Fibrous Dysplasia: Craniofacial Care

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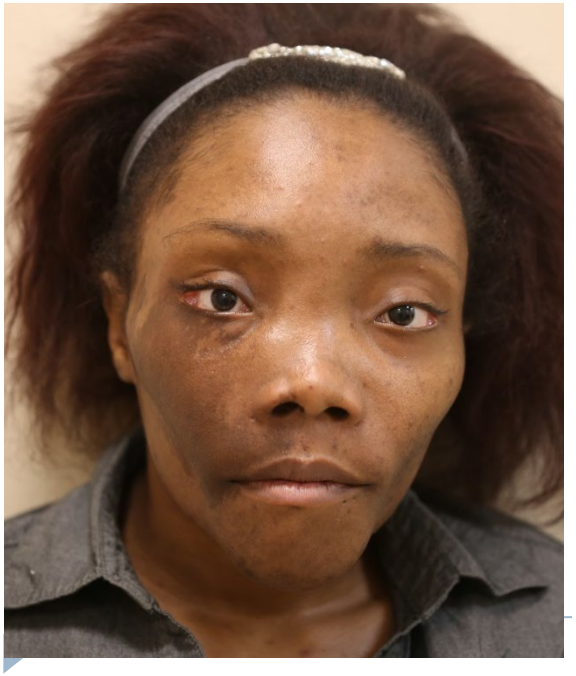
How is CF-FD Treated?

- Craniofacial Multi-disciplinary Team
  - Craniofacial surgeon(s)
    - Oral & Maxillofacial Surgeon
    - Plastic & Reconstructive Surgeon
    - Otolaryngologist (ENT)
    - Neurosurgeon
    - Oculoplastic Surgeon (Ophthalmology)
  - Pediatrician
  - Pediatric Dentist
  - Orthodontist
  - Audiologist
  - Speech & Language Pathologist
  - Geneticist
  - Psychologist
  - Social Worker
  - Nursing (various specialties)

How do CF lesions show up?

- Incidental findings on x-rays
- Asymmetry of face (eyes, forehead, cheeks, nose, jaws)
  - Degree of asymmetry varies
  - MAS most severely affected, particularly when associated with untreated/inadequately treated growth hormone excess



CF-FD Symptoms

Facial asymmetry (86%)
Growth/Mass (64%)

Headache (20%)

Sinusitis (8%)
Epistaxis (3%)

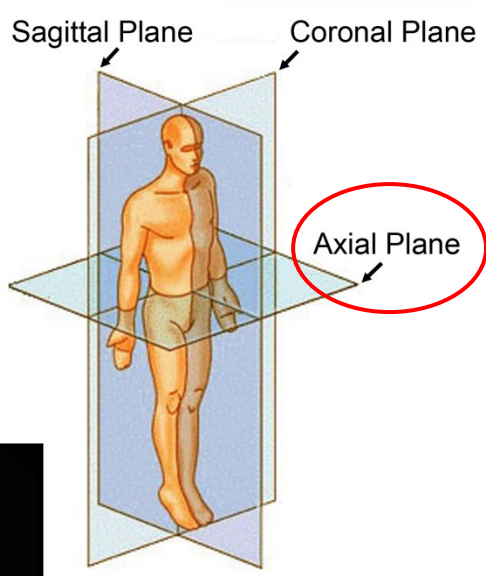
Blurred vision (24%)
Eyelid position abnormalities (10%)
Loss of visual field (8%)
Diplopia (8%)
Epiphora (7%)

Hearing Loss (3%)

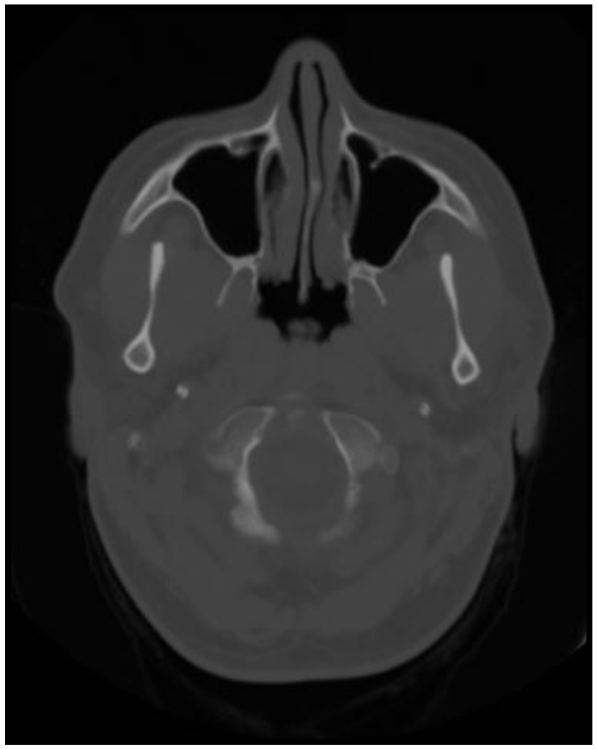
Amit 2011

Computed Tomography (CT)

