

Oral and Dental Health Care and Anesthesia for Persons With Fibrodysplasia Ossificans Progressiva

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Abstract

Fibrodysplasia ossificans progressiva (FOP) is a devastating genetic disorder with profound oral and dental health care issues. Most patients with FOP eventually develop heterotopic ossification of the chewing muscles with resultant ankylosis of the temporomandibular joints. Preventive and restorative dental care, as well as endodontic, periodontic, and orthodontic care present therapeutic challenges to the dental practitioner, the anesthesiologist, and to the FOP patient throughout his or her life.

Key Words: Fibrodysplasia ossificans progressiva (FOP); dental health care; oral health care; anesthesia.

Introduction

Fibrodysplasia ossificans progressiva (FOP) is a rare autosomal dominant disorder characterized by congenital malformation of the great toes and by progressive postnatal heterotopic ossification (HO) of soft tissues in characteristic anatomic patterns (1–4). Postnatal HO generally appears within the first decade of life following either spontaneous or trauma-induced flare-ups (1,4,5). Progressive episodes of HO lead to ankylosis of all major joints of the axial and appendicular skeleton, including the temporomandibular joints (TMJs), rendering movement impossible (6–8). Overstretching the jaw, mandibular anesthetic blocks, and surgical trauma associated with resection of heterotopic bone leads to catastrophic

episodes of robust new bone formation (1,4,9). Even in the absence of HO, developmental abnormalities in the TMJs are often noted early in life (7).

Dental Problems in Patients Who Have FOP

Dental issues in patients with FOP are similar to those in the general population. Patients with FOP develop caries, gum disease, and periodontal disease. If the TMJs are functional, individuals with FOP do not exhibit more dental problems than in others. However, if the TMJs are ankylosed, then the teeth cannot be brushed on the occlusal or lingual surfaces, and caries, gingivitis, and periodontitis are likely to develop with higher incidence (10).

Many patients with FOP are unfortunately not treated for caries early in life owing to the fear that dental manipulation may worsen the condition. Other reported reasons for nontreatment are lack of

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knowledge on how to access decayed teeth for restoration and lack of knowledge about proper local and general anesthetic care (10). Luchetti reported severe restriction in jaw movement following routine injection of local anesthetics for mandibular blocks in patients who have FOP (9). Obviously, mandibular blocks and overstretching of the jaw must be assiduously avoided in patients with FOP.

TMJ surgery in patients with FOP with ankylosed TMJs has had variable success. Most of the morbidity is the result of anesthetic issues and, in some cases, surgical issues. Crofford reported failure of jaw surgery and isotretinoin to restore jaw immobilization in FOP (11). Herford reported a successful oral opening in a FOP patient with an ankylosed jaw through an osteotomy approach, although the TMJs reankylosed in an open position (12). The osteotomy was performed to provide oral access for sustenance and dental care, and the patient was satisfied with the outcome. At the present time, there is no effective approach to the remobilization of an ankylosed jaw in a patient with FOP.

Preventive Oral Health Care Recommendations

Preventive oral and dental health care measures are essential in patients with FOP, especially during childhood years. Periodontic and preventative oral care is crucial to prevent long-term dental and oral complications in patients with FOP. Fluoridation of water is suggested for all patients who have FOP. The use of high-dose fluoride toothpaste is recommended, along with use of fluoride gels and rinses to help prevent the need for restorative dental care. Chlorhexidine rinses are encouraged to prevent gingivitis and tooth decay. Frequent flossing and brushing are as necessary in patients with FOP as in unaffected individuals, but may be difficult on account of limited jaw opening as the FOP progresses. Patients with FOP who are still able to open their mouths can be treated with normal dental instruments as in unaffected individuals, but assiduous care must be exercised to prevent overstretching of the TMJs during dental procedures. In patients who have ankylosed TMJs, professional instrumentation and special toothbrushes may be helpful, but are often limited to use on the buccal surfaces. Antimicrobial and fluoride rinses may be the only method to reach the lingual and palatal surfaces (10).

