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COMMUNICATIONS

HEREDITARY CORNEAL DYSTROPHY*

BY

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ABERDEEN

HEREDITARY corneal dystrophy was first described by Groenouw in 1890. Since then cases have been found in most European Countries, in America and in Japan, but no case has yet been published of the disease occurring in a negro patient. The condition, however, is relatively rare, and in this country at least, must be quite unfamiliar to many ophthalmologists.

The essential features of the disease are—(1) Its hereditary and familial character, (2) its bi-lateral distribution, (3) its situation in the cornea, occupying the axial zone only, in the case of the dominant varieties, and (4) the absence of vascularization.

The terms used in describing the lesion have been many and varied, and I suspect that the difference was mostly due to the authors' descriptive powers and literary ability, and not to any fundamental difference in the clinical picture.

Historical

Groenouw first gave a description of this disease in 1890. He described the eye condition of two patients, and called the corneal disease "nodular corneae." It has since been proved by Bucklers

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that the first patient suffered from granular dystrophy, and the second from macular dystrophy.

In 1898 Groenouw described the corneal condition of his original patient, then 49 years old, in the following words—"On both eyes the conjunctivae are normal. The periphery of the cornea is completely clear for a breadth of about 3 mm. while the centre is taken up with very numerous grey spots. These opacities are irregularly rounded with small prongs and endings. They could best be compared with small crumbs of dry bread. The opacities lie partly separate and partly fused into groups and streaks. They are about the size of a poppy seed. Between them lie a large number of even finer points which are only perceived with a magnifying glass. Also several quite fine grey streaks and strokes are visible between the opacities. With the exception of these grey, sharply defined deposits, the corneal tissue is clear." As will be seen later this description could be applied to the majority of the affected members of my pedigree.

The description of the other patient was as follows—a girl aged 17 years came to have her eyes examined owing to gradual dimness of vision. "Both corneae show in their centre a great number of grey, roundish opacities, which are fused partially together. The opacities are more numerous than in the first named patient. They are situated in the outer corneal layers and push forward the epithelium sporadically in small humps. The surface of the cornea is otherwise smooth and well reflecting. Between these larger spots are quite small points visible only with the magnifying glass. They occupy the entire cornea to the margin and appear as soft, nebulous opacities to the naked eye. These small points extend also to the deeper layers of the cornea."

The chief differences between the corneal lesions of the two patients are (a) The distribution of the opacities. The opacities in No. 1 being in the central region of the cornea while in No. 2 the opacities extend to the limbus. (b) The effect on vision. Patient No. 1 came at the age of 47 years not complaining about poor vision, but owing to conjunctivitis, while No. 2 came at the age of 17 years owing to poor vision.

The first description of reticular dystrophy was given by Biber (1890). He described three patients who were unrelated to each other. With transmitted light he saw a fine mist-like opacity which was composed of long and short fine lines. "They run in various directions, and sometimes cross each other. They remind one of a birch twig branching here and there. Ledge-like projections on the surface of the cornea appear to correspond to those lines." By means of oblique illumination he observed that these superficial ledges were partly transparent and that consequently these grey lines were caused through irregular refraction on the part of these ledges.

Haab (1899) presented other four patients with this disease all belonging to one family, three in one generation and one in a younger generation. He writes about the condition "the lines show in general a radial arrangement, especially the longer ones. Moreover they are often forked, the fork being open towards the centre of the cornea. Still one sees here and there a fork that is open towards the periphery. Some of the lines bend after running towards the centre of the cornea forming an arc of a large circle. The lines have throughout a somewhat wrinkled or undulating course and their arrangement reminds one of twigs or figures which one sees on a frozen window. Between the lines one sees small grey spots and points." He goes on to say that "the figures are transparent and light refracting, so that formations that look like drops of water correspond to the small spots, and the lines are like ledges which resemble threads of crystal." All these changes were most numerous in front of the periphery of the pupil.

Dimmer (1899) described three members of a family who had lattice-like opacities of the cornea. The central corneal region showed nodular and crystallized raised parts. Surrounding this region were very delicate anastomosing lines. These lines become visible only with special illumination (reflected light). From this time onwards many cases were published, some agreeing with Groenouw's first case, and others with the Biber, Haab, Dimmer type. Others again, Fleischer (1905) especially, maintained that all cases of corneal dystrophy were of the same type, and should be classed under the name "Familial Corneal Degeneration."

Some English investigators Spicer (1904) Doayne and Stephenson (1905) were of the opinion that the two forms could be seen in the same family and that transition types could be demonstrated. Doayne and Stephenson (1905) stated "The two diseases, then, agree in several particulars—as for example that they begin about the period of puberty; that they are accompanied by insignificant signs of inflammation; that they attack at first chiefly the central parts of the cornea; that they are slowly progressive; that they probably represent not an inflammatory but a degenerative process. Then, lastly there is the fact that the sensitiveness of the cornea, particularly as regards the central region may be impaired; and, finally that both conditions tend to run in families so to speak. In short there is so close an analogy between the "nodular" and the lattice-like conditions that it is difficult to avoid a suspicion that they are at the root one and the same affection."

An American investigator Judd (1933) was also of the opinion that nodular and lattice-like degeneration of the cornea could be present in the same family.

