British Journal of Ophthalmology, 1977, **61**, 135–140

Cataract in childhood: photographic methods in assessment

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SUMMARY The slit-lamp examination of the child's lens with cataract can yield useful prognostic signs, especially when this is supplemented by slit-image photography and measurement. A normal lens size for the subject's age is a good prognostic sign and is usually associated with a static cataract or with very slow deterioration in a developmental cataract. A small lens and absence of the anterior subcapsular clear zone are each associated with progressive cataract. Nuclear lamellar cataracts are shown to become smaller in diameter with time, which favours medical treatment with mydriasis.

The technique of quantitative slit-image photography of the lens (Fig. 1) (Brown, 1972a, b) provides information on the dimensions of the lens, including sagittal width and radius of curvature of the surfaces and of dimensions and situation of the cataract. This information is not easily obtained by any other technique, and the photographs have also the advantage of providing a permanent record with which to compare future examinations. A number of features in the photographs have prognostic value, in particular the observation of the anterior subcapsular clear zone and of lens size. This technique has also provided new information on normal lens growth and on the natural history of cataracts.

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Lens growth

The lens grows at a steady rate throughout life, at least between the ages of 10 and 80 (Fig. 2) (Francois, 1959; Brown and Tripathi, 1974), but there are insufficient data below the age of 10 to say whether the growth rate is any different then. The growth is mainly cortical, and there is very little change in the width of the nucleus with age (Fig. 2), when the limit

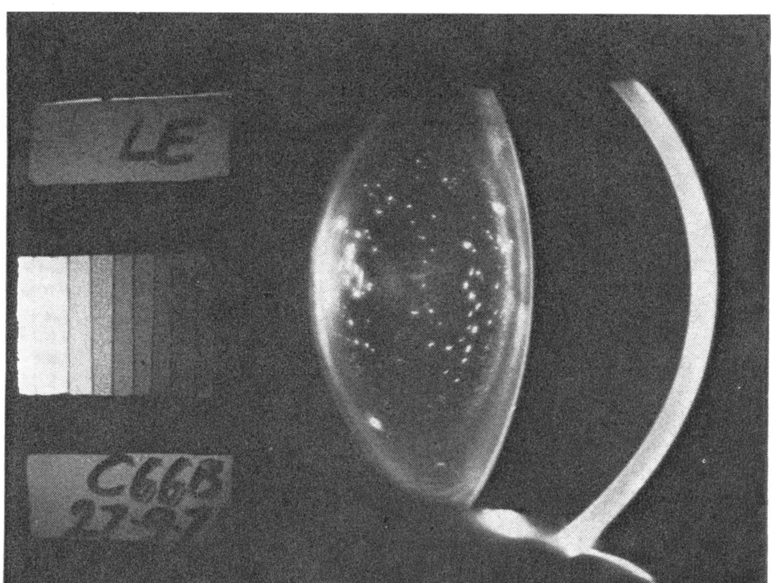

Fig. 1 *Slit-image photograph of the lens of a year-old girl showing developmental cataract*

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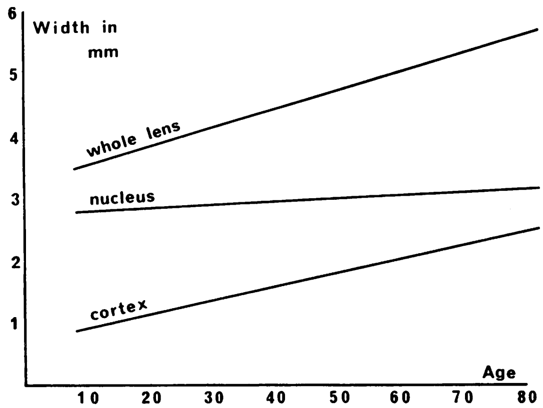

