









Criss-cross heart, dextrocardia and transposition of the great arteries – comprehensive management of a complex cardiovascular defect and rare morphology

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Artykuł jest tłumaczeniem pracy: Haponiuk J, et al. Wada serca typu *criss-cross heart* współwystępująca z malpozycją wielkich naczyń i dekstrocardią – kompleksowe leczenie złożonej wrodzonej wady układu sercowo-naczyniowego o rzadkiej morfologii. *Folia Cardiol.* 2020; 15(1): 72–75.

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Abstract

The criss-cross heart congenital defect (CHD) concomitant with transposition of the great arteries has an incidence of less than 0.1% of all inherited heart anatomy abnormalities. It derives from the anomaly of cardiac rotation around its long axis after ventricular septation, and results in an atypical relation between the ventricles and the atria. Furthermore, such patients may also present with ventricular septal defect (VSD), large vessel malposition, double outflow and/or right ventricular hypoplasia (RV), pulmonary artery stenosis (PA), and tricuspid valve hypoplasia.

A 7-month-old patient after a transcatheter pulmonary balloon valvuloplasty and a surgical unilateral modified Blalock-Taussig (BT) shunt was admitted to the paediatric cardiac surgery department for further treatment.

CHD had been diagnosed prenatally and confirmed after birth. Echocardiographic study revealed dextrocardia and a criss-cross heart pathology with discordant atrioventricular (AV) and ventriculoarterial relations. Intracardiac pathology was concomitant with transposition of the great arteries [aorta originating from the RV, and PA from above the VSD], subvalvular and valvular pulmonary stenosis (gradient RV/PA approx. 90 mm Hg), and unobstructed BT shunt. Systemic cyanosis was not observed, and the child's mental and physical development was undisturbed. After diagnostic catheterisation, the patient was referred for further surgery. Using extracorporeal circulation, a bidirectional Glenn anastomosis was performed, and the BT shunt as well as the PA were closed and dissected. The postoperative course was uncomplicated.

More than 90% of criss-cross heart co-occur with levocardia and the concomitance with the AV-related transposition of the great arteries and dextrocardia, to the best of our knowledge, is the second described report in the literature. Thus, the diagnosis of a rare congenital heart defect should be always considered, especially in prenatal diagnostics, and precisely defined during qualification for further surgical treatment.

Key words: criss-cross heart, dextrocardia, transposition of the great arteries, congenital heart defects, paediatric cardiac surgery

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Introduction

The recent, global development of prenatal diagnostics has resulted in an increased number of live births of neonates with a congenital heart defect (CHD). Between 2010 and 2017, that number reached 9,410 per 10,000 births [1]. A criss-cross heart (CCH)-type congenital heart defect comorbid with atrioventricular (AV) cross-flow is an extremely rare and complex anatomical defect in which, during foetal development, the apex of the heart is displaced 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 [2, 3]. According to current data, the incidence of this defect is less than 0.1% of all CHDs, and does not exceed 8/1,000,000 live births [4, 5]. Criss-cross heart can take one of three forms: complete transposition of the great arteries (also known as dextro-transposition of the great arteries); congenitally corrected transposition of the great arteries (also known as levo-transposition of the great arteries); and functionally normal heart. Physiology and symptoms depend on the inflow tracts crossing presence and degree, on the morphology of the AV connection, and on comorbid heart defects [5].

CCH patients most commonly present with ventricular septal defects (VSDs), transposition of the great arteries, double outlet and/or hypoplastic right ventricle (RV), pulmonary artery (PA) stenosis, and tricuspid valve hypoplasia [4].

Therapeutic management of CCH depends on whether it is possible to preserve the function of both heart ventricles; however, physiological palliative pathway cardiac surgery treatment aimed at achieving a single functional systemic ventricle is the most common approach [6].

Case report

A 7-month-old female presented with a CHD in the form of CCH currently undergoing a multistage palliative treatment, following a single ventricle pathway physiology. The patient was born via vaginal delivery, achieving Apgar score 10 and uncomplicated adaptation. The CHD was diagnosed prenatally and confirmed after delivery. Percutaneous pulmonary valvuloplasty and right-side modified Blalock-Taussig systemic-to-pulmonary shunt (BT dex) were performed after birth, during the neonatal period.

Follow-up echocardiography confirmed a CCH-type CHD – cross-flow and abnormal AV connections (RV connected via tricuspid valve to the left atrium and LV connected via mitral valve to the right atrium). In addition, the examination also showed valvular and subvalvular PA stenosis (RV/PA gradient approx. 90 mm Hg), massive VSD,

transposition of the great arteries (aorta originating from RV and PA from over the VSD), and unobstructed BT dex shunt as shown in Figure 1A and 1B. Despite slight cyanosis, the infant showed no signs of dyspnoea, the psychomotor development was adequate. After diagnostic heart catheterisation, the patient was admitted to the further cardiac surgery treatment (Figure 1C). Bidirectional Glenn procedure (superior vena cava to right PA shunt) was performed under extracorporeal circulation. During this procedure, the BT dex shunt was removed and the PA trunk was closed. The perioperative and postoperative course was uncomplicated (Figure 1D).

