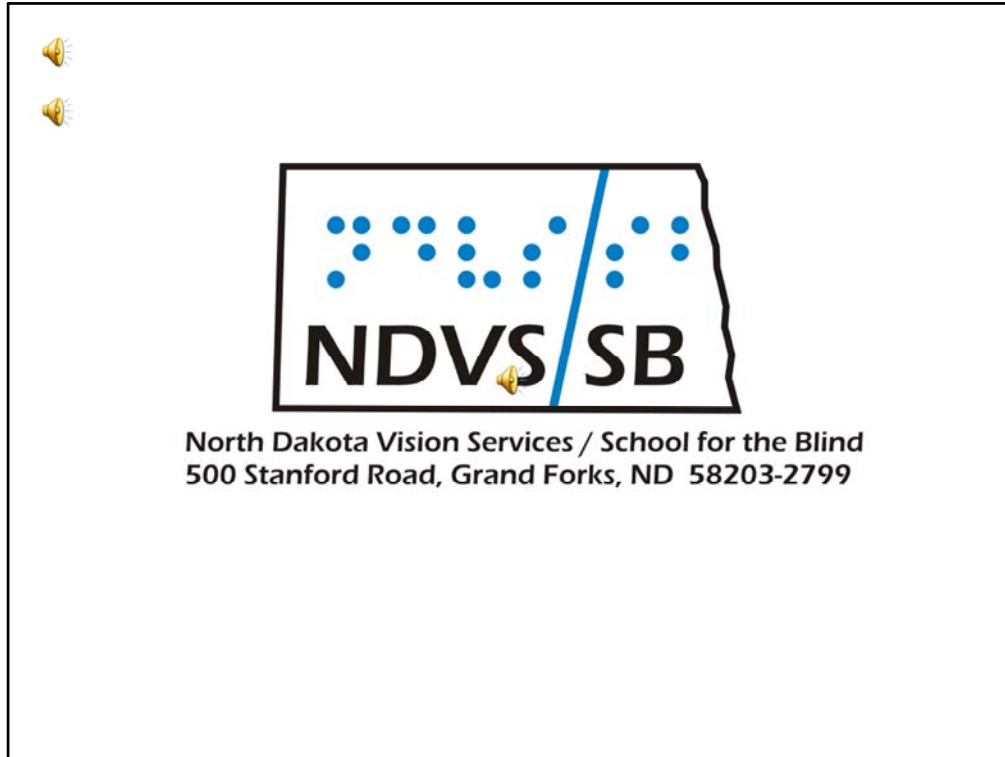


Welcome to “Introduction to Vision Loss.” This module will introduce the participant to educational considerations for children who are experiencing vision loss ranging from mild loss to total blindness. There are many causes of visual impairment, but this module will present information about several of the leading causes as well as provide guidance on several issues relating to normal eye health and function. Most importantly it will serve as a resource for parents, teachers and other professionals who want to gain a greater understanding of the implications that a visual impairment has on children. This presentation is a just starting point for becoming more knowledgeable about vision loss and its implications for children and their families.

This module was compiled by Paul Olson from North Dakota Vision Services/School for the Blind.

North Dakota Vision Services/School for the Blind is a state agency that is a division of the Department of Public instruction.



**The narrative in this presentation is meant to compliment the text. In addition to this further audio description of some pictures and other graphic information will be included to make the presentation more accessible to individuals with visual impairment.**

The logo for North Dakota Vision Services/School for the Blind is illustrated in this slide. The logo is an outline of the state with capitalized letters “NDVS/SB” depicted in both print and simulated braille font. Under the logo the the address of North Dakota Vision Services/School for the Blind is shown. The address is 500 Stanford Road, Grand Forks, ND 58203-2799.



## Normal Eye Health & Function

- The eyes are amazing, complex organs that are hardwired to the visual cortex of the brain.
- The eyes are comprised of numerous internal structures that work together to send millions of simultaneous impulses to the brain where the images are interpreted.
- When the structures of the eye and the brain work in harmony the result is the miracle we know as sight.

Any discussion of vision loss must first begin with a description of normal eye health and function. From this perspective, it is easier to understand the forms of dysfunction that can occur as the result of trauma, genetic abnormality or disease, infection or cellular tissue damage.

The eyes are directly connected to the brain. Normal function involves both the health of the eye and the accurate interpretation of images by the brain.



## Major Eye Structures

- Cornea
- Pupil
- Lens
- Iris
- Sclera
- Vitreous
- Retina

The health of the eye is highly dependent on numerous substructures. Some of the major structures of the eye include the cornea, the lens, the iris, the sclera, the vitreous and the retina.

A common analogy for the eye is that it acts like a camera. Light comes in through the cornea, a clear cover that functions like the glass of a camera's aperture. The amount of light coming into the eye is controlled by the pupil which opens and closes a little like a camera shutter. The iris, the colored part of the eye, is muscle that dilates and constricts to change the size of the pupil. The sclera is the outer portion of the eye that is white in color.

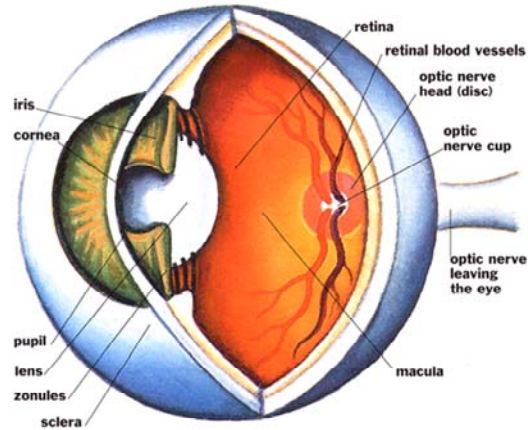
The lens rests behind the pupil. It has the appearance of a miniature magnifier lens. In young children, the lens is very elastic and can change in thickness as they look at objects that are close and far away.

