New Concepts and Names of Visual System in Early Intervention and Rehabilitation of Infants and Children

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Abstract

Awareness of vision problems in infants and children with other disabling conditions has meant rapid increase in the number of children with impaired vision, assessed as visual acuity and the size of the visual field, and even greater increase in the number of children with atypical, disabling visual functioning due to brain damage. Therefore we need further training in assessment of all visual pathways and vision related functions for the evaluation of visual functioning.

Discussion: The changes in the different parts of the visual system are so many that the information for clinical and educational assessment has to be gathered from the parents, all medical and optometric specialists, therapists and teachers for a thorough evaluation. To structure the questions, schools and medical (re)habilitation teams have created long lists of functions that should be observed and assessed in different conditions so that a *Profile of Visual Functioning*[1] can be described for each child.

Conclusion: *Transdisciplinary approach* in the assessment of visual functions and functioning requires development of a common language and approach in the assessment so that each child's needs in early intervention and education are met early and then regularly as a normal part of medical care and education. The Profile of Visual Functioning is instrumental also in the exchange of information between the teams in a well-structured way.

Keywords: Assessment of Impaired Vision, Profile of Visual Functioning, ICF-CY, CVI. Transdisciplinary Approach

Rehabilitation centers have been aware of vision problems in infants and children with motor or intellectual disability and different syndromes since 1970s, yet early diagnosis of impaired vision is not well developed in many countries and early intervention is not yet an integral part of treatment of eye disorders or vision problems related to brain damage. This paper covers, as a short overview, the varying atypical visual functions related to changes in the *eyes, optic pathways to the lateral geniculate nucleus and the tectal pathways, the optical radiation, visual cortices in the occipital lobe of the brain and in the higher associative cortices* that combine vision with other sensory information, motor functions, and memory. In each case we should ask ourselves whether we have covered all parts of the visual system. This is particularly important in infants who have retinopathy of prematurity (ROP) and mild or no motor problems, in all infants with difficulties in early visual communication, in all *hypotonic infants* and children, infants and children with delays in cognitive development, and children with autistic-like behaviors.

Visual pathways

Visual information can become distorted in 1) the eyes due to corneal changes, cloudiness of the lens or vitreous, and in the complex retinal functions, 2) in the long pathways that transfer the information but may also change it by filtering, and/or 3) in the brain during the final processing of visual information (Fig.1).

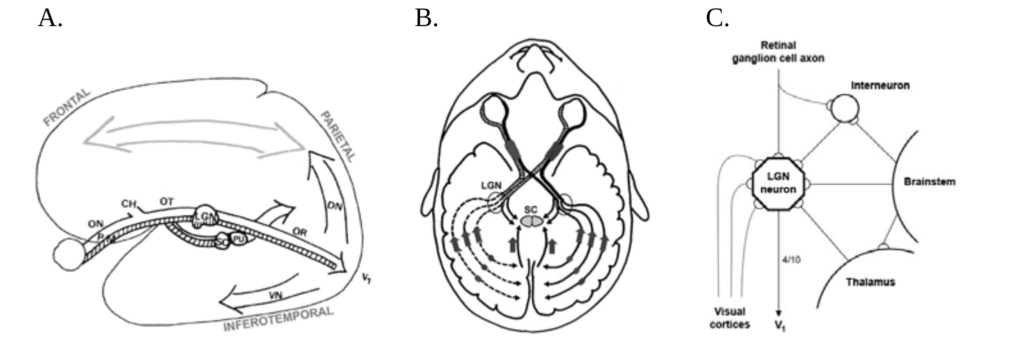

Figure 1. Visual Pathways. A. Main structures of the retinocalcarine pathway are: the eye, optic nerve (ON), optic chiasm (CH), optic tract (OT), lateral geniculate nucleus (LGN), optic radiation (OR), and primary visual cortex (V_1). The letter P stands for parvocellular and the letter M for magnocellular pathway. The tectal pathway leaves the optic tract just before the LGN and transfers information via the superior colliculus (SC) to pulvinar (PU) and to the parietal lobe. The big arrow between parietal and frontal lobes depicts the role of executive functions. B. The retinocortical pathway transfers information through the lateral geniculate body (LGN) to the primary visual cortex. Note that in optic radiation information flows toward the primary visual cortex (small arrows) and at the same time there is a greater flow from the primary visual cortex toward the LGN (large arrows).C. This diagram stresses the importance of the LGN in the selection of only a part of visual information for further transfer to the primary visual cortex.

