

orphananesthesia

Anaesthesia recommendations for patients suffering from **Fibrodysplasia ossificans progressiva**

Disease name: Fibrodysplasia ossificans progressiva

ICD 10: M61.1

Synonyms: FOP

Fibrodysplasia ossificans progressiva (FOP) is a rare, progressive, and disabling autosomal dominant disorder of extraskeletal endochondral ossification. The diaphragm, tongue, extraocular, and laryngeal muscles are spared from heterotopic ossification; cardiac and smooth muscles are also not affected. Individuals with FOP appear normal at birth except for malformations of the great toes. During the first decade of life, inflammatory soft tissue swellings, also known as flare-ups, transform skeletal muscles and connective tissues into a second skeleton of heterotopic bone that progressively immobilizes all of the joints of the axial and appendicular skeleton. Flare-ups may occur spontaneously, but can be precipitated by misplaced venipuncture, soft tissue injury, muscle fatigue, intramuscular injection, biopsy or excision of heterotopic bone, viral illnesses, routine dental therapy, and injection of local anaesthetics during dental procedures.

Disease flare-ups are episodic; immobility is cumulative. Heterotopic ossification usually begins by ten years of age with neck and shoulder involvement. The temporomandibular joints may be involved early and are vulnerable to trauma at any age. The median lifespan is 40 years; death results most commonly from complications of thoracic insufficiency syndrome or pneumonia. A few deaths have been attributed to complications of general anaesthesia. At the present time, no medical or surgical intervention can alter the natural history of FOP. Anaesthetic management for patients with FOP is challenging. Cervical spine fusion, ankylosis of the temporomandibular joints, thoracic insufficiency syndrome, restrictive chest wall disease, and sensitivity to oral trauma complicate airway management and anaesthesia.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Typical surgery

Numerous anaesthetics for dental rehabilitation, teeth extractions, and oral abscess drainages have been reported. Less common surgeries that have been reported include dilation and curettage, eye evisceration repair, femur and spine fracture correction, hysterectomy, pelvic mass excision, craniotomy for brainstem lesion, and osteomyelitis debridement.

Surgeries undertaken to correct complications caused by heterotopic bone formation are not advised unless the deformity is extraordinarily life or limb threatening. Interventions to correct bony formations and joint deformations often result in worsening of the disease due to reactionary heterotopic bone formation at the surgical sites. FOP patients have, however, undergone successful corrective surgeries for chin-on-chest deformity, thoracic spinal canal stenosis, ulna-carpal bar, and hip ankylosis. These surgeries can be technically challenging and have been known to result in large amounts of blood loss. Furthermore, any surgery where connective tissue is manipulated may result in postoperative heterotopic ossification. Risks and benefits must be carefully weighed before proceeding with surgeries.

Type of anaesthesia

FOP patients often have temporomandibular joint ankylosis and cervical spine fusion; their airways can be difficult to manage. Three cases of perioperative "cannot ventilate, cannot intubate" situations have been reported. Sedation should be performed with extreme caution, and general anaesthesia with an endotracheal tube is recommended. Traditional airway manoeuvres (i.e. jaw thrust) to alleviate obstruction or mask ventilate may not be successful. Laryngeal mask airway management placement may be impossible in cases of limited mouth opening. Additionally, patients with restrictive lung disease may have limited physiological reserve. Additionally, direct laryngoscopy with a blade may traumatize temporomandibular joints and cause a flare-up, which may permanently decrease mouth opening. For the above reasons, an awake nasal fiberoptic intubation before induction of general anaesthesia is advised.

Neuraxial anaesthesia is contraindicated in FOP patients due to the risk of heterotopic ossification in the spinal cord area precipitated by trauma from spinal or epidural needle insertion. Regional anaesthesia is relatively contraindicated; heterotopic ossification surrounding the area of needle placement is a concern. A successful and uncomplicated ultrasound-guided ankle block has been performed in an FOP patient. The anaesthesiologist limited local anaesthetic injection to the epifascial tissue layer. Regional anaesthesia may be a reasonable choice if the technique does not disrupt skeletal muscles or connective tissues, which are susceptible to pathologic ossification.

