We've Got a Bone to Prevent: Cesarean Delivery in a Unique Fibrodysplasia Ossificans Progressiva (FOP) Patient

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- FOP is a rare genetic disorder of ACVR1/ALK2 gene on chromosome 2
- Causes progressive heterotopic bone formation of ligaments, tendons, skeletal muscle
- Results in severe joint immobility, especially in jaw, c-spine, rib cage
- 3% have mild variant with minimal symptoms
- Pregnancy in FOP patients is rare and has risk of rapid disease progression, worsened pulmonary status, miscarriage, preterm delivery, DVT, C/S, fetal inheritance (50%)

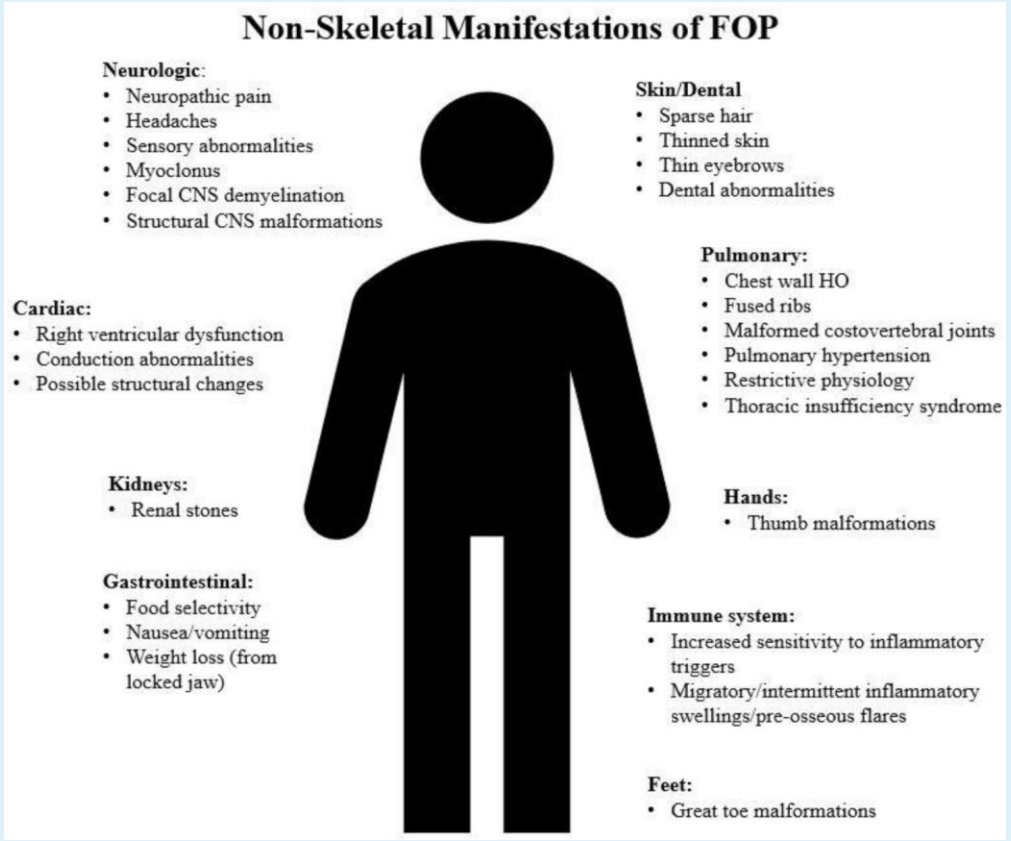

Figure 1: Non-skeletal FOP symptoms

Khan F, Yu X, Hsiao EC. Cardiopulmonary and Neurologic Dysfunctions in Fibrodysplasia Ossificans Progressiva. *Biomedicines*. 2021.

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Case Presentation

- 33 y/o F G1P0 at 37w1d s/f C/S in setting of new gHTN and FGR
  - PMH: anxiety, depression, GERD, hypothyroidism
  - Asymptomatic mild variant FOP (heterozygous ACVR1 mutation)
- Multidisciplinary management plan
- Positioning: supine on air transfer mattress with padded IV, BP cuff, and compression stocking sites
- Anesthetic: GETA with standard monitors
  - RSI with propofol and succinylcholine, maintenance with propofol, nitrous oxide, and rocuronium
  - Video laryngoscopy with McGrath #3 blade
- IV solumedrol, IV tranexamic acid, oxytocin infusion, and rectal misoprostol given
- Uneventful C/S with delivery of 2579 g neonate (APGARs 3^{1}, 6^{5}), QBL 1.36 L
- Pain control: B/L Transverse abdominis plane blocks with 0.25% ropivacaine, IV ketorolac, hydromorphone
- 3-day course of PO prednisone, discharged on POD#4, no FOP flares or soft tissue ossifications since C/S

Teaching Points

- Multidisciplinary planning (anesthesia, OB, neonatology, FOP experts) is key

- Goal is minimizing invasive interventions and triggers
  - Vaginal birth contraindicated
  - Regional anesthesia and direct laryngoscopy not recommended
  - Surgery can exacerbate FOP
  - Careful positioning, padding soft tissues, avoiding IM injections, steroid prophylaxis

- FOP treatment is symptomatic
  - Steroids, NSAIDs, COX-2 & leukotriene inhibitors, mast cell stabilizers

- Unclear whether this mild variant patient will develop future FOP sequelae from anesthetic management during C/S

References

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4. 4. Am J Case Rep 2021;22:e931614