The two parallel visual pathways make it possible that certain areas of the brain may get normal visual information via the tectal pathway when the main pathway is damaged in its posterior part. The two main directions of the flow of the visual information from the occipital cortex are called the dorsal networks toward the posterior parietal lobe where we have visual functions related to orientation in space and eye–hand coordination and the ventral networks toward the inferior temporal lobe where we have numerous recognition related functions (Fig.1).

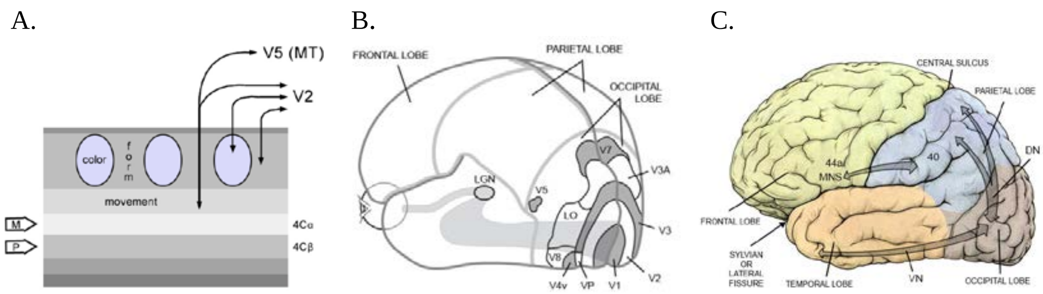

Figure 2. A. In the primary visual cortex (V_1) color information, contrast edges, lines and other form related information are coded in specific groups of cells, motion information in a separate layer. B. The primary visual cortex (V_1) and other visual cortices in the occipital lobe. **C.** The cortical visual networks, the ventral network in the inferotemporal area, the dorsal network in the posterior parietal area, and the mirror neuron system (MNS) are depicted here on the surface of the brain. *Note that the arrows depicting the flow of information are larger toward the occipital lobe (top-down) than from it.*

The important features to know are 1) the parallel visual pathways make it possible for some parts of the brain to receive visual information when other parts are damaged. This is especially important in damage to the optic radiation that causes hemianopia or hemiamblyopia if motion information is available via the tectal pathway because attention to the hemiplegic side can and should be activated right from the start[2]. 2) The brain cortex has numerous specialized areas for handling specific parts of visual information and associative functions that combine visual and other types of sensory information with motor functions. All these functions are affected by *attention* and complex *executive functions* in the frontal lobe (Fig.1.A).

For the assessment we consider 1) eyes as the receiver of visual information, the camera part; 2) the long pathways as a part of active processing, and 3) the brain as the structure processing visual information. Visual processing in the brain begins in the occipital lobe as the Early Processing and continues in the numerous functions of the Ventral and Dorsal Networks, and the Mirror Neuron System.

The transdisciplinary assessment has three parts:

- Assessment of ocular motor functions
- Assessment of sensory functions
- Assessment of visual processing functions

A transdisciplinary assessment should always be completed with evaluation of all compensatory sensory functions, especially hearing, tactile and haptic information, and memory functions as well as gross and fine motor functions.

Thorough clinical examination is the foundation for a comprehensive functional assessment by the (re)habilitation team and the educational team, often working together to find a solution in the more complex cases. Ocular motor functions that should be reported by the doctors are: *fixation to penlight and at details of small pictures, tracking, saccades, convergence, miosis during convergence, accommodation, and nystagmus.* Clinical assessment of visual sensory functions is often limited to measurement of distance visual acuity (with single optotypes only) and the other important measurements are forgotten:

- recognition acuity at close distances with single optotypes, line tests and more crowded line tests,
- visual acuity at low contrast levels,
- grating acuity at full and low contrast levels,
- reading acuity,
- visual field,
- color vision,
- visual adaptation and
- motion perception.

Most of these clinical tests can and should also be *used at kindergarten and school* so that the results of clinical examinations can be understood and compared with the measurements by the educational team.

