COMPLICATED CATARACT EXTRACTION IN FUCHS'S HETEROCHROMIC UVEITIS*†

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CATARACT is a well-recognized complication of Fuchs's heterochromic uveitis. Coles (1964) considers it to occur in 15 per cent. of cases. Perkins (1962) suggests that 75 per cent. of all cases are eventually affected.

Many authorities consider that cataract extraction in this type of case is an uncomplicated procedure with a smooth post-operative course. Duke-Elder (1940) considered that "cataract extraction is easy and the operative prognosis good". Coles (1964) said that cataract extraction was well tolerated. Stallard (1965) pointed out that the intracapsular operation is well tolerated and healing of the wound proceeds smoothly. Huber (1961) suggested that the removal of the cataract was usually without complication.

Hyphaema following paracentesis of the anterior chamber in cases of heterochromic uveitis has been recognized by the continental authors Amsler and Verrey (1946), François (1954), and Franceschetti (1955). This sign was recently discussed by Goldberg, Erozan, Duke, and Frost (1965). Amsler (1948) re-stated his opinion that a post-paracentesis filiform hyphaema was pathognomonic of Fuchs's heterochromic uveitis. Becker and Shaffer (1965) said that lens extraction in this type of case was well tolerated in spite of recurrent hyphaema. Stallard (1965) pointed out that a post-operative hyphaema in cases of extraction of complicated cataract might be slow to absorb.

Huber (1961) noted a peculiarity of the disease whereby there is a not infrequent abrupt rise in pressure following anterior chamber puncture.

Secondary glaucoma as a late complication of heterochromic uveitis has been increasingly recognized recently since its further description by Huber (1961). Although described from time to time by many authors since Fuchs (1906), it is only recently that it has become apparent that a quiet and insidious type of glaucoma supervenes in a considerable number of cases, as Duke-Elder (1940) suggested. De Rosa (1959) found that secondary glaucoma occurred in 14 per cent. of cases and that the visual results of drainage operations were poor. Coles (1964) considered that it developed in 15 to 25 per cent. of all cases, usually in eyes with cataract; when present the glaucoma frequently disappeared after cataract extraction, but usually responded well to non-surgical therapy if cataract extraction was not indicated. Huber (1961) also suggested a 25 per cent. incidence, more frequent in bilateral cases.

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The object of the present investigation is to challenge the view that the operative prognosis in cases of cataract complicating heterochromic uveitis is usually good. While the belief that operation is technically straightforward is not contested, it is pointed out that extraction can be followed by severe post-operative complications, including intra-ocular haemorrhage, persistent vitreous opacity, and a particularly serious rise in ocular tension.

Selection of Patients and Case Histories

It recently became obvious that patients undergoing extraction of complicated cataracts secondary to Fuchs's heterochromic uveitis did not always experience a smooth post-operative course.

A search of the recent records was instituted and the following nine cases were considered to merit description.

The scope of this investigation is only to establish the occurrence of complications following cataract extraction in patients with heterochromic uveitis. No attempt is made to assess the incidence of these complications. However, all the cases described except No. 1 have undergone operation within the past 3 years.

It is important to note that the extractions were performed by five different surgeons using differing techniques.

Case Reports

Case 1, a man aged 38, presented with typical left heterochromic uveitis and a mature left cataract. He worked as a miner and cataract extraction was indicated in order to increase his field of vision.

An elective extracapsular extraction after a von Graefe section was performed with full iridectomy. The operation was uncomplicated.

A small post-operative hyphaema was followed on the fifth day by a total hyphaema. Also the nasal pillar of the iridectomy prolapsed but was adequately covered by the conjunctival flap and was allowed to remain undisturbed. The hyphaema resolved spontaneously over the following 2 weeks and a large vitreous haemorrhage was revealed.

