

PERIOPERATIVE MANAGEMENT FOR WOMAN AFFECTED BY OSTEOPENIA SUBJECTED TO CAESAREAN SECTION

Tommaso Ciacca; Marco Trippetti; Laura Donati*

U.O. Anesthesia and Intensive Care Hospital of Spoleto, Italy

*U.O. Obstetric and Gynecology Hospital of Spoleto, Italy

Indirizzo per corrispondenza: Dott. Tommaso Ciacca

U.O. Anesthesia and Intensive Care Hospital of Spoleto

via Silvio Pellico 2, 06126 Perugia (Pg) Italia

tel: +39 07536226; Fax: +39 0763 307345; e-mail: tommaso.ciacca@libero.it

ABSTRACT

We report the case of a woman affected by Osteogenesis Imperfecta (OI) subjected to caesarean section in our maternity hospital. We describe the anaesthetic and obstetric reasons which suggest the perioperative way that had ardly engaged the team to have good result and satisfaction for the patient.

Key words: *Osteogenesis Imperfecta; Caesarean Section*

RIASSUNTO

In questo articolo viene descritto l'approccio ostetrico e anestesiológico relativamente al caso clinico di una gestante affetta da osteogenesi imperfetta sottoposta a taglio cesareo effettuato alla trentaquattresima settimana di gravidanza. La paziente ha una forma di osteogenesi imperfetta caratterizzata da bassa statura (altezza di 112 cm), severa cifoscoliosi, osteoporosi diffusa, pregresse fratture multiple, prollasso mitralico. In tali condizioni, che certamente rappresentano una rarità, è necessaria una precoce valutazione e la massima collaborazione tra i professionisti coinvolti che devono mettere a punto un piano operativo condiviso. Non sono riportati molti casi analoghi in letteratura e con la descrizione di questo caso clinico si offre l'opportunità per un approfondimento sulle difficoltà che possono presentarsi nel prendersi in carico gestanti affette da osteogenesi imperfetta, prevenendo rischi e complicanze della fase peroperatoria.

Parole chiave: *Osteogenesi Imperfetta; Taglio Cesareo*

BACKGROUND

Osteogenesis Imperfecta (OI) is a rare inherited disease where the basic pathology is a defective maturation of collagen, resulting in very fragile bones and multiple fractures.

Depending on the inheritance pattern of OI, the disease can result in a spectrum of severity.

According to recent data the prevalence of OI in Europe is 6.5/100.000 (1).

Main abnormalities associated with this disease include: short stature, kyphoscoliosis, decreased bone density; other symptoms included: blue sclera, poor dentition, conducting hearing loss, coagulative problems. In many patients can be observed involvement of other connective tissues : thin skin,

joint laxity, congenital heart disease, valvular heart disease, cor pulmonale.

For unclear reasons some patients develop increased metabolism resulting in hyperthermia, hyperhydrosis, elevated tiroxine levels (2).

OI is more common in women, and pregnant patients with OI. need careful antenatal care in a multidisciplinary approach. Preterm delivery by a caesarean section is the method of choice.

The preoperative anaesthetic management includes early clinical evaluation of the patient in order to decide further examinations , the best anaesthetic techniques, the best post-partum monitoring.

Precautionary measures are required in order to avoid bone fractures during placing on the bed, difficulties in tracheal intubation, possible heart

and pulmonary problems, possible inadvertent dural puncture during regional anaesthetic technique cause of kyphoskoliosis.

Patients affected by OI seem to be at risk for increased body temperature and metabolic acidosis in perioperative time, but both features are not correlated to malignant hyperthermia (3).

In literature there are some cases of pregnant women affected by OI underwent a caesarean section under general or regional anesthesia. (4-5-6)

CASE REPORT AND DISCUSSION

A 43 year-old , gravida I, with severe type III O.I. presented at our Obstetric and Gynaecologic Department for her first observation at 16 weeks gestation believing instead to be in menopause.

The patient underwent amniocentesis for alpha-fetoprotein (NTDs), karyotyping, and some genetic diseases such as cystic fibrosis, x-frangible syndrome, muscle dystrophy, deaf-mutism, and research for gene 1a1 of type 1 a OI. All results were negative, serial ultrasound scanning showed a regular fetal growth (50° centile) excluding anatomic anomalies.