Bucklers (1938) published the result of an investigation that he

undertook for the German Government under the sterilization of the unfit order. This study proved genetically and clinically that there are three types of hereditary corneal dystrophy, a dominant granular type, a dominant lattice-like type and a recessive macular type. A total number of 129 affected patients were examined, 91 were of the granular type, 32 the spotted type, and 6 the lattice-like type. Affected patients previously examined by Fleischer, Tritscheller and Gilch were re-examined and re-classified in certain instances; this was comparatively easy when there were other cases of the same pedigree for comparison. Bucklers states that the investigators who maintain that the granular type and lattice-like type occur in the same family and pedigree never actually examined a genuine case of lattice-like corneal dystrophy, as there is no similarity between the two lesions. Doubt might arise about the diagnosis between an isolated case of well-marked granular corneal dystrophy and the macular type; but as the transmission is different and the distribution of the opacities is different, no real difficulty should now be encountered.

An attempt has been made by many authors to classify the disease, but as the classifications were based mainly upon the shape of the opacities it can be understood that few had any real scientific foundation. The chief difficulty appears to have been that the rarity of the disease made it impossible for the various authors to see any other cases. They had therefore to depend upon written descriptions and drawn illustrations for comparison. It will be of interest to study some of the published classifications and the nomenclature of the disease as used by the various authors.

Nomenclature of the Disease as used by the Author

- AURAND (1935).—Grill-like keratitis.
 BONNET and BUSSY (1935).—Nodular degeneration (Groenouw).
 BRAV (1935).—Familial corneal degeneration.
 DIMMER (1899).—Superficial lattice-like corneal opacity.
 DOYNE and STEPHENSON (1905).—Familial degeneration of cornea.
 FEHR (1904).—Familial spotted corneal dystrophy.
 FISHER (1936).—Congenital corneal opacities.
 FLEISCHER (1905).—Familial corneal dystrophy.
 FUCHS (1915).—Nodular form of corneal opacity.
 GREENWOOD (1931).—Lattice keratitis.
 GROENOUW (1890).—Noduli corneae.
 GUNN (1902).—Keratitis nodosa.
 HAAB (1899).—Lattice-like keratitis.
 JUDD (1933).—Nodular degeneration of the cornea.
 KOBY (1927).—Superficial reticular degeneration.
 MACRAE (1933).—Tesselated opacity of the cornea.
 PATERSON (1907).—Reticular keratitis.
 ROLLET (1933).—Hereditary and familial keratitis and corneal degeneration.
 SATANOWSKY (1932).—Keratitis punctata profunda.
 SPICER (1904).—Nodular opacities of the cornea.
 SRINIVASAN (1932).—Groenouw's dystrophy.
 UHTHOFF (1915).—Keratitis urica.

- VEASY (1904).—Familial macular degeneration of the cornea.
 VOGT (1934).—Corneae-guttae.
 WORTON (1912).—Punctate crystalloid deposits in both corneae.

CLASSIFICATION

DEUTSCHMANN (1908).

1. Spotted Corneal Dystrophy.
 (a) Familial. (b) Non-familial.
2. Nodular Corneal Dystrophy.
 A—Familial. (a) Lattice-like opacities. (b) Fine punctate.
 B—Non-familial. (a) Fine punctate. (b) Irregular, mixed (dots and twigs).

AXENFELD (1917).

Corneal Dystrophies.

- (a) Nodular (Groenouw). (b) Lattice-like (Haab, Dimmer).
- (c) Familial (Fleischer).

BACHSTEZ (1919).

1. Lattice-like (Biber, Haab, Dimmer).
2. Nodular (Groenouw, Fuchs).
3. Spotted, familial (Fehr).
4. Fleischer's cases—3 groups.
 (a) Opacities smaller than Groenouw type.
 (b) Opacities similar but more pronounced.
 (c) Discs and small dots, and in addition a large number of circles which lie in the deepest layers only.

LUGLI (1931).

1. Nodular degeneration of Groenouw.
2. Lattice-like keratitis (Biber, Haab).
3. Familial spotted degeneration of Fehr.
4. Bachstetz type.
5. Leucomatous nodular dystrophy of the cornea of Yoshida.
6. Salzmann type.

CHOU (1932).

1. Groenouw type.
2. Fleischer type.

WARDENBURG (1932).

Familial corneal degeneration.

1. Nodular (Gunn, Fleischer).
2. Spotted (Fehr, Rosenberg).
3. Crystalline (Tritscheller, Von Went, Weibaut, Von Hippel).
4. Lattice-like (Freund, Fleischer).
5. Mixed and transition types (Lowenstein, Landenberger and Dimmer).

VAN DER HEYDT (1937).

1. Fleischer type.
2. Haab-Dimmer type.
3. Groenouw type.

BUCKLERS (1938).

1. Granular corneal dystrophy (Groenouw (1), Fleischer (2)).
2. Spotted corneal dystrophy (Groenouw (2), Fuchs, Fehr, Fleischer).
3. Lattice-like (Haab, Dimmer).

DUKE-ELDER (1939).

1. Nodular type (Groenouw).
2. Reticular or lattice-like type (Biber, Haab, Dimmer).
3. Ring-shaped type (Fleischer).

FRANCESCHETTI and STREIFF (1940).

