We have heard about the many ways in which cerebral disorders can affect visual behavior. I have been asked to speak about their functional classification.

To do so, we must look at different aspects of vision and vision loss. What do we mean when we speak of aspects? Let me give an analogy. Do you see three objects here? Or could it be that you are just looking at three aspects of an unfamiliar object? Turning the object around to look at it from different viewpoints, we see that what seemed different objects may just be different aspects of the same object.

I like the concept of ASPECTS, because it informs us not only about properties of the object, but also gain insight in the point of view of the beholder.

Consider a patient with AMD, who comes to make an appointment. The front desk will think about when to schedule her. The doctor will think about which treatment to select. The office manager may worry whether the insurance may pay. The daughter worries whether mother can still drive. Those are all very different aspects of the same clinical case.

Similarly, depending on our point of view, we can consider many different aspects of vision loss when we discuss visual functioning.

First we may consider how various causes may result in structural changes at the organ level, such as scarring, atrophy or loss. Here we need the pathologist to look at the structure of the organ.

However, the structural changes do not tell us how well the eye can function. We need to widen our view to include functional changes, and measure aspects such as visual acuity, visual field and contrast sensitivity. Knowing how the eye functions, does not tell us yet how the person functions. So we need to widen our perspective again to consider tasks, such as reading, mobility, face recognition. Here we need various low vision professionals to work with the patient, in this picture an elderly patient with macular degeneration.

Beyond that, we need to look at the consequences in a societal context. Do these changes impact on the person’s participation in society, causing job loss or reduced quality of life? How can we be sure of a happy patient, which is the end goal of all our interventions?

For today’s discussion it is useful to draw a line in the middle; on the organ side we speak of visual functions, on the person side of functional vision.
★ Various activities may cover more than one aspect. When we consider READING, ★ the measurement of print size and reading speed belong in the organ function column. Yet, it is not only important how many words per minute we can read, but also how many hours per day. ★ Reading endurance belongs in the ability column. ★ Reading enjoyment, finally, belongs with the aspect of Quality of Life.

★ The traditional medical focus is on the organ of vision, its structure and its functioning. ★ The rehab focus extends this to the functioning of the person.

★ For the traditional focus on the causes of CVI, we need an anatomical classification. We consider eye conditions, which can involve either the optics or the retina. We also consider cerebral conditions, which can involve either the pathways or the cortical structures. That cerebral causes can involve more than just the cortex is a clear reason why we should speak of CEREBRAL, rather than CORTICAL visual impairment.

★ To discuss the consequences of CVI we need a functional classification. That will be my topic. In the area of VISUAL FUNCTIONS, we have good standardized methods to measure things such as Acuity, Field and Contrast Sensitivity. We need to extend that to the area of cognitive dysfunctions. We need to define the problems and standardize the assessment.

★ In the area of the ABILITIES of the person, we can look at the four categories promoted by Lea Hyvärinen. We again need to look in more detail at cognitive problems for each activity.

★ Going back to the earlier distinction between visual function and functional vision, ★ we use the term VISUAL IMPAIRMENT to indicate deficits in how the eye and the visual system function. ★ I prefer to use the term VISUAL DYSFUNCTION for deficits in functional vision. I prefer these separate terms to emphasize the differences in approach.

★ At the cerebral level, visual impairments deal with lower visual functions, mainly in retinotopically organized cortical areas. Visual dysfunction refers to higher visual functions in areas with conceptual and cognitive organization.

★ The assessment of the two areas is very different. ★ On the impairment side, we typically test one parameter at a time, in an artificial environment. We record the stimulus level that results in a fixed endpoint: the threshold level, where the responses are 50% correct. ★ On the dysfunction side, we present a fixed task and record the variable performance of the subject. That performance must be at a sustainable level, which always is well above 50% correct.
Here I show three familiar threshold tests, each measuring a different parameter. Visual acuity, contrast sensitivity and dark adaptation. Each test varies only one parameter at a time.

In real life, however, we always have to deal with multiple parameters. Detail varies, as does contrast, as does illumination. Furthermore, we ask for sustainable performance.

In visual functioning, we can very broadly define three stages, with very different properties: the optical, the retinal and the neural stage. In the optical stage, an image of the outside world is projected onto the retina. In the retina this optical image is translated into neural impulses. The neural stage processes these signals, beginning in the inner retina and proceeding to the visual cortex and higher centers to finally allow visual functioning.

