

Anesthetic Management in Fibrodysplasia Ossificans Progressiva: A Case Report

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INTRODUCTION

- Fibrodysplasia ossificans progressiva (FOP) is a rare, progressive, and disabling genetic disorder¹.
- FOP affects 1 per 1-2 million individuals globally².
- FOP is characterized by progressive heterotopic ossification (HO) of skeletal muscle, fascia, tendons and ligaments¹.
- HO typically begins at 10 years of age².
- HO is caused by soft tissue inflammation or “flare-ups” which progressively transform skeletal muscle and connective tissue into a second skeleton².
- Ossification can be spontaneous or the result of minor trauma such as intramuscular injections, soft tissue injury, viral illnesses, or routine dental therapy².
- Individuals typically wheelchair bound by age 30, median life expectancy is 40³.
- Death most commonly from thoracic insufficiency syndrome or pneumonia².
- No available surgical or medical cure¹.
- **Purpose:** Review anesthetic recommendations for managing a patient with FOP and disseminate knowledge to anesthesia practitioners

CASE SUMMARY

- 53-year-old male, physical status 4, history of FOP, asthma, HTN
- Scheduled for robotic bilateral inguinal herniorrhaphy
- Limited mobility, able to ambulate with walker
- No cervical spine range of motion
- Mouth opening approximately 1 cm, nutrition intake via a straw
- Minimal expansion of thoracic cage with deep inhalation
- Airway management: awake nasal fiberoptic intubation
- Intravenous hydrocortisone (100 mg) given intraoperatively and continued for 4 days
- Positioned on operating room table with extra padding and pillows
- Regional anesthesia techniques avoided
- Insufflation with poor view, converted to open procedure
- Unable to reduce herniated bowel, 1 foot resected and right orchiectomy performed
- Sugammadex administered for neuromuscular blockade antagonism
- Fully emerged from anesthesia for extubation; eye opening and following commands, transferred to intensive care unit
- Postoperative course significant for atelectasis
- Discharged postoperative day 8 to home on oxygen
- The TCU IRB determined the case report is exempt from IRB/IACUC approval as defined by 45 CFR 46.102.

METHODS

- PubMed, Embase and MEDLINE Complete searched
- Search terms “*fibrodysplasia ossificans progressiva*”, and “*anesthesia*”
- Dental procedures and pediatrics excluded
- Search yielded 12 results from the 3 databases
- 10 relevant case reports were assessed and synthesized: treatment guidelines by International Clinical Council on FOP included in recommendations

Fibrodysplasia Ossificans Progressiva is a rare, progressive disease that poses multiple anesthetic challenges.

