Fibrous Dysplasia: Management of the Optic Canal

Thomas S. Satterwhite, M.D.
Glenn Morrison, M.D.
John Ragheb, M.D.
Sanjib Bhatia, M.D.
Chad Perlyn, M.D.
S. Anthony Wolfe, M.D.

Miami, Fla.

Background: Fibrous dysplasia is an abnormal growth of bone that can lead to severe facial disfigurement. A dreaded outcome is compression of the optic nerve, leading to blindness. Controversy has surrounded the role of optic nerve unroofing for circumferential involvement of the optic canal. At present, many neurosurgeons unroof the nerve therapeutically in the setting of optic nerve dysfunction. Prophylactic unroofing (i.e., unroofing the nerve prior to the development of visual symptoms) has been previously proposed, although reported outcomes have been mixed. The authors present their long-term results of patients who have undergone optic nerve unroofing.

Methods: From 1975 to 2012, patients with fibrous dysplasia were investigated. Their age, demographics, operative procedure, optic nerve involvement (radiologically and clinically), and long-term outcomes and complications were recorded.

Results: Over 37 years, the senior author (S.A.W.) operated on 32 patients with fibrous dysplasia. Average follow-up was 5 years. Nine patients underwent optic nerve unroofing. Two patients had bilateral unroofing. Three patients who underwent therapeutic optic nerve unroofing ultimately went on to complete vision loss. The remaining seven patients who underwent prophylactic unroofing had no immediate postoperative visual compromise.

Conclusions: Therapeutic optic nerve unroofing is advocated in fibrous dysplasia patients with continuous deterioration of vision. However, the authors believe prophylactic unroofing is safe, and it should be performed not necessarily as a primary surgical procedure, but as a procedure along with excision of fibrous dysplasia in the anterior skull base during the same operation performed for orbitocranial deformity. (*Plast. Reconstr. Surg.* 135: 1016e, 2015.)

CLINICAL QUESTION/LEVEL OF EVIDENCE: Therapeutic, IV.

Fibrous dysplasia is a "benign" overgrowth of bone, but its effects can be disastrous if it is left untreated. Fibrous dysplasia can affect one bone (monostotic), which occurs in 70 percent of cases, or it can affect multiple bones (polyostotic), where the ribs and femur are the most involved noncraniofacial bones, and is associated with McCune-Albright disease, with manifestations of hyperendocrinopathy and café-au-lait spots.[1] In its mildest presentation, there can be aesthetic contour abnormalities, while its most severe presentation includes devastating facial deformities. One of the most dreaded complications of fibrous dysplasia is vision loss. Controversy surrounds the etiology of visual compromise in the fibrous dysplasia patient, clinical progression, treatment, and timing for surgical intervention. We investigated our series of patients with fibrous dysplasia with involvement of the optic canal. We retrospectively reviewed data involving patients who had undergone prophylactic or therapeutic unroofing to determine safety, efficacy, and long-term results. We also provide a review of the pertinent surgical literature.

The nature of this disease has been studied for over a century. Pathologically, it is described as an overgrowth of nondescript metaplastic fibrous material. This is caused by a mutation in the G-signaling protein in osteoblastic cells, causing a disorganized deposition of bony matrix.[1,2]

Fibrous dysplasia typically starts early in life. In the majority of patients, it presents within the first

From the Divisions of Plastic Surgery and Neurosurgery, Miami Children's Hospital.
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three decades. Its course of progression is variable. The initial presenting sign is swelling or deformity of the affected area. Functional deficits can ensue if fibrous dysplasia involves sensitive areas, leading to conductive hearing loss, nasal obstruction, oral incompetence, loss of teeth, and orbital involvement. One of the most devastating complications of orbital involvement is vision loss.

Diagnosis is typically made on clinical examination, with confirmation made radiographically. On computed tomography scans there is a ground glass appearance with a mix of fibrous and osseous components.[3] It can be classified as sclerotic, cystic, or mixed. The sclerotic type is most prevalent in thick cranial bones.