The vitreous cleared only slowly over the next 5 months, at the end of which time the corrected visual acuity was 6/12; 3 years later a marked vitreous haze was considered responsible for a corrected visual acuity of only 6/18. The optic disc was normal and the tension not raised.

Case 2, a man aged 50, had been attending for some years with right heterochromic uveitis. Increasing lens opacity developed and when the cataract was mature, extraction was considered advisable. The visual acuity was light perception with accurate projection. The tension of the eye was normal.

Intracapsular extraction was performed without difficulty, using keratome and scissors section with preplaced sutures, peripheral iridectomy, and α -chymotrypsin zonulolysis. The anterior chamber was reformed with air. The operation was straightforward.

On the first post-operative day a gross hyphaema was present, with increase in ocular tension. Paracentesis and evacuation of blood clot was performed on the fifth and tenth post-operative days. With clearing of the anterior segment, it was apparent there was also a large vitreous haemorrhage.

One month later the anterior chamber appeared clear on slit-lamp examination, but gonioscopy showed blood in the angles. The angle was open except in the area of the paracentesis where peripheral anterior synechiae were present. The tension remained raised (32 mm. Hg) for 4 months post-operatively, at the end of which time a spontaneous fall to below 20 mm. Hg occurred. A marked vitreous haze persisted, however, and the corrected visual acuity was 6/12.

Case 3, a woman aged 44, attended with right heterochromic uveitis for 7 years before operation. The initially posterior cortical lens opacity progressed to a mature cataract, which was removed for cosmetic reasons. The ocular tensions were recorded as normal before operation.

Straightforward intracapsular extraction was performed after keratome and scissors section and complete iridectomy. Enzyme zonulolysis was not used. A small hyphaema was present for 3 days post-operatively. It resolved uneventfully.

For 2 months after operation an active anterior uveitis persisted with tensions of above 30 mm. Hg (applanation). With decrease of activity of the uveitis, the tension increased to 50 mm. Hg and the optic disc showed cupping. Corrected visual acuity at this stage was 6/12. Gonioscopy showed the angle occluded by peripheral anterior synechiae with debris all round.

The secondary glaucoma was not controllable by medical therapy and a Scheie-type drainage procedure was performed 4 months after the extraction. Operation was followed by a moderate hyphaema associated with a shallow anterior chamber and choroidal oedema. These complications subsided spontaneously and the tension was adequately controlled.

However, by this time there was gross cupping and advanced field loss. The corrected visual acuity was only 3/60.

Case 4, a woman aged 34, presented with left heterochromic uveitis and a mature left cataract. The ocular tensions were 17 mm. Hg in the right eye and 37 mm. Hg in the left (applanation). The angles of both eyes were open and appeared normal.

Over a period of 4 months the tension of the affected eye showed large swings of 20 to 60 mm. Hg (applanation).

Left cataract extraction with complete iridectomy was performed. Intracapsular extraction was attempted but the lens capsule ruptured. However, most of the capsule appeared to have been removed.

Post-operatively a heavy flare and gross vitreous haze were present. The tension increased to 36 mm. Hg over the next 4 weeks. The optic disc was very poorly seen but appeared to be normal.

Over the first six post-operative months, the tensions fluctuated from 26 to 44 mm. Hg. Gonioscopy showed extensive peripheral anterior synechiae. The corrected visual acuity was 6/18 and the marked vitreous haze precluded any further improvement.

A year later the disc was markedly cupped. Advanced field loss was present. The ocular tensions were barely controlled at levels of 20 to 25 mm. Hg on maximal medical therapy.

Case 5, a man aged 55, attended for 25 years with right heterochromic uveitis. When last seen as an out-patient a year before operation, he had a mature right cataract and normal tensions.

He presented as an emergency with right acute secondary glaucoma of a lens-induced type.

Emergency intracapsular extraction was performed after hypotensive therapy with oral glycerol. The operation was uncomplicated; a peripheral iridectomy was performed and enzyme zonulolysis was used.

