Fact Sheet

Aniridia

This information was taken from http://www.aniridia.net/index.html and http://www.aniridia.org/conditions/index.html

Aniridia involves an absence or partial absence of the iris, which is the colored portion of the front of the eye. The iris is the part of the eye we refer to when we say a person has green, blue, brown or grey eyes. It is a small circular muscle with a hole in the middle, called the pupil. The iris controls the amount of light that enters the eye by contracting in bright light making the pupil small and dilating in dark conditions making the pupil large. An eye with little or no iris allows in more light than a normal eye. This causes photophobia or light sensitivity which is a difficulty to see due to glare and discomfort in bright light conditions.

Aniridia can be present in one or both eyes. A person can have total aniridia or no visible iris in one or both eyes. A person with totally absent irises appears to have very dark eyes this is because they only have a large pupil. However, the iris is never completely absent; the root of the iris is still present but the muscle which creates the visible iris is missing. A person may have a partial Aniridia or part of one or both irises is missing. A person with partially absent irises appears to have very large and/or misshapen pupils with a small ring of color around the edge of the pupil. In these cases the iris will not contract and dilate when appropriate. Finally a person may have atypical aniridia or the iris may appear normal but it may be thinner than normal or have connective tissue defects. Ectopia pupillae may be a form of atypical aniridia.

Aniridia is rare. Its incidence is between 1/50,000 and 1/100,000. The genetic condition has been found to be a deletion on chromosome 11. It is an autosomal dominant condition, in which one parent also has aniridia, in 2/3 of the cases.

Aniridia may be more than just being born without an Iris. Many people with aniridia have other eye conditions that will affect their vision. Other visual conditions may include: cataract, lens dislocation, foveal dysplasia, optic nerve hypoplasia, corneal pannus, glaucoma and nystagmus. All of these aforementioned conditions will contribute to the reduction in visual
A person is born with aniridia and possibly other eye or medical conditions. It is not a condition that occurs after birth. Most children born with aniridia have visual acuities around 20/200 and are considered usually to be legally blind. The inner structures of the eye such as the retina, optic nerve and iris are not fully developed.

Other possible medical conditions that coexist with a diagnosis of aniridia include glucose intolerance, diabetes, obesity, and WAGR syndrome. One third of those with aniridia will have WAGR Syndrome which means they will have one or more of these aspects in addition to having aniridia: Wilms tumor (tumor of the kidney usually before age eight), genitourinary abnormalities, gonadblastoma and various levels of learning challenges.

This condition will require ongoing eye care and consultation with low vision specialists for needed magnification and sun protection devices. A teacher certified in the area of visual impairment will likely be a member of the child’s educational team.

Resources:

Aniridia Foundation International is a 501(c)3 non-profit charitable organization dedicated to assisting those with low vision or blindness due to aniridia. Our members consist of those with aniridia, their families, physicians, researchers, and teachers. Aniridia Foundation International headquarters are located at the Hamilton Eye Institute at the University of Tennessee in Memphis, TN.

Aniridia Network is a support website for individuals with Aniridia and their families. The web link is [http://www.aniridia.org/conditions/aniridia.html](http://www.aniridia.org/conditions/aniridia.html)

For more information about the CO Services for Children and Youth with Combined Vision and Hearing Loss Project and the project’s services specific to Colorado children who are deafblind, please contact:

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Fact Sheets from the Colorado Services to Children and Youth with Combined Vision and Hearing Loss Project are to be used by both families and professionals serving individuals with vision and hearing loss. The information applies to children, birth through 21 years of age. The purpose of the Fact Sheet is to give general information on a specific topic. More specific information for an individual student can be provided through personalized technical assistance available from the project. For more information call (303) 866-6681 or (303) 866-6605. Updated: 10/10