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CASE REPORT

Intrauterine Repair at 25 Weeks For Myelomeningocele. Case Report With Video

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Abstract

Background: Neural tube closure defects, such as myelomeningocele, represent serious congenital malformations that can result in significant neurological sequelae. Intrauterine surgical correction has emerged as a promising strategy to improve fetal prognosis by treating the defect before birth.

Case Report: This study reports a healthy 35-year-old patient with gestational age (GA) of 25 weeks plus 4 days, with no history of anesthetics, physical status ASA 1 who underwent surgery for intrauterine correction of myelomeningocele under balanced general anesthesia associated with continuous epidural anesthesia. Preoperatively, omeprazole 40 mg, metoclopramide 10 mg, cimetidine 100 mg and, as pre-anesthetic medication, midazolam 3 mg were administered. The epidural puncture was performed in L1-L2 intervertebral space after sedation with midazolam and fentanyl, in a single attempt with a Touhy needle, infused with 0.1% ropivacaine, followed by the passage of an epidural catheter 20G for postoperative analgesia. Anesthetic induction was in rapid sequence with propofol 150mg, lidocaine 80mg, remifentanil Minto´s model target controlled and rocuronium and maintenance of anesthesia with 3 to 4% Sevoflurane. After hysterotomy and exposure of the fetal gluteal region, anesthesia and fetal immobility were guaranteed with intramuscular injection of fentanyl 12µg and pancuronium 80µg. Maternal hemodynamic stability was guaranteed with a systolic target above 100mmHg with crystalloid infusion and norepinephrine bolus. At the end of the procedure, the amniotic fluid was replaced with warmed 0,9% saline solution. The procedure was completed without complications and the patient was sent to the Intensive Care Unit for immediate post-operative care with continuous infusion of ropivacaine into the epidural catheter for analgesia.

Conclusion: Anesthesia for fetal surgery involves anesthesia with an emphasis on the care of the mother-fetus binomial, the preservation of good uteroplacental flow, the adoption of tocolysis measures and the prevention of anesthetic and surgical complications. There is a need for multidisciplinary monitoring, with a team of anesthesia, neurosurgery, pediatrics and obstetrics.

Keywords: Myelomeningocele, Fetal Anesthesia, Intrauterine Surgery, Neural Tube Defect.

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1. Introduction

Congenital malformations during pregnancy and neural tube closure defects originate in embryonic life and appear in the first four weeks of pregnancy. The etiology is multifactorial and encompasses genetic and nutritional aspects, with maternal folic acid deficiency remaining the main modifiable factor of the disease [1]. Naturally, it has a higher incidence in underdeveloped countries, with Brazil ranking fourth in prevalence according to the World Health Organization [2].

Myelomeningocele is an embryonic malformation resulting from a failure in the closure of the neural tube resulting in a vertebral, musculofascial, cutaneous and dural opening with protrusion and exposure of the spinal cord, resulting in functional alteration in varying degrees, depending on the medullary level of the lesions, with greater neurological damage if located in more cranial medullary segments [3].

Myelomeningocele is diagnosed prenatally by means of obstetric ultrasound (US) and constitutes a surgical emergency at birth and should ideally be corrected within the first 24 hours of life [4]. It is associated with varying degrees of neurological deficit and the prognosis is worse the longer the treatment delay and the more cranial the lesion [5,6]. Recently, new surgical techniques for intrauterine correction of the defect have stood out on the world stage for reducing infant morbidity [6].

A study comparing repair of myelomeningocele, in utero repair with standard postnatal repair, showed reduced the need for shunting and improved motor outcomes at 30 months, but the early intervention was associated with both maternal and fetal morbidity, which requires a multidisciplinary team approach and presents unique challenges to the anesthesiologist [7].

Anesthesia for fetal surgery involves two beings, mother and fetus, and anesthetic management requires maternal-fetal safety, fetal anesthesia and immobility, uterine relaxation, prevention of premature labor, and postoperative analgesia [6]. This case report shows the first anesthetic procedure performed at the Hospital das Clínicas of the Federal University of Goiás (HC-UFG) for intrauterine correction of myelomeningocele in a fetus of 25 weeks of gestational age, with success and post-birth follow-up.

2. Case Report

This case report was registered in the Plataforma Brasil (CAAE 85314824.6.0000) and approved by the CEP/CONEP (number 7,284,616) of the Hospital das Clínicas of the Federal University of Goiás/EBSERH, and the Free and Informed Consent Form was signed by the mother and father after the explanation of the entire anesthetic procedure to be performed on the mother-fetus binomial and authorized for publication in an indexed medical journal.

A 35-year-old primigravida patient, gestational age (GA) of 25 weeks plus 4 days, ASA physical status II, with no gestational comorbidities, no known allergies and no predictors of difficult airway, underwent balanced general anesthesia associated with continuous epidural for intrauterine surgery to correct fetal myelomeningocele. The anesthetic procedure was performed according to the anesthesia protocol for intrauterine myelomeningocele developed by the Department of Anesthesiology of the Federal University of São Paulo.

