Complex criss-cross heart malformation: Completed comprehensive multistage hybrid treatment

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A criss-cross heart (CCH) congenital heart defect (CHD) concomitant with atrioventricular (AV) cross-flow is a complex and extremely rare heart defect. CCH develops due to abnormal rotation of ventricles around the long axis of the heart while the base of the heart and the AV valves remain fixed. Due to its rarity (less than 0.1% of all CHDs), it is often misdiagnosed. The treatment depends on the anatomical possibilities of preserving the biventricular heart, otherwise, Fontan palliation following a single ventricle pathway is chosen [1–3]. Current literature on the completion of successful treatment of complex CCH-type CDH is limited.

We present a unique report of a 4-year-old girl who presented with prenatally diagnosed CHD in the form of CCH concomitant with dextrocardia, malposition of the great arteries, and pulmonary stenosis, and who has recently accomplished multistage, hybrid-surgical palliative treatment following a single ventricle pathway.

The child was born at 38 weeks of gestation in good condition. A chest X-ray showed the heart located medially, heart apex facing right, abdominal aorta and stomach on the left side of the spine, inferior vena cava, and liver on the right (Figure 1A). Echocardiography showed cross-flow and abnormal AV connections: the right ventricle (RV) connected via the tricuspid valve to the left atrium, the left ventricle (LV) connected via the mitral valve to the right atrium, accompanied by valvular and sub-valvular pulmonary artery (PA) stenosis (PS-RV/PA gradient approx. 90 mm Hg), unrestricted ventricular septal defect (VSD), malposition of the great arteries (aorta originating from RV and PA overriding VSD) (Figure 1B, Supplementary material, *Video S1*). The final diagnosis of congenital corrected malposition of the great arteries with atrioventricular discordance, VSD, and sub-pulmonary stenosis was made.

Due to unrepairable, multilevel stenosis of the right ventricular outflow tract (RVOT) which after double-switch correction should be systemic ventricular outflow tract, the child was referred for single-ventricle, multistage, and palliative treatment. First, percutaneous pulmonary valvuloplasty and right-side Blalock-Taussig shunt (BT dex) were performed during the neonatal period, without complications. At the age of 6 months, the bidirectional Glenn procedure (superior vena cava to right PA shunt) was executed under extracorporeal circulation (Figure 1C). After 2 years, due to the narrowing of the left pulmonary artery (LPA) to 6 mm (9 mm at PA ostium), successful percutaneous balloon dilatation of LPA was performed. Next, total extracardiac Fontan (total cavopulmonary connection [TCPC]) operation was executed with intraoperative, hybrid, balloon dilatation of LPA to ensure unobstructed pulmonary circulation. Echocardiography showed a good surgical outcome (Figure 1D, Supplementary material, *Video S2*). However, the postoperative course was

