

# PROPOSTA DI PROTOCOLLO STOMATOLOGICO IN PAZIENTI AFFETTI DA FOP:REVISIONE DELLA LETTERATURA

5<sup>th</sup> MEETING F.O.P.  5<sup>th</sup> F.O.P. CONFERENCE

**FIBRODISPLASIA OSSIFICANTE PROGRESSIVA**  
**FIBRODISPLASIA OSSIFICANS PROGRESSIVA**

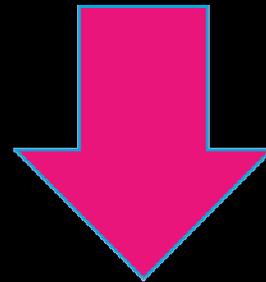
**ROMA - ITALY MARZO 2011 MARCH**  
**GIOVEDI 24, VENERDI 25, SABATO 26**  
**THURSDAY 24, FRIDAY 25, SATURDAY 26**

**SALA ANFITEATRO**  
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FOP

Fibrodysplasia ossificans progressiva



Disordine del tessuto connettivo con  
progressiva ossificazione ectopica di tendini,  
legamenti, muscoli facio-scheletrici

**“IL PZ FOP NON HA PROBLEMATICHE  
ODONTOIATRICHE CHE DIFFERISCONO  
DALLA POPOLAZIONE NORMALE BENSÌ HA  
LIMITATE POSSIBILITÀ DI ESSERE  
TRATTATO CON CONSEGUENTE RISCHIO DI  
EVOLUZIONI PATOLOGICHE”**

## KEY WORDS: FOP IN DENTISTRY

MANOVRE TERAPEUTICHE VELOCI E NON  
INVASIVE:

- CONTROINDICAZIONI ALL' ANESTESIA  
LOCALE
- LIMITATA APERTURA DELLA BOCCA

# PREVENZIONE INDIVIDUALIZZATA

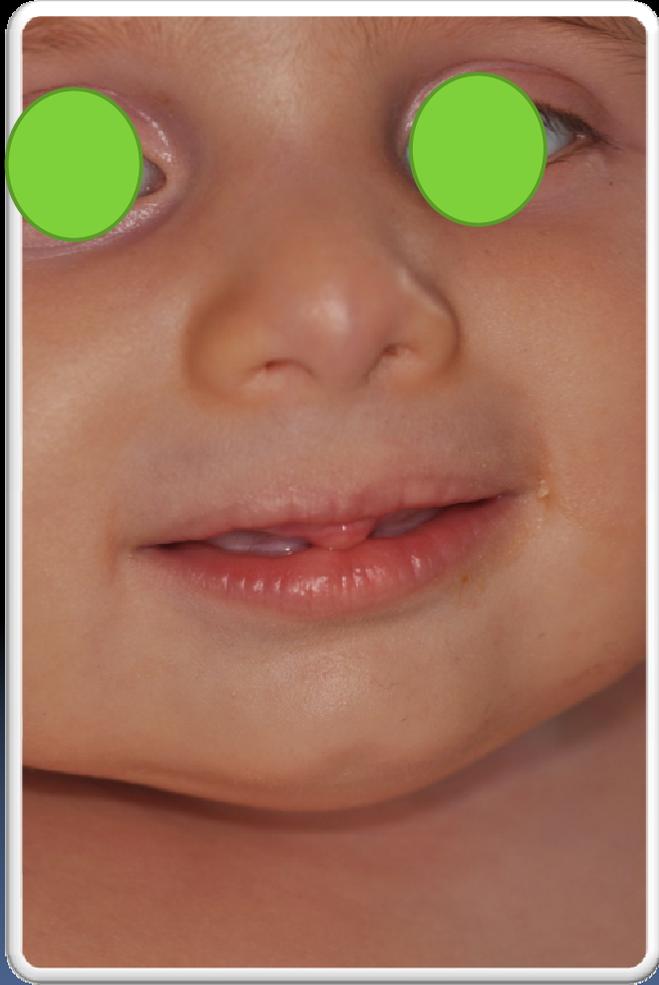


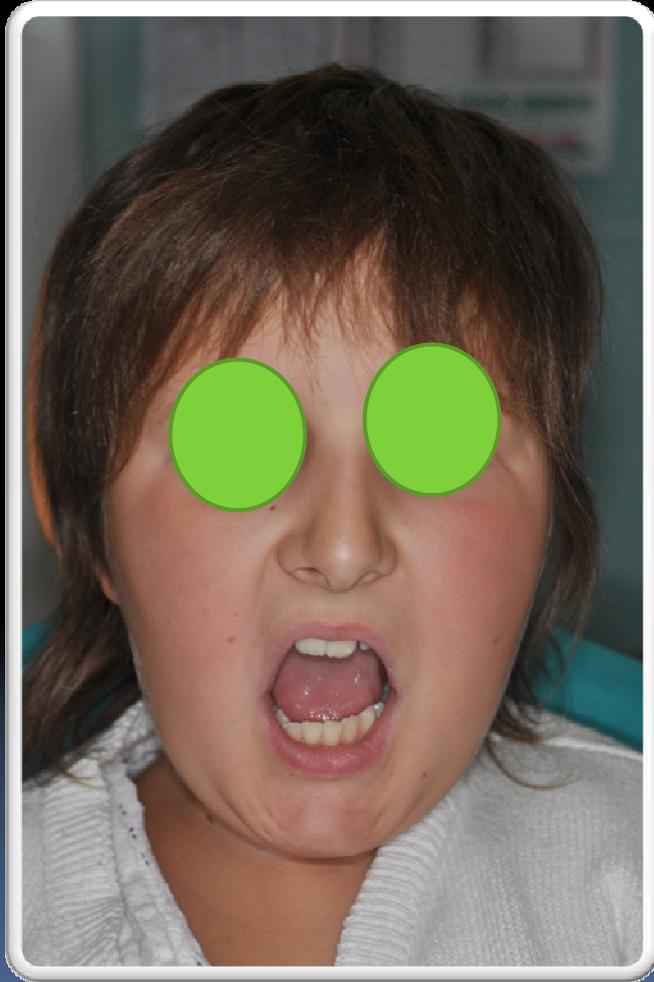
- DIFFICOLTA' NELL ESEGUIRE MANOVRE  
TERAPEUTICHE DI ROUTINE

# PREVENZIONE PATOLOGIA CARIOSI, DELLE MALATTIE PARODONTALI e DELLE MALOCCLUSIONI:

- -alimentazione corretta
- -igiene orale domiciliare
- -controllo odontoiatrico
- -sigillature dei solchi in dentatura decidua e permanente
- -intercettazione malocclusioni















# SIGILLATURE DEI SOLCHI

- Le sigillature sono indicate per tutti i bambini. La loro efficacia nel prevenire la carie è massima se vengono applicate nei 2 anni successivi all'eruzione e la loro integrità va controllata ogni 6-12 mesi.





# REVISIONE DELLA LETTERATURA

Severe restriction in jaw movement after routine injection of local anesthetic in patients who have fibrodysplasia ossificans progressiva.

Luchetti W, Cohen RB, Hahn GV, Rocke DM, Helpin M, Zasloff M, Kaplan FS.

Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1996 Jan;81(1):21-5.

Department of Orthopaedic Surgery, University of Pennsylvania School of Medicine, USA.

- CONCLUSION: Injections of local anesthetic during dental procedures pose serious and immediate risk for inciting heterotopic ossification and ankylosis of the jaw in patients who have fibrodysplasia ossificans progressiva and should be assiduously avoided

**Fibrodysplasia ossificans progressiva: report of a case with guidelines for pediatric dental and anesthetic management.**

Nussbaum BL, O'Hara I, Kaplan FS

ASDC J Dent Child. 1996 Nov-Dec;63(6):448-50.

Children's Hospital of Philadelphia, USA

- Permanent ankylosis of the jaw may be precipitated by minimal soft tissue trauma, a potentially devastating complication following routine dental care during childhood. Assiduous precautions are necessary in administering dental care to children who have FOP as exemplified in this case report. **Routine dental prophylaxis is also necessary in order to minimize the need for invasive procedures.**

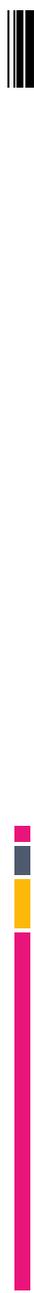
## Ankylosis of the jaw in a patient with fibrodysplasia ossificans progressiva

Alan S. Herford, DDS, MD,<sup>a</sup> and Philip J. Boyne, DMD, MS, DSc,<sup>b</sup> Loma Linda, Calif  
LOMA LINDA UNIVERSITY

A case of fibrodysplasia ossificans progressiva (FOP) is presented. This uncommon connective tissue disease tends to produce progressing ectopic osteogenesis. Because there are no reported curative procedures for TMJ ankylosis occurring in this condition, a palliative surgical approach is described. Etiology, diagnosis, and prognosis of the disease is reviewed. Recent research in BMP cytokine-induced bone repair may allow new approaches to treating this debilitating disease in the future. (*Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003;96:680-684)



Fig 2. A, CT 3-dimensional generated image showing elongated coronoid process (*arrow*) with fusion to the zygoma. B, Ankylosis of the right coronoid to the zygoma (*dark arrow*). Note ossification of the trapezius muscle in the background (*light arrow*).



