

Anaesthetic strategy required in fibrodysplasia ossificans progressiva

DP Crain

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ABSTRACT

Fibrodysplasia Ossificans Progressiva (FOP) is a very rare hereditary condition that causes extraskeletal connective tissue ossification. By their third decade

of life, many people who have been affected have become fully immobile. Among the many anaesthetic issues, managing the airway of patients with FOP may prove to be the most difficult.

Key Words: Tendons, ligaments, skeletal muscle, connective tissue, regional anaesthesia.

INTRODUCTION

Extraskeletal ossification of connective tissue such as tendons, ligaments, and the connective tissue in skeletal muscle is a rare hereditary condition known as Fibrodysplasia Ossificans Progressiva (FOP). By their third decade of life, affected people are often entirely immobilised. Thoracic insufficiency syndrome can cause difficulties as the disease worsens. The greatest anaesthetic problem in young children, however, may be airway management. Only a few studies have been published on the anaesthetic care of young children with FOP. The anaesthetic care of a three-year-old child with FOP is described in this case report.

Fibrodysplasia ossificans progressiva (FOP) is a severe connective tissue illness marked by progressive heterotopic ossification of skeletal muscles, ligaments, tendons, fascia, and aponeuroses, as well as deformities of the great toes. Heterotopic ossification is preceded by painful repeated episodes of soft tissue swelling (flare-ups). Flare-ups commonly start in the first decade of life, and before heterotopic ossification appears, many individuals with FOP are misdiagnosed as having soft tissue tumours or aggressive fibromatosis [1]. They are sometimes subjected to risky and needless diagnostic treatments that result in heterotopic ossification, which causes irreversible damage and impairment [2]. To avoid additional iatrogenic injury or trauma, early clinical identification and confirmation genetic testing of FOP are critical [3].

Patients with the classic FOP phenotype, which accounts for about 92 percent of cases and all of whom have the canonical *ACVRI/ALK2* gene c.617G>A (p.R206H) mutation, have both congenital malformations of the great toe and progressive HO, as well as other common but variable FOP features such as tibial osteochondromas, conductive hearing impairment, age-dependent increased risk of hypercalcaemia-related nephrolithiasis, sparse hair and eyebrows, and neuroimaging abnormalities especially involving the pontine region.

Differential diagnoses for FOP without hallux deformities (HMO, POH, METCDS), and Brachydactyly Type B1 (BDB1) include Hereditary Multiple Osteochondromas (HMO), Progressive Osseous Heteroplasia (POH), Metachondromatosis (METCDS), and Brachydactyly type B1 (BDB1) (HMO, METCDS and BDB1). Hallux abnormalities can also be caused by solitary congenital malformations or juvenile bunions, as well as tumor-like swellings caused by sarcomas, desmoid tumours, aggressive juvenile fibromatosis, or lymphedema [4-6].

FOP is inherited in an autosomal dominant form, which means it can be genetically counselled. The majority of those affected have simplex instances caused by a *de novo* pathogenic mutation in the *ACVRI/ALK2* gene. Only a small percentage of people with FOP have a parent who is also affected.

Prenatal tests, such as foetal ultrasound, can detect a hallux valgus deformity as early as week 23 of pregnancy. Prenatal testing for a pregnancy at elevated risk and preimplantation genetic testing are both possible if an *ACVRI/ALK2* gene pathogenic variation is detected in an affected family member.

AIRWAY MANAGEMENT

The effects of FOP on The Temporomandibular Joints (TMJs) and the cervical spine cause the majority of the difficulty with airway control in patients with FOP. TMJ ankylosis is frequent in FOP, and jaw overstretching can give enough damage to the TMJs to produce a localised flare-up and additional ossification. Neck stiffness can appear early in the life of a kid with FOP, and it usually occurs before heterotopic bone development. Tall, thin vertebral processes, as well as big pedicles and spinous processes, have been found as anatomical anomalies of the cervical spine in patients with FOP. Early infancy is when the facet joints and spinous processes between the second and seventh cervical vertebrae fuse [7].