Discussion

More than 90% of cases of CCH-type congenital defect co-occur with levocardia. Concomitance with AV-related transposition of the great arteries and dextrocardia, to the best of our knowledge, is the second time described, after Kasar et al., in the literature.

The choice of procedure for CCH depends on a thorough assessment of the individual morphology of the defect and its haemodynamics. The final decision is determined by the possibility of both ventricles preservation. Possible procedures include: the Rastelli procedure, arterial (Jatene) switch procedure, Glenn shunt, or central systemic-to-pulmonary shunt formation. However, analysis of several cases of adult patients with a CCH morphology CCH indicates that clinical manifestation depended predominantly on the underlying hemodynamic abnormalities and the results of surgical management rather than on congenital criss-cross anatomy [7].

The diagnosis of CCH should be suspected in echocardiographic study when the parallel arrangement of the AV valves and ventricular inlets cannot be achieved, and the two valves are not spontaneously visualised on apical 4 chamber view [5]. In addition, diagnostic procedures for CCH also describe the use of magnetic resonance imaging (MRI) to accurately assess morphology of the defect before surgery [8].

Conclusions

To conclude, the diagnosis of a rare congenital heart defect should be always considered, especially in prenatal diagnostics, and precisely defined during qualification for further surgical treatment.

Conflict(s) of interest

The authors declare no conflict of interest.

