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Case Report

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Cesarean section in a patient with Ebstein's anomaly: A multidisciplinary approach to high-risk obstetric management

La cesárea en una paciente con anomalía de Ebstein. Abordaje multidisciplinario del alto riesgo obstétrico

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Abstract

We present the case of a 37-year-old woman with Ebstein's anomaly and a prior tricuspid valve replacement. At 38.2 weeks of gestation, she underwent an elective cesarean delivery without complications. We propose a schematic representation of a standardized workflow system for pregnant women with heart disease in obstetric critical care units. Ebstein's anomaly is a rare congenital heart defect, with a prevalence of less than 0.3%, and limited evidence regarding its prognosis during pregnancy. Management requires a multidisciplinary approach, with primary emphasis on controlling heart failure. Implementation of the proposed workflow reduces indirect maternal mortality. Systematizing evidence-based interventions minimizes human error in complex scenarios and transforms empirical management into a protocol-driven strategy that ensures maternal and fetal safety.

Keywords: cardio-obstetrics, heart failure, high risk pregnancy

Resumen

Se presenta el caso de una paciente femenina de 37 años con una anomalía de Ebstein y antecedentes de reemplazo de la válvula tricúspide. A las 38,2 semanas de gestación, se le realizó cesárea programada, la cual transcurrió sin complicaciones. Se propone la representación esquemática de un sistema de trabajo estandarizado para mujeres embarazadas con cardiopatías en unidades de medicina crítica obstétrica. Esta es una cardiopatía congénita rara, con una prevalencia inferior al 0,3% y escasa evidencia sobre su pronóstico durante la gestación. Su abordaje exige un enfoque multidisciplinario y su objetivo principal es el control de la insuficiencia. La implementación del esquema de trabajo que se representa, reduce la mortalidad materna indirecta. Sistematizar las intervenciones basadas en evidencia minimiza el error humano en escenarios complejos y transforma la conducción empírica en una protocolizada que garantiza la seguridad del binomio madre-hijo.

Palabras clave: *cardio-obstetricia, insuficiencia cardíaca, embarazo de alto riesgo*

Introduction

Cardio-obstetrics is an interdisciplinary field focused on the care of pregnant women with preexisting cardiovascular disease—including congenital heart defects—or those who develop cardiac complications during pregnancy.[1]

The implementation of a Comprehensive Maternal Cardiac Care Model, in which the patient is at the center of a synchronized multidisciplinary team, involves specialists in maternal–fetal medicine, cardiology, anesthesiology, neonatology, nursing, social work, and pharmacology working collaboratively to optimize maternal and fetal outcomes.[2]

Management of pregnant women with heart disease in the Critical

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Care Unit (CCU) requires a multidisciplinary approach, strict monitoring, and careful consideration of pregnancy-related physiological changes to optimize outcomes.[3]

The most relevant components of this approach include:

1. Risk assessment and stratification: Maternal morbidity and mortality risk are stratified using the World Health Organization (WHO) risk classification, which guides decision-making regarding pregnancy continuation and the required level of care.[4] Preconception counseling—including genetic counseling and family planning—is essential to assess risks and discuss potential complications.[5]
2. Multidisciplinary management: Care should be delivered by a specialized team including obstetricians, cardiologists, anesthesiologists, and critical care specialists with expertise in cardiac disease during pregnancy. Referral to centers with specialized cardiology programs is recommended.[6]
3. 3. Strict hemodynamic monitoring: Physiological changes during pregnancy, such as increased blood volume and cardiac output, along with decreased systemic vascular resistance, may precipitate decompensation. Continuous monitoring, including advanced hemodynamic parameters available in tertiary care centers, is crucial for early detection of heart failure or arrhythmias.[7,8]
4. 4. Pharmacologic management: Medication regimens must be reviewed and adjusted to avoid contraindicated drugs during pregnancy. Anticoagulation is critical in conditions such as pulmonary hypertension or the presence of mechanical heart valves; in these cases, agents with established safety profile in pregnancy, including low-molecular-weight heparins, are recommended. Associated conditions such as anemia must also be treated, as they may exacerbate cardiac stress.[9]
5. 5. Labor and puerperium management: The mode and timing of delivery, vaginal or cesarean, must be carefully planned by the multidisciplinary team. An anesthesiologist experienced in cardiac disease should be present. From the second trimester onward, the patient should be positioned in the left lateral decubitus position to prevent aortocaval compression and improve venous return. The puerperium is a critical period due to abrupt hemodynamic shifts following delivery. Oxytocin can be administered as a slow infusion to prevent postpartum hemorrhage; however, agents such as ergometrine and prostaglandin F analogues should be avoided due to their potential cardiovascular adverse effects. Thromboembolism prophylaxis is a standard recommendation.[10]
6. 6. Emergency considerations: In the event of maternal cardiac arrest, cardiopulmonary resuscitation should be initiated with pregnancy-specific modifications, including lateral uterine displacement and, in advanced gestation, consideration of perimortem cesarean delivery when indicated.[11]