On the first post-operative day a scattered hyphaema was present. This cleared only to recur on the fourth day. After further clearing a more severe hyphaema associated with a slight vitreous haemorrhage occurred on the tenth day. The media appeared clear 2 weeks later.

Two months post-operatively the corrected visual acuity was 6/6 in the right eye. The ocular tensions were 20 mm. Hg in the right eye and 15 mm. Hg in the left (applanation).

Gonioscopy of the right eye showed an open angle with peripheral anterior synechiae inferiorly. A few fine abnormal vessels were present below.

Case 6, a man aged 42, presented with heterochromic uveitis and immature cataract. The ocular tensions were 14 mm. Hg in the right eye and 40 mm. Hg in the left (applanation). Gonioscopy showed open angles with no peripheral anterior synechiae. There were keratic precipitates in the angle of the left anterior chamber.

A combined cataract extraction and iris inclusion drainage operation was performed the following week after acetazolamide hypotensive therapy. The lens capsule ruptured during delivery despite α -chymotrypsin zonulolysis. However, the capsule remnants appeared to have been completely removed.

Post-operatively for a week a moderate hyphaema and marked vitreous opacity were present.

One month post-operatively the anterior chamber was clear but marked vitreous opacity persisted with raised ocular tension. The drainage procedure did not appear to be functioning freely and acetazolamide and pilocarpine therapy was required to maintain the eye in a normotensive state.

It was a year post-operatively before the vitreous became clear. At this stage the disc was cupped; there was gross loss of nasal field and the corrected visual acuity was 6/24. The drainage bleb was encysted. Pilocarpine and acetazolamide therapy was necessary.

Two years later the drainage bleb was reconstructed. The conjunctiva was noted to be abnormally thickened around the bleb. The tension, however, remained raised and the patient required long-term pilocarpine therapy.

Case 7, a man aged 45, attended for years with left heterochromic uveitis and increasing lens opacity showing the typical clinical signs. The iris vasculature was unusually easy to see because of marked iris atrophy. The tensions were normal. Cataract extraction was performed for optical reasons, particularly to increase the field of vision. Intracapsular extraction using the irisophake with full iridectomy was performed. At operation the conjunctiva appeared to be abnormally thickened and hyperaemic. The operation was straightforward.

Post-operatively a moderate hyphaema and a marked vitreous haze were present.

The hyphaema persisted for 2 weeks with a moderate rise in tension to 30 mm. Hg; the tension returned to normal 2 weeks after clearing of the anterior chamber and has since remained normal.

However, owing to a marked persisting vitreous haze, the corrected visual acuity was only 6/12 after 6 months observation.

Case 8, a woman aged 66, attended for 14 years with left heterochromic uveitis and increasing lens opacity. Eventually the cataract appeared to be hypermature and the ocular tensions, which had previously been recorded as normal, were 20 mm. Hg in the right eye and 25 mm. Hg in the left (applanation). Gonioscopy showed normal angles but that in the affected eye was rather narrower.

Cataract extraction was performed by the intracapsular method with peripheral iridectomy and keratome and scissors section. Enzyme zonulolysis was used. It was noted at operation that the conjunctiva appeared thickened and bled easily. On evacuation of the anterior chamber after keratome section, haemorrhage was observed to arise from the angle below to be followed immediately by many haemorrhages arising multi-centrally around the iris. The operation was straightforward.

Despite three sutures, on the first post-operative day, a small iris prolapse was noted in the temporal side of the section. The anterior chamber was clear. On the second post-operative day the prolapse was much larger and was abscised with a small vitreous loss. The iridectomy was completed. Following this operation the anterior chamber remained clear but a moderate vitreous haze was present.

The tension of the eye showed a gradual rise till on the 14th post-operative day the readings were 18 mm. Hg in the right eye and 36 mm. Hg in the left (applanation).

4 months later the tension remained raised to this level despite acetazolamide therapy. The wound was ectatic and a gross vitreous haze was present. The corrected visual acuity was only 2/60.

