

Newborn with Tricuspid Valve Dysplasia and Pulmonary Atresia : A Diagnostic and Therapeutic challenge : A Case Report and Review of the Literature

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Received date: July 21, 2025; **Accepted date:** August 05, 2025; **Published date:** August 19, 2025

Citation: Z.El Abasse, (2025), Newborn with Tricuspid Valve Dysplasia and Pulmonary Atresia : A Diagnostic and Therapeutic challenge : A Case Report and Review of the Literature, *J Clinical Cardiology and Cardiovascular Interventions*, 8(13); DOI:10.31579/2641-0419/500

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Abstract

A 5-day-old newborn, born at 36 weeks of gestation to a mother with preeclampsia, presented with poor initial adaptation, including no movement, no cry, and a heart rate below 100 bpm. Echocardiography revealed a major defect in the atrioventricular valve with torrential regurgitation, tricuspid atresia with absence of flow through the right ventricular outflow tract, and a well-developed pulmonary artery. The newborn was started on prostaglandin therapy, which allowed for pulmonary perfusion with oxygen saturation between 85-90%. A multidisciplinary team discussed three possible management options: dilation of the right ventricular outflow tract, Starnes procedure with Blalock shunt, or discontinuation of prostaglandin therapy.

Kew Words: temporary cardiac pacing; percutaneous coronary intervention

Case Presentation

- *Gestational Age:* 36 weeks
- *Maternal History:* Preeclampsia
- *Newborn Presentation:* Poor initial adaptation, no movement, no cry, heart rate below 100 bpm
- *Echocardiography Findings:*
 - Major defect in atrioventricular valve with torrential regurgitation
 - Tricuspid atresia with absence of flow through the right ventricular outflow tract
 - Well-developed pulmonary artery
 - Large atrial septal defect (ASD) with right-to-left shunt
 - Large patent ductus arteriosus (PDA) with left-to-right shunt
 - No left-sided anomalies
- *Prostaglandin Therapy:*

