Functional Visual Loss in Adults and Children

Patient Characteristics, Management, and Outcomes

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Objective: To compare the characteristics of functional visual loss (FVL) in adults and children.
Design: Retrospective chart review.
Participants: One hundred forty patients diagnosed with FVL over a 5-year period.
Methods: Medical records of these patients were reviewed and data analyzed using statistical software.
Outcome: Demographics, underlying organic and psychiatric disease, concomitant psychosocial events, and resolution rates were studied.
Results: Functional visual loss, with or without functional overlay, was initially diagnosed in 140 patients and was, in retrospect, a correct diagnosis in 138. There were 56 (40.6%) children and 82 (59.4%) adults (mean age, 13.4 and 40.0 years). The gender ratio, incidence of concomitant psychosocial events, incidence of functional overlay, prevalence of migraine or facial pain, and proportion referred for counseling were similar in the 2 groups. Concomitant psychosocial events were primarily social in children and related to trauma in adults. Thirty-two (39.0%) adults had a history of psychiatric illness, versus 10 (17.9%) children ($P = 0.008$). Symptoms were bilateral in 65.0% of cases. Functional visual loss manifested as visual acuity (VA) loss only occurred in 26.1% of patients, FVL manifested as visual field (VF) loss only was present in 28.3% of patients, and FVL with loss of both VA and VF occurred in 45.6% of patients. There was no significant difference in children versus adults in the proportion of VA, VF, or both being affected. Functional visual loss with coexistent organic disease (functional overlay) was present in 16.7% of patients. Follow-up information was available for 26.1% of patients. Normalization of any one parameter occurred in 58.3% of patients and was more likely in children. Three patients (2.2%) originally felt to have solely functional disease were subsequently diagnosed with organic disease.
Conclusion: Functional visual loss is most common in teenagers, is typically bilateral, and involves both VA and VF. Normal VA was proven half the time at initial consultation. At all ages, patients were predominantly female, and one fifth had migraine, facial pain, or coexistent organic pathology. Concomitant psychosocial events were mainly social in children and related to trauma in adults. Psychiatric disease was twice as likely in adults. Normalization of visual function occurred in a majority of patients. Early-onset macular dystrophies and hereditary optic neuropathies may be misdiagnosed as FVL. *Ophthalmology 2005;112:1821–1828 © 2005 by the American Academy of Ophthalmology.*

Incidences of functional visual loss (FVL) in an outpatient ophthalmology clinic setting have been reported to be approximately 1.75% in children and 5.25% in adults.[1,2] Many ophthalmologists, however, are often reluctant to make a diagnosis of FVL; on the one hand, there is no objective pathology to support the diagnosis; on the other, there is the possibility of missing an organic, treatable cause of visual loss. Recognizing FVL and managing it appropriately minimize patient distress, inappropriate referrals, and unnecessary health care and disability expenditures. The purpose of this article is to describe and compare the clinical characteristics of FVL in children and adults and outline our management principles for these patients. We also report the association of FVL with chronic pain syndromes (e.g., migraine, trigeminal neuralgia). In addition, we review cases in which organic pathology was initially misdiagnosed as FVL.

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Materials and Methods

After Oklahoma University Health Sciences Center Institutional Review Board approval, we performed a retrospective chart review

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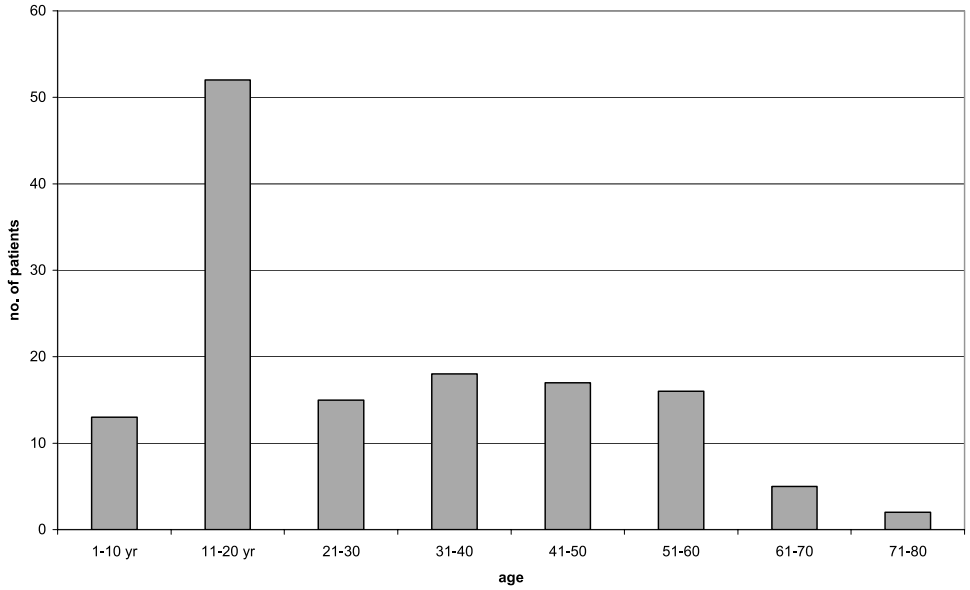

Figure 1. Age (years) distribution of patients with functional visual loss.

of all patients seen by the Neuro-ophthalmology Service at the Dean A. McGee Eye Institute with a diagnosis code of visual disturbance, unspecified from May 1, 1999 to April 30, 2004 (International Classification of Diseases 9, 368.10). Out of a total of 1486 cases, we identified 140 patients who were diagnosed clinically with FVL.