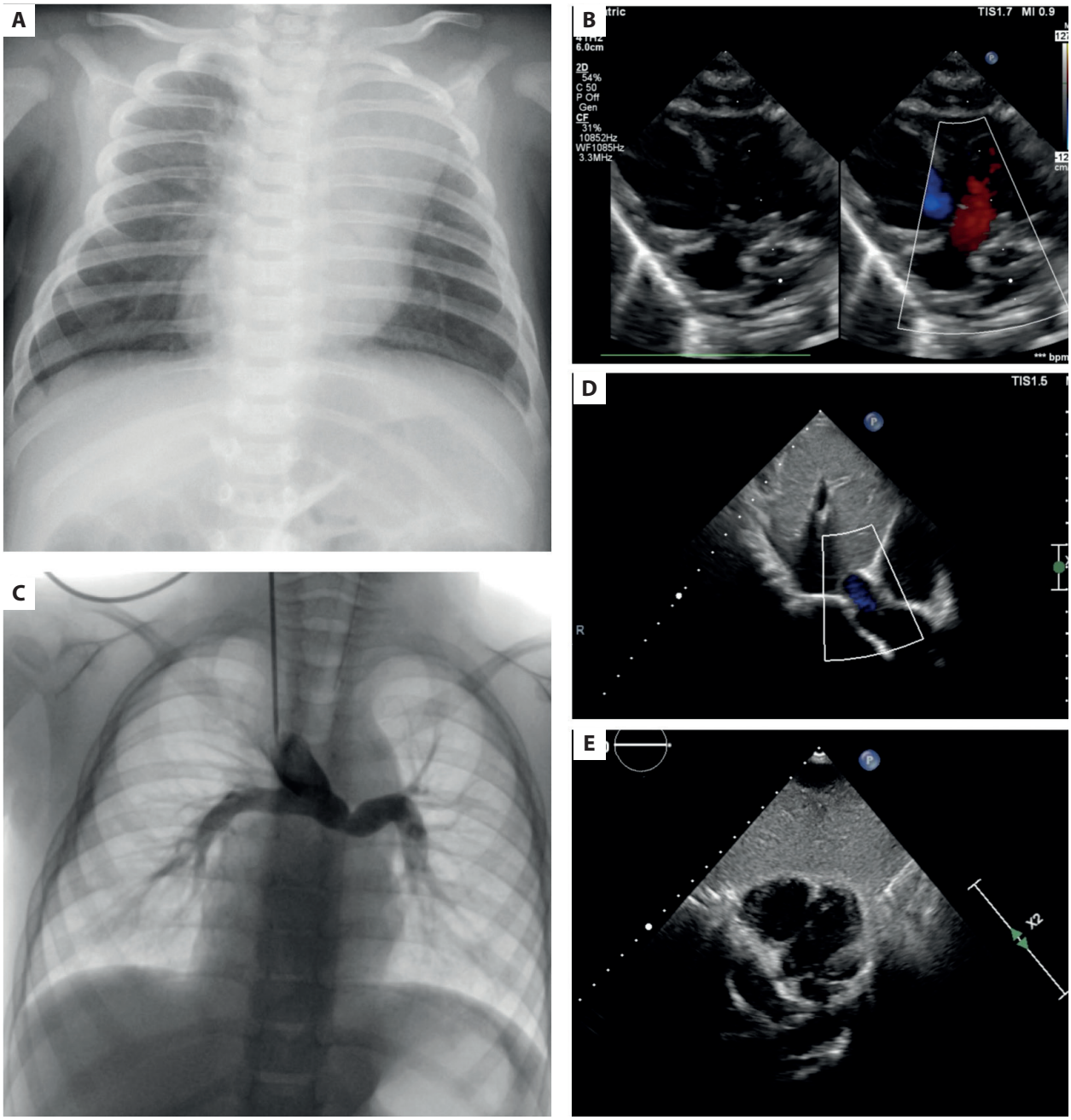

Figure 1. A. Chest X-ray (neonate period) — heart located medially, heart apex facing right. **B.** Echocardiography (neonate period) of the criss-cross heart morphology — cross-flow and abnormal AV connections — more in Supplementary materials, *Video S1*. **C.** Heart catheterization after the bidirectional Glenn procedure at the age of 6 months. **D.** Echocardiography after Fontan (TCPC) operation — more in Supplementary materials, *Video S2*. **E.** Control echocardiography at the age of 4 years old — good function of the functionally univentricular heart, effective Fontan flow with respiratory-dependent variability — more in Supplementary materials, *Video S3*

Abbreviations: AV, atrioventricular, TCPC, total cavopulmonary connection

complicated by right diaphragm paralysis and chylothorax. Therapy included somatostatin, total parenteral nutrition, anti-inflammatory drugs, and steroids. After recovery, the child was discharged home in good general condition, with efficient cardiac performance and diaphragm function. Acetylsalicylic acid and sildenafil were prescribed.

The recent examination of the 4-year-old girl showed satisfactory development. Echocardiography confirmed good function of the functionally univentricular heart, effective Fontan flow with respiratory-dependent variability, with good contractility (Figure 1E, Supplementary material, *Video S3*).

More than 90% of CCH patients present with levocardia, while the concomitance of AV-related malposition of the great arteries and dextrocardia is the second case ever described [4]. This report is a rare description of the completed treatment of this complex CCH morphology, with good results and promising follow-up.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska

Article information

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