1. The recessive type of dystrophy of the cornea, *dystrophia corneae maculosa* (recessive form of Groenouw's familial corneal dystrophy, macular corneal dystrophy of Fehr).
2. Dominant type of dystrophy.
 - (a) *Dystrophia corneae granulosa nodulosa* (nodular). Dominant form of Groenouw's familial corneal dystrophy, granular crumb-like corneal dystrophy.
 - (b) *Dystrophia corneae reticularis*. Lattice-like corneal dystrophy (Biber, Haab, Dimmer). Lattice-like keratitis.

Groenouw's original title of "nodular" occurs frequently. This is unfortunate and there is no doubt that the use by Groenouw of this word, misled many investigators into thinking that the nodule was the distinguishing feature of the disease, while in fact it is a common feature in all three types. Several call the disease Groenouw's corneal dystrophy. As Bucklers (1938) proved that Groenouw's original two cases were of a different type, it is impossible, unfortunately, to give to any one type the name of the original discoverer of the disease, and at the same time prevent confusion between the granular and macular types. This mistake of Groenouw's also undoubtedly confused future investigators, as cases were encountered which differed from the original description of the granular type. "Keratitis" crops up frequently. Surely this term is improper having regard to the fact that, in several instances, there have been no attacks of pain or redness at all; in other instances in which attacks of inflammation have been noted, such attacks have been transitory and have succeeded, not preceded, the opacities.

Heredity of the Disease

That the disease is familial was first noted by Fuchs (1901). Both males and females are affected and the disease can be transmitted through either. Pedigrees of considerable extent have been published, showing that the granular and the reticular types are transmitted in a dominant manner. Descendants of the healthy are, and always remain healthy. Pedigrees showing the disease directly transmitted through the following number of generations have been published.

Three generations.—Freund (1904), Doyne and Stephenson (1905), Reigel (1933), Hess (1905), Judd (1933), Saeboe (1935), Freiburger (1936).

Four generations—Pillat (1922), Groenouw (1933), Bucklers (1938).

Six generations.—Tritscheller (1921), Frykholm (1935).

Sporadic cases of corneal dystrophy have been published and shown at ophthalmological meetings, while again several affected

were brothers and sisters, with healthy parents and offspring. Macnab, (1907 and (1914), Evans, (1930), Fehr, (1904), Moxon, (1914), Cardell, (1931). It would appear therefore that corneal dystrophy is not always transmitted in a dominant way. Unless parents and children of affected cases were actually examined by an ophthalmologist, however, the statement made by a patient that there was nothing wrong with their parents' or children's eyes cannot carry much weight, as the vision of those affected with granular corneal dystrophy varies little from normal until well over middle age, so there is a probability that several cases published were transmitted in a dominant way, and were genuine cases of granular corneal dystrophy. Bucklers (1938) investigated 7 pedigrees of corneal dystrophy and found that in 5 of the pedigrees all the affected siblings belonged to the one generation, no affected ancestors or descendants could be found. In two pedigrees, besides one parent, several children suffered from the disease. It was found that both the grandparents and the parents had married blood relations. In all the pedigrees investigated, consanguinity was present. The blood relationship in some of the pedigrees was very obscure and had to be traced back 8 to 11 generations.

As two modes of transmission are now known, hereditary corneal dystrophy can be divided into two main classes, the classes being sub-divided into their respective groups. As the aetiology of the condition is still quite unknown, a word describing the opacities is possibly still the best term to use for the sub-groups. As so much confusion has arisen through using the term nodular, I would advocate the use of the descriptive term of "granular" dystrophy and suggest that the term "nodular" be abandoned because it has caused so much confusion.

HEREDITARY CORNEAL DYSTROPHY

Dominant transmission	Recessive transmission
Granular (Groenouw 1)	Macular (Groenouw 2, Fehr).
Reticular (Haab). ...	

Granular Corneal Dystrophy.—This is a dominant disease, affecting both male and female. The disease begins in the first decade the youngest reported case being 2 years old (Saeboe). The opacities are first seen as fine dust-like dots, discs or circles, situated below the epithelium in the axial region of the cornea.

These dots often form radial lines, hence the confusion between early granular and lattice-like types. Over a number of years the opacities multiply and coalesce to form clumps. The clumps are of a greyish colour, irregular in outline, and have been compared to "crumbs of bread." These clumps are individually sharply circumscribed and occupy a disc-shaped area of about 5-7 mm. in diameter in the central region of the cornea. They are mostly situated in the region of Bowman's membrane, but are also found in the parenchyma as deep as Descemet's membrane. The cornea between the particles and also a marginal zone of 3-5 mm. is quite clear. Vision in youth is unimpaired.

In the second decade the refraction usually changes, becoming astigmatic, mixed or myopic, but vision can be kept near normal with glasses. In middle age vision becomes affected but there is no definite irresistible progress of the disease, vision in some cases being remarkably good, even in old age. In old age the surface of the cornea becomes slightly irregular. Sensitivity in the majority of cases is normal.

Reticular Corneal Dystrophy is transmitted in a dominant way. It also commences in the first decade of life. Until middle age is reached, no corneal abnormality can be seen by macroscopic examination. The opacities are first seen in the axial region of the cornea as transparent nodules like "drops of water" (Haab) which raise the surface epithelium. In the intermediate zone opposite the periphery of the pupil, fine anastomosing branching lines form a lattice-like network. Unlike the opacities of the granular type, the lines are channels and fissures filled with a clear substance having a different refractive index from the surrounding cornea. These lines can only be seen with high magnification and reflected light (Dimmer). Between the lines are interspersed fine glassy, reflecting spots. The surface of the cornea becomes irregular at an early age, and it is this surface irregularity that causes the diminished vision at an early age (Bucklers). Corneal sensation is greatly diminished.

