



**UZ
LEUVEN**



Functional visual loss ?

Malingering ?

Prof. Dr. Werner SPILEERS

Definitions:

- 5% in general ophthalmological practice ?
- Functional visual loss; non-organic visual loss; conversion, somatisation, ...
- Malingering
- Aggravation: concurrently with organic illness

Functional visual loss (FVL)

- The patient is simulating poor vision (acuity, visual field), when in fact their vision is normal

ptosis, blepharospasm, diplopia, convergence spasm, voluntary nystagmus, convergence insufficiency are also possible symptoms

may be antecedent mild trauma, ...

(children: psychosocial stress, school, home, ...)

Non-organic visual loss

- Malingering:
a psychologically well patient who feigns visual loss
for some material benefit
with full awareness that the reported complaints are false
- Conversion disorder (“hysteria”):
often teenager or young adult with significant
psychological or social problems (school or work
stress, parental conflicts, ...)

Functional visual loss

- The intent is: ruling out of **organic** pathology
- Try and “trick” the patient into demonstrating normal visual function
- “*diagnosis by exclusion*” after extensive investigation is not as reliable
 - false positive: some patients can accurately and repeatedly “fake” abnormal testing
 - false negative: in genuine but early and subtle disease !
- Non-organic overlay !! In addition to genuine organic visual loss

Diseases often misdiagnosed as functional visual loss

- Pituitary tumors with early compressive signs
- Leber's hereditary optic neuropathy
- Bilateral retrochiasmal disease
- Early cone dystrophy or Stargardt's disease
- Retrobulbar optic neuropathies
- CAR, MAR, ...
- Small occipital infarcts

Unexplained visual loss

- BCVA
- Stenopeic vision: pinhole test
- Full eye examination: cornea, lens, tear film, ...
- Pupils: RAPD !!
- Colour vision
- Exclusion of amblyopia
- Eye fundus: macula: OCT, angiography
- Appropriate electrophysiology: VEP, ERG
- Visual fields of both eyes
- Neuro-imaging taking into account the clinical findings

