

Functional Visual Loss in an Israeli Pediatric Population

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ABSTRACT: **Background:** Pediatric functional visual loss (FVL) is the loss of vision in a child that cannot be explained by an organic pathology. In the last decade, only a few studies on pediatric FVL have reported long-term patient follow-up.

Objectives: To report the characteristics of pediatric FVL with long-term follow-up in Israeli children.

Methods: We conducted a retrospective chart review of the medical records of patients with FVL from 2000 to 2010. Only children with adequate follow-up (at least 2 months) were included.

Results: Of the 12 patients identified, 9 were females. Mean patient age was 10.5 ± 4.4 years (range 3.5–17 years). Most children (75%) had bilateral visual loss. One patient had a history of psychiatric illness and in three patients a preceding psychosocial event/trauma was identified. Brain imaging and electrophysiology testing (if done) were normal in all cases. No medications were prescribed to any of the patients, and all were reassured that there was a high chance of spontaneous resolution. The follow-up time was 2–108 months (mean 23.8 months, median 6). During the follow-up period 9 of the 12 had complete resolution and 2 had relief of symptoms. Three patients reported a recurrence of symptoms. No organic disease was ever diagnosed in this group.

Conclusions: FVL may occur in all age groups, including children. In cases of visual loss, it is usually bilateral and can involve both acuity and visual field loss. In the present report most of the patients experienced normalization or relief of their symptoms without medical treatment.

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KEY WORDS: functional visual loss, non-organic visual loss, malingering, conversion, visual field

Functional visual loss is the loss of vision that cannot be explained by a pathology of the eye or visual pathway [1]. This entity is not uncommon and can involve up to 5% of the patients in a general ophthalmology practice [2]. Several terms have been used to define this condition, including FVL, hysteria, hysterical visual loss, malingering, non-physiologic visual loss, factitious visual loss, non-organic visual loss,

FVL = functional visual loss

psychogenic visual loss, Munchausen syndrome, hypochondriasis, and conversion disorder of vision [3].

Practitioners often hesitate to 'tag' a patient as having FVL since some patients also manifest true organic disease. Moreover, previous studies have shown that up to 53% of FVL in adults and 25% in children occur in the setting of a concomitant organic disease [4,5], which makes it even more challenging to diagnose. Scott and Egan [6] found that central scotomata were always associated with organic pathology.

Pediatric FVL has been thought to be different from malingering, where there is a deliberate fabrication or exaggeration of symptoms in order to acquire "secondary gain" [7]. However, a desire to wear glasses or contact lenses may be considered a type of innocent secondary gain in children; in a series of 71 pediatric patients with FVL only 14% had no identified psychological/psychiatric cause for the complaint [8].

Several studies have shown that the cause of pediatric FVL is a major psychosocial/psychiatric disorder, such as amblyopia [9], conversion disorder [10], a neurotic conflict (children who wish to express hostility towards their parents) [11], prepubertal stress [12], school/family difficulties [13], and a stressful event [14]. However, in the last 10 years relatively few studies have had (essential) long-term patient follow-up [6,8,15-17].

In the present study we report our experience with pediatric FVL in Israeli children. We also briefly cover the major techniques for the office diagnosis of FVL.

PATIENTS AND METHODS

After approval of this study by the ethics committee of the Sheba Medical Center we reviewed retrospectively the charts of all pediatric patients with the diagnosis of FVL in the database of our hospital's ophthalmology department, who were examined between 2000 and 2010. FVL was defined as an afferent or efferent abnormality in the visual pathway that could not be explained after a detailed eye exam, visual field testing (Humphrey or Goldmann), neuroimaging, or electrophysiology. Additional workup was performed when necessary.

Exclusion criteria for this study included age older than 18 years, patients or families not consenting to investigation of organic causes of a disease, or a follow-up period of less than 2 months. The following data were collected:

- Patient demographics: age, gender
- Medical and psychiatric history
- Presence of psychosocial event: e.g., stressors at school, sexual abuse, physical trauma
- Type of FVL: visual acuity loss only, visual field loss only, or both
- Workup performed prior to diagnosis of FVL
- Follow-up: duration, rate of cure or relief of symptoms, and recurrences or normalization of symptoms.

