

POSTOPERATIVE MANAGEMENT OF SEVERE RIGHT VENTRICULAR DYSFUNCTION FOLLOWING ONE-AND-A-HALF VENTRICULAR REPAIR IN A CHILD WITH DOWN SYNDROME AND ATRIOVENTRICULAR CANAL DEFECT WITH EBSTEIN'S ANOMALY: A CASE REPORT**Dhananj Shivganesh B. R.*, Ankur Dogra, Kanika Prashar**

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INTRODUCTION

Down syndrome is strongly associated with atrioventricular canal defects (AVCD), with up to 40% of affected children presenting with this lesion. The coexistence of Ebstein's anomaly is rare but significantly complicates surgical repair due to the high risk of postoperative RV dysfunction.^[2,3] One-and-a-half ventricular repair (1.5VR), achieved by combining intracardiac repair with a BD Glenn, provides an effective strategy for unloading the dysfunctional RV.^[1-4] However, postoperative management remains challenging and is often the key determinant of outcome. Here, we present a case where vigilant intensive care interventions, including inotropic support, fluid optimisation, and airway protection, resulted in a favourable recovery.

CASE PRESENTATION

An 8-year-old boy with Down syndrome presented with complete AVCD and significant displacement of the septal tricuspid leaflet, consistent with Ebstein's anomaly. He underwent intracardiac repair.

Intraoperative findings and procedure: After completion of cardiopulmonary bypass, severe RV dysfunction was evident. A BD Glenn was constructed, converting the repair to a one-and-a-half ventricular strategy.^[2-4]

Immediate postoperative course: The patient was transferred to the ICU on multiple inotropes: Milrinone 0.3 µg/kg/min, Norepinephrine 0.1 µg/kg/min, Adrenaline 0.1 µg/kg/min, Vasopressin 10 mU/kg/hr.

Echocardiography showed severe RV and moderate LV dysfunction. Clinically, he developed marked facial and upper limb oedema, suggestive of venous congestion.

POD 1: The patient was extubated but developed inspiratory stridor. Serial arterial blood gases demonstrated hypercapnia. He was electively reintubated to prevent worsening respiratory acidosis.

Subsequent management: A continuous furosemide infusion was initiated. Over 48 hours, a negative fluid balance of 1000 ml was achieved, resulting in resolution of oedema. The patient was then extubated successfully. With gradual improvement in ventricular function, inotropes were weaned off.

Outcome: The child recovered well and was discharged from ICU in stable condition.

DISCUSSION

This case highlights the complexity of postoperative management following 1.5VR in patients with AVCD and Ebstein's anomaly.

1. Inotropic and vasoactive support: Children with postoperative RV dysfunction benefit from multimodal inotropic support. Milrinone has been shown to reduce pulmonary vascular resistance and improve RV performance, making it a cornerstone in congenital cardiac postoperative care.^[5] Combination therapy with norepinephrine, adrenaline, and vasopressin provided additional circulatory support in this case, consistent with current paediatric cardiac ICU practice.^[6,7]

2. Fluid and diuretic management: In Glenn physiology, excessive preload or venous congestion can compromise both systemic and pulmonary circulations.^[1,2] In our case, facial and upper limb oedema, along with stridor, were manifestations of venous congestion. A continuous furosemide infusion facilitated a safe negative fluid balance and rapid clinical improvement.

3. Airway management: Airway oedema after Glenn procedures may be exacerbated by venous hypertension. Although early extubation is desirable in congenital cardiac surgery, timely recognition of stridor and CO₂ retention prompted elective reintubation in our patient, preventing severe respiratory compromise.

Previous series have emphasised the importance of meticulous postoperative monitoring in children undergoing 1.5VR.^[1-4] Our case reinforces that tailored ICU management is as critical as surgical technique in determining outcomes.

CONCLUSION

One-and-a-half ventricular repair offers a valuable surgical alternative in children with AVCD and Ebstein's anomaly complicated by RV dysfunction. However, the postoperative course can be challenging. This case underscores the pivotal role of judicious inotrope use, aggressive fluid management, and proactive airway protection in achieving a favourable recovery.

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