Macular Corneal Dystrophy.—This disease is transmitted in a recessive manner, consanguinity being found by Bucklers in the 7 pedigrees he investigated.

The disease commences in the first decade as a fine, diffuse opacity rapidly occupying the whole cornea. In the axial region the opacity consists of greyish spots of various shapes. The spots are larger and whiter than in the granular type. The opacities in the central region are mainly superficial but near the periphery they are mostly in the deeper parts. The epithelium is irregular and often traversed by pigment lines (Bucklers). Sensitivity is greatly diminished. Vision is early affected and in middle age is reduced to finger-counting.

FIG. 1.

Granular corneal dystrophy.

Figs. 1, 2 and 3 are copied from *Die Erbliehen Hornhautdystrophien* by Bucklers.

FIG. 2.

Reticular corneal dystrophy.

FIG 3.
Macular corneal dystrophy.

FIG. 4.

VI(42) as seen with the luminous ophthalmoscope and plus 20D. lens in the aperture. Opacities seen black against the red fundus reflex.

FIG. 5.

Photograph of V(34). Opacities seen as a central plaque macroscopically.

FIG. 6.

VI(70) as seen with the slit-lamp and corneal microscope.

Common Factor

1. All are bi-laterally affected, the eyes having within reasonable limits symmetrical opacities.
2. All commence in the first decade of life.
3. All types have nodules causing irregularity of the corneal surface.
4. Vascularization is absent.
5. Attacks of pain are common.
6. The region of the cornea furthest from the limbus is first affected, and in the case of the dominant forms is the only region affected.
7. The aetiology is unknown.
8. All are resistant to any form of medical treatment.
9. None is associated with any other known disease or defect.

CHIEF DIFFERENCES

	Granular	Reticular	Macular
Shape of opacities ...	Irregular discrete granules	Interlacing lines and nodules	Spots being so numerous as to form a diffuse opacity
Situation of opacities	Axial region	Axial region	Whole cornea but more dense in axial region
Vision ...	Good until middle age	Early affected	Early affected. Reduced to finger-counting in middle age
Transmission ...	Dominant	Dominant	Recessive
Corneal Sensation ...	Good	Defective	Very defective

My Own Investigations*

This investigation began in 1938 and fortunately considerable progress had been made before the outbreak of War. Since hostilities started contact has been kept up with several of the

* The expenses of this investigation were defrayed by a grant from the Medical Research Council.

affected members, but owing to transport difficulties and War duties, it has not been possible to interview certain members of the pedigree.

The investigation commenced subsequent to the diagnosis of hereditary corneal dystrophy in a 31 year old unmarried woman VI(70). She came to the out-patient Department, Chalmers Hospital, Banff, complaining of sore eyes. I had not previously encountered an instance of this disease, and the diagnosis was arrived at through comparing the slit-lamp appearances with the descriptions of nodular corneal dystrophy given in Koby's Slit-lamp Microscopy. When it was realised that the condition was hereditary, I visited the woman's mother and found that she was suffering from a similar disease. This woman was most helpful and her information was found to be very reliable. She was aware that several of her relations suffered from an eye defect of a peculiar nature, and every one she named as possibly being affected, actually suffered from this disease. Reliance could therefore reasonably be placed on her opinion of the eye condition of deceased relatives, upon whom no eye examination had been made by an ophthalmologist. Most of the members of the pedigree are fisher people and fortunately reside in the North East corner of Scotland, in fact twelve of the affected members reside in the same town within half a mile of each other. No difficulty was encountered in interviewing or examining any of the members, all were very friendly and willing to co-operate.

Method of examination.—Miss M., VI(70), the first diagnosed patient, was admitted into the Eye Ward, Aberdeen Royal Infirmary, and a complete investigation was carried out. The corneal opacities were examined by—

1. Naked eye.
2. Lister-Morton luminous ophthalmoscope with +20 D. lens in the aperture.
3. Oblique illumination supplied by the luminous ophthalmoscope with the battery of lenses removed, and the binocular loupe.
4. Slit-lamp and corneal microscope.

In reviewing the literature it was realised that a considerable amount of confusion arose from the fact that no uniform method of examination and magnification was used. When a description of the opacities was given, often it was not stated whether the description was that of low or high magnification. Obviously a more detailed description of the shape and nature of the opacities can be given when examined by the slit-lamp than with the binocular loupe. It was soon realized that the pedigree was of considerable extent and that many could not be examined with the slit-lamp, so with this original patient an effort was made to familiarize

the appearance of the corneal opacities by some means that was easily transportable, and allowed of the patients being examined in their own homes. It was soon found that the best method for diagnosing the condition was by method 2.

The opacities could be easily seen as black opacities against the red fundus reflex (Fig. 4). Their shapes such as disc, circle, club, comma, irregular, fine lines, and their distribution and number, could be easily estimated. In no case was any difficulty found in saying whether the cornea was clear or otherwise, nor was there ever any reason to classify any of the affected patients as reticular corneal dystrophy.

Fortunately few cases were encountered who were below the age when the opacities are known to become visible, and who would possibly have the disease. These young members, actually eight in number, were examined with the slit-lamp with negative result.