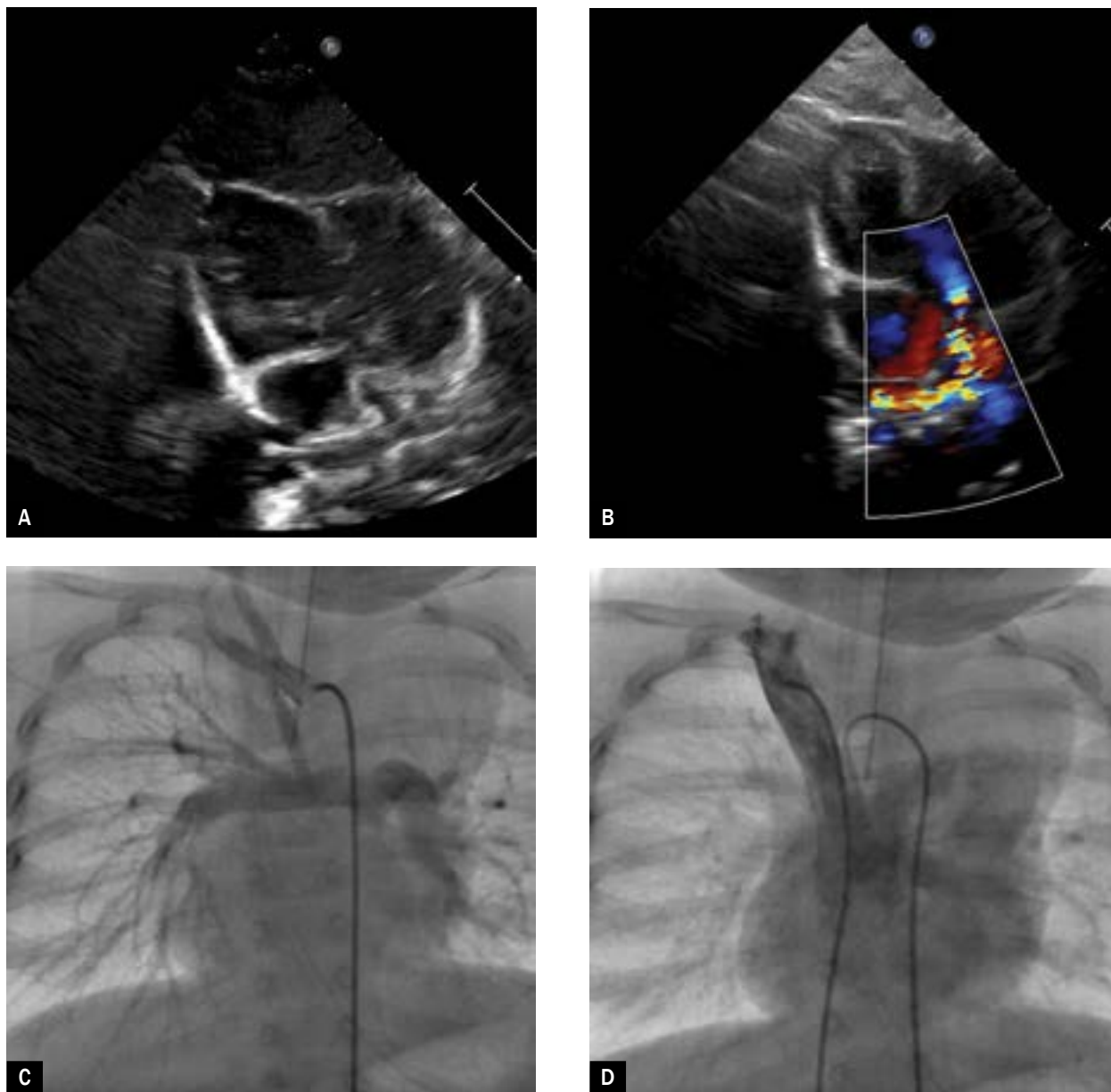


Figure 1A. Echocardiography, subcostal view (short axis): atrium with right atrial morphology connected via tricuspid valve to left ventricle and atrium with left atrial morphology connected via mitral valve to right ventricle; **B.** Echocardiography, apical 5-chamber view: pulmonary artery (PA) stenosis; **C.** Diagnostic cardiac catheterisation: inflow from right subclavian artery via unobstructed modified Blalock-Taussig systemic-to-pulmonary shunt to PA; **D.** Post-operative cardiac catheterisation: inflow from superior vena cava to pulmonary arteries, dextrocardia – apex of heart oriented right

References

1. Liu Y, Chen S, Zühlke L, et al. Global birth prevalence of congenital heart defects 1970-2017: updated systematic review and meta-analysis of 260 studies. *Int J Epidemiol.* 2019; 48(2): 455–463, doi: [10.1093/ije/dyz009](https://doi.org/10.1093/ije/dyz009), indexed in Pubmed: [30783674](https://pubmed.ncbi.nlm.nih.gov/30783674/).
2. Anderson RH, Shinebourne EA, Gerlis LM. Criss-cross atrioventricular relationships producing paradoxical atrioventricular concordance or discordance. Their significance to nomenclature of congenital heart disease. *Circulation.* 1974; 50(1): 176–180, doi: [10.1161/01.cir.50.1.176](https://doi.org/10.1161/01.cir.50.1.176), indexed in Pubmed: [4835263](https://pubmed.ncbi.nlm.nih.gov/4835263/).
3. Manuel D, Ghosh G, Joseph G, et al. Criss-cross heart: transthoracic echocardiographic features. *Indian Heart J.* 2018; 70(1): 71–74, doi: [10.1016/j.ihj.2017.03.008](https://doi.org/10.1016/j.ihj.2017.03.008), indexed in Pubmed: [29455791](https://pubmed.ncbi.nlm.nih.gov/29455791/).
4. Kasar T, Ayyıldız P, Türkvan A, et al. Criss-cross heart with dextrocardia and transposition of the great arteries: a rare pathology. *Turk*

- Kardiyol Dern Ars. 2016; 44(1): 91, doi: [10.5543/tkda.2015.00057](https://doi.org/10.5543/tkda.2015.00057), indexed in Pubmed: [26875142](https://pubmed.ncbi.nlm.nih.gov/26875142/).
5. Taksande AM. Echocardiographic recognition of a criss-cross heart with double outlet right ventricle. *Images Paediatr Cardiol*. 2013; 15(2): 3–7, indexed in Pubmed: [23847656](https://pubmed.ncbi.nlm.nih.gov/23847656/).
 6. Danielson GK, Tabry IF, Ritter DG, et al. Surgical repair of criss-cross heart with straddling atrioventricular valve. *J Thorac Cardiovasc Surg*. 1979; 77(6): 847–851, indexed in Pubmed: [374883](https://pubmed.ncbi.nlm.nih.gov/374883/).
 7. Hoffman P, Szymański P, Lubiszewska B, et al. Crisscross hearts in adults: echocardiographic evaluation and natural history. *J Am Soc Echocardiogr*. 2009; 22(2): 134–140, doi: [10.1016/j.echo.2008.10.004](https://doi.org/10.1016/j.echo.2008.10.004), indexed in Pubmed: [19056236](https://pubmed.ncbi.nlm.nih.gov/19056236/).
 8. Cantinotti M, Bell A, Hegde S, et al. A segmental approach to criss-cross heart by cardiac MRI. *Int J Cardiol*. 2007; 118(3): e103–e105, doi: [10.1016/j.ijcard.2007.01.063](https://doi.org/10.1016/j.ijcard.2007.01.063), indexed in Pubmed: [17400312](https://pubmed.ncbi.nlm.nih.gov/17400312/).