STUDIUM PRZYPADKU / CLINICAL VIGNETTE

Successful hybrid management for a patient with tricuspid atresia and innominate vein obstruction

Skuteczne leczenie hybrydowe pacjenta z atrezją zastawki trójdzielnej i zakrzepicą żyły bezimiennej

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A 2-month-old and 5.65-kg baby, diagnosed with tricuspid atresia type IB with pulmonary stenosis and an unrestrictive ventricular septal defect, underwent a modified Blalock-Taussig shunt as an emergency due to intractable critical cyanosis. Even though he took aspirin 5 mg/kg/day after the modified Blalock-Taussig shunt, 11 months after the shunt, angiogram revealed complete occlusion of the innominate vein. The collateral veins drained into the superior vena cava through the haemiazygos vein and the azygos vein, and the multiple small collaterals also drained into the inferior vena cava (Figs. 1A, B). We, therefore, took the hybrid approach to include venous drainage from the left upper extremity into the planned right bidirectional cavopulmonary shunt. In the catheterisation laboratory, the coil embolisation of the distal accessory haemiazygos vein and the small collaterals was performed by a paediatric cardiologist. On the same day, the patient was transferred to the operating room to undergo a bidirectional cavopulmonary shunt. This bidirectional cavopulmonary

shunt was successfully done including venous flow from the left upper trunk into the pulmonary bed leaving the azygos vein open. The patient was discharged without any clinical problems. Postoperative computed tomography (CT) showed U-shaped large azygos and accessory haemiazygos vein (Fig. 2). At 5 years of age, a Fontan operation was successfully performed using a 20-mm extracardiac Gore--Tex conduit. Aspirin and warfarin were used as anticoagulant medication after the Fontan operation. Good run-off to the previous Glenn shunt with the azygos vein open-wide was confirmed. The patient has been followed-up for 5 years without any problems taking the aspirin and warfarin. Usually, the azygos vein must be ligated during the bidirectional cavopulmonary shunt procedure. We, however, had a rare experience in which the azygos vein was an important venous route in a patient with innominate vein thrombosis in a bidirectional cavopulmonary shunt. In this case, the thrombosis was caused by a central venous line placed in the previous operation and angiogram showed that venous flow was completely occluded by the innominate vein thrombosis, which was drained via the haemiazygos azygos veins. We then successfully treated the innominate vein thrombosis in the patient facing a bidirectional cavopulmonary shunt with the trans-catheter procedure, followed by a bidirectional cavopulmonary shunt on the same day.







Figure 2. Coils were placed to occlude the collateral vessels (**A**). Follow--up CT. The distal accessory haemiazygos veins and small collaterals were completely occluded using coils. The U-shaped large azygos and accessory haemiazygos veins drain to the superior vena cava (SVC) (white arrows) after a bidirectional cavopulmonary shunt (**B**); LSVC — left superior vena cava; v — vein

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