Peripartum management of Eisenmenger syndrome : A case report

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Abstract

Eisenmenger syndrome is defined by elevated pulmonary vascular pressures and a right-to-left shunt across a defect connecting the systemic and pulmonary circulation. During pregnancy, this condition results in significant morbidity and mortality.

We report the case of a 36-year-old patient admitted to our facility at 35 weeks of gestation for the management of Eisenmenger syndrome due to a patent ductus arteriosus during pregnancy.

The end of the third trimester, labor, delivery by epidural cesarean section, and the postpartum period were uneventful and without impact on the patient.

Patients in this situation should be managed by a multidisciplinary team, with adequate monitoring and medication management, including the avoidance of factors that aggravate pulmonary hypertension and the anticipation of complications related to this condition.

Key words

Eisenmenger syndrome, case report, Anesthesia, Peripartum, Pulmonary arterial hypertension.

Introduction

Pregnancy causes physiological changes, particularly cardiovascular ones. It is contraindicated in cases of WHOm IV heart disease, and its termination should be discussed [1,2]. If the pregnancy continues, multidisciplinary monitoring is required. Management includes numerous anesthetic implications, particularly beyond the 30th week of amenorrhea, during labor, and the postpartum period.

We present the case of a 36-year-old patient being monitored for severe pulmonary hypertension, who was diagnosed with Eisenmenger syndrome due to a patent ductus arteriosus at the end of the third trimester of pregnancy.

Case report

Patient Information : We report the case of a 36-year-old patient admitted at 35 weeks of gestation to the emergency department of the Souissi Maternity Unit of the Ibn Sina University Hospital in Rabat-Salé for severe intrauterine growth restriction.

She has been treated for pulmonary hypertension since the age of 14 and was given sildenafil 20 mg once daily orally.

Pregnancy was diagnosed on May 20, 2024, at 13 weeks of gestation, with concomitant diagnosis of a patent ductus arteriosus and severe pulmonary hypertension. The decision was made to discontinue sildenafil the same day. The evolution of the pregnancy was marked by the appearance of intrauterine growth restriction which justified hospitalization at 27 weeks of amenorrhea, then the occurrence of community-acquired pneumonia without cardiac decompensation at 31 weeks of amenorrhea treated with antibiotics with good clinical and obstetrical evolution.

Clinical Findings : The examination revealed a conscious patient with normal conjunctiva. A gynecological examination revealed a uterine height of 27 cm, an estimated fetal weight of 1500 g, and no abnormalities in the umbilical and cerebral Dopplers. The patient had a heart rate of 80 beats per minute, blood pressure of 116/65 mmHg, and no orthopnea or signs of

acute heart failure. Auscultation revealed a left subclavicular murmur in a eupneic patient, with no signs of respiratory struggle, and ample and symmetrical breathing. Furthermore, the patient presented with digital clubbing, with a difference in pulse oxygen saturation in the four limbs, shown in Figure 1.

Timeline : (Figure 2).

Pre-anesthetic assessment: The evaluation found the patient exhibiting no signs of severe pulmonary hypertension, including dyspnea, orthopnea, chest pain, syncope, or signs of right heart failure.
The electrocardiogram Found a right axis deviation. Cardiac enzyme markers were negative. Furthermore, the workup revealed no hepatic or renal involvement.
A transthoracic echocardiogram was performed the day after admission and revealed a patent ductus arteriosus, with severe pulmonary hypertension (PAPS at 78 mmHg, PAPD at 60), significant dilation of the right cavities, and functional tricuspid regurgitation, with a right-to-left shunt.
Our evaluation also revealed polycythemia at 16.9 g/dL, with a hematocrit of 50.8%.

Anesthetic preparation and delivery procedure: Our preparation consisted of reintroducing sildenafil at 20 mg per day orally, oxygen therapy for vasodilatory purposes, and avoiding factors aggravating pulmonary hypertension.
She underwent multiple daily monitoring of her heart rate, respiratory rate, pulse oxygen saturation, and blood pressure. Her blood volume was closely monitored clinically, biologically and using ultrasound.
At 37 weeks of gestation, fetal heart rate showed distress pattern, justifying the indication of an emergency cesarean section.