The vitreous is the clear gel like substance that fills the eye and helps maintain the shape of the eye. It also helps bend the light as it passes through the eye on the way to the retina.

Light focuses on the retina, a series of light-sensitive cells lining the back of the eye. The retina acts like camera film, reacting to the incoming light and sending a record of an image via the optic nerve to the brain.



# Anatomy of the Human Eye



The anatomy of the eye is very complex. This picture shows a cutaway section of the eye and its internal structures. The retina is red in color. The arteries and capillaries that carry the nourishment to the eye are visible. The health of the eye absolutely depends upon good blood flow for nourishment and the removal of waste products that build up in the eye.

The head of the optic nerve is a large circular area in the back of the eye that appears to be on the surface of the retina. The macula is the area of the retina with the highest concentration of cone cells which give us the best possible vision for discerning detail.

Much more must be learned about the retina. The retina is made of multiple layers of two types of photoreceptive cells, known as rods and cones. Multiple sources of information can provide a more detailed description of the anatomy and physiology of the human eye. Visit the National Eye Institute website for further information .

## **Developmental Milestones in the** **First Few Months of Life**

- Stares at surroundings when awake
- Holds gaze on bright lights or objects
- Eyes begin to move with less head movement
- Begins to watch own hands
- Eyes turn inward while inspecting hands or toys
- Visually responds to smiles and voices of others

As with all physical development, milestones for visual development have been identified. A great deal of normal variation exists between children. For children with an identified visual impairment delays in reaching these milestones can be expected. Parents should be aware of the major developmental milestones for vision, yet understand that every child progresses at his or her own pace.

Parents—especially mothers—look for that first obvious visual recognition from their baby. This visual recognition serves to strengthen the emotional bond between the two. When this visual connection is delayed or absent, parents may become concerned. If visual recognition is delayed or absent, parents are encouraged to be patient and to tap into other senses to make an emotional connection with their child.



## Developmental Milestones for Toddlers and Preschoolers

- Interested in simple pictures
- Smiles when viewing favorite objects and people
- Watches and imitates other children
- Uses eyes and hands together
- Draws and names pictures
- Colors within lines
- Copies simple forms and letters

Learning certainly takes place using all senses, but vision is often the primary sense associated with academic learning. Learning and play are one and the same for children. Parents and educators look for developmental milestones in their toddlers and preschoolers because they place a great deal of importance on early learning. Toddlers demonstrate a universal interest in pictures and objects. Drawing and naming pictures, coloring and copying simple forms and letters are just a few of the skills that parents and educators expect preschoolers to exhibit.

Play is fueled by the curiosity to reach, touch and manipulate. When typical skills do not develop or develop atypically, parents and educators may need to take a more active role. Facilitating the development of these skills may include making necessary adaptations to support the child in developing them. When there is a concern with any of these milestones it may be appropriate to consider getting professional advice.



## What if Milestones are Delayed or Absent?

- Every child should have a clinical evaluation by an Optometrist or Ophthalmologist.
- The eye doctor will check the health of the eyes and observe the child's functioning.
- If major problems are identified a referral to a retinal specialist or a pediatric ophthalmologist may be necessary.
- Treatment and intervention can vary greatly. Referrals to other professionals (like early childhood educators) may also be beneficial.

Parents are usually the first to notice what might be a delay or an atypical visual behavior. Sometimes a family member or a medical provider will notice the delay or atypical behavior. Parents should not become alarmed when a delay or atypical behavior occurs. Instead a prudent course of action would be making a call to an eye care specialist such as an optometrist or ophthalmologist to discuss their concerns . In recent years, increased efforts to screen infants more thoroughly during their first year of life have been made. Early detection of eye anomalies increase the effectiveness of medical treatment. When delays or atypical behaviors occur, eye doctors may recommend a thorough evaluation of the child's ocular health.



## Clinical Eye Exams

- A basic examination focuses on ocular health.
- Diagnostic equipment is used to screen for anomalies.
- Refractive errors are corrected by prescribing glasses, contacts, etc.
- Health concerns are addressed through medication, surgery or other medical treatments.

During a clinical eye exam, the eye doctor might ask the parents about the child's general health. Then, he or she typically will perform an eye exam that measures: acuity, color vision and depth perception as well as early indicators of potential problems, such as glaucoma, retinal issues, or cataracts.

How can vision be checked in a baby or young child? After the dilating drops are given, the doctor shines a light in the eye and looks at the focus of the light that has been reflected through the eye. The doctor then places different lenses in front of the eye until the focus looks right. Once the proper lens power is determined, the doctor decides whether the patient needs help keeping things in focus -- if so, glasses will be prescribed



# Ophthalmoscope



The ophthalmoscope is one of the devices that may be used in a clinical examination. It is used by eye doctors to view the inside of the eye to assess the health of the retina and other structures.

Because it is normal for children to be fearful of medical professionals using unfamiliar tools (like the ophthalmoscope) it is important to take the process slowly and help the child become comfortable. This picture shows a young boy sitting with the eye doctor slightly bent over holding the hand-held ophthalmoscope to her eye about 3-4 inches in front of the child for optimal viewing. Again, this instrument allows the doctor to look into the child's eye.



