

# Cesarean Section of a 25 3/7 Week Parturient with Fibrodysplasia Ossificans Progressiva: A Case Report

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## Abstract

Fibrodysplasia ossificans progressiva (FOP) is a rare autosomal dominant disease characterized by progressive ossification of tendons, ligaments, fascia, and skeletal muscle. Ossification can occur either spontaneously or as a result of tissue trauma beginning in childhood. Joints typically ankylose and patients frequently become immobile in their 20s. Patient's frequently die before their 40s because of restrictive pulmonary failure as a result of intercostal muscle involvement.(1) The temporomandibular joint (TMJ) and cervical spine also often ossify resulting in immobilization of the jaw and neck, making traditional airway management impossible.(2) Although there are case reports relating to anesthesia for dental procedures, there are no case reports related to the anesthesia care of a parturient with FOP. Our patient is a 24 y.o. G1P0 who presented at 22 weeks for a prenatal consultation. Initial evaluation revealed a 5'0" tall, 58 kg female with a fetus developing appropriately. The patient was successfully delivered at 25 3/7 weeks under GETA, with awake nasal intubation, due to preterm labor and vaginal bleeding. Particular attention was made to prevent further muscular trauma to avoid exacerbation of the disease.

## INTRODUCTION

Fibrodysplasia ossificans progressiva (FOP) is a rare autosomal dominant disease characterized by progressive ossification of tendons, ligaments, fascia, and skeletal muscle. Beginning in childhood, ossification can occur either spontaneously or as a result of tissue trauma. (3) Spontaneous flare-ups present as large, painful swellings resulting in heterotopic ossification. Joints typically ankylose and patients frequently become immobile in their 20s. Affected individuals frequently die before their 40s because of restrictive pulmonary failure and cor pulmonale as a result of intercostal muscle involvement.(1) There are currently fewer than 150 known patients with this disease in the United States. (4)

Individuals with FOP will frequently have ossification of their temporomandibular joint (TMJ) resulting in jaw immobility. Additionally, these patients often develop severely decreased range of motion in their neck. Spinal and epidural anesthesia should be avoided in the patient with FOP. Their scoliosis and paravertebral ossification make placement difficult. Also, trauma from needle placement can lead to further progression of their disease and lead to additional painful ossification. (5) Finally, with severely limited TMJ range of motion, conventional emergency airway access techniques can be difficult or impossible. (6)

## CASE REPORT

A 24yo Caucasian G2 P0010 presented at 22 weeks EGA for prenatal care after being referred from a community hospital. The patient weighed 58 kg and had significant musculoskeletal abnormalities including: severe levokyphoscoliosis causing severe restrictive pulmonary disease; a fused temporomandibular joint; a fused and flexed neck; fixation of her shoulders; as well as fused contractures of the elbows, knees, hips; and left wrist. Mouth opening was approximately 1 cm at the widest point.

Peripartum testing included pulmonary function tests as well as transthoracic echocardiography (TTE). The TTE revealed an essentially normal exam with an EF of 55-60%, but was limited by patient positioning. Pulmonary function tests were consistent with severe restrictive lung disease. All other prenatal testing was normal. A multidisciplinary team including obstetricians, obstetrical anesthesiology faculty, an ENT surgeon created a comprehensive plan for Cesarean delivery and bilateral tubal ligation. Vaginal delivery was not an option given the patient's relative hip immobility. General anesthesia with awake fiberoptic intubation was deemed the safest method of anesthesia. Regional anesthesia was to be avoided due to concerns of additional spinal ossification and inability to secure the airway in an emergency. At this time our preoperative concerns included:

difficulty obtaining an airway, difficulty with ventilation, difficulty with positioning and prevention of trauma or ossification, difficulty and risk vs. benefit of IV and arterial access, unknown surgical conditions and potential for increased length of surgery and bleeding.

**Figure 1**

Figure 1. X-ray obtained of patient prior to her pregnancy. Note the severe kyphoscoliosis and the heterotopic calcifications present throughout her body.



At 25 4/7 weeks EGA, the patient presented with contractions and cervical dilation to 1 cm. The patient's contractions were tocolyzed using subcutaneous terbutaline and oral indomethacin. Oral dexamethasone was administered to promote fetal lung maturity. This treatment plan was successful for 3 days. However, on hospital day 4, the patient began to have active contractions and cervical dilation to 6 cm. Additionally, the patient experienced approximately 200 mL vaginal bleeding. The patient was transferred to the operating room for a classical cesarean section with bilateral tubal ligation.