The diagnosis of FVL was a clinical one made by the attending neuro-ophthalmologist. Diagnoses of functional visual acuity (VA) loss and functional visual field (VF) loss were made if either or both of the visual complaints could be proven nonorganic. The following inclusion criteria were used: (1) normal structural ocular examination results or abnormalities unrelated to the VA or VF loss and (2) clinical evidence that visual function was better than that claimed, or a nonphysiologic response to testing in at least one modality. Functional overlay was defined as abnormal structural ophthalmologic examination results but with visual loss either out of proportion to or unexplained by the abnormality noted on the examination. We excluded patients who had (1) inadequate chart documentation, (2) insufficient evidence for nonorganic visual loss (e.g., unable to prove that they see better than claimed), or (3) unreliable but not clearly proven to be nonorganic VFs.

We employed a variety of commonly used clinical tests to diagnose FVL. For example, functional VA loss was often demonstrated via optokinetic responses, acuity testing with fogging, stereoacuity, or a slow tedious refraction with encouragement. To demonstrate functional VF loss, we showed spiraling/crossing of isopters on Goldmann perimetry, persistence of a unilateral field defect under binocular conditions, or a nonexpanding (tunnel) confrontation VF at 1- and 2-m testing distances.

The following data were collected:

1. Patient demographics: age, gender, and type of insurance coverage (private vs. public assistance).
2. Presence of concomitant psychosocial events (e.g. physical trauma, sexual abuse, stressors at school, sibling rivalry).
3. History of psychiatric disease. These data were obtained via a routine review-of-systems questionnaire that all patients at our institution complete before initial consultation.
4. Presence of FVL with functional overlay (see definition above).
5. Referral by the ophthalmologist for psychiatric/ psychological counseling.
6. Type of FVL, classified as FVL with VA loss only, FVL with VF loss only, and FVL with both VA and VF loss.
7. Follow-up and frequency of resolution of FVL. Inclusion criteria for patients with follow-up data include (1) patient seen by attending neuro-ophthalmologist in clinic with adequate chart documentation; (2) patient communication with the attending neuro-ophthalmologist by e-mail or telephone conversation, with adequate information to address the problem; or (3) subsequent records from another ophthalmologist or optometrist documenting improvement of visual function.
8. Presence of migraine and/or trigeminal neuralgia, as ascertained by patient history.

Analysis of the data was performed using statistical analysis software (SPSS, SPSS Inc., Chicago, IL). We divided our patients into 2 age groups: children ($\leq$17 years old) and adults ($\geq$18 years old).

Results

Functional visual loss was initially diagnosed in 140 patients. In 3 patients, a subsequent diagnosis of organic disease was made at a later date. One of these 3, who had FVL with functional overlay at presentation, is included in the following analysis, which consisted of 138 patients (56 children [40.6%] and 82 adults [59.4%]).

The age distribution of our patients is shown in Figure 1. The age range was from 7 to 76 years, with a mode of 17.0 years. Of the 138 patients, 52 were in the second decade of life. Further analysis of these 52 patients revealed that 22 were 16 or 17 years old. There was an even distribution of patients from the third decade to the sixth decade, after which the frequency decreased.

The gender distribution, frequency of patients on private insurance, presence of concomitant psychosocial events, history of psychiatric illness, proportion with FVL with functional overlay, and number referred for psychiatric or psychological counseling are shown in Figure 2. Comparison of these parameters in children and adults revealed that the only significant difference was in the

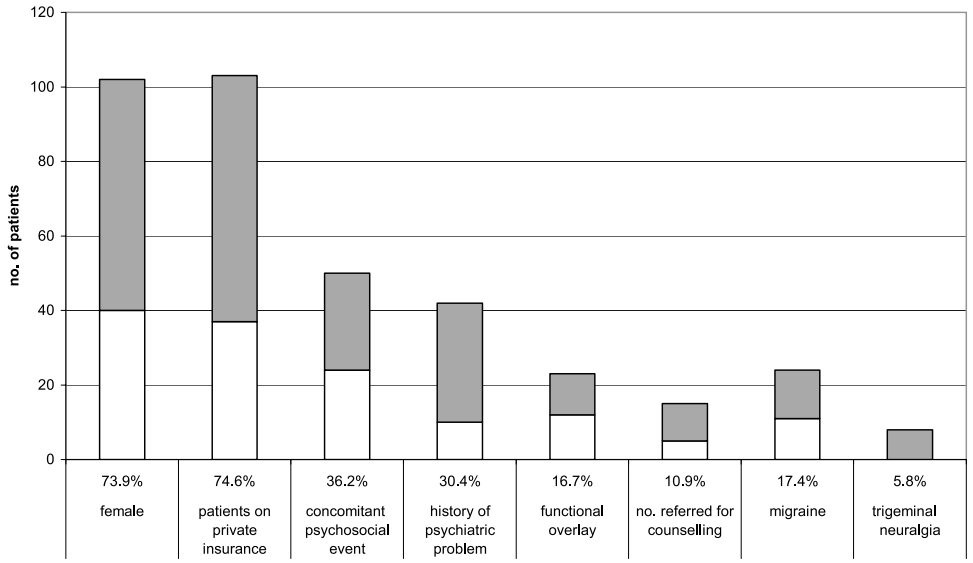

Figure 2. Selected demographics of patients with functional visual loss. □, child; ▨, adult.

presence of underlying psychiatric history, which was higher in adults (17.9% vs. 39.0%, $P = 0.008$).

