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Vision Specific Quality of Life in Children with Optic Pathway Gliomas

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Abstract

Children with optic pathway gliomas (OPGs) frequently experience vision loss from their tumors. Most pediatric OPG research has focused on radiographic and visual outcomes, yet the impact of vision loss on quality of life (QOL) in children with OPGs has not been studied. The present study prospectively recruited children ≤ 10 years of age with sporadic or neurofibromatosis type 1 (NF1) -related OPGs. Vision specific QOL was assessed by parent proxy using the Children's Visual Function Questionnaire (CVFQ), and scores were analyzed according to magnitude of visual acuity (VA) loss and presence of visual field (VF) loss. Thirty-six subjects completed the study (53% female) with median age of 4.6 years. Children with mild, moderate and severe vision loss have lower CVFQ subscale scores, indicating a lower vision specific QOL, compared to those with normal vision. Lower Competence scores were noted in participants with more profound vision loss ($P < .05$), reflecting a decreased ability to complete activities of daily living (e.g. feeding, grooming). Children with two visually impaired eyes were rated as having greater difficulty with social interactions and pleasurable activities (Personality subscale, p=.039) compared to those with only one impaired eye. In summary, our findings demonstrate that children with vision loss secondary to their OPG have a decreased vision specific QOL compared to those with normal vision. Measuring vision specific QOL may be considered a meaningful secondary outcome measure for pediatric OPG clinical trials.

Introduction

Health-related quality of life (HRQOL) measures play an increasingly important role in evaluating both short and long term outcomes in children with chronic illness, including tumors of the central nervous system [1]. Investigators have been forced to use broad based measures of QOL in children given the diversity of tumor type, tumor location and treatment regimens [1], although recent instruments have focused on symptoms specific to brain tumors [2–5]. Children with optic pathway gliomas (OPGs), low-grade gliomas involving only the afferent visual pathway (i.e., optic nerve, chiasm and tracts), are a somewhat more

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homogeneous group of patients who have a relatively high long term survival rate and where preservation of visual function can be the primary treatment goal [6–8].

Vision loss in both sporadic- and neurofibromatosis type 1 (NF1)- related OPGs typically occurs between one and ten years of age, with a median incidence from three to five years old [6, 9]. Many children with OPGs experience permanent and sometimes profound visual acuity (VA) loss from their OPGs, ranging from mild deterioration (e.g., 20/40) to complete blindness. OPGs can also result in significant visual field (VF) loss, even in the context of normal VA. Vision loss during adulthood may have a profound impact on QOL, mortality and employment [10, 11]. In children, vision loss can significantly affect the development of academic and social abilities, in addition to their acquisition of skills related to self-care, mobility, and independent function. Vision loss in children with brain tumors likely confers additional risk for poor school performance and social functioning in a group already at higher risk for cognitive and learning difficulties [12–14]. Since new or progressive vision loss is frequently a compelling factor to initiate treatment of OPGs, the impact of vision loss on quality of life (QOL) is clearly coupled to this decision, yet it has not been studied.

Therefore, our objective was to examine prospectively vision-related QOL in children with OPGs using a measure developed to evaluate this construct in young children. Since vision loss secondary to OPGs and treatment for OPGs typically occurs between one and ten years of age, we investigated the impact of vision loss on vision specific QOL in this age group. We hypothesized that caregivers of children with VA and VF deficits would report poorer vision-specific QOL, and that QOL outcomes would worsen along with the extent of visual impairment.

Methods

Patients

Subjects between one and ten years of age with previously identified OPGs were recruited during their routine neuro-ophthalmology clinic visit at Children's National Medical Center (Washington, D.C.). An OPG, as determined by a pediatric neuroradiologist, was defined as abnormal enlargement and or signal change (T2, FLAIR or contrast) involving any of the following structures: optic nerve, optic chiasm and or tracts. Children with NF1-related OPG were required to have both a diagnosis of NF1 based on established NIH criteria and an MRI of the brain demonstrating the presence of an OPG [15]. Children with sporadic OPGs were diagnosed by MRI findings characteristic of a low grade OPG and/or diagnostic biopsy results. Patients were excluded if they had a history of ophthalmologic disease, other than an OPG, that could have affected their VA, VF or their optic nerve function (e.g., retinopathy of prematurity, amblyopia, glaucoma). To avoid selection bias and to obtain a representative sample, recruitment was initiated immediately after obtaining institutional review board approval and stopped once the enrollment goal of 36 subjects was achieved. The institutional review board at Children's National Medical Center approved the protocol and the parent/guardian provided informed consent prior to study enrollment.

