Corneal Dystrophy

Factsheet

What is Corneal Dystrophy?
The globe of the eye is made of five layers and the cornea is the transparent front portion. It is also the most sensitive structure in the body because of the density of nerves.

The cornea owes its transparency to the presence of a regular lattice structure of collagen fibres. Anything which affects this regularity results in loss of the transparency which is essential for good corneal function and health.

Corneal dystrophies form a group of rare disorders which usually affect both eyes. They may be present at birth, but more frequently develop during adolescence and progress gradually throughout life. Some forms are mild, others severe.

The Causes of Corneal Dystrophies
This group of disorders tends to run in families and the causes of most corneal dystrophies will lie in individual genetic make-up.

Effects of the three main types of Corneal Dystrophy
Although age of onset, symptoms and progression differ in the various dystrophies, most cases of corneal dystrophy fall into three well defined clinical and genetic types, classified by their inheritance pattern and appearance. These are Dominant Granular Dystrophies, Recessive Macular Dystrophy and Dominant Lattice-like Dystrophies.

1. Dominant granular dystrophy usually starts at around 5 years of age. This can be seen as small white dots in the centre of the cornea or may take the form of lines radiating from the centre. These signs can increase in size and number and by 50 years of age, opacities are visible to the naked eye.

2. Recessive macular dystrophy usually starts in the first decade of life and appears as a thin superficial corneal veil with isolated opacities when seen with a slit lamp. It is the least common type of dystrophy. Acute, short lived, attacks may be experienced and there is increasing haziness of the central part of the cornea and increasing isolated opacities.

3. Dominant lattice-like dystrophy can develop in infancy but more usually during the second decade of life. This is seen as a cobweb of fine lines which develop into a lattice-like pattern. By 40 years of age onwards, the centre of

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the cornea can become irregular with ill-defined opacity. Although in some people the pattern of progress is less severe, acute attacks are experienced which can contribute to relatively early onset of sight loss.

Among other types Fingerprint Dystrophy, Fuch's Dystrophy, Meesman Dystrophy and RET-Buckler Dystrophy are most encountered.

Genetic Inheritance
Dominant, single gene diseases result from one of a pair of matched autosomal genes having a disease and the other being normal.

With each pregnancy there is a 1 in 2 chance of the disease appearing in the offspring.

Recessive single gene disease requires both parents to carry the condition and this results in a 1 in 4 inheritance risk in each pregnancy.

Only siblings within a single generation are affected, unless members of that generation create offspring with another carrier of the specific gene.

It is valuable to seek genetic advice on all conditions, which have an hereditary cause in order to identify how this may affect individual family members. Information about local genetic services will be available from your General Practitioner or the hospital eye specialist.

Prognosis
Although there are many more forms of corneal dystrophy, essentially there are three inherited classical varieties and the progress and likely outcome varies with each.

1. Dominant Granular dystrophies are usually mild and may be unnoticed by those with the condition. In some cases sight is not affected even in later years.

2. Recessive Macular Dystrophy is a severe dystrophy which may cause considerable damage by 30 years of age. 3. Dominant Lattice-like Dystrophies can be either mild or severe and from middle-age these may cause acute attacks, capable of causing serious sight loss.

Treatment

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In some conditions corneal grafting offers a good prospect of visual improvement.

Low Vision Services

Low Vision Services exist to optimise vision by the use of techniques, aids, lighting and general information. The eye specialist will be able to advise you on the nearest Low Vision Clinic within the Hospital Eye Service where a loan service will be available on a variety of sight enhancing equipment.

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