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Surgical Management of Polyostotic Craniofacial Fibrous Dysplasia: Long-Term Outcomes and Predictors for Postoperative Regrowth

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Abstract

Background—The mainstay of treatment for craniofacial fibrous dysplasia is surgical; however, optimal indications and techniques are poorly understood, particularly in polyostotic disease and McCune-Albright syndrome. This study investigated surgical indications and risk factors for recurrence in a large cohort.

Methods—One hundred thirty-three craniofacial fibrous dysplasia subjects in a natural history study were evaluated. Radiographic studies, operative reports, and clinical records were reviewed.

Results—Thirty-six subjects underwent 103 craniofacial procedures (mean, 2.8 operations per subject), with 13.5 ± 10.5 -year follow-up (range, 0 to 39 years). The most common indication was craniofacial deformity ($n = 61$ operations), including 36 initial operations (59 percent) and 26 reoperations (41 percent). Mean time to reoperation was 3.4 ± 3.2 years (range, 0.3 to 13.3 years). Re-growth occurred after 42 operations (68 percent), and was more frequent after operations in subjects with McCune-Albright syndrome growth hormone excess [22 of 25 operations (88 percent)] than without growth hormone excess [15 of 36 operations (58 percent); $p = 0.02$]. Of 11 subjects with growth hormone excess, nine (82 percent) were undiagnosed at the time of their initial operation. Regrowth was more frequent after debulking procedures [31 of 38 (82 percent)] than after more aggressive reconstructions [nine of 20 (45 percent); $p = 0.007$]. Eleven subjects underwent treatment for aneurysmal bone cysts, with recurrence in one subject. Eleven subjects underwent biopsies and none had complications or regrowth.

Conclusions—Craniofacial fibrous dysplasia regrowth and reoperation are common, particularly after debulking procedures. Outcomes are favorable for aneurysmal bone cysts and biopsies. McCune-Albright syndrome growth hormone excess is a risk factor for regrowth, and may be underdiagnosed in surgical patients. Surgeons should be aware of appropriate screening for

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endocrinopathies in fibrous dysplasia. These findings highlight the importance of a multidisciplinary approach to craniofacial fibrous dysplasia, and individualized care with long-term follow-up.

CLINICAL QUESTION/LEVEL OF EVIDENCE—Therapeutic, IV.

Fibrous dysplasia of bone is an uncommon skeletal disorder in which bone and bone marrow are replaced by fibro-osseous tissue. It arises from somatic activating mutations in *GNAS*, which lead to constitutive activation of the α -subunit of the G_s -coupled protein receptor.¹ In the skeleton, these mutations lead to proliferation of undifferentiated skeletal progenitor cells, resulting in bone that is weak and prone to expansion, deformity, and pain.² Lesions may affect one bone (monostotic) or multiple bones (polyostotic), and may occur in isolation or in association with café-au-lait macules and hyperfunctioning endocrinopathies, termed McCune-Albright syndrome.³

The craniofacial bones are the most frequently affected in patients with fibrous dysplasia.⁴ Similar to fibrous dysplasia in the axial and appendicular skeleton, craniofacial fibrous dysplasia occurs along a broad spectrum of severity, ranging from an asymptomatic incidental lesion to severe disabling disease. The current mainstay of treatment is surgical; however, optimal indications, techniques, and timing are poorly understood. The best defined surgical indication is optic nerve encasement, in which large cohort studies have demonstrated the superiority of expectant management over prophylactic decompression^{5,6} and established untreated growth hormone excess as an independent risk factor for vision loss.⁷ In contrast, there is little evidence to inform outcomes for other surgical indications in craniofacial fibrous dysplasia.

Surgical techniques in craniofacial fibrous dysplasia are controversial. Previous studies have advocated radical subtotal resection with or without reconstruction over more conservative debulking or recontouring techniques; however, the optimal approach has not been established.^{8–11} The extent of bone involvement and anatomical location largely determine operative approach. Although partial resections may be complicated by postoperative regrowth and recurrence of the deformity, it is not feasible to treat most cases of extensive craniofacial fibrous dysplasia by total resection secondary to increased morbidity and risk of complication.

