TRANSCATHETER PERFORATION OF ATRETIC CORONARY SINUS OSTIUM IN A COMPLEX SINGLE VENTRICLE CASE: RARE BUT CRITICAL ANOMALY FOR UNIVENTRICULAR REPAIR

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History and physical:

A 5-year-old female patient was hospitalized for planned catheterization. She was diagnosed as single ventricle, common atrioventricular valve, pulmonary atresia and major aortopulmonary collateral arteries (MAPCA) at birth. Uniforcalization of MAPCAs and right modified BT shunt procedure was performed when she was 7 months old. At the age of 4, due to insufficient pulmonary blood flow and uncontrollable AVV regurgitation, central shunt addition and artificial valve replacement were performed. She was 103cm tall and weigh 15.0kg. Her SpO2 was 80%. At the catheterization, her single ventricle end-diastolic pressure (SVEDP), central venous pressure (CVP) and ejection fraction (EF) were 11mmHg, 12mmHg, and 45% respectively. It was diagnosed for the first time that her coronary sinus (CS) ostium was atretic and most of coronary vein blood flow drained via persistent Marshall vein to the innominate vein. Besides, Marshall vein was stenotic in the middle of it and there was fistula communicating to left pulmonary artery (LPA).

Imaging:

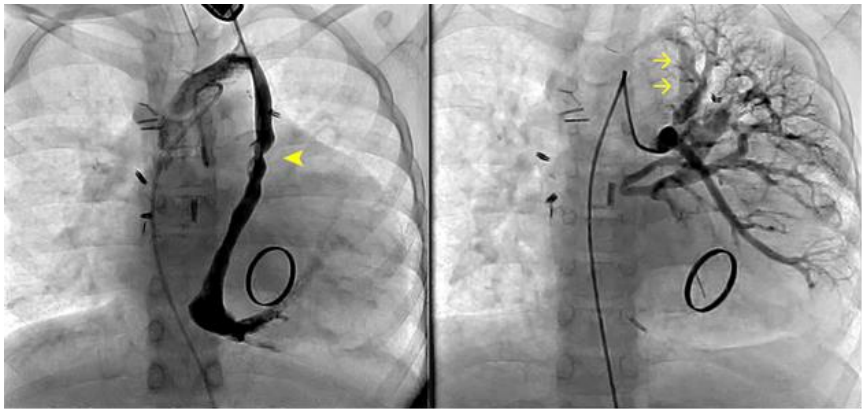

Figure 1. : Stenotic position of Marshal vein (arrowhead) at which LPA communicated to Marshal vein found in the late phase of LPA angiogram (arrows showing Marshall vein draining into innominate vein).

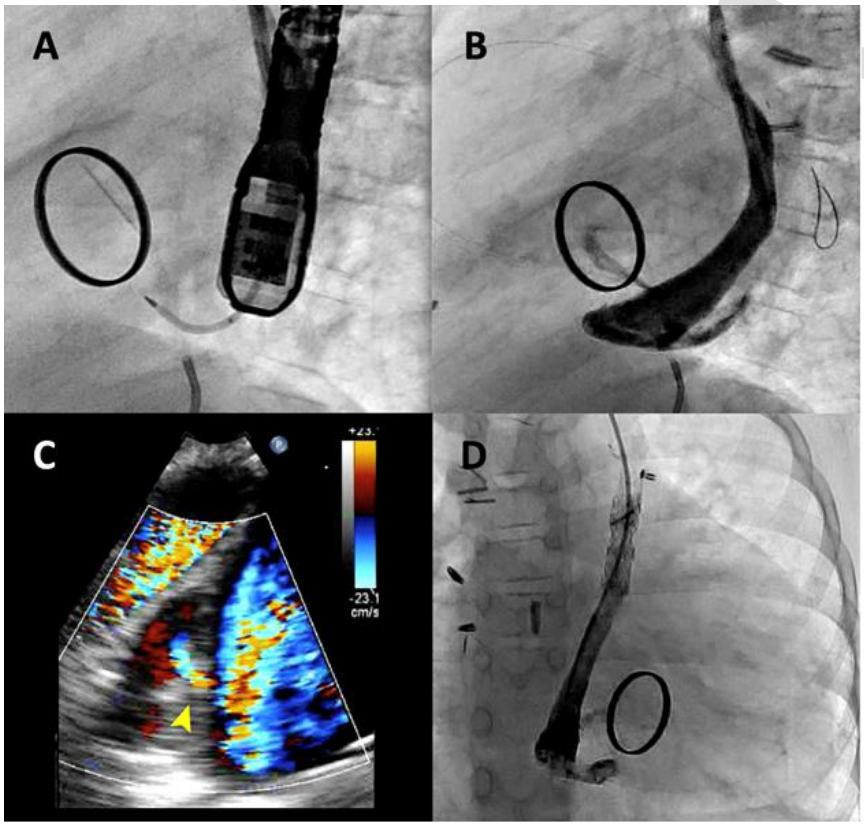

Figure 2: : Fluoroscopic position of successful radiofrequency perforation of CS ostium. (B) Created CS ostium after perforation and balloon dilation. (C) TEE image showing communication between CS and right atrium through the thickened CS wall (arrowhead). (D) Dilated Marshall vein stenosis after VIAVAHN covered stent placement.

Indication for intervention:

Transcatheter perforation of atretic CS ostium and simultaneous covered stent implantation for Marshall vein stenosis and fistula was planned to improve coronary circulation and ventricular function certainly. Surgical intervention was hesitated because of reduced ventricular function.

Intervention:

CS angiography revealed atretic CS ostium and stenotic Marshal vein of which minimum diameter was 3.4mm. 7F destination long sheath (Terumo, Tokyo, Japan) was advanced close to CS ostium. Then, 4F JR catheter and 2.9F Leonis Mova steerable micro catheter (Sumitomo Bakelite, Tokyo, Japan) with Nykanen RF wire (Baylis Medical, Ontario, Canada) system was advanced and its tip was pushed against CS ostium and radiofrequency power was delivered under TEE guidance. However, perforation was not achieved after several attempts. Given the TEE view showing thickened CS wall, the radiofrequency settings was changed from pulse mode to constant mode so that the radiofrequency power was able to be delivered longer time. Finally, perforation was achieved, and the hole was subsequently dilated with 2mm Coyote (Boston scientific, MA, US), 5mm Sterling, and 8mm Mustang (Boston scientific, MA, US) balloon catheters. After that, 7mm x 29mm VIAVAHN VBX covered stent (W.L. Gore & associates, DE, US) was placed at the position of stenosis and fistula. 1 month later, her catheterization data showed improved CVP, SVEDP and EF as 7mmHg, 8mmHg and 63%, respectively.

Learning points of the procedure:

CS ostium atresia is an extremely rare anomaly. In the process of univentricular repair, it would aggravate ventricular function as a consequents of Marshall vein stenosis or Glenn procedure by elevating CS pressure. Transcatheter radiofrequency puncture was feasible even for thickened CS wall.