If a child has *medications* that affect wakefulness/alertness, all measurements should record the time of the day. Results of all these sensory tests depict brain functions that may be *modified by changes in the anterior part of the visual system* or also by *posture or communication problems*. The tester should also know the limitations of tests, for example, *if contrast sensitivity is low then low contrast color vision screening tests are likely to give wrong results*. An anomalous finding in a color vision screening test should lead to assessment with a quantitative sorting test.

Recognition acuity tests and grating acuity tests revealing different problems
In normally sighted people recognition acuity and grating acuity measurements result in values that are closely related. From the measurements of contrast sensitivity *in visually impaired clients with macular degenerations we learned in late 1970s that grating acuity value usually was higher than recognition visual acuity (optotype acuity)*, and that it was defined by the size of the grating stimulus. *In an impaired visual system these two values can be widely apart*. At the first Finnish Vision Rehabilitation Center at the Finnish Federation of Visually Impaired we experienced this in 1977 when a 6-year old lively boy did not recognize the 30M LH-optotypes at 30 cm distance, except the “ring” that he seemed to trace with eye movements. His recognition acuity was thus less than 0.01 (20/2000).Yet at the same distance he had no difficulty in discriminating thin lines corresponding to 4 or 5 cpd (Fig. 3A). This finding was so perplexing that the boy was examined also in a vision research laboratory at the Department of Psychology of the University of Helsinki where the results were confirmed: the boy had close to normal contrast sensitivity at very low spatial frequencies but the slope came down at 4 cpd (Fig.3B). The grating acuity value was thus 4 cpd, which was ten times higher than the visual acuity of barely 0.01. Such a visual acuity would usually lead to classification “profound visual impairment” or “near blindness” but this boy moved clearly based on visual information, recognized large landmarks and even persons.

A. B. C.

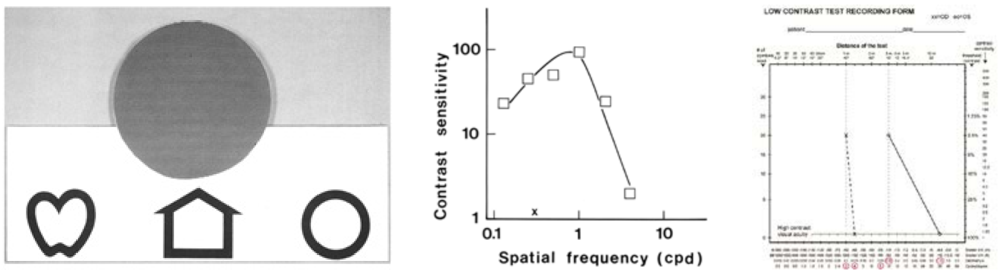

Figure 3. A. Recognition acuity was not measurable, yet grating acuity measured with fine lines was 4 cpd. B. Measurement of contrast sensitivity curve using computer generated grating stimuli showed nearly normal function at low spatial frequencies (0.1 to 1 cpd) and a slope ending at 4 cpd. C. In this case recognition acuity is normal (1.25) at 90% contrast and 0.32 at 2.5% contrast whereas the lines in the grating test at 90% contrast were seen only in its periphery at a distance corresponding to 8 cpd and as straight lines also in the center at a distance corresponding to 4 cpd; the 2.5% grating was seen at 3cpd.

A few months after the assessment of the boy, the first news were heard about monkeys that had visual functions after their visual cortices had been carefully removed. Their grating acuity was also 4 cpd[3]. The boy was referred to further clinical studies and was found to have rudimentary occipital cortex probably caused by a circulatory failure. Later another boy has been diagnosed with similar low recognition acuity and grating acuity 4 cpd[4]. In that case the boy had only a narrow peripheral remnant in the lower visual field where even normally sighted persons have very low visual acuity.

The *tectal pathway* is the pathway that also normally sighted people use for fast overview of environment, especially in traffic.

Children with normal recognition acuity and atypical problems in reading often perceive the center of a fine grating (8 cpcm) as having *distorted lines, moving lines or spots* as if there was poor coding of lines at the V_1 level. Some children with severe reading problems despite normal or near normal (0.63) recognition acuity perceive *nothing at the center of the grating*. When these children grow to young adults it may be possible to investigate in detail where in the Early Processing the information related to the very fine lines is distorted or totally lost. Sometimes the distortion or moving of the grating lines disappear and the lines look straight and darker if the grating is covered so that only a small central part of the grating is visible. This could be interpreted so that the surrounding information disturbs the information in the center (similar to increased crowding effect) but these two phenomena are not always present in the same children.