Fibrodysplasia ossificans progressiva (FOP) is a rare genetic disorder of connective tissue characterized by skeletal malformations at birth and progressive ectopic ossification throughout life.<sup>1-4</sup> The temporomandibular joint (TMJ) is involved in 71% of FOP patients and is often the last osseous articulation to be affected. How-

## CONCLUSION

This report describes a case of a debilitating progressive disease, fibrodysplasia ossificans progressiva. A surgical method of treatment is described. Although surgery is not routinely recommended, it may improve quality of life in selected patients. The unusual nature of this disease together with a description of possible methods of treatment in the future is presented. Hopefully further elucidation of the BMP cytokine pathways will lead to new approaches to intervene medically as well as surgically in this severely debilitating disease.

# Fibrodysplasia ossificans progressiva. A unusual cause of restricted mandibular movement

EH van der Meij, AG Becking, I van der Waal

Oral Disease 2006 12, 204-207

“...controindicata l’anestesia tronculare mandibolare  
.....anestesia intraligamentaria possibile senza  
complicanze....”

“...no controindicazioni al trattamento ortodontico nel  
pz FOP facendo pero’ attenzione alle manovre  
applicative”

# Oral management in a patient with fibrodysplasia ossificans progressiva

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Burton L. Nussbaum, DDS<sup>3</sup>

<sup>1</sup>Chief Resident, Department of Oral and Maxillofacial Surgery, Thomas Jefferson University Hospital, Philadelphia, Pennsylvania; <sup>2</sup>Chairman, Department of Oral and Maxillofacial Surgery and Dentistry, Thomas Jefferson University Hospital; <sup>3</sup>Special Needs Dentist, Thomas Jefferson University Hospital, and private practice, Cherry Hill, New Jersey. \*Corresponding author e-mail: Justin.young@jefferson.edu

*Spec Care Dentist* 27(3): 101-104 (2007)

## Permanent

ankylosis of the temporomandibular joint (TMJ) is a common late finding, but is usually preceded by specific inciting trauma. Extreme care must be exercised when carrying out routine dental care to prevent soft tissue trauma. Oral health care for patients with FOP is usually complex. There are additional considerations when oral surgical procedures, such as extractions, are needed. Maintaining a balance between oral health and disease progression in these patients is challenging, as they frequently present with *advanced oral disease*. Fiber optic-assisted placement of the nasoendotracheal tube is the standard of care. Intramuscular injections including mandibular blocks must be avoided.

# A description of two surgical and anesthetic management techniques used for a patient with fibrodysplasia ossificans progressiva

*Spec Care Dentist* 30(3): 106-109, 2010

Rose Wadenya, BDS, DMD, MS;<sup>1\*</sup> Megan Fulcher, DMD;<sup>2</sup> Tal Grunwald;<sup>3</sup> Burton Nussbaum, DDS, M SND RCS Ed;<sup>4</sup> Zvi Grunwald, MD<sup>5</sup>

Currently, there is no effective treatment for FOP. Thus, attention should be directed to the prevention of dental caries and periodontal disease to minimize the need for invasive dental procedures. Where oral rehabilitation is necessary in a patient with a neck and

temporomandibular joint that are ossified and locked in position, treatment can be provided through the buccal approach. This is a less invasive approach that provides access to the posterior teeth while minimizing the risk and morbidity associated with osteotomy procedures. The challenge of safely securing the airway is met through an awake or sedated fiber-optic endotracheal intubation. A multidisciplinary team approach comprising a dentist, an oral and maxillofacial surgeon, an anesthesiologist, and an otolaryngologist is necessary to achieve therapeutic goals and reduce the risks, cost, and morbidity associated with routine surgical management of patients with FOP.

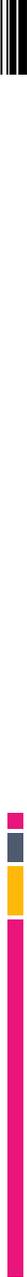
# PREVENZIONE INDIVIDUALIZZATA



- DIFFICOLTA' NELL ESEGUIRE MANOVRE  
TERAPEUTICHE DI ROUTINE

# PROTOCOLLO STOMATOLOGICO PER IL PZ AFFETTO DA FOP

- - Istruzioni ai genitori per alimentazione corretta
- - Istruzioni igiene orale domiciliare
- - Controlli odontoiatrici frequenti (ogni 3 mesi) al fine di intercettare carie allo stato iniziale curabili senza utilizzo di anestetico locale
- - Sigillature dei solchi dei denti da latte e dei permanenti
- - fluoroprofilassi
- - Sedute di igiene professionale ogni 3 mesi
- - Intercettazione malocclusioni e patologia TMJ con metodiche ortopedico funzionali non traumatiche al fine di prevenire carie secondarie ad affollamenti dentali e successive scelte terapeutiche estrattive .



**GRAZIE PER L'ATTENZIONE**

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