- Axial/transverse plane
- Bone windows

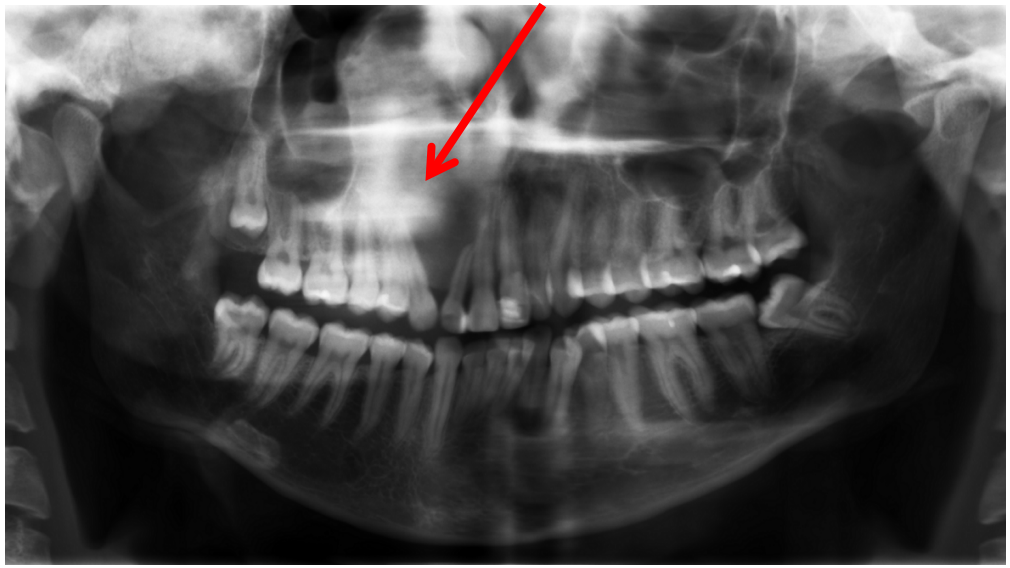


Fibrous Dysplasia



No Involvement

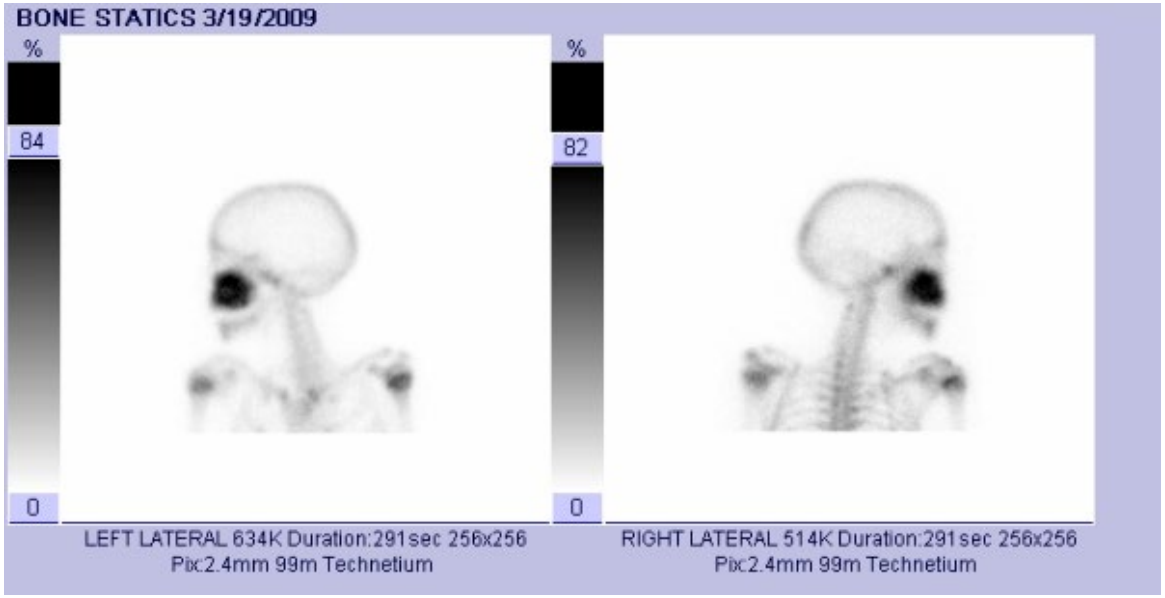
Plain X-rays: Panorex



FD

No involvement

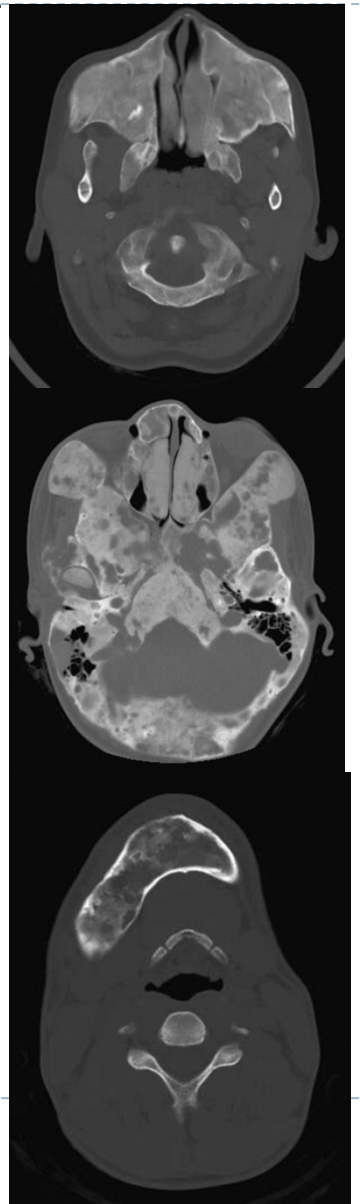
Bone Scan



Monostotic
Fibrous Dysplasia

Craniofacial Fibrous Dysplasia on CT

- Obliteration of normal architecture and landmarks
  - Cortex of bone remains intact
- Variable radiographic appearance
  - Lytic/Lucent
  - Ground glass/mixed
  - Sclerotic
  - Cystic component
- Not uniform
- Appearance changes over time



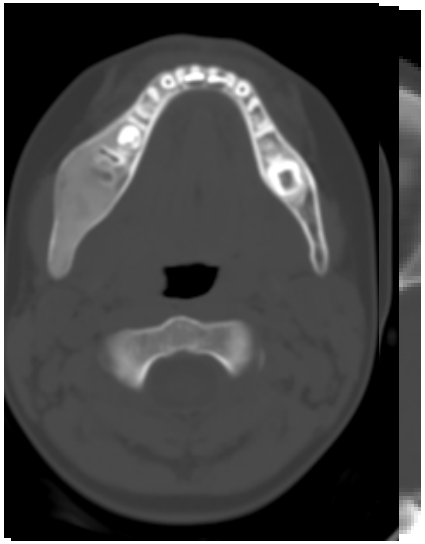
Groundglass

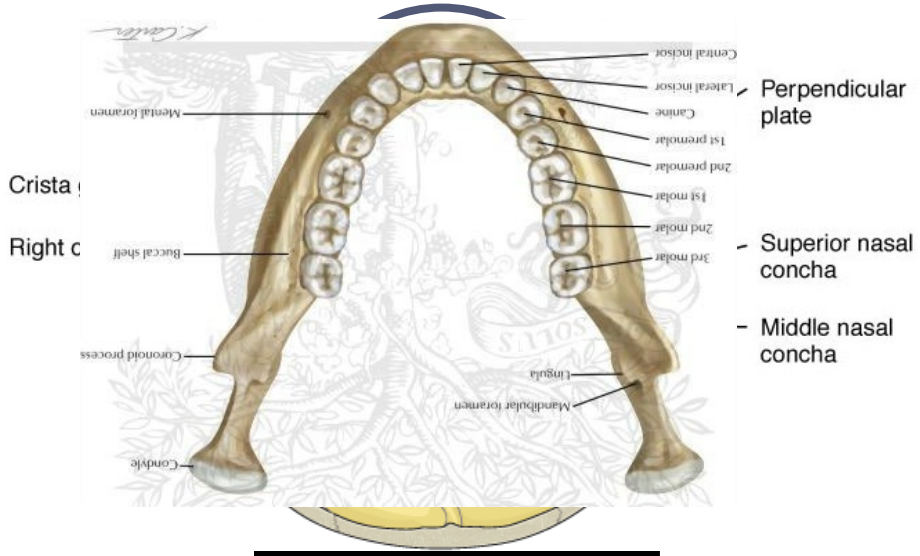
Mixed

Lytic

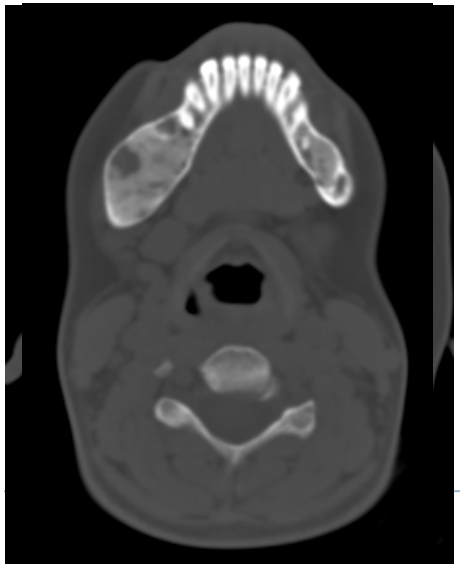
How do FD lesions progress?