Unexplained visual loss

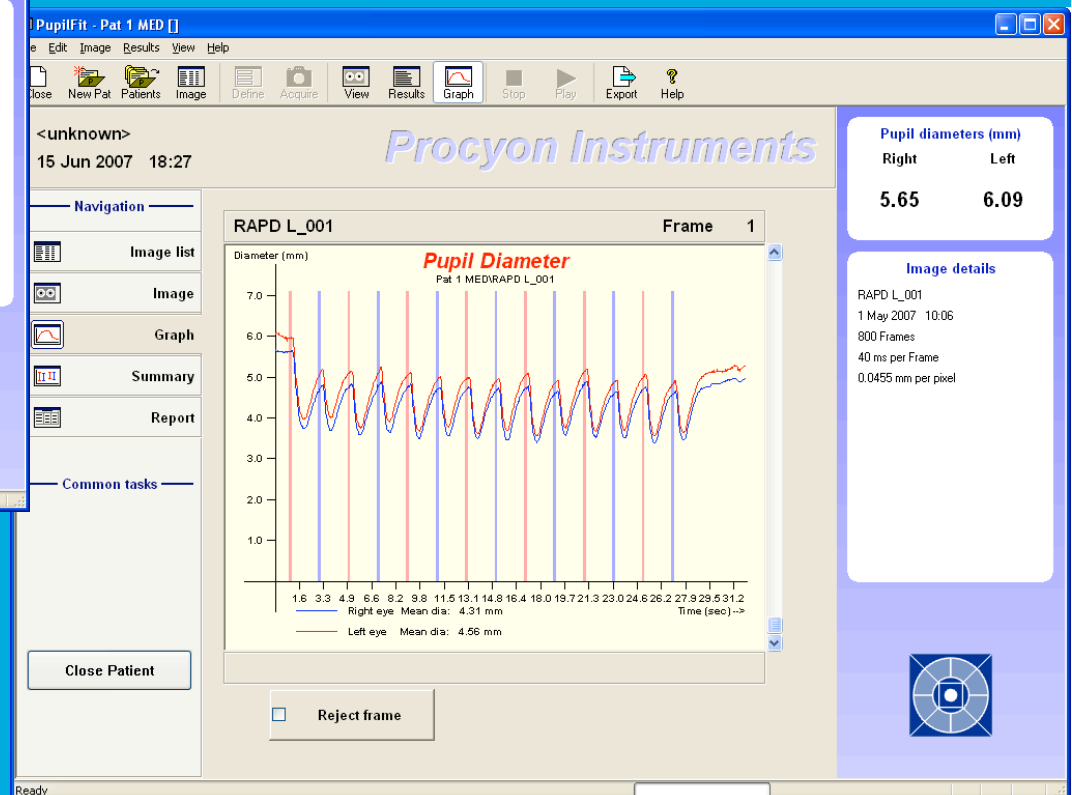
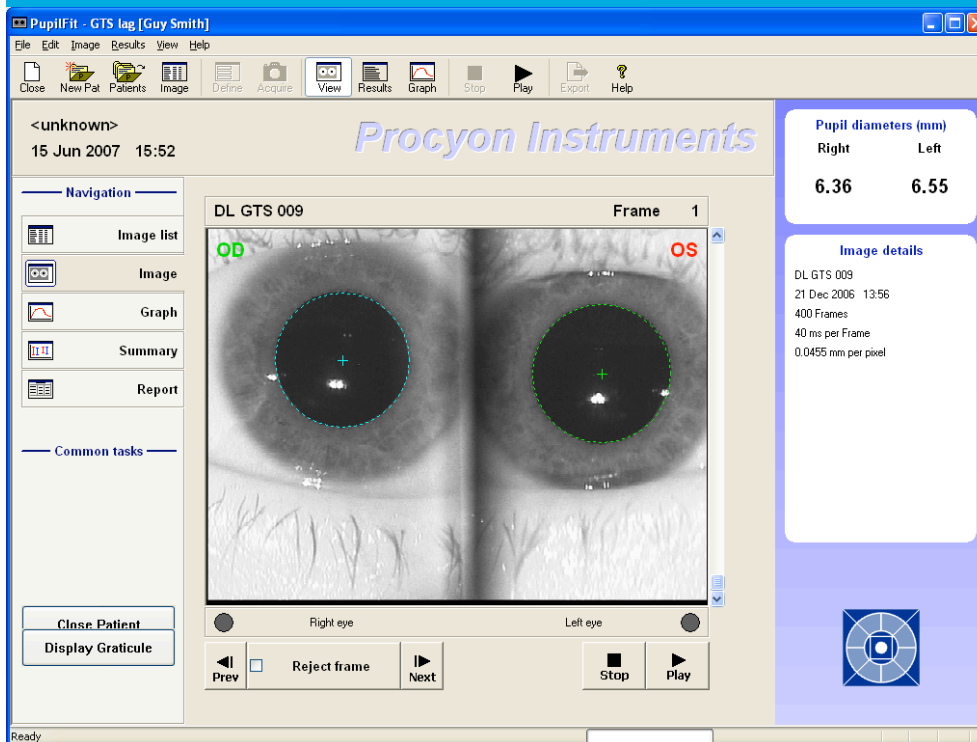
- First exclude organic visual loss !!
- Be aware of clues that a patient's visual loss may be non-organic:

RAPD ?!

Relative afferent pupillary defect

- Reliable and sensitive indicator of asymmetrical **optic nerve dysfunction**
- Absence of RAPD should prompt reevaluation of a working diagnosis of optic neuropathy or consideration of bilateral involvement
- A relatively small lesion of the optic nerve results in a large RAPD

Relative afferent pupillary defect



Relative afferent pupillary defect

- A retinal lesion must be substantially larger: retinal artery occlusion; widespread chorioretinal lesions; retinal detachment: R/ eye fundus examination !
- Chiasmal lesion if fibers of optic nerves are involved asymmetrically
- Optic tract lesion: mild RAPD in contralateral eye (= eye with temporal field loss) due to more crossed fibers

Relative afferent pupillary defect

- LGN, radiatio optica, cortical lesion: no RAPD
- An RAPD should never be attributed to media opacities only
- (amblyopia: only very mild RAPD possible, versus important acuity loss)
- NOT PRESENT in functional visual loss

Visual electrophysiology

- Pattern-VEP: denotes abnormality along the visual pathway BUT DOES NOT give the localisation of the deficit !!
- Flash-ERG: is a mass retinal response; ganglion cell activity does not contribute to the waveforms obtained

Perimetry

- Do not forget the manual perimeter of Goldmann !!
- (automated perimetry)

Clinical “tricks” to try and demonstrate normal vision

- Bilateral complete blindness (rare)
- Unilateral complete blindness
- Blurred vision in both eyes
- Blurred vision in one eye
- Constricted (altered) visual field in one or both eyes
- *The more severe and “one-eyed” the complaint, the easier it is to diagnose FVL*

Bilateral complete blindness (rare)

- Pupils:

bilateral blindness due to severe bilateral retinal, optic nerve or chiasmal disease: both pupils “sluggishly” reactive to light

if both pupils are briskly reactive to light, the only cause for true bilateral blindness is bilateral retrochiasmal disease

- Observation:

in the waiting room; walking out of the hospital, hand shake, ...