The optical stage has been known for many centuries. Retinal diseases have been known since the invention of the ophthalmoscope, a century and a half ago. Neural processing has often been overlooked and is gaining more attention only in the last decades.

How do we get from the retinal image to visually guided action? A significant intermediate step that is often insufficiently studied is that of the MENTAL MODEL of the environment. The initial transition from retinal image to mental model is largely subconscious and involuntary. The transition from mental model to visually guided action involves perception and cognition and conscious decision making.

We know that this intermediary step exists, because we cannot interact directly with a retinal image that changes with every eye movement, we need a stable model of the environment to plan our actions and to execute our locomotion through and our manipulation of that environment.

Modern imaging techniques have given us a better idea of where various brain areas involved with these process are located. I will not discuss those details. I just want to point out that it speaks to the incredible plasticity of the brain that, while the neural processing undoubtedly is the most complex part of vision, it also is the part that most rarely presents problems, except in the cases that we are discussing today.

The previous slide depicted the many brain areas that are involved in different components of functional vision. Yet, the processing from all those areas is seamlessly integrated. We would never know that all this integration occurs, if it were not for cases where it fails.
Brain damage-related Vision Loss – Functional Classification
August Colenbrander, MD, April 2008

- Vestibular and visual cues are normally integrated. ★ But if they contradict each other, we experience motion sickness.
- ★ The images from the right and the left eye are integrated into a single perception. ★ If that fails, we experience diplopia.
- ★ Information from the left and the right hemisphere is integrated. ★ However, we cannot detect any boundary, except in cases of hemianopia.
- ★ Information from the ventral stream (the “what” stream) and the dorsal stream (the “where” stream) is similarly integrated. ★ But we notice problems only in cases of brain damage.

- ★ Let us look a little closer at the differences between the retinal image and the mental model.
- ★ We already mentioned that the retinal image is constantly changing, but that the mental model is stable.
- ★ Also, the retinal image is eye-centered, whereas the mental model is environment-centered.
- ★ We can simulate what a video camera sees when it scans the room. That shifting image looks unnatural and may make you feel sea sick. ★ When we simulate the same with roving eye movements across a stable environment, the image looks far more natural.

- ★ Another important difference is that the retinal image is made up of 2-dimensional figures, whereas our mental model is filled with 3-dimensional objects.
- ★ In this image from Escher, we see a pond with the reflection of some trees above, we see leaves floating on the surface and we see a fish swimming under the surface. On the retina, these three components are intermixed, but in our mental model they represent three distinct domains.

- ★ Different activities have different visual demands.
- ★ In reading the emphasis is on recognition of finely detailed, 2-dimensional letter and word shapes. ★ The question of WHAT we see is relatively more important than WHERE we see it.
- ★ In mobility, on the other hand, we want to avoid obstacles. ★ To determine WHERE a 3-dimensional obstacle is, is relatively more important than determining WHAT it is.
- ★ In the manipulation of objects, the 2-dimensional detail and the 3-dimensional shape and location are both important.

- ★ The retinal image is strictly visual. However, the mental model is multi-sensory. ★ Even the blind have a mental model of their environment, built entirely from non-visual information.
When we shift our attention, we do that in the mental model. Those attention shifts then drive our eye movements and visual search.

Looking at Escher’s picture again, we can shift our attention from the trees to the leaves, or to the fish, because they represent different domains in the mental model. We could never do that in the retinal image, where the three are completely interwoven.

Finally, the retinal image ends at the limits of our visual field and has gaps where the retina has blind spots. The mental model extends around us, although the area behind our backs has vague details, because we have gathered less information about that area. The retinal image changes from moment to moment. The mental model is persistent and filled with the best available information, accumulated in memory.

The vision of people with a central scotoma is often simulated with a picture as shown here. In reality, people never see their central scotoma as a hole in their vision. The stationary picture is unnatural because it eliminates eye movements. When we introduce eye movements, we see that the scotoma disappears from our awareness, as long as it is moving. As soon as it stops, we see it again as one big gaping hole.

Having discussed the various stages, we can now summarize the steps in the visual process.

The first step is image formation. It is an optical process. Its clarity is judged mainly by the fovea.

The next step is image detection in the outer retina. It introduces possible topographic variations due to the distribution of retinal disease. This introduces the importance of the visual field.

The next steps, starting in the inner retina, involve image processing. After analysis of the components in various brain centers and the synthesis of the result, it provides us with a visual perception, the mental model of our environment.

