RESEARCH PAPER

Transcending the Natural History: A Rare Case of Successful Pregnancy in a Woman with Unrepaired Truncus Arteriosus Type I

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Abstract

Background

Pregnancy in women with uncorrected truncus arteriosus Type I complicated by Eisenmenger physiology is associated with extremely high maternal mortality and is generally contraindicated. Survival into reproductive age without surgical correction is uncommon, and only a handful of pregnancies in women with unrepaired truncus arteriosus have been reported in the literature.

Case Presentation

We report a case of successful delivery in a 27-year-old primigravida with uncorrected truncus arteriosus complicated by severe pulmonary arterial hypertension and Eisenmenger physiology. Despite counselling in early pregnancy regarding the extremely high maternal risk, the patient opted to continue the pregnancy. She presented at 34 weeks of gestation with severe preeclampsia and profound hypoxemia, with oxygen saturation as low as 59% on room air. Following stabilization with multidisciplinary care, elective preterm cesarean section was performed under epidural anesthesia. Postoperatively, the patient required intensive care management with non-invasive ventilation and oxygen support. She was discharged in stable condition on postoperative day 8 with advice regarding long-term home oxygen therapy and close cardiology follow-up.

Conclusion

This case highlights the importance of coordinated multidisciplinary management in an exceptionally high-risk pregnancy complicated by uncorrected truncus arteriosus and Eisenmenger physiology. Favorable maternal and neonatal outcomes, though rare, may be achievable with meticulous perioperative care, intensive monitoring, and timely intervention.

Keywords: Truncus arteriosus, Eisenmenger syndrome, Pulmonary arterial hypertension, Preeclampsia

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Introduction

Truncus arteriosus is a rare cyanotic congenital cardiac anomaly characterized by a single arterial trunk arising from the heart and supplying the systemic, pulmonary, and coronary circulations. Survival into adulthood without surgical correction is uncommon because of the early development of severe pulmonary arterial hypertension, heart failure, and Eisenmenger physiology.[1]

Only a handful of cases of pregnancy in women with unrepaired truncus arteriosus have been reported in the literature. Most published reports describe pregnancies in women with surgically corrected truncus arteriosus or comparatively stable cardiac status, while only very few reports describe successful pregnancies in women with unrepaired truncus arteriosus and pulmonary hypertension.[2–8]

In patients with longstanding uncorrected disease, progressive pulmonary vascular remodeling may eventually lead to Eisenmenger physiology, which is associated with extremely high maternal mortality during pregnancy due to the physiologic hemodynamic changes in pregnancy. According to the World Health Organization (WHO) classification, such patients fall under class IV cardiac risk, where pregnancy is generally contraindicated due to maternal mortality rates ranging from 30% to 50%.[9]

The coexistence of uncorrected truncus arteriosus, Eisenmenger physiology, pregnancy, and severe preeclampsia represents an exceptionally high-risk

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clinical scenario requiring meticulous multidisciplinary management.[10]

Case Description

A 27-year-old primigravida was referred from a peripheral hospital at 18 weeks of gestation in view of truncus arteriosus with ventricular septal defect with severe pulmonary arterial hypertension for further evaluation and management. She was diagnosed to have truncus arteriosis Type I since childhood, with severe pulmonary arterial hypertension, for which no corrective surgery had been performed. After evaluation by a multidisciplinary team, the patient and her family were extensively counseled regarding the extremely high maternal risk. Despite this, the patient opted to continue the pregnancy.

She received antenatal care at a private hospital. She only attended our hospital again at 34 weeks and 2 days of gestation with complaints of progressive breathlessness since two days. On admission, the patient was conscious and oriented but appeared dyspneic. Vital parameters revealed tachycardia (114 beats/min), tachypnea (34 cycles/min), and severe hypertension (BP 170/100 mmHg). Oxygen saturation was critically low at 59% on room air, improving only to 68% with supplemental oxygen. Clinical examination showed central and peripheral cyanosis, grade II clubbing, raised jugular venous pressure, and bilateral pedal edema, suggestive of advanced Eisenmenger physiology.

Obstetric examination revealed a uterus corresponding to approximately 30 weeks of gestation with a live fetus in cephalic presentation. The non-stress test was reactive. Abdominal wall edema was noted. Laboratory investigations were consistent with severe preeclampsia, and obstetric ultrasound demonstrated late-onset fetal growth restriction.

