HYDROPHTHALMIA

BY

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An exhaustive study of this disease has recently (1939) been set forth in a monograph by Ringland Anderson, who suggests that the term buphthalmia should be reserved for megalocornea, in which the tension remains normal, and that the term hydrophthalmia should be applied to cases of infantile (congenital) glaucoma. There was not at any time any doubt about the classification of our case and we therefore adopt Ringland Anderson's nomenclature.

The subject of this communication is a boy now aged 14, who was seen for the first time at the Ophthalmic Department of the Glasgow Royal Infirmary, on December 19, 1932, when he was 4 years of age. His eyes were said to have been large since birth, and they presented the typical appearances of hydrophthalmia. There was general oedema of the cornea of the left eye, and evidence of ruptures of Descemet's membrane in both. The right
optic disc was clearly seen and was deeply cupped and greyish-blue in colour. The left fundus could not be seen owing to the corneal oedema. The diameter of the cornea was 15 mm. The tension as estimated immediately before the operations was, unfortunately, not noted on the case sheet.

On January 8, 1932, both eyes were trephined, and the pressure in the left eye was so great that the aqueous spouted on the completion of the trephining. After recovering from the operation it was found that the left optic disc was also deeply cupped and greyish-blue.

The boy was brought at intervals to the Out-Patient Department, and the mother reported that he was leading a normal life with sufficient sight to enable him to look after himself whilst playing with other boys in the streets.

There was doubtful perception of light in the left eye, and the distant vision of the right eye was never better than 6/36, but he was able to read his school books. He went to an ordinary school, and his progress was so good that he was chosen for transfer to a higher grade school. These plans, however, were upset by an unfortunate accident in March 1940. The left eye was hit by a snowball, and a fortnight later he was brought to the hospital. There was a complete hyphaema, intense pericorneal injection, with photophobia and lacrimation. The condition improved somewhat, and he was discharged from hospital on May 4, but he was not brought again until July 4, when the other eye was found to be the seat of an intense irido-cyclitis. When asked when the right eye had begun the mother said that she was quite unaware of it, because the boy had made no complaint and had constantly kept his head down on account of the photophobia.

The Wassermann reaction was negative and a diagnosis of sympathetic ophthalmitis was made, and the left eye was removed on July 27, 1940. This has not resulted in any improvement in the condition of the other eye. It is now quite blind, the iris is completely adherent to the lens capsule, and the pupil is full of exudate.

The excised eye was hardened in formalin and embedded in celloidin, sections were stained with haematoxylin, eosin, van Gieson, Mallory, orcein, and Weigert.

Sections made from the left eye showed a great variety of pathological changes due to hydrophthalmia and to the superadded sympathetic ophthalmitis, but we confine ourselves chiefly to a description of Bowman's membrane and Descemet's membrane, both of which showed unusual features.

The corneal epithelium is not affected. It has a smooth surface and is nowhere detached.
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Bowman's Membrane

A great number of delicate lines across Bowman's membrane attracted attention. The majority were perpendicular to the surface (Fig. 1), but some were oblique (Fig. 2), and one was branched making a Y shape. They are not artefacts, and the corneal epithelium was normal in outline. In one place there was a fracture in a backward direction of the more deeply stained anterior part of the membrane with overlapping of the two ends. (Fig. 1.) Here again there was no change in continuity of the epithelium overlying this fracture. On each side of this

Fig 1.
Hydropthalmus congenital haematoxylin—Eosin. 10 eyepiece, 40 objective Zeiss. Cracks and fracture in Bowman's membrane.

Fig 2.
Acute bend with cracks without fracture.
fracture the cross lines were more numerous, and this suggested the possibility that they may be minute cracks or creases in the membrane.

At another point there was an acute bend backwards of the anterior margin of the membrane without any change in the continuity of the posterior margin directly opposite (Fig. 2). At the bend the basal epithelial cell appears to be squeezed at the apex of the bend, that is to say, it is a longer narrower cell than the other basal epithelial cells, and its posterior end is pointed.

At another point there was a longitudinal slit in the membrane (Fig. 3), partly lined with elongated nuclei. The glass membrane itself can be distinctly traced passing along in front of and behind the slit.