Fig. 2 *Graph showing the growth in sagittal width of the normal lens with age*

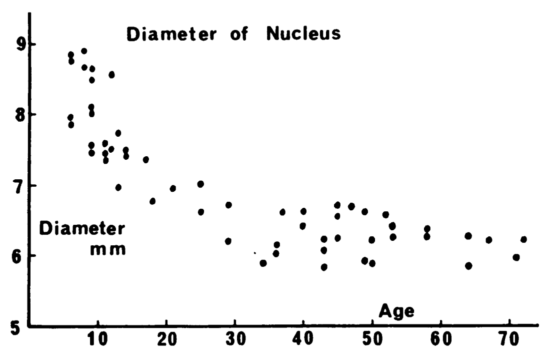

Fig. 3 *Graph showing the reduction in equatorial diameter of the nucleus with age*

of the nucleus is defined by the sudden change in optical density which is seen in the slit-image pictures. While the width of the nucleus remains nearly constant, the equatorial diameter is actually becoming less with age (Fig. 3) and is most rapid in childhood. This has an effect on the size of nuclear cataracts, causing them to become smaller with age.

A small lens size for the subject's age is seen in lenses developing certain forms of cataract, in particular progressive anterior or posterior subcapsular cataract (Brown and Tripathi, 1974). A normal lens size is seen in static cataracts such as congenital nuclear cataracts and congenital anterior and posterior polar opacities. Thus, a normal lens size in an eye with cataract is a good prognostic sign. This is particularly valuable in developmental cataracts, which are discussed separately below.

A small lens size can also be seen following trauma (Fig. 4) either blunt or perforating. This is commonly seen in those lenses which have been examined within a few days of the trauma. The size recovers to normal in those lenses in which recovery of growth takes place (Brown, 1976). In these lenses the opacity remains static in size and sinks into the lens with time. The lenses which fail to recover to normal size are liable to deteriorate to mature cataract, which in children can be a very rapid process reaching maturity within a few weeks.

Lens curvature

The radius of curvature of the lens surfaces is easily measured from the slit-image pictures (Brown, 1973) but is very difficult to assess with the slit lamp alone.

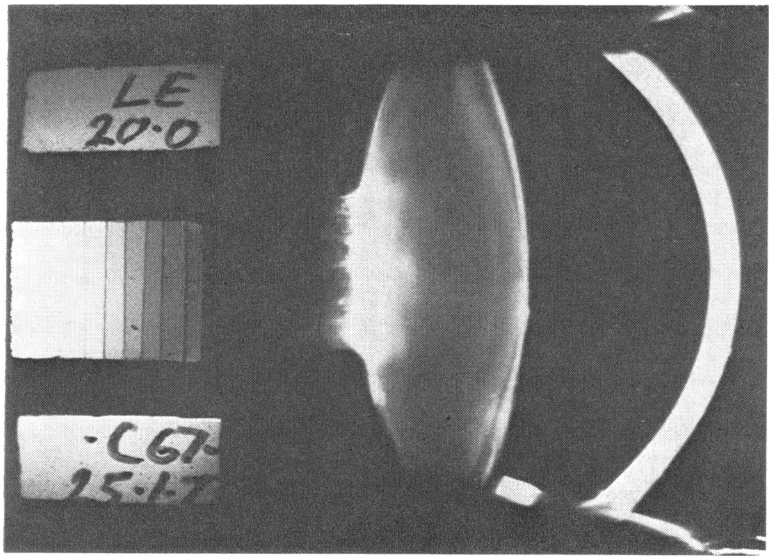

Fig. 4 *A child's lens affected by non-perforating trauma showing small size with flattening of the posterior surface*

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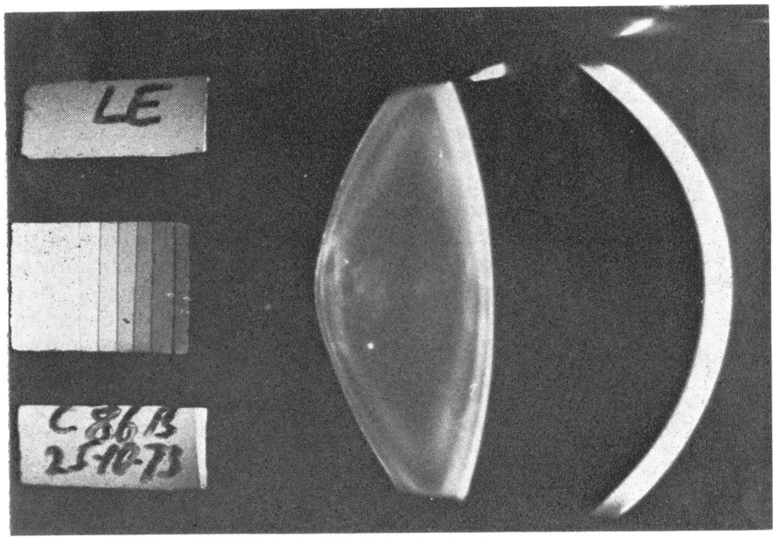