Routine injections of local anaesthetic for dental procedures, especially mandibular blocks, should not be used because they have been shown to precipitate flare-ups and cause fusion of the temporomandibular joints.

Necessary additional diagnostic procedures (preoperative)

FOP patients are susceptible to restrictive lung disease, pneumonia, thoracic insufficiency syndrome, and right-sided heart failure. Depending on the functional status of the patient and the risk level of the surgery, pulmonary and cardiac evaluations may be necessary to guide management decisions. Chest radiographs, pulmonary function tests, and echocardiograms are commonly obtained. A baseline pulse oximeter reading should be obtained on the day of surgery and before preoperative sedation is administered. A preoperative evaluation by the

anaesthesiologist prior to the day of surgery is advised. The evaluation can impart the need for diagnostic procedures and aid in preparation.

Particular preparation for airway management

There have been three reported cases of anaesthesia providers not being able to ventilate or intubate an FOP patient. The recommendation is that an awake, nasal fiberoptic intubation before induction of general anaesthesia be considered the preferred, primary approach. This recommendation is based on: 1) frequently challenging airway anatomy and 2) the potential risk of trauma and heterotopic ossification of the temporomandibular joints from overstretching of the jaw with direct laryngoscopy.

Successful tracheal intubations after induction of general anaesthesia using a GlideScope® have been reported. These patients had normal mouth opening, and care was taken not to overstretch their jaws. Patients for this technique must be carefully selected. Their normal range of jaw motion should be preoperatively measured preoperatively and not exceeded during airway instrumentation.

Emergency airway equipment should be available at the start of every anaesthetic. An otolaryngologist should be immediately available to assist with airway management during the procedure and to perform an emergency tracheostomy if needed.

Particular preparation for transfusion or administration of blood products

Corrective orthopaedic surgeries may result in increased blood loss.

Particular preparation for anticoagulation

None known or reported.

Particular precautions for positioning, transport or mobilisation

Positioning consideration is essential; patients' bodies are often fused in a rigid position. All pressure points must be padded and the neck supported. If a patient's cervical spine is fused in flexion, steep Trendelenburg positioning is often needed for adequate exposure for a dental procedure (the most common of surgeries). Positioning specifics for Trendelenburg include padding the patients' shoulders and securing the patients to the bed to ensure that their bodies do not shift on the table.

Probable interaction between anaesthetic agents and patient's long term medication

No unique considerations known or reported.

Anaesthesiologic procedure

Succinylcholine should be avoided because of risk of hyperkalemia due to patient immobilization.

A four-day perioperative course of high-dose corticosteroids should be administered and begin prior to the start of the surgery to mitigate heterotopic ossification. A typical dose of prednisone is 2 mg/kg/day (up to 100 mg), administered as a single daily dose. If a patient is not administered a PO dose of prednisone on the morning of surgery, an equivalent dose of IV dexamethasone should be given preoperatively.

Particular or additional monitoring

None known.

Possible complications

Possible complications include:

- 1) Difficult airway management
- 2) Exacerbation of thoracic insufficiency syndrome, restrictive lung disease, or right-sided heart failure
- 3) Heterotopic ossification due to misplaced intravenous catheter, needle trauma, or tracheal intubation
- 4) Heterotopic ossification or neuropathy due to positioning pressure
- 5) Hyperkalemia from succinylcholine administration.

Postoperative care

Patients with severe cardiac or pulmonary disease may not be candidates for surgery or may require intensive care monitoring postoperatively.

Information about emergency-like situations / Differential diagnostics

None known or reported.

Ambulatory anaesthesia

Ambulatory anaesthesia is an option if the surgery is minor and the patient is relatively healthy. Many patients have been safely discharged to home the same day of a general anaesthetic for dental procedures. It is still recommended, however, that the anaesthesiologist be informed of a patient with FOP before the day of surgery and have an opportunity to evaluate the patient. The ambulatory surgery should take place at a tertiary care institution with adequate resources.

Obstetrical anaesthesia

Neuraxial anaesthesia is contraindicated, as previously described in these recommendations. A case of an FOP patient who underwent a Caesarean section for fetal distress under local infiltration with lidocaine has been reported.

Literature and internet links

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