Concomitant Psychosocial Events

A list of concomitant psychosocial events is presented in Table 1. Of the 138 patients, a concomitant psychosocial event was elicited in 50 (36.2%). There was no significant difference in the proportion of children versus adults (42.9% vs. 31.7%, $P = 0.18$) or the gender distribution of patients with a concomitant psychosocial event (47.2% male vs. 32.4% female, $P = 0.11$). However, there was a difference in the type of psychosocial event in children versus adults. Social problems at home or in school were the major associations in children (45.8%), whereas physical trauma (57.7%) predominated in adults.

Of great significance, review of the social problems in children revealed that 2 were related to sexual abuse. In one patient, FVL was an initial presentation of the incident and brought the crime to light. In the second case, known sexual abuse had occurred some time earlier, and the patient developed FVL when the case was being tried. In adults, one episode was related to sexual abuse;—in this case, the patient's daughter being the victim. The other social events in children were diverse; there were 3 related to school stress and 1 related to each of the following: divorce, new guardian, mother's new "friend" living in, conflict with dad, finding a summer job, mother admitted to drug rehabilitation program, and death of sibling. The last patient came from a family with a history of Pelizaeus–Merzbacher disease and revealed that she felt she did not receive enough of her parents' attention.

Table 1. Types of Concomitant Psychosocial Events in Children and Adults

	Children* (n = 24)	Adults† (n = 26)
Physical trauma	6	15
Social (schoolwork or home environment stressors)	11	1
Systemic or ocular pathology	7	8
Sexual abuse	2	1
After surgery	0	3 (LASIK, blood donation, spine surgery)
Wants glasses	1	0

*Three patients had more than 1 trigger.
†Two patients had more than 1 trigger.

Closed head trauma accounted for 10 of the 21 cases of physical trauma (47.6%). There was more work-related trauma in adults than in children.

In 15 patients, a systemic or ocular pathology preceded the FVL and was thought to be associated as a psychosocial event that precipitated the FVL. This occurred with equal frequency in both age groups. In children, coexistent ocular or systemic diseases included neuroretinitis (n = 1), retinal dystrophies (n = 2), migraine with visual aura (n = 2), Crohn's disease (n = 1), and a viral illness requiring hospitalization (n = 1). In adults, entities were LASIK surgery with suboptimal outcome (n = 1), optic nerve drusen (n = 1), undiagnosed amblyopia (n = 2), thyroid eye disease (n = 1), migraine with aura (n = 2), and ovarian cancer (n = 1).

Physical trauma was an associated factor in 15 adult cases (18.3% of total adult cases) and 6 pediatric cases (10.7% of cases). Eight of 15 adults (53.3%) were involved in litigation with attention to their purported visual loss. None of the pediatric cases involved nonaccidental trauma, and abuse was not suspected in any of these cases.

Migraine and Trigeminal Neuralgia

The number of patients with migraine and/or trigeminal neuralgia is shown in Figure 2. There was no difference in the proportion of children and adults with migraine. All patients with trigeminal neuralgia were adults.

Eleven of the 24 patients with migraine were children. Two of the 11 children were boys, and the mean age of this group was 11.3 years (range, 8–17; mode, 16). Nine of these 11 children had migraine associated with visual aura. Functional visual loss due to VA loss only was present in 2 children, and FVL due to VF loss only was present in 3, whereas 6 children had FVL due to both VA loss and VF loss.

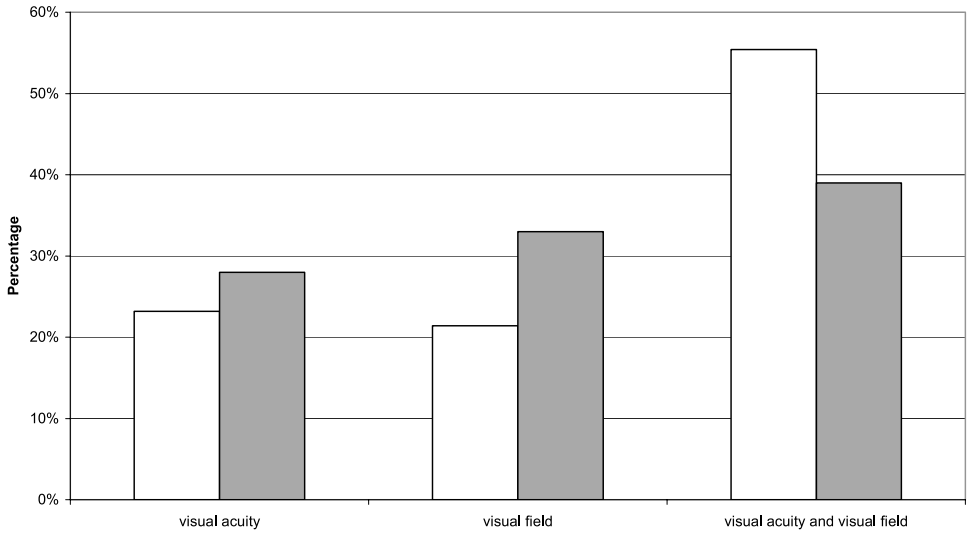

Figure 3. Type of functional visual loss. □, child; ▨, adult.

Migraine was present in 13 adults, of whom 12 were female. The mean age was 34.1 years. Visual aura was present in 7 patients. Functional visual loss due to VA loss only was present in 2 patients, FVL due to VF only was present in 5, and FVL due to both VA loss and VF loss was present in 6.

