COMMON VISUAL DISORDERS IN EARLY CHILDHOOD

There are a large variety of eye conditions that affect young children. They can have a variety of causes, including damage to the eye, disease, or an inherited condition. This list includes some of the most common conditions in early childhood. Organizations that provide information and support about each of these conditions are listed in the Resources section at the back of this book.

The three most common visual diagnoses in early childhood, according to the American Printing House for the Blind's Babies Count project, a national registry of young children, ages birth to 36 months, with visual impairments (www.aph.org/advisory/babies count.html), are cortical visual impairment, retinopathy of prematurity, and optic nerve hypoplasia. The frequency of these and other visual disorders are displayed in "Most Frequent Visual Diagnoses in Babies Count Project." Visual disorders are described here in the order of the frequency in which they occur:

Cortical visual impairment (CVI) s caused by damage to part of the brain related to vision. Although the parts of the eye appear normal, the brain cannot properly process the information it receives. The degree of vision loss can vary greatly, ranging from mild to severe. People with CVI may have trouble recognizing faces, interpreting drawings, perceiving depth, or distinguishing between background and foreground. CVI is also known as cerebral visual impairment, neurological visual impairment, and brain-injury–related visual impairment. Children with CVI are often not diagnosed until they are 6 months old or even older, and their visual abilities vary over time. A large portion of children with CVI also have additional disabilities, but many do not.

Retinopathy of prematurity (ROP) s a condition associated with premature birth and low birth weight in which the regular growth of normal blood vessels in the retina stops and abnormal blood vessels develop. As a result, the infant has an increased risk of retinal detachment, which means the retina can no longer transmit visual information to the brain. Retinopathy of prematurity was formerly called retrolental fibroplasia and once resulted in total blindness. Today, because it is usually diagnosed and treated in the hospital before the infant goes home, ROP rarely results in total blindness but may result in reduced or severely limited vision, particularly in the peripheral visual fields. Many babies with ROP are developmentally delayed, but only about one-fifth have additional disabilities other than developmental delay.

Optic nerve hypoplasia (ONH) s a condition, present at birth, in which the optic nerve, the part of the eye that transmits visual information from the eye to the brain, is underdeveloped, so that adequate visual information is not carried from the retina to the brain. The effects of optic nerve hypoplasia have a broad range, from little or no visual impairment to near-total blindness. The condition may affect one or both eyes. The majority of children with ONH are diagnosed before the age of 6 months, experience severe vision loss, but do not usually have additional disabilities.

Other eye conditions are much less frequent in early childhood, and there are many that are extremely rare. The following are a few of them:

Albinism s a hereditary condition characterized by a variable lack of pigment in the eyes, skin, and hair. People with albinism may have pale pink skin and blond to white hair, but there are different types of albinism, and the amount of pigment varies. The irises of the eyes may be blue, violet, or even hazel in color. They are sensitive to bright light and glare and commonly have other vision problems, such as nystagmus and strabismus. Although some people with albinism can see well enough to drive, many have impaired vision and may even be legally blind. Because of the pigmentation and nystagmus, babies

with albinism are usually diagnosed around 3 months of age, and a very small portion of them also have additional disabilities.

Nystagmus, as suggested above, is more of a characteristic than a specific condition. It often occurs with other eye conditions, such as albinism. It involves involuntary, rapid, repetitive movements of one or both eyes from side to side, up and down, or in a circular motion, resulting from the brain's search for the best point of focus. Nystagmus may be present at birth or, less commonly, may result from disease or injury. We know little about when nystagmus is diagnosed or whether additional disabilities are common to it because it is rarely a sole diagnosis and usually leads to a primary diagnosis of some type.

Cataracts re a clouding of the lens inside the eye. The lens is normally clear and the cataract can make the child's vision seem cloudy or blurry. If cataracts are present at birth, they are called congenital cataracts, and, if formed in the first 6 months, they are called infantile cataracts. Cataracts can come in different forms and locations on the lens, and the density of the cataracts impacts the degree of vision loss. Young children with cataracts must be followed closely by their eye care specialist to ensure the cataract is not harming the child's visual development. Sometimes cataracts do not cause problems, but, if they do, surgery to remove the cloudy lens is the only effective treatment. After the surgery, young children are frequently given contact lenses. In older children, the cloudy lens can be replaced with a lens implant (an intraocular artificial lens).

Retinoblastoma s a malignant tumor (cancer) of the retina, which is usually first detected in infancy or early childhood. Retinoblastoma is usually hereditary and may affect one or both eyes. Retinoblastoma has a cure rate of over 90 percent if treated early. Without prompt treatment, the cancer can spread to the eye socket, the brain, and elsewhere and can cause death. Depending on the size and location of the tumor, treatment options include laser surgery, cryotherapy (a freezing treatment), radiation, and chemotherapy. In some cases, the affected eye may need to be removed.