RESULTS

Full data with appropriate follow-up were available for 12 patients. Patients' characteristics are summarized in Table 1. The follow-up period ranged from 2 months to 9 years, with a mean and median follow-up period of 23.8 months and 6 months, respectively. The study included 9 females (9/12, 75%). Mean patient age was 10.5 ± 4.4 years (range 3.5–17 years). In three cases the FVL was unilateral; all other patients complained of bilateral visual loss. One patient (#10) had both VA and VF loss (concentric deterioration that disappeared during follow-up).

One patient had a history of psychiatric illness (attention deficit hyperactivity disorder) and one patient was treated with acetazolamide after being diagnosed as having idiopathic intracranial hypertension, but during follow-up the diagnosis was proven wrong. Patient 11, who suffered from congenital glaucoma, had undergone bilateral trabeculotomy and subsequent bilateral trabeculectomy at the age of 3–6 months. His vision was stabilized at 6/60 in his right eye and 6/30 in his left eye until the age of 11 years. At this point his vision suddenly deteriorated to hand motions in the right eye and 1/60 in the left eye. He underwent extensive workup including several evaluations under anesthesia, imaging and electrophysiology testing, but no reason for this phenomenon was found. In three patients a preceding psychosocial event/trauma was identified. In one case (#5) the patient had been in a motor vehicle accident and had mildly banged her head against her sister. Another patient (#1) developed the visual disturbances after a bone marrow transplant due to thalassemia major. Patient 8 sustained a minor ocular trauma from a nail, with conjunctival laceration prior to referral. He was treated with prednisone for 5 days (for a reason unknown to the authors and no explanation could be found in his chart) and complained of a sharp deterioration in vision after the steroid drops were discontinued. None of the patients seemed to simply "want glasses."

Of the children who presented with decreased vision, at presentation the VA in the worse eye ranged from 6/6 to no perception of light. All children except one (#12) experi-

enced normalization or improvement in VA at the end of the first visit or during follow-up. Patient 12 is still being followed at our clinic but no improvement in VA has yet been documented for reasons unknown to us. Two patients had improvement in VA without complete normalization. In both cases VA at presentation was of no light perception that markedly improved to 6/40 and 6/20. Moreover, VA in both cases was fluctuating, even reaching 6/9 on some visits.

In four cases there was an associated complaint of headaches. In two cases there was a concomitant complaint of monocular and binocular diplopia, and in one case it was purely monocular. Ptosis and unspecified sensory lower limb disturbances were associated in one case and in another case there was a complaint of "ptosis," but physical examination revealed a decreased palpebral fissure rather than true ptosis. One patient suffered from blepharospasm. There were no other objective components in any of the patients after comprehensive clinical examination.

Humphry visual field was performed in nine cases and Goldmann visual field in three. In none of the cases was there a pattern of a central scotoma. Brain imaging was performed in six patients and interpreted as normal in all. In five cases electroretinography was undertaken and in four cases visual evoked potentials were recorded. All results were within normal limits.

Treatment in all patients consisted mainly of reassuring the patient and family. One patient, a 16½ year old girl, was warned that the presence of VA would probably prevent her from getting a drivers license (obtainable at age 17). No prescription for any drug was given. Two patients were referred for a psychological evaluation.

The follow-up period ranged from 2 months to 9 years. Nine patients had complete resolution of symptoms. Recurrence was documented in three cases. One patient had three recurrences 3, 4 and 7 years after the original episode; two other patients had recurrences after 3 and 7 years, respectively. No organic disease was ever diagnosed during the subsequent follow-up.

DISCUSSION

Functional visual loss is not a rare diagnosis in the pediatric population and obtaining the diagnosis is not a simple task. It combines a thorough eye examination and interview of both the child and the family. It is quite easy to miss the subtle signs of a child's or parents' discomfort and the diagnosis is often a lengthy process [15,18-20]. It takes a skilled interviewer to ask the appropriate questions (questions that are open-ended and specific yet non-offending). Once FVL is suspected the physician must document that the patient's vision is better than that reported. For the diagnosis of severe bilateral visual loss (hand motions or worse) there are simple office techniques:

VA = visual acuity
VVF = visual field

Table 1. Characteristics of Israeli children with functional visual loss (FVL)

| Age (yrs) | Gender | Laterality | Type | Preceding event | Other symptoms | Medical history | Workup | Treatment | Psychologist referral | Follow-up (mos) | VA presentation | VA end of follow-up | Course | Recurrence |
|-----------|--------|-------------------------|----------|-----------------|--|-----------------------------------|--|-----------------------|-----------------------|-----------------|-----------------|---------------------|-----------------------------|--------------------------|
| 1 | F | Bilateral | VA↓ | BMT | Diplopia (monocular+binocular) | Thalassemia | CT | Reassurance | Yes | 36 | BE 6/60 | BE 6/6 | Normalization | Yes (after 3 yrs) |
| 2 | F | Bilateral | VA↓ | None | Headache | None | None | Reassurance, warming* | No | 9 | BE 6/15 | BE 6/6.6 | Normalization | No |
| 3 | F | Bilateral | VA↓ | None | Diplopia (monocular+binocular) | None | None | Reassurance | No | 6 | BE 6/15 | BE 6/7.5 | Normalization | No |
| 4 | F | Bilateral | VA↓ | None | Headache | IIH [§] | CT, MRI, ERG | Reassurance | No | 10 | BE 6/90 | BE 6/7.5 | Normalization | No |
| 5 | F | Bilateral | VA↓ | Car accident | None | None | None | Reassurance | No | 2 | BE CF | BE 6/6 | Normalization | No |
| 6 | F | Bilateral | VA↓ | None | Headache | None | None | Reassurance | No | 108 | BE 6/15 | BE 6/6 | Normalization | Yes (after 7 yrs) |
| 7 | F | Unilateral | VA↓ | None | "Ptosis" [#] , sensory disturbance in lower limbs | Asthma, reflux, ADHD, tachycardia | MRI x3, MRA, laboratory for hypercoagulability | Reassurance | No | 87 | RE 6/6 | RE 6/4.5 | Normalization | Yes x3 (after 3,4,7 yrs) |
| 8 | M | Unilateral | VA↓ | Ocular trauma | Ptosis | None | ERG, VEP | Reassurance | No | 3 | LE NLP | LE 6/40 | Improvement | No |
| 9 | F | Bilateral | VA↓ | None | Headache | None | ERG, VEP, CT | reassurance | No | 4.5 | BE 6/15 | BE 6/7.5 | Normalization | No |
| 10 | F | Unilateral [°] | VA↓ +VF↓ | None | Monocular diplopia (fellow eye) | None | None | Reassurance | Yes | 6 | RE NLP | RE 6/20 | Improvement | No |
| 11 | M | Bilateral | VA↓ | None | Blepharospasm | Congenital glaucoma | EUA, MRI, ERG, VEP | Reassurance | No | 18 | RE HM LE 1/60 | RE 6/60 LE 6/30 | Normalization (to basic VA) | No |
| 12 | M | Bilateral | VA↓ | None | None | None | CT, MRI, VEP, ERG, OCT, NMO Ab, mDNA | Reassurance | No | 4 | BE 6/30 | BE 6/30 | No improvement | No |

*Patient was told that due to the VA she will not get a drivers license

[§]Diagnosis of idiopathic intracranial hypertension (IIH) uncertain (see text)

[#]Physical examination revealed decreased palpebral fissure but no actual ptosis

[°]Considered unilateral due to main complaint of decreased VA+VF. The monocular diplopia at the fellow eye was a minor transient symptom.

VA = visual acuity, BMT = bone marrow transplantation, MRI = magnetic resonance imaging, MRA = magnetic resonance angiogram, CF = counting fingers, HM = hand motion, BE = both eyes, RE = right eye, LE = left eye, CT = computed tomography, ERG = electroretinogram, IIH = idiopathic intracranial hypertension, VEP = visual evoked potentials, EUA = evaluation under anesthesia, OCT = optical coherence tomography, NMO Ab = neuromyelitis optica antibodies, mDNA = mitochondrial DNA, ADHD = attention deficit hyperactivity disorder, VF = visual field, NLP = no light perception

- **Observation of the patient's navigation in the examination lane and outside:** a truly blind patient will move slowly and try to feel objects while a FVL patient may deliberately bump into objects [21]
- **Achieving nystagmus:** using the optokinetic drum or tracking eye movement using a mirror
- **Proprioception test:** while a truly blind patient can touch their own nose with their finger a FVL patient might deliberately miss, thinking it is a test for vision.