Defining surgical management in craniofacial fibrous dysplasia is also limited by the lack of surgical outcome predictors. For example, it is unknown whether craniofacial surgery during childhood, when fibrous dysplasia lesions tend to be more active, is associated with higher rates of regrowth. Furthermore, there are few case reports describing the management of aneurysmal bone cysts in association with craniofacial fibrous dysplasia, but the prevalence and long-term outcomes in these patients are unknown.⁸ Current literature focuses largely on case reports detailing the management of isolated monostotic lesions rather than polyostotic disease or the effect of McCune-Albright syndrome–associated endocrinopathies. In this study, we retrospectively investigated long-term outcomes from craniofacial surgery in a large cohort of subjects with polyostotic fibrous dysplasia/McCune-Albright syndrome to determine the surgical indications, recurrence rates, and reasons for reoperation, and to identify risk factors for postoperative regrowth.

PATIENTS AND METHODS

Subjects in a longstanding fibrous dysplasia/McCune-Albright syndrome natural history database at the National Institutes of Health were evaluated. This study was approved by the National Institute of Dental and Craniofacial Research Institutional Review Board, and all subjects and/ or their guardians gave informed consent/assent. Subjects were evaluated at the National Institutes of Health Clinical Center between 1998 and 2015, where they underwent history and physical examination, biochemical testing, vision and hearing evaluation, photography, and radiographic imaging. Subjects were evaluated for McCune-Albright syndrome-associated endocrinopathies, including the following: precocious puberty (bone age examination and sex steroid hormone levels), hyperthyroidism (thyroid function tests and thyroid ultrasound), growth hormone excess (insulin-like growth factor-1, oral glucose tolerance testing, and serial overnight growth hormone testing), neonatal hypercortisolism (history and physical examination), and hypophosphatemia (serum and urine phosphorus levels). Craniofacial surgical history was obtained from subject and/or parent report, and operative reports and surgical records were collected and reviewed. Surgical outcomes were determined by review of serial computed tomographic scans in addition to subject interview and review of records. Fisher's exact test and the unpaired *t* test were used to make comparisons between subject groups and to identify predictors of postoperative fibrous dysplasia progression. Statistics and figures were prepared using GraphPad Prism 6 for Windows, Version 6.02 (GraphPad Software, Inc., San Diego, Calif.).

RESULTS

Subject Characteristics

Of 169 subjects with polyostotic fibrous dysplasia/McCune-Albright syndrome, 133 (79 percent) had craniofacial fibrous dysplasia and, of these, 36 subjects underwent at least one craniofacial surgical procedure. Subject characteristics are listed in Table 1. Subjects who underwent craniofacial surgery had more craniofacial fibrous dysplasia involvement than nonsurgical subjects as determined by skeletal disease burden score, a validated tool to quantify fibrous dysplasia bone involvement⁴ ($p = 0.01$) (Table 1). The most common locations for craniofacial fibrous dysplasia involvement were the calvaria and skull base, followed by the maxilla and mandible. There were no differences in fibrous dysplasia location between the two groups (surgical and nonsurgical), with the exception of the maxilla, which was affected more frequently in the surgical group ($p = 0.04$). Subjects who underwent surgery had a higher prevalence of vision loss ($p = 0.0002$) and facial deformity ($p = 0.004$) in comparison with nonsurgical subjects. There was no difference in hearing loss between the two groups, and no difference in craniofacial pain, which occurred commonly in approximately 40 percent of subjects. The prevalence of endocrinopathies was not different between the surgical and nonsurgical groups.

Surgical Procedures

One hundred three craniofacial operations were performed on 36 subjects (mean, 2.8 ± 2.1 procedures per subject; range, one to nine). Sixty-two (60 percent) of all operations were initial operations, and 41 (40 percent) were reoperations. Surgical indications were

determined by subject report and review of available surgical records. Operations were classified based on the following indications: aneurysmal bone cyst, biopsy, deformity, optic nerve decompression, and miscellaneous (Table 2). Fourteen procedures were performed for more than one indication (e.g., treatment of aneurysmal bone cyst in addition to correction of deformity at a separate site); in these procedures, outcomes for each indication were analyzed separately.

The most frequent surgical indication was correction of craniofacial deformity (60 percent of all procedures). Of procedures performed to treat deformities, the majority were performed for correction of facial asymmetry (70 percent), with a smaller minority performed to treat functional impairments including nasal airway obstruction (11 percent), malocclusion (9 percent), and otic canal obstruction (6 percent) (Fig. 1). Eight procedures (8 percent) were performed for miscellaneous indications, including the following: Chiari decompression ($n = 2$), cranial nerve palsy ($n = 2$), debulking of fibrous dysplasia–associated sarcoma ($n = 3$), and dental implant placement ($n = 1$).