Of the 8 patients with trigeminal neuralgia, 2 had posttraumatic disease, whereas the other 6 were idiopathic. One had FVL due to VA loss only, 4 had FVL due to VF loss only, and 3 had FVL due to both VA loss and VF loss. Six patients had FVL ipsilateral to the side affected with trigeminal neuralgia, whereas 1 patient had FVL in both eyes. The remaining patient initially presented with FVL ipsilateral to the side with trigeminal neuralgia, but progressed to develop bilateral symptoms.

Functional Visual Loss with Functional Overlay

Functional visual loss with functional overlay was present in 23 of the 138 patients with FVL (16.7%), 12 of 56 children (21.4%), and 11 of 82 adults (13.4%) ($P = 0.22$).

In patients with FVL and functional overlay, 9 patients had unilateral ocular pathology: amblyopia (n = 6) and optic neuropathy (n = 3) (optic neuritis [n = 1] and morning glory anomaly [n = 2]). Fourteen patients had bilateral disease, 12 with the same condition in each eye (optic neuropathy [n = 5], neuroretinitis [n = 1], keratoconus [n = 3], cone dystrophy [n = 2], and foveal hypoplasia [n = 1], whereas 2 patients had different pathology in each eye.

Seven patients had unilateral organic disease with FVL in the contralateral normal eye. The conditions responsible in these cases were optic neuropathy (n = 3), macular scar (n = 2), amblyopia (n = 1), and enucleation (n = 1).

Type of Functional Visual Loss

The type of FVL (FVL with VA loss only, FVL with VF loss only, or FVL with both VA loss and VF loss) present in our patients is shown in Figure 3. There was no difference in the distribution of the 3 types of FVL in children versus adults.

Of the 138 patients with FVL, 99 had functional VA loss (71.7%). Of the 99, we were able to demonstrate normal VA (20/30 or better) during the initial office visit in 46 (46.5%). We were unable to prove 20/30 or better acuity in 34 patients (34.3%) but demonstrated that they could see better than claimed and found no pathology to account for their symptoms. In another 19 patients in whom we were unable to prove normal acuity in the clinic (19.2%), underlying organic disease partially accounted for subnormal VA. Bilateral VA loss was present in 63 patients (63.6%), and 36 were unilateral (36.4%). Of the 36 unilateral cases, 19 were in the right eye (52.8%) and 17 in the left eye (47.2%).

Formal VF testing was performed in 110 of these 138 patients. Of these, 102 had functional VF loss (92.7%). Of these 102 patients, we were able to demonstrate nonphysiologic fields by Humphrey or Goldmann VF testing (Table 1) in 37 (36.3%). Fifteen patients (14.7%) had organic disease with functional overlay. In the remaining 50 patients (49.0%), we were able to demonstrate tunneling of VFs on confrontation testing. Bilateral VF loss was present in 69 patients (67.7%), and 33 were unilateral (32.3%). Of the 33 unilateral cases, 19 were in the right eye (57.6%) and 14 in the left eye (42.4%).

Follow-up

Follow-up data were available for 36 patients (26.1%) (13 children, 23 adults). The duration of follow-up ranged from 1 to 60 months (mean, 31.5). Of the 13 children, 10 had FVL due to VA loss only, 2 had FVL due to VF loss only, and 1 had FVL due to both VA loss and VF loss. Of the 23 adults, 12 had FVL due to VA loss only, 5 had FVL due to VF loss only, and 6 had FVL due to both VA loss and VF loss. Resolution of any one parameter occurred in 21 patients (58.3%) (11/13 children [84.6%] and 10/23 adults [43.5%]). This difference was statistically significant ($P = 0.016$).

For all 99 patients with functional VA loss, we had follow-up information on 29 (29.3%). Resolution occurred in 19 patients (65.5%): 10 of 11 children (90.9%) and 9 of 18 adults (50%) ($P = 0.025$). Four of 9 males (44.4%) had resolution of VA, compared with 15 of 20 females (75%) ($P = 0.109$).

For all 102 patients with functional VF loss, we had follow-up data on 14 (13.7%); only 7 of these had resolution (50%): 2 of 3 children (66.7%) and 5 of 11 adults (45.5%) ($P = 0.52$). None of the 4 male patients had resolution, whereas 7 of the 10 females did (70%) ($P = 0.018$).

Misdiagnosis

Three of our patients were subsequently diagnosed with organic disease that was missed on initial consultation (2.2%). One patient clearly had FVL with functional overlay on initial consultation and was included in our analysis of FVL (case 1 below), whereas the other 2 patients were thought to have FVL on initial presentation but, in retrospect, had organic pathology accounting for all of their visual loss. We present these case reports here.

Case 1. A 16-year-old female complained of decreased vision over several years—specifically, of difficulty seeing the board at school and problems with contrast. Old records documented VAs of 20/20 in each eye 3 years before our consultation. She had been seen by a neurologist and told she may have multiple sclerosis.

Initial corrected VAs were 5/200 in the right eye (−7.50 +2.00 ×095) and 20/300 in the left eye (−8.00 +2.50 ×070). Manifest refraction yielded acuities of 20/100 in the right eye (−11.25 +2.00 ×095) and 20/50 in the left eye (−11.75 +3.50 ×070). Her VA varied between 1/200 and 20/100 in the right and 2/200 and 20/50 in the left by changing the test distances and target size. At near, she read Jaeger 1 at 10 inches binocularly but had no improvement with +1.50 sphere lenses in front of each eye. Stereoacuity on the Titmus test was 40 arc seconds, and Worth 4-dot testing showed fusion at both near and distance. Initial color vision testing using the Ishihara color plates revealed that 7 of 15 plates were correct in the right eye and 14 of 15 plates were correct in the left eye, which improved to 13 of 15 plates in each eye on repeat testing. Pupils were briskly reactive and there was no relative afferent pupillary defect, and the rest of the neuro-ophthalmological examination was normal. Confrontation VF testing was normal in each eye, whereas Goldmann perimetry revealed crossing of isopters in the right eye.