The patient received epidural anesthesia. This was performed in the operating room in a seated position, with blood pressure, right hand pulse saturation, and maternal and fetal heart rates monitored, in a heated room under oxygen, after an IV filling by 200 cc of isotonic saline.
The puncture site, at L4-L5, was infiltrated with 3 cc of 2% xylocaine. The epidural space was identified by loss of resistance. We administered 3 cc of 2% xylocaine every 5 minutes for a total of 280 mg, with 25 micrograms of fentanyl. There was no vascular penetration of the local anesthetics, and the patient did not experience hypotension or heart rate abnormalities.

The cesarean section resulted in the delivery of a 2100 g male newborn. The APGAR score was 8, 9 and 9/10, with respiratory distress at 1/10 on the Silverman scale on examination. The patient received 15 IU of syntocinon by slow intravenous infusion, with no observed hypotension or tachycardia, combined with antibiotic prophylaxis for Oslerian endocarditis. The procedure was not complicated by bleeding or volume overload. This was prevented by administering furosemide at a dose of 0.5 mg/kg directly intravenously to compensate for the effects of autotransfusion following delivery.

Postpartum and outcome: The patient was transferred to intensive care with warming, multimodal analgesia, monitoring, and thromboprophylaxis using intermittent pneumatic compression, followed by medication after removal of the epidural catheter.

Her stay was not marked by the occurrence of any rhythmic or thromboembolic complications. Furthermore, she did not experience any decompensation of her heart disease, with a follow-up echocardiogram not revealing any notable changes. She was discharged on the 6th day of intensive care with a favorable outcome and a clean wound.
The evolution of her newborn was favorable after 2 days of non-invasive ventilation. His lactate level ruled out the diagnosis of acute fetal distress, and an echocardiogram ruled out the presence of congenital heart disease. He was released to his family on the 2nd day.

Discussion

Eisenmenger syndrome is defined as elevated pulmonary vascular resistance and a right-to-left shunt across a defect connecting the pulmonary and systemic circulation.
This defect is responsible for pulmonary hyperflow, resulting in histological lesions of the pulmonary vasculature. These lesions are irreversible and lead to a loss of vasodilation capacity, arterial stiffness, and increased pulmonary vascular resistance [3].
During pregnancy, numerous physiological changes occur. Circulating volume increases, and there is a decrease in systemic vascular resistance. This, combined with the lack of reduction in pulmonary vascular resistance, leads to an exacerbation of the right-to-left shunt during pregnancy [4].
The consequences are numerous for the mother and fetus and are summarized in Table 1.

These changes require avoiding factors that aggravate pulmonary hypertension during pregnancy and the peripartum period. This requires optimizing vasodilator therapy, which may include inhaled nitric oxide, prostacyclins (including epoprostenol), and phosphodiesterase type 5 inhibitors. All of these agents have been used successfully during pregnancy, unlike endothelin receptor antagonists (Bosentan), which are teratogenic but can be initiated postpartum.
Finally, due to the inherent thromboembolic risk of pregnancy and the postpartum period, which is increased by pulmonary hypertension, anticoagulation should be discussed during pregnancy and is mandatory during the postpartum period [4].

There are numerous anesthetic implications during labor for a woman suffering from Eisenmenger syndrome. One of these concerns the route of delivery:
The various available case studies do not allow us to determine the best route of delivery. A 1998 study considers cesarean section to be an independent factor in maternal mortality. Indeed, it is associated with greater variations in blood volume, greater blood loss, and increases the risk of thromboembolic events. However, a 2009 review specifies that the route of delivery does not represent an additional risk factor. It should be noted that the vaginal delivery is accompanied by Valsalva maneuvers concomitant with contractions, which reduce right ventricular preload and worsen the right-to-left shunt [5, 6]
Finally, the ESC recommended in 2018 that cesarean sections be preferred in cases of severe pulmonary hypertension [7].

During labor and delivery, factors that aggravate pulmonary hypertension must be avoided. Pain is one of these factors, and it is therefore necessary to consider an anesthesia that provides optimal analgesia with the least possible hemodynamic impact. Cold, hypoxia, acidosis, and hypercapnia are other factors that aggravate pulmonary hypertension, and they must be actively prevented [3, 5].

