CATARACT **MEDICAL APPENDIX**

DEFINITION

1. The term **cataract** refers to any opacity in the lens of the eye or its capsule. Many different factors may result in cataract formation.

2. The lens of the eye is composed of three layers: the **capsule**, the **epithelium** and the **lens substance.** The lens substance comprises fibres derived from the epithelium, which are formed in concentric layers as the lens develops. The lens is avascular. Its fibres are unable to divide and are incapable of inflammatory reaction, but can be affected by metabolic change. Its function is to focus light on the retina, so its transparency is essential.

3. Opacification may be caused by any factor that disturbs the critical equilibrium of water and electrolytes or deranges the colloid system within the fibres. Disturbances of the developing lens result in loss of transparency in the particular part of the lens being formed when the disturbance occurred. Once the lens is fully developed, internal or external insults result in degeneration of the formed lens fibres with opacification.

4. Cataracts may be classified according to their aetiology or their morphology, which in some cases indicates their aetiology. They may affect the capsule, subcapsular region, cortex or nucleus (the firmer centre) of the lens.

CLINICAL MANIFESTATIONS

5. The symptoms are entirely visual, the cardinal symptom of juvenile or adult cataract being progressive, painless loss of vision. Early complaints may include seeing stationary spots before the eyes (in the same position in the visual field, whatever the position of the eye) and, due to irregular refraction by the lens, seeing double or triple images with the affected eye, coloured halos around objects and glare in bright light.

6. As the opacity extends and increases in density, the acuity of central vision suffers, the deterioration depending upon the density and position of the opacity. When deterioration is progressive, vision steadily deteriorates until only perception of light remains. However, not all cataracts progress. Many reach a certain level of density and are then static for long periods. Cataracts affecting the nucleus of the lens result in increasing refractivity, and hence increasing myopia, so that a presbyopic individual may initially notice an improvement in near vision.

7. Congenital cataracts are present from birth or appear shortly afterwards.

8. Treatment is by surgical extraction of the lens. The resulting aphakia may be corrected with spectacles or, which is more usual nowadays, with an intraocular lens made of plastic or silicone. Extracapsular lens extraction is now the procedure of choice. It may be followed by posterior capsule opacification (treatable by laser). More serious complications are now uncommon. The possibilities include infection, haemorrhage, prolapsed iris, glaucoma, corneal decompensation, cystoid macular degeneration, and retinal detachment.

AETIOLOGY

AGE-RELATED OR SENILE CATARACT

9. This is the most common form of cataract. It is rare in persons below the age of 50, but almost universal to some degree in persons over the age of 70 years. It occurs equally in men and women and is usually bilateral, although often developing earlier in one eye than the other.

10. There is a strong genetic influence on its incidence and, in hereditary cases, it may appear at an earlier age in successive generations.

11. Senile cataract is usually nuclear (nuclear sclerosis), but cortical and anterior or posterior subcapsular opacities may also occur with ageing.

TRAUMATIC CATARACT

12. Although the eye has some protection from the lids and the projecting margins of the orbit, it is not exempt from injury and any of its structures may be involved. Trauma is the most common cause of unilateral cataract in a young individual. The following types of injury may cause lens opacities:

    Direct penetrating injury
    Concussion/contusion injury to the eye
    Electric shock or lightning
    Heat
    Radiation

13. Following penetrating injury or concussion/contusion injury, cataracts usually appear in close time relationship to the injury and certainly within 2 years.

14. **Direct Penetrating Injury**

    14.1. Cataract may occur following a direct wound to the lens. If the initial wound to the lens is large, opacification is rapid. In young patients, the lens substance may escape and be dissolved by the aqueous humour, invariably causing iridocyclitis, probable secondary glaucoma and sympathetic ophthalmitis. In older patients the nucleus does not dissolve in this way but the other complications may occur.

    14.2. Trauma to the lens may be unnoticed if the wound is small, for example by a thorn, needle or tiny metallic fragment. A small wound to the capsule results in entry of aqueous humour and localised opacification. At the same time irrespective of the site of the wound, there are changes in the posterior cortex which eventually develop into a rosette-shaped cataract, as in concussion cataract. If the capsular wound heals, these changes may become static, but they are usually progressive until a complete cataract is formed.