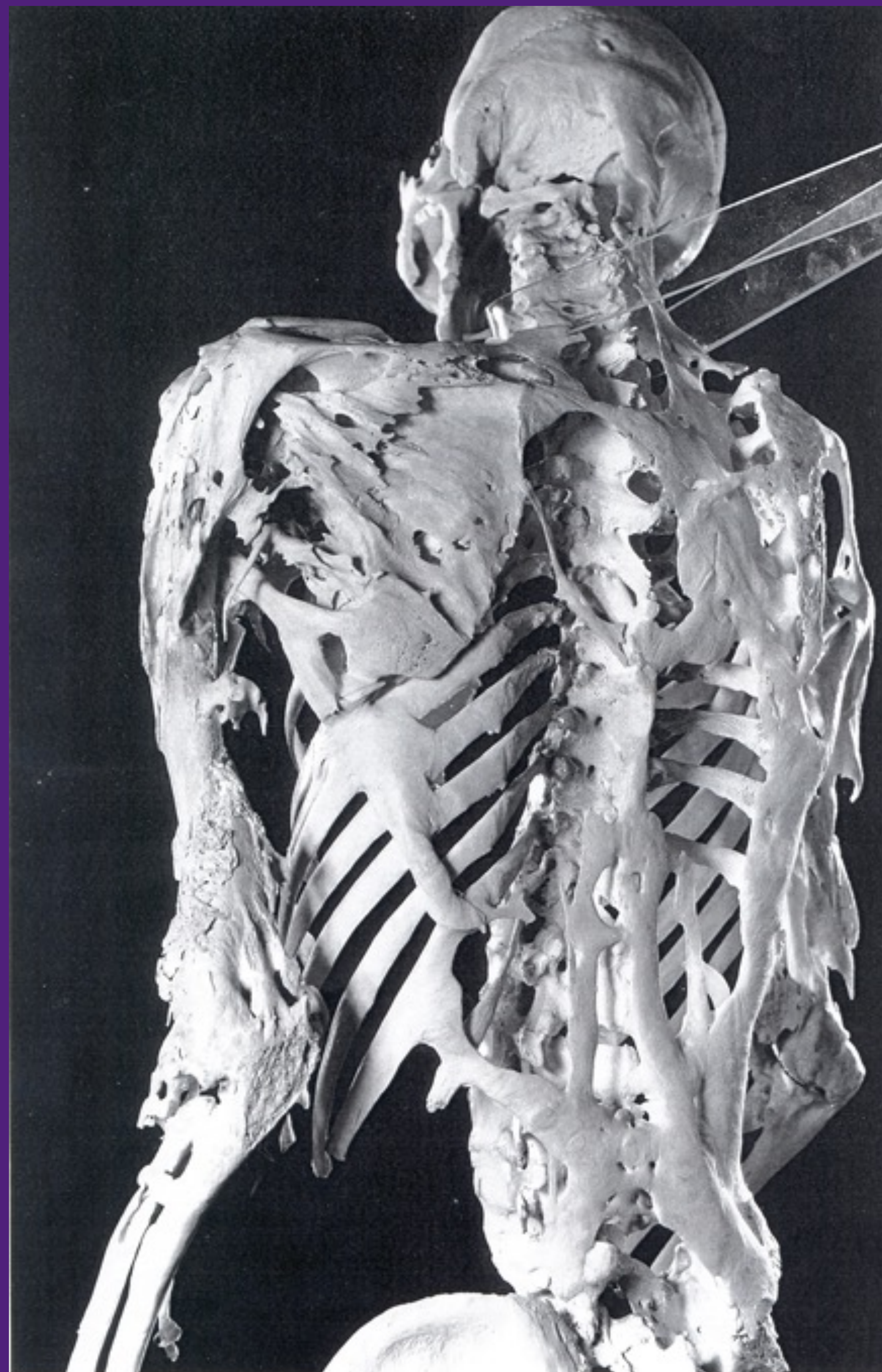


Figure. A photo of Harry Eastlack’s skeleton which is on display at The Mutter Museum of the College of Physicians in Philadelphia. Mr. Eastlack suffered from FOP and donated his body to science to help others affected with the disorder.¹³



DISCUSSION / SUPPORTING EVIDENCE

Anesthesia Plan	Case studies using technique	Methods
Airway	7/10 instrumented airway ²⁻⁸	<ul style="list-style-type: none"> • 1 direct laryngoscopy⁴ • 1 direct video laryngoscopy³ • 3 awake nasal fiberoptic^{2, 5-6} • 1 awake oral fiberoptic⁷ • 1 blind nasal intubation⁸
Steroids	4/10 administered steroids ^{3,5-6,9}	Steroid administration: <ul style="list-style-type: none"> • Postoperative and x 6 days³ • Preoperative and x 3 days⁶ • Single dose intraoperative to supplement patient regimen⁵ • Administered in nerve block catheters⁹
	2/10 avoided steroids ⁷⁻⁸	Rationale: <ul style="list-style-type: none"> • No effect⁸ • Unproven benefit⁷
Regional	3/10 used regional anesthesia technique ^{6, 9-10}	Ultrasound-guided blocks: <ul style="list-style-type: none"> • Ankle block—foot surgery¹⁰ • Femoral and sciatic nerve—lower leg amputation⁹ • Bilateral transversus abdominus plane (TAP) block—supracervical hysterectomy, appendectomy⁶

Table. Individualized Anesthesia Management Techniques in 10 Case Studies.²⁻¹¹

Recommendations for Practice

Location	Major medical center with practitioners and equipment capable of caring for complex cases
Anesthesia	General anesthesia due to airway challenges
Airway	Awake nasal fiberoptic intubation considered optimal Otolaryngologist present, prepared to place emergent airway
Avoid tissue injury	Pillows, padding to minimize soft tissue injury Minimize tourniquet time for peripheral IV placement
Medications	Minimize opioid administration to avoid respiratory depression Acetaminophen, NSAIDs are recommended
Regional	Avoid mandibular blocks—known to cause fusion of TMJ Recommendations for regional anesthesia are lacking 1 case report of a TAP block and 1 case report of femoral and sciatic block—no HO formation reported Intracutaneous or subcutaneous injections are tolerated USG, subcutaneous infiltration of LA may be a viable option

Table 2. Treatment Guidelines by International Clinical Council on Fibrodysplasia Ossificans Progressiva and Case Reports.^{8,9,11,12}
Abbreviations. HO, heterotopic ossification; IV, intravenous; LA, local anesthetic; NSAIDs, non-steroidal anti-inflammatory drugs; TAP, transversus abdominus plane; TMJ, temporomandibular joint; USG, ultrasound-guided.

CONCLUSIONS

- FOP is a rare disease that presents numerous challenges for the anesthesia professional
- Future research in the utility of specific ultrasound-guided blocks is needed