Fig. 5 *The lens of a 14-year-old child showing posterior lenticonus*

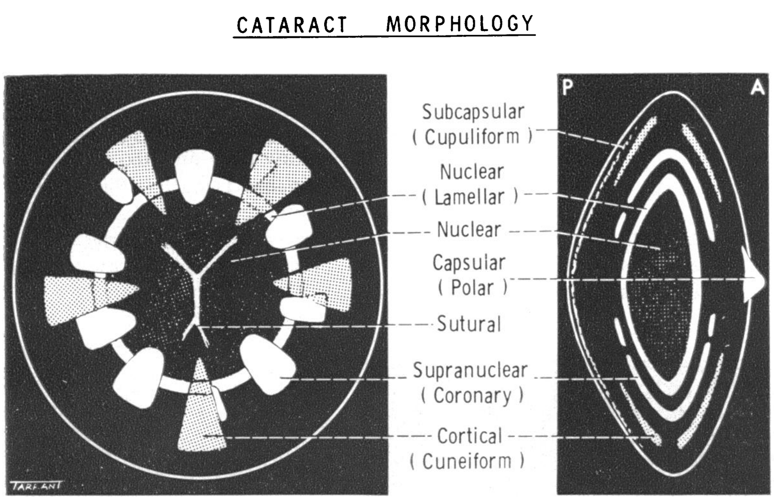

Fig. 6 *The morphological classification of cataract*

An abnormal lens curvature is associated with childhood cataract in posterior lenticonus. The measurement of the posterior radius of curvature is therefore a useful exercise in children with posterior polar cataract and in their siblings (Fig. 5) and may help in the identification of Alport's syndrome.

Cataract situation

The site and character of the cataract can be determined quite well by the clinical use of the slit lamp, which is sufficient for assigning the cataract to one of the various morphological groups of cataract (Fig. 6). Slit-image photography is important in defining the exact depth of the cataract, as measured from the capsule. Many forms of acquired childhood cataract are initially subcapsular—traumatic (Vogt, 1922), hypoparathyroid (Goldmann, 1929), diabetic, complicated, steroid, and radiation induced. It would appear that the youngest nucleated lens fibres which lie in the subcapsular clear zone are the most vulnerable to noxious influences, except in radiation cataract, where the fault appears to be primarily in the lens epithelium. When the noxious influence is not severe and is present for a limited time, the lens recovers normal growth and the opacities are seen to sink into the lens (Fig. 7). The rate at which the opacities sink in is actually faster than the lens is growing, which implies that the lens is undergoing central compression while growing by surface

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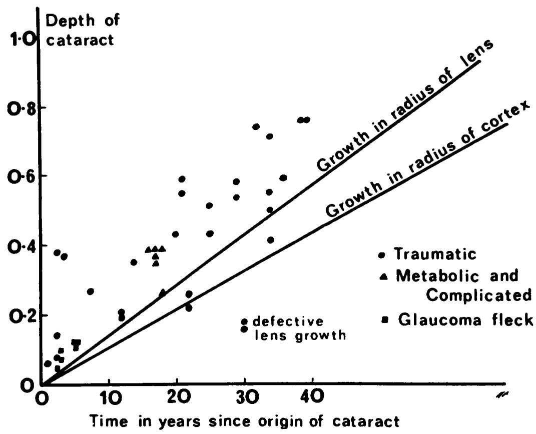

Fig. 7 *Graph showing the depth of lens opacities compared with the rate of growth of the lens*

accretion. The rate of movements is greater nearer the surface (Brown, 1976). The compression effect appears to be largely a cortical phenomenon, and cataracts never sink into the nucleus (Nordmann, personal communication), which I confirm.

