Case report

# Surgical treatment of tricuspid valve dysplasia in the neonatal period

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**Tricuspid valve dysplasia, other than Ebstein's anomaly, is a very rare congenital heart defect. During the prenatal and/or the neonatal periods the clinical picture is very critical. We here report on a newborn infant with severe tricuspid valve dysplasia and 4/4 tricuspid regurgitation, giant right atriomegaly, functional pulmonary atresia with ductal-dependent pulmonary blood flow. The child was successfully submitted to implantation of a 15 mm pulmonary stentless heterograft valve using the top-hat technique.**

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## Introduction

Dysplasia of the tricuspid valve responsible for severe regurgitation is a rare occurrence and a surgical challenge. Different surgical options have been proposed for children and adult patients[1-5]. However, in case of newborns, no successful treatment has been described.

We here report on a newborn infant who underwent pulmonary stentless heterograft valve replacement in the tricuspid position (top-hat procedure).

## Case report

Our patient had a prenatal diagnosis of pulmonary atresia with an intact interventricular septum at 20 weeks of gestation. He had severe cardiomegaly with no signs of overt heart failure.

At birth, he required intubation and mechanical ventilation for cardiopulmonary failure. The cardiothoracic ratio was 0.9. Preoperative echocardiography showed severe tricuspid valve dysplasia and 4/4 tricuspid regurgitation (Fig. 1), giant right atriomegaly, functional pulmonary atresia with ductal-dependent pulmonary blood flow. The ductus arteriosus was maintained patent by means of prostaglandin $E_1$ infusion. Attempts to wean him from drugs and ventilation after 10 days were unsuccessful. At 12 days of age, he underwent surgery including reduction atrioplasty and tricuspid valve replacement. No major complications were observed during the postoperative course.

At discharge, the cardiothoracic ratio was 0.6. Echocardiography (Fig. 2) showed that there was neither tricuspid regurgitation nor stenosis and that the biventricular function was satisfactory. Neurologic assessment was normal.

He was discharged on day 30 with phenobarbital 15 mg twice daily, furosemide 3 mg twice daily, and aspirin 25 mg/die. After 3 months of follow-up he is clinically well.

**Operative technique.** The surgical procedure included standard cardiopulmonary techniques with moderate systemic hypothermia and anterograde cold blood cardioplegia for myocardial protection.

To create more space so as to allow the lungs to expand postoperatively and to create a more efficient right atrium, a sizable segment of the right atrial free wall was resected using an elliptical incision. The tricuspid valve was trimmed. On the basis of echocardiographic analysis, a 15 mm pulmonary stentless heterograft valve withdrawn from a tube Tissumed was chosen.

Using the top-hat technique, the valve was sutured to the annulus and to the valve remnants, taking care to avoid injury to the conduction system.

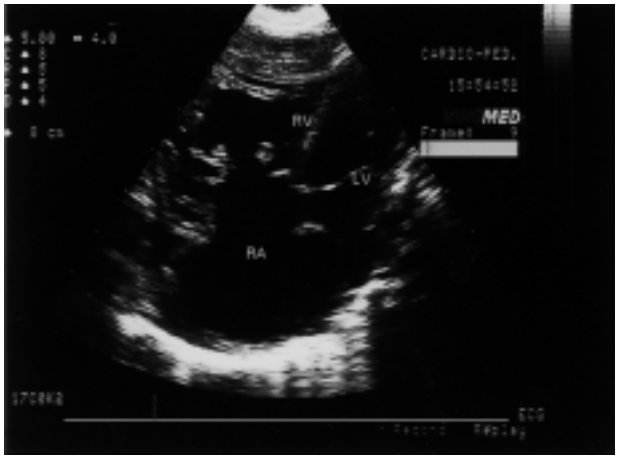


**Figure 1.** *Apical 4-chamber view in systole showing severe tricuspid dysplasia, no coaptation of the tricuspid leaflets and huge right atriomegaly. LV = left ventricle; RA = right atrium; RV = right ventricle.*