Case 9, a girl aged 13, presented with right heterochromic uveitis and early lens opacity. The tension in the right eye was moderately raised to 25 to 30 mm. Hg (applanation). Gonioscopy showed fundamentally open angles with some abnormal tissue and many abnormal radial and circular vessels. The tension was controlled on gutt. pilocarpine 1 per cent. twice daily. 3 years later the lens was opaque and was subluxated.

Vectis extraction and complete iridectomy were performed after a von Graefe section. There was a slight vitreous loss.

A marked vitreous haze persisted for 4 weeks after surgery, at which stage the ocular tension was found to be 40 mm. Hg (applanation). The glaucoma was uncontrolled by medical means and cyclodialysis was performed 3 months later. Following this operation there was a large hyphaema with vitreous haemorrhage.

With clearing of the anterior chamber, the tension remained elevated and a Scheie-type drainage operation was performed 4 months later. A small hyphaema was again present post-operatively.

The fistula did not drain freely and the patient has since been maintained on long-term acetazolamide therapy.

A marked vitreous haze persisted throughout. The disc remained normal. However, the eye was amblyopic and the corrected visual acuity was only counting fingers at 2 metres.

Recent gonioscopy showed the angle basically open with multiple peripheral anterior synechiae. A remarkable finding was that the gonioscopy procedure appeared to rupture a blood vessel and pulsatile haemorrhage was observed for some minutes from a small vessel in the angle above.

It is worth mentioning in passing that five of this series of nine patients showed skin sensitivity to atropine. Presumably this is due simply to the common practice of prolonged and probably unnecessary use of atropine drops in this type of case.

Discussion

Nine patients underwent extraction of the cataract associated with Fuchs's heterochromic uveitis. In each case a complication, or several complications of various degrees of severity, occurred. These complications appear to fall into three main groups:

- (1) An intra-ocular haemorrhage occurring immediately post-operatively.
- (2) A persistent vitreous haze.
- (3) A serious elevation of ocular tension which persists for a considerable time post-operatively and is particularly resistant to therapy.

Hyphaema occurred in seven of the nine patients following cataract extraction. This complication occurred on ten occasions after thirteen operations among this group. Vitreous haemorrhage occurred in four of the nine patients.

The hyphaema was slight in three cases, moderate in two, and severe in two. The last two patients also suffered marked vitreous haemorrhages.

Apart from being mentioned by Becker and Shaffer (1965), intra-ocular haemorrhage has not been specifically described previously in relation to cataract extraction in heterochromic uveitis.

This complication is of particular interest in that it should be quite predictable. Amsler and Verrey (1946), François (1954), and Franceschetti (1955) have mentioned a filiform hyphaema following paracentesis of the anterior chamber in this type of case. The first author considers this sign to be pathognomonic. Goldberg and others (1965), in an important paper on the cytopathological and histopathological aspects of heterochromic uveitis, mention that they found this sign of post-paracentesis hyphaema in three of four cases before cataract extraction. They noted a post-operative hyphaema in only one case. In this connexion, one of our cases is of interest in that, after evacuation of the anterior chamber before extraction, haemorrhage was observed to occur initially at the base of the iris below and then to arise multi-centrally around the iris. Another of our cases showed the ease with which intra-ocular haemorrhage can occur, as a haemorrhage from an angle vessel was precipitated merely by gonioscopy. Becker and Shaffer (1965) noted that hyphaema was a frequent complication of minor trauma.

As regards the aetiology of the haemorrhage, this could well be bleeding from fragile sclerotic uveal vessels. Histopathologically, Goldberg and others (1963) found that, in iris specimens in cases of heterochromic uveitis, there was definite hyalinization of the arterioles in addition to the normal anatomical "cuffing".