Management varies from patient to patient, depending on anatomic involvement and disease progression. With minimal to mild involvement, serial examinations can be an appropriate and safe treatment plan. Attempts at medical management are quite limited. There are reports of using medications intended to limit growth, such as bisphosphonate, calcitonin, and etidronate, but success has not been shown to validate their regular use.[4,5] Systemic steroids are important in the temporary management of acute visual loss from optic nerve involvement.[6]

Surgical intervention is the primary treatment modality for fibrous dysplasia.[7–10] Surgery can involve burring and contouring of involved bone to aggressive resection with autologous bone reconstruction.[11,12] With extensive resections, some authors advocate autoclaving, recontouring, and reimplanting the involved bone,[13,14] although we do not advocate this technique because the viability of the bone is unpredictable with the potential of being reabsorbed. Some think that early radical intervention can be prophylactic in preventing further growth,[15–18] whereas others think that radical resection should be reserved until the fibrous dysplasia has ceased to progress, particularly in patients with polyostotic disease.[11]

Close clinical assessment of the optic nerve is important in patients with involvement of the orbit. An ophthalmologist should be closely involved in the care of the patient. Red color desaturation and visual fields should be serially monitored as they are the first to be affected by optic nerve involvement.[19] With progressive deterioration, visual acuity then becomes affected. More complex examinations include visual-evoked potentials, which measure the functional integrity of the visual pathway, and optical coherence tomography, which captures micrometer-resolution three-dimensional images of the optic nerve.[17] Regardless, the most reliable means of observing patients is with close clinical examination by a neuro-opthalmologist.

The etiology of vision loss is thought to be due to progressive compression of the optic nerve from canal stenosis, resulting in chronic ischemic optic neuropathy.[18,20,21] However, the etiology can be multifactorial. Other causes can include ischemia (with venous and arterial compromise) and nerve traction. Causes of more acute visual changes can be attributed to cystic lesions, mucoceles, or hemorrhage.[22–24]

Therapeutic unroofing is indicated in patients with visual compromise.[20,25,26] Acute visual changes warrant urgent intervention.[23,27] Gradual visual loss is also an indication for surgical unroofing,[20,26] although the timing of intervention is not standardized. Prophylactic optic nerve unroofing is supported by some authors,[15,17,18] while others think that unroofing should only be used when visual symptoms have developed.[28–31]

Prophylactic unroofing (before symptoms have developed) is advocated in some reports.[16,18,32] However, there is concern that prophylactic interventions can lead to unnecessary injury to the optic nerve due to direct trauma, injury from burring, thermal damage, traction, loss of blood supply, and vascular thrombosis.[33] Iatrogenic injury leading to blindness is the worst-case scenario.[28]

On the other hand, there can be morbidity in waiting until symptoms have developed. There is the concern that once visual symptoms have developed, much of the damage to the optic nerve, which is part of the central nervous system, has become irreversible. Animal studies have shown that optic nerve dysfunction can be restored when only demyelination has occurred, but more involved axonopathy can produce an irreversible state.[34] Waiting for symptoms to develop is especially dangerous in patients with poor compliance and who may have inadequate follow-up. In such cases, we may be following up these patients to a most unfortunate fate: blindness.

PATIENTS AND METHODS

From 1975 to 2013, all patients with fibrous dysplasia were investigated. Their age at operation, demographics, operative procedure, optic nerve involvement (radiologically and clinically), and long-term outcomes and complications were recorded in a retrospective fashion. Patients had been referred by their primary physician or pediatrician. Preoperative computed tomography

was performed to assess their degree of disease. For those with radiologic or clinical optic nerve involvement, ophthalmology and neurosurgery services were co-consulted.

Operative technique varied based on degree of involvement. In general, a coronal approach was required to fully access the involved bone. Aggressive efforts were made to remove as much of the dysplastic bone as possible, and in some cases, complete removal was possible. Bony defects were reconstructed with autogenous bone grafts—split calvarial, rib, or iliac crest. Optic nerve unroofing was performed by the neurosurgical team.