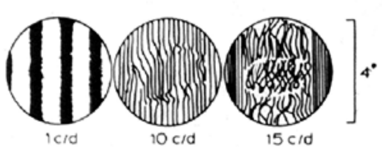

Figure 4. An adult subject's drawing depicting how he saw the gratings with his amblyopic eye. Changes vary from tangled lines or a white area in the middle of the grating to fast moving spots.

This phenomenon of distortion of grating lines was described already in late 1970s and drawn by an adult person with an amblyopic eye[5]. It has not been studied how common it might be. *We apparently lose important information by not routinely using grating tests* in the assessment of atypical visual functioning.

Early Visual Processing in the Occipital Lobe

When the neural activity in the optic radiation reaches the primary visual cortex it is coded based on the type of the neural activity (Fig.2.A): color related activity in the roundish "blobs", contrast edges and lines in the "interblob" areas and motion related activity in another layer of the primary cortex. This encoded information is then used in the visual cortices for definition of color shades, line lengths and orientations, filling in of gaps in visual information, figure-ground and object-background perception, direction and speed of movements etc. If fusion of information from the two eyes is good enough in infants, binocularity and stereovision can develop. Deviations from typical development in the use of vision during the first year of life should therefore be corrected without delay; "wait-and- see"-attitude may lead to loss of binocularity and stereovision.

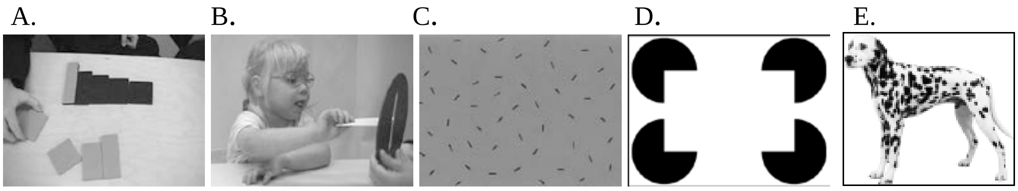

Figure 5. Assessment of some functions in Early Processing of visual information looks like simple play situations. A. LEA Rectangles Game, a pediatric variant of Efron's rectangles, allows observations on the development of the purely visual comparison of length of objects and grasping objects with good planning of goal directed reach. B. LEA Mailbox Game measures the ability to perceive and use information related to orientation of objects for the eye-hand-coordination: if a child does not perceive the orientation of the card given in the direction opposite to the direction of the slot, the card hits the "Mailbox/Postbox" in the original orientation. C. Visual closure is required for the gaps between the short lines to perceive and recognize the ring in this low contrast picture. D. Kanizsa's illusory rectangle depicts the function of completing a form drawn with a gap between two lines in exactly same direction. E. Pepi-test is two tests in one: 1) a test of

perception of motion, figure-in-motion and recognition of the figure, and 2) observing the eye movements: fixation, saccades, and following movements starting at the age of 4 months.

Inclusion of a few Early Visual Processing tasks in the neuropsychological assessment and in the games of occupational therapy makes the tester aware of loss of basic visual functions and helps to avoid an erroneous diagnosis of "delayed cognitive development" when a detail in visual functions has not developed. When observing infants and children with low visual acuity and contrast sensitivity, picture material has to be modified to meet the visual needs of each child. Also, drawing forms with dashed lines with the child before testing visual closure shows whether the child can perceive forms in this easier situation where the forms are discussed with the child. If the task requires hand functions, the test situation has to be modified to allow use of poor grasping, for example by pasting magnets on the puzzle pieces of the LEA Puzzle and on a champagne bottle cork so that the child can move the pieces like the other children.

After assessing the early processes in perception and recognition of forms (Fig.6), it is easier to become aware of the rare, unexpected total face blindness in a child who functioned normally with nice interaction in the tests of basic structures of form and then did not understand anything related to pictures of faces.

Figure 6. A. Careful, correct comparison of length of rectangles. B. Good planning of hand movement to turn the card in correct orientation before it touches the Mailbox. C. In Colorama Game the child must simultaneously compare color and form. D. Although several visually guided motor functions and purely visual functions were normal, the child did not understand how to look at pictures of faces, even when her finger was used to draw along the lines. Videos @www.lea-test.fi

In some brain damages, the difficulties in visual functioning are limited to a few areas or only one of the higher visual functions is lost. It is so difficult to imagine that a child who performs normally in all other areas cannot use vision in a few simple tasks and the child may be punished "until he/she stops refusing to do as being told".