Interestingly, after our diagnosis of FVL, correspondence with her referring physician revealed that the patient's brother was a star athlete. The family acknowledged some sibling rivalry to be present. She followed up with this physician for another 18 months, during which time her VA continued to worsen and she was referred for repeat neuro-ophthalmolgical examination.

At this second evaluation, the patient stated that she saw better in dim light than in bright light. A history of blindness in her maternal grandfather was also elicited. Initial corrected VAs with contact lenses were 20/250 in the right eye and 20/100 in the left eye. Overrefraction with her contact lenses revealed VAs of 20/125 in the right eye (plano) and 20/50 in the left eye (−0.50 sphere). Testing with the potential acuity meter showed acuities of 20/200 in the right and 20/50 in the left. Stereoacuity was 70 arc seconds, and Worth 4-dot testing showed fusion at both distance and near. Color vision was abnormal in both eyes (0/14 correct in the right and 5/14 correct in the left) using the Ishihara color plates. After instillation of cycloplegic drops and with her cycloplegic refraction, VAs were 20/30 in the right and 20/50 in the left. However, due to her complaints of hemeralopia and the family history of legal blindness, eletroretinography was performed, which showed a consistent decrease in amplitudes under scotopic conditions. The 30-hertz flicker response also showed decreased amplitudes and delayed latencies consistent with a diagnosis of cone dystrophy.

Case 2. A 21-year-old female was referred for sudden painless sequential loss of vision in each eye 1 month apart. She initially presented with counting fingers vision in her left eye, with a questionable relative afferent pupillary defect and mild left disc swelling. She had been diagnosed and treated for optic neuritis, but magnetic resonance imaging of the brain, spinal fluid analysis, and a workup by a neurologist were normal. An immunologic workup was also negative, except for a positive antinuclear antibody to a titer of 1:80. Subsequently, her fellow eye experienced visual loss.

Neuro-ophthalmological examination by us 3 months after onset revealed uncorrected VAs of 20/400 in the right eye and 20/250 in the left eye. Binocular VA varied between 3/100 and 5/25 as her distance from the screen and target size was changed. She was unable to perceive any of the targets on the Jaeger card at any distance, even with +2.50 or +4.00 lenses in front of each eye. The pupils reacted briskly to light without a relative afferent pupillary defect. Visual field testing initially showed bilateral superior altitudinal defects but was normal when repeated and, at a later date, showed bilateral inferior altitudinal defects. Goldman perimetry showed spiraling. Slit-lamp examination was normal, and dilated fundus examination showed a normal right optic disc and questionable temporal pallor of the left disc. Cycloplegic retinoscopy showed minimal refractive error in both eyes. Fluorescein angiography was normal bilaterally.

An initial diagnosis of old optic neuritis in the left and functional overlay due to her inconsistent responses was made, although normal VA could not be proven. We repeated her antinuclear antibody, which was positive at a titer of 1:360. A repeat anti–double-stranded DNA titer was negative, and serum B_{12} and folate levels were normal.

Reexamination 1 week later showed improvement in each eye (20/160 in the right and 20/80 in the left). At near she read J10 at 10 inches binocularly. Electroretinography showed normal rod and cone responses, but visual evoked responses were abnormal bilaterally.

Neurological and hematological evaluations were normal. On follow-up 2 months later, VAs were 5/200 in the right and 20/400 in the left. Goldmann perimetry under good test-taking parameters demonstrated bilateral central scotomas. Genetic testing for Leber's hereditary optic neuropathy showed a mutation at the 14484 locus.

Case 3. An 11-year-old female suffered an episode of contact lens overwear and secondary bacterial keratitis, after which she complained of decreased vision in both eyes. She had consulted with several ophthalmologists and finally presented for neuro-ophthalmological evaluation 18 months later. Her mother said that she had never seen 20/20, and old records showed best-corrected VA in the 20/25 to 20/30 range bilaterally.

Initial corrected VA was 20/70 bilaterally, but her responses to repeat testing were very inconsistent, with VA ranging from 20/40 equivalent to 20/200. Near VA was 20/40 bilaterally. Color vision was abnormal by Ishihara (6/15 plates correct in both eyes), but she reported that she could not see the test plate in either eye. Stereoacuity was 200 arc seconds, and Worth 4-dot testing showed that she had fusion at both distance and near. The 4-diopter base-out prism test showed a normal refixation response in each eye while the 20/20 line was viewed. Confrontation VF testing was normal, but Humphrey perimetry revealed bilateral central scotomas, although she had many fixation losses in each eye. Goldmann perimetry revealed constriction of the I2e isopter bilaterally. Slit-lamp examination showed mild central stromal scarring bilaterally. Fundus examination revealed normal retinas, but there was mild temporal disc pallor in each eye.

She was suspected of having functional overlay due to the inconsistent responses. Magnetic resonance imaging of the anterior visual pathway, obtained because of the disc pallor, was normal. Subsequent retinal evaluation plus fluorescein angiography showed hyperfluorescent flecks in the macula and a dark choroid, consistent with a diagnosis of Stargardt's disease.