Cataract size

STATIC CATARACTS

Cataracts due to an external influence such as trauma or metabolic disturbance which is short-acting tend to remain constant in size, though they do change in position as described above. Congenital cataracts, which are necessarily nuclear, since there is no cortex present at birth, are usually regarded as static but now may have to be considered as shrinking cataracts.

INCREASING CATARACTS

A cataract may increase by (i) the laying down of new lens fibres, some of which are opaque at the time of formation, as in developmental cataracts; (ii) the spread of the cataract into previously clear lens fibres, which is seen typically in senile cuneiform cataract and in children with severe traumatic cataract and in the final stage of maturation of developmental or complicated cataract; and (iii) the laying down of granular material in the subcapsular clear zone by a lens which has lost the ability for normal growth, which is seen in complicated cataract, steroid cataract, and radiation cataract, and implies the production of defective material by a diseased epithelium which has ceased normal mitosis (Brown and Tripathi, 1974). These lenses are commonly small for the subject's age. Slit-image photography is useful in identifying progressive cataract, particularly in measuring lens size and in recording the state of the anterior subcapsular clear zone. Attention has already been directed to the loss of the clear zone (Brown and Tripathi, 1974), which signifies a progressive cataract, and it is also seen in developmental cataracts which have changed from the gradual addition of new opacities to a more rapid form of deterioration.

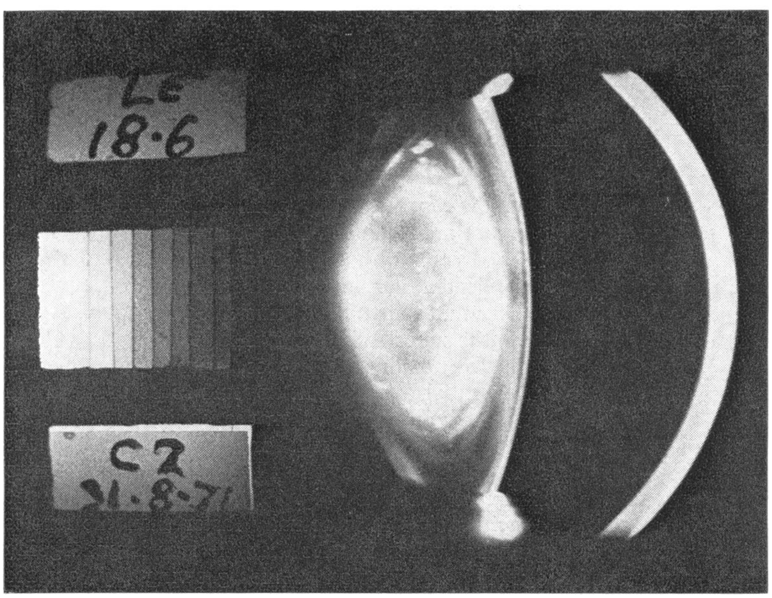

Fig. 8 *A congenital lamellar cataract in a girl aged 9*

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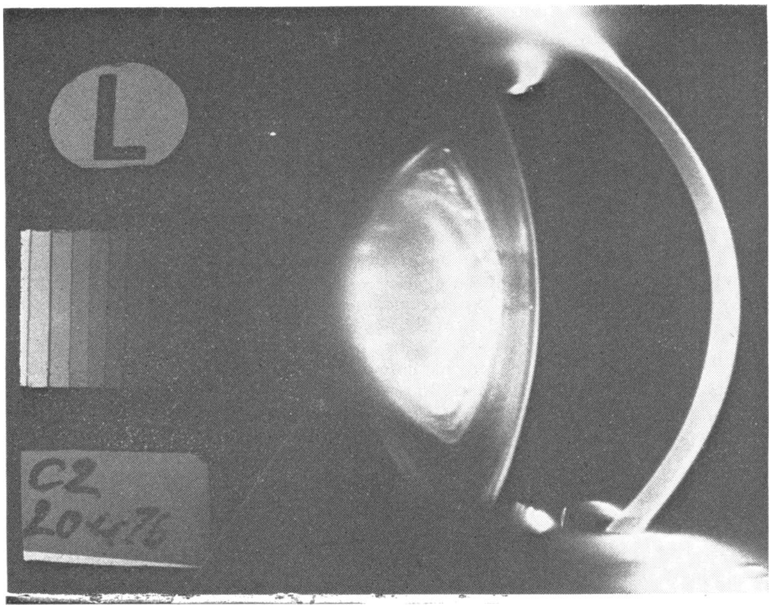