RESULTS

Over the course of 37 years, the senior author (S.A.W.) treated 32 patients with fibrous dysplasia. Nine of these patients underwent optic nerve unroofing. Patient data are presented in Table 1. The average age was 21 years (range, 7 to 45 years), and the average follow-up was 5 years (range, 1 to 10 years). All patients had radiological evidence of optic canal involvement.

Among the nine patients who underwent optic nerve unroofing, three patients underwent therapeutic unroofing due to progressive visual compromise. Two patients with polyostotic disease underwent bilateral therapeutic unroofing. Seven patients underwent prophylactic unroofing. One patient underwent prophylactic twice, followed by therapeutic unroofing. There was no visual compromise after the prophylactic unroofing. Postoperative blindness developed in the three patients who underwent therapeutic unroofing: one who was having a precipitous drop in visual acuity preoperatively went on to bilateral blindness despite therapeutic unroofing; in the other two, the visual loss was gradual.

When broken down by optic canal (Table 2), therapeutic unroofing was performed five times. Among those five cases, immediate postoperative improvement was seen in two cases. Immediate postoperative vision loss was seen with two optic canals. Long-term vision loss was seen in two optic canals. Prophylactic optic nerve unroofing was performed on seven optic canals. These were cases in which vision was normal preoperatively. Six optic canals maintained normal vision. One optic canal developed long-term vision loss (in a patient who underwent prophylactic followed by therapeutic unroofing). None of the optic canals that underwent prophylactic unroofing developed immediate postoperative visual compromise.

CASE REPORTS

Case 1

An 18-year-old man presented with left frontal bossing, hypoglobus, and proptosis (Fig. 1, *above, left*). Fibrous dysplasia involved his left frontal bone, sphenoid, and orbit (Fig. 1, *above, center*). The optic canal was involved circumferentially, but he had no visual deficits. Neurosurgery and ophthalmology services were consulted preoperatively. The patient underwent a bifrontal craniotomy to remove the involved bone. Efforts were made to resect all involved areas in the frontal bone, sphenoid, and orbit. The frontal bone was noted to be more than 4 cm thick (Fig. 1, *below, left*). The optic canal was identified, and the nerve was unroofed with the assistance of the neurosurgery team (Fig. 1, *below, center*). Split cranial bone grafts were used to reconstruct the frontal bone as well as the orbital roof (Fig. 1, *below, right*). Pathologic analysis confirmed the diagnosis of fibrous dysplasia. The patient had no postoperative complications. The patient subsequently returned to the operating room a year later for removal of hardware and recontouring of the supraorbital ridge and orbital roof. Seven years later, he was doing well with no visual compromise, and was a practicing orthopedic surgeon (Fig. 1, *above, right*).

Case 2

A 17-year-old boy with McCune-Albright syndrome was noted to have fibrous dysplasia since the age of 3 (Fig. 2, *above, left*). He was being closely observed by the ophthalmology service because of bilateral involvement of the optic canals (Fig. 2, *above, center*). Over the course of 6 months, progressive visual impairment developed in his right eye. He underwent resection of fibrous dysplasia from the right orbit, left mandible, and therapeutic optic nerve unroofing. This helped his visual symptoms. Two years later, progressive visual impairment developed in his left eye. He underwent resection of involved bone in the orbit, maxilla, and mandible, as well as left therapeutic optic nerve unroofing. Diseased bone was more than 3 cm thick (Fig. 2, *below, left*). The maxilla and orbit were reconstructed with split calvarial and iliac crest bone grafts. Optic nerve unroofing was performed by the neurosurgery team, using a microscope and intraoperative navigation system. The superior orbital fissure was identified, as well as the bony strut separating the orbital fissure and optic canal. The nerve was skeletonized superiorly, laterally, and inferiorly (Fig. 2, *below, right*). Microcurrettes were used to carefully remove tissue from the optic nerve dura without violating it. The patient was lost to follow-up in our clinic and ophthalmology service. Two years later, he had complete vision loss of the left eye (Fig. 2, *above, right*).