Case Report

A 37-year-old primigravida with a history of Ebstein's anomaly and systemic arterial hypertension treated with 50 mg of metoprolol twice daily. She had undergone tricuspid valve replacement one year earlier and is under hematology follow-up for thrombophilia, plus a deficiency of protein S diagnosed in 2019, being treated with 80 mg of enoxaparin every 12 hours subcutaneously.

She is also under endocrinology care for primary hypothyroidism diagnosed three years earlier, treated with levothyroxine 100 mg every 12 hours. She is being followed by the Obstetric Critical Care department and treated with 850 mg of metformin due to a history of gestational diabetes. The patient reports allergies to B-complex vitamins, amlodipine, and ceftriaxone. Under multidisciplinary management, she reached 38 weeks of gestation.

Cardiological evaluation via transthoracic echocardiogram reveals:

RIGHT VENTRICLE: severe dilation of the right ventricle (basal diameter 52 mm, mean diameter 32 mm, longitudinal diameter 67 mm; wall thickness 5 mm). The mobility is preserved.

Right ventricular systolic function measured by TAPSE is 19 mm.

With a myocardial performance index of 0.54

Ending diastolic volume: 126 mL; Ending systolic volume: 79 mL

Right ventricle ejection fraction: 37%; combined systolic–diastolic right ventricular dysfunction; fractional shortening 26%; RV/LV ratio 1.37.

Bioprosthetic valve is present in tricuspid position; no leaks or abnormalities observed.

LEFT VENTRICLE: Left ventricular geometry abnormal due to concentric remodeling; left ventricular diastolic function grade II, left ventricular ejection fraction: 64%.

Maternal–fetal medicine evaluation reports:

A single live fetus with average fetometry within the normal percentile, longitudinal lie, breech presentation, heart rate of 145 bpm, normoinserted placenta with no structural al-

terations. Doppler flowmetry of the umbilical artery pulsability index (PI): 1.03, middle cerebral artery PI 1.61, and cerebro-placental ratio 1.56, all within normal limits.

An elective cesarean section was scheduled and performed without intraoperative complications. The patient had an uneventful recovery and continued postpartum surveillance. She was discharged with appropriate treatment. One month later, at outpatient obstetric critical care follow-up, the patient was asymptomatic, with laboratory control tests within normal limits, and was discharged from the service. During the cesarean section, bilateral tubal occlusion was performed as definitive contraception.

Discussion

Management of pregnancy in women with Ebstein's anomaly requires a multidisciplinary approach involving obstetric intensivists, cardiologists, and anesthesiologists due to maternal and fetal risks. The decision to perform cesarean section and disease-specific treatment depends on the severity of the patient's condition and the hemodynamic stability12.

Most women with Ebstein's anomaly tolerate pregnancy well; however, those with pre-pregnancy heart failure symptoms are at increased risk of major adverse cardiovascular events and require thorough counseling13.

Vaginal delivery is generally considered the preferred mode of delivery, as cesarean section has been associated with a higher risk of complications, including postpartum heart failure and adverse fetal outcomes. Nevertheless, in this case, cesarean delivery was performed with minimal morbidity14.

Approximately 10% of pregnant women with Ebstein's anomaly experience severe adverse events, most commonly heart failure or arrhythmia. Therefore, classification as WHO risk class II appears reasonable. Preconception counseling is essential to identify additional risk factors15.

