TRANSCATHETER FONTAN COMPLETION IN AN ADOLESCENT WITH CRISS CROSS HEART, SITUS INVERSUS-DEXTROCARDIA

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History and physical:

17 years old, 38 kg, female with a situs inversus-dextrocardia, Criss Cross heart, double outlet right ventricle, ventricular septal defect and significant pulmonary stenosis, had undergone BT shunt twice was referred for advanced cyanosis with spO2 %55. At first hemodynamic assessment was suitable and she was carried over bidirectional cavopulmonary connection (BCPC) with preparation for a subsequent Transcatheter Fontan completion procedure after the cardiac unit interdisciplinary council.

Imaging and intervention:

BCPC and preparation for Fontan: The one side of an 20 mm PTFE tube was sutured end to end to the stump of the IVC. A fenestration was made on the atrial side of the PTFE tube and sutured to the right atriotomy to create a nonrestrictive communication. The BCPC was performed by anastomosing (end to side) the transected SVC to a longitudinal incision on the superior border of the RPA. After performing right BCPC, incision was made on the inferior RPA, and a patch of autologous pericardium was prepared and sewed to the top of the PTFE graft that revealed the IVC return exclusively to the atrium.

After the BCPC there was a short stay in ICU, she was discharged home with spO2 %75. She called for an Transcatheter Fontan completion procedure eight months later. Under general anesthesia a 6F sheath was placed in the left internal jugular vein (LIJV) and a 8F sheath in the left femoral vein (LFV) with the Seldinger technique. Firstly 22 mm covered CP stent (NUMED, Inc.) was implanted by using 8x20 mm Z Med ballon at the stenotic part of the RPA. Angiography was performed simultaneously via a 6F Pigtail catheter in the IVC and a 5,2F Pigtail catheter (Cordis Corporation Miami, USA) in the SVC. It demonstrated a 3-mm thick plate of the Corematrix patch in the conduit dividing the upper and lower portions of the previously placed PTFE graft. Widely patent BCPC, branch pulmonary arteries and conduit fenestration to

the RA were confirmed (Movie, part 1). The loop of a snare catheter was positioned in the inferior part of the PTFE conduit from the LFV to provide a target zone. The Corematrix membrane occluding the PTFE conduit was perforated with a 0,018'' Halberd hydrophilic guidewire guided by a 5,2F multipurpose catheter passed from the LIJV (Movie Part 2). The Halberd (ASAHI, Intecc, USA.INC.) hydrophilic wire was snared. The catheter was then passed across the membrane and the Halberd wire exchanged for a 0.035'' Extra Stiff Amplatzer guidewire (Boston Scientific, Marlborough, Massachusetts) which was extracted from the internal jugular vein sheath to create a stable wire track. A 18F Mullins sheath (Cook Medical, Bloomington, USA) was placed anterogradely from the IVC. A 57-mm Optimus XL PTFE Covered Vascular Stent (AndraTec, GmbH, Germany) was mounted with a 22x50 mm Z-Med-II ballon deployed within the conduit after checking that it completely occluded the fenestration to the RA. (Movie part 3). After stent implantation pulmonary anterograde closure was performed with 11mm Amplatzer ASD device (Movie part 4). Hemodynamics after Fontan demonstrated an SVC pressure of 13 mmHg and s02 in the high 90s. Angiography showed a widely patent channel from the IVC to the RPA with no residual fenestration. Patient was transferred to the intensive care unit overnight; pleural drainage was not needed during the follow-up. She was discharged home on post procedure day 10 with s02 in the high 90s .

Learning points of the procedure:

Glenn operation for single ventricle palliation and transcatheter Fontan preparation were performed in the single ventricle patient presenting late with severe hypoxemia. Repetitive surgical interventions are required for single ventricle patients; therefore, transcatheter Fontan preparation is an effective and safe method in patients with late admission..

Movie:

ZB movie.mp4