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Visual Outcomes in Children With Neurofibromatosis Type 1 and Orbitotemporal Plexiform Neurofibromas

ROBERT A. AVERY, EVA DOMBI, KELLY A. HUTCHESON, MARIA T. ACOSTA, ANDREA M. BALDWIN, WILLIAM P. MADIGAN, ANDREA GILLESPIE, EDMOND J. FITZGIBBON, ROGER J. PACKER, BRIGITTE C. WIDEMANN

Departments of Neurology (R.A.A., M.T.A., R.J.P.), Pediatrics (R.A.A., M.T.A., R.J.P.), and Ophthalmology (K.A.H., W.P.M.); Gilbert Family Neurofibromatosis Institute (R.A.A., M.T.A., R.J.P.); and Center for Neuroscience and Behavior (R.A.A., M.T.A., R.J.P.), Children's National Medical Center, Washington, DC; Pediatric Oncology Branch, National Cancer Institute, Bethesda, Maryland (E.D., A.M.B., A.G., B.C.W.); and National Eye Institute, Bethesda, Maryland (E.J.F.)

Abstract

- **PURPOSE:** To describe the visual outcomes and volumetric magnetic resonance imaging (3D MRI) in children with neurofibromatosis type 1 (NF1) and orbitotemporal plexiform neurofibromas.
- **DESIGN:** Multicenter retrospective case series.
- **METHODS:** Two institutions with dedicated NF1 clinical research programs queried their established clinical databases for children with orbitotemporal plexiform neurofibromas. Visual acuity, refractive error, amblyopia, and treatment history were abstracted. Extent of orbitotemporal plexiform neurofibroma involvement was assessed clinically and with 3D MRI analysis. Children with optic pathway gliomas or ocular causes of decreased visual acuity (ie, cataracts, glaucoma) other than strabismus or anisometropia were excluded.
- **RESULTS:** Twenty-one children met inclusion criteria (median age 8 years, range 0.33–23 years). Orbitotemporal plexiform neurofibroma location was classified as isolated eyelid ($n = 6$), eyelid and orbit ($n = 7$), orbit and temporal region ($n = 7$), or diffuse orbit ($n = 1$). Three subjects had bilateral orbital involvement. Amblyopia secondary to the orbitotemporal plexiform neurofibroma was present in 13 subjects (62%) and was caused by strabismus ($n = 2$, 10%), occlusion from ptosis ($n = 9$, 43%), or anisometropia ($n = 9$, 43%), or a combination of factors ($n = 6$, 29%). MRI-derived volumes were measured in 19 subjects (median 41.8 mL, range 2.7–754 mL). All subjects with amblyopia had orbitotemporal plexiform neurofibroma volumes greater than 10 mL.
- **CONCLUSION:** In our series, amblyopia occurs in more than half of NF1 children with orbitotemporal plexiform neurofibromas, most commonly because of ptosis and anisometropia.

The 3D MRI analysis allowed for sensitive measurement of orbitotemporal plexiform neurofibroma size, and larger volumes were associated with development of amblyopia.

NEUROFIBROMATOSIS TYPE 1 (NF1) IS A MULTI-system genetic disorder occurring in approximately 1:3000 births.^{1,2} Although optic pathway gliomas are the most common ophthalmologic condition in children with NF1, plexiform neurofibromas that infiltrate the orbit, temporal region, and/or eyelids, termed orbitotemporal neurofibromas, are less common but potentially vision threatening. Most orbitotemporal neurofibromas are believed to be congenital, grow during early childhood, and have the potential to become malignant in a small number of cases.² Orbitotemporal neurofibromas can cause strabismus and proptosis, alter globe length, and obscure the visual axis secondary to infiltration and edema of the orbit and eyelids—putting young children at risk for amblyopia.³

Most research on orbitotemporal neurofibromas has focused on the surgical treatment, primarily in adult cohorts, rather than visual outcomes in young children at risk for amblyopia.^{3–6} Long-term success of surgical resection and ptosis repair is reduced by the potential for recurrence of orbitotemporal neurofibromas, especially in young children. Recurrence of incompletely resected orbitotemporal neurofibromas is now better understood as it is likely a result of the inflammatory environment created by surgery, resulting in increased recruitment of mast cells.⁷

We describe the visual outcomes associated with orbitotemporal neurofibromas and propose initial guidelines for the evaluation and management of orbitotemporal neurofibromas in children.