Cardiology evaluation during the current admission showed truncus arteriosus Type I with a large subtruncal ventricular septal defect, severe pulmonary arterial hypertension, and bidirectional shunting, consistent with Eisenmenger physiology (Fig. 1). The patient was categorized as cardiac class IV risk, indicating extremely high perioperative and maternal risk.[9]

The patient was managed with a multidisciplinary approach involving senior consultants from obstetrics, cardiology, respiratory medicine, endocrinology, and intensive care teams. Initial stabilization included administration of antihypertensives, oxygen therapy, diuretics, and careful fluid restriction, with continuous hemodynamic monitoring. Respiratory support was optimized with high-flow oxygen, and close monitoring for signs of decompensation was undertaken.

In view of worsening maternal condition and severe preeclampsia, after completing the dexamethasone course, an elective preterm lower segment cesarean section at 34 weeks 4 days was performed after a multidisciplinary assessment. The procedure was performed under epidural anesthesia to minimize hemodynamic fluctuations and to facilitate postoperative pain management. There were no intraoperative complications noted.The neonate weighed 1.7 kg, with APGAR scores of 6 and 8 at 1 and 5 minutes, respectively, and was shifted to the neonatal intensive care unit for respiratory support and management of low birth weight.

Postoperatively, the patient was shifted to the cardiac intensive care unit for close monitoring. She developed pulmonary edema and was managed aggressively using non-invasive ventilation along with high-flow oxygen support, cardiac medications and diuretics. Thromboprophylaxis with low-molecular-weight heparin was initiated. Careful attention was paid to fluid balance and oxygenation. Over the next 24 hours, her condition gradually improved, and ventilatory support was weaned off.

The patient was subsequently shifted back to the obstetric ICU, where the remainder of her hospital stay was uneventful. She was discharged on postoperative day 8 in stable condition. Her saturation was maintained between 48 – 74% on room air. She and the family were counseled about the need for long-term home oxygen therapy along with medications.

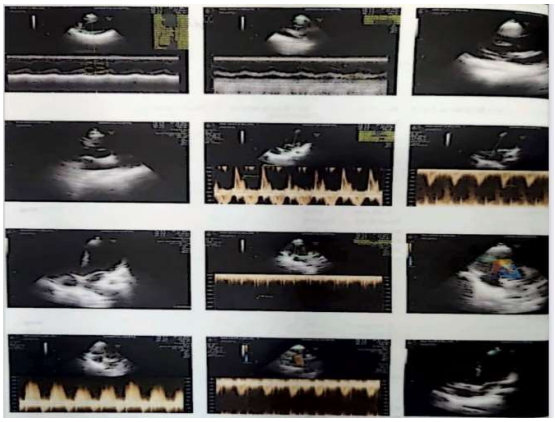

Figure 1: Echocardiographic findings

Composite transthoracic echocardiographic images demonstrating features of truncus arteriosus with a single arterial trunk, large ventricular septal defect, and severe pulmonary arterial hypertension. Doppler studies show abnormal flow patterns consistent with Eisenmenger physiology.

Discussion

Truncus arteriosus is a rare cyanotic congenital cardiac anomaly accounting for only 0.7% of all congenital heart lesions. It characterized by a single arterial trunk supplying systemic, pulmonary, and coronary circulations. Survival into adulthood without surgical correction is uncommon due to the early development of severe pulmonary arterial hypertension and heart failure.[1]

In patients who remain uncorrected, longstanding pulmonary vascular changes may eventually result in Eisenmenger physiology, which is associated with extremely high maternal mortality during pregnancy.[10] The physiological increase in blood volume and cardiac output during pregnancy exacerbates right-to-left shunting, resulting in worsening hypoxemia and increased risk of sudden cardiac decompensation.[11]

Current ESC (European Society of Cardiology) guidelines strongly advise against pregnancy in patients with Eisenmenger physiology (WHO class IV), recommending early termination due to high maternal mortality.[9]

In the present case, longstanding uncorrected truncus arteriosus Type I resulted in severe pulmonary arterial hypertension and development of Eisenmenger physiology.[12] The patient demonstrated profound hypoxemia, with oxygen saturation as low as 59% on room air, indicating advanced disease. Furthermore, she was categorized as WHO class IV cardiac risk, where pregnancy is strongly contraindicated.[9]

The superimposition of severe preeclampsia further increased the hemodynamic burden by elevating systemic vascular resistance, thereby worsening shunt dynamics and increasing the risk of right ventricular failure. This dual pathology significantly increased both maternal and fetal risk.