Restorative and Surgical Dental Care

When jaw-opening ability is limited, restorative dental care and fillings are technically difficult to perform in patients with FOP. Preparation of the tooth and removal of the decay may need to be modified for patients with limited oral opening. Access to the tooth from the buccal surface may be necessary for patients with FOP who are unable to open their jaw adequately. In limited space, it may be possible to remove tooth decay using a slow dental drill. The use of fluoride-releasing filling material is recommended. These filling materials bond to the tooth and release fluoride into the surrounding tooth structure, preventing further decay.

In patients with FOP who have ankylosed TMJs, dental extractions pose an extreme problem. It may be necessary, for example, to approach the tooth entirely from a buccal direction as forceps cannot be placed on top of the tooth in the usual manner. In a closed-mouth individual with an ankylosed TMJ, it may be necessary to section the decayed tooth into pieces before removal. Formation of a window on the buccal side may be necessary to remove a wisdom tooth or to create access to the dental roots. A dental instrument must be placed lingually or palatally to prevent tooth debris from falling inside the mouth. A soft, malleable neurosurgical retractor may be fitted between the teeth to create a shield.

Dental Anesthesia in Patients With FOP

Patients with FOP have limited options for dental anesthesia. Mandibular blocks are forbidden because they will lead to ossification of the pterygoid muscles and rapid ankylosis of the TMJ (9). Infiltration anesthesia is difficult in the mandibular posterior molar areas for permanent teeth. Successful anesthesia in mandibular primary teeth can be achieved by infiltration through the dental pulp. Interligamentary infiltration may be helpful, if performed carefully (13); however, in some patients, this type of local anesthesia may not be possible for age-related behavioral reasons or for access reasons. In those cases, general anesthesia may be necessary.

General anesthesia is a particularly dangerous matter in patients who have FOP. Overstretching of the jaw for intubation may cause additional trauma to

the TMJs, and lead to disease flare-ups. Because the TMJs may be ankylosed in older patients who have FOP, oral access for intubation may not be possible. General anesthesia for dental care in patients with FOP should be accomplished through a fiberoptic nasal intubation while the patient is awake but under light sedation, so that the patient can control secretions. This type of procedure should be performed by well-trained anesthesia teams who are familiar and experienced with this type of procedure (10).

Orthodontics and FOP

Most people seek orthodontic care for aesthetic and functional reasons. For the FOP population, self image is as important as in the general population. Orthodontic therapy can be safely performed on patients with FOP who have normal or nearly normal oral opening (9).

Patients who have FOP often develop mandibular hypoplasia with a maxillary overbite and, therefore, orthodontic therapy may be considered. However, many patients find that the overbite provides a means of access for eating, as well as for oral and dental hygiene. Posterior and anterior dental cross-bites can have an effect on the TMJs and should be corrected. For children with functional TMJs and with anterior open bites that are less than 15 mm, orthodontics is not recommended because the overbite will facilitate nutrition and subsequent dental care if the TMJ does eventually ankylose.

When orthodontic care is considered, brief appointment times are recommended to lessen stress on the TMJs. The use of nonextraction therapy is also recommended. To prevent the need for extractions in patients with FOP, it may be advisable to align the anterior segments for aesthetics, leaving posterior dental crowding untreated. Crowded posterior teeth may be a better alternative than the risks of flare-up and TMJ ankylosis that can accompany an extraction.

Submandibular Swelling in Patients With FOP

In addition to flare-ups involving the TMJs, flare-ups involving the submandibular region occur in patients who have FOP and can complicate dental care (14). Submandibular flare-ups were initially noted in 12 of 107 FOP patients (11%), and were

mistaken initially in seven patients for mumps, angioneurotic edema, abscess, mononucleosis, or neoplasm. In our experience, the prevalence of patients with FOP who have had submandibular flare-ups approaches 30% of the known adult patients with FOP worldwide. Most patients survived following assiduous precautionary measures. One patient who required emergency tracheostomy and ventilatory support also survived. Another patient died of inanition from chronic swallowing difficulty. Submandibular swelling in patients who have FOP can be a medical emergency, and requires intensive precautionary measures to avoid catastrophic clinical deterioration. These measures include avoidance of lesional manipulation, airway monitoring, aspiration precautions, nutritional support, and glucocorticoid therapy (14).

