Instructions:

• Each speaker will prepare a syllabus that must be submitted through the online submission system.
• The length of the syllabus will be no shorter than 4 single spaced pages in essay (not point) format, plus references.
• Use single spaced, 11 point type and (if possible) Times New Roman font.
• When typing the text use word wrap, not hard returns to determine your lines.
• If headings and subheadings are used, these may be highlighted by using all caps and bold.
• Do not use the header or footer feature or endnotes in preparing the text.
• The submission must be submitted online.

Title:
Non-organic Visual Loss

Learning Objectives:

1. At the end of the session, the attendee will have gained knowledge about epidemiology of patients with non-organic visual loss.
2. At the end of the session, the attendee will have reviewed the common presentations of patients with non-organic loss of vision.
3. At the end of the session, the attendee will have gained knowledge on the psychological profile of patients with non-organic loss of vision.

CME Questions:

1. A non-organic cause of visual field loss can be ruled out if:
   a. Visual field defects are reproducible
   b. Reliability indices of the visual field are good
   c. a. + b.
   d. None of the above

2. Which visual field defect is generally not suggestive of a non-organic etiology?
   a. Homonymous defect
   b. Central scotoma
   c. Bitemporal defect

3. Which statement is not correct regarding non-organic loss of vision?
   a. Unilateral involvement is common in children
   b. Psychiatric disturbances are more frequent in adults
   c. There is a predominance of women in adults

Keywords (Max 5):

1. Non-organic
2. Loss of vision
3. Visual field
4. Visual acuity
5.
Introduction

The non-organic nature of signs and symptoms in some patients has been recognized for almost 4000 years by the ancient Egyptians. Hippocrates coined the term hysteria as he believed that the misplaced wandering of a uterus was responsible for such manifestations. Other terms were given by famous physicians like Jean-Martin Charcot (“La belle indifférence”), Sigmund Freud (conversion syndrome), or Jozef Jules François Félix Babinsky (pithiatism).

Homer’s description of a warrior who lost sight upon seeing an enemy may have been the first case of non-organic visual loss (NOVL). (Culpin) However, the first report of NOVL in the medical literature was written by Albrecht von Graefe in 1855 who described two techniques to demonstrate the non-organicity of visual loss in some patients. (Graefe)

Body

The true incidence of non-organic signs and symptoms is not known and varies depending on medical specialty. It represents 1-4% of all diagnoses in a general hospital and can be as high as 30% of neurological patients. Between the early twentieth century and its end no drastic change in the incidence of non-organic manifestations occurred and this was demonstrated in different countries, namely UK, Greece, and Switzerland. (Frei, Stefanis, Stephens) However, a change in the spectrum of clinical manifestations occurred, as a net decrease of the dramatic manifestations of hysteria was observed, whereas an increase of more minor manifestations was noted by these authors.

The incidence of NOVL was reported to be 5.25% amongst adults (Schlaegel) and 1.0-3.08% in the pediatric population. (Bain, Mantyjarvi, Munoz) Recent epidemiological studies are lacking and it is generally accepted that NOVL accounts for 1-5% of diagnoses amongst ophthalmological patients. (Kathol, Schlaegel) NOVL can occur in patients at any age, and from both sexes. For any age group there is a female predominance with even a higher proportion of girls in the younger group (≤ 14 years-old). (Behrman, Kathol, Keltner, Krill, Lim, Yasuna) Children tend to present more frequently with bilateral symmetric visual loss (Clarke, Catalano, Mouriaux, Toldo, Munoz) and adults can either present monocular or binocular loss of vision. (Weller)