Age 5





Age 15



Uninvolved

Will more FD lesions develop?

- CF-FD lesions are earliest to occur, but can remain "silent" until deformity or growth occurs

- Lesions can expand and may change over time
  - Radiographic changes
    - Suggests changes in the activity of the abnormal bone-forming cells as a function of age
    - Most dramatic change occurs in the second decade (11-20 yrs)
      - Patients undergoing growth and hormonal changes
  - Become associated with other rapidly growing lesions
    - i.e. aneurysmal bone cysts
  - Extremely rare transformation to malignancy, <1%
  - Growth Hormone excess can exacerbate

Fibrous Dysplasia and Vision Loss

- Frequent anterior cranial base involvement

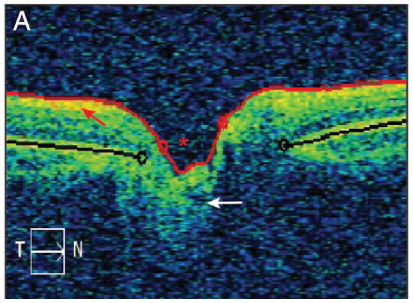
- Assumption:
  - FD around the optic nerve inevitably leads to blindness → prophylactic optic nerve decompression is necessary

Fibrous Dysplasia and Vision Loss

- **Prophylactic optic nerve decompression is not recommended based on radiographic findings alone since these findings DO NOT correlate with vision loss**

- FD is NOT a progressive condition which results in inevitable blindness

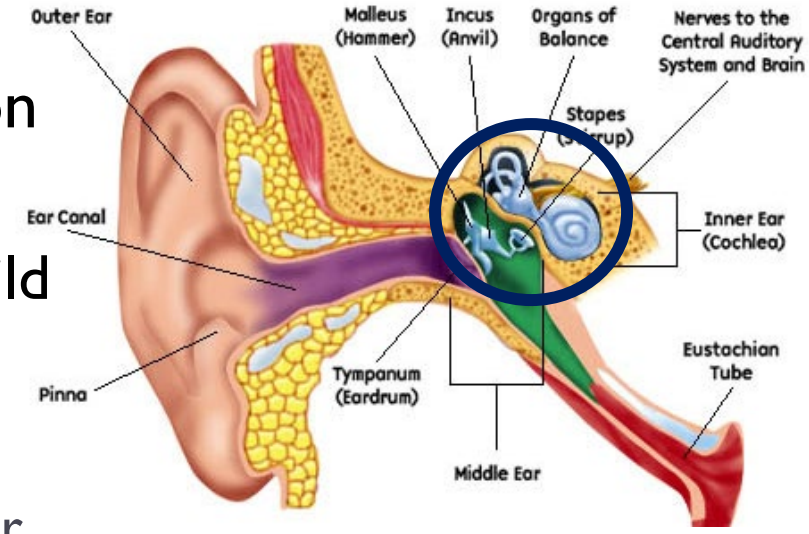
OCT uses high-resolution cross-sections of the retina to determine the retinal nerve fiber layer (RNFL) thickness – risk stratify for optic neuropathy



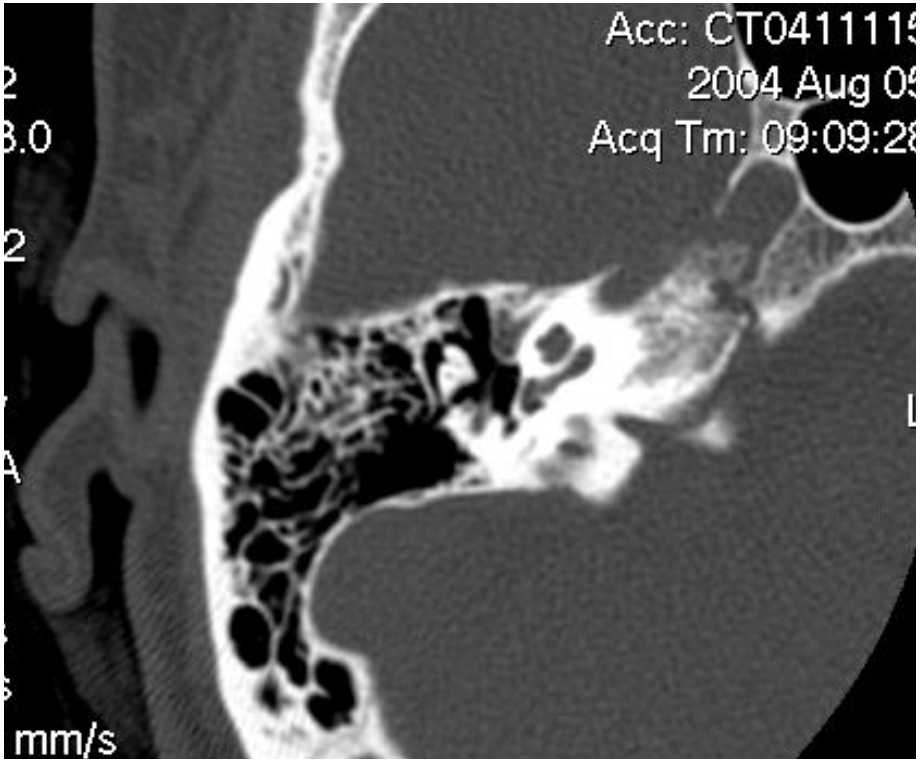
Lee JS et al, NEJM, 2002
Pan 2020 JBMR

Fibrous Dysplasia and Hearing Loss

- Abnormal tympanogram is the most common audiologic finding in PFD
- Ear canal narrowing most common
- Hearing loss occurs, but is mostly mild
- Ear canal cholesteatoma – rare
  - Abnormal skin growth in the middle ear behind the eardrum
  - Secondary to narrowing
  - Can lead to destruction of bones and hearing loss