At 30 weeks gestation she was visited by our anaesthetist for an early, appropriate evaluation: on physical examination she presented short size (she was 110 cm tall and weighted 48 Kg , 10 kg more than her habitual weight), kiphoskoliosis, osteoporosis, mitrale prolapse. She had personal history of multiple fractures during childhood followed by orthopaedic intervention. Pulmonary function tests at midtrimester revealed significant restrictive disease, sinus tachycardia (mean heart rate of 120 bpm), discomfort for supine position , dyspnoea. Ecocardiographic scan confirmed mitral valve prolapse with moderate valve insufficiency. She was on treatment with pindolo 2.5 mg/daily. Patient was fully collaborative and had no important difficulties in deambulation; her haematology and coagulative study resulted regular. She denied any adverse reaction to previous anaesthetics. A 34 weeks gestation an elective caesarean section was decided cause of increasing respiratory and deambulatory difficulties of the woman, who was forced in the last days to sleep in a sitted position.

Betamethasone 12 mg i.m was administrated during gestation in order to accelerate fetal lung maturity. Generally patients affected by OI have difficulties for ventilation and endotracheal intubation (7-8-9).

In this particular case the patient had a Mallam-

pati –class II airway, a 6 cm thyromental distance, a quite good dentition with intercisor distance of 3,5 cm. The anaesthesiologic choice is based on following reasons: 1) patient was at that time of gestation unable to tolerate supine position cause of severe respiratory distress 2) possible difficulties in performing perimedullar block that could be extended unexpectedly high so to decrease respiratory compliance 3) hemodynamic unbalance in tachycardic patient with symptomatic caval compression 4) possible surgical difficulties and complications 5) better hemodynamic and respiratory control through general approach 6) increased anxiety of our patient.

Preoperatively 500 ml of lactated Ringer's solution, metoclopramide 10 mg iv, ranitidine 50 mg iv and antibiotic prophylaxis for bacterial endocarditis were administered. Patient was positioned in a modified supine position with her head elevated approximately 30 °, and preoxygenation for 10 minutes was started. General anaesthesia was induced with sodium thiopental 200 mg, and Rocuronio 35 mg and maintained with Sevoflorane 1,2 MAC and N2O (50%); ventilatory support was instituted using tidal volumes of 270 ml at 13 breath rate /min. Intubation was performed with a 7 cuffed endotracheal tube. Intraoperative monitoring included ECG, NIMB, Sat o2, body temperature monitoring with endoesophageal sensor .

For the particular contracted pelvic size of the patient obstetrician decided to perform an infraumbilical midline vertical incision and to allow the quick delivery of the infant without difficulties and fundal pressure the so called classical incision was performed (vertical incision into the body of the uterus above the lower uterine segment and reaching the uterine fundus).

A baby girl 2050 gr in breech presentation was extracted , apgar 9/9. In addition to oxytocin (20U/L normal saline), methylergonovine was administered for control of uterine atony and post-partum hemorrhage.

During operation cadiorespiratory parameters were stable except a brief episode of decreased saturation at 92%. Intraoperative EGA was performed. After 55 minutes the patient was brought to intensive care unite with intubated trachea and she was extubated after one hour, according with extubation criteria. After 12 hours patient was transferred to her recovery room, no maternal complications occurred and she was discharged home 6 days after caesarean delivery.

CONCLUSION

Cases of pregnant women affected by OI are rare and their management requires full collaboration between anaesthetists and obstetricians in order to decide the best approach to the patient, considering level of ex-

perience of both teams, and the severity of conditions of patient (skeletal anomalies, hemodynamic and respiratory state, possible difficulties for intubation and ventilation ..). Perioperative monitoring is very important and a period of post-operative observation in intensive care unit must be considered.

REFERENCES

1. http://www.orpha.net/orphacom/cahiers/docs/IT/Prevalenza_delle_malattie_rare_in_ordine_decrescente_o_casi.pdf
2. Harrison, *Principi di Medicina Interna*, Vol. 2, pag. 2481-2483 Ed. italiana della 12ª edizione originale, maggio 1992
3. Gregory A. et al. Perioperative Care of the Patient With Osteogenesis Imperfecta, *Orthopedics* 2007;30:1043
4. Vogel TM et al. Pregnancy complicated by severe osteogenesis imperfecta: a report two cases. *Anesth Analg* 2002;94:1315-1317
5. Rudolf B. et al. Anesthesia for caesarean section in a patient with Lobstein's syndrome *Anaesthesist* 2006;55:655-9.
6. Yeo ST et al. Regional anaesthesia for multiple caesarean sections in a parturient with osteogenesis imperfecta. *Int J Obstet Anaesth* 1999; 8:284-7
7. Rodrigo C Anesthesia for Maxillary and Mandibular Osteotomies in Osteogenesis Imperfecta. *Anesth Prog* 1995; 42:17-20
8. Kostopanagiotou et al. Anaesthesia using a laryngeal mask airway in a patient with osteogenesis imperfecta. *Anaesthesia* 2000; 55: 506.
9. Santos ML et al. Airway Management with ProSeal LMA in a Patient with Osteogenesis Imperfecta. *Anesthesia & Analgesia* 2006; 103: 794.