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Perioperative echocardiography in a newborn with severe tricuspid dysplasia operated on with CorMatrix tube reconstruction

Short title: Neonatal tricuspid valve dysplasia

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Severe tricuspid valve regurgitation (TVR) is a serious problem in prenatal period leading to cardiac compromise and non-immune hydrops fetalis requiring urgent surgical interventions in neonatal period [1–4]. In this case, TVR and functional pulmonary atresia was diagnosed prenatally

at midgestation (Supplementary material, *Figure S1*). Progressive fetal distress was an indication for immediate delivery and cardiac surgery.

A 1-day-old male newborn was delivered at 36 weeks of gestation by cesarean section due to fetal distress and weighing 2490 g. At birth the newborn had tachycardia 150/min, tachypnea, low arterial saturation (SaO_2 45-60%) and hypotension (mean arterial pressure 27 mm Hg). Respiratory support with FiO_2 1.0 improved SaO_2 up to 75%. Chest X ray showed cardiomegaly (cardio-thoracic ratio 1.0, Figure 1A). Transthoracic echocardiography (TTE) revealed irregular thickening of the TV leaflets, shortening of the chordae tendineae without septal displacement resulting in severe TVR (IV degree) (Figure 1B–C; Supplementary material, *Videos S1* and *S2*) with a systolic pressure gradient 33 mm Hg. Significant right atrium and right ventricle enlargement were found with dilated TV diameter (22 mm, z score +6.8). Right-to-left shunt through patent foramen ovale (Supplementary material, *Video S3*) and left-to-right shunt through the arterial duct were recorded. The left ventricle was diminished with normal contractility (EF 73%; Supplementary material, *Video S4*). Moderate pulmonary regurgitation was demonstrated (Figure 1D; Supplementary material, *Video S5*) without antegrade flow to the pulmonary trunk.

Initial therapies with respiratory support, inhaled nitric oxide (20 ppm), PGE1, milrinone, and pressors (dopamine and noradrenaline) infusions were insufficient. Blood gases showed progressive metabolic acidosis (lactic acid 12 mmol/l, n <1.6) with hypoxemia (pO_2 46 mm Hg in arterial blood; n >83 mm Hg) and coagulation disturbances (INR 4.0, n <1.3). Low body weight and type of TV anomaly restricted the possibility of surgical interventions.

The child was referred for cardiac surgery in cross clamp circulation. Intraoperative TV evaluation showed dysplastic leaflets and primary repair with valvuloplasty or annuloplasty was unfeasible. Therefore, valve reconstruction with Cormatrix tube was performed (Figure 1E; Supplementary material, *Videos S6* and *S7*). Postoperative transient complete atrio-ventricular block appeared which released with normal sinus rhythm and atrial premature beats. The clinical course was difficult with gradual improvement of cardio-pulmonary compromise and multi-organ dysfunction. TTE showed effective flow through the competent reconstructed TV. Holter electrocardiogram showed single atrial premature beats without episodes of tachy- or brady-arrhythmias. The child was discharged home. In the second month, a check-up revealed complete atrioventricular block, requiring implantation of an epicardial DDD pacemaker. In the first year of follow-up, the condition is good, without symptoms of cardiac compromise in multi-drug therapy,

including beta-blocker, angiotensin-converting enzyme inhibitor and diuretic. TTE showed right atrial enlargement, preserved TV function (mean pressure gradient 4 mm Hg) with normal myocardial contractility.

This is the first case in the literature describing surgical TV reconstruction with CorMatrix tube in a critically ill newborn. We admit, that this is a palliative procedure for newborns with heart failure and severe TVR, but providing spectacular improvement of general condition. In follow-up progressive tricuspid stenosis and regurgitation are expected which may require further surgical interventions including heart transplant [5].

Supplementary material

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

Article information

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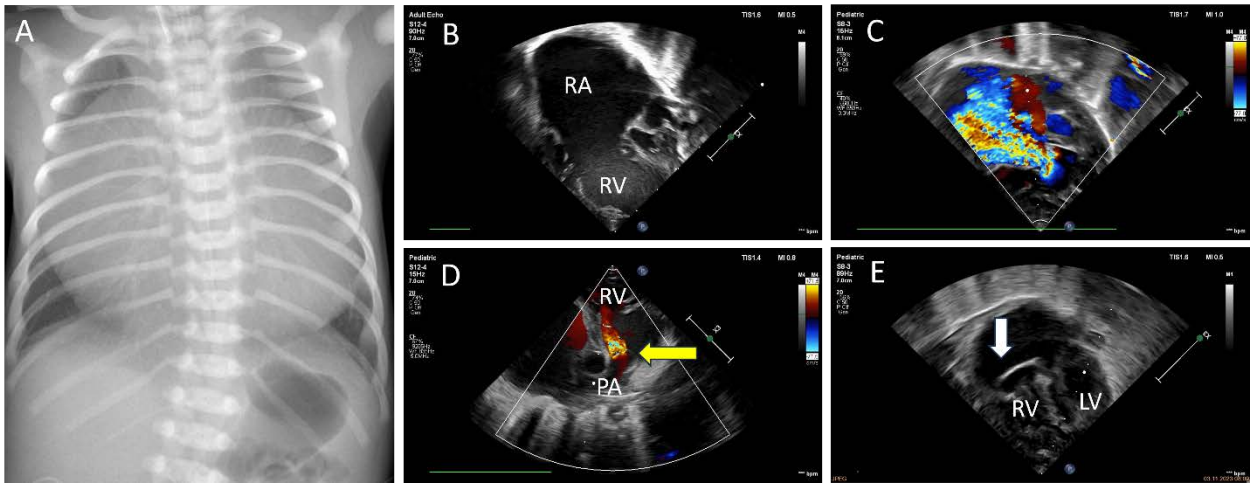

Figure 1. Severe tricuspid valve regurgitation in a 1 day-old-newborn operated on with tricuspid valve reconstruction using CorMatrix tube. **A.** Chest X ray in anteroposterior view. Cardiomegaly with pulmonary compression. **B.** Transthoracic echocardiography (TTE). 2DE in apical 4-chamber view showing severely dilated right atrium and right ventricle with left heart compression. **C.** TTE. 4-chamber view with color Doppler flow showing severe tricuspid valve regurgitation with right heart volume overload. **D.** TTE. Short parasternal view with color Doppler flow showing pulmonary regurgitation. **E.** TTE. Apical 4-chamber view in 2DE showing reconstructed tricuspid valve in systole (white arrow)

Abbreviations: LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle