricular outflow tract widening at neonatal period. Later, BCPS and right pulmonary artery angioplasty was done. There was no evidence of P-AVFs in pre-Fontan evaluation. Fontan operation with 4mm fenestration was done at 20 months of age. Because of left pulmonary artery (LPA) stenosis, balloon angioplasty was done at seven years of age. He complained of dyspnea on exertion at 19 years of age and his SpO2 was 85-91%. Heart CT revealed severe stenosis at LPA

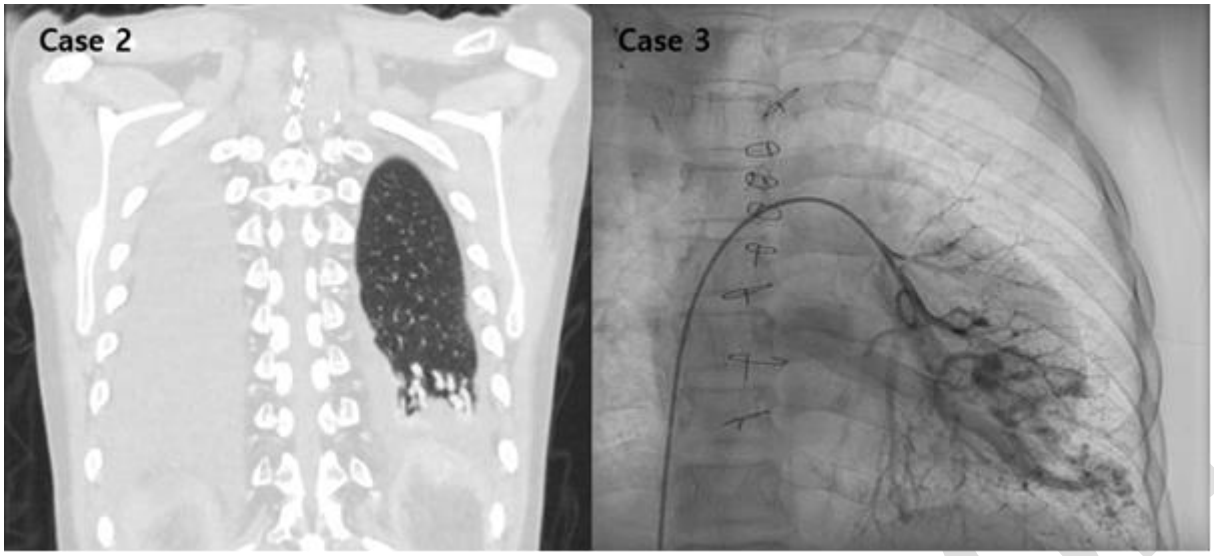
origin. Therefore, he underwent LPA stent insertion. Later, he was diagnosed with hepatocellular carcinoma (HCC) at 23 years of age. Progressive desaturation occurred and evident P-AVFs in left lower lung fields were identified at 25 years of age. Because of recurred HCC and lung metastasis, the patient have been followed-up without intervention.

Case 3

The patient was diagnosed with tricuspid atresia, mild pulmonary stenosis. Noonan syndrome was diagnosed with gene study. Pulmonary artery banding was done in neonatal period. Bilateral BCPS was done at nine months of age. There was no P-AVFs in pre-Fontan evaluation. Extracardiac conduit Fontan with 5mm fenestration was done. The patient had received GHT from six years of age. She was admitted for progressive desaturation at 12 years of age. Her SpO2 was the late 70%. There was no Fontan pathway obstruction. Multiple p-AVFs were identified at left lower lung field. P-AVFs were embolized with multiple devices including Piccolo and Concerto detachable coils. Her SpO2 increased to 85-86% after the procedure.

Imaging:





Indication for intervention:

Evaluating the causes of desaturation after Fontan operation.

Intervention:

Diagnostic catheterization and embolization.

Learning points of the procedure:

P-AVFs can develop and progress after the Fontan operation despite balanced hepatic flow. P-AVFs should be considered as a potential cause of desaturation after Fontan operation.