The patient's spinal deformity and contractures presented unique positioning challenges. In FOP any prolonged pressure can cause further ossification. All operating tables at Grady Memorial Hospital are equipped with thick three inch soft foam pads, but are flat. Our patient's deformity necessitated that we pad raised areas to minimize pressure

points and create as level of an operative site as possible. Disposable foam padding was cut to stabilize the patient's contractures and deformities in a neutral state. To further balance safety and minimization of ossification, non-invasive blood pressure monitoring was started at 5 minute intervals, additional IVs and invasive monitors were avoided until necessary.

General anesthesia and awake nasal fiberoptic intubation was performed. IV glycopyrolate was administered to decrease oral secretions. The nares were topicalized and vasoconstricted using lidocaine and oxymetazoline spray. Additionally, the oropharynx was topicalized using nebulized lidocaine via a facemask atomizer. Finally, vocal cord topicalization and additional trachea topicalization was performed by injecting lidocaine through the bronchoscope sideport once the vocal cords were visualized. After the endotracheal tube was placed and positive CO2 was confirmed, propofol and rocuronium were administered. Transtracheal and other typical nerve blocks were avoided to prevent ossification.

The surgery was performed through a Pfannensteil skin incision, and a classical cesarean uterine incision was made. There was difficulty obtaining abdominal entry and uterine visualization due to calcified connective tissues which resulted in the rectus muscles having the appearance of being tense despite adequate neuromuscular blockade. The muscles were also significantly more difficult to dissect due to ossification. A 760 gm male infant was born with 1 and 5 minute Apgar scores of 1 and 8 respectively, 25 minutes after skin incision. Uterine atony was noted following delivery and increasing amounts of oxytocin were administered as well as methylergonovine. Total estimated blood loss was 2,800 mL after 4 hours and 7 minutes of surgery. This was attributed to uterine hemorrhage secondary to atony, and resultant coagulopathy. Five liters of Lactated Ringers, 3 units of Packed Red Blood Cells, and 2 Units of FFP were administered Total urine output was 550 mL. Methylprednisolone 125mg was given shortly after the baby was removed to help prevent postoperative ossification. The patient remained intubated overnight and was successfully extubated the following day. She was discharged on POD#6 and recovered uneventfully.

**DISCUSSION**

Fibrodysplasia ossificans progressive is an exceptionally rare autosomal dominant disorder, estimated to occur at a frequency of 1 case per 1.64 million births.(1) There are an

estimated 150 patients in the United States with FOP and anesthetic literature is limited mainly to case reports of dental restoration. Ossification frequently begins in the cervical and upper paravertebral muscles, eventually immobilizing all major joints. (1)

Successful delivery of the patient with fibrodysplasia ossificans progressiva is dependent on thorough preoperative preparation. Because the patient presented early in the 2<sup>nd</sup> trimester, we were able to perform several diagnostic tests to guide our further management. Pulmonary function testing quantified the degree of restrictive lung disease caused by her profound scoliosis and intercostal muscle calcification. Transthoracic echocardiography evaluated myocardial function and looked for evidence of right heart failure secondary to the patient's severe restrictive lung disease.

Intraoperatively, three units of packed red blood cells were required due to blood loss of approximately 2800mL. Inability to exteriorize the uterus following delivery and resistant uterine atony despite multiple doses of oxytocin was the likely cause. Several case reports describe calcification at sites of I.M. injections following routine immunizations in children. For this reason, our team insisted on avoiding traditional I.M. injections of Methergine and Hemabate. The uterus however was expected to become calcified after completion of the surgery. Consequently, the Obstetricians injected Methergine directly into the uterus to treat the continued uterine atony. Alternatively, intramuscular injection could have been performed in the patient's thighs since she had preexisting mobility limitations and further ossification would not have added additional morbidity in our patient.

Because safely securing the airway is paramount for the provision of anesthesia to the parturient with FOP, a nasal fiberoptic intubation was performed. Laryngeal nerve blocks were not performed out of consideration for additional tissue trauma and desire to avoid worsening her disease. While the literature does not contain reports of any patients experiencing laryngeal muscle calcification, the literature does recommend avoiding tracheostomy to prevent airway obstruction secondary to traumatic calcification.

Surgical time for this patient was approximately 4 hours. This was over four times as long as our average primary general caesarean section by the same surgical service. The obstetricians attributed this to very difficult surgical conditions from an ossified abdomen. The degree of preoperative IV access should be balanced with risk of ossification and possibility of large blood loss. It is important to avoid placement in joints that are still mobile, such as the antecubital fossa and wrist in this case.

Thorough preparation for the parturient with FOP is essential. Considerations include not only the patient's airway, but also respiratory mechanics, potential for ossification with vascular access, difficult surgical conditions, blood pressure monitoring and positioning. A complete preoperative evaluation and optimization before the onset of labor was very helpful. We were fortunate to have adequate time to collaborate with other services and create an appropriate and successful delivery plan.

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