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Case Report

Peripartum Anesthetic Management of Patient with Osteogenesis Imperfecta: Case Report and Literature Review

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Introduction

Osteogenesis Imperfect (OI) (Brittle Bones Disease) is the most common hereditary connective tissue disease. The main clinical forms of OI are quantitative and qualitative anomalies of type I collagen, the most common protein in bones [8].

In modern literature, cases of successful delivery of patients with imperfect osteogenesis are described, however this disease is considered a contraindication to pregnancy, and when it is advised, an artificial interruption is recommended. Such tactic is associated with the occurrence of a significant number of complications, during pregnancy and delivery, sometimes fatal for both the mother and the fetus [8].

Caesarean section is conducted more often in such patients due to maternal pelvic deformity, cephalopelvic disproportion, abnormal fetal position variants [1,8]. In literature, there are few cases of an anesthetic aid for Cesarean Section (CS) in patients with OI (types 3-4).

The clinical observation that is presented, describes features of labor management of a patient with OI type IV.

Case Report

Patient B. 35 years old was admitted to the National Medical Research Center of Obstetrics, Gynecology and Perinatology named after Academician V.I. Kulakov with a diagnosis of pregnancy 31-32 week. Head previa. OI type 4. Condition of multiple fractures from the age of 2.

Pregnancy first. Has come spontaneously. In the I trimester – mild pregnancy anemia. In the II trimester, correction of anemia of the pregnant with iron preparations (Maltofer). At 24 weeks in-patient treatment of the threat of preterm birth was carried out at the place of residency.01/09/2017 An attempt to abort pregnancy. There were 3 unsuccessful attempts to incubate the trachea using endoscopic equipment. Further attempts of abortion were declined due to the fear of the patient for her life. III trimester - threatening premature birth.

In the aftermath, she was repeatedly consulted by the Center's specialists through telemedicine communications.

For delivery, the patient was admitted to the National Medical Research Center of Obstetrics, Gynecology and Perinatology named after Academician V. I. Kulakov on 09.11.2017. Objectively: the condition was satisfactory, no complaints. Disproportional short stature (height - 92 cm, weight - 23.5 kg). Cannot move independently. Sharply intensified thoracic kyphosis and lumbar lordosis. The general muscular tonus is lowered. Limitation of movement in all limbs. Triangular shaped skull, lower jaw is underdeveloped. Sclera of a blue hue. Airway assessment revealed bad mouth opening (Mallampati class 4) and bad dentition.

There are no respiratory and hemodynamic disorders. In the clinical analysis of blood mild anemia was observed: (hemoglobin 109g/l) other indicators within the limits of the norm. In the biochemical analysis of the blood, moderate hypoproteinemia was observed (Protein 54.9g/L), urine analysis, an ECG - without pathology.

Genetic consultation: the form of the disease is hereditary (Mother – OI type 4).

An ultrasound of the fetus revealed no abnormalities. The fetus corresponds to the 31st week of gestation. Pelvic presentation, transverse position of the fetus. Normal fetoplacental and uteroplacental blood flow.

The indication for operative labor was the increase of dyspnoea at night of the 35-year-old primipara with severe extragenital pathology: OI type IV and transverse position of the fetus. Considering the severe extragenital pathology and the wishes of the woman, a ligation of the fallopian tubes is shown. It was decided to carry out the delivery of the patient by a caesarean section operation in a planned manner.

21/11/2017 a Pfannenstiel incision was conducted, a caesarean section in the lower uterine segment.