The decision for elective preterm cesarean section was based on progressive maternal deterioration and the need to minimize hemodynamic fluctuations. Regional anesthesia was preferred over general/spinal anesthesia due to the need for hemodynamic stability. It also had the added advantage of providing adequate postoperative pain relief.

Postoperative management in an intensive care setting is crucial, as patients with Eisenmenger physiology are particularly vulnerable to fluid shifts, hypoxia, and pulmonary edema in the immediate postpartum period, as seen in our case. The requirement of non-invasive ventilation and high-flow oxygen support in our patient reflects the severity of cardiopulmonary compromise.

Although pregnancy with truncus arteriosus has been described in the literature, most reported cases involve women with surgically repaired truncus arteriosus or comparatively stable cardiac status. Perry et al. reported successful childbirth in a woman with surgically repaired truncus arteriosus, while Steckham et al. described eight pregnancies in four women with repaired truncus arteriosus, with no adverse maternal cardiac events reported. In contrast, pregnancies with uncorrected truncus arteriosus are exceedingly rare, as survival into reproductive age without repair is uncommon.[2,3]

Among the few reported cases of uncorrected truncus arteriosus in pregnancy, Wilton et al. described anesthetic management for cesarean section in a 25-year-old primigravida with uncorrected truncus arteriosus, while Bosatra et al. reported cesarean delivery in a 28-year-old primigravida with uncorrected truncus arteriosus Type IV. More recently, Prathep et al. reported successful general anesthesia for cesarean section in a pregnant woman with truncus arteriosus and severe pulmonary hypertension, using intraoperative transesophageal echocardiographic monitoring. In comparison, our patient had uncorrected truncus arteriosus Type I with Eisenmenger physiology, profound hypoxemia, superimposed severe preeclampsia, and required postoperative ICU care with non-invasive ventilation and long-term oxygen therapy. This emphasizes the uniqueness of the present case and highlights the importance of individualized anesthetic planning and intensive postoperative surveillance.[4–6]

Despite these challenges, a favorable maternal and neonatal outcome was achieved. This can be attributed to coordinated multidisciplinary management. Cautious fluid balance, oxygen therapy, and timely intervention were crucial for the favorable outcome. Unlike many reported cases with poor maternal outcomes, our patient survived despite severe hypoxemia and superimposed preeclampsia, highlighting the critical role of intensive perioperative and ICU care in influencing outcomes.[13]

Previous studies have reported maternal mortality rates ranging from 30% to 50% in Eisenmenger physiology during pregnancy, despite advances in multidisciplinary care and critical care support.[9,14] In contrast to previously reported cases, our patient had uncorrected truncus arteriosus complicated by Eisenmenger physiology, severe preeclampsia, and persistent postoperative hypoxemia requiring ICU support, making this an exceptionally high-risk yet successfully managed pregnancy.

This case also emphasizes the need for preconception counselling of the patient and family and highlights the challenges faced when high-risk pregnancies are continued despite medical advice.

Conclusion

Uncorrected truncus arteriosus in pregnancy is an exceptionally rare and extremely high-risk condition due to the early development of severe pulmonary arterial hypertension and Eisenmenger physiology. Maternal and fetal outcomes are often poor, and pregnancy is generally contraindicated in such patients. However, in cases where pregnancy is continued, individualized,

meticulous multidisciplinary care, early recognition of complications, anesthetic planning, and intensive postoperative monitoring may improve outcomes. This case highlights that successful maternal and neonatal outcomes, though uncommon, are achievable even in exceptionally high-risk pregnancies with uncorrected truncus arteriosus.

Clinical Significance

This case underscores the importance of preconception counseling and highlights that continuation of pregnancy in WHO class IV cardiac conditions requires highly specialized multidisciplinary care and intensive monitoring to achieve favorable outcomes.

Ethical Considerations

Written informed consent was obtained from the patient for publication of this case report and accompanying clinical details.

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