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

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Severe tricuspid valve regurgitation (TVR) is a serious problem in the prenatal period, leading to cardiac compromise and non-immune hydrops fetalis, requiring urgent surgical interventions in the neonatal period [1–4]. In this case, TVR and functional pulmonary atresia were diagnosed prenatally at midgestation (Supplementary material, *Figure S1*). Progressive fetal distress was an indication for immediate delivery and cardiac surgery.

A male newborn weighing 2490 g was delivered at 36 weeks of gestation by cesarean section. 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 tricuspid valve (TV) leaflets and shortening of the chordae tendineae without septal displacement, which resulted in severe TVR (IV degree) (Figure 1B–C;

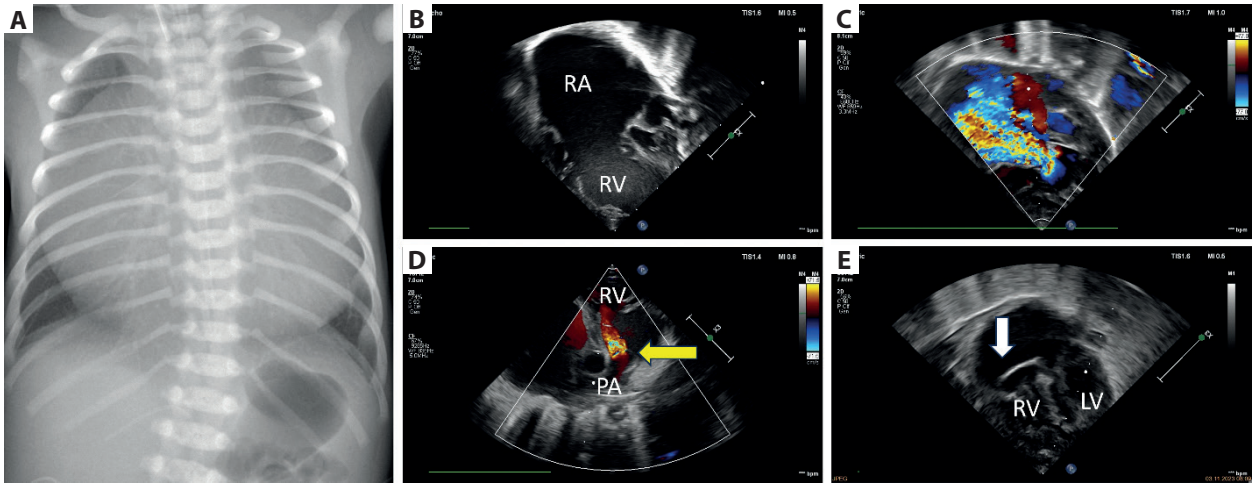

Figure 1. Severe tricuspid valve regurgitation in a newborn boy operated on with tricuspid valve reconstruction using a CorMatrix tube. **A.** Chest X-ray in the anteroposterior view. Cardiomegaly with pulmonary compression. **B.** Transthoracic echocardiography (TTE). 2DE in the apical 4-chamber view showing a severely dilated right atrium and the 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 the reconstructed tricuspid valve in systole (white arrow)

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

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 the 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 (ejection fraction 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 pressor (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 possible 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 a 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 visualised single atrial premature beats without episodes of tachy- or brady- arrhythmias. The child was discharged home. In the following month, a check-up showed a complete atrioventricular block, requiring implantation of an epicardial DDD pacemaker. In the first year of follow-up, the patient condition was good, without symptoms of cardiac compromise, and he was kept on multi-drug therapy, including beta-blockers, angiotensin-converting enzyme inhibitors and diuretics. 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 a CorMatrix tube in a critically ill newborn. We admit that this is a palliative procedure for newborns with heart failure and severe TVR, but it can provide spectacular improvement in general condition. In follow-up, progressive tricuspid stenosis and regurgitation are expected, which may require further surgical interventions including heart transplantion [5].

Supplementary material

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

Article information

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