## Slit-lamp Microscope



The slit lamp microscope is an instrument consisting of a high-intensity light source that can be focused to shine a thin sheet of light into the eye. The lamp facilitates an examination of the frontal structures and posterior segment of the eye which includes the eyelid , sclera, conjunctiva, iris, lens, and cornea.

This picture shows a child sitting with his chin and head resting in the device and the eye doctor looking through the microscope from the other side to see into the child's eye.



## What is Strabismus?

- Strabismus is a visual disorder where the eyes are misaligned and point in different directions. This misalignment may be constantly present, or it may come and go. Sometimes, only one eye is affected — turning inward (esotropia), outward (exotropia) or downward — while the other eye is directed straight ahead.
- Strabismus is a common condition among children. Normal alignment of both eyes during childhood allows the brain to fuse the two pictures into a single 3-dimensional image. Strabismus or abnormal alignment can block this normal binocular development and may cause “amblyopia” or reduced vision in one eye

At this point in the study of vision, it is appropriate to mention a condition called strabismus. Strabismus can be of great concern for parents. It is noteworthy in the discussion of vision impairment, but it in itself is not a major cause of vision loss. It is a noticeable condition that may prompt others to ask questions about it. Parents can find these questions difficult to answer. Eye doctors are well prepared to help parents make decisions regarding whether treatment is necessary.

Statistics indicate that up to 5% of children may have some detectable form of strabismus.

In infants, it is often difficult to determine the difference between eyes that appear to be crossed and true strabismus. Young children often have a wide, flat nasal bridge and a fold of skin at the inner eyelid that tends to hide the eye when looking to the side, thus causing the eyes to appear crossed.

Treatment can involve a process of patching as well as surgical intervention. Sometimes the strabismus will correct itself over time.



## Strabismus



In this picture, the left eye turns inward toward the nose. If the strabismus persists, the left eye is at risk of losing some function—resulting in a condition known as amblyopia.

Why do we need properly aligned eyes?

Proper alignment between the eyes makes 3-D vision (or depth perception) possible. When both eyes look at the same object at the same time, they each send a separate (and slightly different) picture to the brain. Within the brain are specialized brain cells ( called binocular cells) that fuse the two separate pictures together into a single three-dimensional picture.



## Visual Acuity

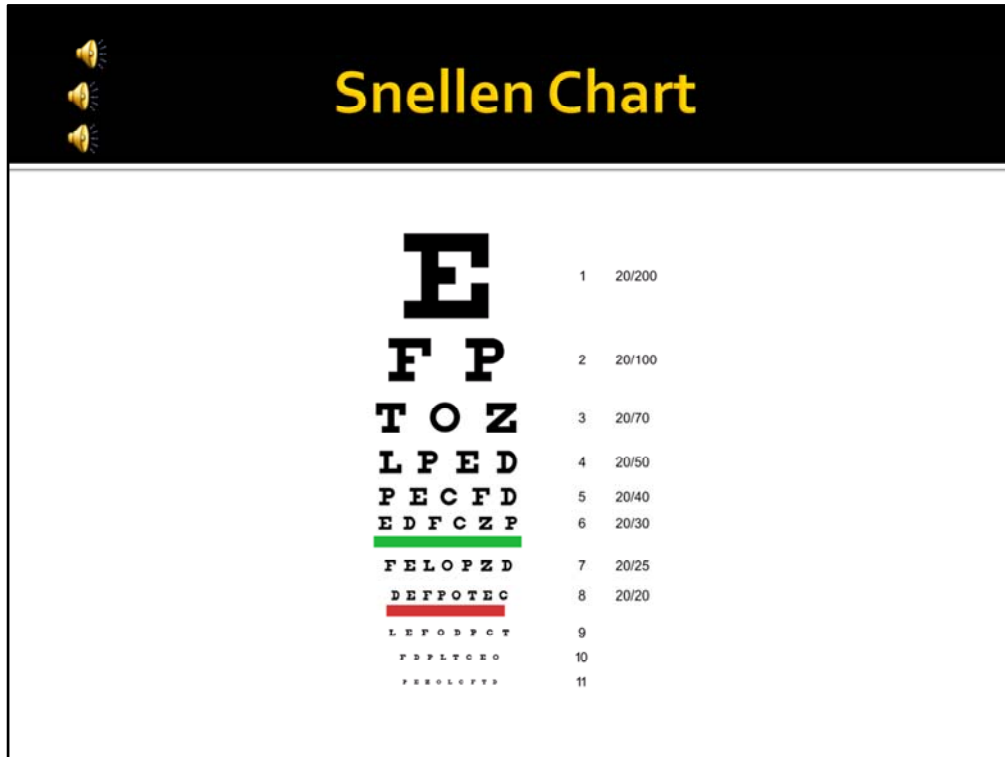
- Visual acuity is the ability to see fine detail clearly.
- The most common way to test acuity is with a traditional eye chart (or Snellen chart).
- Your score is based on how many lines or letters you can read down the chart from 20 feet away.
- The score is often written as a ratio. 20/20 is a benchmark for normal acuity.

One of the first things we think about when talking about vision is acuity.

Visual acuity is a measurement that allows us to make comparisons between individuals and better understand the visual functioning of these individuals. It is important to note that acuity is a rough approximation. Many other factors affect the overall perception of individuals. Two people with exactly the same acuity may function quite differently.

It is important to note that visual acuity problems cannot always be solved by prescription eyewear. Only acuity problems associated with the shape of the cornea and power of the lens can be improved through corrective lenses.

There are many conditions responsible for poor acuity that cannot be easily improved or corrected. The retina, the tissue at the back of the eye, is critical to visual acuity. Numerous pathologies can damage the retina. When the retina is damaged, the visual acuity cannot be significantly improved by glasses or contact lenses.