As there were no contraindications, a fasting abbreviation protocol was performed with administration of 200 ml of maltodextrin 2 hours before admission to the surgical center. One hour before the procedure, still in the ward bed, an infusion of omeprazole 40 mg, metoclopramide 10 mg and cimetidine 100 mg was performed in a prior 22G venoclysis in the right hand. Upon admission to the surgical block, 3 mg of intravenous midazolam was administered as pre-anesthetic medication.

After performing the safe surgery checklist, the presence of the entire multidisciplinary team was confirmed, including anesthesiologist, neurosurgeon, obstetrician, pediatrician, ultrasound specialist, psychologist, and nursing staff. The patient was monitored with cardioscope, plethysmography, bispectral index (BIS), sequence of four stimuli (TOF), and indwelling urinary catheter. An anesthetic button was then placed with 1 ml of 2% lidocaine without vasoconstrictor, and the right radial artery was punctured for continuous invasive blood pressure measurement. In the first Ringer's lactate, 2 g of cefazolin and 2 g of magnesium sulfate were infused.

After obtaining 2 peripheral venous accesses of 16G in the left and right cubital fossa, the patient was sedated with 2 mg midazolam and 75 µg fentanyl, positioned sitting and underwent epidural puncture in the L1-L2 space, asepsis with 0.5% alcoholic chlorhexidine, anesthetic button was performed with 2% lidocaine, 18G Touhy needle in a single attempt with Dogliotti's technique, 20 ml of 0.1% ropivacaine was injected and a 20G epidural catheter was passed for continuous analgesia in the postoperative period.

The patient was placed in the supine position and the extent of the epidural block was assessed. Then, general anesthesia was induced in rapid sequence with 150 mg propofol, 70 mg lidocaine, remifentanil in target-controlled infusion according to the Minto pharmacokinetic model at 7 ng/ml, and 80 µg rocuronium. The patient was intubated with a 7.5 mm cuffed tube under direct laryngoscopy in a single attempt and attached to mechanical ventilation (Mindray A7 anesthesia machine) controlled by volume (VCV, total volume (VT) 400 ml and respiratory rate (RR) 20 respiratory incursions per minute and Positive End Expiratory Pressure (PEEP) 6 cm H20. Anesthesia was maintained with sevoflurane 2 minimum alveolar concentrations (MAC), remifentanil 7 ng/ml, magnesium sulfate 2 g/h and intermittent bolus of 10 µg rocuronium guided by TOF. Terbutaline was administered as a continuous infusion (2.5 mg of terbutaline in 500 ml of 5% glucose solution - 60 ml/h) throughout the anesthetic procedure aiming at tocolysis.

Hemodynamic targets were adjusted to maintain BIS 40-55, systolic blood pressure (SBP) above 100 mmHg, TOF 0, oxygen saturation greater than 94% and diuresis above 1 ml/kg/h. During the anesthetic procedure, it was necessary to perform a bolus of norepinephrine 8 µg to correct a transient episode of hypotension.

For anesthesia and immobility of the fetus (estimated weight by US of 800 g), 3 syringes containing 12 µg of fentanyl (15 µg/kg), 16 µg of atropine (20 µg/kg) and 80 µg of pancuronium (100 µg/kg) were prepared in a sterile manner and administered intramuscularly in the fetal gluteus, after hysterotomy (Video 1). Syringes of atropine 0.02 mg.kg and epinephrine 1 µg.kg were also available to rescue fetal hemodynamics. After adequate analgesia and fetal muscle blockade, the myelomeningocele was exposed and corrected without complications.

Video 1. *During surgical procedure.* **CLICK HERE**

The fetal heart rate (FHR) was monitored with intermittent ultrasound every 5 minutes. The minimum fetal heart rate recorded was 130 bpm and there was no need for additional doses of atropine. Warmed 0.9% saline was added to replace the amniotic fluid lost during the procedure. The amniotic membrane and uterine tissue were sutured uneventfully.

At the end of the procedure, the patient was extubated, the arterial puncture was removed and she was transferred to the intensive care unit (ICU) in hemodynamic stability, normotensive without vasoactive drugs, eupneic in room air and with continuous analgesia through the epidural catheter with ropivacaine 0.2% 10 ml/h in a Terumo TE-LM 830 continuous roller infusion pump, for 24 hours, with adequate pain control without the need for intravenous rescue with opioids. After this period, the epidural catheter was removed uneventfully, and the patient was transferred to the ward. The patient was discharged one week after the procedure and remained under follow-up in the high-risk prenatal service of HC-UFG.