Conclusion

Implementation of a standardized management workflow in the critical care unit represents an effective strategy to reduce indirect maternal mortality. Systematizing evidence-based interventions minimizes human error in high-complexity scenarios and transforms empirical management into a protocol-driven strategy that ensures maternal and fetal safety while optimizing institutional resources.

Proposal

Management of pregnant women with heart disease in the Obstetric Critical Care Unit (OCCU) should focus on a multidisciplinary approach, intensive monitoring, and careful consideration of pregnancy-related physiological changes. Care requires a specialized team including obstetricians, cardiologists, anesthesiologists, intensivists, and neonatologists, all formally trained in cardio-obstetrics. Successful management demands a proactive strategy, ideally beginning with preconception counseling and continuing with coordinated follow-up throughout pregnancy and the puerperium. The establishment of dedicated cardio-obstetrics units is strongly recommended, given that cardiometabolic con-

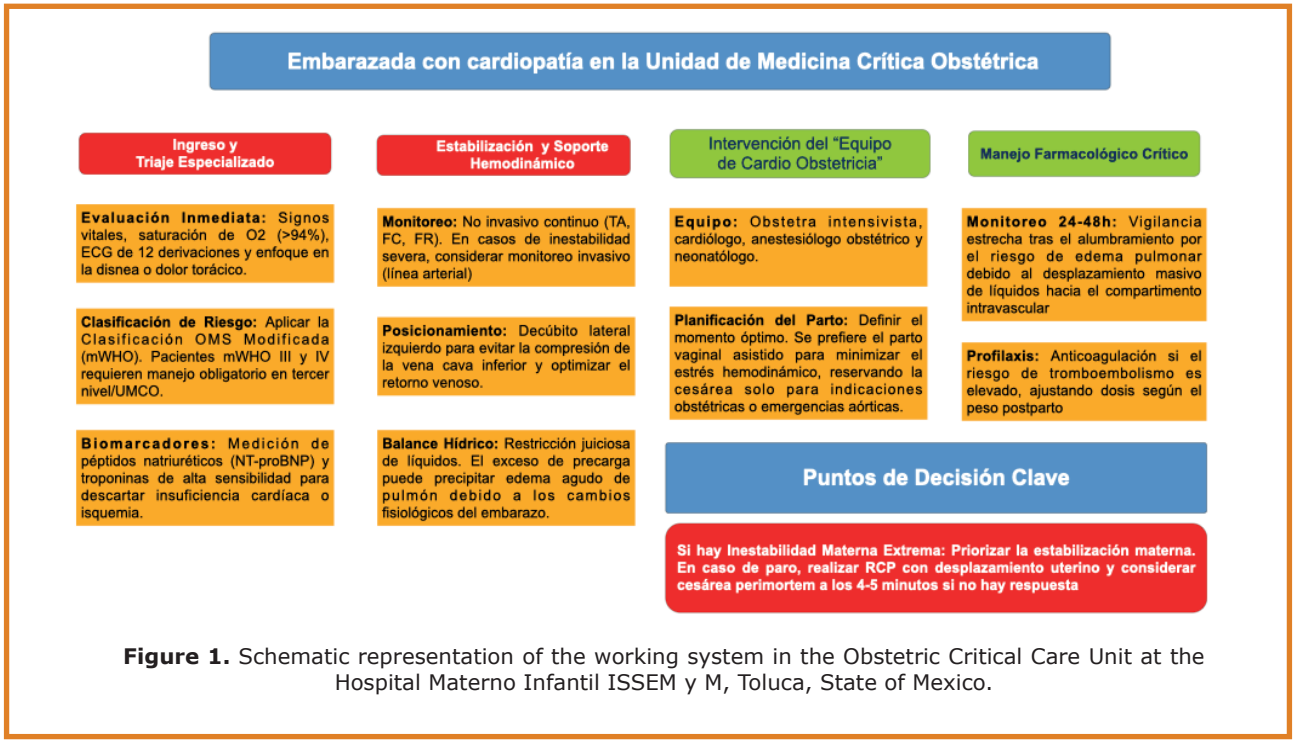

Figure 1. Schematic representation of the working system in the Obstetric Critical Care Unit at the Hospital Materno Infantil ISSEM y M, Toluca, State of Mexico.

ditions remain leading causes of maternal mortality. **Figure 1** illustrates the workflow implemented in the management of this patient.

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