

Anisocoria and Horner's Syndrome

WHAT IS THE PUPIL?

The colored part of the eye is called the iris. It is a circular muscle, similar in shape to a donut. The empty hole in the middle, which allows light to enter the eye, is called the pupil. When in a bright room or outdoors the pupil usually gets smaller (or constricts); conversely when in a dark room the pupil usually gets bigger (or dilates) to allow more light to enter the eye [See figure 1].

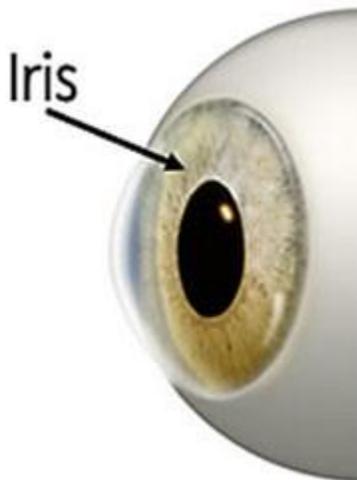


Fig. 1: The iris is the colored part of the eye.

IS IT NORMAL TO HAVE PUPILS OF DIFFERENT SIZES?

Normally the size of the pupil is the same in each eye, with both eyes dilating or constricting together. The term anisocoria refers to pupils that are different sizes at the same time. The presence of anisocoria can be normal (physiologic), or it can be a sign of an underlying medical condition.

WHEN IS ANISOCORIA NORMAL?

Up to 30% of the normal population has anisocoria. The amount of anisocoria can vary from day-to-day and can even switch eyes. Anisocoria that is NOT associated with or due to an underlying medical condition is called physiologic



anisocoria. Typically, with physiologic anisocoria, the difference in pupil size between the two eyes does not exceed one millimeter. In physiologic anisocoria, the difference in pupil size does not change under bright or dim light.

HOW DOES THE DOCTOR DETERMINE WHETHER ANISOCORIA IS DUE TO AN UNDERLYING MEDICAL PROBLEM?

Certain characteristics, such as when the anisocoria was first noted, whether it is more noticeable in bright or dim light, and whether there was an event that occurred in the past that could have caused it, will help determine the underlying cause. A complete eye examination is performed by a pediatric ophthalmologist or neuro-ophthalmologist to evaluate vision, eyelid position, how the eyes move, and the health of the front and back portions of the eyes (among other things). The doctor will evaluate the size of the pupils and how they react to bright and dim light. Based on the evaluation, the doctor may wish to perform additional tests with eyedrops or perform laboratory or radiologic testing.

HOW DOES THE DOCTOR KNOW IF THE BIG PUPIL IS 'TOO BIG' OR THE SMALL PUPIL IS 'TOO SMALL'?

One of the most important parts in the evaluation of anisocoria is determining which pupil is abnormal. If the difference in size between the pupils increases in the dark, then the smaller (miotic) pupil may not be dilating well and could be the abnormal one. On the other hand, if the difference in pupil size increases in bright light, then the larger (mydriatic) pupil may be the abnormal one because it is not getting small (or constricting) normally.

WHAT ARE SOME CAUSES OF AN ABNORMALLY LARGE (DILATED OR MYDRIATIC) PUPIL?

After trauma to the eye, the colored part of the eye (i.e. the iris tissue) can be injured causing the pupil to not get small (or constrict) to bright light normally. Another possible cause is Adie's tonic pupil syndrome. This is a condition most common in young adult females, which usually begins in one eye. The pupil is slow to react to light. Many people with this condition will also have diminished deep tendon reflexes and they can have trouble focusing at near. The condition is usually not associated with any serious conditions. Some eyedrops, nasal sprays, or other medications can have a dilating effect on the pupil. There have been cases of prescription anti-perspirant wipes that have accidentally gotten in the eye and caused temporary pupil dilation. Finally, an abnormality of the third

cranial nerve (a nerve that comes from the brain to the eye and controls eyelid position, eye movement, and pupil size) can cause an abnormality of the pupil. In this condition, there is often droopiness (otherwise known as ptosis) of the upper eyelid on the same side as the larger (dilated) pupil. In addition, the eye may not move normally, and an older child might complain of double vision. A third cranial nerve palsy can be a sign of a serious condition, and the doctor may want to order immediate testing, including imaging studies of the brain.

WHAT ARE SOME CAUSES OF AN ABNORMALLY SMALL (MIOTIC) PUPIL?

Inflammation within the eye, whether from trauma or another cause, can result in a small (miotic) pupil. Horner's syndrome also causes a small pupil in the affected eye.

WHAT ARE THE SIGNS OF HORNER'S SYNDROME?

Horner's syndrome is caused by injury to the sympathetic nerves, which are responsible for dilating the pupil and raising the eyelid on the same side of the face. In Horner's syndrome, the pupil in the involved eye is smaller and does not get bigger (dilate) as well as the other eye. The difference in pupil size between the two eyes is more noticeable under dim light. The child may have mild droopiness (ptosis) of the upper eyelid [See figure 2]. Sometimes the lower eyelid may be slightly higher than normal (known as inverse ptosis). When the upper eyelid is slightly lower than normal and the lower eyelid is slightly higher than normal, the eye may appear smaller.

If the Horner's syndrome developed during the first year of life, the colored part of the eye (iris) on the affected side may appear lighter in color than the uninvolved side (heterochromia). Sometimes, the pressure in the eye is lower in the affected eye and sometimes there is decreased sweating or flushing of the skin on the face on the affected side (anhidrosis).



Fig. 2: In Horner's syndrome, ptosis may occur.

WHAT ARE THE CAUSES OF HORNER'S SYNDROME IN CHILDREN?

Horner's syndrome is caused by injury to the sympathetic nerves that are located in the brain, neck, or upper chest on the same side as the smaller pupil. Horner's syndrome can be divided into congenital (occurring in the first 4 weeks of life) and acquired cases. Congenital Horner's can result from neck/shoulder trauma during birth and can result in injury to the shoulder, arm or hand on the same side, which is due to injury of the nerves called the brachial plexus. Often there is no known cause for congenital Horner's syndrome. Acquired cases can be due to neck trauma, neck surgery, or an abnormality in the chest, neck, or brain. In children, Horner's syndrome may be caused by a tumor called neuroblastoma, which can arise in other parts of the body and spread to affect the sympathetic nerves that control the pupil. Although rare, the risk of neuroblastoma is significantly greater with acquired Horner's syndrome than it is with congenital cases.

WHAT TESTS MAY BE CONSIDERED WHEN HORNER'S SYNDROME IS SUSPECTED?

When clinical findings point towards a diagnosis of Horner's syndrome, additional testing may be necessary. There are tests that the eye doctor may perform in the clinic to confirm a diagnosis of Horner's syndrome, in which the doctor will test the response of the pupils to different lighting conditions and certain eye drops. When Horner's syndrome is diagnosed in a child, the doctor may order additional tests including imaging studies and urine tests.

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