Strabismus s a condition in which each eye is not directed toward the same point simultaneously. Strabismus occurs when eye muscles are not working properly together. It is most commonly an inherited condition but may also be caused by disease or injury. If diagnosed early, strabismus can usually be corrected. The condition may be treated with corrective eyeglasses, eye-muscle exercises, surgery, or a combination of these approaches. Young children with this condition may need to wear an eye patch over their stronger eye to force their weaker eye to function correctly. Children whose strabismus is not corrected may develop amblyopia.

Amblyopia s a condition in which a person's vision does not develop properly in early childhood because the eye and the brain are not working together. Amblyopia, which usually affects only one eye, is also known as "lazy eye." A person with amblyopia experiences blurred vision in the affected eye. The brain suppresses the blurred image, and the child "sees" only the image from the unaffected eye. However, children often do not complain of blurred vision because this seems normal to them (they've never known anything different). Early treatment is advisable because, if left untreated, this condition may lead to permanent vision problems or even partial blindness. Treatment options include vision therapy exercises, prescription eyeglasses, or surgery. People with amblyopia may need to wear an eye patch over their stronger eye in order to force the affected eye to function as it should.

Structural disorders are changes or abnormalities in the formation of the eye. They are usually diagnosed soon after birth because they typically are noticeable. As a group, they affect only about 5 percent of the population of young children with visual impairment. The most common structural

disorders are the following:

Aniridia s the partial or complete absence of the iris of the eye. This rare condition, usually present at birth, results in impaired vision and sensitivity to light. Children with aniridia are also at high risk for certain other eye conditions, such as glaucoma, nystagmus, and cataracts. Children with aniridia are often sensitive to light and may benefit from wearing tinted contact lenses, sunglasses, or hats with a brim.

Coloboma s a cleft or gap in some part of the eye, such as the iris, lens, or retina, that is caused by a defect in the development of the eyeball. How much your child's vision is affected by a coloboma depends on the size and location of the cleft and whether it occurs in one or both eyes. For example, a child with only a tiny defect in the iris may have normal vision. However, if there are large defects in the retina and optic nerve, he or she may have quite limited vision.

De Morsier syndrome s a rare disorder, present at birth, in which the optic nerve is underdeveloped, the pituitary gland does not function properly, and often a portion of brain tissue is not formed. Also known as septo-optic dysplasia, De Morsier syndrome may cause blindness in one or both eyes and is often accompanied by nystagmus and various other symptoms. Some children with De Morsier syndrome have additional disabilities, whereas others do not.

Glaucoma s a disease in which the pressure of the fluid inside the eye is too high, resulting in a loss of peripheral vision. Without treatment, the increased pressure can damage the optic nerve and, eventually, lead to total blindness. Glaucoma is not easily diagnosed because it does not have any symptoms or warning signs. Making sure your child has regular eye examinations with the pressure measured by the doctor (and treatment prescribed, if necessary) is the key to preventing loss of vision. Your child may need daily medication (usually eye drops) or surgery, in addition to whatever treatment she receives for her main visual diagnosis.

Microphthalmia s a rare disorder, usually inherited, in which one or both eyes are abnormally small. The degree of visual impairment varies, from reduced vision to blindness. Extreme microphthalmia resembles some forms of anophthalmia (absence of the eye), where one or both eyes do not form during the first trimester of pregnancy.

Optic nerve atrophy s a degeneration of the optic nerve. Children are not born with optic nerve atrophy; as the name implies, the optic nerve weakens or wastes away, due to injury, disease, or lack of use.

There are also a group of eye conditions that affect the retina. These are about twice as common as structural disorders and are first diagnosed closer to 6 months of age.

Achromatopsia s a rare, inherited vision disorder in which a child has little or no ability to see color. People with achromatopsia commonly experience some vision loss, especially in bright light, to which they are extremely sensitive. Children may need to wear brimmed hats or sunglasses to handle glare. Children with achromatopsia do not see the world in black and white, but rather in shades of gray, which means that color will not be experienced the same way that someone with "normal" vision experiences it. Differences in color, however, can still be distinguished. Achromatopsia becomes more of an issue for school-age children, when so many of the print and digital textbooks utilize color as ways to highlight text.

Leber congenital amaurosis, n inherited condition, is thought to be caused by degeneration of the

retina. An infant with this condition is either born blind or develops severe vision loss soon after birth. Children with Leber congenital amaurosis typically also have nystagmus, and some may have additional disabilities.

Macular degeneration s a disease that impairs the macula, the area of the retina that is responsible for the sharpest central vision, used for such daily activities as reading, driving, and recognizing faces and colors. It is the leading cause of blindness in adults but is found in children as Stargardt disease (juvenile macular degeneration) and Best disease (a hereditary, progressive atrophy of the macula). These eye conditions cause blurred, distorted, or dim central vision or a blind spot in the center of the visual field. Peripheral vision is generally not affected.

Retinitis pigmentosa, degeneration of the retina, results in decreased night vision, a gradual loss of peripheral vision, and, in some cases, loss of central vision. The degeneration progresses over time and can lead to total blindness. Retinitis pigmentosa is a rare, inherited disease for which there is currently no treatment or cure.