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Cesarean Delivery in Severe Pulmonary Hypertension: A Case Report

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Summary. *Pregnancy in a woman with pulmonary hypertension carries a prohibitively high risk of maternal mortality, and pregnancy is contraindicated in such patients. Some women decide to continue with their pregnancy despite being aware of possible fatal maternal outcome. The management of pulmonary hypertension in pregnancy is a challenge and requires a multiprofessional approach. We report the case of a patient with severe pulmonary hypertension, who successfully underwent elective cesarean section under epidural anesthesia at 38 weeks of gestation and discuss major issues associated with the obstetric and anesthetic management of pregnant patients with pulmonary hypertension.*

Introduction

Pulmonary hypertension (PH) is a rare pathology encountered in pregnant women, which carries a high risk of maternal mortality. PH manifests with a gradual increase in pulmonary artery pressure (PAP) accompanied by pulmonary arterial wall changes of various degree, endothelial dysfunction, vasoconstriction, and thrombosis, which ultimately cause right ventricular insufficiency (1–5). The mean PAP greater than 25 mm Hg or the peak systolic PAP greater than 40 mm Hg is interpreted as pulmonary hypertension. The disease is usually classified as severe when the mean PAP exceeds 45 mm Hg or the peak systolic PAP exceeds 60 mm Hg (6). Initial symptoms are often nonspecific, and PH is only diagnosed in late pregnancy or as a casual finding during transthoracic echocardiography (TTE) (1, 2). In the presence of PH, a physiological increase in blood volume that occurs during pregnancy causes volume overload in the right heart and problems of hemodynamic compensation that may have serious consequences, such as thromboembolic events, cerebrovascular accidents, or blood hyperviscosity syndrome (7). Anesthetic management of a parturient with PH undergoing cesarean section is a challenge as maternal mortality rate remains 30%–50% irrespective of adequate medical care (1, 8–10).

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We report the case of a patient with severe PH, who successfully underwent an elective cesarean section under epidural anesthesia at 38 weeks of gestation and discuss major issues related to the obstetric and anesthetic management of pregnant patients with PH.

Case Report

A 34-year-old multiparous woman, second pregnancy and labor, presented with hemoptysis at 36 weeks of gestation and was urgently hospitalized in our perinatology center suspecting pulmonary artery embolism. The patient also complained of general weakness and dyspnea. Ten years ago, she had a cesarean delivery due to fetal hypoxia under general anesthesia without any complications.

Our parturient had a congenital heart defect diagnosed in childhood, which, however, was not corrected possibly due to parental negligence. Cardiac catheterization and specification of the diagnosis were performed only at the age of 32 years. The aforementioned examination showed a significant atrial septal defect with left-to-right shunt, right atrial pressure of 90 mm Hg (reference range, 0–7 mm Hg), PAP of 90/48 mm Hg (reference range, 17–32/4–13 mm Hg), and mean PAP pressure of 63 mm Hg (reference range, 9–19 mm Hg) suggesting severe PH. Cardiac surgeons considered the risk-to-benefit ratio of surgical treatment in such an advanced disease unfavorable. The woman was discouraged from pregnancy, and strict recommendations for contraception were given.

On examination, cyanotic lips at rest were observed, and our patient could only sleep in a semi-sitting position. Rhythmic cardiac tones with an accentuated pulmonary component of the second heart sound were found on auscultation, heart rate was 76 beats per minute, and arterial blood pressure was 109/50 mm Hg. Electrocardiography showed sinus rhythm and complete right bundle-branch block suggesting overload of the right heart. Lung function test revealed third-degree lung restriction. TTE revealed an undisturbed function of the left ventricle with an ejection fraction of 60%, pronounced right ventricular and atrial dilation, third-degree tricuspid leakage, peak systolic PAP of 80 mm Hg, and pulmonary artery dilation of up to 4.6 cm.

Inpatient follow-up TTE showed a mildly reduced left ventricular ejection fraction of 50%, first-degree mitral leakage, insufficient systolic interventricular septum contraction, third-degree tricuspid leakage, and progression of PH (peak systolic PAP, 95 mm Hg). Arterial blood gas test showed hypoxemia (pO_2, 85 mm Hg), hypercapnia (pCO_2, 48 mm Hg), and pH 7.35. Oxygen therapy (2–3 L/min through nasal catheters) was initiated, and treatment with low-molecular-weight heparin (LMWH) (subcutaneous nadroparin, 0.3 mL b.i.d.) was administered.

Based on relatively stable maternal and good fetal conditions, our multiprofessional team decided to extend the gestation to 38 weeks under close fetal and maternal monitoring and to schedule an elective cesarean section. At 38 weeks of gestation, the patient underwent a cesarean section under epidural anesthesia (EA).

Internal jugular vein and radial artery were catheterized in the operating room for invasive hemodynamic monitoring, as the risk of cardiovascular decompensation was critical. The patient started with a blood pressure of 110/70 mm Hg, heart rate of 84 beats per minute, central venous pressure (CVP) of 18 mm Hg, and oxygen saturation of 98% with a facemask (1:1 oxygen and air mixture). A moderate intravenous infusion of 500-mL Ringer lactate was started. An epidural catheter was inserted in the L1/L2 interspace 5 cm cephalad; test dose, negative. An epidural block was established with the incremental doses of bupivacaine/fentanyl mixture: the dose of 10 mL 0.5% bupivacaine and 0.025 mg of fentanyl was followed by a top-up dose of 5 mL 0.5% bupivacaine and 0.025 mg of fentanyl after 10 minutes. Block level was estimated 10 minutes after each dose by temperature test until it reached T4 segmental level. Intraoperative hemodynamic parameters remained stable with an arterial blood pressure of 105–125/55–60 mm Hg, heart rate of 74–80 beats per minute, and CVP of 20 mm Hg. A total volume of 1000-mL crystalloids was infused during the surgery. Estimated intraoperative blood loss was approximately 400 mL; urine output, 300 mL. Neither vasopressors nor inotropes were necessary. A full-term male neonate was born (weight, 2580 g; Apgar score, 9/9 at 1 and 5 minutes).