Discussion

A summary of various reported FVL studies is presented in Table 2. In the literature, there is a female preponderance in

Table 2. Summary of Published Reports on Functional Visual Loss

Authors	Age Group	Total	Female (%)	Male (%)	Trigger (%)	Underlying Psychiatric Problem (%)	Functional Overlay (%)	FVA (%)	FVF (%)	Follow-up (%)	Resolve (%)	Misdiagnosis (%)
Yasuna et al*	Adult + child	19	15 (79)	4 (21)	NI	NI	1 (5)	19 (100)	19 (100)	NI	NI	NI
	Adult	9	8 (89)	1 (11)	NI	NI	NI	NI	NI	NI	NI	NI
	Child	10	7 (70)	3 (30)	NI	NI	NI	NI	NI	NI	NI	NI
Schlaegel et al†	Adult + child	42	27 (64)	15 (36)	NI	NI	15 (36)	17 (40)	42 (100)	NI	NI	NI
	Adult	NI	NI	NI	NI	NI	NI	NI	NI	NI	NI	NI
	Child	NI	NI	NI	NI	NI	NI	NI	NI	NI	NI	NI
Friesen et al‡	Adult	11	NI	NI	NI	NI	1 (9)	NI	NI	11 (100)	5 (45)	NI
Krill et al§	Adult + child	59	35 (59)	24 (41)	16 (27)	6 (10)	6 (10)	48 (81)	55 (93)	NI	NI	2 (3)
	Adult	25	NI	NI	NI	NI	NI	NI	NI	NI	NI	NI
	Child	34	17 (50)	17 (50)	NI	NI	NI	NI	NI	NI	NI	NI
Behrman et al‖	Adult + child	14	12 (86)	2 (14)	NI	5 (36)	NI	14 (100)	14 (100)	10 (71)	5 (50)	NI
	Adult	3	2 (67)	1 (33)	NI	NI	NI	NI	NI	2 (66)	1 (50)	NI
	Child	11	10 (91)	1 (9)	NI	NI	NI	NI	NI	8 (73)	4 (50)	NI
Rada et al¶	Child	20	NI	NI	NI	NI	NI	NI	NI	18 (90)	11 (61)	NI
van Balen et al#	Child	31	24 (77)	7 (23)	NI	NI	NI	31 (100)	2 (5)	28 (90)	16 (57)	NI
Mantyjari**	Child	52	48 (92)	4 (8)	4 (8)	NI	NI	NI	NI	46 (88)	33 (71)	NI
Kathol et al††	Adult + child	42	33 (79)	9 (21)	NI	22 (52)	11 (26)	23 (55)	38 (90)	42 (100)	19 (45)	1 (2)
	Adult	32	28 (88)	4 (12)	NI	NI	NI	NI	NI	NI	NI	NI
	Child	8	5 (63)	3 (37)	NI	NI	NI	NI	NI	NI	NI	NI
Keltner et al‡‡	Adult + child	84	54 (64)	30 (36)	54 (64)	NI	45 (54)	61 (73)	43 (51)	32 (38)	3 (9)	NI
	Adult	59	36 (61)	23 (39)	29 (49)	NI	NI	NI	NI	NI	NI	NI
	Child	25	18 (72)	7 (28)	25 (100)	NI	NI	NI	NI	NI	NI	NI
Catalano et al§§	Child	23	16 (70)	7 (30)	21 (91)	NI	NI	23 (100)	NI	23 (100)	22 (96)	NI
Clarke et al‖‖	Child	54	38 (70)	16 (30)	NI	NI	NI	54 (100)	NI	NI	NI	NI
Present study	Adult + child	140	102 (74)	36 (26)	50 (36)	42 (30)	23 (17)	99 (72)	102 (74)	36 (26)	21 (58)	3 (2)
	Adult	82	62 (76)	20 (24)	26 (32)	32 (39)	11 (13)	NI	NI	23 (28)	10 (43)	NI
	Child	56	40 (71)	16 (29)	24 (43)	10 (18)	12 (21)	NI	NI	13 (23)	11 (85)	NI
Total		591										
Range (%)			59–92	8–41	8–91	10–52	5–54				9–96	2–3
No. of studies			13	13	5	4	7				9	3
Mean (%)			63	22	45	32	22				50	2

FVA = functional visual acuity loss; FVF = functional visual field loss; NI = no information.
*Yasuna ER. Hysterical amblyopia in children and young adults. Arch Ophthalmol 1951;45:70–76.
†Schlaegel TF, Quilala FV. Hysterical amblyopia. Arch Ophthalmol 1955;54:875–84.
‡Friesen H, Mann WA. Follow-up study of hysterical amblyopia. Am J Ophthalmol 1966;62:1106–15.
§Krill AE, Newell FW. The diagnosis of ocular conversion reation involving visual function. Arch Ophthalmol 1968;79:254–61.
‖Behrman J, Levy R. Neurophysiological studies on patients with hysterical disturbances of vision. J Psychosom Res 1970;14:187–94.
¶Rada RT, Krill AE, Meyer GG, et al. Visual conversion reaction in children. II. Follow-up. Psychosomatics 1973;14:271–6.
#van Balen ATM, Slijper FEM. Psychogenic amblyopia in children. J Pediatr Ophthalmol Strabismus 1978;15:164–7.
**Mantyjarvi MI. The amblyopic schoolgirl syndrome. J Pediatr Ophthalmol Strabismus 1981;18:30–3.
††Kathol RG, Cox TA, Corbett jj, et al. Functional visual loss: Follow-up of 42 cases. Arch Ophthal 1983;101:729–35.
‡‡Keltner JL, May WN, Johnson CA, et al. The California syndrome: Functional visual complaints with potential economic impact. Ophthalmology 1985;92:427–35.
§§Catalano RA, Simon JW, Krohel GB, et al. Functional visual loss in children. Ophthalmol 1986;93:385–90.
‖‖Clark WN, Bariciak M. Functional visual loss in children: A common problem with an easy solution. Can J Ophthalmol 1996;31:311–13.

this condition, both in children and adults (mean, 63% of all studies), as in our series.