15. **Contusion or Concussion Injury to the Eye**

15.1. In a contusion injury, the cornea is thrust inwards. It may be forced against the lens and the iris. In turn these structures may be pushed backwards, striking the retina and choroid. When the concussion wave rebounds from the back of the eye, the lens and iris are thrust forwards again. This may lead to severe damage and complications, one of which is **concussion cataract**, which arises either due to the direct mechanical effect of the injury on the lens, or due to damage to the capsule causing leakage of aqueous humour into the lens. Capsular damage may result in small, discrete punctate opacities that do not progress or, if the leak is not sealed off, the cataract will progress to involve the entire lens.

15.2. Concussion cataract often has a characteristic rosette-shaped appearance. This may appear early in relation to the injury, when it may be located in the anterior or posterior cortex, or in both. A late, smaller rosette-shaped cataract may also appear in the posterior cortex up to two years after a contusion injury. Concussion injury may also lead to the development of a Vossius' ring on the anterior capsule of the lens due to imprinting of the iris pigment when the iris is forced back against the lens.

16. **Electric Shock and Lightning**

Cataracts due to electrical discharge are rare, but may develop after the passage of any powerful electric current through the body. The rate of development depends on the strength of the current and its path. Lightning passing through the head can cause cataract within days, but cataracts appearing several months after a shock are not considered to be related to the event.

RADIATION (Non-ionising and ionising)

17. **Heat**

17.1. Heat (infrared) cataract was seen industrially in workers exposed for long periods, for example metal smelting or mill operatives and glass workers.

17.2. Light of visible wavelengths from whatever source has a lesser heating effect on the eye because the normal lens is transparent to this light, so it is far less cataractogenic than is infrared.

17.3. Other heating effects are described in Paragraphs 19-22 below.

18. Ultraviolet (UV) light

18.1. UV light is classified by its wavelength: that of UVA is 315 - 400 nm (nanometers), UVB 280 - 315 nm and UVC 200 - 280 nm. All three are damaging to tissues and are present in sunlight, their intensity increasing with altitude. UVC does not reach the earth's surface. UVB, a component of both sunlight and electric arc light, is the most harmful to the lens, as it is absorbed by the lens. UVA, the predominant component of the light of sunbeds etc is almost all transmitted by the lens.

18.2. Exposure to artificial sources of UVB causes cortical lens opacities in laboratory animals, both after short-term high intensity exposure and chronic exposure. Even with very high doses that damaged the cornea, UVA did not have the same effect.

18.3. The relevant exposure for humans is the long-term chronic exposure to UV radiation in sunlight. Cataracts are more common in tropical countries, leading to the postulate that their increased prevalence is due to greater sunlight exposure. Confounding factors make the measurement of sunlight exposure very difficult, in respect of both individuals and populations.

18.4. Some studies in relatively temperate zones have attempted to quantify sunlight exposure. They have shown an association which is probably causal between high personal exposure levels and cataract, but in wider populations the increased risk solely attributable to ambient exposure was barely significant.

18.5. Generally accepted indicators of high exposure to UVB in sunlight include residence at high altitude and at latitudes closer to the equator, outdoor work and sunbathing. None of these is associated with an increased risk of cataract, once other variables are eliminated. Surprisingly, one study found that the control group had been sunbathers more frequently than had the cataract cases.

18.6. Other indicators are high prevalence of the ocular conditions pinguecula, pterygium and climatic keratopathy, which are caused by high exposure. None of these conditions is associated with an excess incidence of cataract. For climatic keratopathy, there is actually an inverse relationship.

18.7. There is no evidence of a causal association between nuclear cataracts and exposure to solar UVB.

18.8. On balance, there is not a proven relationship between the prevalence of cataract in populations and their exposure to sunlight, independent of dietary, genetic, socio-economic and other factors mentioned below.

19. **Radiofrequency electromagnetic waves (Microwaves and Radar)**

19.1. The lens of the eye is one of the organs most sensitive to heating by microwave radiation. The risk is related to frequency, power and time of exposure, but may also be cumulative from repeated low exposures and may be partly due to a property of the radiation other than its thermal effect. Although less penetrative, radar waves have a similar effect. In each case, the operation of properly shielded equipment carries no risk.