The pregnancy proceeded uneventfully and elective cesarean section was performed at 39 weeks of gestational age. The newborn showed good vitality and obtained APGAR scores of 9 and 10 in the first and fifth minutes of life, respectively. There were no signs of myelomeningocele at birth. Today, the child, aged 10 months, remains under follow-up in the pediatrics department of the hospital with good neuropsychomotor evolution and no apparent deficits.

3. Discussion

A study comparing prenatal and postnatal correction demonstrated that children undergoing intrauterine correction of myelomeningocele is an intervention that stands out for its ability to modify the prognosis of a serious neural tube condition, preventing or significantly reducing neurological and motor complications [7]. Myelomeningocele is characterized by the exposure of neural tissue to amniotic fluid, which causes continuous damage to nerve fibers and results in motor and sensory deficits [7]. Studies indicate that prolonged exposure of neural tissue not only causes direct injury but also impairs the overall development of the nervous system, aggravating the sequelae throughout pregnancy [8]. In the present case, intrauterine intervention allowed successful correction of spinal dysraphism and the absence of evident lesions at birth.

Myelomeningocele presents lower rates of hydrocephalus and less need for ventricular shunt after birth, in addition to a greater probability of independent motor development [7]. These findings indicate that fetal surgery can prevent the progression of neurological lesions, enhancing neuromuscular

development and future quality of life. The infant in this report, now 10 months old, remains under follow-up at the service and has not shown signs of hydrocephalus, without neurological impairment.

In a recent systematic review shows great variance in the anesthetic management during fetal operation procedures, reporting intraoperative fetal monitoring and fetal hormonal responses to external stimuli, are necessary to identify the best anesthetic approach [9]. The choice of balanced general anesthesia associated with continuous epidural was strategic to ensure hemodynamic stability and pain control, both during the procedure and in the postoperative period. General anesthesia was necessary to maintain control of the maternal airway and allow the administration of anesthetic agents that minimize stress to the mother and fetus. The option to use remifentanil in target-controlled continuous infusion ensures adequate intraoperative analgesia and reduces the incidence of respiratory side effects in the postoperative period due to the pharmacokinetic profile of the drug. Continuous epidural analgesia was included to provide a more comfortable recovery and reduce the need for opioids, an important strategy to minimize postoperative side effects, the catheter being removed in the ward.

Fetal anesthesia played a crucial role in the procedure. The use of analgesic agents and neuromuscular blockers directly on the fetus reduces stress responses and prevents movements that could compromise the exposure and correction of the myelomeningocele, ensuring safety for the nervous tissue during the intervention [10]. For intrauterine cardiac surgery, a detailed fetal echocardiographic assessment is essential to identify the appropriate fetal cardiac interventions candidate is essential [11]. In the present case, there was no diagnosis of cardiac alterations by US, which is why echocardiography was not performed.

Pancuronium and fentanyl were administered intramuscularly in the fetal gluteus using a 30x7mm needle. The surgery was performed uneventfully, and intermittent monitoring of the fetal heartbeat did not reveal the need for intervention in the hemodynamics of the fetus, which remained stable and immobile throughout the procedure [12].

The management of tocolysis with continuous terbutaline and the use of vasopressors to control maternal blood pressure were equally essential to avoid complications, such as premature labor and hypotension, which could compromise the supply of oxygen and nutrients to the fetus [13,14]. In this case, a single bolus of norepinephrine was necessary to correct a momentary episode of hypotension observed shortly after anesthetic induction. The combination of continuous terbutaline with sevoflurane at 2 MAC was sufficient to maintain uterine relaxation and prevent the occurrence of premature labor.

Intrauterine correction of myelomeningocele decreases the incidence of hindbrain herniation and shunt-dependent hydrocephalus in children with spina bifida [15]. Intrauterine repair of myelomeningocele offers a unique window of intervention before birth, allowing the fetal nervous system to be protected and progressive neurological deficits to be mitigated. Scientific evidence suggests that this intervention results in better prognoses compared with neonatal repair, promoting more adequate motor function, in addition to a reduced need for postnatal interventions [16]. Intrauterine surgery between the 24th and 26th week of gestation is essential for the success of this surgery.

4. Conclusion

Repair of the neural tube defect takes place between 24 and 26 weeks' gestation to allow adequate time for the fetal repair to be effective [13]. In this case, the procedure was performed at 25 weeks of gestation, resulting in birth without neurological impairment. Surgery is performed by obstetricians and paediatric neurosurgeons. Conduct of anesthesia for the mother should be scheduled before induction, during the procedure and postoperatively. On the other hand, anesthetic conduct for the fetus involves not only that received via the maternal circulation but, before the repair, the fetus receives an intramuscular injection of fentanyl, neuromuscular blocker and atropine to achieve anesthesia, analgesia, homeostasis and immobility. The development of anesthetic techniques in this area should focus on minimizing maternal risk and preserving normal neural development of the fetus.

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