In children, FVL is commonest in the prepubertal age group. In the literature, Mantyjarvi found a peak at 9 to 11 years of age,[1] and Clarke found a mean age of 10.2 years (mode, 9).[3] In our series, the mean age of our children was 13.4 years (mode, 17). This discrepancy can be explained as follows: many patients in the 16- to 18-year-old age range may see either an adult neuro-ophthalmologist or a pediatric ophthalmologist, leading to a skewed population if patients were seen by either. At our institution, we have both pediatric and adult neuro-ophthalmologists; thus, our study may have a better representation of the spectrum of patients with FVL. We have demonstrated that FVL is most common in 16- and 17-year-olds and is evenly spread out over the ages 20 through 50, declining thereafter.

In our experience, a minority of patients with FVL have a major psychiatric illness requiring treatment; <15% were on psychiatric medications, and only a small minority were referred for psychiatric or psychologic evaluation. An even smaller proportion of patients are involved in litigation (<10% of adults, and only one half of adults with physical trauma as an antecedent event). Nevertheless, one third of patients reported symptoms or feelings of stress, anxiety, or depression. This stress, anxiety, and depression (SADness) can originate from chronic pain syndromes (migraine and trigeminal neuralgia), trauma, and the other psychosocial stressors stated earlier. Despite this, only 10.9% of our

patients were referred for counseling, the remainder needing only reassurance.

We are the first to report the association of types of chronic pain syndromes and FVL. A significant number of patients were migraineurs (19.6% of children and 15.9% of adults). The majority of patients with migraine had associated visual aura (81.8% of children and 53.8% of adults). In addition, trigeminal neuralgia was also noted in 8 adults with FVL (9.8% of adults). In all cases of trigeminal neuralgia, the FVL was ipsilateral to the side of the pain. With respect to trauma, an association between closed head injury and FVL already has been noted. Sabates et al reported that 41.6% of their patients with closed head trauma had functional (tunnel) VFs.[4] This correlates with the results from our series, in which 47.6% of patients with physical trauma as an antecedent event had closed head injury.

It is not surprising that >30% of our patients reported underlying depression and/or anxiety. This anxiety may be worsened by physicians who cannot find an organic diagnosis but are unwilling to make a diagnosis of FVL. Subsequent anxiety or fear of the unknown only worsens the visual loss and leads to depression, which further perpetuates the cycle. This association is by no means new. Rovner et al report data in patients with macular degeneration suggesting that, as depressive symptoms increase over time, there is a corresponding decline in visual function independent of change in VA.[5] In addition, Casten et al report that 33% to 50% of patients who are blind have depression.[6] An important caveat is that of children, when neglect or abuse may lie behind the FVL; 3.6% of the children in our series had suffered sexual abuse.

The greatest concern in making a diagnosis of FVL is that of missing organic disease and withholding appropriate treatment. Of Krill and Newell's 59 patients, organic disease was later diagnosed in 2, one with macular degeneration and the other with unilateral optic neuropathy, presumably secondary to multiple sclerosis.[7] In Kathol et al's series, 1 patient with functional VFs was later diagnosed with normal-tension glaucoma (Table 2).[8]

Three patients were diagnosed with organic disease that was missed at initial consultation: cone dystrophy, Stargardt's disease, and Leber's hereditary optic neuropathy. All 3 patients were 21 years or younger. The important keys that eventually led to the correct diagnosis were that normal visual function could not be proven and the fact that their examination was not completely normal, although patient responses were inconsistent and suggested functional overlay. Egan has reported the importance of a central scotoma on VF as an indication that the visual loss is due to an organic pathology.[9] This is supported in our third example, where, although the VF testing was inconsistent, a central scotoma was detected on a Humphrey VF.

Retinal diseases are more easily missed in children and young adults because, early in the disease course, fundus findings may be absent or very minimal. Leber's hereditary optic neuropathy occurs in females, although rarely, and may spare pupillary fibers preferentially, even in the presence of severe visual loss.[10,11]

Intracranial lesions are less likely to be missed, as most ophthalmologists are aware of this possibility and many patients have undergone neuroradiological investigation even before they are referred to a neuro-ophthalmologist. However, Moster et al reported 2 patients with occipital lobe lesions who were initially diagnosed with FVL.[12] In these patients, neither computed tomography nor magnetic resonance imaging adequately demonstrated the occipital lobe pathology, but single proton emission tomography or positron emission tomography delineated the abnormalities. Hence, we propose that when normal vision is not demonstrated in the clinic, macular dystrophies, Leber's hereditary optic neuropathy, small occipital infarcts, retrobulbar optic neuropathies, paraneoplastic optic neuropathy or retinopathy, and acute zonal occult outer retinopathy should be considered in the appropriate setting.