Another feature noted were round bodies (Figs. 3, 4 and 5) situated on the anterior limit of Bowman’s membrane. They are mostly

![Fig. 3. Longitudinal splitting of Bowman’s membrane.](image)

![Fig. 4. Hemispherical bodies at the anterior borderline of Bowman’s membrane. Weigert stain for elastic tissue. 10/40.](image)
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Hemispherical, but some are slightly conical, and a few have flattened tops. Their straight base lies on the membrane. They vary in size, some being as large as a halved erythrocyte, while others are so small that they are discovered only with a closed down illumination. They are conspicuous when stained with orcein, and take the stain more deeply than the nucleus of a lymphocyte. They appear to be quite structureless. They were described by von Hippel (1928); and Talbot (1938) states that in an examination of 32 consecutive eyes he found these bodies in 14, all of them glaucoma. None were found in non-glaucomatous eyes. Talbot then examined 27 old slides, and in 11 of these, in which the tension had been raised, the bodies were present.

Talbot considers that they are found only in cases in which increased tension has existed for more than a few days, and in which the tension has been increased up to the time of excision of the eye. This condition, however, is not absolute, because we have found the bodies in eyes in which the tension had for a long time been sub-normal, as in our case, and finally we detected them in an eye of a 65-year-old woman with no history of glaucoma, normal function, and normal fundi. Her globe had been excised on account of a melano-sarcoma of the conjunctiva.

It is difficult to determine the cause of their presence and their significance.

Talbot refers to explanations which have been offered.

1. That they are nuclei of corneal epithelial cells, in the posterior layer, depressed into Bowman's membrane. He considers this to be unlikely because he found the hemispherical bodies in one case in which the corneal epithelium had become detached.
2. The number in a typical section corresponds to the number of nerves which penetrate Bowman's membrane and Wolff (1938) suggests that they are connected with these fibrils. We could not detect any connection with corneal nerve fibres visible with closed down illumination.

Although we can offer no satisfactory explanation we think that their appearance and situation suggest that they are the products of disordered epithelium. In Talbot's case they may have been formed before the epithelium separated from Bowman's membrane.

In cases of high tension we do find small dark-stained circular corpuscles (Fig. 6) mostly in the basal cells of the corneal epithelium and the hemispherical bodies may result from these.

FIG. 6.
Spheroidal dark bodies within the basal cells of corneal epithelium in glaucoma absolutum. Haematoxylin—Eosin. 10/oil immersion.

Whatever may be their actual origin and mode of formation we may suppose that they are an indication of disordered metabolism of the cornea in glaucomatous eyes and that the cracks or fissures, and the fractures, and the splitting of Bowman’s membrane may be placed in the same category.

The only changes seen in the corneal parenchymatous lamellae are newly formed blood vessels and lymphocytic infiltration, but Descemet’s membrane reveals most interesting features.

Descemet's Membrane

Ruptures of Descemet's membrane are well known as a feature of hydrophthalmia. They were seen clinically in both eyes of this boy when he first came under observation nine years ago at the age of four.
Their pathology was studied by George Coats with his usual care and attention to detail when he described twelve cases of it in 13 cases of hydrophthalmic eyes.

In our case the rupture, which was probably ten years old, to which we devoted most attention was situated 4.6 mm. from the corneo-iridic angle, and the first thing to attract attention was the presence of two knobs (Figs. 7, 8 and 9) on the posterior surface of the cornea. These are the boundaries of the rupture and are 1.215 mm. apart.

**FIG. 7.**

**FIG. 8.**
Elastic ribbon

Second Desc. m.

Third Desc. m.

Elastic ribbon

Second Desc. m.

Third Desc. m.

Distance between \( a \) and \( b = 1.215 \) mm.

**Fig. 9.**

It was these knob-like excrescences which attracted the attention of earlier observers before their significance was understood. Thus (quoting Coats' Paper) Gama Pinto (1886) noted on the posterior surface of the cornea small ovoid projections covered with endothelium, with a concentric structure, like Pacinian corpuscles, and Durr and Schlegdendal (1899) described "small excrescences of the membrane of Descemet" in one case and "nodular and tumour-like thickenings and projections" in another.

Although the condition was well-known anatomically, the first certain clinical recognition of ruptures in hydrophthalmia is due to Haab (1899).

The reason why the edges of the rupture take this knob-like form becomes more apparent, we think, from a study of the membrane itself.

There appear to be three distinct layers in Descemet's membrane because there are three degrees of staining with haematoxylin, orcein, Weigert, etc.