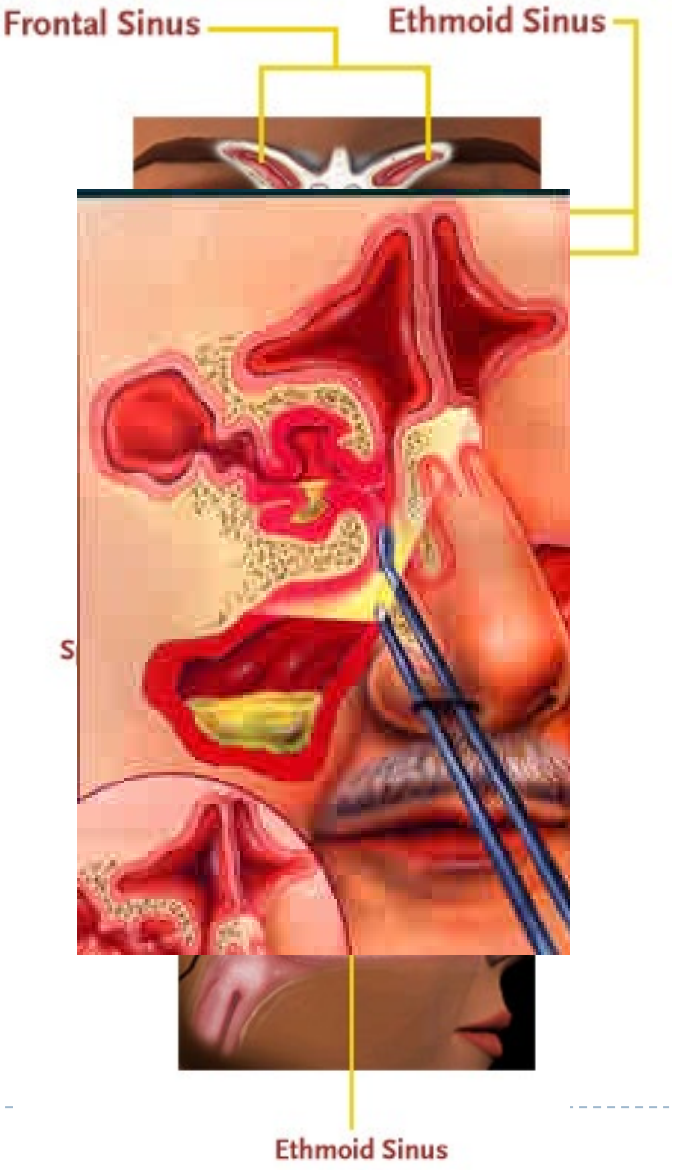


Fibrous Dysplasia and Hearing Loss



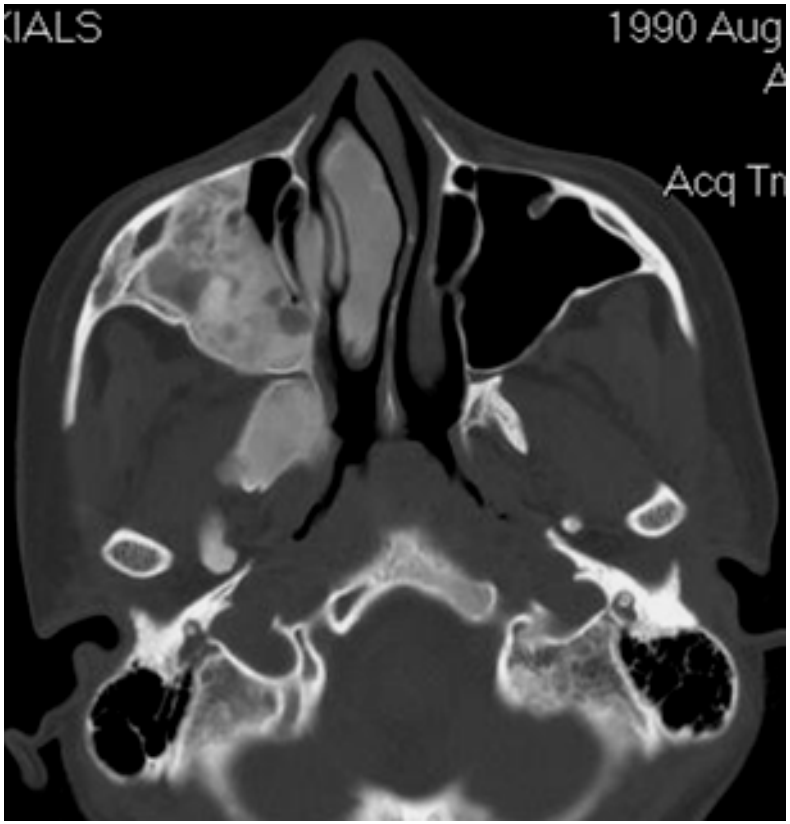
Fibrous Dysplasia and Congestion

- Well-documented that nasal cavity and sinuses are affected in FD
- Nasal congestion is the most common paranasal sinus problem in FD
- True sinus infections are not as frequent as previously thought

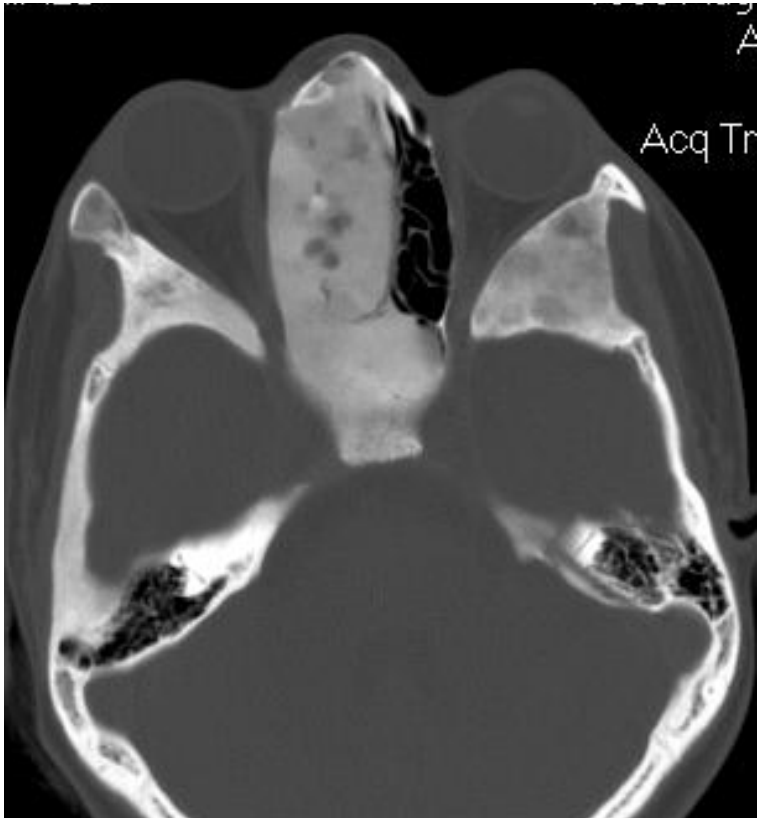


Fibrous Dysplasia and Congestion

Maxillary sinus & turbinate

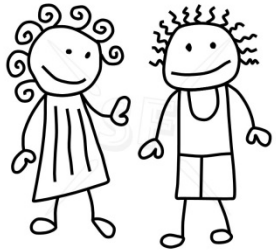


Ethmoid & Sphenoid sinuses



Types of Dental Specialists

- Pediatric Dentist



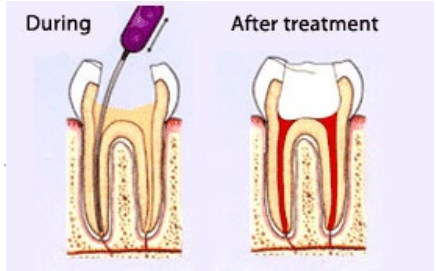
- Orthodontist (dentofacial orthopedics)