Triggering factors may be present but are not always found. They include physical trauma, conflicts at school or home, environment stressors, prescription of glasses to a sibling or friend, or, rarely but more worrying, physical or sexual abuse. (Bain, Catalano, Kathol, Lim, Mouriaux, Rada) Mouriaux reported that a rather trivial precipitating factor could be disclosed in 64% of their pediatric patients. (Mouriaux) During periods of extreme stress, such as wars, there is an increased incidence of NOVL, usually manifesting as bilateral and severe loss of vision, resulting from either malingering or a true conversion syndrome. Such an example of NOVL during wartime is provided by Corporal Adolf Hitler who, at the end of the first World War, was able to avoid active duties claiming bilateral blindness after gas exposure. (Maranho-Filho)

The true prevalence of psychosocial disturbances in patients with NOVL is not known. Not only does it vary between adults and children, but it varies markedly from publication to publication. Psychological/psychiatric disturbances were present in 27% of children and significant home/school stress was uncovered in another 31%. (Taich) Sixty percent of children were reported to have social problems (school, family, exams). (Bain) Similarly, family problems, difficulty in school and mild psychological problems were prominent in a series of 58 children, but no serious psychiatric disorder was disclosed. (Toldo) The incidence of true psychiatric disorders, for which psychiatric management is necessary, is overall low, and lower in the pediatric group. Mantyjarvi reported that 8% of their cohort presented psychiatric disturbances. (Mantyjarvi) In Clarke’s experience and from a series of 54 children with NOVL, most were emotionally stable and did not have a specific psychologic conflict. (Clarke) Catalano believed that psychiatric referral was not necessary for children with NOVL, but underlined that the possibility of evidence of sexual or physical abuse should be sought in any case. (Catalano) In adults, Kathol et al reported no evidence of psychiatric syndrome or personality disorder in almost 50% of their cases, and advised psychiatric referral only if non-organic symptoms other than visual were detected. (Kathol) Lim et al reported an incidence of psychiatric problems in 18% of children, whereas the incidence was 39% in adults. (Lim)

The psychological mechanisms leading to NOVL may be psychogenic, unconscious, or deliberate and conscious. Patients with NOVL have been categorized by Thompson as ranging on a spectrum of “deliberate malingerers”, “worried impostors”, “impressionable exaggerators”, and “suggestible innocents”. NOVL in all
these patients result from a mixture of fraud and suggestibility, the extremes being mostly fraud (deliberate malingerer) and mostly suggestibility (suggestible innocent). (Thompson HS) As a general rule, true malingerers are more difficult to handle (as they are worried to be discovered as non-organic) and fortunately represent a minority of NOVL patients. However, depending on cultural background, countries, and socio-economical situations, the psychomechanisms of NOVL can vary. For example, in California, Keltner et al reported that 86% of 59 adults with NOVL were malingerers seeking financial gain, faking or exaggerating their symptoms consciously. Noteworthy, 56% of these 59 patients were unemployed. (Keltner) Coexisting organic disease is always a possibility and exhaustive investigations are necessary to rule out that possibility. Amongst published series, the frequency of functional overlay averages 22%, ranging from 5% to 54%. (Bain, Behrmann, Kathol, Keltner, Krill, Lim, Schlaegel, Scott, Yasuna) Underlying pathologies included amblyopia, keratoconus, congenital or acquired optic neuropathies, congenital or acquired retinopathies. (Lim) The fear of the physician is to mistakenly diagnose NOVL when a real pathology was responsible of the visual loss. Such a misdiagnosis carries potential medico-legal implications. The rate of misdiagnosis appears to be overall low (2-3%), most likely reflecting the caution that physicians adopt when facing a suspicion of NOVL. (Kathol, Krill, Lim) Occult retinopathies (early Stargardt’s disease, cone dystrophy) and hereditary optic neuropathies (Leber’s hereditary optic neuropathy) are the most frequent causes of misdiagnoses. (Lim)

3. Clinical presentation
NOVL can manifest as decreased visual acuity, altered visual field, or dyschromatopsia. Examination frequently reveals a combination of these. Around two-thirds of NOVL patients present a combination of decreased VA and altered VF; less frequently they present either isolated VA loss or VF loss. (Barris, Keltner) Dyschromatopsia is rarely a primary complaint in NOVL but abnormal color vision results can be found in up to 50% of cases. (Keltner, Toldo, Yamade)