Management of patients with FVL is crucial. Kathol et al report that reassurance alone was significantly more likely to result in recovery than the addition of nonspecific treatments like glasses or eyedrops.[13] Of the 4 patients in their series who received psychotherapy, none felt that it helped. Thompson writes that he is careful not to mix reassurance with pills, eyedrops, convergence exercises, or eyeglasses, as the patient may conclude that he or she may in fact have a problem so awful that the doctor feels the need to keep it from the patient.[14]

In our practice, once we are convinced that the visual loss is functional, we approach the patient by first telling him or her and family members that we see absolutely no reason in their eye examination to explain the visual loss. The entire examination is reviewed in a positive manner with the patient, particularly the absence of any brain tumor or blinding eye disease. We discuss in detail the toll that SADness takes on many people daily, as manifested by heart attacks, strokes, headaches, or gastrointestinal dysfunction, and explain that this same SADness may also cause mild to severe visual loss without their conscious awareness of it. Many patients respond that they understand the effect SADness has on health. At this point, we ask the patient if there are any areas of stress in his or her life that could contribute to their symptoms. It is very helpful if a family member close to the patient is present, as sometimes the patient will deny any problem but the family member will attest to pertinent underlying issues. Occasionally, it may even be necessary to speak to the patient and family member separately. Special care must be taken with the pediatric patient, with a targeted history and cognizance of the association between sexual abuse or trauma and FVL, and the physician must be prepared to contact appropriate authorities whenever such suspicion arises. The patient is told that the first step to healing is to identify problem(s) and try to resolve them; a pastor, counselor, social worker, etc. is suggested as a convenient or familiar resource that may be helpful. If any evidence of child abuse is suspected, appropriate authorities need to be notified. When the patient accepts that SADness can indeed be the cause of his or her problem, he or she can begin to regain the vision that stress and anxiety had stolen.

Limitations to this study are as follows: (1) retrospective nature of review, (2) lack of a gold standard for diagnosis of FVL, (3) lack of long-term follow-up to strengthen diagnosis of FVL (organic pathology may be recognized over time), and (4) ascertainment bias and limitations of self-reporting for

conditions like migraine and trigeminal neuralgia, depression, and presence of a concomitant psychosocial event.

In conclusion, FVL is a condition most prevalent in teenagers between 16 and 17 years of age and is 3 times more common in females. Only 25% of patients are on public assistance; thus, all socioeconomic groups are susceptible, and physicians should not discriminate on this basis. Underlying SADness is identifiable in one third of patients. In children, social or emotional issues at school or at home are the most frequent psychosocial association, whereas in adults, physical trauma is the main culprit. Care should be taken to identify victims of child abuse or neglect. The physician should be aware of these and inquire about them at the first patient encounter. Pain resulting from migraine and trigeminal neuralgia may also be associated with FVL. Management of this condition requires a sympathetic yet firm approach to the patient, explaining the effects that SADness has on visual loss and helping them to recognize the source of their fear. Compassion and reassurance are the mainstays of treatment, and nonspecific treatments are discouraged. Resolution occurred in over half of our patients and was more likely in children. The rate of misdiagnosis in a consultative neuro-ophthalmology practice is low (2.2%), and the patient will request a return visit more often than patients with purely functional disease. At that juncture, diagnoses to be considered include macular dystrophies and hereditary optic neuropathies, as shown here. However, small occipital infarcts, retrobulbar optic neuropathies (inflammatory or infectious), paraneoplastic syndromes (carcinoma-associated retinopathy, melanoma-associated retinopathy, optic neuropathy), and acute zonal occult outer retinopathy should also be considered.

References

1. Mantyjarvi MI. The amblyopic schoolgirl syndrome. J Pediatr Ophthalmol Strabismus 1981;18:30–3.
2. Schlaegel TF Jr, Quilala FV. Hysterical amblyopia: statistical analysis of forty-two cases found in a survey of eight hundred unselected eye patients at a state medical center. Arch Ophthalmol 1955;54:875–84.
3. Clarke WN, Noel LP, Bariciak M. Functional visual loss in children: a common problem with an easy solution. Can J Ophthalmol 1996;31:311–3.
4. Sabates NR, Gonce MA, Farris BK. Neuro-ophthalmological findings in closed head trauma. J Clin Neuroophthalmol 1991; 11:273–7.
5. Rovner BW, Casten RJ, Tasman WS. Effect of depression on vision function in age-related macular degeneration. Arch Ophthalmol 2002;120:1041–4.
6. Casten RJ, Rovner BW, Tasman WS. Age-related macular degeneration and depression: a review of recent research. Curr Opin Ophthalmol 2004;15:181–3.
7. Krill AE, Newell FW. The diagnosis of ocular conversion reaction involving visual function. Arch Ophthalmol 1968;79: 254–61.
8. Kathol RG, Cox TA, Corbett JJ, et al. Functional visual loss: II. Psychiatric aspects in 42 patients followed for 4 years. Psychol Med 1983;13:315–24.
9. Egan RA. Functional visual loss. Ophthalmol Clin North Am 2004;17:321–8, vi.
10. Wakakura M, Yokoe J. Evidence for preserved direct pupillary light response in Leber's hereditary optic neuropathy. Br J Ophthalmol 1995;79:442–6.
11. Nikoskelainen EK, Huoponen K, Juvonen V, et al. Ophthalmic findings in Leber hereditary optic neuropathy, with special reference to mtDNA mutations. Ophthalmology 1996; 103:504–14.
12. Moster ML, Galetta SL, Schatz NJ. Physiologic functional imaging in "functional" visual loss. Surv Ophthalmol 1996; 40:395–9.
13. Kathol RG, Cox TA, Corbett JJ, Thompson HS. Functional visual loss: follow-up of 42 cases. Arch Ophthalmol 1983; 101:729–35.
14. Thompson HS. Functional visual loss. Am J Ophthalmol 1985;100:209–13.