DISCUSSION

In our series, we found that prophylactic unroofing resulted in no acute optic nerve injury. Visual compromise or any other major complications did not develop in the immediate postoperative period. Therapeutic unroofing, however, resulted in more immediate and long-term complications. In our hands, prophylactic unroofing proved to be safe.

Controversy surrounds many aspects of fibrous dysplasia. Regarding visual compromise, controversy and disagreement surround the etiology,

Table 1. Patient Data

Patient	Initials	Age at OR (yrs)	Eye Involved	Optic Canal Inolvement Radiologically?	Optic Nerve Symptomatic?	Vision Preoperatively	Type of ONU	Vision Postoperatively	Follow-Up (yrs)	Involvement/Procedure
1	M.A.	45	Right	Y	N	Normal	Prophylactic	Unchanged	1	Right orbit, frontal bone/split calvarial bone graft
2	D.B.	8	Left	Y	N	Normal	Prophylactic	Unchanged	10	Left skull base, orbit, frontoparietal bone/split calvarial bone graft
3	S.B.	17	Right	Y	Y	Abnormal	Therapeutic	Improved	4	Polyostotic disease; extensive involvement of maxilla, mandible, frontal bones, both orbits/split calvarial bone graft
			Left	Y	Y	Abnormal	Therapeutic	Worsened to blindness		
4	F.B.	18	Left	Y	N	Normal	Prophylactic	Unchanged	7	Left frontal bone, orbit/split cranial bone graft
5	G.C.	8	Right	Y	N (initially 1982)/Y (later with recurrence 1990)	Normal (prior to prophylactic); abnormal (prior to therapeutic)	Prophylactic, followed by therapeutic	Unchanged after prophylactic; progressed to blindness after therapeutic	9	Right frontal bone, orbit, maxilla/iliac bone and split rib graft
6	T.F.	7	Right	Y	N	Normal	Prophylactic	Unchanged	1	Right frontal bone, orbit/split calvarial bone graft
7	S.G.	32	Right	Y	N	Normal	Prophylactic	Unchanged	7	Right maxilla, orbit/split calvarial bone graft
8	C.I.	23	Right	Y	N	Normal	Prophylactic	Unchanged	3	Right frontal bone, orbit/rib and iliac bone grafts
9	T.K.	29	Right	Y	Y	Abnormal	Therapeutic	Worsened to blindness	1	Polyostotic disease; bilateral orbits and frontal bone
			Left	Y	Y	Abnormal	Therapeutic	Worsened to blindness		

N, no; Y, yes; OR, operation; ONU, optic nerve unroofing.

Table 2. Results per Optic Canal

Type of Decompression	Total No. of Procedures	Immediate Postoperative Improvement	Unchanged	Immediate Postoperative Vision Compromise	Long-Term Vision Compromise
Therapeutic (impaired vision)	5	2	0	2	2
Prophylactic (normal vision)	7	N/A	6	0	1

N/A, not applicable.