3.1. Visual acuity loss
VA loss can vary from minimal to complete blindness and can affect one or both eyes. Sometimes discrepant results between near and distance VA are present in NOVL patients, hinting at the non-organic cause of visual loss. When visual loss affects only one eye and is profound, the situation is relatively easy to handle. More difficult is the presence of moderate and bilateral symmetrical loss of VA. Many tests can be used to diagnose NOVL, and it is not the purpose of this paper to review them all. Excellent reviews and textbooks provide an exhaustive list and explanations of these techniques. (Walsh & Hoyt) A selection of the tests frequently used in patients with either monocular or binocular non-organic loss of visual acuity is listed in the Table. Recently, Mojon and Flueckiger designed a very useful test for detection of NOVL. It is a pocket optotype chart in which the optotype minimal angle of resolution is independent of its size; patients with organic visual loss identify correctly all the optotypes whereas non-organic patients tend to stop at the large optotypes. (Mojon)

When VA is decreased, mERG, pattern ERG, EOG and pattern VEP are the most useful tests. Results of these tests are often considered as objective and reliable, but it is worth keeping in mind that results of VEPs can be altered by the patient, voluntarily or not. (Morgan, Uren) Normal VEP results are very helpful, but abnormal results are less reliable.

The past 15 years have seen the revolution and evolution of optical coherence tomography (OCT). With OCT, the physician can now appreciate in vivo whether the anatomy of either the retina or the optic nerve is intact. Measurement of the retinal nerve fiber layer thickness, assessment of the macular thickness, and, more recently, evaluation of the ganglion cell layer are invaluable tools for the investigation of NOVL.

The mimickers of non-organic loss of VA are the followings: keratoconus, amblyopia, subtle maculopathies, and early optic neuropathies. The following are the most common mimickers of NOVL with tips on how to diagnose them:

- Keratoconus: decreased VA, normal color vision, no RAPD. Normal OCT, abnormal corneal topography;
- Occult maculopathy: decreased VA, sometimes abnormal color vision, no RAPD. Possibly metamorphopsia, possibly abnormal VEP, abnormal macular OCT, abnormal pPERG or mERG;
- Early Leber’s hereditary optic neuropathy: decreased VA, abnormal color vision, absence of RAPD at the early stage, abnormal VEP, abnormal pPERG, normal mERG.

3.2. Visual field loss
Distinctive and various patterns of VF loss are present in NOVL. The patterns of VF loss can also vary according to the technique of perimetry that was used. Keane reviewed his own experience of 454 non-organic patients and found 142 patients with non-organic visual field loss. Whereas the majority (91/142) exhibited tubular fields, a surprising 36% displayed hemianopias. This high number of hysterical hemianopias was explained by the confrontational technique of counting fingers which encourage the patient to choose one side. On the other hand, centripetal testing (tangent screen, Goldmann perimetry) tends to produce constriction of
Nowadays, as most patients are examined with computerized static perimetry, the most frequent defect is a concentric loss of sensitivity. With manual kinetic perimetry, there is a bigger variety of VF loss, concentric loss being the most frequent presentation, but also spiral fields and intersecting isopters. With both perimetric techniques, other type of VF loss can be encountered either bilaterally, sometimes mimicking a neurological deficit (bitemporal or homonymous VF defects), or purely monocularly (monocular temporal, nasal or altitudinal VF loss). As a rule, the finding of either central or arcuate scotomas imply an organic disorder, and is not suggestive of non-organic pathology.