This is a picture of a Snellen Chart. The top line shows the large letter E. If this line is the only line that can be identified, the individual would have a recorded distance acuity of 20/200. The line underlined in green is near the middle of the chart (several lines down) and has significantly smaller print. If this line can be read without error, the individual will have a recorded acuity of 20/30. Further down the chart there is a red line. The print above this line signifies 20/20 acuity.

There are numerous types of eye charts that are used to record acuity. The Snellen is the best known. Other charts may be appropriate for younger children. Often pictures are presented instead of letters. This is certainly a better choice for children who cannot yet identify their letters.

Some charts are more detailed allowing for the eye doctor to measure acuity more accurately. Notice on the Snellen chart that there is a huge gap between the 20/100 line and the 20/200 line. It may be very relevant to know whether a child has an acuity of 20/120 or 20/160. Getting an accurate measure of acuity is not possible with this particular chart. Therefore, eye doctors may use charts that allow for better levels of accuracy.



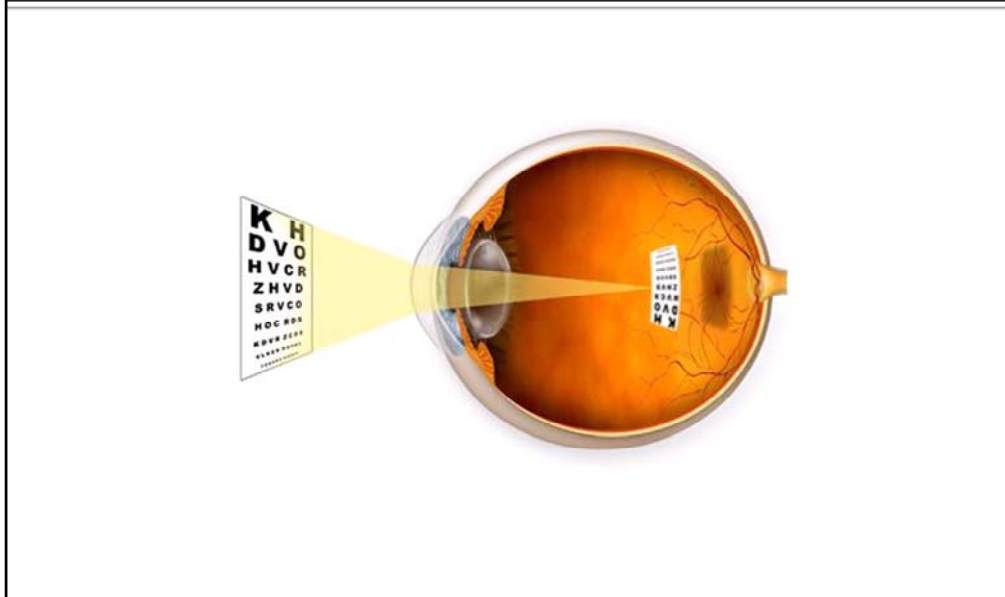
## Myopia (Nearsightedness)

- With uncorrected myopia, distance vision is blurred, while objects held closer are clearer.
- In effect, light enters the eye on a more or less parallel plane, but the cornea and lens bend the light too much.
- The image that is supposed to bend to a point directly on the retina in the back of the eye comes to a point in front of the retina.
- The power of the eye is too great.

Young children are generally nearsighted as they go through the various developmental stages. Newborns and toddlers are normally myopic, but some people are naturally predisposed to be myopic because of the shape of their cornea. The curvature of the cornea (the outer clear covering of the eye) has a great deal to do with how the light is bent as it enters the eye. If the curvature bends the light too much, the image forms in front of the retina and the image is not clear. In effect the cornea and lens together are too strong.



## Optics of Myopia



This picture illustrates the condition known as myopia. In this slide, a side view of the human eye is shown. The light rays are shown entering the eye and then narrowing as they pass through the eye. An eye chart is depicted in front of the eye. A smaller version of the chart forms slightly in front of the retina. The bending power of the lens is too strong which causes the image to fall short of the retina.



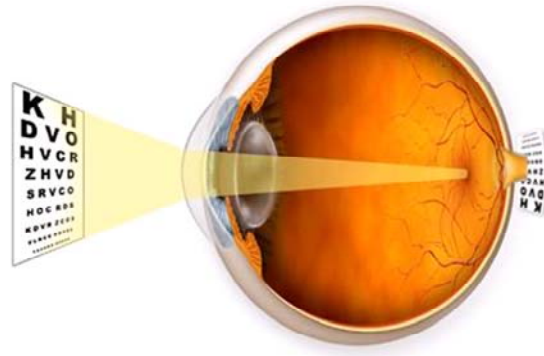
## Hyperopia (Farsightedness)

- An abnormal condition of the eye in which vision is better for distant objects than for near objects. It results from the eyeball being too short from front to back, causing images to be focused behind the retina