Method 3 was then used to ascertain the colour of the opacities and their approximate depth in the cornea. The polish and smoothness of the cornea were observed by daylight in front of a window, and at the same time the naked eye appearance of the cornea was noted as to the visibility or otherwise of opacities. Only in a few cases could the opacities be seen macroscopically. The vision was recorded and the refraction estimated. Corneal sensation was tested by means of a pointed piece of cotton-wool. A careful medical history was taken of all cases examined irrespective of whether they had corneal dystrophy or not. It was soon found that the majority of the affected members had periodic attacks of inflammation and pain, and the description of the attacks was so similar, that they are undoubtedly part of the clinical picture. Note was made of the colour of the iris; a large number had blue eyes, not a single instance was encountered where an affected case had brown eyes. One member had one iris blue and the other brown, but she had not unilateral corneal dystrophy.

With the help of the Galton Laboratory, London, the blood groups of 52 members were determined, no correlation was found between the blood group and the corneal dystrophy.

BLOOD GROUPS

<i>Pedigree No.</i>	<i>Blood Group</i>
IV, 7	A 2 M
V, 5	A 1 N
V, 6	A 1 M N
V, 15	O M
V, 18	O M
V, 20	A 1 M N
Wife of V, 20	A 2 M N
V, 23	O N
Husband of V, 23	O M
V, 24	A 2 B M

BLOOD GROUPS—*continued*

<i>Pedigree No.</i>	<i>Blood Group</i>
V, 27 ...	BM
Wife of V, 27 ...	BMN
V, 29 ...	A2BMN
Wife of V, 29 ...	BM
V, 31 ...	A1M
V, 33 ...	A1MN
V, 34 ...	OMN
V, 37 ...	OM
Wife of V, 37 ...	ON
V, 45 ...	ON
VI, 11 ...	A1MN
VI, 14 ...	ON
VI, 16 ...	A1N
VI, 17 ...	A1MN
VI, 18 ...	A1MN
VI, 32 ...	OMN
Wife of VI, 32 ...	OMN
VI, 35 ...	OM
VI, 40 ...	ON
VI, 42 ...	OMN
VI, 50 ...	OMN
Husband of VI, 50 ...	A1BN
VI, 51 ...	OMN
VI, 53 ...	OMN
VI, 54 ...	OMN
VI, 55 ...	OMN
VI, 57 ...	BMN
VI, 59 ...	BM
VI, 61 ...	OMN
VI, 67 ...	BN
VI, 70 ...	A1MN
VI, 72 ...	OM
VI, 74 ...	OMN
VI, 75 ...	OMN
VI, 76 ...	OMN
VII, 30 ...	OM
VII, 31 ...	ON
VII, 32 ...	ON
VII, 39 ...	BN
VII, 43 ...	OMN
VII, 44 ...	OMN
VII, 57 ...	A1M

MEMBERS WHO HAVE A
PIGMENTED IRIS

<i>Generation</i>	<i>Pedigree Number</i>
VI	4, 7, 9, 56, 66
VII	4, 5, 6, 10, 31, 36, 37, 56, 57, 59, 64, 66, 76, 77, 79, 80, 81, 82

Report on Cases

1. A. L., age 90 years, IV (7). He states that his vision has been bad for a long time and that he has been unable to read the newspapers for at least 30 years.

He was a fisherman up to about 40 years of age, and said that he had good sight up to that time, and could read small print quite well without glasses. He then left the sea and started kippering. From then, and up to about the age of 60, he had frequent attacks of pain, redness and photophobia of the eyes, and his vision gradually got dim. He blames the smoke used to kipper the herrings as the cause of his inflammation and of the subsequent dimness.

When War was declared in 1914 he offered his services as a mine-sweeper, and although he says that he could only read the large letter on the test card, he was passed as fit for mine-sweeping. He was in the Navy for three years and says he managed to perform his duties quite satisfactorily.

Ophthalmological examination: The palpebral fissures are narrow and there is slight palpebral conjunctival redness. No abnormality of the corneae can be seen with the naked eye. The pupils are small and react to light. The tension is normal and the anterior chambers are of medium depth.

R.V.: 1/60, not improved with lenses. L.V.: 3/60, not improved with lenses. There is a central scotoma for red and green. He smokes 2 ozs. black tobacco per week.

The central areas of both corneae are seen to be studded with dozens of small grey spots of various sizes, mostly round discs, but some in the form of rings with clear centres, a few fine lines and some aggregations of dots forming irregular clumps. All are well forward in the cornea. There are several raised nodules on both, making the surface slightly irregular. The area occupied by the opacities is circular and entirely covers the pupil. There is a round white corneal opacity, about 3 mm. in diameter near the limbus down and out in the right eye. Apart from this isolated opacity the peripheral zones of both corneae are entirely transparent and clear from opacities. The corneal sensation is normal.

He states that when young he had three attacks of rheumatic fever and was in bed on each occasion for nine weeks. He has had no other illnesses.

2. JAMES L., age 62 years, baker, V (20). He has known for many years that his vision was poor. He was rejected for military service during the Great War owing to poor eyesight. He has been continuously employed as a baker, and is still working at his trade. He has been unable to read the newspapers since 1930.

He was examined in the Eye Department, Aberdeen Royal Infirmary, in 1932, and the following description is abstracted from the notes.

Pupils unequal, left being greater than the right. Both react poorly to light. Tension normal.

