Osteogenesis Imperfecta In Pregnancy: Case Report

Maryam Rabiee; M.D.¹, Mahin Rasi Etemadi; M.D.²

1 Department of gynecology and obstetrics, Faculty of medicine, Shahed university, Tehran, Iran
2 Department of gynecology and obstetrics, Faculty of medicine, Shahed university, Zaynab Hospital, Tehran, Iran

Received September 2010; Revised and accepted February 2011

Abstract
Osteogenesis imperfecta is a rare inherited Connective tissue disorder with an expression that varies from mild to severe disease affecting bone, Sclera and middle ear. Fertility is preserved, especially in those patients with type 1. We present hereby a pregnant woman with Osteogenesis imperfecta that had over 30 fractures in long bones and vertebrae. The object of this report was to determine choice of delivery method, maternal and neonatal Complications and prenatal diagnosis.

Keywords: Osteogenesis imperfect, Osteogenesis imperfect in pregnancy, Complications in osteogenesis imperfect, Anesthesia in osteogenesis imperfecta

Introduction
Osteogenesis imperfecta is a rare inherited Connective tissue disease with incidence between $\frac{1}{20000} - \frac{1}{60000}$. It is thought to be secondary to mutations within the genome that encodes for type I collagen (1). The most Common Classification for OI was developed by silence and Colleagues divided OI to four types. Type I is the most common and the mildest form. In this type of OI, The sclera is blue and most fractures occur before puberty. Vertebral fractures are typical and can lead to mild scoliosis. Connective tissue malfunction, such as thin skin, hernias and generalized Joint hyper mobility are present.

Type II is lethal in uterus or shortly after birth. Types III and IV are intermediate in severity between types I and II (2, 3) Fertility is preserved, especially in those patients with type I of disease and pregnancy can be carried to term. Patient with OI suffer from skeletal deformities and chronic pain secondary to repeat fractures. The physiological changes of pregnancy also predispose to musculoskeletal pain. Vertebral fractures occur in 4.2% of pregnant OI female (1, 3, 4).

The risk of hypertension has been highlighted in non–pregnant patients with OI, but the risk of pre–eclampsia in OI Patients has not been adequately assessed. Mode of delivery remains Controversial and should be individualized. Uterine rupture and atony may be accurate in pregnant OI (1, 5, 6). In this paper we describe a Case of pregnant OI whom admitted to the maternity ward on the 39th weeks with labor Contractions.

Case report
A 27–year–old woman, primi–gravida with OI (probably type I) with a background history of over 30 long bones (2 fractures in head of right femur) and Vertebral Fractures, reduce spinal mobility, intermittent back
pain, mild deformity in hand and legs with blue Sclera admitted in educational Zeynab hospital of Tehran. She was on 39th of pregnancy and had good Contractions. In pelvic examination, 4 cm dilatation, 60% effacement, –2 Cephalic station, intact membrane with good inlet, mid and outlet of pelvic were defected. She delivered after 6 hours pain without any stimulation of labor and application instrument. A girl newborn with \( \frac{10}{10} \) apgar in 5 and 10 minute with 3600 weight was born. Uterine rupture was assessed and for preventing of uterine atony, 30 unites oxytocine that was added to 1 liter ringer serum, was given to her.

Mother and newborn was assessed carefully for 48 hours after delivery but we didn’t find any new fracture in them. In past medical history, the antenatal ultrasonic scans revealed no fetal fracture and abnormality. Aminiocentesis in order to exclude fetal OI was offered her and declined. In her families, her father and brother suffered as same as disease.

She was used to eat 1000 mg Calcium plus 800 IV vit. D daily from her childhood.

**Discussion**

Three findings emerged from this study:

There was a high incidence of fetus involvement, Complications associated with pregnant women with OI, and finally what is the best mode of delivery in them. Type I is the most common, and is inherited as autosomal dominant trait with variable expressivity.

Prenatal diagnosis of affected fetus can be based on chorionic villous samples and amniotic fluid test. Affected fetus may be having reduced bone density, multiple bone fractures and skeletal deformity.

In lethal and more severe type of OI, other findings may occasionally be found including polyhydramnios, oligohydramnious, hydropsfetalis and small for gestational age (2, 7, 8). In our case, prenatal diagnosis of fetus by amniocentesis was refused by parents.

Fortunately, we hadn’t any report about reducing bone density and not fracture in fetus of our patient.

Patients with OI suffer from skeletal deformities and chronic pain secondary to repeat fractures (Figure 1). The physiological changes of pregnancy also predispose to musculoskeletal pain. One study reported severe back pain causing disruption to the daily life in up to 13.1 % of OI pregnancies but the risk of fractures isn’t increased during pregnancy (9).

Our patient had mild back pain and scoliosis but she hadn’t new fractures during pregnancy thus she did not report severe back pain causing disruption to daily life. On the other band, In some studies, the risk of pre–eclampsia in OI patient was reported 20% (1) but our paint hadn’t any increase blood pressure during pregnancy. Out of the most important issues in counseling parents of fetuses suspected of having OI is whether Cesarean delivery improves survival and decreases morbidity for the infant. In mild forms where fractures are infrequent, Cesarean delivery might give little protection against fractures.

Of course, if a severe but non lethal form of OI is suspected, delivery in a tertiary center is recommended. Method of delivery should be based on obstetric Considerations. In vaginal delivery, instrumentation probably should be minimized (7, 9, 10).

If cesarean section was chosen, the anesthesia are multi–factorial and early antenatal assessment is required. Bon fragility, scoliosis with restrictive lung disease, unstable cervical spine, micrognathia, brittle teeth and malignant hyperthermia are the most problems that anesthesiologist must be attend them. Uterine rupture and atony may be accrue in patients with OI (1, 4, 11, 12). Our patient had not any complications after delivery. We visited her the day and 3 weeks after delivery. She hadn’t any new fractures in hers and her fetus and her episiotomy was normal without any disruption.

**Conclusion**

In general, pregnancy in women with OI carries a high risk and should be managed by a multidisciplinary team. Patients need to be a ware of the risks, so that they can balance them against the benefit of having a
Osteogenesis Imperfecta in pregnancy

child mode of delivery usually depends on obstetric indications and we remind that Anastasia in this people have some difficulties and complication.

References