Many children have changes in all four main areas of visual processing (Fig.2. B and C): 1) *Early Processing* in the occipital lobe, 2) *Mirror Neuron System* (MNS), 3), *Recognition functions* in the networks of the inferotemporal lobe (the ventral networks, VN) including face recognition, recognition of concrete objects and their pictures, recognition of abstract figures like Roman letters, numbers and geometric forms, and recognition of landmarks for route based orientation, 4) *Visual functions for doing* in the networks of the parietal lobe (the dorsal networks, DN), like awareness of directions and distances, which in O&M are called *spatial awareness; map based orientation in space; body awareness; visual attention; and eye-hand coordination*. These large networks are interconnected in normal brains. *In a brain that has had an atypical development of pathways and connections, some functions may not be present even in the hemisphere where they usually are. This can also happen in the brain of an elderly person*[6] after a vascular accident and is therefore likely to happen also in the developing brain if there is an early damage to a part of the pathways. Therefore it is wise not to stress the anatomic locations of recordable damages in the brain but discuss each student's specific functional abilities and problems listed in the Profile of Visual Functioning.

Mirror neuron functions

Mirror neurons are neural cells that are active during a motor activity and active when a person watches visually that activity in another person. These cell groups were first found in 1992 in the premotor cortex (marked MNS in Fig. 2.C) by di Pellegrino and later in numerous visuomotor areas of the brain. These multi-sensory action-observation systems enable individuals to (re) learn impaired motor functions through the activation of internal action-related representations[7,8]. Mirror neurons are found also in the Broca's area, which is involved in language processing and speech production, and in cortical areas that mediate visceromotor emotion-related behaviors.

A. B. C. D.

Figure 7. A. Imitation, copying facial expressions and tongue movements can be present in newborn babies. B. At the age of eight weeks visual communication can be active and at the age of 12 weeks leads to enjoyable interaction and supports bonding. C and D. Infants enjoy watching other infants' hand movements and copy them during "Color bath" in the Art Museum of Pori.

Imitation of facial expressions may be a sign of beginning function in the mirror neurons of newborn infants. In a few weeks it develops into a most enjoyable communication that uses both vision and vocalization. At the age of 3 months infants recognize smiling in the low contrast smiling Heidi pictures and respond with a typical social smile, which allows measurement of distance for visual communication. At the age of 3 to 4 months infants start to watch lip movements and goal directed hand movements for reaching and learn to copy other infants hand movements (Fig.7).

Observations on the development of *eye contact, visual interaction and social smile* are important in the follow-up of early communication. If these functions do not appear at the typical ages, vision must be thoroughly examined: anatomic structure of the eyes, refractive errors, accommodation, and eye movements. If the infant has high hyperopia or weak accommodation, near correction spectacles are prescribed. If the infant is found visually impaired, early intervention and training of parents in communication should be started, because otherwise visual deprivation may lead to abnormal development of communication and interaction. If eyes are normal, a pediatric neurologist assesses brain functions without delay.

Tabel 1.

CASE		N	I	P
CLINICAL FINDINGS, ocular motor functions				
A Fixation				
B Following movements				
C Saccades				
D Nystagmus				
E Strabismus				
F Accommodation, convergence				
CLINICAL FINDINGS, sensory				
G Binocularity				
H Visual Acuity				
I Grating Acuity				
J Contrast sensitivity, optotype				
K Contrast sensitivity, grating				
L Colour Vision				
M Adaptation speed				
N Photophobia				
O Visual field, central				
P Visual field, peripheral				
Q Motion perception, Pepi-test				
R Biological motion, Walking Man				
S Depth perception				
T				
U				
EARLY PROCESSING				
V Length of lines				
W Orientation of lines				
X Objects/figures on a patterned background				
Y Visual closure				
Z Textures and surface qualities				
AA Short time memory				
DORSAL STREAM				
A Perception of near and far space				
B Observation of surrounding				
C Orientation in space, map based				
D Route based orientation				
E Simultaneous perception				
F Eye-hand coordination				
G Length of lines				
H Direction of lines				
I LEA-Puzzle				
J Grasping and throwing objects				
K Drawing, free hand				
L Copying from blackboard				
M Spatial problems in math				
N Spatial problems in drawing				
O Spatial problems with thresholds, shadows				