Fig. 9 *The same eye as in Fig. 8 at the age of 14. (The other eye had a cataract extraction and is amblyopic)*

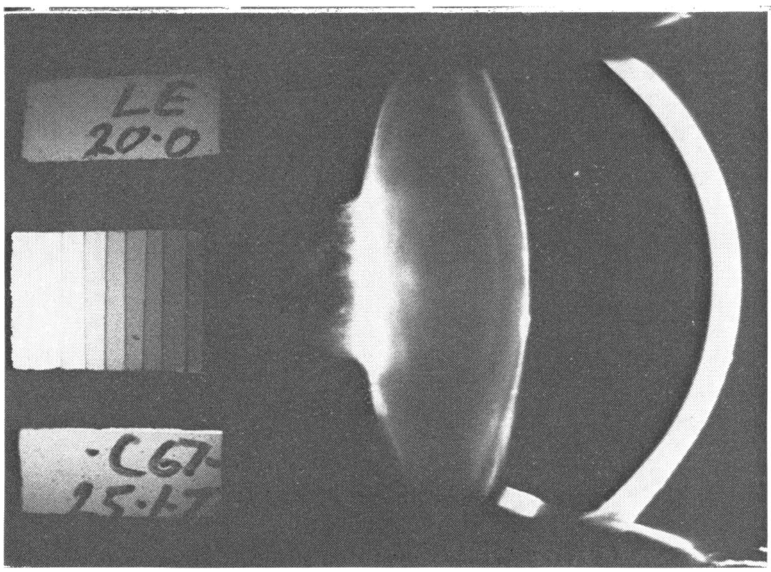

Fig. 10 *A developmental cataract which has reached the stage of rapid deterioration with loss of the anterior subcapsular clear zone*

SHRINKING CATARACTS

After the correction of an acute metabolic disturbance causing cataract, such as diabetes, there is some improvement in lens clarity and reduction in lens swelling, but no true reduction in the size of the cataract. A true reduction in cataract size probably occurs regularly in congenital nuclear cataracts, which was first suggested by the observation (Fig. 3) that the equatorial diameter of the nucleus becomes less with age. Partial confirmation has been obtained from two families with hereditary nuclear cataract, which is seen to be smaller the older the individual. Reliable confirmation (Figs. 8 and 9) is now to hand in a single lens, which has had the longest follow-up (5 years) and in which the cataract is measurably shrunken during the period of observation.

Some specific types of cataract

CONGENITAL NUCLEAR CATARACTS

The classical lamellar cataracts become compressed with time so that the affected lamella becomes

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thinner and smaller in diameter. The situation is only one of deterioration when developmental opacities are superadded. A clear cortex, clear subcapsular clear zone, and normal lens size should be sought as good prognostic signs. Since these cataracts commonly improve with time, medical treatment with mydriatics should be preferred unless the diameter of the cataract is initially as large as the dilated pupil.

POLAR CATARACTS

These may be flat discs or conoid (pyramidal) projections from the poles which are present from birth. These cataracts commonly change little with time. As the lens grows, further discs of opacities are commonly deposited beneath the polar opacity; they do not interfere further with vision except when their diameter is greater, and this does sometimes occur to the extent of needing extraction.

DEVELOPMENTAL CATARACTS

These hereditarily determined cataracts (Fig. 1) are commonly represented at birth by a nuclear cataract to which are gradually added punctate opacities and later coronary opacities with the laying down of a mixture of clear lens fibres and opaque fibres as the lens grows. This results in a very gradual deterioration, but may take a sudden change for the worse with failure of the subcapsular clear zone and posterior subcapsular cataract (Fig. 10). Most patients will not need a cataract extraction until adult life. The lens size is normal for the subject's age except in those lenses which are deteriorating rapidly which are small for the subject's age.

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