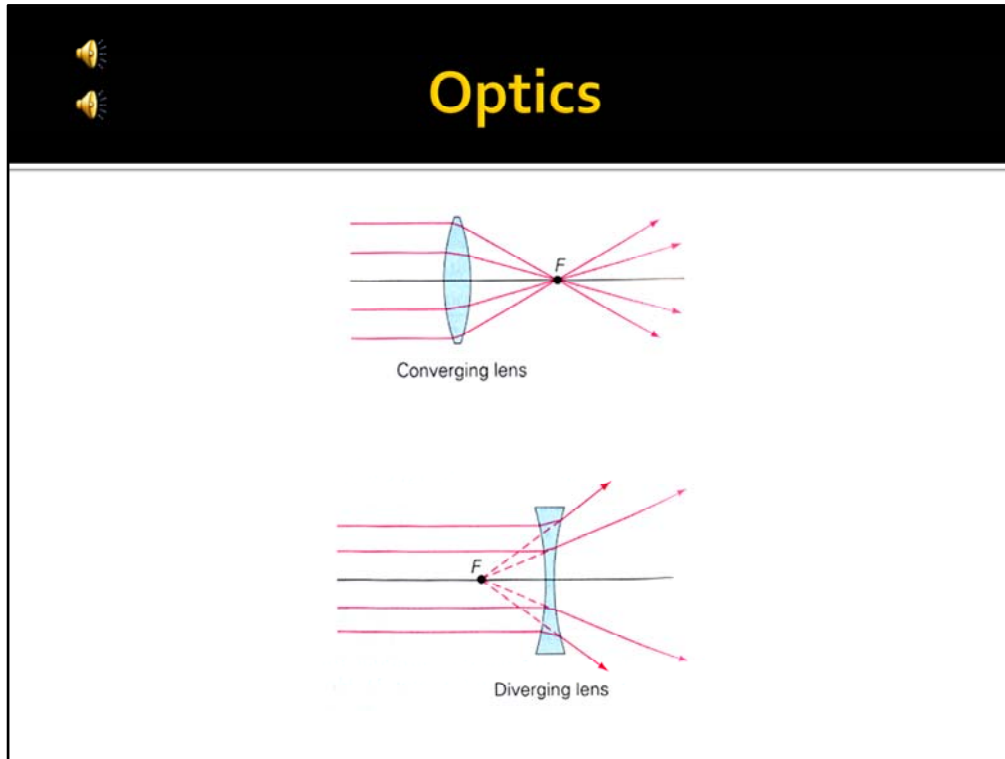
On the other side of the spectrum, some people are predisposed to be hyperopic. This is known as being “farsighted.” In this case the combined power of the cornea and lens do not have enough strength to bend the light sufficiently, thus creating a blurry image on the retina.



# Optics of Hyperopia



This picture illustrates the condition known as hyperopia. This side view of the human eye depicts how the light's rays enter the eye and narrow as they pass through it. When the light reaches the retina, a blurry image of the eye chart in front of the eye forms because the lens does not have sufficient power to bend the light and form the image on the retina. Instead, the light forms an image of the eye chart slightly behind the retina.



The solution to myopia and hyperopia is often prescription optics.

The lens shown at the top is a double convex lens meaning it has two surfaces that curve outward. This lens adds power by converging the light rays. This would be the type of lens needed if one has hyperopia or farsightedness. It helps people form better images at close range—such as for reading.

The lens shown at the bottom is a double concave lens. As the light rays pass through the lens, they diverge or spread apart. This type of lens is used to remedy nearsightedness. If the lens is too strong and converges the rays too much, it will bring the image to a point on the retina that is ideal for formation of a clear image.



The process of determining whether a child is nearsighted or farsighted is generally very effective. When one of these conditions exists eye doctors must determine the degree of “refractive error” in order to correct it.

The phoreopter is the device that eye doctors generally use to determine the lens prescription needed to correct the refractive error. Again, refractive error is simply the result of the light passing through the eye not bending at the best angle for clear vision.

In this picture, a child is seated and the phoreopter is on a movable arm that can be positioned in front of the child.

The child then looks through the phoreopter to read an eye chart. The eye doctor then asks the child which lens makes the image clearer. This process is generally very effective, but it does rely on accurate verbal responses from the child.

There are other methods of determining refractive error and the appropriate prescription when a child cannot accurately provide verbal information to the eye doctor.



Prescription eyewear will greatly improve the problems associated with refractive error. There are two pictures on this slide. The one on the left shows a woman wearing standard eye glasses. The picture on the right is a close up of a standard, clear contact lens on the tip of a finger. Again, prescription eyewear helps bring light into the eye at the best possible angle. It is difficult for many people to understand that eyewear does not solve more complex cases of visual impairment.

Nevertheless, individuals with a true visual impairment may benefit from prescription eyewear. The degree of improvement varies greatly from one individual to the next. For example, an individual may have a retinal disease that limits the extent of improvement that eyewear can achieve.

Any degree of functional improvement may be beneficial. A person identified as legally blind (20/200 acuity) may improve his or her acuity to 20/150 with the correct eyewear. This level of acuity is still certainly a significant challenge, but eliminating the refractive error is desirable in most cases. If the degree of refractive error is very small, it may not improve vision substantially especially when the vision loss results from a severe eye disease. In any case, it is important to discuss these issues with an eye doctor and make decisions based on the unique needs of each child.

## What if prescription eyewear does not solve the problem?

- There are numerous reasons for taking the process one step further for some students.
- A **Functional Vision Evaluation (FVE)** may be an appropriate consideration.
- A recommendation for (FVE) may come from the IEP process.

Prescription eyewear is important, but it may only be one part of a multi-faceted treatment approach. In order to fully understand a child's visual strengths and needs, it may be necessary to do a functional vision evaluation.

## What is a Functional Vision Evaluation (FVE)?

- A process which is aimed at determining how a student with a possible visual impairment typically uses their remaining vision (or other sensory channels) in educational settings, home settings and in the community.
- Builds on eye-medical information to determine how the student uses their vision to function.

A functional vision evaluation goes beyond the information provided by a clinical assessment. First, the evaluation is conducted in locations that are pertinent to the child—like the home and the classroom. The acuity of each eye is very important to note from the clinical assessment, but that acuity may be greatly affected by the lighting in different settings. It may also be affected by fatigue and other factors.