Parents/guardians of the study subjects were approached after their child's neuro-ophthalmology clinic visit had been completed. If they agreed to participate and provided consent, they were seated in an unsupervised waiting room or exam room and asked to complete a general information sheet (requesting basic medical and demographic information) followed by the Children's Visual Function Questionnaire (CVFQ). Caregivers also were asked to report on the presence of mental health or learning issues (ADHD, anxiety disorder, learning disabilities), and current formal school accommodations (e.g., IEP or 504 plan). Clinical information was abstracted from the patient's current examination including VA, VF (scored as normal or abnormal) and diagnosis (sporadic vs. NF1-related

OPG). All study information and CVFQ scores were collected during a single visit for this cross-sectional design.

Children's Visual Function Questionnaire

The CVFQ is a vision specific QOL measure that was specifically developed and validated in children seven years of age and younger since most pediatric eye diseases present and are treated during this age range [16, 17]. Given the young age of children being assessed, the CVFQ was designed as a proxy assessment to be completed by caregivers. Items were developed to evaluate vision-related QOL in four domains: Competence, Personality, Family Impact and Treatment Difficulty, all of which have been demonstrated to be valid and reliable when examining a variety of pediatric eye conditions [17]. The Competence scale assesses the impact of vision loss on physical activities (i.e., walking, biking, ambulating in public places) and activities of daily living (i.e., grooming, dressing). The Personality scale evaluates the child's demeanor, interaction with peers, and enjoyment of common activities (i.e., television, books, crafts, playing with peers). The social consequences on both an individual level (i.e., teased because of vision problems, commentary from others about the child's disability) and family level (i.e., time away from other children or spouse, time spent on care) are addressed in Family Impact subscale. The Treatment subscale of the CVFQ was not analyzed as the majority of the questions were not applicable to children with OPGs.

Parents of children three and older completed the 39-item CVFQ , whereas those with children younger than three years completed the 34-item version of the CVFQ (which excludes questions that were not age appropriate) [17]. Each item for the subscales described above is rated on a four-point Likert scale, anchored by 1("best") and 0("worst"). Additional items relating to General Vision are rated on a 5-point scale. Scores for each subscale are summed and divided by the number of items in each domain, allowing for total and subscale scores from the younger and older version of the CVFQ to be included in the same analysis.

Definition of Clinical Outcomes

VA was categorized identically to previously-published reports using the CVFQ [17] as normal, borderline (within 0.1 logMAR of age-based norms), mildly impaired (within 02. to 0.3 logMAR of age-based norms), moderately impaired (within 0.4 to 0.9 logMAR of age-based norms), or severely impaired (≥ 1.0 logMAR of age-based norms). However, for the analyses presented below, we considered children with ratings of normal or borderline visual acuity in the same category as compared with those classified as having either mild, moderate, or severe visual acuity impairment.

Statistical Analysis

Demographic and clinical characteristics were summarized by standard descriptive statistics. Sample size was determined by estimating the number required to detect a CVFQ subscale difference of 0.2 or greater between groups with a standard deviation of 0.15 at 80% power. Given that subscale scores on the CVFQ have a restricted range and our sample was derived from a clinical population, we first examined the distribution of CVFQ outcome variables of interest. Although the Personality and Family Impact subscales were broadly normally distributed, the Competence subscale was characterized by both kurtosis and negative skew. Therefore, nonparametric Kruskal-Wallis and Mann Whitney tests were used to compare differences in the three CVFQ subscales of interest (i.e., Competence, Personality, Family Impact) as a function of visual acuity (number of affected eyes and impairment category), visual field deficit (impairment category), NF1 diagnosis, and past or current history of chemotherapy.

