CHILDHOOD GLAUCOMA

FACT SHEET

(The term 'glaucoma' refers to a characteristic pattern of damage to the optic nerve)

Childhood Glaucoma describes the different types of glaucoma sometimes referred to separately as Congenital, Infantile or Juvenile glaucoma. There is usually an abnormal development of the eye's aqueous fluid outflow system. Most patients with this condition come under treatment in infancy or early childhood, however, some may not have their glaucoma diagnosed until adulthood.

Childhood glaucoma is uncommon but when it occurs the effect on vision can be devastating. Early diagnosis and appropriate treatment can make a significant difference to the child's sight. There is a large variation in severity in this subtype of glaucoma, some mild and others severe, however any vision during a child's development is worth fighting for.

There are three major types of childhood glaucoma:

1. Developmental diseases of the eye, where the aqueous fluid drainage pathways are imperfectly developed;
2. Eye diseases such as inflammation, or an injury that can lead to glaucoma and;
3. Glaucoma that follows cataract surgery as a young child (usually <1y.o.)

How does childhood glaucoma reveal itself? High eye pressure in babies stretches their softer eyeball so that the eye increases in size. The child often dislikes light and, if older, may bury its head into a parent's arms to escape light. Watering of the eye is also often noticed. Sometimes the condition is only noticed when the deepest layer of the stretched cornea splits, with the aqueous fluid entering the cornea, turning it cloudy or white. If the glaucoma is particularly severe the child may be born with cloudy corneas, indicating that the eye pressure has been significantly raised in the womb.

Assessment of the possibly-affected child can be difficult in babies or little children who usually do not co-operate, thus examination under anaesthetic is often required. In examination, the size and shape of the eye is measured, the eye pressure is recorded and the structures of the tissues in the front and back of the eye are assessed.

Once a firm diagnosis has been made and the baseline measurements are recorded, treatment can be planned. Inflammation, injuries and growths that are the cause of the glaucoma are treated specifically and these measures should lower the eye pressure.

If there is an abnormal development of the drainage pathways this may be managed firstly by drops and/or liquids by mouth to reduce eye pressure, and then by surgery. If the cornea (the window of the eye) is normal in structure and clear, a goniotomy is usually performed.

This consists of passing a special knife-needle across the front of the eye (the anterior chamber) and gently cutting the abnormal tissue that is blocking the flow of aqueous fluid from draining from the anterior chamber of the eye through the trabecular meshwork (the drainage tissues).

The drain is a ring that extends for 360° around the inside of the front of the eye. In a single goniotomy operation about 120 degrees of the abnormal tissue can be cut. If one operation does not lower the eye pressure sufficiently, a second and even a third goniotomy may be needed for each eye.

If the cornea is cloudy and the surgeon cannot see across the anterior chamber to cut the abnormal tissue, then another operation called a trabeculotomy is needed. In this procedure the tiny canal into which the aqueous should be flowing is identified, an instrument is inserted into it and delicately swept into the anterior chamber. This opens the drain into the anterior chamber, and allows the aqueous fluid an easier exit. A trabeculotomy too may need to be performed more than once to lower eye pressure effectively.

Occasionally these operations cannot be performed for technical reasons, or they do not succeed in reducing the eye pressure adequately. Then either a trabeculectomy drainage operation or the placement of an artificial plastic tube and plate on the outside of the eye is needed. Each operation depends on the condition of each eye.

The success of any of these operations has to be reviewed for the life of the child. Initially this means regular checks and sometimes with examinations under anaesthesia until the child can co-operate fully. On each occasion the structures of the tissues of the eye and the pressure are measured. The cornea should be clear and it should only be increasing in size in line with expected growth. The eye pressure should be in the normal range of 10-20 mm Hg, and the shape of the nerve fibres in the optic disc at the back of the eye should be stable.

The need for glasses is assessed and if one eye is affected more severely than the other, the child may need to wear an eye patch for a period of time each day. The end result sought is not just a normal eye pressure (although this is the first important goal without which nothing much else can be achieved) but a well-seeing eye in a happy child. Treatment of childhood glaucoma is therefore complex and care and attention is needed for the life of the patient. **There are no short-cuts.** The range of severity of these diseases is wide. Some affected children will achieve and maintain normal vision; some will achieve partial but effective sight, whereas some will lose their sight no matter what is tried for them.

There are several genes and inheritance patterns in childhood glaucoma. In general and in the absence of a family history, the chance of two unrelated parents with an affected child having another affected baby is between 3% and 15%. If other family members have glaucoma or parents are related the risk may be much higher. If a child is diagnosed as having glaucoma, then all brothers and sisters need to be examined as well. A consultation with a clinical geneticist and possibly genetic testing can be arranged to further detail these risks to other family members.