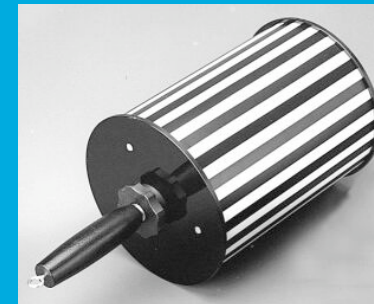
Bilateral complete blindness

- Tests that are independent of vision:
proprioception



(signature test)

- Optokinetic nystagmus
- (Mirror test)



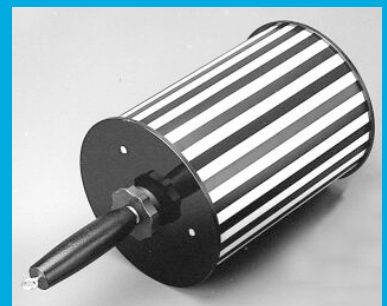
Bilateral complete blindness

- Optokinetic nystagmus:

can be generated in eyes with acuity of count fingers as long as a degree of visual field remains

suppressing the reflex is difficult (but possible)

unquantifiable nature of the response



Unilateral complete blindness

- Pupils: RAPD !
- (Mirror test)
- OKN
- “fogging” or “crossed cylinder technique” with trial frame
- Stereopsis



Figure 8. Titmus Fly Stereotest

requires binocularity

“fogging” or “crossed cylinder technique” with trial frame

- Trial frame
- +4 cyl and -4 cyl on the same axis for both eyes: neutral
- Read the VA chart
- Slowly rotate one of the cyls in the “good” eye = blurring the “good” eye
- Patient may close one eye to check what is going on

Stereotest



Figure 8. Titmus Fly Stereotest

Table 1

The degree of stereopsis in arc seconds may be converted into visual acuity¹⁹

Stereopsis (arc second)	Visual acuity
40	20/20
52	20/30
60	20/40
78	20/50
94	20/70
124	20/100
160	20/200

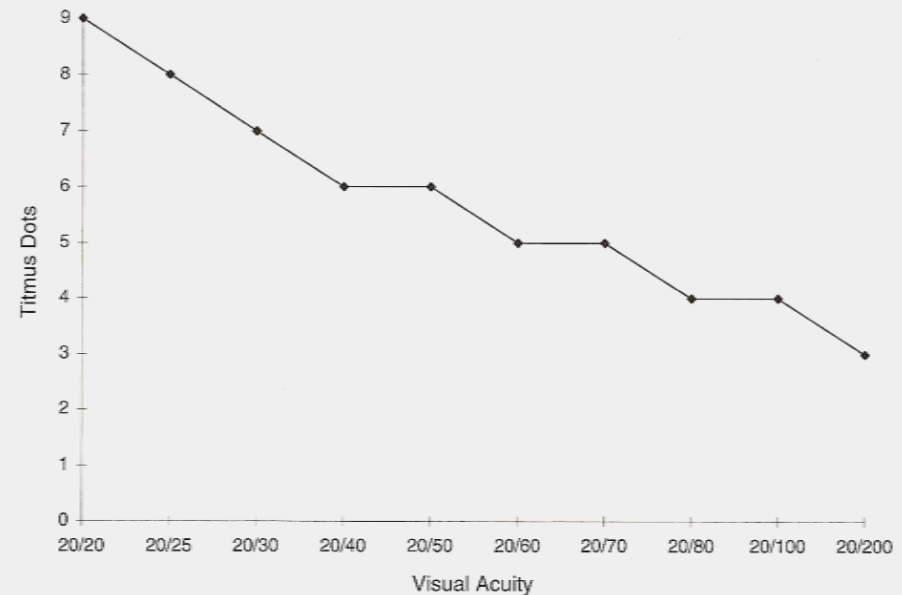


FIGURE 11-2. Correlation of level of stereoacuity with visual acuity. Individuals who can perceive the eighth of nine Titmus stereo dots must have at least 20/25 acuity in each eye. Similarly, the ability to perceive the first six of nine stereo dots correctly requires at least 20/50 in each eye. (Adapted from Levy NS, Glick EB: Stereoscopic perception and Snellen visual acuity, Am J Ophthalmol 1974;78:722-724, with permission from Elsevier Science.)