Results

Thirty-six subjects completed the study (See Table 1 for demographic and clinical information). Recruitment commenced on March 1, 2012 and concluded on July 11, 2012 once the enrollment goal of 36 subjects was achieved. No parent/guardian refused to complete the CVFQ. Nine children with NF1 related OPG were identified as having mental health/learning disabilities (ADHD = 4, Learning disability = 5), 5 of which received formal school accommodations. Five children with sporadic OPG had mental health/learning disabilities (Anxiety = 1, Learning disability = 4), 4 of which received formal school accommodations. Table 2 lists the VA and VF results. One participant's VA and VF loss could not be accurately quantified or categorized due to the patient's development delay secondary to NF1, therefore this participant was eliminated from subsequent analysis.

Children who were categorized as having mild, moderate or severe VA loss had significantly lower perceived functioning on the Competence ($U = 35.5$, $p = .001$) and Family Impact ($U = 67.5$, $p = .022$) subscales than children with normal or borderline VA (See Table 3). Participants with two affected eyes were rated as having significantly lower Competence ($H = 14.7$, $p = .001$) and Personality ($H = 6.5$, $p = .039$) scores compared to those with either no VA problems or those with VA loss affecting only one eye (See Figure 1). Post-hoc analysis of multiple comparisons for Competence scores did not reach significance when comparing participants with VA affecting one versus zero eyes ($p = 0.44$) and trended towards significance when comparing those with VA affecting one versus two eyes ($p = 0.14$). Post-hoc analysis of multiple comparisons for Personality scores did not reach significance when comparing participants with VA affecting one versus zero eyes ($p = 0.9$) and trended towards significance when comparing those with VA affecting one versus two eyes ($p = 0.06$). Examination of visual functioning in those with and without visual field deficits indicated significant differences in ratings only for perceived Competence ($U = 78.5$, $p = .026$). Finally, we also examined whether medical variables were associated with differences in parent ratings of visual functioning. Neither NF1 diagnosis nor history of past or current chemotherapy were associated with significant differences in vision-related Competence ($p = 0.51$ and $p = 0.33$, respectively) or Personality ($p = 0.84$ and $p = 0.53$, respectively). Diagnosis of NF1 was not associated with Family Impact ($p = 0.55$), though there was a trend for those who had received chemotherapy to be rated as having had greater Family Impact ($U = 99.5$, $p = .053$).

Discussion

Using the CVFQ, we demonstrated that children with vision loss from their OPGs have a lower vision-specific QOL than children with the same tumors but normal vision. VA loss was significantly predictive of poorer functioning in the Competence and Family Impact domains, but not the Personality subscale. The Competence subscale addresses common activities of daily living such as the ability to brush teeth or feed independently, or to recognize others, whereas the Family Impact subscale addresses the parents' concern about the child's vision as well as the impact of impaired vision on the child's social functioning. Finally, the Personality subscale describes parents' perceptions of their child's enjoyment of common social and recreational activities (e.g., visiting with friends and family, watching television). Not surprisingly, those children with vision loss in both eyes had significantly lower Competence and Personality scores compared to those with either no VA loss or those with VA loss affecting only one eye. These findings indicate that bilateral VA loss may impact not only young children's acquisition and independent performance of daily living skills, but also the level to which they engage and take pleasure in many social and recreational activities. In contrast, visual field loss was associated with lower Competence ratings only. Thus, while children with both VA and VF loss may be seen by their caregivers

as struggling with autonomy related to self-care and socialization, only children with OPG-related bilateral VA loss are perceived as being limited in their enjoyment of a number of common childhood activities.

