General Optical Council Approved CET For Optometrists
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Cataract

Part 1 – Aetiology, morphology and classification

In the first of a five-part CET series looking at cataract, **Louise Stainer** and **Michelle Hanratty** describe the nature and observation of the different types of cataract. CET Module C13717, one general CET point, suitable for optometrists and dispensing opticians

Cataract is the term used to describe opacification of the lens in the eye. It is still the leading cause of blindness worldwide.[1]

Lens anatomy and physiology

The lens is a biconcave avascular structure surrounded by the lens capsule which is secreted by the cells within the lens. On the anterior surface of the lens there is a single layer of cuboidal epithelial cells. Throughout life, anterior epithelial cells divide, proliferate at the lens equator and differentiate. Cells on the posterior surface lose their nuclei during embryogenesis and elongate to form primary lens fibres. These concentric sheets of cells or lamellae push older lens fibres towards the centre of the lens. The original primary lens fibres become the fetal lens nucleus. Lines form at the anterior and posterior poles of the embryonic lens nucleus surrounding it from about eight to nine weeks of foetal development. These are known as sutures and are where the ends of the more peripheral fibre cells meet. The addition of more cell layers continues until birth, forming a Y-pattern anteriorly and an inverted Y-pattern posteriorly. After birth, suture formation becomes more complex and irregular.[2]

The structure of the lens facilitates the transmission and focusing of light onto the retina in a number of ways. The orientation of the secondary fibres is highly specific to produce a diffraction grating with destructive interference to minimise scattering of light[3] This stray light increases with age – studies have shown a significant increase after the age of 40 to 50 years.[4] This can be perceived as considerable glare – particularly in demanding visual conditions such as driving at night.

Lens transparency is achieved by degradation of all the organelles including the nucleus within lens fibres during the later stages of their differentiation. Most of the metabolic, synthetic, and active transport components in the lens are restricted to surface cells that still contain a nucleus. The survival of the fibre cells and the maintenance of lens transparency is dependent on the activity of epithelial cells and on intercellular communication via gap junctions.[5]

Structural water soluble proteins called crystallins (α, β and χ) are also synthesised within the lens fibres and account for 90 per cent of all lens proteins.[5,6] α-crystallin is made up of constantly associating and dissociating sub-units.[6] The ability of α-crystallin to act as a chaperone, suppressing the aggregation of lens fibre proteins denatured by factors including oxidation and heat hinges on its dynamic structural nature. β and χ crystallins have similarities in their structures and their association also help reduce light scatter within the lens.[7]

Glycation of α-crystallin as a result of uncontrolled diabetes reduces the efficiency of its chaperoning ability due to the formation of cross-linkages between subunits of the protein. This results in the aggregation of denatured proteins increasing light scatter and loss of lens transparency.[8]

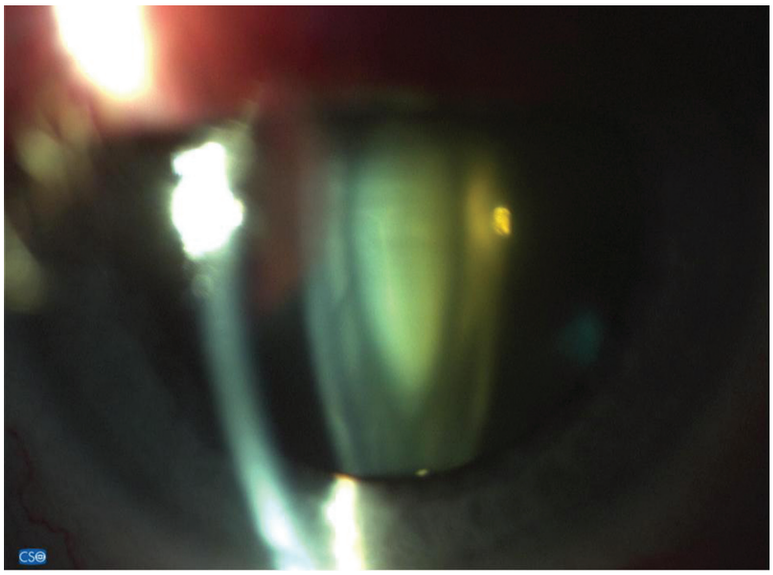

Figure 1
Nuclear cataract

TABLE 1
Classification of cataract

Aetiology	Morphology
Age-related	Nuclear
Congenital	Cortical
Secondary	Sub-capsular (anterior or posterior)
Traumatic	Polar (anterior and/or posterior)

Classification of cataracts

Cataracts can be classified in a number of ways. They can be described in relation to their aetiology or cause. Alternatively cataracts can be classified according to their location within the lens (morphology).