Duke-Elder (1940) mentioned that the arterioles were thickened. Makley (1956) noted in identical twins with heterochromic uveitis that, pathologically, instead of the thick walls associated with normal iris vessels, the walls were unusually thin and irregular and seemed in places to be fragmented.

Huber (1961) thought that the most important diagnostic sign of heterochromic uveitis was vascularization of the angle with radial or looping vessels.

Bleeding from abnormal vessels in the filtration angles, as discussed by Kimura, Hogan, and Thygeson (1955), could be an aetiological factor in our cases although abnormal vessels were seen in only two of our nine cases.

Henkind (1964) discussed the difficulty in assessing "abnormal" vessels of the angle in various ocular disorders. He pointed out that the radial vessels in the trabecular zone which Franceschetti (1955) and Vannini (quoted by Franceschetti) mentioned as occurring frequently in heterochromic uveitis, might also occur in non-diseased eyes.

Chatterjee (1960) classified normal and abnormal radial gonio-vessels.

The possibility of decreased sympathetic tone being responsible for post-paracentesis hyphaema was mentioned by Amsler and Verrey (1946) but was considered unlikely as this sign did not occur in neurogenic heterochromia. Also this would appear to be excluded by the work of Goldberg and others (1965), who found different cytopathological aspects of the cells from the anterior chamber in cases of Fuchs's heterochromic uveitis and in cases of heterochromia secondary to Horner's syndrome and status dysraphicus.

Vitreous opacity, a well-known feature of heterochromic uveitis, was noticed after operation in eight of the nine cases. In four cases it appeared to be responsible for a poor visual result. In four cases the opacity was preceded by haemorrhage into the vitreous which in one patient proceeded to an organized mass. However, in the remaining cases, the opacity took the form of a vitreous haze of various degrees of density, usually with the addition of many larger floating opacities.

There is no simple way of assessing the clarity of the vitreous before extraction. However, unless we consider that the cataract is a causal agent, it would be reasonable to assume that surgical interference does increase vitreous opacity, as there was in all cases a gradual clearing of the media in the post-operative period.

The aetiology of the opacity was obscure except in those four cases in which it apparently followed haemorrhage. A peripheral choroiditis has been noted by Duke-Elder (1940). Coles (1964) said this was frequently present. Welch, Maumenee, and Wahlen (1960) described "pars planitis" which causes anterior vitreous opacity. Perkins (1961), however, found no evidence of such a condition in his series of cases.

Although it would be reasonable to assume that a low-grade inflammatory or degenerative reaction was responsible for the vitreous haze in our cases, there was no evidence of peripheral choroidal or cyclitic changes.

Of the nine cases described, eight showed a post-operative rise in ocular tension.

Five of the eight patients showed raised ocular tension before surgery. Two of these had glaucoma of what appeared to be a lens-induced type. Müller (1963) described a similar case and discussed it as a clinical presentation of a phacolytic type of glaucoma. However, the cataracts of these eyes, although mature, were not

of the degree of hypermaturity which is associated with this type of glaucoma in normal eyes. The significance of this feature is discussed further.

With regard to the post-operative course of the eight cases, all showed a rise in ocular tension to a greater or lesser extent for at least part of the subsequent period. One showed a rise only as relative to the normal eye (20 as compared with 15 mm. Hg). Two showed a post-operative rise with spontaneous return to normal. Two patients, despite maximal medical therapy, continue to maintain high ocular tensions and are requiring a drainage procedure.

The remaining three patients have all required drainage operations in order to control serious persistent ocular hypertension. In two of these the eyes have suffered permanent damage in the shape of advanced field loss and poor visual acuity. It is not possible to assess the other case as the eye has amblyopia ex anopsia.

It is apparent that the secondary glaucoma is probably the most serious and most common condition associated with cataract in heterochromic uveitis. In some cases this carries a poor visual prognosis.