Finally, hypovolemia (absolute or relative) and volume overload also represent a decompensation factor, hence the need to choose an anesthetic technique that preserves hemodynamic stability [6].
The various advantages and disadvantages of anesthetic techniques are summarized in Table 2.
In our case, we preferred epidural anesthesia. It provides analgesia that can be titrated according to pain, and has minimal hemodynamic impact.

Hemodynamic variations during labor require close hemodynamic monitoring. It should be noted that bleeding can cause hypovolemia, and autologous transfusion following delivery can cause overload, which can be fatal in the setting of Eisenmenger syndrome. Echocardiographic and invasive blood pressure monitoring are recommended in these situations. The benefit of pulmonary catheterization is debated, particularly given its invasive nature [8].

Finally, the postpartum period is the period with the highest mortality and morbidity rates. This can be explained by volume overload, the incidence of pain, and the occurrence of thromboembolic events, which explains the need for 2 weeks of monitoring in an intensive care unit or intensive care unit, with optimization of blood volume, vasodilator therapy, and chemical or mechanical thromboprophylaxis [9].

The limitations in our case are numerous. The attending physician did not propose termination of the pregnancy at 13 weeks of gestation as recommended. Vasodilator treatments aren't available. Intrapartum hemodynamic monitoring was neither invasive nor complete due to the urgency of the extraction.
Finally, the obstetricians and cardiologists did not offer antenatal advice to the patient regarding the prevention of a future pregnancy.

Conclusion

The association of Eisenmenger syndrome and pregnancy can be responsible for significant mortality and comorbidities. This explains the need for pregnancy prevention in patients with this syndrome and for multidisciplinary and close management of these patients in the event of pregnancy, particularly in resource-limited countries where vasodilatory treatment is not available.

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Tables and figures

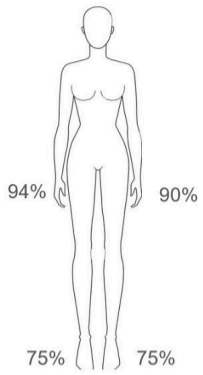

Figure 1 : Difference in pulse oxygen saturation in the four limbs.

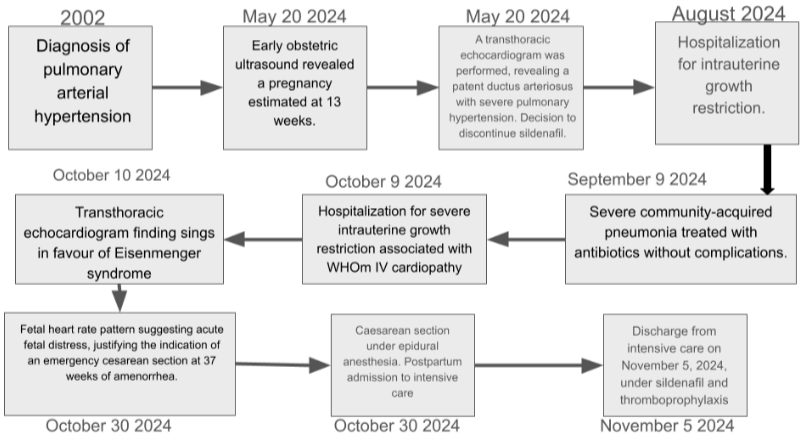

Figure 2 :Timeline.

	Maternal	Fetal

Complications	Arrhythmia Ventricular dysfunction Ventricular failure (left, right, or global) Sudden death Myocardial infarction Thromboembolic events	Prematurity Intrauterine growth restriction Intrauterine fetal death Abortion Congenital heart disease

Table 1 : Eisenmenger Syndrome and pregnancy related complication for the mother and the foetus

Techniques	Advantages	Disadvantages
Epidural anesthesia	Possible titration of analgesia Hemodynamic stability compared to other techniques	Neurological injuries Difficulty of the technique, failure, and need for conversion to general anesthesia Dural rupture Slowness of technique execution
General Anesthesia	Speed Better airway management	Use of vasodilatory or negative inotropic drugs Neonatal respiratory distress Inhalation Uterine atony
Spinal Anesthesia	Effective sensory block Speed of execution	Decreased systemic vascular resistance Worsening of the right-to-left shunt Neurological damage Difficulty of the technique, failure, and need for conversion to general anesthesia

Table 2 : Comparison of different anesthetic techniques for cesarean delivery.

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