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Plast Reconstr Surg. Author manuscript; available in PMC 2017 June 01.

Published in final edited form as:

Plast Reconstr Surg. 2016 June ; 137(6): 1060e–1061e. doi:10.1097/PRS.0000000000002210.

Response to "Fibrous Dysplasia: Management of the Optic Canal":

Submission to: Plastic and Reconstructive Surgery

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In Dr. Satterwhite and colleagues' article, "Fibrous Dysplasia: Management of the Optic Canal" (1), the authors reported their experience with prophylactic optic nerve decompression (OND). Long-term visual compromise developed in 2/5 fibrous dysplasia (FD) patients undergoing therapeutic OND and 1/7 patients undergoing prophylactic OND, with 3/12 (25%) having adverse outcomes. They concluded that FD patients who underwent prophylactic OND had better outcomes, and that optic nerve unroofing is "safe in our [*sic*] hands."

We disagree with the authors' belief that, "optic canal involvement is a predictor for impending optic nerve damage." Our National Institutes of Health (NIH) cohort (2) reported 13 patients with optic neuropathy – of which 10 had growth hormone excess (GHE) (7/13, 54%) or associated aneurysmal bone cyst (ABC) (3/13, 23%). We know GHE in FD is associated with macrocephaly and vision loss, with some patients developing blindness. Recent analysis of the NIH cohort indicates that early diagnosis and treatment may prevent GHE-associated morbidity, specifically vision loss (3). Furthermore, OND in symptomatic patients has been shown to have high peri-operative morbidity. We advocate for close observation (frequent ophthalmologic exams with OCT and evoked potentials, medical treatment of GHE when present) as the preferred approach to patients with FD-encasement of the optic canal.

Thus, we feel that confounding factors such as untreated GHE and ABCs, *not long-term involvement of the optic canal*, are predictors of visual compromise in the McCune-Albright population. This case series did not state whether patients had GHE or associated ABC. We stress the importance of a complete workup in FD patients prior to surgery, since GHE can be treated non-operatively, reducing the risk of optic neuropathy.

Amit et al. performed a meta-analysis (4) on patients from the NIH: a total of 368 nerves were included in the meta-analysis; long-term follow-up revealed that surgery in

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Disclosures: None of the authors has a financial interest in any of the products, devices, or drugs mentioned in this manuscript.

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asymptomatic patients is associated with worse prognosis, as compared to patients managed expectantly. Moreover, GHE was not found to have an association with optic neuropathy because young subjects with reported excess underwent early treatment. Visual loss was only seen in subjects whose GHE diagnosis and treatment were delayed until adulthood, as none of the subjects treated in childhood had visual disturbances (5). There is no evidence that FD progression can be predicted, and it is not possible to foresee which patients would benefit from OND surgery.

We agree that a randomized, prospective trial would be informative, though difficult. Although the authors report a lower risk in prophylactic decompression at their center, they admit that, “Other authors have reported blindness with prophylactic unroofing, but in our hands, this never happened.” In light of the meta-analysis performed, it is unreasonable to create a standard of care based on the abilities of a few surgeons who have had rare complications. Further, an outcome of 3/12 (25%) patients developing visual compromise is still relatively high. We strongly recommend against prophylactic OND based upon the aforementioned meta-analysis data and our experience from the natural history protocol ongoing at the NIH.

Acknowledgments

This research was supported by the Intramural Research Program of the NIDCR, NIH.

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