Prism shift test

- 10 D base-out prism in front of the “blind” eye
- If normal binocular vision: movement of both eyes towards the apex of the prism, followed by a shift of both eyes back to the centre

Diplopia test

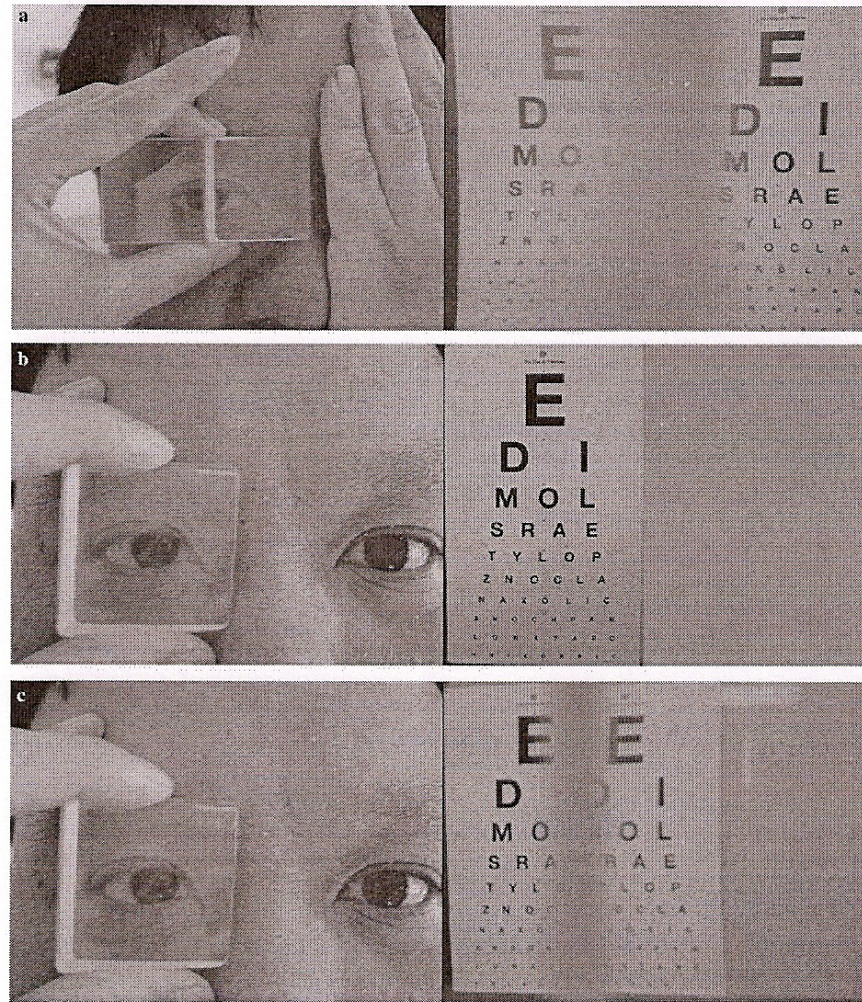


Fig. 4. The diplopia test.²⁰ (a) The “blind eye” is occluded and a strong prism is placed over the “good eye” to produce monocular diplopia. (b) The prism is then placed over the good eye. If only the good eye is seeing, it will see a single displaced image. (c) If both eyes are seeing, there will be two images, one from the displaced image of the “good eye”, the second image from the alleged “blind eye”. Hence if the patient admits to diplopia, he/she is seeing out of both eyes.

Blurred vision in both eyes

- Testing visual acuity at different distances: no improvement at closer distances is inconsistent with organic visual loss
- Reading vision and distance vision
- Ishihara plate color testing: test plate
- Perimetry ! Cave: small central scotoma missed on 30-2/24-2: also perform 10-2

Blurred vision in one eye

- Pupils
- “fogging” or “crossed cylinder technique” with trial frame
- Stereopsis
- Monocular vertical prism dissociation test

Monocular vertical prism dissociation test

- “The good eye is being tested”
- Look at acuity chart with both eyes open
- 4 diopter prism is placed base down in front of the “good” eye
- “What do you see?”
 - *Normal: two equally distinct rows*
 - *Genuine VA loss: one row (or the lower image is less distinct)*
 - *FVL: two equally distinct rows*

Constricted visual field in one or both eyes

- Manual perimeter of Goldmann !