METHODS

• PATIENTS:

The NF1 clinical research programs at Children’s National Medical Center (Washington, DC) and the Pediatric Oncology Branch of the National Cancer Institute (Bethesda, Maryland) performed a retrospective case study by querying their established NF1 clinical databases for children who met the following criteria: confirmed orbitotemporal neurofibromas by neuroimaging (magnetic resonance imaging [MRI]/computed tomography) and ophthalmologic records. Children with optic pathway gliomas or ocular causes of decreased visual acuity (ie, cataracts, glaucoma) other than strabismus or anisometropia were excluded. Those subjects meeting inclusion criteria had the following data abstracted from their first visit’s clinical chart onto a standardized data form: visual acuity (VA), refractive error, presence of amblyopia, and treatment (medical or surgical) history for their orbitotemporal neurofibromas. Amblyopia was defined as a greater than or equal to a 0.2 logarithm of the minimal angle of resolution (logMAR) difference in the age-based VA and attributed to strabismus, occlusion from ptosis, and/or anisometropia, defined as greater than 1.5 diopters (D) myopia, 2.5 D hyperopia, or 1.5 D astigmatism.⁸ Amblyopia treatment was defined as any intervention aimed at improving VA or alignment such as patching, atropine drops, or surgical repair of strabismus. Orbitotemporal neurofibroma location was classified as isolated eyelid, eyelid and orbit, orbit and temporal region, or

diffuse orbit (Figure 1). Treatment indications included amblyopia, cosmetic appearance, exposure keratopathy, or a combination of factors.

The Institutional Review Boards from both institutions approved the clinical databases used in the study. All subjects provided written consent to allow their protected health information to be included in the database. No identifying information was exchanged between institutions.

• VOLUMETRIC MAGNETIC RESONANCE IMAGING ANALYSIS:

Extent of orbitotemporal neurofibromas was assessed clinically and with MRI, including volumetric MRI (3D MRI) analysis, if feasible; 3D MRI analysis was performed as previously described using MEDx (Medical Numerics Inc, Germantown, Maryland).^{9,10} For each MRI image, the tumor is roughly outlined manually, including a rim of low-signal-intensity normal tissue, followed by automated histogram analysis of signal intensity pixel by pixel, definition of threshold that distinguishes high-signal-intensity tumor from normal tissue, and determination of tumor contours (Figure 2).

• STATISTICAL ANALYSIS:

Demographic and clinical characteristics were summarized by descriptive summaries (eg, means and standard deviations for continuous variables such as age and percentages for categorical variables such as sex). Wilcoxon rank sum was used to compare variables between eyes affected by orbitotemporal neurofibromas and unaffected eyes. Spearman rank correlation determined the relationship between orbitotemporal neurofibroma volumes and magnitude of amblyopia (logMAR).

RESULTS

TWENTY-EIGHT CHILDREN WITH NF1 AND ORBITOTEMPORal neurofibromas were identified (median age 8 years at first evaluation, range 0.33-23 years). Seven subjects were excluded for coexistent optic pathway glioma (n = 6) or glaucoma (n = 1), resulting in 21 evaluable subjects. Ten of 21 subjects (48%) were female; 2 subjects were African American, 1 subject was of multiple races, and 18 were white. Discovery of orbitotemporal neurofibromas occurred between birth and 5 years of age, with most being diagnosed within the first 2 years of life. Thirteen subjects experienced amblyopia (mean = 0.5 logMAR difference, range 0.2-1.5 logMAR). Fifteen subjects received corrective eyewear, of which 14 of 15 (93%) were for myopia. The amount of spherical refractive error was greater for the orbitotemporal neurofibroma eyes (median -2.63 D, range -10.00 to +1.50) compared with the unaffected eye (median = -0.125 D, range -7.25 to +1.50; $P = .025$). The amount of astigmatism affected by orbitotemporal neurofibromas (median = 0.875 D, range 0.00 to 6.00) was slightly greater but not statistically different than the unaffected eyes (median = 0.25 D, range 0.00 to 1.25; $P = .063$).