Next is interpretation of that mental model and cognitive understanding. It involves memory, recognition and integration with other senses.

Finally, we are ready for a motor response to interact with the mental model of our environment.

We can now summarize the steps in the visual process in these four boxes.

- Image formation and image detection.
- Basic image processing.
- Synthesis to a visual perception and its cognitive interpretation, and finally,
- Interaction and motor response.

At this point it is important to note that our functional and anatomical divisions do not coincide. This is because the eye is not just a peripheral organ, in its development the retina is part of the brain.
We are now ready to talk about terminology. We can apply the term visual impairment to the top two boxes. And the term visual dysfunction to the next, rather complex, group of steps that lead to cognitive interpretation. Adding adjectives, we can divide the impairment box into Ocular Visual Impairment (OVI) and Cerebral Visual Impairment (CVI), to make an anatomical division.

At the visual dysfunction level we could speak of cerebral visual dysfunction. However, this would be a pleonasm, since all higher functions are cerebral. I prefer to use the term Cognitive Visual Dysfunction (CVD) to emphasize that dysfunctions are qualitatively different from impairments. Using a separate term also solves the dilemma with some professionals who do not want to use the term CVI unless there is visual acuity or visual field loss. We can now say that disorders such as prosopagnosia or simultanagnosia are visual dysfunctions, rather than simple visual impairments.

Adding the interaction with the environment, we add the possibility of motor dysfunctions.

This slide shows some examples for each of the categories. Ocular Visual Impairments (OVI) include cases of optical scatter (cataracts) or defocus (refractive error) and of retinal scars and dystrophies. Cerebral Visual Impairments (CVI) include sensory functions, such as visual acuity (amblyopia), field (hemianopia) and motion perception and oculo-motor functions such as fix and follow. So far, these functions are entirely visual.

Moving to visual dysfunction, other senses come into play as well. Examples include: visual neglect, hand-eye coordination (foot-eye coordination in mobility), figure-ground separation, color naming, etc.

In the motor dysfunction box we can mention that children in these groups often exhibit inconsistent behavior and are easily tired.

Looking again at visual acuity, we see that it can play a role at all levels.

At the optical level it plays a major role, but as we proceed the role of visual acuity lessens. Field defects outside the fovea, even a hemianopia, do not affect acuity.

Acuity describes the magnification requirement, which is irrelevant for cognitive problems.

Normal cognition can coexist with total blindness; abnormal cognition can also coexist with normal visual acuity.

At the motor level, eccentric viewing may disrupt hand-eye coordination and acuity, but the acuity loss does not cause the eccentric viewing.

These are additional reasons to reject the automatic association of the word impairment with visual acuity deficits.
Some quick notes about interactions.

- OVI, CVI and CVD may well coexist.
- They may also interact. That interaction can go both ways. For example: Deprivation amblyopia is caused by lack of input. Suppression amblyopia is caused by negative feedback from a higher level.
- Early visual impairments may limit a child’s general development if not properly addressed by the Blind Babies Foundation or other groups.
- In cases where visual dysfunction and general dysfunction coexist, we may say that Cognitive Visual Dysfunction exists if the visual developmental age is less than the general developmental age. That statement, of course, assumes that we have adequate scales for both.

In summary, we have provided you with functional categories to describe visual functioning.

- Those categories must be combined with ability ranges to describe whether the deficit is mild, moderate, severe or profound. Our time does not allow us to go into detail in this area.
- Additionally, these categories need to be coordinated with our developing anatomical insights, and also with the rehabilitative needs of the children or adults involved.

This slide summarizes the terms I propose to use.

- We distinguish between visual functions, which describe how the eye and the visual system function and functional vision, which describes how the person functions.
- When visual functions are disturbed, we speak of visual impairment. When functional vision is disturbed, we speak of visual dysfunction.
- On the impairment side we distinguish between ocular and cerebral impairments.
- On the dysfunction side, we speak of cognitive visual dysfunctions to emphasize the differences.

What should be next on our agenda? Creating better definitions will be helpful for many groups.

- Researchers need to know not to lump dissimilar impairments and dysfunctions into one group.
- Educators, who have predominantly learned how to deal with visual impairments, need guidance to distinguish visual dysfunctions as a separate group that needs a very different approach.
- Finally, the guidelines need to be simple enough to be understood by administrators, insurance and other third parties to prevent inappropriate denial of services.

Thank you for your attention.