1. An anterior darkly stained thin layer in contact with the corneal lamellae.
2. A broad middle layer which stains a bluish pink.
3. A narrow posterior layer which stains pinkish. (Figs. 7, 8 and 9.)

The first layer does not appear in the floor of the rupture. At each edge it has curled up in an anterior direction in the shape on one side of a reversed S, and on the other side in the shape of the head of a Bishop's staff. It is this curl which is the basis of the knob-like prominence, and the other two layers form a cap over it. These two layers, and especially the middle layer, are much broader over the prominence than elsewhere, and there is irregularity of their width in the floor of the gap.

In our case there had been an intense irido-cyclitis for four months prior to the removal of the eye, and it is therefore not surprising that across the rupture there is loss of endothelium, which is replaced by many irregular flat nuclei and heaps of red blood corpuscles—the remains of the hyphaema.
The ability of endothelium to spread widely is well-known and therefore it will quickly cover the gap in the cases in which it also had given way. Coats states that in all his cases, in which there had been no complications of an inflammatory nature, the endothelium was intact over the gap, but he cites other cases from the literature in which its regeneration was irregular and incomplete.

The opacity of the cornea which accompanies the ruptures of Descemet's membrane is due to contact of the aqueous with the corneal parenchyma. The degree and permanence of the opacity depends upon the duration of that contact. We think it possible —nay probable—that not all ruptures involve the whole depth of the membrane in which case there would be no opacity at the site of the rupture.

**Regeneration of Descemet's Membrane**

We have stated that the staining reactions of these 9 years old ruptures suggest that there are three layers in Descemet's membrane proper. This view is supported by a study of the regenerated membrane.

Regeneration appears to be confined to the two posterior broader layers which can be traced as a seemingly continuous sheet from each side of the rupture, over each knob, and across the floor of the gap (Fig. 9). But examinations with the highest power reveal that there is not always end to end continuity but that a minute gap may separate the ruptured ends of the two posterior layers. (Fig. 10.)
On the other hand there is no evidence of regeneration of the thin anterior darkly stained layer, the truly elastic layer. When it ruptured its edges sprang apart, and have remained so, and there is no sign of it in the floor of the gap.

Over the knobs there is increased thickness of the broader homogeneous layers plastered over the divided ends of the thin anterior layer, comparable to the excess of callus thrown out around a badly opposed fracture of bone.

Ranvier (1898) in studying Descemet's membrane which had been ruptured experimentally considered that regeneration took place from the convexity of the curl thus filling the gap. But we show that it is only the elastic layer which is curled and that the other layers form a continuous sheet over the curled anterior layer and across the gap. But it must be noted that Ranvier studied fresh ruptures whereas our ruptures are probably 9 years old.

The thickening of the two posterior layers on the cap over the curled up anterior elastic layer may be due to a bunching up by the edges of the ruptured elastic layer as they sprang apart.

It may be that the broader posterior layers are tougher than the thin anterior layers and that they do not always give way as the eyeball stretches. It would be interesting to know if it is these two layers, rather than the whole membrane, which prove so resistant to rupture in perforating ulcer of the cornea, and we intend to pursue enquiry on that point as pathological material offers.

In describing the development of these parts Ida Mann states that the endothelium is formed by a flattening and re-arrangement in situ of mesodermal cells in the posterior aspect of the cornea and that it does not grow in from the periphery, that the structureless layer (Descemet's membrane) is homogeneous from the first, and that elastic fibres are recognised in the deepest layers of the parenchyma directly in contact with Descemet's membrane. This last statement is interesting in view of our suggestion that the only elastic part of the membrane is a thin sheet of it in direct contact with the substantia propria of the cornea.

Descemet's membrane is thought to be produced by a secretion from the endothelial cells, whereas Bowman's membrane is simply a condensation of the anterior corneal fibrillae (Seefelder, quoted by Wolff). For that reason Descemet's membrane can be reformed whereas Bowman's never regenerates.

But, as stated above, it is only the two posterior layers which are regenerated. The anterior layer, like Bowman's membrane, is not regenerated. May the explanation of this be that this layer is developmentally related to the posterior layers of the corneal parenchyma, and that it is a condensation of the elastic fibres contained therein just as Bowman's membrane is thought to be
formed by condensation of the fibrillae of the anterior layers of the cornea.