Taking into account 3 unsuccessful intubation attempts in the past, it was decided to conduct a cesarean section with a provision of a combined spinal epidural anesthesia, in the presence of endoscopic

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Table 1:

Year	Author	OI type	Age (years)	Weight (kg)	Height (sm)	Gestation (weeks)	Airway Examination	Anesthetic technique	Anesthetic Details
1977 [7]	Bullard	No data							
1984 [9]	Cunningham	No data	27	52	154	38	No data	Epidural	L3-L4.12 ml 0.5% Bupivacaine
1986 [10]	Allanson	No data						GA	No data
1992 [11]	Cho	No data	20	No data	No data	38	No data	No data	No data
2002 [12]	Vogel	4	30	No data	91	24	MP I, no neck flexion or extension, poor dentition	Epidural	Grade II view on DL; LOR at 3 cm; 2% lidocaine 13 mL for T4 block
2002 [12]	Vogel	3	35	No data	86	32	MP II, cervical immobility, TMD 4 cm, poor dentition	GA	Grade III-IV view on DL; awake nasal fiberoptic intubation conducted
2006 [13]	Rudlof	3	21	No data				SA	No data
2009 [14]	Hwang	4	28	41	116	36	MP II, normal neck extension, TMD 3 finger breadths, poor dentition	SA	Bupivacaine 8 mg produced T4 block in 5 min
2010 [15]	Murray	3	24	25	91	35	MP I, normal mouth opening, normal dentition	CSE	L3-4, LOR 3.5 cm, hyperbaric bupivacaine 4 mg with fentanyl 5 μg produced T12 block after 15 min; 0.5% bupivacaine 5 mL produced T4 block
2010 [16]	Lyra	Mild Form	23	No data	No data	38	No data	SA	L3-L4 0,5% hyperbaric bupivacaine 10 mg + Morphini 60 μg
2011 [17]	Fiegel	3	27	36	130	31	MP 2, limited neck mobility, normal bite , poor dentition.	GA	RSI, LOR.
2015 [18]	Dinges	3	37	36	111	32	MP 4, mouth opening3 sm, limited neck mobility , protruding mandibula .	GA	Video laryngoscopy with difficulties Induction: remifentanil, propofol, rocuronium 35 mg. maintenance of anesthesia : nitrous oxide + sevoflurane .
2015 [18]	Dinges	3	37	42	92	28	MP3, Mouth opening 4 sm, normal neck extension	CSE	L3-L4 with ultrasound navigation. hyperbaric bupivacaine 5 mg + Fentanil 10 mcg; 2 Lidocaini 2% bolus 3 ml + bicarbonate 0.2 mEq/ml + Epinephrine 5 μg/ml – produed T4 block.

service.

The initial hemodynamic parameters of the patient: BP 140/78 mm Hg. Heart rate at 104 beats/minute.

Premedication - Dexamethasone 2 mg.

With the patient on the left side, under local anesthesia Sol. Lidocaini 2% - 3 ml, an 18-gauge Tuohy needle was inserted at L2-3 and, using the loss of resistance to saline, the epidural space was identified at 3.0 cm. A 27-gauge pencil-point spinal needle was inserted through the Tuohy needle and after clear cerebrospinal fluid was observed, hyperbaric bupivacaine 0.5% 4 mg was injected. A catheter was then threaded 2 cm into the epidural space. After 7 min sensory block had reached T11 - T12.

Medication: 250 mg Tranexamic acid for bleeding prophylaxis, uterotonic medication - Oxytocin 4 IU, infusion therapy - 380 ml saline, constant norepinephrine infusion at a dosage of 0.01 to 0.08 μg/kg/min was used to maintain hemodynamics.

Surgery was allowed to be proceeded and a female infant weighing 1800 g was delivered with Apgar scores of 7 at 1, 7 at 5 min.

The total duration of the operation was 30 minutes.

Postoperative pain was managed initially with epidural boluses of Ropivacaini 0.2% 2-3 ml and with regular i/v injections of tramadol and dexketoprophen.

The antibacterial prophylaxis was conducted intraoperatively after clamping the umbilical cord -Amoxiclav1.2 g.

Mother and baby were discharged home after seven days.

