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PREGNANCY AT TERM IN EBSTEIN ANOMALY WITH SEVERE TRICUSPID REGURGITATION AND PULMONARY HYPERTENSION: SUCCESSFUL MULTIDISCIPLINARY MANAGEMENT AND ASSISTED VAGINAL DELIVERY

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ABSTRACT

Background: Ebstein anomaly is an uncommon congenital tricuspid valve malformation with a highly variable clinical spectrum. Pregnancy may increase right-sided volume load and aggravate tricuspid regurgitation, heart failure and arrhythmia. **Case presentation**: A 35-year-old gravida 2, para 1 woman with known Ebstein anomaly presented at 38 weeks and 6 days. She remained clinically compensated under obstetric and cardiology supervision and received metoprolol and digoxin. Echocardiography demonstrated right atrial and right ventricular dilatation, severe tricuspid regurgitation and reported severe pulmonary artery hypertension, with preserved left ventricular systolic function (ejection fraction 55–60%). Following multidisciplinary assessment, vaginal delivery was planned. Forceps-assisted vaginal delivery was performed to shorten the second stage. A male infant weighing 2.0 kg was delivered with satisfactory immediate adaptation. The mother underwent 24-hour intensive-care observation and had an uncomplicated postpartum course; mother and infant were discharged in stable condition on the tenth postnatal day. **Conclusion:** Favourable pregnancy outcomes may occur in selected women with Ebstein anomaly despite substantial echocardiographic abnormalities when clinical status remains stable and care is coordinated by a multidisciplinary team.

KEYWORDS

Ebstein anomaly; pregnancy; congenital heart disease; tricuspid regurgitation; pulmonary hypertension; assisted vaginal delivery; multidisciplinary care.

INTRODUCTION

Ebstein anomaly is a rare congenital malformation of the tricuspid valve characterized by apical displacement of the septal and posterior tricuspid leaflets, atrialization of a portion of the right ventricle and variable tricuspid regurgitation. Its clinical spectrum ranges from minimally symptomatic disease to severe right-sided dysfunction, cyanosis, arrhythmia and heart failure.

Pregnancy produces major cardiovascular adaptations, including increased plasma volume and cardiac output. In women with Ebstein anomaly, these changes may increase right-sided volume loading and exacerbate tricuspid regurgitation or precipitate heart failure and arrhythmia. Contemporary prospective evidence from the Registry of Pregnancy and Cardiac Disease (ROPAC) demonstrates that most women tolerate pregnancy, but maternal morbidity remains clinically important when additional risk factors are present.

We report a woman with Ebstein anomaly who had severe tricuspid regurgitation and reported severe pulmonary artery hypertension but remained clinically compensated, reached 38+6 weeks and underwent successful assisted vaginal delivery. The case illustrates the importance of integrating clinical functional status with echocardiographic findings and individualizing delivery planning.

Case Presentation

Patient history

A 35-year-old gravida 2, para 1 woman with known Ebstein anomaly presented to the Department of Obstetrics and Gynecology, B.K.L. Walawalkar Rural Hospital, Dervan, Ratnagiri, Maharashtra, at 38 weeks and 6 days of gestation for planned institutional delivery. She had been under obstetric and cardiology supervision throughout pregnancy and remained clinically compensated. Metoprolol and digoxin were continued under specialist supervision.

There was no documented clinical evidence during pregnancy of overt cardiac failure, significant arrhythmia or progressive clinical deterioration. Pregnancy was therefore continued with multidisciplinary surveillance.

Clinical and cardiac assessment

At admission, the patient was clinically stable without evidence of acute cardiac decompensation or respiratory distress. Obstetric examination showed a singleton cephalic pregnancy with reassuring fetal heart rate.

Transthoracic echocardiography demonstrated right atrial and right ventricular dilatation, severe tricuspid regurgitation and severe pulmonary artery hypertension. Left ventricular systolic function remained preserved, with an ejection fraction of 55–60%.

Multidisciplinary management and delivery

- The case was reviewed by the obstetric, cardiology, anaesthesia, intensive-care and neonatal teams.
- Because the patient remained clinically compensated, vaginal delivery was selected. Close maternal
- haemodynamic and fetal surveillance was maintained. Forceps-assisted vaginal delivery was undertaken to shorten the second stage and limit prolonged maternal expulsive effort.
- The delivery was completed without documented intrapartum cardiac or major obstetric complication.