Surgical Outcomes

The mean length of follow-up for the group of all operations was 13.5 ± 10.5 years (range, 0 to 39 years). Surgical outcomes are listed in Table 2. Indications and outcomes for optic nerve decompressions in this cohort were previously reported in detail in a meta-analysis by Amit et al.⁵ and were therefore not investigated further in the current analyses.

Operations to correct craniofacial deformities were associated with high rates of fibrous dysplasia regrowth, affecting 68 percent of procedures. Reoperations were performed after 43 percent of initial procedures, with a mean time to reoperation of 3.4 ± 3.2 years (range, 0.3 to 13.3 years). Outcomes for aneurysmal bone cysts were favorable, with recurrence and reoperation after only one procedure (9 percent). Biopsy procedures also showed favorable outcomes, with no fibrous dysplasia regrowth at surgical sites and no complications requiring reoperation after any procedure.

Predictors of Regrowth and Reoperation

Procedures to correct craniofacial deformities were analyzed to determine correlations between clinical and surgical features and the prevalence of reoperation and postoperative regrowth, respectively. Because of the low rates of regrowth/recurrence after aneurysmal bone cyst and biopsy procedures, these were excluded from the analyses. The following clinical features were investigated: (1) presence of McCune-Albright syndrome–associated endocrinopathies, including growth hormone excess, hyperthyroidism, precocious puberty, and phosphate wasting; (2) age at operation; (3) fibrous dysplasia location, classified according to zones of involvement defined by Chen and Noordhoff⁹ (zone 1, facial area above the maxillary alveolar bone; zone 2, hair-bearing cranium; zone 3, central cranial base, petrous, mastoid, and pterygoid region; and zone 4, maxillary alveolar bone and mandible); (4) postoperative treatment with bisphosphonates; and (5) surgical technique, broadly defined as debulking (partial removal and/or recontouring of fibrous dysplasia) and reconstruction (partial resection of fibrous dysplasia bone with introduction of hardware and/or grafting material). Documentation was not sufficient to confidently categorize fibrous

dysplasia location for one procedure, or to categorize debulking versus reconstruction for three procedures; these were therefore excluded from the outcome analyses. Total resection of fibrous dysplasia was not performed in any subject because of the extent of disease.

Growth hormone excess was associated with a higher prevalence of postoperative fibrous dysplasia regrowth ($p = 0.02$) (Fig. 2). There was no association between other endocrinopathies or age and fibrous dysplasia regrowth or reoperation. There was no association between fibrous dysplasia location and postoperative regrowth; however, reoperation was more frequent after procedures performed in zone 4 (maxillary alveolar bone and mandible) ($p = 0.04$). Postoperative bisphosphonates, including zoledronic acid, pamidronate, alendronate, and risedronate, were administered to nine subjects after a total of 13 procedures. There was no difference in reoperation or postoperative regrowth in treated versus untreated subjects. Reconstruction procedures were associated with a lower prevalence of both regrowth and reoperation in comparison with debulking procedures ($p = 0.007$ and 0.03 , respectively) (Fig. 2).

DISCUSSION

Studies of the surgical management of craniofacial fibrous dysplasia have been limited by multiple challenges, including the rarity of the disease, the heterogeneity of the patient population, and diagnostic uncertainties. The goals of surgical treatment include correction of functional problems and/or aesthetic enhancement. The existing literature focuses primarily on treatment of craniofacial deformity, reporting variable recurrence rates ranging from 25 to 80 percent.^{10–14} In this series, we found a high prevalence of postoperative regrowth and reoperation in 68 and 43 percent of procedures, respectively. Given these dissatisfying outcomes, there is a critical need to identify reliable preoperative prognostic indicators to guide surgical planning. We demonstrated that procedures in subjects with McCune-Albright syndrome–associated growth hormone excess had a significantly higher prevalence of postoperative regrowth. This is a novel finding that provides practitioners with a means of preoperatively identifying a subset of patients at increased risk for regrowth. It also demonstrates the importance of multidisciplinary collaboration between surgical and medical practitioners to ensure that patients are evaluated and treated for endocrinopathies as part of routine preoperative care. In our cohort (where the majority of subjects underwent surgery at outside centers before referral), surgeons were frequently unaware of McCune-Albright syndrome–associated endocrine comorbidities: of subjects with growth hormone excess, nine of 11 (82 percent) were not diagnosed or treated before undergoing their initial craniofacial procedure. To prevent these inconsistencies in care, we recommend that all patients with suspected features of fibrous dysplasia/McCune-Albright syndrome undergo a staging evaluation, typically performed by an endocrinologist, at the time of initial diagnosis to establish the extent of disease involvement. Specific guidelines are reported by Boyce and Collins.³ Patients with fibrous dysplasia/McCune-Albright syndrome are frequently seen by surgeons for their initial presentation to care. Increasing awareness of the critical need for endocrine screening among surgical practitioners thus has the potential to significantly improve postoperative and other clinical outcomes.