There are numerous grey spots situated in the substance of both corneae, covering rather more than the pupillary area. In places the opacities form streaks and are at varying depths. Some are like commas, some cones and some strands.

R.V.: 5/60, not improved with lenses, with +8.0 D. sph. J. 10. L.V.: 6/60, not improved with lenses, with +8.0 D. sph. J. 10.

Corneal sensation normal.

With the ophthalmoscope there is a dull red reflex, but no details of fundi can be seen. Fields with hands full, no central scotoma for colour.

When I examined him in 1938 he was suffering from angular conjunctivitis. The pupils were small, the left greater than the right. Both reacted actively to light. Tension was normal.

R.V.: 5/60, not improved with lenses, hypermetropia about 2.0 D. sph. L.V.: 4/60, not improved with lenses, hypermetropia about 2.0 D. sph. With +5.0 D. sph. added J. 20.

Corneal sensation normal. Surface irregular. Corneal opacities seen with naked eye, occupying the central area of both corneae and covering the pupils. The area of the opacities is roughly circular and occupies an area of about 2/3 of the cornea. With the ophthalmoscope the opacities are seen to consist of small discs and rings mostly, but there are a number of larger irregular clumps with crenated edges. Some of the aggregations of dots have a feathery-like formation. Most are greyish-white, but some are slightly brown. All the opacities are situated in the superficial layers of the cornea.

No details of the fundi could be seen with the ophthalmoscope.

3. JANE C., age 30 years V (23). (Dr. Usher's Notes—May 12, 1902.)

Peculiar Corneal Opacities.

Comes on account of small swelling in R.

R.V. : 6/6p. Hm. +0.5. J.1 at 10 ins. L.V. : 6/6p. Hm. +0.5. J.1 at 10 ins.

R. nearly circular semitransparent 2 mm. diameter hemispherical jelly-like swelling at sclero-corneal margin.

Pupils active, size $3\frac{1}{2}$ mm., anterior chambers perhaps slightly too deep.

Something noticed on R. by someone 6 months ago. It had same appearance as at present. It gave no trouble until 1 or 2 months ago. Since then she has had slight attacks of redness, with slight photophobia, with a little matter at the canthi.

L. has not been affected. H. and c. to both. Pupils well dilated and circular. There is no evidence of old iritis. No opacities in the vitreous. In the L. there is a small patch on the upper and inner quadrant of the disc.

Both Corneae : The spots are symmetrical, they are all white and sharply defined, all are much about the same size, but vary in shape. They are thickly and evenly distributed over most of the surface of the cornea, but there is quite a definite clear part of the cornea separating them from the corneal margin. In both this clear area is broadest above. The dots are quite superficial and are mostly circular. Some of the circles are of uniform whiteness throughout looking like small discs. Others consist of a small ring enclosing a clear part of the cornea. Other discs have simply a minute transparent part in the centre. There are a few crescents and one or two linear opacities. These are connected with one or two of the discs. There are also a number of minute white dots. Cornea quite smooth on surface.

She has been a healthy woman. Typhoid, otherwise quite healthy. Married and has 5 of a family, no miscarriages. Patient is oldest of a family of 8. All well. No eye trouble. Both parents alive and well. No relation to each other.

July 12, 1902. There is no change in the condition, surface of cornea not raised. There is no haze of cornea between the dots. Thyroid gland not enlarged.

My own Examination : Eyes last examined by Dr. Usher in 1902. Since then there have been frequent attacks of pain and inflammation of both eyes. Pain was

very severe and she had to go to bed. Attack lasted for about a week. Last attack 18 months ago. Vision has been very dim for several years.

At junction of middle and inner $\frac{1}{3}$ of Right cornea there is a dense white vertical opacity. Between this opacity and edge of cornea there is a vascularized less dense opacity. There is a well marked complete arcus senilis which is dented opposite the dense opacity.

R.V. : 6/60, no improvement with lenses. L.V. : 5/60, no improvement with lenses with +3.0 D. sph. J.2.

Dr. Usher re-examined this patient the same day and said that the central corneal opacities occupy more of the clear cornea than they did in 1902.

The opacities consist of discs, circles and lines but mostly irregular clumps. Some of the opacities are pigmented. Opacities seen with naked eye. Surface of cornea smooth. Corneal sensation normal.

4. ALEXANDER MCC., age 67 years. V (25). He declares that his eyes have always been good, but lately does not see so clearly at a distance. He uses someone else's glasses for reading.

No pain or redness of eyes.

In the centre region of both corneae there are the typical opacities.

—0.5 D.sph.

R.V. : 6/12 with $\frac{\text{---}0.5 \text{ D.sph.}}{\text{---}0.5 \text{ ax. } 90^\circ} = 6/9$. L.V. : 6/12 with —1 D.cyl. $90^\circ = 6/9$. With

+2.5 D. sph. J.1.

5. WILLIAM MCC., age 62 years, fisherman. V (27). This patient has not had his eyes examined previously. He has no complaint about vision, has not previously worn glasses and can read the newspapers easily with the naked eye. No history of any attacks of pain or inflammation. Eyes quiet. Pupils active.

R.V. : 6/60, with —3.5 D. sph. 6/12. L.V. : 6/18, with —1.0 D. sph. 6/6.