The child was advised by a geneticist - a blood test was conducted to find mutations in the genes COL1A1, COL1A2, 27.11.17 - no

mutations were detected. Blood was taken for genetic research (targeted sequencing) - the result was negative.

Discussion

The frequency of OI is approximately 1 per 20,000 newborns. People with OI caused by mutations of collagen have a 50% risk of transmitting the disease to the child. The proportion of cases caused by the de novo mutation varies depending on the severity of the disease [1,2].

Osteogenesis imperfect is divided into 8 types, the most common are 1-, 2-, 3- and 4-th. The first type of OI is the lightest and most common form of the disease, after which by severity of complications follow 2-, 3- and 4-th types. Recently, types 5, 6, 7 and 8 have been classified, which have the same clinical features as the 4th, but each has unique histological and genetic data [3,4,5].

Along with a high frequency of bone fracture osteogenesis imperfecta is also characteristic of otherclinical manifestations: short stature, scoliosis, a triangular face configuration (large arch of the palate, small lower jaw), deformation of the skull, hearing loss, blue sclera due to reduced collagen content, resulting in a visible pigmentation of the choroid, incomplete tooth formation, loss of connective tissue of ligament apparatus and the heart valves [1].

We found 13 cases of an anesthetic aid for CS in patients with OI (types 3-4) from which: 4 - general anesthesia; 2 combined spinal epidural anesthesia; 2 - epidural anesthesia; 3 - spinal anesthesia (Table 1).

We are describing the case of delivery of a patient with type OI type 4 through caesarean section. Because of the increase of the size of the pregnant woman's womb with OI, she does not usually reach full-term pregnancy and requires an early cesarean section. Pregnancy negatively affects the strongly altered organism of such patients, which is associated with a number of complications and requires a careful consideration of the variant of anesthesia. [19].

With severe deformations of the skeleton of the mother or fetus, with a significant disparity between the fetal head and the mother's pelvis, and in cases of severe pain in the joints and bones of the mother, in most cases (54%) a cesarean section is conducted. [3,10] Some authors consider it possible in patients with OI to maintain spontaneous delivery [19,20].

Cases of uterine ruptures in patients with the 1st type of OI are described [4,5], which may be due to the decreased collagen content in uterine myometrium in comparison with the healthy pregnant women [6]. Complications that develop during gestation include bleeding, bone fractures, uterine ruptures, uterine atony, uterine bleeding, hypothyroidism, aortic aneurysm ruptures. Regardless of the type of imperfect osteogenesis, pains in the back area are most common during pregnancy or after childbirth [3].

Patients with OI should undergo genetic counseling and prenatal diagnosis. Pregnancy during this disease is contraindicated [16].

Preparation for childbirth takes into account the risk of anesthesia.

With patients with OI, caution is necessary during the intubation of the trachea to avoid injury and fractures of the vertebrae, lower jaw and teeth, due to the decrease in the height of the cervical vertebrae, malformation of the teeth, and underdevelopment of the upper jaw. If difficulties arise during the visualization of the larynx, the use of the fibrooptic technique is recommended [6].

Regional anesthesia is limited by the degree of kyphoscoliosis, the problems that arise when creating the necessary position of the body, during puncture due to previous fractures of the ribs and spine, and also by the small height of the patients [6].

In the postpartum period there can be a tendency to bleeding, an increased risk of spinal hematoma [6]. Need to be particularly attentive to the possible hemorrhagic complications as patients with OI prone to spontaneous non-traumatic subdural hematoma [21], even in the case of normal hemostasis and normal platelet counts. [22] In this regard, we recommend that additional hemostasis tests should be performed before choosing an anesthetic method.

Conclusion

Given the severity and variety of clinical manifestations in patients with OI type 4, anesthesia should be adapted to each individual case.

In the absence of contraindications, CSE combines the benefits of SA with EA capabilities. A contingency plan for general anesthesia should always be prepared, if neuroaxial anesthesia is not possible.

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