DEFINITION

Optic Nerve Atrophy (ONA) is a permanent visual impairment caused by damage to the optic nerve. The optic nerve functions like a cable carrying information from the eye to be processed by the brain. The optic nerve is comprised of over a million small nerve fibers (axons). When some of these nerve fibers are damaged through disease, the brain doesn’t receive complete vision information and sight becomes blurred. Atrophy (wasting away) may be partial in which some axons are damaged or profound in which most axons are damaged. A child’s ability to see clearly (visual acuity) is affected due to nerve damage that occurs in the central part of the retina responsible for detail and color vision (macula). These areas of the eye are more vulnerable to the effects of atrophy. ONA is the end result of damage to the optic nerve. It can affect one or both eyes. It may also be progressive, depending on the cause.

CAUSE

Many diseases and conditions may lead to optic atrophy. Tumors of the visual pathways, inadequate blood or oxygen supply (hypoxia ischemia) before or shortly after birth, trauma, hydrocephalus, heredity, and rare degenerative diseases have been identified as causes of ONA. When hereditary, the pattern is dominant. This means that one parent with the condition would pass the gene to 50% of his/her children. If caused by a tumor, the process of ONA may be halted by removal of the tumor.

DIAGNOSIS

ONA in children is diagnosed by a pediatric ophthalmologist in a number of ways, including:

- Visual acuity and color vision may be found to be abnormal (if testable).
- Pupil reactions to light are diminished.
- The optic nerve, when examined with an ophthalmoscope, has a gray-white appearance, which may not be apparent for 4-6 weeks from time of optic nerve injury.
- Optic atrophy which occurs in both eyes from time of birth (bilateral and
congenital) may cause rhythmic, involuntary eye movements (nystagmus).

CHARACTERISTICS

Although there are several types of ONA, the following characteristics are common to most:

- Central vision is affected.
- Color vision deficits may be evident.
- It may be difficult for children with ONA to discriminate contrast, due to damage in the area of the eye responsible for detailed vision (macula).
- A wide range of acuity loss exists in this population.
- Onset of ONA may be gradual or sudden depending on the cause.
- A general decrease of sensitivity in all visual fields (depressed visual fields) may occur, also dependent on the cause of ONA.
- Many children with ONA have additional neurologic problems, such as seizures, developmental delays or motor problems, and Cortical Visual Impairment (CVI). (When CVI coexists with ONA, it may be difficult to determine which diagnosis is responsible for specific visual problems).

VISUAL AND BEHAVIORAL CHARACTERISTICS

- Visual acuity may range from nearly normal to totally blind.
- Children with bilateral central blind spots (scotomas) may "overlook" in order to see a person or object.

MYTHS

The following statement is NOT TRUE according to current research:

- ONA can be corrected with glasses.

TEACHING STRATEGIES

- Ongoing evaluation, and communication among family, medical and education specialists is essential to develop the best home and school program for the child with ONA. Assessment and services from a pediatric ophthalmologist, a teacher of the visually impaired, and a specialist in Orientation and Mobility who keep in close communication with caregivers will ensure maximum development for the child.
- Carefully observe a young child with ONA to gather valuable information about the way she sees best. Determine the best position for the child and her toys to accommodate for central field loss.
• The physical demands of looking at an object or toy for a long period of time may cause eye fatigue. Allow a child with ONA to rest between activities requiring vision.
• Use touch and spoken description to tell a child about present and future activities. The use of additional senses are necessary to enrich the learning process.
• Good contrast and lighting are essential for the child with ONA to see objects in the environment clearly. For example, offering dark colored food on a light plate, or a light toy against a dark background provides good contrast.
• Using bold colors (red, yellow, green, blue) and simple, clear pictures will help the child to see more clearly.
• Use familiar and real objects to encourage visual attention. Change one characteristic of a familiar object only after the child is able to recognize it consistently. For instance, after the child is able to recognize a cup that is blue consistently, change the cup to red.
• When introducing unfamiliar objects to the child, relate them to familiar objects and settings.
• Note: If a child with ONA also has Cortical Visual Impairment (CVI), strategies effective with the CVI population should be used (see CVI Fact Sheet).

GLOSSARY

1. Axon: single projection from a nerve cell that under normal conditions, carries nerve impulses away from the cell body.
2. Optic Nerve Hypoplasia: refers to underdevelopment of the optic nerve during pregnancy.

RESOURCES


Hoyt, C., Good, W. (1 992). Do We Really Understand The Difference Between Optic Nerve Hypoplasia And Atrophy?, Eye, 6,201-204.


ACKNOWLEDGMENTS

Project Coordinator: Julie Bernas-Pierce, M.Ed.
Dr. William Good, Hsiao-hui Ning, Dennak Murphy, Linda Kekelis, Sandra Nevin, Susana Saeidnia. Reviewers: Dr. William Good, Kathryn Neale Manalo.

The Pediatric Visual Diagnosis Fact Sheets are sponsored by a grant from the Blind Children's Center and with support from the Hilton/Perkins Program through a grant from the Conrad Hilton Foundation of Reno, Nevada.

REPRODUCTION FOR RESALE IS STRICTLY PROHIBITED (1/98 BBF)