In the central region of both corneae there are the typical opacities found in the other members of this pedigree. The opacities are of two main varieties as seen with the ophthalmoscope and plus 20 D. lens, (a) irregularly shaped opacities well spaced and scattered between, (b) small round spots.

6. JOHN MCC., age 40 years (1930). V (29). Copied from Aberdeen Royal Infirmary Notes—February, 1930. Corneal degeneration, familial guttae.

Complaint : Not seeing well with present glasses. Eyes very sore.

R.V. : 1/60, with —10.0 D. sph. = 6/60, without any glasses. J.14 close.

L.V. : 1/60, with —10.0 D. sph. = 6/12, without glasses J.1 close.

Tension normal, pupils equal and active to light. Palpebral conjunctivitis. Both corneae studded with greyish spots, some circular, some curved. The right fundus shows myopic macular changes.

I examined him along with Dr. Usher in his own home on August 21, 1938.

Both eyes slightly red, pupils active, tension normal. Both corneae studded with grey dots, clumps and circles. No roughening of surface. Sensation normal. Periphery of cornea clear.

R.V. : With glasses 1/60. L.V. : With —12.0 D. sph. 6/12.

He says that he has often had attacks of inflammation that last for about a week.

7. JOHN L., age 69 years (1938). V (30). He says that he has had frequent attacks of inflammation of the eyes, an attack lasting for about 3 days. Pain very severe in and above the eye prevents sleep. Attacks were more frequent when snow was on the ground and after being exposed to wind. He has been a fisherman all his life and says that he had no trouble with his eyes up to the age of 20. He has suffered from right-sided trigeminal neuralgia for nine years. He has had no other previous illnesses. The left pupil is greater than the right, both react to light. Central region of both corneae studded with discrete dots and a few clumps. Surface of corneae smooth, sensation normal.

This patient died on April 5, 1943. I was fortunate to get permission from his next-of-kin to remove his eyes and this was done about an hour after death.

8. ANDREW L., age 67 years, fisherman. V (31). He says he never had good eyesight. No attacks of pain or inflammation. Has glasses for reading but never had glasses for outside use. He is a line fisherman and often fishes for crabs. Crab traps have to be placed overnight, and the situation of the trap indicated by means of a buoy and flag. He says he can steer his small boat out and into harbour and can see the trap flags. He has had two sons. One died age 8 months. The other was accidentally killed aged 25 years. According to his father his eyes were examined by a Post Office doctor 2-3 years previous to his death, and were said to be perfect.

Eyes quiet. Pupils active, tension normal, deepish anterior chambers. Corneal sensation defective. Surface of cornea smooth. Opacities not seen with the naked eye.

—1.5 D. sph.

R.V. : 6/60 with $\frac{\text{---}1.5 \text{ D. sph.}}{\text{---}4.0 \text{ D. cyl. } 90^\circ} = 6/12$.

—4.0 D. cyl. 90°

—3.0 D. sph.

L.V. : 2/60 with $\frac{\text{---}3.0 \text{ D. sph.}}{\text{---}5.0 \text{ D. ax. } 90^\circ} = 6/36$. With +3.5 D. sph. added J.1.

—5.0 D. ax. 90°

Corneal opacities consist of numerous grey spots, some irregular clumps and several fine lines. All are in central region of the cornea.

9. ALEXR. L., age 63 years. V (33). Came to have his eyes examined as he could not recognise people in the street. He says his distance vision has been bad for more than 20 years, but that he saw well before the Great War. He has not previously had his eyes examined or worn glasses. For many years he suffered from occasional attacks of pain and redness of the eyes. He thought the inflammation was caused by the wind that was always present in the wheel-house of the drifter

upon which he worked. He admitted, however, that his eyes were more sensitive than his fellow seamen to the cold wind. He is married but has no family.

Ophthalmological Examination: Eyes quiet, pupils equal and active, tension normal. No abnormality of cornea seen with naked eye.

—9.5 D. sph.

R.V.: 4/60 with ————— = 6/12. With + 4.0 D. sph. added, J.I.

—0.5 D. cyl. 180°

—9.0 D. sph.

L.V.: 2/60 with ————— = 6/18. J.I. well.

—0.75 D. cyl. 180°

With the ophthalmoscope it is seen that both corneae are studded in the central region with discrete opacities. Some disc shaped, some circles and several fine lines. All opacities are separated by clear corneal tissue. The periphery of both corneae is clear. Corneal surface is smooth and the sensation is normal. Both lenses clear. Fundi normal.

10. MRS. MARY JANE M., age 58 years. V (34). Dimness of vision for many years. Never previously had eyes examined.

At the age of 16 she had inflammation of the eyes with pain and photophobia. Up to about 40 years of age there was a recurrence of the inflammation about twice yearly. After each attack vision was always worse. General health has been good. She has had three of a family.

Eyes quiet. L. pupil greater than R. Both active to light. Tension normal. Movements full.

Irregular grey opacities seen with naked eye on centre of both corneae occupying a slightly larger area than the pupil. Corneal sensation normal. No irregularities of surface. The corneal opacities of this patient are larger and more numerous than are present in any of the other cases observed; when viewed at a few feet distant the opacities are seen as a grey plaque occupying the centre of the cornea and covering the pupil (Fig. 5).

R.V.: 2/60 with —10.0 D. sph. 6/36 (—1):

L.V.: 2/60 with —10.0 D. sph. 6/36 (—1). Looking sideways J.16.