19.2. The National Radiological Protection Board (NRPB) recommends that acceptable exposures are those which:

(a) Do not raise the body temperature by more than 0.5°C, as shown by skin and rectal temperature.

(b) Do not result in local tissue temperatures exceeding 38°C for the head, 39°C for the trunk or 40°C for the limbs.

Advice is given on the limits of absorption rates (measured in watts/kg) and on limitation of exposure times to avoid such temperatures. However, these recommendations are mainly based on risk of malignant changes in tissues.

19.3. Earlier American research led to the conclusion that there is no risk of cataractogenesis with exposures of intensity less than 100 mW/cm^2.

20. **Lasers**

Lasers have a cataractogenic effect, but only with direct exposure of the eye to the beams. The effect may not be entirely thermal. Patient receiving repeated laser treatment to the retina are at risk and so, to a lesser degree, are those having photorefractive keratectomy (PRK), although this may be partly due to postoperative application of steroids (see below). Operators of laser equipment, including gunsights, are not at risk.

21. **Ionising Radiation**

Radiation cataract may be caused by direct exposure of the eye to X–rays, gamma rays and neutrons and is commonly seen after radiotherapy to tumours of the eye or orbit. It is typically central, posterior and subcapsular.

22. **Ultrasound**

Direct exposure to ultrasonic vibration may induce cataract due to the effects of both heat and high-frequency concussion, which can induce minute cavitation bubbles within cells. The effects are dependent on both power levels and ultrasonic frequency. No ill-effect has been associated with the power level normally used in medical practice, nor with the use of properly maintained equipment in other situations, such as cleaning, welding, soldering, machining or testing materials.

23. **Visual display units**

There is no association between VDU (either CRTs, which emit very small amounts of radiation, or other type) usage and lens opacification.

METABOLIC CAUSES

24. **Diabetes Mellitus**

24.1 **Senile cataracts** appear at an earlier age and may progress more quickly in patients with diabetes mellitus.

24.2 True **diabetic cataract** is a rare condition, typically occurring in young insulin-dependent (type I) diabetics, if the condition is so acute as to grossly disturb the body's fluid balance. Very high blood sugar levels cause osmotic overhydration of the lens and cataract may develop very rapidly, but may resolve completely once the osmotic balance is restored.

25. **Inborn errors of metabolism**

There is increased risk of cataract, particularly with galactosaemia, galactokinase deficiency, mannosidosis, Fabry's disease, Niemann-Pick disease type A and Lowe's syndrome.

26. **Hypocalcaemic Syndromes**

Atrophy or excision of the parathyroid glands may cause cataract, and so may neonatal hypocalcaemia.

TOXIC CAUSES

27. **Steroid-induced Cataract**

27.1 Steroids, both systemic and topical to eye and skin, are cataractogenic. Initially the cataract is posterior subcapsular, then anterior subcapsular opacities develop. The relationship between dosage and duration of treatment and the development of cataract is unclear. It is probable that doses of less than 10mg prednisone equivalent per day or treatment for less than a year do not cause cataract. Some individuals develop cataract on very low doses, probably due to greater genetic susceptibility.

27.2 Recent studies have confirmed an increased risk of both nuclear and posterior subcapsular cataracts in patients using inhaled corticosteroids, even without systemic usage. The use of inhaled corticosteroids at any time has been found to be associated with increased prevalence of nuclear cataracts. A lifetime dose of at least 2,000mg of beclomethasone among current users is associated with a relative prevalence of 5.5 for posterior subcapsular cataracts.

27.3 In most cases, steroid-induced cataracts progress slowly, even after the cessation of steroid therapy.

28. **Other drugs and chemicals**

    Cataracts may develop after the ingestion of substances including naphthalene, thallium, dinitrophenol, phenothiazines, busulphan, amiodarone and dinitrophenol, and also with the long-term use of miotic eye drops, particularly the long-acting cholinesterase inhibitors such as Eserine. Occupational exposure to mercury is a risk factor, and so is the prolonged administration of any eye drops containing mercurial preservations. About 50% of those having gold therapy for 3 or more years develop lens opacities. In most cases the opacities are not severe. Alcoholism and heavy cigarette smoking are also associated with an increased risk.