Table 1 lists the location of orbitotemporal neurofibromas, etiology of amblyopia, type, and indication for treatment. The locations of orbitotemporal neurofibromas were fairly well distributed, although diffuse orbital infiltration occurred in 1 of 21 cases (5%). The etiology of amblyopia occurred most commonly from ptosis (9/21; 43%) or anisometropia (9/21;

43%), although a combination of multiple factors (6/21; 29%) frequently contributed. Treatment of orbitotemporal neurofibromas most commonly occurred because of amblyopia (7/21; 33%), followed by cosmetic appearance (4/21; 19%).

MRI-derived orbitotemporal neurofibroma volumes were measured in 19 subjects (median 39.9 mL, range 2.7-754 mL), with 1 subject contributing volumes from bilateral orbitotemporal neurofibromas (Table 2). Subjects with amblyopia had a greater orbitotemporal neurofibroma volume (median 41.8 mL, range 12.8-754.0) compared with those without vision loss (median 9 mL, range 2.7-66.7), although this did not reach statistical significance ($P = .074$). All 13 subjects with amblyopia had orbitotemporal neurofibroma volumes greater than 10 mL, compared with 3 of 7 subjects without amblyopia (Figure 3). Despite the differences in groups, the correlation between orbitotemporal neurofibroma volume and amount of vision loss (logMAR difference from normal) did not reach statistical significance ($r = 0.259$, $P > .2$).

DISCUSSION

THIS STUDY DESCRIBES THE VISUAL OUTCOMES, SPECIFICALLY visual acuity and refractive error, of orbitotemporal neurofibromas secondary to NF1. More than half of the children with orbitotemporal neurofibromas experienced amblyopia, most commonly attributable to anisometropia or occlusion of the visual axis by ptosis, although multiple factors may have contributed. Amblyopia from orbitotemporal neurofibromas was typically mild, but on occasion some subjects experienced severe vision loss. Although all subjects with amblyopia had orbitotemporal neurofibromas larger than 10 mL, some subjects without amblyopia had orbitotemporal neurofibroma volumes similar to those with amblyopia (Figure 3). Our cross-sectional analysis precluded our ability to determine if those without amblyopia and orbitotemporal neurofibroma volumes >10 mL would ultimately experience vision loss. Because of technical limitations, we compared the entire orbitotemporal neurofibroma volume, rather than just the eyelid volume, to the rate of amblyopia. It is conceivable that the orbitotemporal neurofibroma volume in a particular location (ie, eyelid) may contribute more to amblyopia. Although MRI has been used to measure plexiform neurofibromas elsewhere in the body,^{9,11,12} to our knowledge this is the first study directly examining the relationship between visual outcomes and orbitotemporal neurofibroma volumes. 3D MRI analysis is now considered to be a relevant outcome measure for clinical trials in children with plexiform neurofibromas.^{9,11,12}

Nearly 62% of subjects in our study experienced amblyopia, similar to the frequency reported in 2 other studies.^{6,13} However, vision loss was found to be much more severe in those studies^{6,13} compared to our study, possibly because of their inclusion criteria permitting coexisting optic pathway gliomas, optic neuropathy, and glaucoma. Our study protocol was developed and data collected without knowledge of the study by Oystreck and associates.¹³ To avoid attributing vision loss to etiologies other than orbitotemporal neurofibromas, we made the a priori decision to exclude coexisting optic pathway gliomas and glaucoma subjects from our study. This exclusion criterion, along with our younger age of subjects, analysis of baseline visual function, and much lower incidence of glaucoma compared to other studies,^{3,13} may have contributed to the differences in the severity of

vision loss. Unfortunately, none of the previous studies described the incidence of pupillary defects. Only 1 of our subjects had an infiltrating orbitotemporal neurofibromas that could have potentially caused a compressive optic neuropathy. A post hoc review of our subjects failed to uncover signs of optic neuropathy (eg, relative afferent pupillary defect, decrease in color vision, optic pallor).

Treatment of orbitotemporal neurofibromas likely differs among medical centers with or without dedicated NF1 clinical teams, as most care for relatively few of these patients each year. Fewer than one-third of our patients received either ptosis surgery or debulking, compared with 90% of subjects in the study by Chaudhry and associates.³ Some of our patients received investigational medical therapy as part of a clinical trial in addition to surgical intervention. Since our 2 institutions primarily care for young children with orbitotemporal neurofibromas, it is not surprising that the primary indication for treatment was amblyopia.