While these results may be at least partly intuitive, it is particularly relevant to understand the extent to which VA/VF deficits are associated with adaptive difficulties in children with OPGs because treatment decisions are often predicated on the emergence or progression of vision loss [6, 9]. Because most children with OPGs can be expected to survive the diagnosis [8], timing of treatment, or whether to treat at all, may be based more on the desire to minimize vision loss rather than to maximize survival. In this way, understanding how loss of VA or VF in one or both eyes is likely to impact QOL in both the short- and long-term is at least as relevant to the decision-making process as considering the likelihood of late-effects associated with different treatment modalities. For these reasons, inclusion of the CVFQ as a secondary outcome measure in clinical trials seems reasonable. Specifically, changes in CVFQ scores between trial entry and trial completion could provide a more comprehensive assessment of treatment benefit/failure than primary outcomes such as change in tumor size—which is not predictive of functional visual outcomes[18, 19]. Also, baseline vision specific QOL values could potentially help stratify patients to more or less aggressive treatment arms.

There are conflicting reports on whether monocular vision loss impacts QOL in the setting of a normal-seeing fellow eye. Speculating that the normal-seeing fellow eye can adequately compensate for a visually-impaired eye, Angeles-Han and colleagues reported that monocular vision loss in children with uveitis did not impact vision-related QOL [20]. However, in our study monocular VA loss did result in lower Competence and Family Impact QOL subscales compared to individuals with normal VA in both eyes. Our results are consistent with numerous other studies reporting lower vision-related QOL in subjects with monocular VA loss from multiple sclerosis [21–23], unilateral retinal detachment [24], cataracts [17], pituitary adenoma [25], and retinopathy of prematurity [17].

The VF assessment is notoriously difficult and unreliable in young children. In children under 10 years of age, the VF cannot be quantitated in a majority of cases. Determining partial versus complete quadrant or hemifield defects is typically not possible, therefore many practitioners note the qualitative presence or absence of VF defects. Given the inherent difficulties in quantitating VF loss in this young age group, it is difficult to determine what magnitude of VF loss is required to impact QOL. Furthermore, only 4 subjects had VF loss in the setting of normal VA, therefore limiting our ability to separate the impact of combined VF and VA loss on vision-specific QOL.

Children and adolescents with NF1 are known to have a lower health-related QOL compared to reference populations [26, 27]. In our study, a diagnosis of NF1 was not associated with CVFQ scores, suggesting that visual impairment alone was responsible for the lower vision-specific QOL scores. This hypothesis is further supported by the fact that both children with sporadic OPGs (5/12, 42%) and NF1 related OPGs (9/24, 38%) in our study had similar frequencies of reported mental health and or learning disabilities. It should be noted that the diagnosis of ADHD and learning disabilities may not always be established in the very young age. Also, OPG treatment with chemotherapy did not influence the Competence or Personality subscales, again supporting a close relationship between visual impairment and vision-specific QOL measures. Not surprisingly, OPG treatment did show a trend that neared statistical significance for influencing the Family Impact score, suggesting that receiving OPG-targeted chemotherapy may affect parents' concerns about their child's visual functioning over time. Additional research is needed, however, to clarify whether these concerns are mitigated with treatment efficacy or time. Also, future research in older

children with OPGs could evaluate the association between caregiver's perceptions of vision related difficulties and actual vision ability.

The study was limited by a relatively small sample size and reliance on a QOL measure that was evaluated by parent proxy. It is notable that very few vision-specific QOL and vision ability questionnaires have been developed for young children [17, 28–30]. Although self-report measures exist, they have either been developed for adult samples or measure visual ability rather than vision-related QOL [16, 17]. While the self-reported format may be preferred, the young age of at which most patients experience OPG-related vision loss may make it difficult to acquire reliable self-report data in this population. Indeed, our inclusion criteria for age was based on the fact that most OPGs are discovered, monitored and when necessary, treated between the ages of 1 and 10 years of age [6, 9]. Although young children have been able to independently complete some broad-based QOL measures [31], the parent proxy method has been validated in medically ill children [32, 33]. Moreover, while some visual ability questionnaires use self-reporting, the only two vision specific QOL measures for children use the proxy method [17, 29].