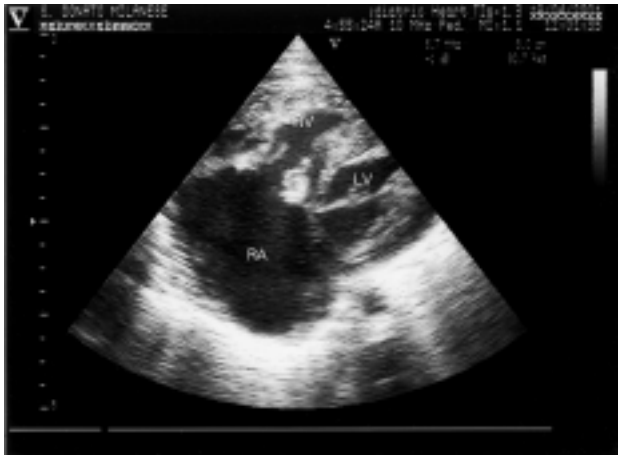


**Figure 2.** *Apical 4-chamber view 1 week after surgery showing the opening of the leaflets of the heterograft valve in the tricuspid position. Right atrial dilation persists. LV = left ventricle; RA = right atrium; RV = right ventricle.*

In accordance with the top-hat technique, the semilunar stentless valve was implanted at the level of the tricuspid annulus in a reverse position. In fact, the opening side was towards the right atrium and not towards the right ventricle as would have been with the pulmonary semilunar valve (Fig. 3).

The patient was weaned off cardiopulmonary bypass in sinus rhythm and excellent hemodynamics. The bypass time was 130 min. The aortic cross-clamp time was 36 min.

## Discussion

Tricuspid valve dysplasia, other than Ebstein's anomaly, is a very rare congenital heart defect.

The essential morphologic defect involves a mucoid dysplasia of the leaflets associated with shortened or absent chordae tendinae. These anomalies are responsible for severe tricuspid regurgitation and functional pulmonary atresia. During the prenatal and/or the neonatal periods the clinical picture is very critical with signs and symptoms of right heart failure and respiratory distress.

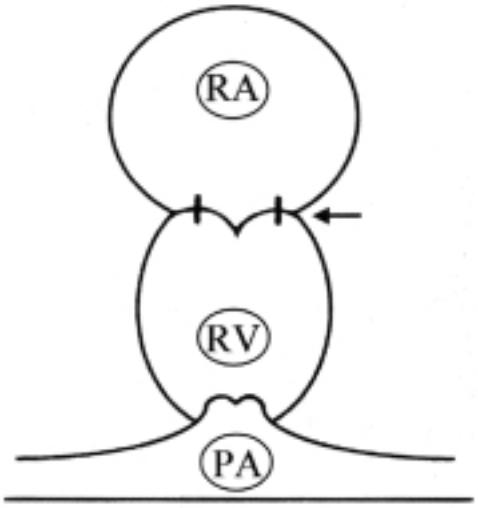


**Figure 3.** *Schematic view of the top-hat technique. See text for details. PA = pulmonary artery; RA = right atrium; RV = right ventricle.*

The prognosis depends on the severity of the valvular dysfunction.

Different techniques have been reported for children and adults surviving the neonatal period or in patients with mild tricuspid dysplasia.

Replacement of the malformed tricuspid valve with mechanical valves has been reported[4]. Chordal replacement or augmentation with an expanded polyfluoroethylene suture was reported by Reddy et al.[5] and by Katogi et al.[2]. Miyagishima et al.[1] proposed the use of a cryopreserved mitral homograft for drug abusers with tricuspid endocarditis. Kumar et al.[3] reported the use of an unstented semilunar homograft to replace the tricuspid valve in 3 children with Ebstein's malformation.

However, such approaches have never been described in newborns with tricuspid valve dysplasia.

Furthermore, it is not possible to find a mitral valve homograft or artificial chordae tendinae suitable for use in newborns. We decided not to use an artificial valve due to the technical problems related to the reduced space in the neonatal heart. Finally, lifelong anticoagulation is required, and this may be a relevant limitation in a growing child.

In our patient, we chose a pulmonary stentless valve using the top-hat technique. In the case reported, this technique has provided a competent valve with a central laminar flow with no stenosis. The short-term follow-up confirms the satisfactory outcome.

However, it is highly probable that in the future due to somatic growth the infant will develop a stenosis at the level of the tricuspid prosthesis. Hence, it should be

possible to implant a larger prosthesis and eventually a definitive mechanical prosthesis.

In conclusion, we show that the top-hat technique can be performed successfully in newborns with severe tricuspid dysplasia.

## References

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