While the results of our study and others describe the contributing factors for developing vision loss in children, they also highlight the fact that functional rather than surgical outcomes in these patients have not been well studied.¹³ Since only a portion of orbitotemporal neurofibromas will cause vision loss, guidelines are needed to determine the appropriate frequency of monitoring as well as indications for surgical or medical intervention. Consensus recommendations for the approach to children with NF1-related optic pathway gliomas have been developed and refined,^{14,15} but no such guidelines exist for children with orbitotemporal neurofibromas. Until a panel composed of pediatric NF1 specialists, pediatric ophthalmologists, neuro-ophthalmologists, and oculoplastic surgeons can convene, monitoring recommendations similar to those for optic pathway gliomas could be adopted. For example, any child with an identified orbitotemporal neurofibroma should have a complete ophthalmologic examination every 6 months, which includes quantitative, not qualitative, visual acuity testing, motility evaluation, intraocular pressure measurement, and cycloplegic refraction. Given the associated cognitive and behavioral problems of children with NF1,² it is conceivable that a quantitative visual acuity or a reliable pupil examination may not be feasible because of patient cooperation. If the child is old enough, evaluations of stereopsis and color vision should be included until visual maturation has been achieved. The frequency of ophthalmologic monitoring could be increased in cases of declining visual acuity, growth of the orbitotemporal neurofibroma, or new ophthalmologic symptoms. Use of these proposed guidelines may provide the basis for the development of an expert consensus on research priorities and monitoring/management guidelines.

Ideally, a standardized multicenter observational study would better elucidate the timing of changes in refraction and acuity and examine long-term ophthalmologic and quality-of-life outcomes. The addition of 3D MRI analysis may contribute to these outcomes and provide an objective metric in which to evaluate therapeutics.^{16,17} It is paramount that agreed-upon standardized clinical, imaging, and surgical outcome measures are developed as new biologic therapies are being introduced for plexiform neurofibromas, in order to allow for the optimal comparison of therapeutic and clinical outcomes between studies and centers.

16,17

A number of study limitations should be considered when interpreting our study results. Most notably, our statistical power is significantly limited by our small sample size. Despite both of our institutions having well-established NF1 clinical and research programs, our retrospective study design may have resulted in a selection bias. If selection bias did occur, it would likely favor enrolling more clinically symptomatic patients. However, the disease severity of our patients was less compared to other studies.¹³ Multiple clinicians, all experienced in caring for children with NF1, evaluated the study subjects during the routine clinical visits or research study visits. Different techniques in determining visual acuity, especially in the younger children, as well as variability in determining refractive error may have influenced the results. Fortunately, the differences in refraction between eyes affected by an orbitotemporal neurofibroma and the unaffected eyes were robust, so it is unlikely that any differences between clinicians significantly affected the outcome. Lastly, the variability of disease severity, the relatively few studies focusing on young children during visual development, and the lack of longitudinal outcomes in young children with orbitotemporal neurofibromas limits our ability to make formal recommendations about patient care guidelines.

In conclusion, young children with orbitotemporal neurofibromas are at high risk for vision loss, most commonly because of anisometropia and deprivation from ptosis. Frequent ophthalmologic examinations in children less than 10 years old are necessary to monitor for amblyopia and make well-informed treatment decisions. Multicenter prospective studies are needed to develop evidence-based guidelines to manage these challenging cases.

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Biosketch

Robert A. Avery, DO, MSCE, is a pediatric neuro-ophthalmologist at the Gilbert Family Neurofibromatosis Institute and Children's Research Institute within Children's National Medical Center in Washington, District of Columbia. Dr Avery received his medical degree from the Philadelphia College of Osteopathic Medicine, Philadelphia, Pennsylvania. He completed a residency in pediatrics at duPont Children's Hospital in Wilmington, Delaware

followed by fellowships in pediatric neurology and pediatric neuro-ophthalmology at the Children's Hospital of Philadelphia/University of Pennsylvania, Philadelphia, Pennsylvania.

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FIGURE 1.

