Occlusion of Complex Veno-atrial Collaterals in a Fontan Circulation

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Introduction
An 8-year-old boy with Double outlet right ventricle and hypoplastic left ventricle underwent Fontan completion by 4 years of age. He developed progressive cyanosis with resting Oxygen saturations of 65%. Absence of a fenestration made a venous collateral decompressing the Fontan circuit likely. We proceeded to cardiac catheter to assess his fontan circulation.

Case
Mean pressure in the Fontan was 13 mmHg and pathways unobstructed. IVC angiogram showed multiple complex systemic venous collateral channels from the IVC and hepatic veins draining to a common site in the left atrium.

Engaging the collateral from femoral approach was difficult due to the angle and tortuous course. A jugular vein approach was adopted. Multiple stenoses resulted in inability to advance the sheath to the desired position, the LA.

We occluded the major individual channels using a 10 mm(a) and 12 mm(b) Amplatzer Vascular plug II and a 6x6mm(c) Amplatzer ductal occluder II. Angiogram confirmed reduced flow through the collaterals and stable device position. Saturations improved to 86%.

Discussion
Systemic venous collaterals can decompress the higher-pressure Fontan circuit to the lower pressure atrium or pulmonary vein, resulting in symptomatic cyanosis. After bidirectional cavopulmonary anastomosis incidence of systemic venous collaterals was 17% in a series by Gross et al and 31% by Magee et al. Weber detected significant systemic venous collaterals in 18% post Fontan.

Risk factors for collateral development include: gradient between the superior vena cava and right atrium, increased transpulmonary gradient, heterotaxia and high mean PA pressure. Sugiyama et al found 68 systemic venous collaterals in 50 cyanosed patients post Fontan. These were supracardiac in 36 (53%), cardiac in 12 (18%), and infracardiac in 20 (29%). The brachiocephalic vein was the most common site with 30 of the 68 collaterals (44%).

Mehta et al have described accessing the pulmonary venous atrium from the PA floor, a potential route for us to place a device at the LA exit site.

Decompressing systemic venous collaterals continue to be problematic in the Fontan circuit. Longer term data is required to assess the impact of collaterals in patients who have and have not undergone occlusion.