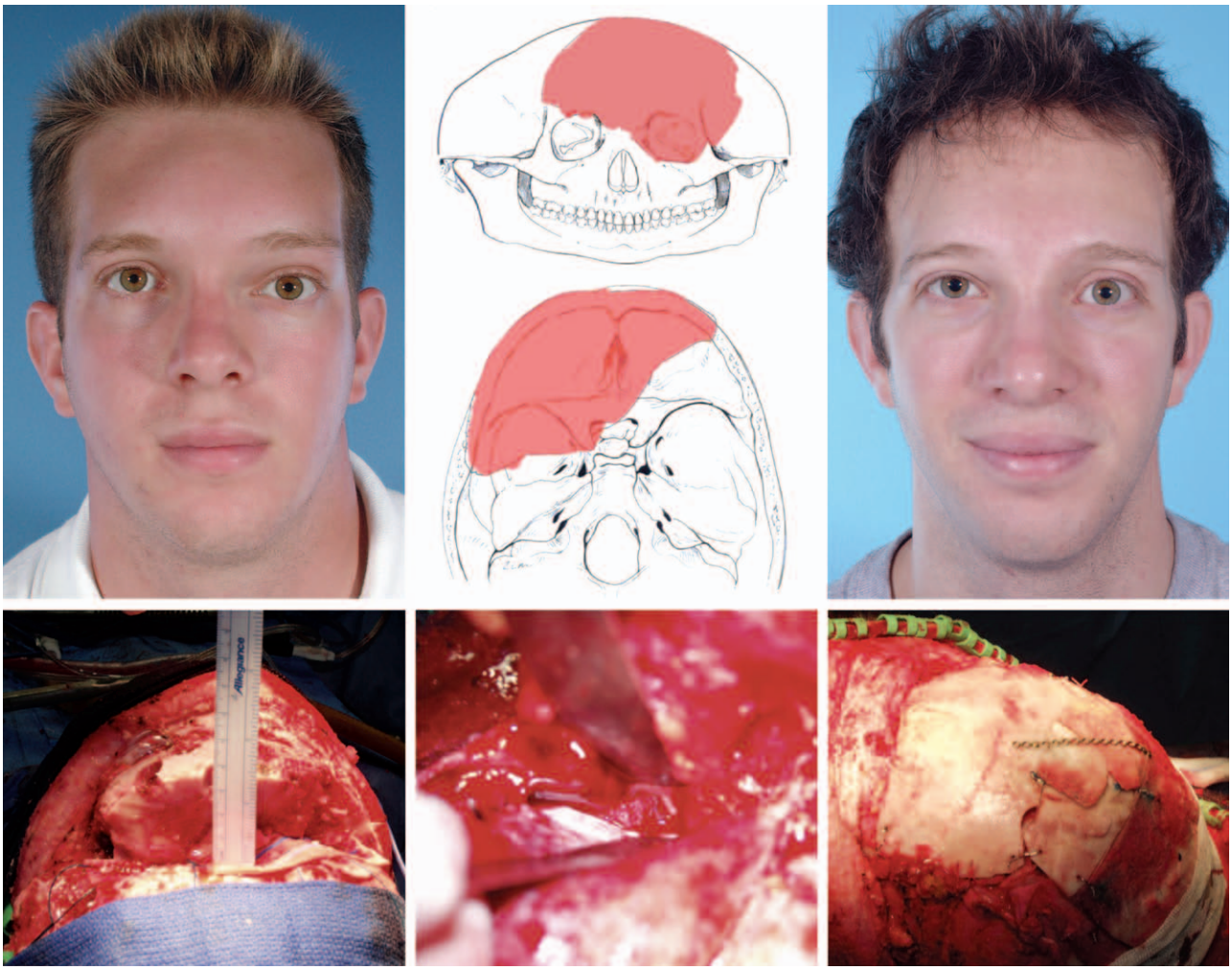

Fig. 1. Patient presented with bossing and hypoglobus (*above, left*), with involvement of left frontal bone and orbit (*above, center*). Thick, diseased bone was resected (*below, left*). The orbital nerve was unroofed prophylactically (*below, center*). The resultant defect was reconstructed with a split calvarial bone graft (*below, right*). Patient is shown 7 years postoperatively with normal vision (*above, right*).

the progression of disease, and the timing of intervention.

Regarding etiology, it has been assumed that fibrous dysplasia is a progressive disease. It typically begins in childhood, and "burns out" by adulthood.[11] With involvement of the optic canal, there is concern that progressive compression of the optic nerve will lead to blindness. Therefore, some authors believe that early unroofing will prevent such disastrous long-term consequences.[27] Several authors have reported favorable results with prophylactic unroofing, with the ability to preserve vision long term.[18,32] However, without side-by-side randomized studies, it is quite difficult to truly ascertain the effectiveness. Moreover, the heterogeneity of this patient population and presentation of disease makes comparisons between prophylactic and therapeutic treatments rather difficult.