Color deficits may be noted in a clinical assessment. Children may respond to some colors in a school setting much better than other colors. These subtle strengths and weaknesses can be recorded after multiple observations. Teachers and parents need to understand any difficulties that the child may be experiencing working with real school materials. Knowing the best print size and font is an extremely important consideration for each child. If reading rate is significantly reduced, overall academic performance can be negatively affected.

The child's ability to track moving objects, determine depths and constantly shift vision from close objects to far objects are all crucial visual skills in a school setting. If there are deficits, recommendations for training or making adaptations can be made.

Professionals qualified to perform FVEs include teachers of the Visually Impaired, Certified Orientation & Mobility Specialists and other related service personnel with additional training in visual impairments.



## Leading Causes of Childhood Visual Impairment

- Retinopathy of Prematurity
- Optic Nerve Hypoplasia
- Cortical Visual Impairment

What are the leading causes of childhood visual impairment? The leading causes are: Retinopathy of Prematurity, Optic Nerve Hypoplasia and Cortical Visual Impairment

Statistics on visual impairment can be found in a number of reliable sources. The American Foundation for the Blind, The American Printing House for the Blind, the National Eye Institute and the World Health Organization are a few of the best know sources.

Across the board, the statistics indicate a much higher incidence rate of visual impairment with adults—especially the elderly. Statistics on children are a little more difficult to find. These statistics will be much different if one narrows the search to visual impairment in the United States and other developed countries versus worldwide. As would be expected there are many causes of visual impairment in other countries that are less prevalent in the US. These causes are minimized in the US due to better health care and disease prevention.



## Retinopathy of Prematurity

- Retinopathy of Prematurity (ROP), is the growth of abnormal blood vessels in the retina that generally begins during the first few days of life and may progress rapidly to blindness over a period of weeks.
- Blood vessels normally grow toward the edges of the retina until the time of birth. When a baby is born prematurely, this normal vessel growth stops and new abnormal vessels begin to grow.
- Over time this vessel growth produces a fibrous scar tissue which attaches to the retina and the vitreous gel that gives the eyeball its shape. If enough scar tissue forms, it can begin to pull the retina, detaching it, and, in some cases causing blindness.

Not all premature babies will develop Retinopathy of Prematurity or ROP. Many babies born with ROP will improve spontaneously. However, due to ROP being "responsible for more blindness among children in this country than all other causes combined" (Watson, 1997), it is important to screen premature babies for ROP. During the examination, doctors use an indirect ophthalmoscope, which allows a wide-angle view of the retina. The examination begins after a drop of topical anesthetic has been applied to the eye to reduce the baby's discomfort. Then the baby's eyelids are held open with a device called a speculum and a special probe holds the eyeball still while the doctor examines it. Because this examination can be stressful to the baby, sometimes the exams are postponed until the baby's medical condition is stabilized. Usually only babies that are at high risk for ROP, those born at a young gestational age and/or low birth weight, are screened. (Ophthalmology Associate Homepage, 1997)

Although a correlation has been established between preemies who receive high levels of oxygen and ROP, additional factors may contribute to the development of the condition. In addition to birth weight and gestational age, other potential contributing factors include: elevated blood carbon dioxide levels, anemia, blood transfusions, intraventricular hemorrhage, respiratory distress syndrome, chronic hypoxia in utero, multiple episodes of apnea or bradycardia, mechanical ventilation, and seizures (Ophthalmology Associates Homepage, 1997).

Another theory hypothesizes that exposure to bright fluorescent lighting in hospitals contributes to the development of ROP. This theory is not supported in the research and therefore, not widely accepted among ophthalmologists (Prevent Blindness in Premature Babies, 1997; Ophthalmology Associates Homepage, 1997). Current thinking hypothesizes that a combination of factors, some occurring in utero and some occurring after the baby is born, lead to the development of ROP.



## Stages of Retinopathy of Prematurity

- Stages 1 and 2 usually do not require treatment
- Stage 3 requires very close monitoring
- Stage 4 represents partial retinal detachment
- Stage 5 is typically associated with complete retinal detachment

The stages of ROP are determined by close observation of the health and physical characteristics of the retina. Certain classifications or stages are used to describe the progression of the condition. The location and degree of retinal scarring present determines the classification and stage of ROP.

In terms of treatment, Stage 1 and 2 ROP require weekly observations until the retina is fully vascularized. Some babies with ROP may recover spontaneously from stage 1 or stage 2 ROP. Infants that have stage 3 ROP typically require treatment with an argon laser to slow or halt the condition's progress. Children with stage 4 and 5 ROP may require "vitroretinal surgery". It is important to stress that not every child with ROP will progress to stage 5.

## Kristi with son, Ethan



Kristi Thiseth describes her experience with her son, Ethan and his diagnosis of ROP .

Hello, my name is Kristi. I have a son Ethan who is two and a half years old. Ethan was born when he was only 23 weeks gestational age. At that time the doctors did not give us much hope for his survival; but Ethan did survive. We are very, very thankful for that. When Ethan was about 30 weeks gestational age, they had the eye doctor come and look at his eyes. The eye doctor was immediately concerned by what he saw. A week or two later, Ethan then had his very first eye surgery. It was a success but there were many more problems so they sent us out to Michigan for more surgeries. We did get to come back home and we were in the hospital for almost five months. We got dismissed one day, got to drive to Minneapolis for more eye surgery the next day. At that surgery, the eye doctor did cataract surgery. They were going to take out a lens and possibly put in a new lens in if the doctor felt his eye was viable. At the surgery the doctor looked at his eyes and saw no blood flow. So he came out of surgery, the doctor told us that Ethan was going to be completely blind. We would be lucky if he even could see light or dark. We were devastated but Ethan had still survived so we were thankful. We were still thankful. The next morning when the eye doctor checked on Ethan again, he did see his blood flow, so we were very excited but he told us not to be too optimistic. He still had no idea what Ethan's visions would be.