Constricted visual field

Spiraling

Crossing of isopters

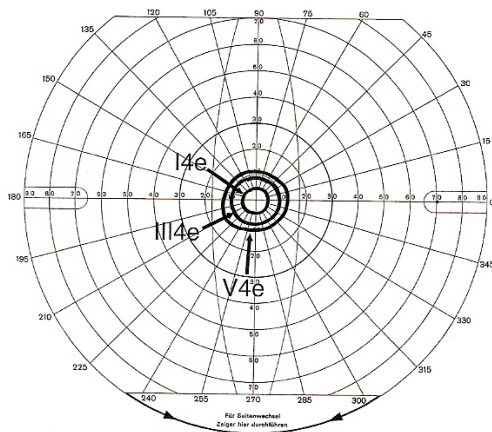


FIGURE 11-4. Goldmann visual field demonstrating generalized constriction. Note that each stimulus is associated with approximately the same size visual field.

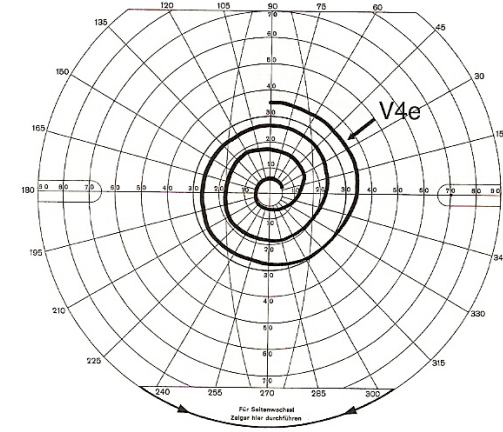


FIGURE 11-7. Goldmann visual field demonstrating non-physiologic spiraling. As the test proceeds along adjacent radii, the patient responds later and later. The field “spirals” to the center.

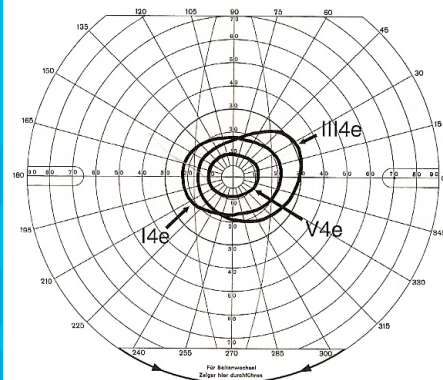


FIGURE 11-6. Goldmann visual field demonstrating “criss-crossing” of isopters. Note that there are parts of the visual field that are larger when testing with the smaller III4e stimulus compared to the larger V4e stimulus, and the I4e field is larger than the V4e field. This is nonphysiologic.

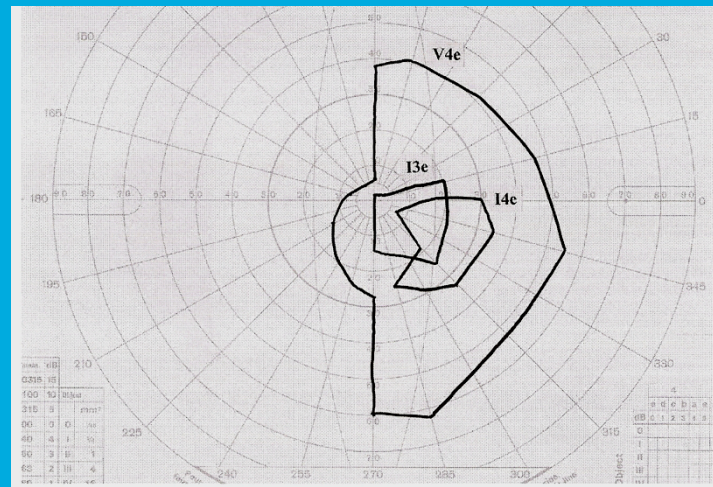


FIGURE 1. Goldmann visual field of the left eye showing nonphysiologic crossing of isopters. There is also a temporal hemianopia in that eye. Fixation was excellent during testing. Note that the I4e isopter actually falls inside the area of the I3e isopter at several points.

