Meet Henry
Henry is a likable young man of fifteen. He does not read fluently in either print or braille, but he uses his vision well for orientation and for avoiding obstacles as he travels. His motor planning skills are not great, and he brings all materials up close to his eyes when he works with them. When he meets with success at school, Henry imitates a drum role with his hands on the table and says, “I’m so proud of you!” (This is how his preschool teacher rewarded the children when he was four.)

Optic nerve hypoplasia (ONH) is a congenital eye condition that was thought to be rare into the twentieth century and which is now known to be one of the three most common causes of visual impairment in children in industrialized countries. It is not yet known why children are born with optic nerve hypoplasia and why there seems to be a great increase in the number of children born with it.

Optic nerve hypoplasia means that the optic nerve did not develop completely. Doctors can see this by looking into the back of the eye through the pupil. An undeveloped optic nerve cannot take visual messages to the brain adequately. The result may be poor visual acuity, fluctuating vision and the loss of visual fields or spots of no vision within the fields. Blurred vision may be corrected to some degree with prescription glasses. Some people who are born with ONH have 20/20 acuity, others have a range of impaired acuities and others may be totally blind.

Optic nerve hypoplasia was an interest of a neurologist named Georges de Morsier in the early twentieth century. De Morsier discovered that a few of the brains he examined from people who had been diagnosed with optic nerve hypoplasia also had compromised development of the septum pellucidum. The septum pellucidum is the part of the brain which divides the two halves of the brain. He named a syndrome septo-optic dysplasia (SOD) which includes both optic nerve hypoplasia and the absence of the septum pellucidum.

Apparently, it makes little difference if the septum pellucidum is not developed fully. However, children who are diagnosed with both optic nerve hypoplasia and an under-developed septum pellucidum are likely to have other anomalies as well. Currently, Dr. Mark Borchert of Children’s Hospital in Los Angeles is doing a longitudinal study of children who are diagnosed with ONH and SOD. He suggests that, since absence of the septum pellucidum is not a problem itself, we would do well to drop the diagnosis of septo-optic dysplasia and concentrate on determining what other anomalies a person who is diagnosed with ONH has.
Several problems may accompany optic nerve hypoplasia whether or not the septum pellucidum is absent. One is a pituitary gland that does not function well enough for typical growth. That means that children who have ONH often need endocrine supplements, such as human growth hormone and synthetic thyroid. Their adrenal glands may not produce enough cortisol. This can be a dangerous health problem if not detected, because without enough cortisol, a simple cold may be life-threatening. Children who have ONH may have a condition called diabetes insipidus which causes them to be constantly thirsty. They may have chronic hypoglycemia or low blood sugar, meaning that they require frequent, nutritious snacks to maintain their energy. Without snacks they become lethargic and may even have seizures or go into a coma in extreme cases.

Children who have ONH may also have poor development of the corpus callosum in the brain. The corpus callosum is the part of the brain which is responsible for communication between the two sides of the brain. This is a diagnosis that makes a difference. With no corpus callosum it is difficult to do tasks that require both hands. The students may appear to be ambidextrous. They use their right hand to reach for objects on their right side, and their left hand to reach for objects on their left side. They do not cross the midline of their bodies without a lot of effort. Likewise emotions and thoughts are not communicated between the sides of the brain and so may not influence each other. Mistaking letters in both print and braille for their mirror images is common in children who have a poorly developed corpus callosum. Mistakes of this kind are called “reversals.”

Communication and social skills of people who have ONH may be on the autism spectrum. Thought processes may be rigid. Some children who have ONH have difficulty doing the same activity in more than one way or following different rules in different situations. They are called “first-time learners.” They may learn something one way and not be able to alter their thinking about it when more information is available. It is important to keep this in mind when teaching these children. For example, if a child like this learns to call using the bathroom “going potty” at three years old, he may be talking about “going potty” into his teens. When working with children who are first-time learners, using language that will be appropriate for life is important from the beginning.

People who have ONH have a wide range of abilities. Some of them are as intelligent, athletic and social as the rest of their family members, and others may have severe physical, cognitive and social disabilities. Most will be somewhere between those two extremes.

Musical talent is frequently present in people who have ONH, no matter how disabled they may be. A hallmark of ONH is perfect pitch! Enjoying playing with language may also be a gift for children who have ONH.
Students who have ONH will need a range of accommodations for academic progress. Most will need to use braille or other tactile systems as their primary learning medium. They are likely to be bothered by glare and to benefit from using sunglasses and a hat with a dark brim outdoors. Good contrast between visual targets and backgrounds will be helpful. Avoiding clutter is important, just as it is for students who have cortical visual impairment.

In summary, people who have optic nerve hypoplasia or have been diagnosed with septo-optic dysplasia are likely to have a wide range of abilities in all areas. The common trait they share is reduced or fluctuating vision that has many of the characteristics seen in people who have other neurological visual impairments, such as cortical visual impairment or optic nerve atrophy. Here is a summary in list form of accommodations that you may need to provide for students who have optic nerve hypoplasia. Talk with the student’s teacher of visually impaired students to see which ones are appropriate and what other accommodations are needed.

- Provide enlarged materials if indicated;
- Protect from glare with sunshades and/or a cap with a brim in brightly lit situations;
- Make sure light sources are behind or to the side of the student who has ONH;
- Facilitate preferential seating when needed;
- Be informed about concomitant medical concerns and what measures you should take if symptoms are appear;
- Watch for fatigue and allow breaks as needed;
- Model language that will be appropriate for life;
- Expect to re-teach skills in each new environment.

Resources


[Optic Nerve Hypoplasia: A Guide for Parents (PDF)](https://example.com), written by Dr. Francine Kaufman, Dr Neal Kaufman, Dr. Mark Borchert and Talia Inlender.