Summary

In summary, patients with FOP have numerous oral and dental healthcare issues that require life-long and assiduous attention to prevention, and to minimization of risks when restorative care is necessary. Patients who have FOP should consult the dental section of *The FOP Treatment Guidelines (The Medical Management of Fibrodysplasia Ossificans Progressiva (FOP): Current Treatment Considerations)* for updated clinical guidelines on oral health issues. These guidelines are updated frequently, and can be accessed on the International FOP Association website at: www.ifopa.org.

References

1. Connor JM, Evans, DA. 1982 Fibrodysplasia ossificans progressiva (FOP): the clinical features and natural history of 34 patients. *J Bone Joint Surg Br* 64:76–83.
2. Cohen RB, Hahn GV, Tabas J, et al. 1993 The natural history of heterotopic ossification in patients who have fibrodysplasia ossificans progressiva. *J Bone Joint Surg* 75-A:215–219.
3. Rocke DM, Zasloff M, Peeper J, Cohen RB, Kaplan FS. 1994 Age and joint-specific risk of initial heterotopic ossification in patients who have fibrodysplasia ossificans progressiva. *Clin Orthop Rel Res* 301:243–248.
4. Kaplan FS, Shore EM, Connor JM. 2002 Fibrodysplasia ossificans progressiva (FOP). In: Royce PM, Steinmann B, eds. *Connective Tissue and Its Heritable Disorders: Molecular, Genetic, and Medical Aspects*, 2nd Ed. Wiley-Liss; John Wiley & Sons, New York, pp. 827–840.
5. Lanchoney TF, Cohen RB, Rocke DM, Zasloff MA, Kaplan FS. 1995 Permanent heterotopic ossification at the

- injection site after diphtheria-tetanus-pertussis immunizations in children who have fibrodysplasia ossificans progressiva. *J Pediatrics* 126:762–764.
6. Connor JM, Evans, DA. 1982 Extra-articular ankylosis in fibrodysplasia ossificans progressiva. *Br J Oral Surg* 20:117–121.
 7. Renton P, Parkin SF, Stamp TC. 1982 Abnormal temporomandibular joints in fibrodysplasia ossificans progressiva (FOP). *Br J Oral Surg* 20:31–38.
 8. el-Labban NG, Hopper C, Barber P. 1995 Ultrastructural finding of vascular degeneration in fibrodysplasia ossificans progressiva (FOP). *J Oral Pathol Med* 24:125–129.
 9. Luchetti W, Cohen RB, Hahn GV, et al. 1996 Severe restriction in jaw movement after route injection of local anesthetic in patients who have fibrodysplasia ossificans progressiva. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 81:21–25.
 10. Nussbaum BL, O'Hara I, Kaplan FS. 1996 Fibrodysplasia ossificans progressiva: report of a case with guidelines for pediatric dental and anesthetic management. *J Dentistry Children* 63:448–450.
 11. Crofford LJ, Brahim JS, Zasloff, MA, Marini JC. 1990 Failure of surgery and isotretinoin to relieve jaw immobilization in fibrodysplasia ossificans progressiva (FOP): report of two cases. *J Oral Maxillofac Surg* 48:204–208.
 12. Herford AS, Boyne PJ. 2003 Ankylosis of the jaw in a patient with fibrodysplasia ossificans progressiva. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 96:680–684.
 13. Webb MD, Wilson C. 1996 The use of intraosseous anesthesia in a patient with myositis ossificans progressiva. *Spec Care Dentist* 16:29–32.
 14. Janoff HB, Zasloff MA, Kaplan FS. 1996 Submandibular swelling in patients with fibrodysplasia ossificans progressiva. *Otolaryngol Head Neck Surg* 114:599–604.