To Operate or Not to Operate?
Quality of Life Outcomes in Craniofacial Fibrous Dysplasia

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INTRODUCTION

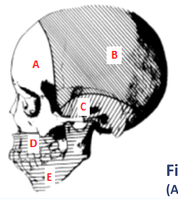
- Fibrous dysplasia (FD), a rare disease of bone marrow stromal cells, arises from somatic activating mutations of *GNAS*.
- The clinical spectrum of FD is broad and lesions may affect one bone (monostotic, MFD), multiple bones (polyostotic, PFD), or may occur in association with café-au-lait macules and hyperfunctioning endocrinopathies in McCune-Albright syndrome (MAS).
- The craniofacial bones are the most frequently affected. Craniofacial FD (CFD) may range from an asymptomatic, incidental lesion to severe deformity of facial features, with potential to cause functional/esthetic problems or pain.
- The mainstay of treatment for CF-FD is surgical. However, regrowth after surgery is common, with subjects requiring multiple operations, leading to associated pain and increased risk of morbidity.
- Given the variable severity of CFD and important psychosocial implications, further investigation is necessary when taking into account demographic data, disease variables and stigma.

AIM

Among patients with CFD, do those treated operatively, when compared to those managed non-operatively, have improved QoL outcomes?

METHODS

- A retrospective cross-sectional study of adult subjects enrolled in the Fibrous Dysplasia Foundation Patient Registry, July 2016-December 2018.
- The **primary predictor variable** was operative treatment (no surgery, one surgery, more than one surgery).
- **Covariates** included: Diagnosis (MFD, PFD, MAS); Craniofacial disease burden (# of CF zones affected, **Figure 1**); Demographic data (age, gender, race/ethnicity, education).
- **Outcome measures** were quality of life (QoL) measures obtained from the validated SF-36 questionnaire
- Univariate analysis of demographics and medical characteristics, SF-36 (8 domains and total scores computed and normed). Bivariate analysis: Mann Whitney, Kruskal Wallis and Spearman's correlation were conducted to determine whether surgery (or multiple surgeries) resulted in significantly different SF-36 domain scores ($p<.05$).



Zone A – frontal, orbital, zygoma, midface, nasal bones
Zone B – cranium, parietal, occipital bones
Zone C – temporal bones, skull base
Zone D – maxilla
Zone E – mandible

Figure 1. Disease Burden – Craniofacial Bones Affected (Adapted from Chen and Noordhoff)

RESULTS

Table 1. Demographic Variables

N=117	Values	Frequencies (%)
Age (Mean = 40 yrs)	18-24 years	17 (15)
	25-34 years	26 (22)
	35-44 years	28 (24)
	45-54 years	27 (31)
	55-64 years	12 (10)
	65-69 years	3 (3)
Gender	men	19 (16)
	women	98 (84)
Race	Caucasian	107 (92)
	Nonwhite	10 (8)
Ethnicity (N=96)	Hispanic	8 (8)
	Non-Hispanic	88 (92)
Participant's Education	K-12	2 (2)
	Graduate HS	23 (20)
	Some college	34 (29)
	Graduated College	39 (33)
	> college	19 (16)

Table 2. Subject Characteristics

	Values	Frequency (%)
FD Diagnosis (N=117)	Monostotic	28 (24)
	Polyostotic	58 (50)
	McCune-Albright Syndrome	31 (27)
Participant-Reported Areas Affected (N = 117)	Zone A Affected	80 (68)
	Zone B Affected	70 (60)
	Zone C Affected	73 (62)
	Zone D Affected	44 (38)
	Zone E Affected	34 (29)
Total CF Disease Burden = # Zones Affected by Subject Report (N = 117)	1 Zone	38 (33)
	2 Zones	28 (24)
	3 Zones	19 (16)
	4 Zones	10 (8)
	5 Zones	22 (19)
Participant-Reported Craniofacial Pain (N = 99)	Yes	92 (93)
Participant-Reported Surgery non-craniofacial bones (N = 109)	Yes	29 (27)

Table 3. Surgical Characteristics

	Values	Frequency (%)
Surgical Variables	**YES CF Surgery**	49 (42)
	1 surgery	24 (21)
	2 or more surgeries	25 (21)
	NO CF Surgery	68 (58)
Mean total # of surgeries	1.11 (SD 1.94)	
Location of Craniofacial Surgery (N = 49)	Zone A treated	29 (42)
	Zone B treated	28 (59)
	Zone C Treated	20 (41)
	Zone D Treated	7 (14)
	Zone E Treated	15 (31)
Participant- Reported Indication for Surgery (N = 40)	Diagnosis	17 (42)
	Aesthetics	21 (52)
	Improve Function	19 (47)
	Pain	17 (42)
	Other	20 (50)

Figure 2. SF-36 QoL Domains

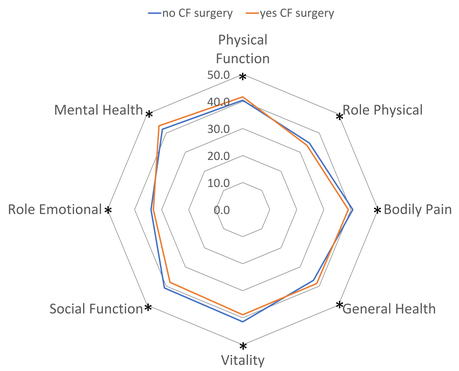


Figure 2. Radar Plot SF-36 Quality of Life Domains comparing craniofacial surgery and no-surgery groups to the 1998 US norms. The craniofacial fibrous dysplasia (CFD) population mean domain scores for the "no surgery" group are represented by the **blue** line; the mean domain scores for the "yes surgery" group are represented by the **orange** line. The national norms are represented by an asterisk (*). Physical parameters include physical function, role physical, bodily pain and general health. Mental parameters include vitality, social function, role emotional and mental health.

- 68% of the sample self-reported a facial deformity.
- Comparison of the surgically treated and nontreated groups revealed race was significantly related to treatment status ($X^2= 4.57$, $p= 0.03$), with nonwhites receiving less surgical treatment.
- Diagnosis (MFD, PFD or MAS) was not significantly associated with surgical treatment.
- Zone A was most reportedly affected by disease (68%), followed by zone C, zone B, zone D and zone E (**Figure 1**). Zone A was also reported to be the most surgically treated region.
- Comparison of the surgically treated and nontreated groups revealed disease burden was significantly associated with treatment ($X^2= 11.069$, $p= 0.026$). Quantity of surgeries was also significantly associated with craniofacial disease burden ($F=4.369$, $p= 0.015$).
- Independent samples T-tests revealed that mean summary and domains scores on the SF-36, when comparing participants who received craniofacial surgery and did not receive craniofacial surgery, were not significantly different. All mean domain scores on the SF-36 for both groups were all significantly below 1998 population norms (**Figure 2**).

CONCLUSIONS

- Craniofacial surgery for FD does not significantly impact quality of life in subjects who have received operative treatment compared to those who have not.
- Limitations include a small sample size for a rare disease and potential bias resulting from patient-reported outcomes.
- Along with surgery, treatment of CFD should address the social effects of the disease. Surgeons need to be aware of the psychosocial implications when making the decision to operate on CFD patients.

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