Emergency Guidelines for Paramedics, A&E Staff, First Responders, Physicians and Dentists

Taken from the International Clinical Council for FOP's Medical Guidelines July 2024

iccfop.org

1. Avoid all IM injections unless necessary for survival of the patient. They will likely cause flare-ups and subsequent ossification.
2. Peripheral IVs are permissible. Use smallest needle possible with brief tourniquet time. Avoid repeated tourniquet use or over-inflation of blood pressure cuffs.
3. Avoid central venous access unless necessary for survival of the patient.
4. In case of major trauma, begin corticosteroids immediately (oral) or IV equivalent of oral Prednisone - 1-2 mg/kg once daily for 4 days.
5. Pad all bony prominences to prevent pressure ulcers and skin breakdown.
6. The cervical spine is often partially or completely ankylosed from FOP. Do not manipulate.
7. The jaw is likely limited in movement or functionally ankylosed. Even if it is mobile, it is extremely susceptible to trauma. Do not passively manipulate. Over-stretching and mandibular blocks are forbidden as they will cause flare-ups.
8. Flare-ups of the anterior neck can compromise breathing and swallowing and should be considered a medical emergency. These submandibular flare-ups require early identification. Provide high dose steroids immediately (Solumedrol 80 mg iv or Dexamethasone 15 mg iv). Avoid additional trauma with lesional manipulation. Airway monitoring, aspiration precautions, nutritional support, and immediate use of corticosteroids are mandatory.
9. Head and neck injuries are common from falls as the arms are rigid from ankylosis of the shoulders early in life and cannot be used to protect the head in case of falls.
10. With head injury, always brace the neck.
11. With any head injury, even without loss of consciousness, a head CT is mandatory to rule out intracranial bleeding due to the high likelihood of an unprotected impact.
12. Flare-ups of the head in younger patients can appear as very large scalp swellings and initially disfiguring. A conservative approach should be taken with scalp flare-ups, with monitoring, and pain control if necessary. Scalp flare-ups will resolve spontaneously over time and disfigurement will be minimal to none as new ossifications are incorporated into the growing skull.
13. Facial swelling due to scalp flare-ups in FOP is uncommon but may occur. Other aetiologies for facial swelling should be considered, such as drug reactions and cavernous sinus thrombosis. A brief course of antihistamine should be considered to exclude allergies in FOP patients who present with facial swelling.
14. It is always advisable to obtain a complete dental examination for swelling or pain of the oral-facial region as it can be difficult to distinguish a swelling of dental origin from a flare-up of FOP. If the dental radiographs and/or pulpal testing (vitality of the nerve of the tooth) indicates no obvious dental origin to the swelling, it is prudent to assume an FOP flare-up and initiate prednisone flare-up dosing. If it is not possible to get a

dental radiograph or do pulpal testing, then prescribing an appropriate antibiotic together with prednisone is warranted until a definitive diagnosis can be made.

15. For "dirty" or contaminated wounds use tetanus hyperimmune globulin. Avoid tetanus immunisation as IM or subcutaneous immunisation unless necessary, as this has a high likelihood of inciting a flare-up.
16. Some hearing impairment is common in FOP. Speak loudly and clearly.
17. Although stable hearing loss is a common feature of FOP in children, acute hearing loss and ear pain is not and should be evaluated and treated as in any child.
18. Dental pain is a common issue in FOP patients and must be evaluated and treated promptly, but only after thorough consultation with an FOP dental expert. Overstretching of the jaw and mandibular blocks are forbidden.
19. Kidney stones are very common in adults with FOP. Keep well hydrated.
20. Fractures are common in normotopic as well as heterotopic bone. Closed immobilisation with splinting and bracing are recommended. Open reduction is contraindicated unless thoroughly discussed with an FOP specialist.
21. With nausea and vomiting in individuals with an ankylosed jaw, cover empirically with antibiotics for aspiration pneumonia.
22. Acute and often severe limb swelling can be seen with flare-ups of FOP, especially of the lower extremities. Due to intense inflammation, angiogenesis and capillary leakage, this swelling may grow to extraordinary and alarming size and lead to extravascular compression of nerves and tissue lymphatics. After excluding a possible deep vein thrombosis, the swelling should be treated conservatively with adequate pain control, elevation, and ultimately with safe lymphedema manipulations. Although signs and symptoms of compartment syndrome may prompt consideration of emergent surgical release of pressure (e.g., fasciotomy), this will exacerbate the flare-up and MUST be avoided. If clinical suspicion of compartment syndrome is high, consider the use of mannitol.
23. In the case of limb swelling that prompts concern for deep vein thrombosis, Doppler ultrasound evaluation of the venous system may be indicated.
24. Ask if patient is enrolled in any FOP Clinical Trials and communicate with principal investigator and regional FOP specialist.
25. In the case of choking and failure to clear throat manually, perform Heimlich manoeuvre if there is no evidence for abdominal heterotopic bone that would prevent attempts.
26. Chest compressions will likely be futile. The chest wall is rigid and immobile.
27. Intubation must be through an awake, fibreoptic nasotracheal approach by an experienced anaesthesiologist.
28. If an emergency tracheotomy is necessary in an individual with anterior neck ossifications, a dental or other drill may be necessary to create an airway.
29. In emergency situations where patients have difficulty clearing secretions, use bronchodilators, mucolytics, and guaifenesin, with a low threshold for mechanical insufflation-exsufflation devices. Hydration should be optimized with intravenous fluids.
30. Avoid unmonitored use of supplemental oxygen to minimize the chance of respiratory failure and death.