Both patients with possibly phacolytic glaucoma showed marked falls in tension after cataract extraction. In one case this was still 5 mm. Hg higher than the normal eye. In the other, having fallen to normal levels for 2 weeks post-operatively, the tension showed a marked prolonged rise and will probably require a drainage operation. This latter case suffered an iris prolapse and had perhaps undergone an abrupt rise in pressure immediately post-operatively similar to that described by Huber (1961) after anterior chamber paracentesis. It is of interest that the only case of the nine reported in which a rise in tension was not recorded also experienced an iris prolapse.

The other three patients who had pre-operative glaucoma each showed marked prolonged post-operative rises in tension; one eventually required a drainage procedure and the other two maximal medical therapy.

Gonioscopy findings in the patients with glaucoma appeared to show that in only two of the eight patients was there sufficient macroscopic evidence in the angles to account for a marked rise in tension—significantly in the only two patients to show a marked post-operative anterior uveitis. In these two patients there were irido-corneal adhesions largely occluding the angles. The remaining patients showed either normal angles or angles with only a few peripheral anterior synechiae.

The mechanism of the rise in ocular tension in these patients is probably not the same in every case. In two cases it appeared to be secondary to inflammatory adhesions occluding the angle. However, in the remaining cases, the aetiology is not so apparent. A hyphaema occurred post-operatively in all but one case. In two patients the tension remained raised while blood remained in the angle and then returned to normal. Presumably blockage of the angle by red blood cells was the chief aetiological factor in these cases, although at the time it was considered that the hyphaema would be insufficient to cause ocular hypertension in a normal eye.

In the remaining patients, the increased tension persisted long after clearing of the anterior chamber. The normal appearance of the angles is similar to that of chronic simple glaucoma.

It is postulated that certain patients with heterochromic uveitis have diminished

aqueous outflow values and that decompensation with consequent secondary glaucoma may be precipitated by various factors. Among important causes may be surgical destruction of part of the angle filtration system at cataract extraction, blockage of the angle structures by red blood cells and breakdown products, and similarly blockage by lens breakdown products. It is possibly significant that two of these patients appeared to develop a lens-induced type of glaucoma and markedly elevated tensions associated with cataracts that did not appear grossly hypermature. Possibly a microscopic leakage of breakdown products from the lens was sufficient to block an already abnormal filtration angle.

Little work appears to have been undertaken on either the clinical or the histopathological aspects of angles and aqueous outflows of eyes with uncomplicated heterochromic uveitis. However, Goldberg and others (1965) have described mononuclear cell infiltration of the base of the iris in specimens of iris removed during cataract extraction. It would seem reasonable to expect a similar infiltration of the angle structures and consequent embarrassment of aqueous outflow. Huber (1961) described a single case with an unusual hyalinization of the angle structures. He also found increased resistance to outflow in cases with glaucoma.

Further studies are obviously necessary on eyes with heterochromic uveitis, and tonographic measurements are being undertaken by the present authors to determine whether there is diminished outflow in normotensive eyes with Fuchs's heterochromic uveitis.

Summary

Nine cases of extraction of the cataract complicating Fuchs's heterochromic uveitis are described. Each of these has suffered one or several complications.

Three types of complication are described:

- (1) *Intra-ocular haemorrhage occurring immediately post-operatively.*—Hyphaema occurred in seven cases and vitreous haemorrhage in four.
- (2) *Vitreous opacity of various degrees of severity.*—This occurred in eight cases and was responsible for a poor visual result in four.
- (3) *Glaucoma present pre-operatively.*—This occurred in four cases, and a post-operative rise in tension probably occurred in all cases. Spontaneous resolution followed in four cases, two cases required medical therapy, and in three cases drainage procedures were necessary.

The aetiology of each complication is discussed.

The operation of lens extraction in Fuchs's heterochromic uveitis is widely held to be a straightforward procedure with a benign prognosis. It is submitted that this is by no means always the case and that the procedure can follow a difficult and complicated course, not infrequently ending with a poor visual result.

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