Prophylactic unroofing, however, is not without consequences. The most disastrous result it

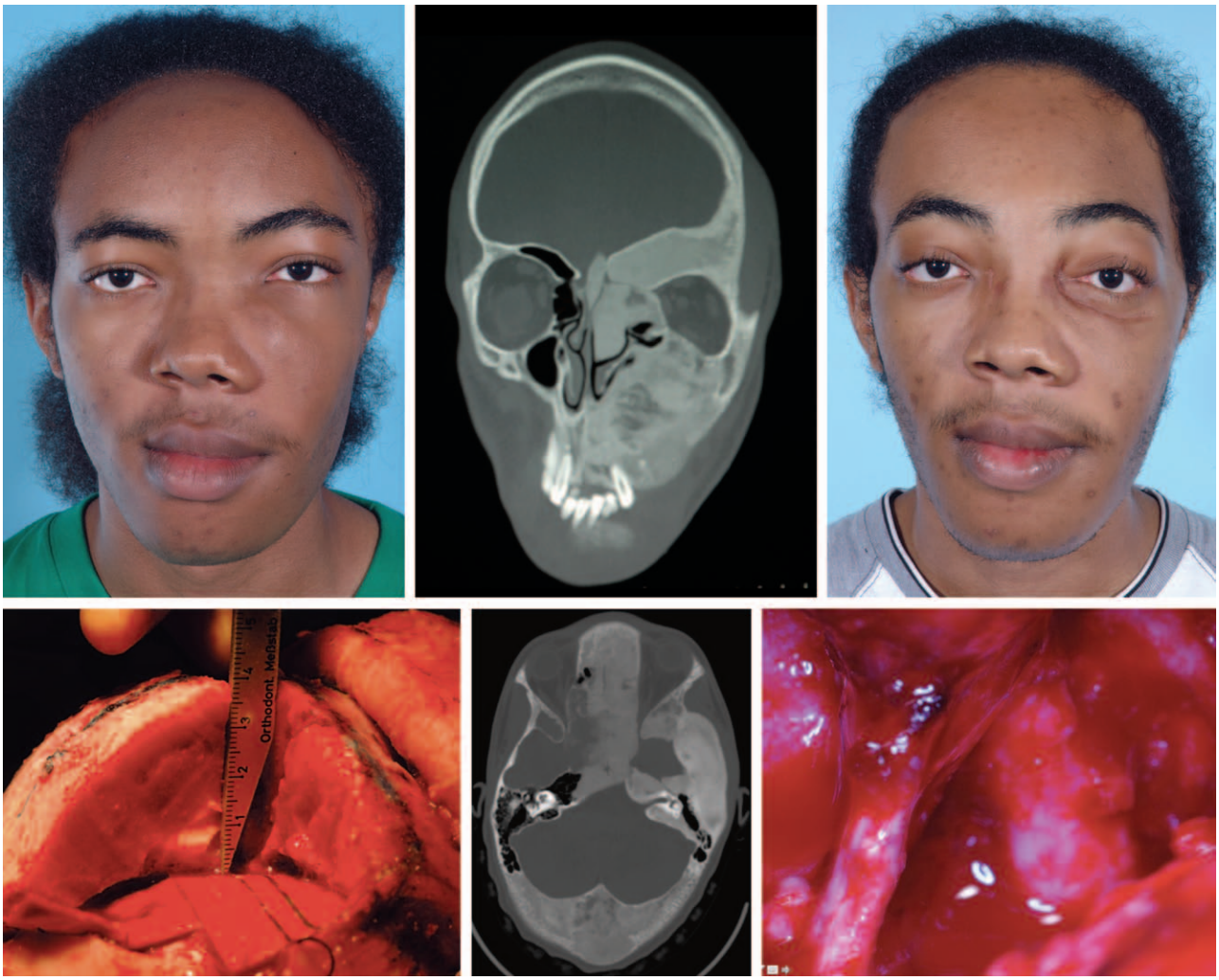

Fig. 2. A 17-year-old boy with McCune-Albright syndrome (*above, left*) with severe polyostotic disease noted on computed tomography scan (*above, center*). Involved frontal bone was noted to be more than 3 cm thick (*below, left*). The patient underwent multiple resections and reconstruction with autogenous bone graft. Initially, he underwent therapeutic optic nerve unroofing on the right, improving his visual compromise (*below, right*). Subsequently, he underwent therapeutic optic nerve unroofing on the left. His vision on the left improved, but he was lost to follow-up, and ultimately developed complete vision loss on the left (*above, right*).

blindness from direct injury to the nerve during the decompressive process. There have been several case reports of this unfortunate result.[27,28]

A more fundamental concern arises: Is visual loss in fibrous dysplasia truly due to progressive compression of the optic nerve? Moore and colleagues had shown that vision loss developed in only two out of five patients with optic canal narrowing.[35] In a more extensive review, Dr. Lee and colleagues at the National Institute of Dental and Craniofacial Research had shown through radiographic analysis in a cross-section of patients that there was no correlation between optic canal diameter and the development of visual compromise and blindness.[30] Of note, the majority of their patients had polyostotic disease or McCune-Albright syndrome. Similarly, in our series we found that the two patients with McCune-Albright syndrome ultimately had the worst outcomes—blindness developed in both patients, even after therapeutic unroofing. Other authors have also reported that visual loss was not caused by nerve compression, but was due to other causes, such as cystic lesions, mucoceles, hemorrhage, and bone cysts.[22]

If progressive compression of the optic nerve is not the underlying etiology for visual deterioration, then one could say that prophylactic unroofing is unnecessary, and, in fact, harmful. However, we believe that optic canal involvement is a predictor for impending optic nerve damage. In their follow-up study, Dr. Lee and colleagues showed

that visual compromise developed in 12 percent of their patients with optic canal involvement.[29] This suggests that long-term involvement of the optic canal can lead to blindness. They went on to report that growth hormone excess is a highly associated risk factor for visual compromise. Dr. Lee stated that with growth hormone excess in patients, "…the fibrous dysplasia growth is massive and puts them at significant risk for complete encasement and optic neuropathies, including blindness."[36] Given these data, it seems that if the optic canal is involved, then there is indeed the real potential to develop blindness. Other authors have further suggested this relationship: they have reported favorable results in potentially preventing the progression of visual compromise, and being able to do it safely.[32,37]