Computerized static perimetry is probably the worst technique to use in NOVL. Not only has it been demonstrated that any kind of VF defect can be faked by a subject but reproducible patterns of VF defects and reliable results can be obtained in NOVL. The physician needs to realize that “reproducible” and “reliable” do not mean “true”. When the patient presents with severe constriction of VF, a very useful technique is the confrontation VF examination. This simple technique can demonstrate non expansion of VF diameter with increasing examination distance or can disclose a much larger VF than by computerized perimetry. Kinetic Goldmann VF examination remains the best examination tool in experienced hands. Demonstrating to the patient that his/her kinetic manual VF is larger than its computerized static counterpart can be of great help, and usually contributes to reassure the patient on his actual VF. Microperimetry disclosed a larger VF than previously measured in one case. Microperimetry can potentially help to assess NOVL patients.

When VF is decreased, full-field ERG and multifocal VEP are the most useful electrophysiological tests. A diffuse retinopathy will produce a significant alteration of the full-field ERG. Multifocal VEP provides an objective mean of assessing the VF. By analyzing the cortical potentials, it is possible to demonstrate the preservation of VF despite apparent significant “loss” by subjective perimetry. Multifocal VEP offers a great potential for investigating NOVL. However, due to the lack of available commercial software, the technique of multifocal VEP is not widely used.

Sometimes NOVL is diagnosed in a patient harboring a real but subtle organic disorder. Hereafter are listed the most common mimickers of altered VF with tips on how to diagnose them:

- Vitamin A deficiency: history, decreased VA, dyschromatopsia, night-blindness, abnormal full-field ERG;
- RP sine pigmento: constricted VF, night-blindness, abnormal full-field ERG;
- Paraneoplastic retinopathy: constricted VF, night-blindness, photopsias, abnormal full-field ERG.

4. Visual prognosis - Recovery

Overall, the rate of improvement or full recovery of visual function varies from 40% to more than 90%. Better prognosis is achieved in the pediatric group with full recovery reaching more than 90% in three series.

5. Conclusions

Neuro-ophthalmologists are in a favorable position when confronted with a patient suspected of NOVL. The physician has knowledge of both the anatomy and the physiology of the visual pathways, whereas the rules governing this specific sensory system are usually not known by the patient. This provides the physician with a certain advantage when investigating NOVL. Further, the use of objective techniques such as electrophysiology and OCT certainly helps the physician to diagnose NOVL with more confidence. The diagnosis of NOVL is not easy and its consequences can be severe for the patient (this labeling will stick to the patient for several years). Facing a patient suspected of NOVL, the goals of the physician are the following:

a. It is necessary to exclude the possibility of any organic cause which could at least partially explain the loss of vision;
b. It is mandatory to demonstrate that the visual function of the patient is superior to what the patient claims;
c. The patient should be informed that his visual potential is unaltered and it should be possible to recover it;
d. Most patients with NOVL need to be reassured without any other therapy. However, some patients may benefit from temporary “external support” in order to help them recover. Psychiatric management remains episodic.
In order to achieve these goals, an empathetic approach is usually most effective; confronting the patient is rarely beneficial.

Table – Selection of tests that are useful for demonstrating the non-organic nature of VA loss

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<tr>
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<th>Unilateral VA loss</th>
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<tr>
<td>Prism test</td>
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<td>Pupils</td>
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<tr>
<td>*Red-green duochrome test</td>
<td>+</td>
<td>-</td>
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<tr>
<td>*Polarized test</td>
<td>+</td>
<td>-</td>
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<td>Rocking mirror test</td>
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<td>Optokinetic drum</td>
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<td>*Titmus stereoscopic test</td>
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<td>Mojon test</td>
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<td>Fogging</td>
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<td>Touch both indexes</td>
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<tr>
<td>Electrophysiology</td>
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*These tests are used with both eyes seeing simultaneously

CME Answers (Use lowercase letters if it’s an a/b/c option; feel free to include a description next to the correct answer):

1. d
2. b
3. a

References: (Author(s) Last Name separated by a Comma, Title/Article, Source (i.e. Journal Name), Volume #, Page #, Year)

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