## Educational Implications

- For students with some functional vision, recognizing facial expressions can be difficult.
- Glare and strong lighting may further impair visual functioning.
- Print materials may either have to be enlarged or provided in braille when the child gets to preschool and school age.
- Mobility skills can be effected.

If the student has a refractive error in addition to the ROP, glasses may be prescribed. This often helps but does not completely solve the problem. Students may require magnification of reading and graphic material. This can be achieved by bringing the material closer to the eye, or by the use of prescribed magnification aids - hand held magnifiers or reading spectacles for near work or a monocular (hand-held telescope) for viewing objects in the distance. Reading material often needs to be modified. Materials may have to be converted to an audio format, enlarged or produced in braille.

Students with ROP need good lighting for reading tasks. Each student should be observed closely to determine how he or she reacts to light. Often children with ROP can be very light sensitive. Students may experience mobility problems. Seeing objects and surface changes (like curbs) can be difficult. If there are concerns an orientation and mobility assessment should be conducted. Some students require instruction in travel safety. This sometimes involves the use of the white cane which can greatly enhance confidence and safety.



## Ethan and Tigger Having Fun



Ethan is a wonderfully active and happy little boy. He has progressed quite nicely in a number of areas. His visual impairment is a factor in his life, but he is not letting it slow him down.

Ethan and his parents receive support from a number of professionals. They have learned along with him. His parents focus more on his abilities than his challenges. Preparing for future challenges requires his parents to learn and experiment with strategies and technologies to determine which can best meet his needs. In the future, Ethan may need adapted materials, technology and even some instruction on how to travel safely and efficiently with a visual impairment. All of these considerations are possible. Being open to these options is important because having a visual impairment is a journey that is different for each person. Ethan like all children is first and foremost a child who needs love, support and high expectations.



## Optic Nerve Hypoplasia (ONH)

- Optic Nerve Hypoplasia is a congenital condition characterized by underdevelopment of the Optic Nerve and adjacent structures of the midline area of the brain.
- For people with Optic Nerve Hypoplasia, the optic nerve is either nonexistent or did not develop properly. Many people with ONH also have malformation (dysplasia) or absence (agenesis) of other structures of the midline of the brain that are physically near the optic nerve.

Hypoplasia means underdevelopment. In this case, the optic nerve is underdeveloped or absent. The optic nerve carries visual information to the brain, so anything interfering with the optic nerve will effect vision.

While only a few cases of this condition were reported in the medical literature before 1970, the numbers of children born with ONH seem to be increasing dramatically.

Some children also have an accompanying condition known as Septo-optic Dysplasia.

Although these conditions sound very serious, the functional level of visual impairment can vary greatly from very mild to more severe.



## Additional Implications

- Optic Nerve Hypoplasia is associated with a host of unique characteristics that distinguish it from blindness or visual impairment due to other causes. Children with ONH can exhibit a wide range of vision, from fairly good visual acuity to total blindness. They often have rapid, involuntary eye movements. This is called “nystagmus.”

Nystagmus, a disorder involving involuntary eye movements, often accompanies optic nerve hypoplasia. Nystagmus has two major presentations: jerk nystagmus, which is characterized by oscillations that occur faster in one direction than another, thereby creating a jerky rhythm; and, pendular nystagmus which is characterized by oscillations to either side that are roughly equal in speed.

Nystagmus is sometimes a sensitive subject for both children and parents. The involuntary movements of the eye are often noticeable and may cause some embarrassment. Children and families are encouraged to openly discuss these topics and to seek information that will help them understand the condition and answer any questions that may arise. It is important to note that nystagmus can accompany a number of other eye diseases. Sometimes there is no known cause for this condition.



## Educational Implications

- Children with ONH can have abnormalities in the structure of the brain, its function, or both. These abnormalities can be minor and have little to no impact on a child's functioning and development.
- In some children, however, these abnormalities can lead to developmental delays and other challenges.
- Children and adults with ONH demonstrate a full range of intellectual functioning from mental retardation to high intelligence.

Many children with ONH have sensory integration dysfunction. Sensory integration is a term that describes how the body processes information received from the senses. In people with sensory integration difficulties, this process doesn't work as it should. Children with ONH and sensory integration difficulties may be highly sensitive to certain textures, sights, sounds, smells, or other features of the environment. They might demonstrate a high pain tolerance and run into dangerous situations with reduced consideration for their safety. They might exhibit inappropriate and stereotyped behaviors, including hand-flapping, body-rocking, head-banging, biting or hitting parts of the body, and eye-pressing. While these behaviors can occur in other children who are blind or visually impaired, they can be much more pervasive and difficult to extinguish in children with ONH. Distractibility, impulsivity, and trouble focusing and staying on task are also typical behaviors in children with ONH that have sensory integration dysfunction.

Music is often a powerful motivator or even a primary means of communication for children with Optic Nerve Hypoplasia. Delays in areas such as adaptive functioning, social skills, gross and fine motor skills are very typical. Global developmental delays may also be prevalent.