- Oral and Maxillofacial Surgeon

- Prosthodontist

- Periodontist

- Endodontist

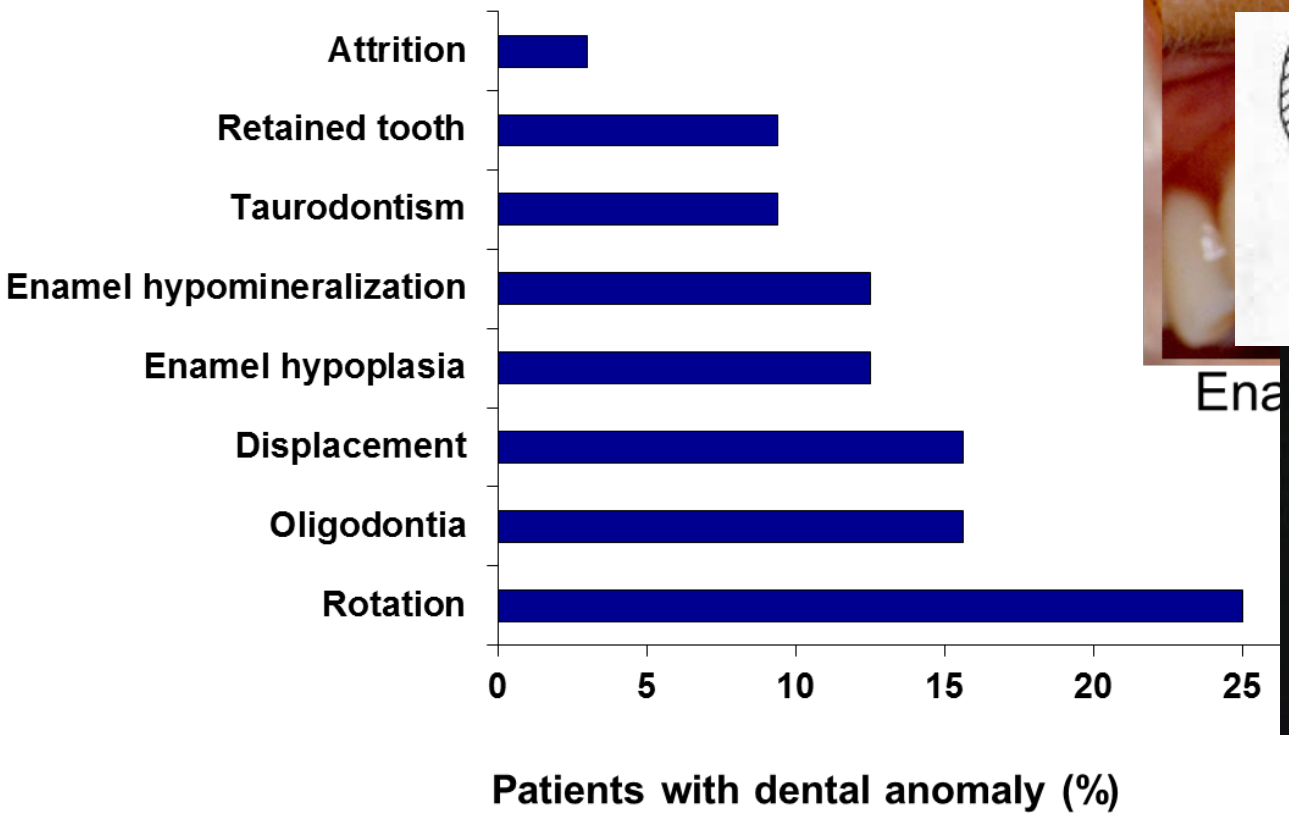


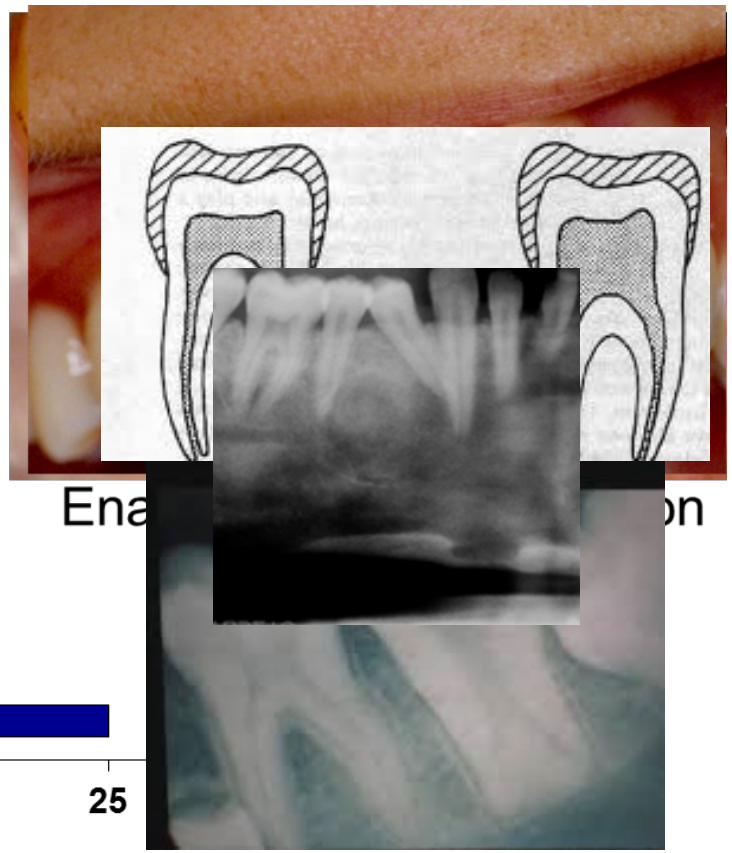
Dental Problems

- Because of often complex co-morbidities, dental aspects are frequently overlooked
  - Variable presentations cause some dental practitioners to delay or avoid dental procedures
- Patients can receive various dental therapies (restorations, root canals, extractions) without exacerbating lesions
- Increased rate of cavities
  - May require more frequent hygiene visits, electric toothbrush, application topical fluoride
- Medication-induced osteonecrosis of the jaws (ONJ)
  - Rare despite higher doses

Akintoye, S. et al, *OOOO*, 2013

Dental Anomalies in FD Patients





43% had dental anomalies
28% anomalies w/in FD bone

n = 23 pts PFD/MAS + 9 pts MFD

Akintoye, S. et al, *OOOO*, 2003

Dental Problems

- Tooth extraction may be needed in cases with teeth "floating," or if they are impacted

- In severe instances, children may require prostheses which need adjustment with growth
  - Can improve function and aesthetics