Morphology

Nuclear cataract

The term nuclear cataract is used to describe opacification within the embryonic and/or the foetal lens nucleus. The addition of lens fibres as the lens ages may result in compression and hardening of the lens nucleus (nuclear sclerosis) and can cause a myopic shift in the individual's spectacle prescription.[9] The lens opacities can vary greatly from small pulverulent or dust-like

opacities to total nuclear opacification[10] (Figure 1).

Higher levels of structural modification and aggregation in lens fibre proteins due to increased oxidative stress can result in overburdening and failure of the α-crystallin chaperone system. This may contribute to the formation of nuclear cataract.[6]

Brunescent cataract and nigra cataract

Brunescent cataracts differ structurally to standard mature cataracts. They are extremely hard and have a shiny brown surface. Normally they consist of a thin capsule, a cortex smaller than the equivalent in a typical mature cataract and a nucleus comprising of three structurally unique layers: a thin epinucleus, an inner nucleus and a hard amorphous central core.[11]

Nigra cataracts, or black cataracts, have a very dense structure, the majority of which is nucleus. They also have minimal cortex, and a brittle capsule.[11]

Phaco-emulsification is the most commonly used procedure for cataract surgery in the UK, generally producing successful outcomes in eyes with standard cataracts. Fortunately brunescent and black cataracts are not commonly seen in this country. They both require significantly more ultrasonic energy to break them down than normally required. Excessive energy dissipation and hard lens fragments can both cause irreparable thermal and mechanical damage to the corneal endothelium. Consequently, phaco-emulsification on these cataract types is more complex. Modified surgical techniques which involve manually breaking the lens down into several small fragments prior to endocapsular phaco-emulsification have been published.[12] It is then easier to perform phaco-emulsification inside the capsular bag without inducing serious intraoperative complications as less ultrasonic energy is required.

Cortical cataract

Age-related cortical cataracts are initially characterised by small circular and radial opacities in the equatorial cortical region of the lens.[13] These are common and have been found to occur in approximately 30 per cent of the population over 45 years of age.[14] However, at this stage these changes are outside the pupillary area and unlikely to have an impact on visual performance. The opacities can slowly progress, extending towards the lens poles and into the pupillary area. Such changes may be based on the fact that the lens fibres in the nucleus are much stiffer than those in the lens cortex and the difference increases with age. Mechanical stress between fibres at the nucleus-cortex interface can result in opacity formation.[13]

Both non-genetic and genetic factors are implicated in the development of cortical cataracts.[15]

Anterior subcapsular cataract

Integrins are a group of transmembrane glycoproteins each made up of one α-subunit and one β-subunit of varying types. Integrins bind to different extracellular matrix proteins situated on the lens capsule. They mediate interactions between lens cells and the extracellular matrix and maintain normal activity during development, cell proliferation and differentiation.

Transforming growth factor β (TGF-β) is a protein situated on lens epithelial cells. In the normal lens it is usually in its latent form. Studies indicate that certain integrins can regulate the action of TGF-β by acting in conjunction with other molecules.[16] Change, inappropriate or loss of function of integrins and a change in the microenvironment are both thought to have a role in the development of anterior subcapsular cataract.[16] Active TGF-β has a role in a significant number of cellular processes including wound healing by stimulation of extracellular matrix production. In the lens it has also been shown to cause adjacent lens epithelial cells to abnormally express an integrin subtype. The action of this receptor results in the differentiation of local epithelial cells into spindle-shaped myo-fibroblasts. These cells are found in anterior subcapsular cataracts.[17] They form sub-capsular plaques underneath the lens capsule and synthesise the contractile protein alpha smooth muscle actin (α-SMA).[16] Myo-fibroblast activity also results in other characteristics of anterior subcapsular cataract such as wrinkling of the lens capsule and an abnormal formation of extracellular matrix (ECM).[18]

Posterior subcapsular cataract

Posterior subcapsular cataracts (PSC) are characterised by cellular abnormality in the equatorial epithelial proliferating zone of the lens. An increase in the number of fibres or Wedl cells occurs in the meridional region of the lens. These cells migrate and cluster at the posterior pole.[19]