In unilateral severe visual loss the examiner can occlude the better eye and use the above tests. For moderate monocular visual loss there is a need to separate the visual ability between the eyes using:

- **Fogging test:** on the good eye while both eyes are opened, making the patient unaware that he/she is actually able to see only with the bad eye
- **Red/green glass (duochrome test):** asking the patient to read the Snellen chart projected through a red/green filter while wearing red/green glasses with the red filter in front of the bad eye. If the patient reads the entire line including the green letters he is obviously also using the "bad" eye
- **Stereopsis:** since this function requires binocular vision it can prove some vision in the bad eye and there are charts for estimating VA on the basis of the number of stereo circles identified on the Titmus test [22]. Normal stereoacuity in the presence of very low VA was found to be the most reliable method for detecting non-organic visual loss in a large series of children [15].
- **Prism dissociation test:** a small vertical prism is used to create diplopia. The patient is asked to comment on the clarity of each image seen. The acknowledgment of diplopia and comments on the clarity of the dissociated images serve as testimony to the actual level of visual acuity in each eye [18].
- **Using the Snellen chart from the smallest letters and slowly increasing in size:** the patient usually "wears down" after several lines (even at the same size) and long silences [7].

In the pediatric population it is thought easier to prove better vision by using the methods above or even simpler approaches because this group of patients is more suggestible and less sophisticated. The examiner can use an anesthetic drop on the bad eye and convince the child that this is a "magic drop" that will temporarily make the eye see better, or using trial frame glasses with little or no refractive correction [21].

In our study the mean age of the patients was 10.5 years. This is compatible with other studies [8,12,15,18]. In a relatively large series of 56 children studied by Lim et al. [16] the mean age was somewhat older – 13.4 years. This discrepancy was explained by the presence of both pediat-

ric ophthalmologists and adult neuro-ophthalmologists in their institute, unlike other institutes where an older child (16–18 years) with FVL can be seen either by an adult neuro-ophthalmologist or by a pediatric ophthalmologist. In our institute there are both pediatric ophthalmologists and adult neuro-ophthalmologists, enabling a real representation of younger age distribution in our institute. Females outnumbered males 3:1. This finding is also compatible with previous studies [4,8,11,13,15-18].

As in other studies, there was a variety of complaints, including blurry vision, diplopia (monocular and binocular), ptosis, and unspecified sensory disturbances. Headaches were also common in our series (25%) and this finding is consistent with the literature [8,15,16]. Our youngest patient was a 3.5 year old girl who complained of blurry vision after mildly banging her head against her sister in a car accident. FVL was previously reported at this young age when a 3 year old girl complained of decreased VA after being reprimanded by her mother [23].

In our series only one-third of the patients had a predisposing factor, and in none of the cases was sexual or physical abuse involved. Furthermore, none of our patients expressed a desire to wear glasses. These data are different from the literature, where conflicts related to family, environment or school were quite common [8,13,15]. On the other hand, there are also studies with low rates of predisposing factors [16]. This also reflected the treatment offered, namely reassurance alone with only two patients referred for psychological assessment. We, of course, cannot exclude any missed predisposing factor; however, since all patients but one had normalization or amelioration of symptoms with reassurance alone and no psychological or social interventions, we feel that major stressors were not missed. It is also possible that children in Israel represent a different "milieu" of pediatric FVL who present without any predisposing factor (e.g., glasses or contact lenses are not appealing to Israeli children). However, due to our small group this conclusion should be taken judiciously.

One of our patients had idiopathic intracranial hypertension. A recent study by Ney and co-workers [1] showed that 6% of patients with this diagnosis had concurrent functional visual loss; however, since the study was conducted in adults and our patient's diagnosis was not definite, no conclusions can be drawn.

To conclude, children with FVL present a challenge to the ophthalmologist. First, a high level of suspicion is needed. Second, one should wisely use the wide range of available ancillary tests to reach the correct diagnosis. Third, one needs to be able to confront the patient and family and ask some basic questions to reveal the underlying cause for the symptoms (or to refer the child and his/her family to a more experienced professional). Finally, the physician should bear in mind that reassurance and an appropriate explanation are

often sufficient and there are probably no clinical parameters that can reliably predict the rate of recovery [15].

Our study suffers from being small and retrospective. Yet, we feel that a long follow-up of the patients, which is lacking in many similar studies, is crucial for understanding the correct prognosis of FVL in children. Moreover, in our series we found that a classical characteristic of FVL – a predisposing factor – was lacking. We cannot, however, rule out that such a factor was missed. We also suggest that reassurance alone is sufficient treatment in most instances, even in the long run, for children with FVL. Larger group analysis is suggested to reach further conclusions.

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