- Allowed for pulmonary perfusion with oxygen saturation between 85-90%

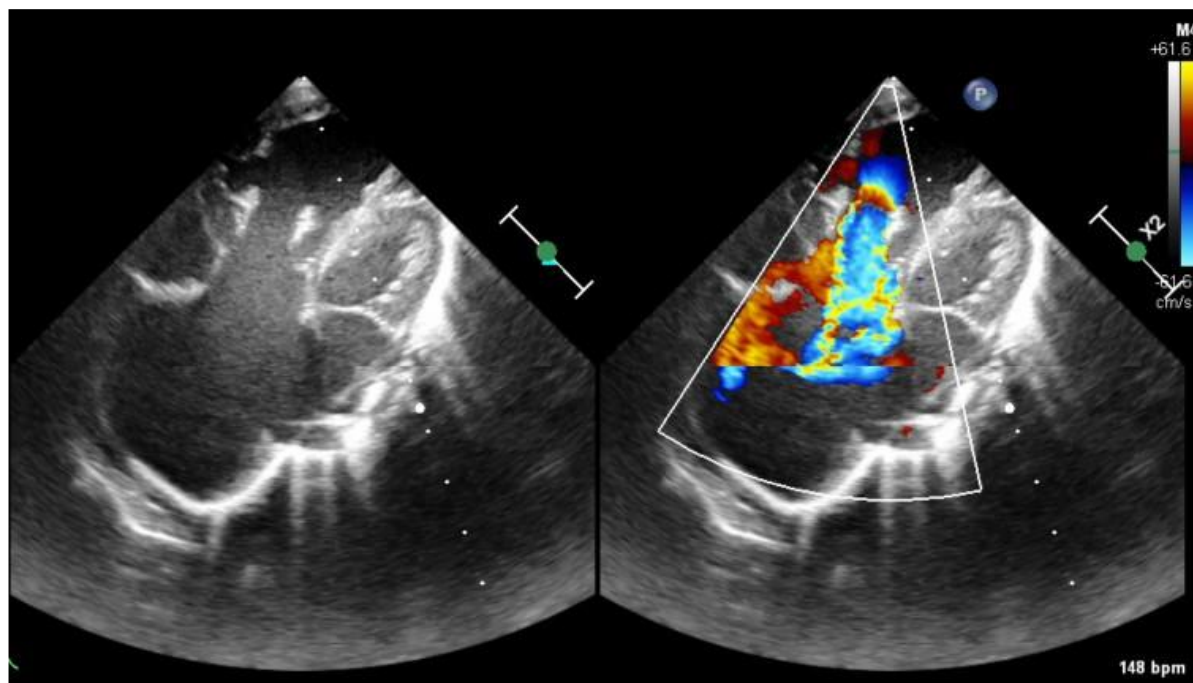
- *Physical Examination:*

- Hepatomegaly without signs of hypoperfusion

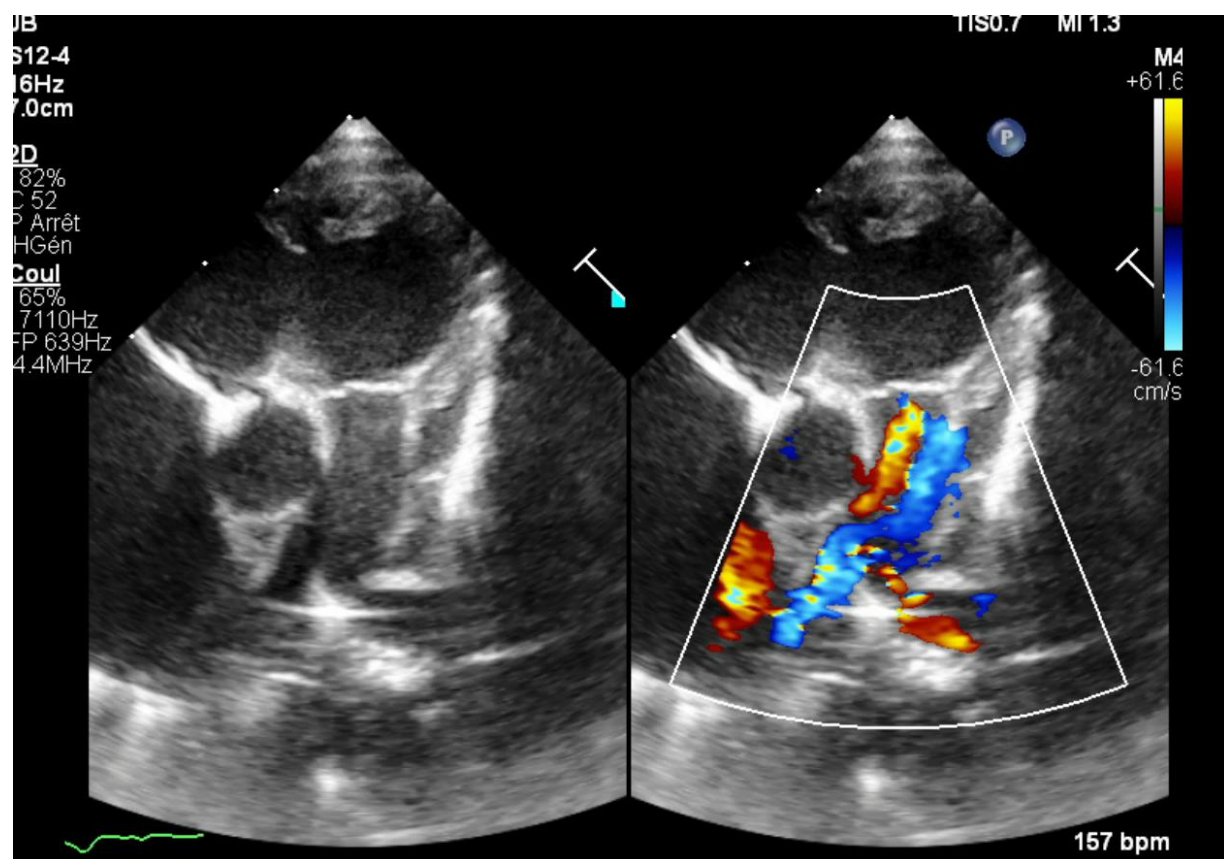
Multidisciplinary Discussion

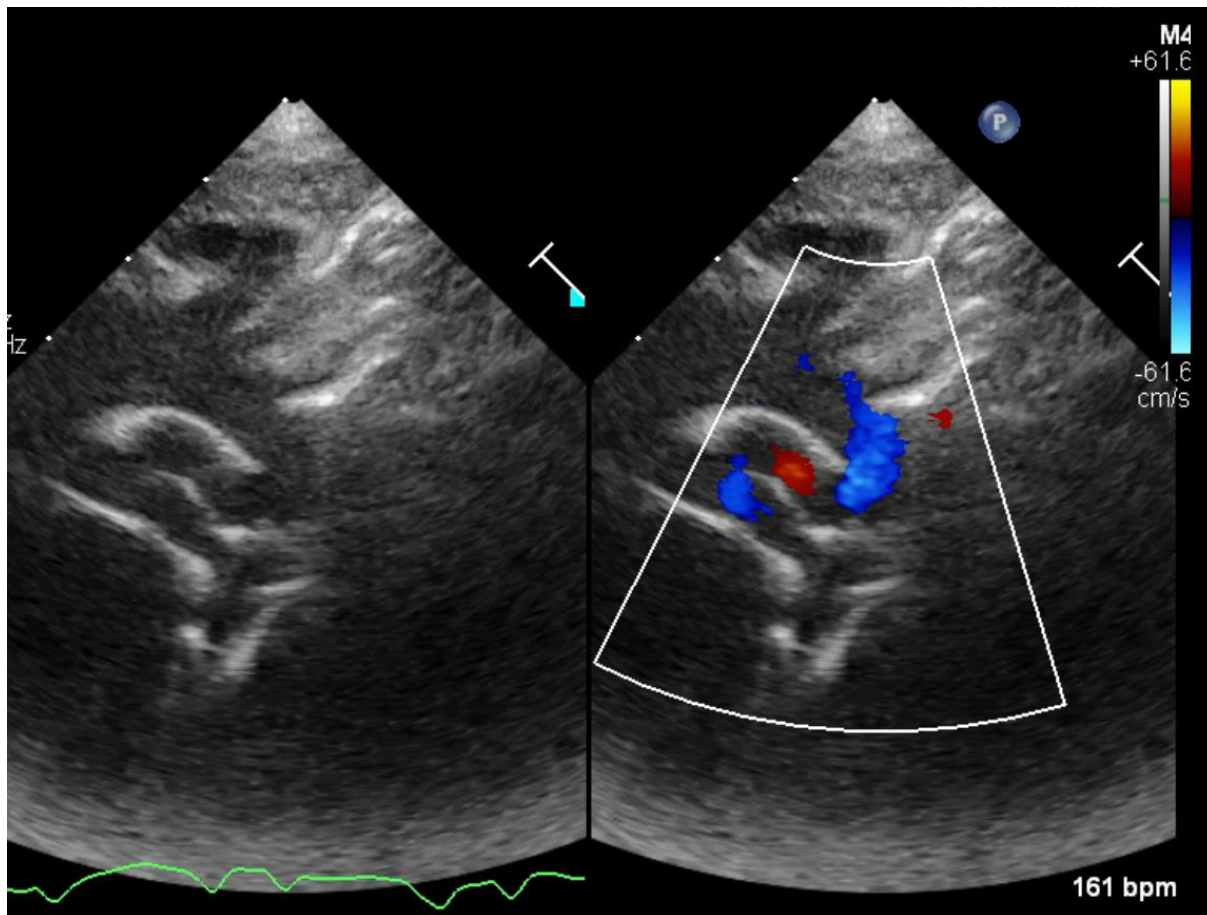
A multidisciplinary team discussed three possible management options for the newborn:

1. *Dilation of the Right Ventricular Outflow Tract:* This procedure aims to improve blood flow to the lungs by dilating the narrowed or blocked right ventricular outflow tract.
2. *Starnes Procedure with Blalock Shunt:* This surgical procedure involves creating a shunt between the subclavian artery and the pulmonary artery to increase blood flow to the lungs.
3. *Discontinuation of Prostaglandin Therapy:* This option would involve stopping the prostaglandin therapy and monitoring the newborn's condition.



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Discussion

Tricuspid valve dysplasia and right ventricular outflow tract obstruction are complex congenital heart defects that require careful management. The use of prostaglandin therapy has improved outcomes for newborns with ductal-dependent lesions. Early diagnosis and surgical intervention are critical in improving survival rates and reducing morbidity in newborns with tricuspid valve dysplasia.

Recent studies have highlighted the importance of multidisciplinary care in managing complex congenital heart defects. A study published in the *Journal of the American College of Cardiology* found that a multidisciplinary approach improved outcomes for newborns with congenital heart disease (1).

Another study published in the *Journal of Thoracic and Cardiovascular Surgery* found that surgical intervention improved survival rates and reduced morbidity in newborns with tricuspid valve dysplasia (2).

A study published in the *European Journal of Cardio-Thoracic Surgery* found that the Starnes procedure with Blalock shunt is an effective surgical treatment for tricuspid valve dysplasia, with good short-term outcomes (3).

A review article published in the *Journal of Cardiovascular Medicine and Surgery* discussed the various surgical options for tricuspid valve dysplasia, including the Starnes procedure and the Fontan procedure (4).

A case report published in the *Journal of Pediatrics and Child Health* highlighted the importance of early diagnosis and surgical intervention in improving outcomes for newborns with tricuspid valve dysplasia (5).

Conclusion

This case report highlights the complexity of managing newborns with tricuspid valve dysplasia and right ventricular outflow tract obstruction. A multidisciplinary team approach is essential in determining the optimal management strategy for these newborns.

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DOI:[10.31579/2641-0419/500](https://doi.org/10.31579/2641-0419/500)

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