Given this web of controversy, the ultimate question arises: What is the safest and most predictable means of preserving vision in a patient with fibrous dysplasia involving the optic canal? For some of us, it is with prophylactic unroofing. Other authors have reported blindness with prophylactic unroofing,[28] but in our hands, this never happened. Admittedly, even among the authors of this article, there are those who support prophylactic unroofing (T.S., G.M., C.P., and S.A.W.), whereas others believe that unroofing should only be performed when symptoms have arisen (J.R. and S.B.). The controversy regarding the management of this disease is so deep that even our own group is in disagreement. Seven patients in our series underwent prophylactic unroofing, and none of them had any major postoperative complications. One patient (patient 5, G.C.) who initially underwent prophylactic unroofing without any complications subsequently underwent therapeutic unroofing years later for visual changes. This improved her vision, but she subsequently was lost to follow-up (due to a variety of social reasons) and finally returned to us years later with blindness. In our series, therapeutic unroofing was performed in three patients. Two patients had bilateral involvement. Outcomes in these patients were less than satisfactory. One patient (patient 9, T.K.) with polyostotic involvement (McCune-Albright) underwent emergent bilateral optic nerve unroofing for rapidly deteriorating visual acuity and blindness developed in both eyes in the immediate postoperative period. It was difficult to determine whether the operation itself caused the visual loss, or whether the involvement was so severe that the operation was unable to prevent the blindness. Another patient (patient 3, S.B.) who also had McCune-Albright syndrome underwent bilateral decompression. Vision improved initially in both eyes, but after being lost to follow-up, he returned to the clinic with vision loss in one eye. The etiology and the time course of the vision loss in our two patients who underwent therapeutic unroofing and who were lost to follow-up remain unknown. Long-term clinical and radiographic studies in patients with severe disease could potentially track progression and determine the efficacy of using interval computed tomography scans to monitor for potential dangerous recurrences.