Constricted visual field in one or both eyes

- Tangent screen visual field at one and two meters

The size of the VF should expand at the 2 meter distance

in functional VF constriction, it is frequent to see the VF remain the same size or actually shrink

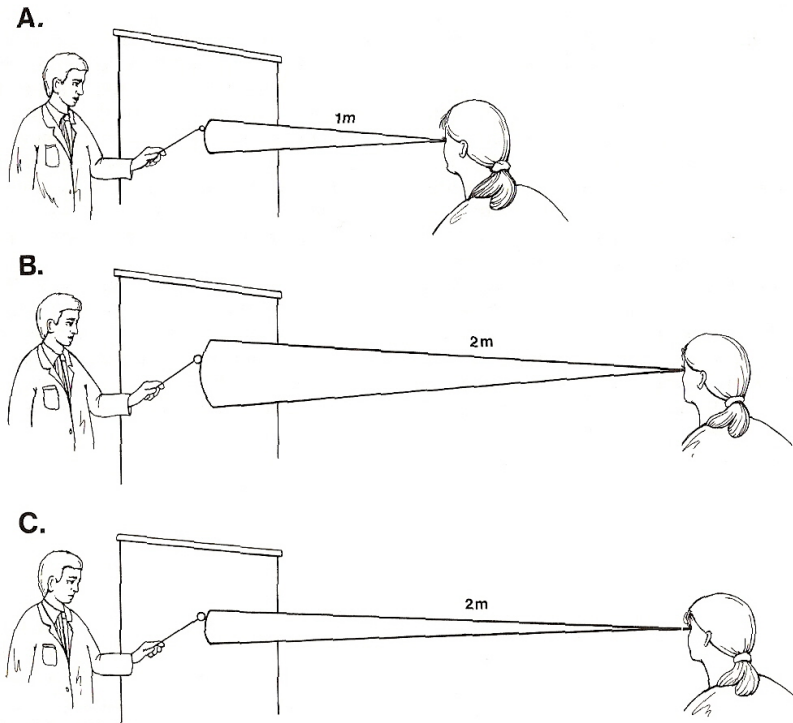
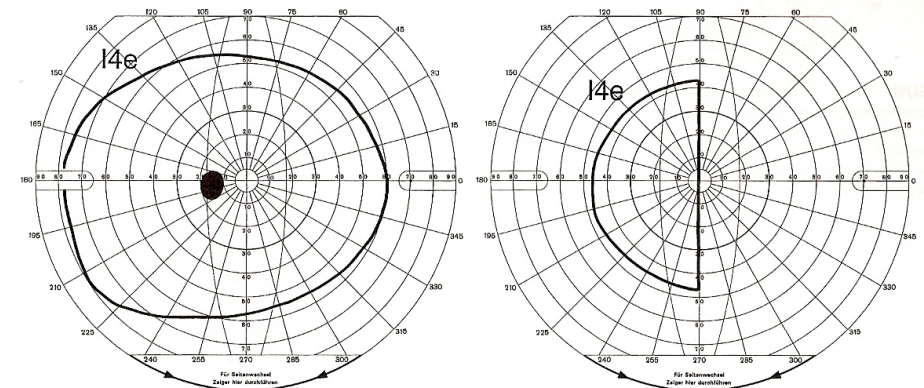
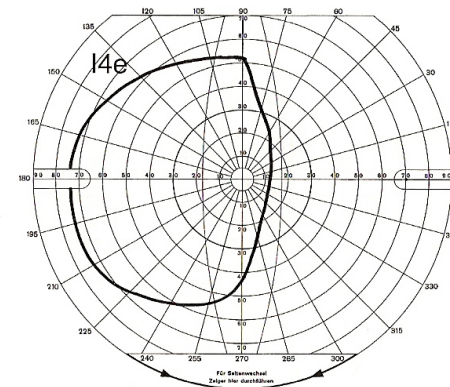


FIGURE 11-5. Testing for tubular visual fields with the tangent screen. **A.** The field is tested at one meter with a 9-mm white object and the results marked on the screen with chalk. **B.** When the patient with organic visual field loss is moved to 2 meters from the screen and the stimulus sized doubled (18-mm white) the field expands to twice the size. **C.** The patient with functional visual loss demonstrates tubular fields as the field does not expand as the testing distance is increased from 1 to 2 meters and the target size is doubled.