Despite limiting the enrollment age to 10 years, a wide range of social, emotional and cognitive differences likely exist within our younger (<3 yo) and older (≥ 3) subject groups. Therefore, some questions from the CVFQ may not be relevant to children within the same age group, such as riding a bicycle. Fortunately, to account for such differences, the CVFQ allows parents to score that feature as "does not apply to my child" or "my child is too young to attempt this." Another limitation of our study is that our hospital serves as a regional referral center for children with NF1, the proportion of these children in our study population may not be representative of other clinical practices.

Conclusion

In conclusion, our study demonstrated that children who have experienced vision loss from their OPG have a lower vision-specific QOL than those children with the same tumor and normal vision. Given the increasing importance of patient-reported and QOL outcomes as endpoints in intervention studies, future research should evaluate the association between improved visual outcomes and QOL in children treated for OPGs. Multi-center collaborative studies are needed to better determine the influence of clinical factors (i.e., tumor location, tumor recurrence, type of chemotherapy, isolated VF loss, median time from vision loss to completing the CVFQ, etc) on vision-specific QOL measures. In addition, subsequent investigations should also assess the relation between vision-specific QOL measured in early childhood and developmental outcomes over time. Given the CVFQ's high test-retest reliability, [17] it may be considered a meaningful secondary outcome measure for OPG clinical trials.

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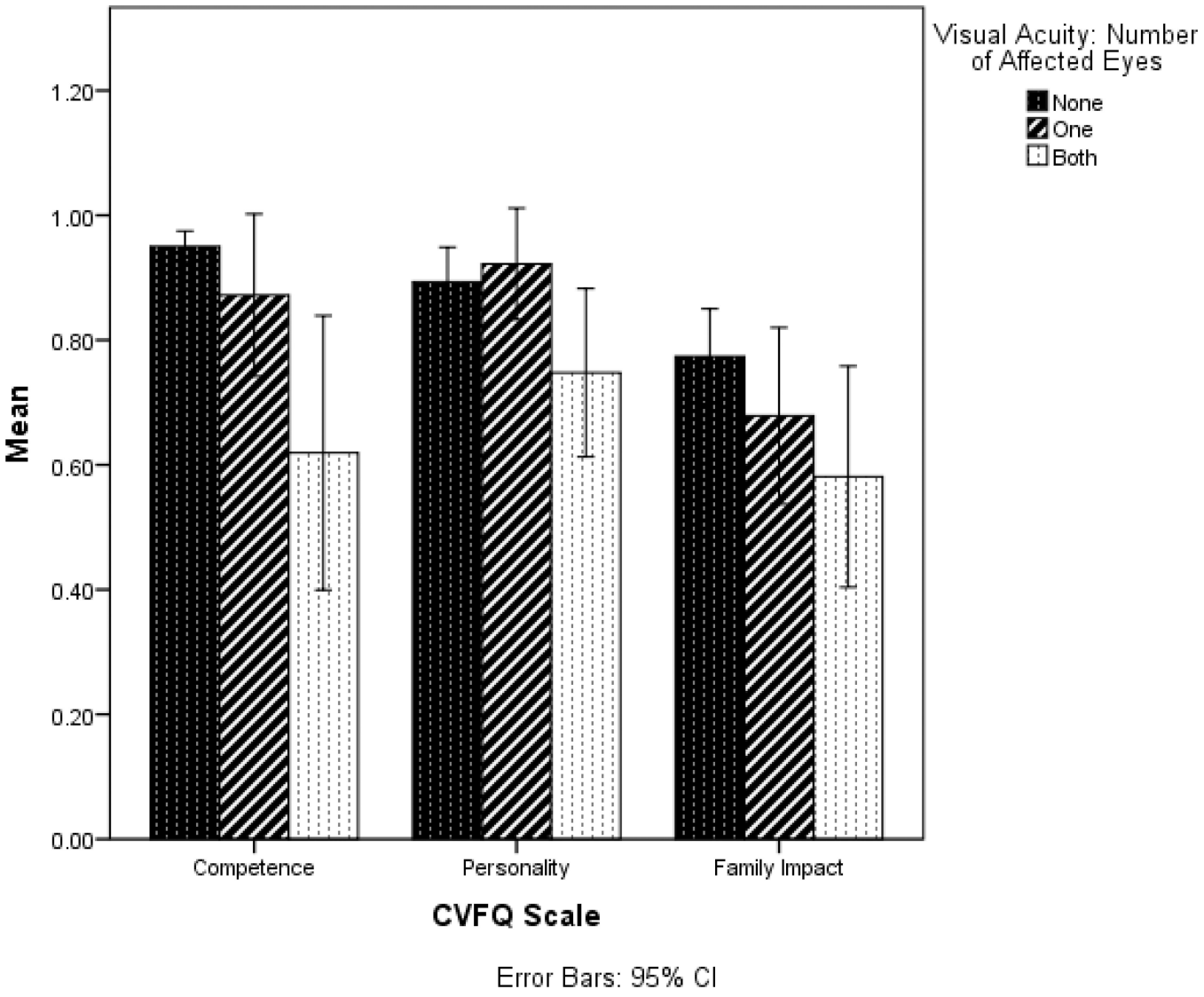