		N	I	P
VENTRAL STREAM				
A Length of lines, purely visual test				
B Direction of lines, purely visual test				
C Recognition of details				
D Noticing errors and missing details				
E Recognition of faces				
F Interpretation of facial expressions				
G Reading body language				
H Landmarks				
I Concrete objects				
J Pictures of concrete objects				
K Abstract pictures of objects of different categ				
L Abstract forms (Roman letters, numbers)				
M Comparison with pictures in memory				
N Cartoons				
O Visual problems in copying pictures				
P Increased crowding effect				
Q Recognition in mathematical tasks				
S Spatial problems in math				
T Communication pictures				
MIRROR NEURON SYSTEM				
A Early communication and interaction				
B Interpretation of emotions and intentions				
C Observation and copying of movements				
D Effect of image quality, motion perception				
E Effect of image quality, contrast sensitivity				
F				
G				
OTHER COMMON PROBLEMS				
M Integration of sensory information				
N Visual and auditory overload				
O Specific memory problems				
P Head control				
Q Body control				
R Hand functions				
S Moving				
T Hearing				
U Executive functions				
V Other				
W Use of devices, categories decided locally				

Table 1. This long list of functions to be observed is for preschool and school age. If child's family, therapists, doctors, and teachers observe and report functions that might be affected by vision, a thorough medical and educational assessment is possible. It makes the assessments in different hospitals and private offices more standardized, which leads to earlier recognition of problems and better medical services and special educational arrangements. Each function is marked either NORMAL (N), IMPAIRED (I), or PROFOUNDLY impaired (P).

Some 80% of children with impaired vision have other disabling conditions
Even in Nordic countries all infants and children with motor or intellectual disorders do not get early evaluation of their vision and also later are not registered as visually impaired, which decreases their possibility to have vision included in their educational (IEP) and learning plans (ILP). Multidisabled infants and children should have vision included in their early intervention, after which it would be natural to build it in their education.

For a vision specialist the great variation of type and severity of other disabling conditions and their effect on the use of vision and devices often causes a great challenge. The assessment is most successful at the child's nursery school/ day care, kindergarten or school and the presence of the child's therapist and teacher is most helpful and also facilitates correct use of instructions. In this part of our work there are hundreds of new words and concepts each year but they are not discussed in this short paper.

A. B. C. D.

Figure 8. During the assessment as well as during activities at home and at school we should observe visual and general ergonomics. A. Right hemiplegic condition, right hemiamblyopia and inability to turn eyes down were compensated with the use of x-y-table where the text is pushed further away to bring it higher up on the screen (because the eyes did not turn down) and to avoid holding text with the only functioning hand. B. If head and body control are so weak that the child must be supported during testing this same weakness affects the use of sight also in the classroom (which is difficult in schools with conductive education). C. Good head support and proper near glasses facilitate use of vision. D. Communication may require an adult who dictates two possible answers in the communication device and the child chooses the one closest to her opinion.
Videos @ www.lea-test.fi/videos/julkaisut/Nordic_2013.

The new concept in assessment of vision impaired infants and children with other disabilities has been used in some places for years: at the hospital, the clinical examination of vision covers the observations and measurements that are necessary for the treatment of the child and those sensory functions that can be reliably performed (the first and second part of the Profile of Visual Functioning). The tests are used as they are used at schools and recorded so that the measured values can be compared with the values measured at the kindergarten, home or school. The educational assessment team is supported once or twice a year by the rehabilitation team and regularly by a special vision teacher. The tests and testing are modified to meet the needs of each child.

Summary: Many words in this paper are written in italics to ask you to pause a moment to think whether that word is well known. If not, study the pictures in detail, preferably with magnification. There are three concepts that I would like you to consider: 1) the *tectal pathway* that we often forget, 2) the *top-down flow* of visual information that organizes the use of inflowing information, and 3) the *variation in the relationship between recognition visual acuity and grating acuity*, which clearly confirms the well-known fact that visual acuity does not alone depict the type or severity of visual disability.
Videos are @ www.lea-test.fi/videos/julkaisut/Nordic_2013; Finnish Translation ready on June 11.

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