Magnetic resonance imaging examples of orbitotemporal neurofibromas. (Top left) Small plexiform neurofibroma of the right upper orbit with typical central dot sign. (Top right) Plexiform neurofibroma invading the left upper eyelid and the lateral rectus muscle. (Bottom left) Diffuse infiltrative plexiform neurofibroma of the left eyelid, the lateral and posterior orbital space displacing the optic nerve and invading the cavernous sinus. (Bottom right) Massive, disfiguring plexiform neurofibroma expanding to the left side of face.

FIGURE 2.

Volumetric (3D) magnetic resonance imaging analysis of orbitotemporal neurofibromas. The area within the tumor border multiplied by image slice thickness is used to calculate the tumor volume (mL).

FIGURE 3.
Comparison of orbitotemporal neurofibromas volumes (mL; y-axis) in patients with and without amblyopia (x-axis).

TABLE 1.**Clinical Characteristics of Children With Orbitotemporal Plexiform Neurofibromas**

Subjects (N = 21)	
Location of OTPN, no. (%)	
Isolated eyelid	6 (29)
Eyelid and orbit ^a	7 (33)
Orbit and temporal region	7 (33)
Diffuse orbit	1 (5)
Etiology of amblyopia, no. (%)	
Strabismus	2 (10)
Occlusion from ptosis	9 (43)
Anisometropia	9 (43)
Multiple etiologies	6 (29)
Treatment for OTPN, no. (%)	
Ptosis surgery	2 (22)
OTPN removal from eyelid	3 (33)
Enucleation	2 (22)
Medication ^b	4 (44)
Reasons for treatment, no. (%) ^c	
Amblyopia	7 (33)
Cosmetic appearance	4 (19)
Exposure keratopathy	1 (5)

OTPN = orbitotemporal plexiform neurofibroma.

^aThree subjects had bilateral involvement.

^bSome attempted medication in combination with surgery.

^cThree subjects because of a combination of factors.

TABLE 2.

Ophthalmologic Outcomes and Volume of Orbitotemporal Plexiform Neurofibromas

Subject	Age (y)	Right Eye Refraction (D) ^a	Left Eye Refraction (D) ^a	logMAR Difference ^b	Affected Eye ^c	MRI Volume (mL) ^d
1	8	0.75 + 0.75	-1.00 + 1.00	0.4	Left	18.9
2	1	-0.25 + 0.00	-3.00 + 0.50	0.2	Left	19.4
3	2	-0.75 + 1.00	-1.50 + 1.25	0.0	Left	8.1
4	0.3	1.50 + 0.00	1.50 + 0.00	0.0	Left	- ^d
5	11	-1.00 + 1.50	0.00 + 0.75	0.3	Right	17.4
6	2	-2.25 + 1.00	-2.25 + 1.00	0.0	Left	48.0
7	8	0.50 + 0.00	0.50 + 0.00	0.0	Left	55.2
8	7	-	0.50 + 0.00	1.5	Right	53.2
9	13	0.00 + 0.00	-6.00 + 0.50	0.2	Left	315
10	17	0.00 + 0.00	-10.00 + 0.00	1.4	Left	81.2
11	18	-3.25 + 0.00	-5.00 + 0.50	0.2	Left	754.0
12	11	-6.50 + 1.25	-6.25 + 0.25	0.2	Left	299.0
13	12	-0.50 + 0.50	-2.25 + 3.50	0.2	Left	400.0
14	23	0.00 + 0.00	0.00 + 0.00	0.0	Right/left	7.4/2.7
15	18	-7.00 + 1.00	-7.25 + 0.75	0.0	Right	9.0
16	5	1.50 + 0.00	1.50 + 0.00	0.5	Left	14.2
17	18	-0.75 + 1.25	-0.50 + 0.50	0.0	Left	- ^d
18	9	0.00 + 0.00	0.00 + 0.00	0.2	Left	41.8
19	1	-1.00 + 0.75	1.25 + 0.00	0.1	Right	66.7
20	3	0.00 + 0.00	-3.00 + 6.00	0.6	Left	36.6
21	2	2.25 + 0.00	-2.50 + 7.25	0.8	Left	33.8

D = diopters; logMAR = logarithm of the minimal angle of resolution; MRI = magnetic resonance imaging.

^aPower plus cylinder.

^bDifference between eyes.

^cEye with orbitotemporal plexiform neurofibroma.

^dVolume not measurable.