Papadaki, *Orphanet* 2012

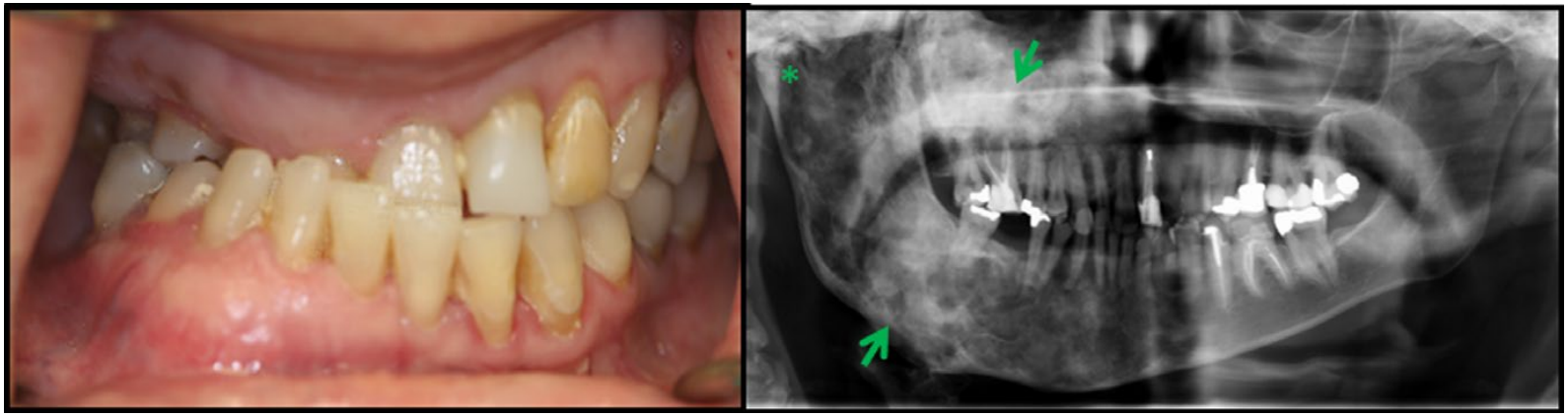
Orthodontic Therapy

- The majority of patients with jaw FD have malocclusion (disorganized arrangement of the teeth)

- Malocclusion seems to be significantly correlated with growth-hormone excess and other endocrinopathies *(unpublished data)*

- Dental malocclusion/crowding
  - Orthodontic treatment is *safe* to perform in FD patients
  - Orthodontic therapy may take shorter (or longer) in FD bone?
  - Relapse may be more common because teeth tend to return to their initial position

Malocclusion



- Delay orthodontics until skeletal maturity?
  - Does not seem to be necessary in FD
- Orthognathic surgery (jaw repositioning) + orthodontics is also an option for severe malocclusion
  - Hold off until skeletal maturity
  - Titanium plates were safe and did not require removal
  - Healing occurred normally after bones were reset

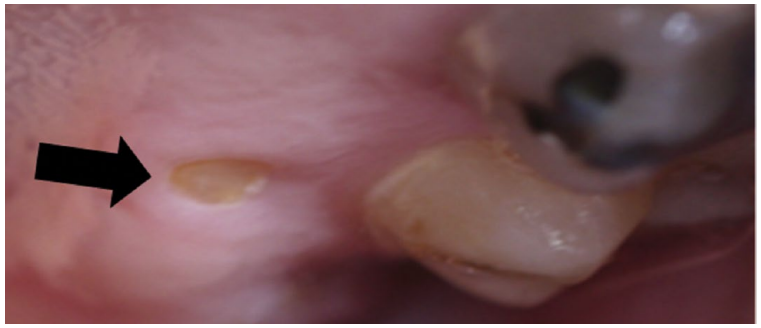
Dental Implants

- Bone healing and integration of the implants occurs
  - May be slower and the quality of bone is thin
- Reported case of 32 yo female w/MAS:
  - Successful integration and loading of dental implants in maxilla and mandible occurred
  - Maxillo-mandibular lesions had been quiescent for 3 years
  - Dental implants were at least 15 mm in length
  - Functional after 5 years
- Literature is limited - unclear risk of implant failure
- Recommend that implant(s) placed after skeletal maturity AND once growth of the FD lesion has subsided

Bajwa, M. et al, *JOMS*, 2008

Medication-Related Osteonecrosis of the Jaws (MRONJ)

- Formerly only BRONJ
  (Bisphosphonate-related)



- What is ONJ?
  - Dead "necrotic" bone due to loss of blood supply
  - Exposed bone for > 8 weeks with history of drug exposure

- Case Reports:
  - Bisphosphonates
  - Denosumab
  - Anti-angiogenic drugs
  - Tyrosine-kinase Inhibitors

- Why does it affect the jaws more???

- NIH Cohort: 5.4% prevalence of BRONJ

Metwally et al. *JOMS*, 2016

How is CF-FD Treated?

- Management based on the extent, aggressiveness and clinical behavior

- Management depends on age/skeletal maturity and clinical findings
  - Lesions can be characterized as:
    - Quiescent (stable with no growth)
    - Non-aggressive (slow growing)
    - Aggressive (rapid growth +/- pain, paresthesia, etc.)

- Monitor, observation and close follow-up
  - Clinical assessment, including photographs
  - Sensory nerve testing
  - Maxillofacial CT – depending on clinical findings

- Medical management – bone pain

Lee, JS et al, *Orphanet*, 2012

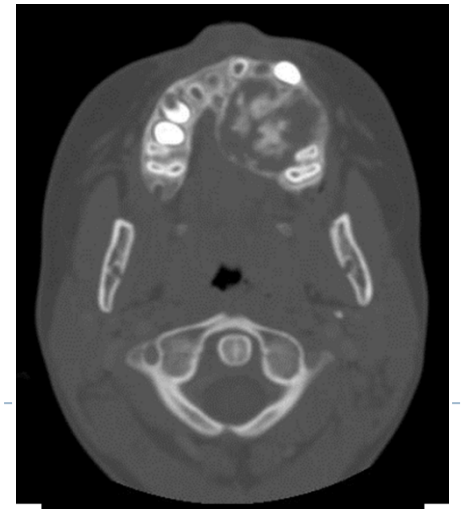
How is CF-FD Treated?

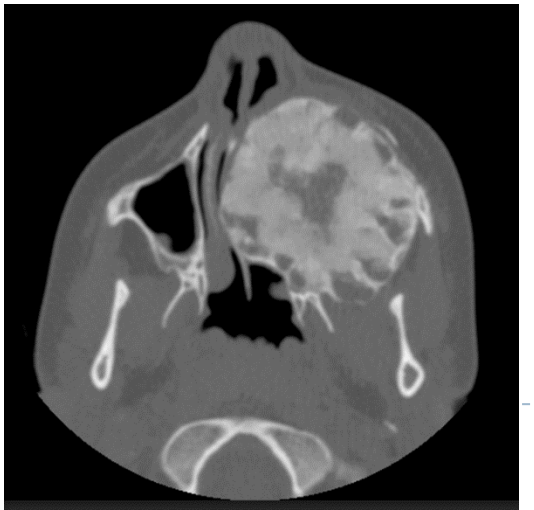
- Indications for surgery:
  - Biopsy if diagnosis is questionable
  - Concern for aggressive growth/atypical or unusual behavior

- Surgical options:
  - "Contouring" – should be done after growth has stopped
  - Resection and reconstruction – if lesion can be completely removed or if there is concern for associated disease/malignancy
  - Orthognathic surgery – correct malocclusion or facial/dental asymmetry
    - No documented contraindication if lesions are quiescent
    - Bone healing is normal

How is CF-FD Treated?