Postoperatively the parturient was transferred to the Cardiology ICU painless with stable hemodynamics. Infusion therapy with crystalloids, oxygen therapy through nasal catheter (oxygen, 3–4 L/min), and postoperative epidural analgesia were continued. A constant infusion of heparin (1000 UA/h) was administered. On the second postoperative day, heparin was changed to LMWH, and bosentan was initiated at a dosage of 125 mg per day. Postoperative hemodynamics remained stable. Eight days later, follow-up TTE showed a left ventricular ejection fraction of 50% and decreased peak systolic PAP to 70 mm Hg. On the postoperative day 14, our patient with her newborn was discharged, and further treatment with warfarin and bosentan was prescribed.

Discussion

According to a recent systematic review by Bedard et al. (10), overall maternal mortality rates for parturients with PH remain prohibitively high although they seem to have decreased from 30%–56% (reported in 1998) (9) to 17%–33% in the last decade. Pregnancy is still discouraged, and termination should be considered if pregnancy occurs (5, 8). Our case, however, reflects that the sense of motherhood may overcome the fear of death. The patient aware of her severe PH became pregnant despite all evidence-based warnings by doctors.

Preoperative Management. In order to ensure the best available care and reduce the risk of complications and death, it is recommended to hospitalize patients at 20 weeks of gestation, when pregnancy-induced hemodynamic changes reach their peak (11–12). Maternal and fetal conditions have to be observed via repeated ultrasound investigations because intrauterine growth retardation due to existing hypoxemia and increased hematocrit level is diagnosed in 33% of cases (12).

If PH is diagnosed in pregnancy, pulmonary vasodilators (calcium channel blockers, prostanoids, phosphodiesterase-5 inhibitors) are usually initiated; oxygen inhalations may be administered (5). Because of the increased risk of venous thromboembolism during pregnancy coupled with the limited cardiorespiratory reserve in patients with PH, anticoagulation is usually recommended. The agent of choice is LMWH (13). Endothelin receptor antagonists (for example bosentan) and warfarin are avoided due to potential teratogenicity. If PH is diagnosed only in the preoperative period and surgery is urgent, disease-specific treatment is initiated as

soon as possible postoperatively (4). We faced the aforementioned situation as the patient never attended any clinic for prenatal care and presented only in late pregnancy with symptoms of severe PH and signs of decompensation.

Considerations Regarding Anesthesia. There is little experience and no clear recommendations addressing the methods of anesthesia for a parturient with PH. Most data are acquired from published clinical case reports (7, 14–15). The choice of anesthesia method undergoing a cesarean section is still an issue because reference can only be made to single clinical cases collected during 10–15 years, while medications and methods used for anesthesia have been improving significantly over years; therefore, such cases are difficult to compare. It is critical to avoid substantial hemodynamic variations, which may induce the sudden emergence of right ventricular insufficiency (4, 12). That is why the mode of delivery and anesthetic management remain debated. Most reported cases have recommended vaginal delivery under epidural analgesia (15).

Nonetheless, a scheduled cesarean delivery is often used. It has the advantage of taking place on the daytime and avoiding the risk of urgent cesarean section in hemodynamically unstable labor that is likely to occur under such conditions. There are some cases reported when general anesthesia was used with a good maternal outcome (7). However, an increased PAP during laryngoscopy and tracheal intubation is reported. Moreover, the adverse effects of positive-pressure ventilation on venous return may ultimately lead to cardiac failure (4). Anesthesia level and pain control might be inadequate at the beginning of the surgery; therefore, the general condition of the patient might impair. Bedard et al. (10) in their systematic review reported that PH patients receiving general anesthesia for cesarean section were 4 times more likely to die compared with patients receiving regional anesthesia.

The number of case reports highlighting the use of regional anesthesia with good outcomes is increasing (14). However, a dense and extended block needed to prevent pain during cesarean delivery may have significant hemodynamic consequences. Specifically, the use of single-shot spinal anesthesia for these patients is discouraged.

Therefore, epidural anesthesia with incremental doses is most often advocated (8, 14). When undergoing EA, a combination of an opioid and a local anesthetic (lowest effective concentration) is recommended to avoid hypotension and to ensure a sufficient sensory block (14). In the light of growing literature basis supporting an epidural anesthetic approach as the best regional technique, we chose EA with the incremental doses of bupivacaine/fentanyl mixture for our cesarean section.

Postoperative Management. All patients should be followed up and treated in an intensive care department for at least 48–72 hours postoperatively, as the risk of complications and sudden death during this period remains very high (12). Postoperative pain is an important issue, and therefore, we consider the possibility to continue analgesia via an epidural catheter as one of the advantages of EA. The administration of lung vasodilators (bosentan, prostaglandins) is initiated or resumed. Unfractioned heparin or LMWH is changed into warfarin, an indirect action anticoagulant (1).

Conclusions

Management of pulmonary hypertension in pregnancy is a very complicated task, which requires close cooperation among obstetricians-gynecologists, anesthesiologists and intensive care specialists, pulmonologists, and cardiologists. Our successful management of the patient with severe pulmonary hypertension under epidural anesthesia with invasive monitoring contributes to the literature advocating epidural anesthesia as the best regional technique for cesarean section.

Statement of Conflicts of Interest

The authors state no conflicts of interest.

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