Reviewing our data, patients who underwent prophylactic unroofing did much better long term than those who underwent therapeutic unroofing. Of course, given the small size of our study population, there are limitations to any definitive conclusions we can draw. It is difficult to ascertain whether the patients who had prophylactic unroofing would have progressed to visual compromise if we had left them alone. In addition, it is difficult to determine whether our three patients who underwent therapeutic unroofing would have benefited from prophylactic unroofing, or was their involvement so severe that they were destined to vision loss regardless of the timing of their intervention? Other authors have also shown that therapeutic unroofing is not always effective and can be more dangerous and complicated.[37] Unroofing once visual deterioration has developed may already be too late. Notably, our most severe cases were those with McCune-Albright disease, which other authors have similarly noted.[25] In these patients, it is difficult to determine whether therapeutic unroofing contributed to their visual compromise. Admittedly, any conclusions we can draw with therapeutic decompression can only be conjecture. In addition, an extensive review of the literature reveals only a small number of case reports, so strong conclusions and recommendations cannot be made. Only with a randomized, prospective trial can we make clear conclusions, but it is quite difficult given the rarity of this disease and its heterogenous and variable presentation.

Despite these shortcomings, we can, with a reasonable degree of confidence, say that prophylactic optic nerve unroofing is safe in our hands. No patient with prophylactic unroofing had any complications or visual compromise in the immediate postoperative period. We agree with Dr. Chen and colleagues: prophylactic unroofing should not be performed as a primary surgical procedure but secondary to an already planned anterior skull base resection.[16,32] We have been able to obtain

safe and predictable outcomes because we work with experienced and well-trained neurosurgeons. We also advocate treating one eye at a time. Prophylactic unroofing is beneficial in our particular patient population. Many of our patients travel internationally or come from difficult social situations that prevent regular follows-ups. Especially in noncompliant patients, prophylactic unroofing is highly recommended—in our patient population, the benefits of prophylactic unroofing outweigh the risks associated with potentially poor clinical follow-up. In our series, the two patients in whom long-term vision loss developed were noncompliant and lost to follow-up, returning to clinic only after complete vision loss occurred.

CONCLUSIONS

The management of the optic nerve in fibrous dysplasia patients who have optic canal involvement is quite controversial. Uncertainties surround the etiology of visual loss and the timing of treatment. Given the rarity of this disease, statistically significant data are almost impossible to obtain. Reviewing our data, we think patients who have involvement of the optic canal should be followed closely by a neuro-opthalmologist. Any visual deterioration should be treated with therapeutic optic nerve unroofing. If a patient with optic canal involvement is being brought to the operating room for anterior skull base or orbital resection, then we recommend concomitant prophylactic unroofing. The majority of patients who underwent prophylactic unroofing appeared to preserve vision and had no immediate postoperative complications. Prophylactic unroofing should only be performed by a highly experienced neurosurgeon. Prophylactic unroofing is particularly beneficial for noncompliant patients and individuals who are at risk of being lost to regular clinical follow-up.

S. Anthony Wolfe, M.D.
Division of Plastic Surgery
Miami Children's Hospital
3100 S.W. 62nd Avenue, Suite 2230
Miami, Fla. 33155
drwolfe@santhonywolfemd.com

PATIENT CONSENT

Patients or parents or guardians provided written consent for use of the patients' images.

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