Maternal and neonatal outcome

A male infant weighing 2.0 kg was delivered with satisfactory immediate neonatal adaptation. The mother was observed in intensive care for 24 hours. No postpartum cardiac decompensation or major complication was documented. Mother and newborn remained stable and were discharged in good condition on the tenth postnatal day.

DISCUSSION

The ROPAC study of 81 pregnancies in women with Ebstein anomaly reported major adverse cardiac events in 9.9% of pregnancies, predominantly heart failure, with no maternal deaths. Pre-pregnancy signs of heart failure were the principal predictor of maternal cardiac events. These findings support individualized assessment rather than treating all women with Ebstein anomaly as having the same pregnancy risk.

Our patient represents a clinically important phenotype: substantial right-sided echocardiographic abnormalities were present, yet she remained clinically compensated and completed pregnancy to term.

The favourable outcome should therefore be interpreted in the context of preserved LV systolic function and absence of documented overt heart failure or significant clinical deterioration, rather than as evidence that severe echocardiographic disease is intrinsically low risk.

Vaginal delivery is appropriate for many clinically stable women with Ebstein anomaly. The ROPAC cohort included vaginal deliveries in more than half of pregnancies, although caesarean delivery remained common. The choice of delivery mode should be individualized according to maternal functional status, haemodynamic risk and obstetric circumstances.

In the present case, assisted vaginal delivery was selected to shorten the second stage. This illustrates a practical principle: when vaginal delivery is appropriate, the conduct of labour can be individualized to reduce avoidable cardiovascular stress while maintaining maternal and fetal safety.

Pulmonary hypertension warrants particular caution. The available case record describes severe pulmonary artery hypertension but does not provide a numerical estimated pressure or invasive haemodynamic confirmation. The qualitative label should therefore be interpreted cautiously and is explicitly acknowledged as a limitation rather than overstated.

The postpartum period is important because rapid haemodynamic shifts after delivery may precipitate cardiovascular deterioration. Twenty-four-hour intensive-care observation provided close surveillance during this vulnerable period.

From a rural tertiary-care perspective, the case demonstrates the value of coordinated obstetric, cardiac, anaesthetic, intensive-care and neonatal support in achieving a favourable outcome in a woman with significant congenital heart disease.

What this case adds

The distinctive clinical message is not simply that pregnancy is possible in Ebstein anomaly. It is that substantial echocardiographic right-sided disease can coexist with clinical compensation, allowing term pregnancy and individualized assisted vaginal delivery when multidisciplinary surveillance is available.

STRENGTHS AND LIMITATIONS

The strength of this report is the clinically relevant combination of Ebstein anomaly, substantial tricuspid regurgitation, reported pulmonary hypertension, term pregnancy and successful assisted vaginal delivery under multidisciplinary care.

The principal limitations are those inherent to a single case. The available record does not provide complete serial quantitative echo-cardiographic measurements, detailed right-ventricular functional indices, exact pulmonary artery pressure values, or a complete longitudinal functional-class assessment. The case should therefore be viewed as an illustrative clinical experience rather than evidence of safety for pregnancy in severe Ebstein anomaly.

Clinical Learning Points

- Ebstein anomaly has a heterogeneous pregnancy risk and should not be treated as a uniform clinical entity.
- Pre-pregnancy or antenatal evidence of heart failure is an important marker of increased maternal risk.
- Significant echocardiographic abnormalities should be interpreted alongside symptoms, functional status and ventricular function.
- Vaginal delivery is appropriate for many clinically stable women; assisted vaginal delivery may be considered when shortening the second stage is clinically desirable.
- Multidisciplinary antenatal, intrapartum and postpartum surveillance is central to safe management.

CONCLUSION

This case demonstrates that a favourable term pregnancy and assisted vaginal delivery can be achieved in a selected woman with Ebstein anomaly despite substantial tricuspid regurgitation and reported pulmonary hypertension when clinical status remains compensated and multidisciplinary care is available. The key lesson is individualized risk stratification, planned delivery and close postpartum surveillance. The case should not be interpreted as evidence that severe Ebstein anomaly or pulmonary hypertension is intrinsically low risk.

Declarations

- **Ethics approval:** Institutional ethical approval was obtained for publication of the case.
- **Consent for publication:** Written informed consent was obtained from the patient for publication of anonymized clinical information and images.
- **Conflict of interest:** None declared.
- **Funding:** None declared.
- **Authors' contributions:** All authors contributed to clinical management, interpretation of the case, manuscript preparation and approval of the final manuscript.

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