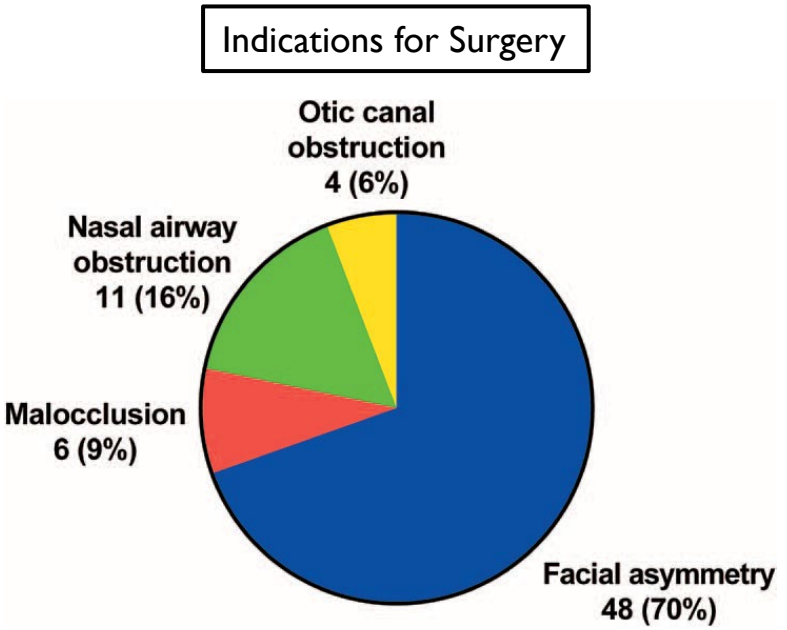
- *However,* there is evidence that lesions can regrow after surgery and even become aggressive
- We cannot predict or prevent regrowth
- Surgery is not typically indicated for cosmetic purposes
  - Dependent on psychosocial situation





NIH CF-FD Outcomes Data

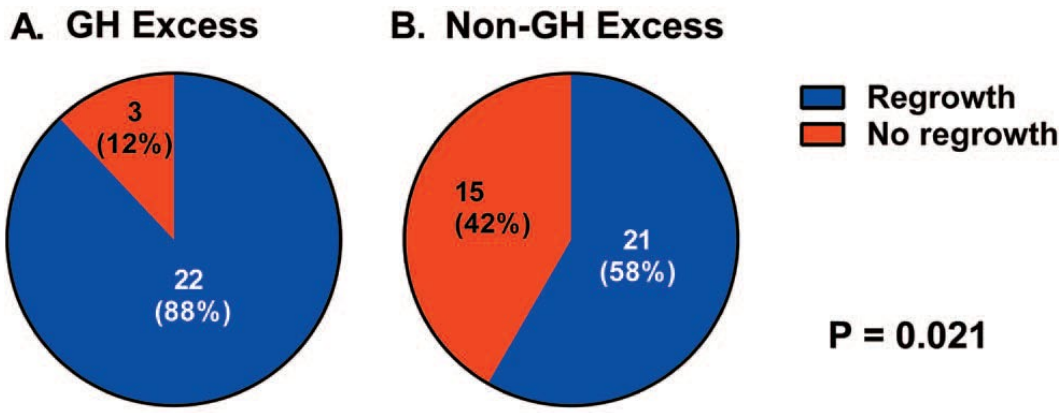
- CF-FD regrowth and reoperation are common, particularly after "debulking" or "recontouring" procedures
- Less regrowth outcomes with Aneurysmal Bone Cysts (ABCs) and bone biopsies
- Bone resection and reconstruction with hardware and/or grafting material may result in less regrowth and fewer re-operations, but has increased morbidity



Boyce et al., *PRS* 2016

NIH CF-FD Outcomes Data

- MAS w/GH-excess is a risk factor for regrowth and may go undiagnosed perioperatively



- Importance of a multidisciplinary approach
  - Surgical and medical practitioners
  - Need for individualized care with long-term follow-up

Boyce et al., *PRS* 2016

Regrowth

$$\text{Tumor growth rate (\%per year)} = \frac{\frac{\text{Mass volume 2}}{\text{Mass volume 1}} - 1}{\text{Age 2} - \text{Age 1}} \times 100$$

Variable	Value	No. of calculations of growth rate	Mean age at diagnosis ± SD, y	Mean tumor volume at diagnosis ± SD, cm^3	Mean growth rate of tumor ± SD, %/y	*P*
Age, y	<16	33	10.4 ± 3.3 (range, 4.6-15.2)	72.7 ± 86.5 (range, 16.8-209.0)	23.1 ± 17.3 (range, 0.1-56.9)	<.001
	≥16	8	24.6 ± 14.2 (range, 16.2-53.2)	85.9 ± 56.6 (range, 39.4-127.2)	2.7 ± 2.7 (range, 0.5-7.9)	
Tumor type	Monostotic	33	14.7 ± 10.9 (range, 4.6-53.2)	53.7 ± 30.4 (range, 16.8-127.2)	19.9 ± 18.1 (range, 0.1-56.9)	.99
	Polyostotic	8	11.8 ± 2.7 (range, 8.4-15.2)	186.1 ± 96.8 (range, 50.1-304.3)	15.7 ± 15.6 (range, 0.6-40.6)	

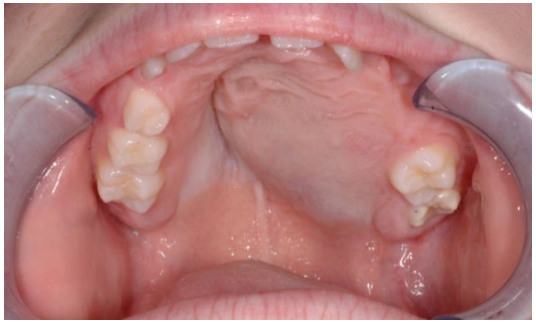
TABLE 3 Comparison of the postoperative regrowth rate of craniofacial fibrous dysplasia according to age at the time of operation

Group	No. of calculations of regrowth rate	Mean age at diagnosis ± SD, y	Mean tumor volume at diagnosis ± SD, cm^3	Mean age at shaving procedure ± SD, y	Mean growth rate of tumor ± SD, %/y	*P*
<16 y old	12	9.3 ± 3.1 (range, 5.0-15.3)	128.4 ± 132.1 (range, 15.2-349.1)	12.1 ± 2.5 (range, 8.3-15.4)	17.1 ± 12.3 (range, 5.4-49.3)	.04
≥16 y old	16	19.7 ± 12.5 (range, 8.3-53.2)	130.9 ± 90.4 (range, 39.0-312.5)	24.0 ± 11.0 (range, 16.0-55.0)	4.7 ± 4.6 (range, 0.1-14.6)	

- Preoperative tumor growth rates were significantly lower in patients aged ≥ 16 years than those aged < 16 years (P < .001)
- Postoperative tumor regrowth rates were significantly greater when a shaving operation was performed at age < 16 years than at age ≥ 16 years (P = .04)

Park et al. 2020

How is CF-FD Treated?