To avoid ectopic ossification of the airway, tracheostomy and transtracheal injections should be avoided in FOP patients. However, if a surgical airway rescue technique is required, an otolaryngologist should be present in the operating room.

In patients with FOP, several studies suggest that nasotracheal fiberoptic intubation in an awake or lightly sedated patient is the safest airway management approach. In juvenile patients, this method is frequently impractical. Kilmartin et al. looked at cases where individuals with FOP needed dental treatment and were given general anaesthetic. In practically every adult case, an awake fiberoptic intubation was performed. Fiberoptic intubation was accomplished under anaesthesia in 14 of their 19 paediatric cases. Prior to nasotracheal fiberoptic intubation, four paediatric instances needed induction of anaesthesia with maintenance of spontaneous ventilation. These four individuals were between the ages of 5 and 10[8-13].

VASCULAR ACCESS AND REGIONAL ANAESTHESIA

In FOP, subcutaneous injections and the cautious placement of superficial intravenous catheters are not prohibited. However, the traumatic insertion of intravenous and arterial lines might result in heterotopic bone development. Any type of intramuscular injection should be avoided since it can trigger an immediate flare-up of FOP at the injection site. Although regional anaesthesia is not recommended, using ultrasound guidance to put needles close to superficial nerves without piercing muscle and connective tissue may be possible.

Patients with FOP may find it more challenging to place needles precisely due to musculoskeletal abnormalities. Patients with severe abnormalities

Managing Editor, *Anesthesiology: Case Report*, Berkshire, UK

Correspondence: DP Crain, Managing Editor, *Anesthesiology: Case Report*, Berkshire, UK, E-mail: anesthesiology@scischolarsjournal.com

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should also be carefully positioned on the operating table, with pressure points padded to reduce soft tissue trauma

NATURAL CLINICAL COURSE

Heterotopic ossification in FOP develops gradually over the first ten years of life, with periodic flare-ups in the axial skeleton that are commonly misinterpreted as soft-tissue sarcoma or aggressive juvenile fibromatosis. Flare-ups can happen as a result of a localised invasion mechanism such as trauma or intramuscular injections that cause bruising, and they can be accompanied by feelings of warmth and pain. In FOP, traumatic damage and surgical intervention cause rapid new bone production. Flare-ups can occur without a known cause and can even be triggered by systemic inflammation caused by viral illnesses like influenza [10].

Heterotopic ossification is a highly personalised process. In certain patients, systemic ankyloses cause difficulty walking and respiratory problems as the condition advances. The volume of heterotopic ossifications and the patient's age are both associated to functional impairment as assessed by patient reports. By their third decade of life, the majority of patients are confined to a wheelchair and require permanent support with activities of daily living. Heterotopic ossification in the temporomandibular joint and surrounding areas frequently causes trismus, which makes eating difficult and resulting in significant weight loss.

SKELETAL MALFORMATIONS

FOP patients appear normal at birth; however, they have a range of bone abnormalities. The most common symptoms of this disorder are deformities of the great toes, which are well-known. Before the advent of heterotopic ossification, the great toe is usually shortened and the hallux valgus is present. The second toe's distal interphalangeal joint is normally proximal to the tip of the great toe. The proximal phalanx is continuously reduced on radiographs and sometimes has a triangular form. The medial side of the metatarsal bone is likewise shortened and pointed, diverging the proximal phalanx laterally from the metatarsal axis. As people get older, they notice a fusion of the proximal and distal phalanx.

CONCLUSION

FOP is a condition that is highly rare and can be fatal. Even in small children, the condition can be severe enough to provide major complications to the attending anaesthetist.

A detailed preoperative examination and anaesthetic strategy are required. A multidisciplinary team should manage all patients with FOP.

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