SECONDARY OR COMPLICATED CATARACT

29. Secondary or complicated cataract develops as the result of some other primary ocular disease, inflammatory or degenerative, which leads to disturbance of the nutrition of the lens.

30. **Chronic anterior uveitis (iridocyclitis)**

    This is the commonest cause of a secondary cataract. Opacification of the lens begins at the posterior pole, progressing to give anterior and posterior subcapsular opacities and eventually opacity of the whole lens. If the uveitis is controlled in its early stage, development of the cataract may be arrested. Because these cataracts affect the visual axis of the lens, visual impairment is often marked.

31. **Inflammatory conditions of the posterior segment of the eye**

    Posterior uveitis and inflammatory conditions of the retina typically cause posterior subcapsular opacities.

32. **Other ocular causes**

    Hereditary fundus dystrophies (for example, retinitis pigmentosa), high myopia (but not simple myopia), retinal detachment and acute congestive angle-closure glaucoma are associated with increased risk of cataract.

HEREDITARY/CONGENITAL CAUSES

33. **Intrauterine infections.** Congenital **rubella** is associated with cataract in about 15% of cases. Other intrauterine infections that may be associated with neonatal cataract are toxoplasmosis, cytomegalovirus, herpes simplex and varicella.

34. **Maternal drug ingestion** during pregnancy, notably thalidomide and steroids, may cause congenital cataracts.

35. **Congenital syndrome.** Cataracts feature in Down's syndrome, Werner's syndrome, Turner's syndrome, Edward's syndrome, Rothmund's syndrome, neurofibromatosis and myotonic dystrophy.

36. **Inborn errors of metabolism:** see Paragraph 25 above.

37. **Hereditary**: one third of all congenital cataracts are hereditary and not associated with any of the metabolic or systemic disorders described. The mode of inheritance is usually dominant.

OTHER CAUSES

38. **Atopic cataracts** may develop in individuals with severe atopic eczema/dermatitis, and in other severe skin disease such as scleroderma.

39. **Developing countries.** The incidence of cataract in developing countries is high, but the reasons for this have not been established. Associated factors are mostly those involved with poverty, including severe diarrhoea (21-fold relative risk with more than one episode in India), vegetarianism, low protein intake, total calorie malnutrition and widowhood. An association with diarrhoea was also found by a case-controlled study of people aged 50 to 79 in Oxfordshire.

40. **Severe dehydration.** Individuals who have suffered severe dehydration in childhood or as young adults may develop cataracts at an early age.

MORPHOLOGY

41. Cataracts may also be classified according to their morphology. Some aetiological factors result in a cataract involving a specific part of the lens:

    41.1. **Capsular cataracts** may be congenital or acquired. Causes include Pseudo-exfoliation syndrome (secretion of fibrillogranular material throughout the anterior segment of the eye), gold, chlorpromazine, trauma (Vossius' ring) and in association with posterior synechiae in anterior uveitis.

    41.2. **Subcapsular cataracts** may be anterior or posterior. Anterior subcapsular cataracts occur in acute angle closure glaucoma, Wilson's disease, miotic therapy and amiodarone treatment. Posterior subcapsular cataracts may be complicated (secondary) or age-related. Other causes include myotonic dystrophy, steroids and irradiation.

    41.3. **Nuclear cataracts** may be age-related or congenital, for example maternal rubella or galactosaemia.

    41.4. **Cortical cataracts** are age-related or congenital. The latter form is very common, does not usually interfere with vision and may be white or deep blue in colour.

    41.5. **Lamellar cataracts,** sandwiched between a clear nucleus and cortex, are invariably congenital.

    41.6. **Sutural cataracts** are common, congenital and usually familial. They are of no visual significance.

41.7. A **mature cataract** is one in which all of the lens protein is opaque, whereas an **immature cataract** has some remaining transparency. If the lens takes up water, it is **intumescent.** If the cortical proteins are liquefied, the cataract is **hypermature.** A hypermature cataract with the lens floating freely in the capsular bag is called a **morgagnian cataract.**

CONCLUSION

42. A cataract is an opacity in the lens of the eye or its capsule. Cataract may be associated with a wide variety of factors, which are listed above.

43. Senile cataract is by far are the most common form of cataract and is due to degenerative changes in the lens fibres, such changes being a consequence of the ageing process, with genetic factors having an important role.

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