Figure 1.
Differences in CVFQ subscale score as a function of Visual Acuity.

Table 1

Demographic and Clinical Characteristics

	Study Subjects (N = 36)
Age[a]	5.09/4.62
Range	(1.75 – 10.58)
Age under 3 years – no. (%)	7(19)
Female sex – no. (%)	19 (53)
Race – no. (%)	
Black/African American	1 (3)
White/Caucasian	30 (83)
Asian	2 (6)
Multiple races	3 (8)
Ethnicity – no. (%)	
Non-Hispanic	31(86)
Hispanic	5 (14)
Diagnosis – no. (%)	
NF1 with OPG	24(67)
Sporadic OPG	12(33)
Mental Health/Learning Disability– no. (%)	
NF1 with OPG	9(25)
Sporadic OPG– no. (%)	5(14)
Treatment of OPG– no. (%)	
Never Treated	16 (44)
Treated	20 (56)
Past Treatment[b]	11
Currently undergoing chemotherapy	9

[a] In years mean/median and (range);

[b] Includes chemotherapy or radiation (N=2)

Table 2

Visual Acuity and Visual Field Results

	Study Subjects (N = 36)[a]
OD Category of Visual Acuity – no. (%)	
Normal	23 (66)
Borderline	4 (11)
Mild impairment	3(9)
Moderate impairment	1(3)
Severe impairment	4 (11)
OS Category of Visual Acuity – no. (%)	
Normal	22 (63)
Borderline	4(11)
Mild impairment	3(9)
Moderate impairment	3 (9)
Severe impairment	3 (9)
Eyes with Abnormal VA[b]	
Zero	24 (69)
Monocular	5 (14)
Binocular	6 (17)
Eyes with Abnormal VF[c]	
Zero	22 (63)
Monocular	6 (17)
Binocular	7 (20)
OD Visual Field	
Normal	26 (74)
Abnormal[c]	9 (26)
OS Visual Field	
Normal	22 (63)
Abnormal[c]	13 (37)

[a] One subject's acuity could not be categorized due to developmental delay and cooperation.

[b] VA classified as mildly impaired or worse.

[c] Visual field loss in one or more quadrant.

Table 3

Vision and CVFQ subscales

	Normal Vision[a] (N=24)	Vision Loss[b] (N=11)
General Health[c]	0.83/0.88	0.68/0.75
	(0.0–1.0)	(0.25–1.00)
General Vision[c]	0.91/1.00	0.50/0.60
	(0.70–1.00)	(0.10–0.80)
Competence[c]	0.95/0.97	0.72/0.82
	(0.75–1.00)	(0.25–1.00)
Personality[c]	0.89/0.95	0.82/0.86
	(0.56–1.00)	(0.45–1.00)
Family Impact[c]	0.77/0.80	0.63/0.63
	(0.25–1.00)	(0.34–0.95)

[a] Patient eyes with a normal high contrast visual acuity (within 0.1 logMAR for age) and normal visual fields.

[b] Includes patients with abnormal VA/normal VF, normal VA/abnormal VF or abnormal VA/abnormal VF.

[c] Mean/median and range in parenthesis.