A Left eye

B Right eye



B Both eyes

FIGURE 11-8. Testing for nonorganic temporal visual field defect using binocular Goldmann visual fields. The patient complains of temporal field loss in the right eye, which is documented in **A**. Note the visual field of the left eye is normal. **B.** The patient is then tested binocularly and believes that the right side of the visual field is seen only by the right eye. Therefore, a persistent temporal defect is present despite the presence of intact right-sided (nasal) visual field in the left eye.

Central visual field

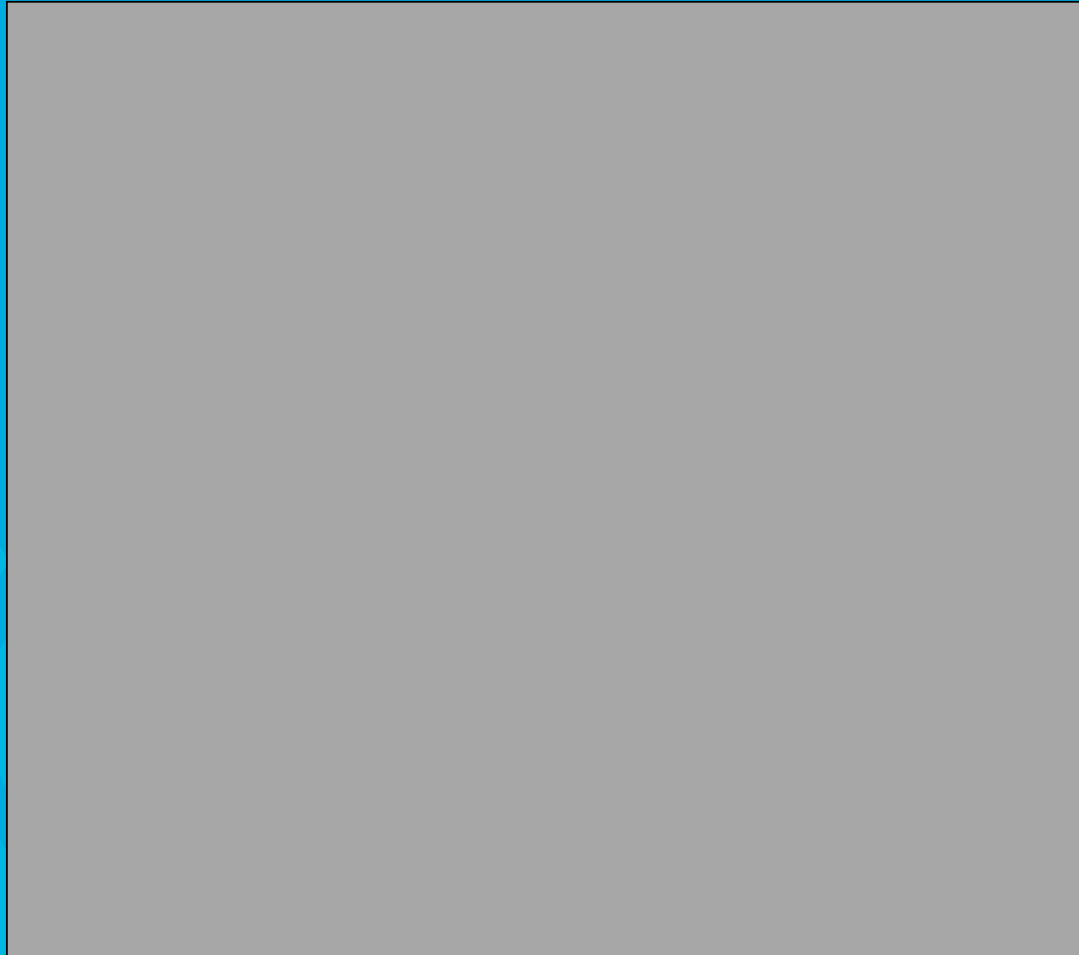
- Central scotoma:

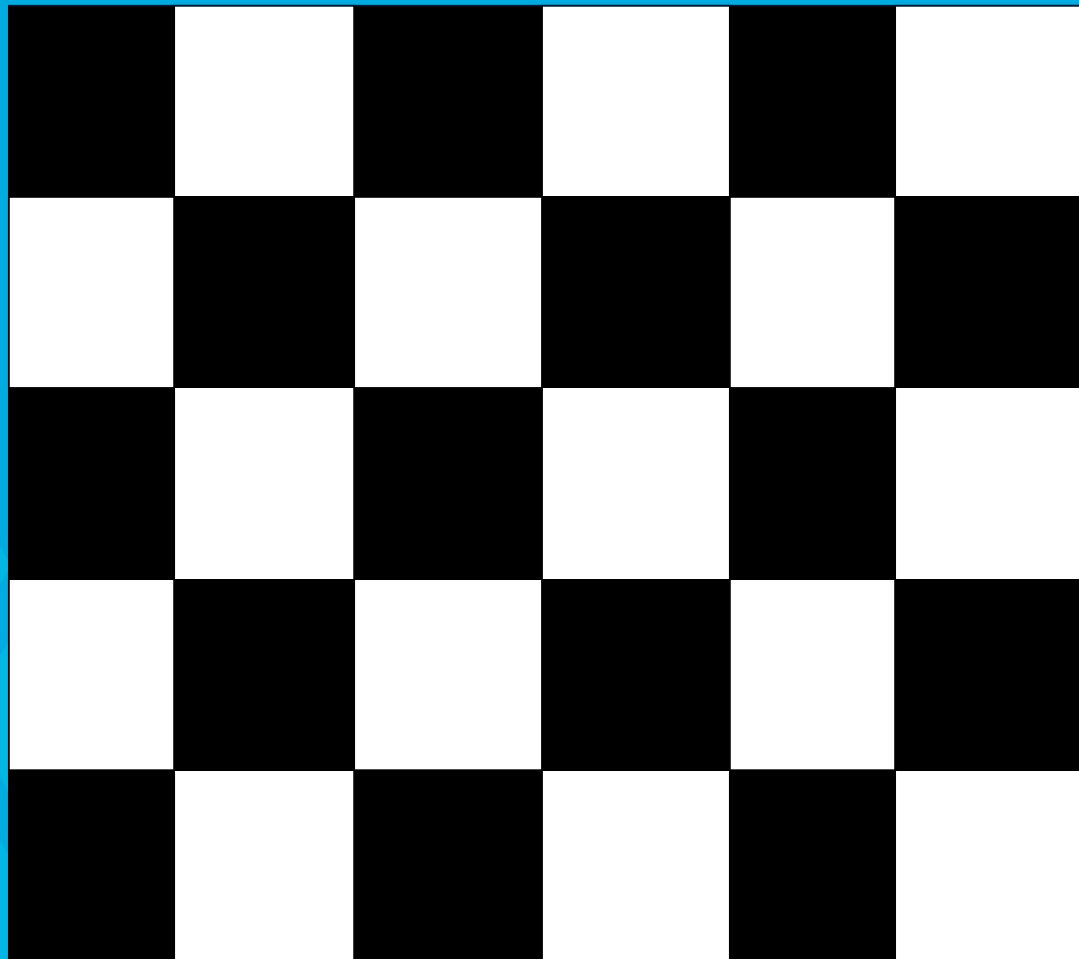
perform careful fundoscopy, OCT, angiography, ERG to rule out subtle maculopathy

perform neuroimaging: most patients with central scotoma on VF testing, have organic pathology !

Pattern-VEP and « objective visual acuity measurement »

- Holder et al., Graefe's Arch Clin Exp Ophthalmol, 2007
- Pattern *appearance-disappearance* 40ms/500 ms
- Different check sizes at different contrast levels
- Minimum check size and contrast level required to elicit a reproducible response of $\geq 5 \mu\text{V}$





Holder et al, 2007

Table 1 A summary table to be used as a quantitative guideline for the estimation of VA in the patients referred with suspected NOVL

PappVep (checksize/ contrast)	Median Snellen VA	Interquartile range
5.5'/20	6/6	[6/5–6/6]
5.5'/40	6/9	[6/6–6/12]
5.5'/80	6/18	[6/12–6/18]
11'/40	6/24	[6/18–6/36]
11'/80	6/36	[6/36–6/60]
22'/40	6/60	[6/36–6/60]
22'/80	6/60	[6/60–3/60]
44'/40	6/60	[6/60–3/60]
44'/80	3/60	[6/60–3/60]

The median Snellen VA is given for the PappVEP response from both the normal subjects and patients with confirmed organic eye disease. Interquartile Snellen acuity range is defined according to the values either side of the median visual acuity

Suspected but unproven functional visual loss

- CAVE: real visual pathway disease being misdiagnosed as non-organic
- Follow-up until you can demonstrate either organic or non-organic disease

Patient with “proven” functional visual loss

- Reassurance
- “good chance to recover” ...
- Leave a way out ...
- Personalize your strategy to the particular patient