Several reports have investigated age at operation as a potential prognostic indicator for surgical outcomes.^{15–17} This is in keeping with the natural history of fibrous dysplasia, where lesions appear to expand during childhood and reach final disease burden around the time of skeletal maturity. Fibrous dysplasia lesion activity is thought to decrease with age because of apoptosis of mutation-bearing mesenchymal cells, leading to histologic improvement¹⁸ and a decrease in fracture rate in older patients.¹⁹ Similar to previous reports, we were unable to definitively demonstrate a relationship between age at surgery and fibrous dysplasia regrowth. Regardless, it is prudent for practitioners to be aware of the natural tendency of fibrous dysplasia to expand in childhood and become more quiescent in adulthood. As such, factoring skeletal growth into surgical treatment planning may avoid multiple procedures and reduce morbidity.

The optimal surgical approach for treatment of craniofacial fibrous dysplasia deformity is an area of controversy. Numerous approaches have been described, including shaving/contouring, debulking, resection, and reconstruction.^{9,11,12,15,20–23} Similar to previous reports, we found a lower prevalence of fibrous dysplasia regrowth after more aggressive reconstruction procedures in comparison with debulking procedures; however, even among reconstructions, regrowth occurred frequently (45 percent of procedures). Encouragingly, there was no postoperative regrowth in any subjects after biopsies of craniofacial fibrous dysplasia lesions.

This study is a retrospective review of procedures at multiple centers over an extended period and is therefore unable to inform strong conclusions regarding technical strategies. However, these findings reinforce the belief that treatment plans should be individualized according to the clinical and psychosocial characteristics of each patient, and that long-term follow-up is of critical importance in all cases.

These analyses also provided the opportunity to investigate surgical outcomes in aneurysmal bone cysts, a rarely reported but highly morbid complication of craniofacial fibrous dysplasia.²⁴ This is the first large series to provide information regarding the prevalence of aneurysmal bone cysts in craniofacial fibrous dysplasia, which affected seven of 133 subjects (5 percent). Two subjects had multiple aneurysmal bone cysts (three each); however, only one of these was the result of recurrence in a previously resected location. These findings are similar to reported outcomes in non-fibrous dysplasia-associated aneurysmal bone cysts, where a recurrence rate of less than 10 percent was seen in a recent large retrospective series of maxillofacial aneurysmal bone cysts.²⁵

Limitations of this study include the retrospective design, the heterogeneity of the patient population, and the multicenter surgical care, which resulted in variability in surgeons providing care, surgical treatment plans, and techniques. The multicenter care also limited our ability to categorize surgical approaches in more detail, such as the magnitude of procedure and the extent of dysplastic tissue removed. A significant limitation was the reliance on subject report for surgical indications in cases where documentation was not sufficiently detailed. Because subjects were evaluated as part of a research study at a tertiary referral center, it is possible this cohort may be biased toward a more severely affected phenotype. This may limit the generalizability of the results; however, it also facilitates

identification of risk factors associated with regrowth and suboptimal surgical outcomes. Strengths of this investigation stem from its design as a natural history study, which allows for extensive clinical and radiographic phenotyping and long-term serial follow-up. To our knowledge, this is the only report of craniofacial surgical outcomes in which all subjects had polyostotic disease, which allows for greater confidence in the diagnosis of craniofacial fibrous dysplasia, as isolated monostotic fibrous dysplasia may be more easily confused with histologically and radiographically similar-appearing fibro-osseous lesions. This is also the first series to investigate the effects of McCune-Albright syndrome–associated endocrinopathies on surgical outcomes.