Photos c/o Dr. Kaban, MGH

Surgical Considerations

- Age
- Rate of Growth
- Anatomic Location
- Type of Involvement
- Functional Disturbance
- Aesthetics

Surgical Options

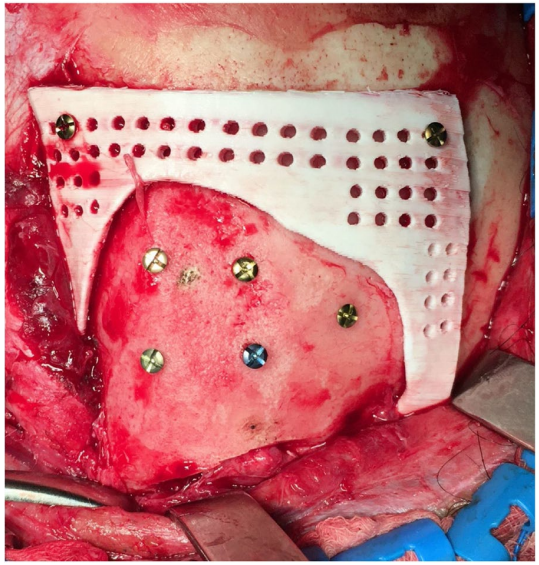
- Deep bone biopsy*
- Recontouring
- Resection +/- Reconstruction

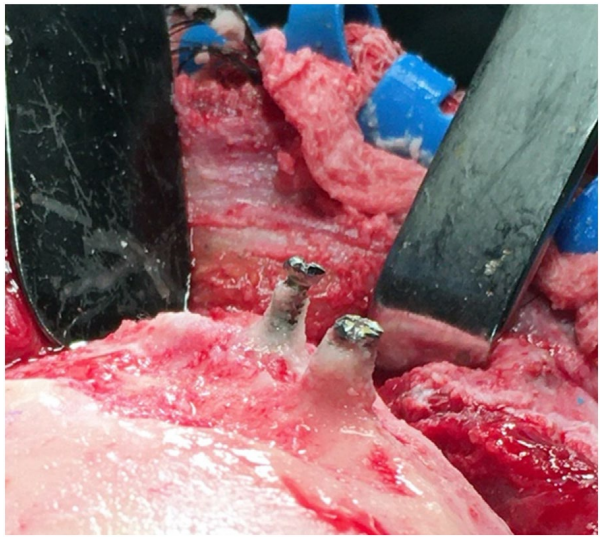
- There are no uniformly accepted guidelines
  - Controversies:
    - prophylactic orbital decompression
    - best surgical management (conservative vs radical approaches)
    - surgical options in patients with immature craniofacial skeleton

Surgical Advances: 3D Recontouring

Three-dimensional surgical contouring of craniofacial fibrous dysplasia via patient-specific cutting guides and depth screws

J. Rengarajoo [a,b,*], S.I. Syed Husman [b], F. Hariri [a] British Journal of Oral and Maxillofacial Surgery 59 (2021) 843–844



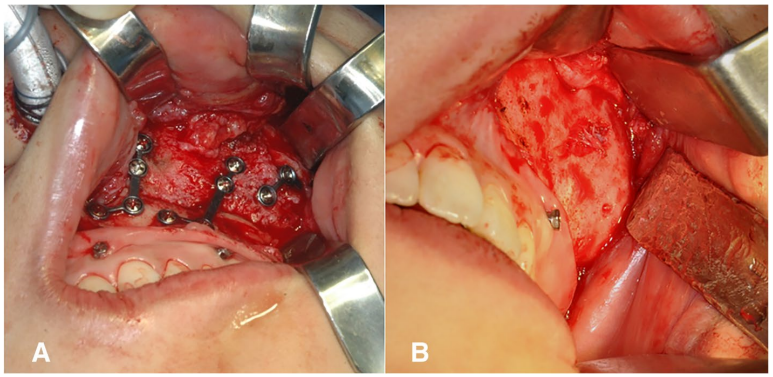


Surgical Advances: Reconstruction

Calvarial bone graft fixed to zygoma and orbital floor

Surgical Advances: Orthognathic Surgery

- Recontouring often carried out at same time
- Difficult to tightly fix the screws and miniplates to the fibrodysplastic bone
- Longterm stability of the osteotomized segments and occlusion
- Osteointegration was observed between the screws and dysplastic bone

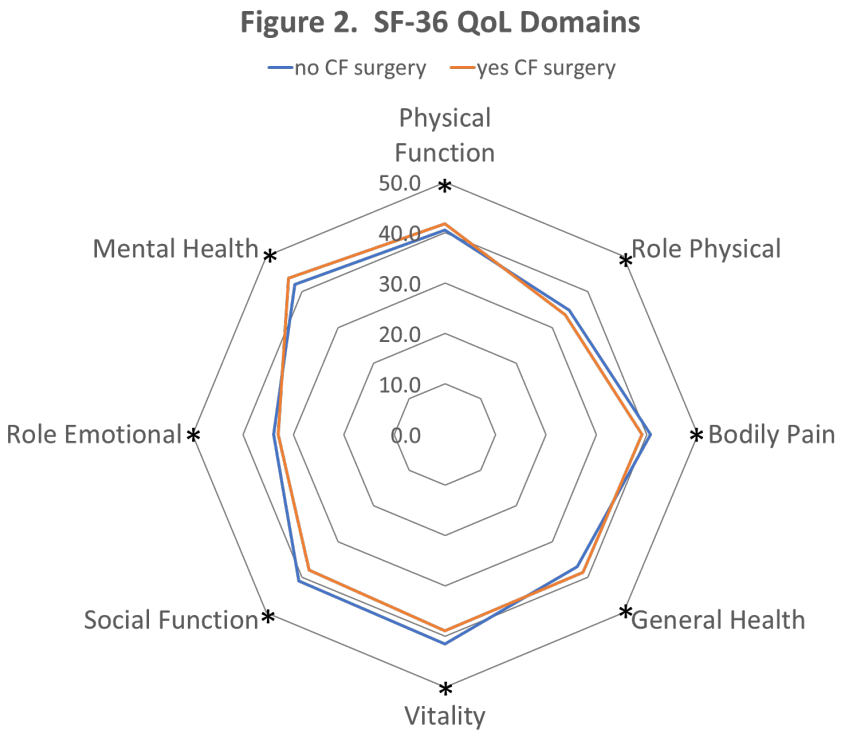


Intraop

2 yrs – hardware removal

4 yrs

Outcomes: QoL



- Craniofacial surgery for FD does not significantly impact quality of life in subjects who have received operative treatment compared to those who have not.
- SF-36 summary values were all significantly lower than the 1998 US population norms.
- Surgical therapy alone, in terms of its occurrence or its number of occurrences, was not significantly related to long-term physical or mental health outcomes.
- Although the bivariate relationship between number of surgeries and mental health was not significant, it suggests a positive association.

Thank you!

Thank you to all of the patients & families

Drs. Michael Collins, Pam Robey, Alison Boyce & Janice Lee
(NIDCR, NIH)

Dr. Jeff Kim
Dr. Sunday Akintoye
Dr. Leonard Kaban
Dr. Janice Lee
Dr. Thomas Dodson