**Elasticity of Descemet's Membrane**

The sections throw some light on the question of the elasticity of the membrane. Coats did not consider it to be elastic, and believed that the fact that it assumes curly or spiral forms when divided is no proof of its elasticity but of its lack of elasticity. If it were truly elastic it would, when divided, thicken and retract but retain its original form, and he pointed out, in support of this contention, that a sheet of stretched India rubber when cut or torn does not curl up like the membrane of Descemet. He regarded the membrane as a sheet of resistant but brittle material.

It is true that the curling of the torn edge is not what one would expect from an elastic membrane. One would expect the curling to be in spiral form, whereas our sections show the curl to be in the shape of a reversed S and some of them resemble the curve on the head of a Bishop's staff.

We consider that this behaviour of the ruptured membrane is due to the three layer formation of Descemet's membrane, that the layer next the parenchyma is elastic, and that it is covered by the rest of the membrane, which is non-elastic. The elastic part is probably less in thickness than one-third of the whole. When ruptured, the non-elastic two-thirds serve to hold the elastic sheet in position and thus prevent it from retracting in the ordinary manner of a cut stretched elastic rubber sheet. In that way it is only a small part of the elastic sheet which is free to change its shape when torn, and it takes the shapes shown in our sections.

In another connection—a case of xanthomatosis bulbi—in which the Sudan stain for fat was used, it was only the anterior dark layer, which we presume to be the elastic one, which took on the red stain, and it did so extensively (Loewenstein, 1941a). Further, in a case of intensive conjunctival argyrosis selective silver staining of the elastic elements is found (Loewenstein, 1941b). Studying such a case with the slit-lamp one finds silver staining of this layer, a further proof of its elastic nature.

These two observations suggest that there is a difference in chemical composition between the anterior third and the posterior two-thirds of Descemet's membrane.

Fig. 11 supports the view of those who consider that Descemet's membrane does not end abruptly peripherally (Ringland Anderson, p. 74). The section shows that the membrane splits in a fan-like manner and the split up layers can be traced as a network into the dense scleral fibres. This is best brought out by Weigert's stain.
The other pathological features found in this eye are the result of the blow with the snowball and the irido-cyclitis which followed it.

(1) The anterior chamber is filled with blood.

(2) The iris is infiltrated and swollen, with great masses of blood in the midst of the iris tissue.

(3) The ciliary body is enormously swollen and densely infiltrated with lymphocytes and a great number of epithelioid and giant cells are seen.

(4) Pigment is scattered all over the ciliary tissue.

(5) The choroid is infiltrated and thickened. Most of the infiltration mass consists of lymphocytes, but there are eosin-stained epithelioid cells and huge giant cells. Parts of the pigment epithelium are destroyed, parts are detached, and in other places it has thickened to several layers producing small and large plaques.

(6) Behind the lens the retina lies completely detached and is in thick folds stuck together round a necrotic centre with large pigmented cells and hyaline degenerative patches.

**Summary**

1. A case of hydrophthalmia is described clinically and histologically.
2. In Bowman's membrane hemispherical bodies, cracks and splits are found.
3. Features are described which suggest that Descemet's membrane consists of three layers, a thin anterior layer and two broader
layers covering the former. The staining reactions and the appearances at a rupture indicate that the thin anterior layer is the truly elastic part of the membrane and that, developmentally, it may be derived from the posterior corneal lamellae, while the two broader posterior layers are the only part derived from the endothelium.

4. The bearing of these observations upon the questions of the regeneration and of the elasticity of the membrane are discussed.

5. When the boy was 12 years of age the eye sustained an accident which was followed by sympathetic ophthalmitis. The numerous pathological signs of this are mentioned.

REFERENCES


LOEWENSTEIN (1941a). — Lipoids within the ocular tissue. Monograph.


CENTRAL RETINITIS IN A GIRL AGED EIGHTEEN YEARS. RECOVERY

BY

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London

The prognosis in choroiditis is usually bad as regards the area affected. As Duke-Elder\(^1\) states, "... despite any type of treatment, the lesion involves a complete and irremediable destruction of the area involved ..." When this area is central, the resultant loss of central vision makes the outlook hopeless.

The aetiology is frequently uncertain, and while we are occupied with the often prolonged search for one of the multiple possibilities of septic foci which may be responsible, the damage is done.

The report of a case in which recovery occurred may therefore be useful.

M.A., aged 18 years, when first seen at the South London Hospital for Women in January 1934, had a small patch of

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