KARDIOLOGIA POLSKA

Polish Heart Journal
The Official Peer-reviewed Journal of the Polish Cardiac Society since 1957

Online first
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ISSN 0022–9032 e-ISSN 1897–4279

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Article type: Clinical vignette

Received: November 1, 2022

Accepted: December 14, 2022

Early publication date: January 10, 2023

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

Short title: Multistage palliation of criss-cross heart congenital heart defect

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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 of less than 0.1% of all CHDs it is often misdiagnosed. The treatment

depends on anatomical possibility to preserve biventricular heart, otherwise Fontan palliation following single ventricle pathway is chosen [1–3]. Current literature on successful treatment completion of complex CCH-type CDH is limited.

We present an unique report of 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 successfully accomplished multistage, hybrid-surgical palliative treatment following single ventricle pathway.

The child was born at 38 weeks gestation in good condition. The 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). The echocardiography showed cross-flow and abnormal AV connections: right ventricle (RV) connected via tricuspid valve to the left atrium, left ventricle (LV) connected via 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 atrio-ventricular discordance, VSD and sub-pulmonary stenosis was made.

Due to unrepairable, multilevel stenosis of the RVOT, which after double-switch correction should be systemic ventricle outflow tract the child was referred for single-ventricle, multistage, 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 (TCPC) operation was executed with intraoperative, hybrid, balloon dilatation of LPA to ensure unobstructed pulmonary circulation. The echocardiography showed good result of the surgery (Figure 1D, Supplementary material, *Video S2*). However, the postoperative course was complicated by right diaphragm palsy 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. Acetylic acid and sildenafil were prescribed.

The recent control of 4 years-old girl showed satisfactory patient's development. The 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 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 completed treatment of this complex CCH morphology, with good result and promising follow-up.

Supplementary material

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

Article information

Conflict of interest: None declared.

Funding: None.

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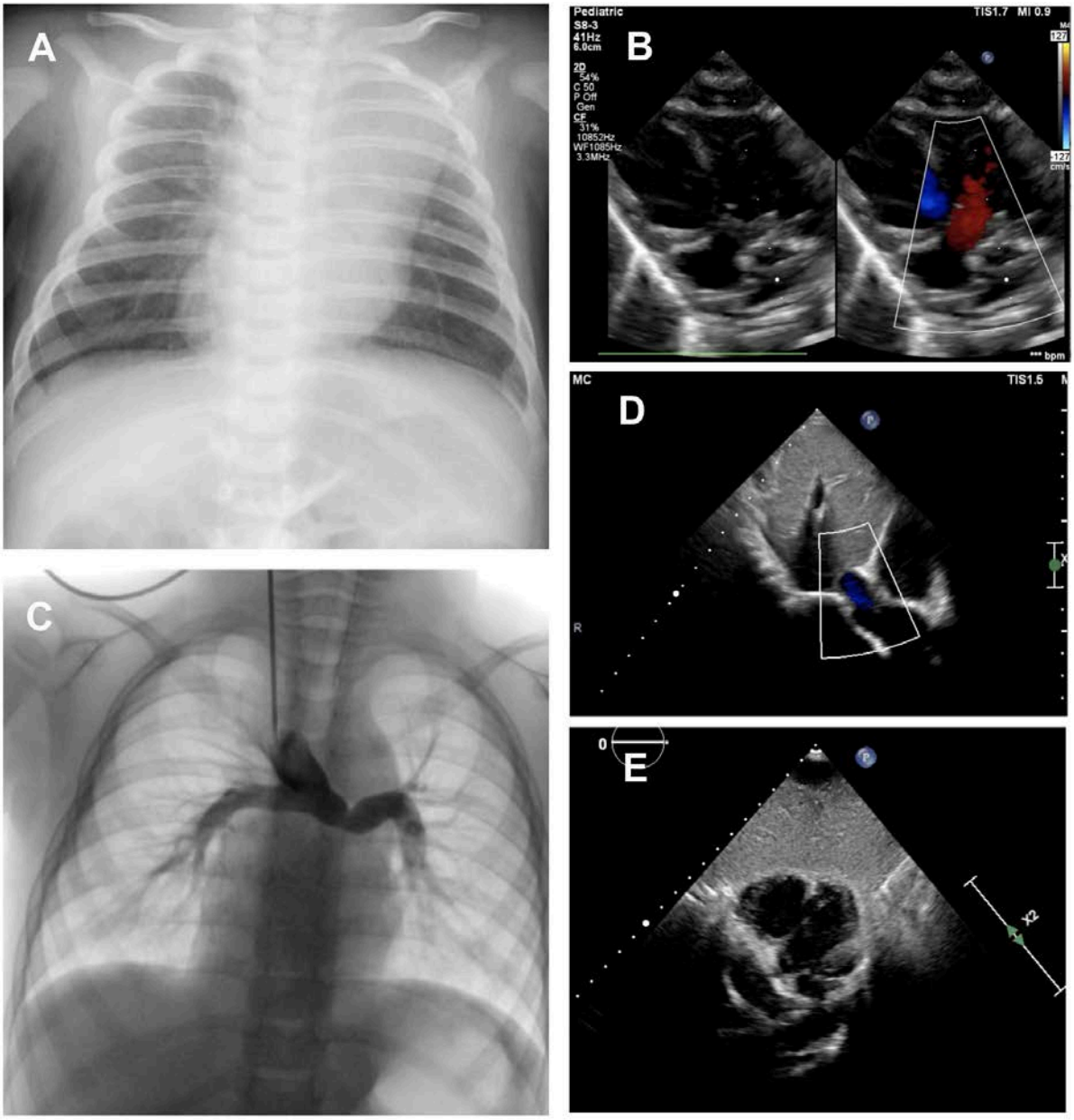

Figure 1. A. The chest X-ray (neonate period) — heart located medially, heart apex facing right. **B.** Echocardiography (neonate period) of the criss-cross heart morphology — cross-flow and abnormal AV connections — more in Supplementary materials, *Video S1*. **C.** Heart catheterization after bidirectional 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, atrio-ventricular, TCPC, total cavopulmonary connection