CONCLUSIONS

Postoperative fibrous dysplasia regrowth and reoperation are common after craniofacial surgery. Patients with McCune-Albright syndrome–associated growth hormone excess are at higher risk for regrowth. Aneurysmal bone cyst and biopsy procedures have favorable outcomes with a low prevalence of regrowth or recurrence. Our study corroborates previous observations suggesting that resection and reconstruction with hardware and/or grafting material may result in less regrowth and fewer reoperations than more conservative debulking and recontouring techniques. These findings highlight the importance of a multidisciplinary approach to craniofacial fibrous dysplasia management, including both surgical and medical practitioners, and the need for individualized care with long-term follow-up.

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Fig. 1. Indications for surgical treatment of craniofacial deformity in fibrous dysplasia. Pie graph showing the number and percentage of procedures performed for specific craniofacial deformities. (Copyright United States Government. For reprint requests, please contact Alison Boyce, M.D.)

Fig. 2.

Clinical features associated with postoperative regrowth in craniofacial fibrous dysplasia. (*Above*) Proportion of procedures with postoperative regrowth (*blue*) and no regrowth (*orange*) in subjects with growth hormone excess (*above, left*) and without growth hormone excess (*above, right*). (*Below*) Postoperative regrowth in debulking (partial removal and/or recontouring of fibrous dysplasia) (*below, left*) and reconstruction procedures (partial resection of fibrous dysplasia bone with introduction of hardware and/or grafting material) (*below, right*). (Copyright United States Government. For reprint requests, please contact Alison Boyce, M.D.)

Table 1

Clinical and Radiographic Characteristics of Subjects with Craniofacial Fibrous Dysplasia

Characteristic	Craniofacial Surgery (%)	No Craniofacial Surgery (%)	<i>p</i>
No. of subjects	36	97	
Female	22 (61)	53 (54)	NS
Age at enrollment, yr			
Mean $\pm$ SD	23.7 $\pm$ 14.1	18.0 $\pm$ 16.3	
Range	5–58	2–80	NS
Mean FD burden score *			
Craniofacial	3.3	2.6	0.001
Total skeleton	24.8	19.5	NS
FD location			
Calvaria	35 (97)	88 (94)	NS
Skull base	36 (100)	88 (94)	NS
Maxilla	32 (89)	66 (70)	0.04
Mandible	25 (69)	55 (60)	NS
Clinical symptoms			
Vision loss	11 (31)	4 (5)	0.0002
Hearing loss	12 (33)	29 (33)	NS
Facial deformity	34 (94)	70 (71)	0.004
Craniofacial pain	15 (42)	38 (39)	NS
Endocrinopathies			
GH excess	11 (31)	17 (18)	NS
Hyperthyroidism	9 (25)	34 (35)	NS
Phosphate wasting	13 (37)	29 (30)	NS
Precocious puberty	20 (56)	53 (54)	NS

NS, not significant; FD, fibrous dysplasia; GH, growth hormone

* From Collins MT, Kushner H, Reynolds JC, et al. An instrument to measure skeletal burden and predict functional outcome in fibrous dysplasia of bone. *J Bone Miner Res.* 2005;20:219–226.

Table 2**Operative Outcomes According to Surgical Indication**

	Deformity (%)	Aneurysmal Bone Cyst (%)	Biopsy (%)	Optic Nerve Decompression (%)[*]
No. of operations	61	11	11	19
Regrowth or recurrence	42 (68)	11 (9)	0	6 (50) [†]
Reoperations	26 (43)	11 (9)	0	4 (21)
Time to reoperation, yr				
Mean $\pm$ SD	3.4 $\pm$ 3.2	3	N/A	5.6 $\pm$ 7.5
Range	0.3–13.3			0.3–19.0
Length of follow-up, yr				
Mean $\pm$ SD	13.9 $\pm$ 10.9	17.1 $\pm$ 9.1	4.1 $\pm$ 5.9	13.1 $\pm$ 10.3
Range	0–39.0	2.4–33.0	0–16	0.3–31

N/A, not applicable.

^{*} Twelve clinically indicated decompressions were performed in subjects with optic neuropathy and preoperative vision loss. Seven prophylactic decompressions were performed in subjects with normal vision preoperatively.

[†] Recurrence is reported here for clinically indicated decompressions presenting with preoperative vision loss. In prophylactic decompressions, five of seven subjects (71 percent) had postsurgical vision loss, as previously reported by Amin et al. (Amit M, Collins MT, FitzGibbon EJ, Butman JA, Fliss DM, Gil Z. Surgery versus watchful waiting in patients with craniofacial fibrous dysplasia: A meta-analysis. *PLoS One* 2011;6:e25179).