PSC has been shown to have a number of causes. Numerous studies have shown an association between use of systemic and topical steroids and formation of this type of cataract.[20] Corticosteroids are hormones with immunosuppressive and anti-inflammatory properties. They are prescribed for the treatment of many conditions ranging from asthma to organ transplants. Cataracts induced by corticosteroids tend to be bilateral. They are typically granular opacities with well demarcated margins that are located in the polar region of the posterior cortex, at the edge of the posterior capsule.[19]

Atopy is also a pre-disposing factor to developing posterior subcapsular cataract. In patients with atopic dermatitis the cataract may form and progress when the condition is active.[21] The sub-capsular opacity associated with this condition often has a characteristic shield-like appearance.

A study has shown that posterior sub-capsular cataracts cause the greatest reduction in visual function and contrast sensitivity compared to equivalent cortical and nuclear cataracts.[22] Deterioration in visual function may be due to increased light scatter which results in reduced retinal image contrast.[22]

Anterior polar cataract

Anterior polar cataract is caused by the proliferation of lens epithelial cells in the anterior sub-capsular lens. In contrast to the normal lens there is no cellular migration to the equatorial area. Several layers of proliferated cells accumulate in the anterior sub-capsule and are limited to the anterior polar region. This phenomenon is different from the mechanism of formation observed with other cataracts.[23] This type of cataract can vary in severity from a small dot to a fibrous pyramid shaped opacity.[24]

Posterior polar cataract

This is a congenital cataract which has a significant impact on the individual's vision due to its location.[25] There may be a small opacity present at birth and progression of the cataract may occur in the third and forth decade. The opacity is circular or rosette shaped and composed of denatured lens fibres; it may be associated with remnants of the hyaloid system.[26] It may become adhered to and cause weakening of the posterior capsule. Consequently, there is increased risk of a capsular tear development during surgery.[26]

Lamellar or zonular cataract

Opacity of the lamellar fibres (the secondary lens fibres) results in a lamellar cataract. Several generations of secondary lens fibres become opacified in response to an insult (such ▶

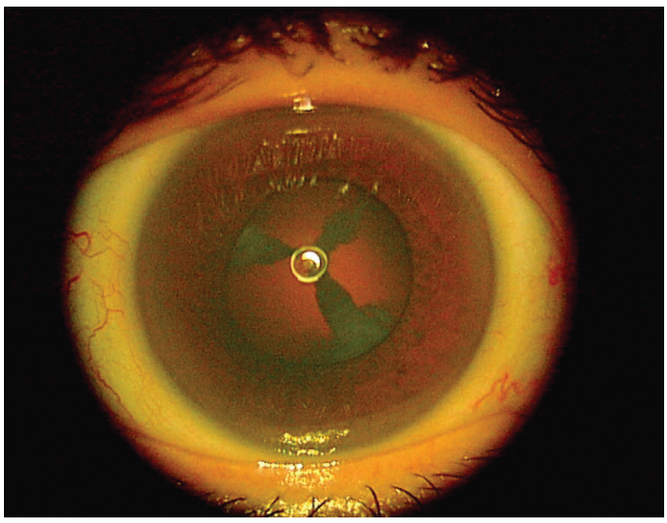

Figure 2 Dense congenital sutural cataract (courtesy of Paul Hutchence)