## Cortical Visual Impairment

- Cortical visual impairment (CVI) is a neurological disorder, which results in unique visual responses to people, educational materials, and to the environment. When students with these visual/behavioral characteristics are shown to have loss of acuity or judged by their performance to be visually impaired, they are considered to have CVI. (APH)
- **Four major causes of CVI:**
  - Asphyxia (lack of oxygen)
  - Brain maldevelopment
  - Head injury
  - Infection

Cortical visual impairment is an unusual condition. Children with a CVI often do not exhibit the obvious or common signs associated with a visual impairment. The health of the eye appears normal, but its visual functioning can be very different. Cortical visual impairment is a neurological disorder that can result from several causes. Any number of medical conditions can inflict damage on key areas of the brain. Lack of oxygen at birth or afterward is thought to be one of the major causes of CVI. If something alters the normal development of the brain during gestation, a CVI can result.

In the past everyone who was considered to be visually impaired had to have reduced or absent visual acuity. In addition, the medical definition of CVI emphasized the loss of the ability to see in the distance (reduced acuity).

The medical definition of CVI is not well understood by non-medical professionals. While acuity testing is difficult in the young and disabled for physicians, it is even more difficult for teachers. Also, many children with normal acuity have visual problems similar to CVI. Over time the visual acuity of children with CVI tends to improve.



## Educational Implications

- Absent or unusual visual reflexive responses
- Focus on objects at very close range only
- Prefers to look at familiar objects versus new objects
- Often turns head away from objects and people

Behaviors associated with CVI can vary greatly. Some of the unique behaviors exhibited by a child with CVI include:

- A preference for looking at old objects, not new,
- A lack of visual curiosity.
- A preference for looking at objects in very close range.
- Difficulty with visual complexity or crowding.
- A distinct color preference, most commonly for red or yellow, but could be any color.
- Slow, often delayed visual responses
- An attraction to movement, especially rapid movements.
- A more unusual characteristic would be an absence or atypical visual reflexive response ( For example, the individual fails to blink at a threatening motion.)

These represent a few of the possible behaviors associated with CVI.



## Magnification & Other Assistive Technology



Dozens of different low vision aids improve the lives of people with visual impairment. Each aid has advantages and limitations. To get the most from a low vision aid, it's important to understand what that device can and cannot do. Since different aids help with different tasks, the person with a visual impairment will likely choose a mix of different low vision aids to accomplish all of his or her goals.

Hand-held magnifiers have been around for many years, but continue to get better. Desktop and portable video magnifiers offer a great deal of versatility in doing tasks at home, at school and at work for adults.

In the picture above, a hand-held video magnifier is held over a book to enlarge the print.

These devices have their own light source and show an undistorted view of the words or pictures that need to be enlarged for better viewing.

This category of device is just one of many types of assistive technology available for people with visual impairment—including blindness.



## Expanded Core Curriculum

- Communication modes (Braille)
- Orientation and mobility
- Social interaction skills
- Independent living skills
- Recreation and leisure skills
- Career education
- Assistive technology
- Sensory efficiency skills
- Self-determination

For students who have a significant visual impairment, there are important curricular considerations. Educators define "core curriculum" as the knowledge and skills students are expected to learn before their high school graduation. Generally, the core curriculum consists of knowledge and skills related to academic subjects. Mastery of the core curriculum is what both parents and teachers stress as essential for academic success in school, and later in life.

The "Expanded Core Curriculum" consists of 9 important topics. Braille may or may not be appropriate for every student, but it should be discussed and seriously considered. Instruction in orientation and mobility helps students learn how to move safely in all environments. Assistive technology is a must for students to have equal access to academic materials and to be productive in this information age. The other areas of the expanded core curriculum include: social skills, independent living skills, recreation, career education, sensory skills and self-determination. Self determination means learning how to be assertive and advocate for accommodations in educational and work settings. All of these skills need to be addressed within the individual education plan (IEP)



## Web Resources for Parents and Educators

- North Dakota Vision Services/School for the Blind  
[www.ndvisionservices.com](http://www.ndvisionservices.com)
- American Printing House for the Blind  
[www.aph.org](http://www.aph.org)
- American Foundation for the Blind  
[www.afb.org](http://www.afb.org)
- Texas School for the Blind and Visually Impaired  
[www.tsbvi.edu](http://www.tsbvi.edu)

Dozens of national and state agencies provide a wealth of information on visual impairments for parents and educators. As a statewide resource, North Dakota Vision Services/School for the Blind provides both outreach services and center-based programming.

American Printing House for the Blind (APH) is the world's largest nonprofit organization creating educational, workplace, and independent living products and services for people who are blind or otherwise identified as having a visual impairment.

The American Foundation for the Blind (AFB) states as its mission: to cooperate with and supplement the work of existing agencies in work for the blind..." In the intervening decades, AFB has become a leading organization for people who are blind or visually impaired by advocating for their rights and interests, broadening access to technology, promoting and facilitating independent living and elevating the quality of information and tools available for families and professionals."

The Texas School for the Blind and Visually Impaired, a residential school, is a national leader in the development of curriculum and other special materials for individuals with visual impairments. TSBVI publishes numerous book, guides, assessments that are available for order on its website.

# References

- Lueck, Amanda H. (Ed) (2004)
  - Functional Vision A Practitioners Guide to Evaluation and Intervention
  - American Foundation for the Blind

Texas School for the Blind and Visually Impaired—[www.tsbvi.edu](http://www.tsbvi.edu)  
Optic Nerve Hypoplasia By Ann Adkins, Education Specialist, TSBVI  
Outreach

Texas School for the Blind and Visually Impaired—[www.tsbvi.edu](http://www.tsbvi.edu)  
Retinopathy of Prematurity by [Kate Moss](#), Family Support  
Specialist, TSBVI Deafblind Outreach