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# THE MEDICAL MANAGEMENT OF FIBRODYSPLASIA OSSIFICANS PROGRESSIVA: CURRENT TREATMENT CONSIDERATIONS



**From The International Clinical Council on FOP (ICC) & Consultants:**

Frederick S. Kaplan, Mona Al Mukaddam, Genevieve Baujat, Matthew Brown, Amanda Cali, Tae-Joon Cho, Corrie Crowe, Carmen L. De Cunto, Patricia Delai, Robert J. Diecidue, Maja Di Rocco, Elisabeth M.W. Eekhoff, Clive Friedman, Zvi Grunwald, Nobuhiko Haga, Edward C. Hsiao, Richard Keen, Joseph Kitterman, Charles Levy, Rolf Morhart, J. Coen Netelenbos, Christiaan Scott, Eileen M. Shore, Michael A. Zasloff, Keqin Zhang, Robert J. Pignolo

Corresponding Editors:

Frederick S. Kaplan, M.D.  
Isaac and Rose Nassau Professor of Orthopaedic Molecular Medicine  
Director, Center for Research in FOP & Related Disorders  
The Perelman School of Medicine - The University of Pennsylvania  
Department of Orthopaedic Surgery  
3737 Market Street – Sixth Floor  
Philadelphia, PA 19104, USA  
Tel: (office) 215-294-9145  
Fax: 215-222-8854  
Email: [Frederick.kaplan@penmedicine.upenn.edu](mailto:Frederick.kaplan@penmedicine.upenn.edu)

Robert J. Pignolo, M.D., Ph.D.  
Chair, Division of Geriatric Medicine & Gerontology  
Robert and Arlene Kogod Professor of Geriatric Medicine  
Mayo Clinic College of Medicine  
200 First Street SW  
Rochester, MN 55905, USA  
Tel: 507-293-6988; Secretary: 507-293-0813  
Fax: 507-293-3853  
Email: [pignolo.robert@mayo.edu](mailto:pignolo.robert@mayo.edu)

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## **VIII. EMERGENCY GUIDELINES FOR 1st RESPONDERS, PHYSICIANS & DENTISTS**

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.
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 (methylprednisolone 100 mg iv or Dexamethasone 4 mg iv). Avoid additional trauma with lesional manipulation. Airway monitoring, aspiration precautions, nutritional support, immediate use of corticosteroids.
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, and other etiologies for facial swelling should be considered. A brief course of antihistamine should be considered to exclude allergies in FOP patients who present with facial swelling.
14. For “dirty” or contaminated wounds use tetanus hyperimmune globulin. Avoid tetanus immunization as IM or subcutaneous immunization unless necessary, as this has a high likelihood of inciting a flare-up.
15. Some hearing impairment is common in FOP. Speak loudly and clearly.
16. 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.
17. 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.
18. Kidney stones are common in adults with FOP. Keep well hydrated.

19. Fractures are common in normotopic as well as heterotopic bone. Closed immobilization with splinting and bracing are recommended. Open reduction is contraindicated unless thoroughly discussed with an FOP specialist.
20. With nausea and vomiting in individuals with an ankylosed jaw, cover empirically with antibiotics for aspiration pneumonia.
21. 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, this will exacerbate the flare-up and **MUST** be avoided.
22. In the case of limb swelling that prompts concern for deep vein thrombosis, Doppler ultrasound evaluation of the venous system may be indicated.
23. Ask if patient is enrolled in any FOP Clinical Trials and communicate with principal investigator and regional FOP specialist.
24. In the case of choking and failure to clear throat manually, perform Heimlich maneuver if there is no evidence for abdominal heterotopic bone that would prevent attempts.
25. Chest compressions will likely be futile. The chest wall is rigid and immobile.
26. Intubation must be through an awake, fiberoptic nasotracheal approach by an experienced anesthesiologist.
27. 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.
28. 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.

### **For Consultation on FOP Anesthesia & Airway Issues:**

#### **Zvi Grunwald, MD**

The James D. Wentzler Professor and Chairman Emeritus

Department of Anesthesiology

Thomas Jefferson University

111 South 11th Street, Suite G-8490

Philadelphia, PA 19107, USA

Tel: 215-955-6161

Cell: 215-206-7362

Fax: 215-923-5507

Email: [zvi.grunwald@jefferson.edu](mailto:zvi.grunwald@jefferson.edu)

**For Consultation on Emergent Dental Care:**

**Corrie Crowe, DDS**

1793 Springdale Road

Cherry Hill, NJ 08003, USA

Tel: 856-258-4025 (Staff Assistant: Lynn)

Cell: 407-701-3210

Fax: 856-504-6179

Email: [drcrowdentistry@gmail.com](mailto:drcrowdentistry@gmail.com)

**Clive S. Friedman, DDS, FAAPD**

389 Hyde Park Road

London, Ontario

Canada N6h 3R8

Phone: cell 519 6576014

Office: 519-679-9860; (private line): 519-679-5473

Home: 519 438-1198

Email: [clive@dentistryforkids.ca](mailto:clive@dentistryforkids.ca) for (home) [clivesf@mac.com](mailto:clivesf@mac.com)

**For Hospital Dentistry and Dental Surgery:**

**Robert Diecidue, MD, DMD, MBA, MPH**

Thomas Jefferson University

Jefferson Medical College

Department of Oral and Maxillofacial Surgery

Chairman and Professor

909 Walnut Street - Suite 300

Philadelphia, Pennsylvania 19107, USA

Tel: 215-955-6215; 215 955 5131

Fax: 215-923-9189

Email: [robert.diecidue@jeffersonhospital.org](mailto:robert.diecidue@jeffersonhospital.org)