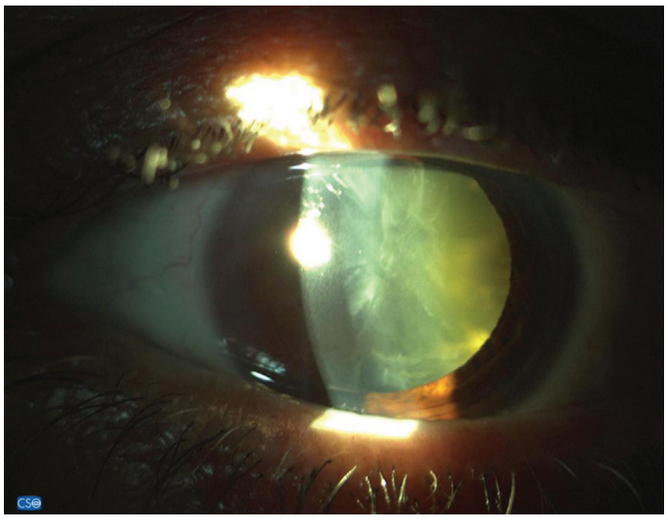

Figure 3 Mixed cataract

as maternal rubella) which coincides with the period of their greatest metabolic activity.[27,28] The rest of the lens tends to be transparent, apart from small radial liner opacities known as riders which may occur close to the equator.[28] Lamellar cataracts are often seen in several members of a family and the pattern of inheritance is usually autosomal dominant.[29] They usually occur bilaterally, but the severity may vary between the two eyes. The degree of cataract and the effect on visual acuity also differs greatly between individuals.[27] Surgical intervention is not always necessary initially, but as this type of cataract is usually progressive may be required at a later stage.[29,30]

Sutural cataract

Sutural cataract is an opacity affecting the whole or part of the anterior or posterior suture of one or both eyes (Figure 2).[31] When both sutures are affected it is known as a stellate cataract.[27] Most sutural cataracts reported are congenital and static, though an age-related cataract could also occur in later life as in any individual.[31]

Mixed cataract

In reality, classification by morphology is not always straightforward as cataracts can be made up of more than one type according to their location. This is described as a mixed cataract as seen in Figure 3.

Mature cataract

A mature cataract can lead to the development of lens-induced secondary angle-closure glaucoma as the intumescent lens physically takes up more space leading to secondary angle closure.[32] The presence of a mature cataract in the affected eye as well as angle closure and intraocular pressure elevation is highly indicative of phacomorphic glaucoma. The swollen lens impedes the passage of aqueous flow from the posterior to anterior eye by blocking the pupil.[33] Apposition between the posterior iris and anterior lens surface may also result in the iris bowing forward (iris bombe) and a progressive reduction in the iridocorneal angle culminating in angle closure.[34] Treatment must result in a rapid reduction of intraocular pressure to prevent further damage to the optic nerve, reverse cornea oedema and prevent synechiae formation.[34]

Hypermature cataract

A hypermature or Morgagnian cataract describes a cataract in which the cortical contents have become liquefied and milky and the hard nucleus is floating freely in a loose capsular bag. The nucleus normally sinks down due to gravity. If phacolysis of the cataract occurs, lens proteins leak into the anterior chamber. An antigenic reaction to these proteins involving macrophages results in inflammation.[33] These proteins and the macrophages themselves may cause phacolytic glaucoma by blocking the trabecular meshwork and impeding aqueous outflow.[33] Resultant elevation of intraocular pressure and inflammation must be managed successfully prior to cataract extraction which needs to be performed as soon as possible to increased the likelihood of a good visual outcome.[33]

Grading techniques

When grading, it is necessary to classify the cataract according to location within the lens, size and density of the opacification. The variability due to differences in subjective grading between clinicians has been greatly improved by the implementation of standardised photographic systems depicting various stages of different types of cataract. The Lens Opacities Classification System has since expanded the set of photographic standards and the Oxford Clinical Classification and Grading System has extended the set of photographic standards and diagrams respectively to allow for more accurate grading.[35,36] Such systems make it easier to determine whether a patient's cataract has progressed in the time between examinations. This, together with discussion with the patient regarding effects on quality of life and any need or wish for further intervention, helps to provide a good standard of care for the patient.

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MULTIPLE-CHOICE QUESTIONS – take part at opticianonline.net

1 Nuclear cataract generally has what effect on the spectacle prescription?
A Emmetropisation
B Increase in astigmatism
C Hyperopic shift
D Myopic shift

2 Cortical cataracts are found in what percentage of the population over 45 years of age?
A 10 per cent
B 30 per cent
C 45 per cent
D 70 per cent

3 What type of cataract has been shown to have the greatest effect on visual function?
A Nuclear
B Cortical
C Blue dot cataract
D Posterior sub-capsular

4 What typically causes cataracts with a classic shield-like appearance?
A Diabetes
B Trauma
C Radiation
D Atopy

5 The mixed cataract in Figure 3 is made up of:
A Nuclear and cortical components
B Sutural and posterior sub-capsular components
C Sutural and anterior sub-capsular components
D Nuclear components

6 Mechanical stress of lens fibres is thought to have a role in the development of which type of cataract?
A Nuclear
B Cortical
C Subcapsular
D Polar

Successful participation in this module counts as one credit towards the GOC CET scheme administered by Vantage and one towards the Association of Optometrists Ireland's scheme.
The deadline for responses is May 20 2010

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