H. and c. to both. Pupils dilated and round. Lenses clear. Fundi appear normal, without lens reads J.2 close. Says she can read the large newspaper type if she shades her eyes.

Advised gutt. atrop. sulph. $\frac{1}{2}$ per cent. twice weekly to right.

Glasses prescribed —10.0 D. sph. R. and L.

One week later: R. red and lids swollen, *atrop. irritation*. Pupils dilated, advised to stop atrop. (Only one application had been used.)

One week later: Eyes quiet, sees much better with her glasses.

11. JAMES L., banker, age 23 years (1906). V (36). Copied from Dr. Usher's notes, Aberdeen Royal Infirmary, January 2, 1906.

Peculiar symmetrical corneal opacities. Dimness of vision, worse in L. for 3 weeks. Previous attacks 3 to 4 years ago. Pain and redness.

+1.0 D. sph.

R.V.: 6/6p. with ————— = 6/6.

—1.5 D. cyl. ax. 120°

+1.5 D. sph.

L.V.: 6/60 with ————— = 6/12.

—2.5 D. cyl. ax. 30°

Pupils circular and react to light. Homatropine to both. Pupils dilated and circular. Oph. fundi normal. No choroiditis, examined far forwards. No vitreous opacities. Symmetrical punctate opacities of corneae. They occupy nearly the whole of the cornea, but the margin is quite free from spots.

The opacity consists of a large number of faint grey dots with a diffuse very faint opacity between dots. On magnification the dots are found to be circular and in some cases ring shaped. They do not appear to be raised. None of them is pigmented.

May 29, 1906. With his glasses—R.V. 6/6, L.V. 6/12.

12. MARGARET L., age 50 years, music teacher. V (45). She has known that something peculiar was wrong with her eyes since she was 18 years of age. At that time she was examined at the Aberdeen Royal Infirmary and glasses were prescribed. Since then she has had occasional attacks of inflammation with great pain. There were years between the attacks, and an attack lasted from one to two weeks. Pain was so severe that it kept her from sleeping, and she was most comfortable in darkness. She has had no serious illnesses. On naked eye examination her eyes appear normal.

$$\text{R.V. : } 6/12 \text{ with } \frac{+0.75 \text{ D. sph.}}{+0.5 \text{ D. cyl. } 60^\circ - 0.75 \text{ D. sph.}} = 6/9 \text{ mostly.}$$

$$\text{L.V. : } 6/18 \text{ with } \frac{-0.75 \text{ D. cyl. } 30^\circ}{-0.75 \text{ D. cyl. } 30^\circ} = 6/12 \text{ mostly. With } +1.5 \text{ D. sph. added J.I.}$$

With the ophthalmoscope and 20 D. lens the pupillary area of each cornea is seen to be covered with small discrete dots with clear cornea between. There is no generalized haze. The affected area is circular and the peripheral zone is quite clear. Most of the dots are grey coloured but some have a brownish tint. The corneal sensation is normal and the surface regular. Both lenses are clear and the fundi normal. She had two brothers, one is still alive, age 58 years, and has 6 of a family. His eyes are clear and likewise the eyes of all the family. Another brother died, age 32 years, of some obscure cerebral disease. On examining the Aberdeen Royal Infirmary's notes I found that his temperature was 108° F. and that he was paralysed on the left side. His eyes were examined then by Dr. Usher and apart from a left internal strabismus nothing abnormal was seen.

13. MRS. C., age 40 years. VI (36). This patient came to see me at my request. Eyes not previously examined.

She has had several attacks of inflammation of the eyes with severe pain and photophobia. The attacks always come on when she is not feeling well. The last attack was 4 years ago, when she was pregnant. She has no complaint about vision.

Eyes quiet, no opacities seen by naked eye examination. Deepish anterior chambers. Pupils equal and active. Tension normal.

$$\text{R.V. : } 6/6p. \text{ with } +0.5 \text{ D. sph.} = 6/6 \text{ mostly.}$$

$$\text{L.V. : } 6/9p. \text{ with } \frac{+0.5 \text{ D. sph.}}{+0.75 \text{ D. cyl. ax. } 105^\circ} = 6/9.$$

In the axial region of both corneae there are numerous grey opacities. Some are very small and round, but mostly they are irregular shaped, some club shaped. No lines seen. Surface of cornea is regular and sensation normal.

14. MRS. C., age 38 years. VI (37). Came to have her eyes examined at my request.

Is a very nervous woman and thinks all the family suffers from tuberculosis, as that was the disease that her father died of. She has had no serious illnesses, no trouble with her eyes, sees well both far and near and has never worn glasses. Is married and has one girl.

Eyes quiet. Pupils active. Anterior chambers deeper than normal. Corneal opacities seen with naked eye. Spots very numerous, irregular shaped mostly, a number of fine lines present some right-angled formation but no reticulation. Periphery of cornea clear. Surface of cornea smooth. Sensation normal.

Daughter's eyes examined and found to be normal.

15. MRS. P., age 36 years. VI (38). She came to have her eyes examined at my request. Says she sees well and has had no trouble at all with her eyes.

$$\text{R.V. : } 6/9p. \text{ with } -0.75 \text{ D. cyl. ax. } 150^\circ = 6/6p.$$

$$\text{L.V. : } 6/12 \text{ with } -1.0 \text{ D. cyl. ax. } 30^\circ = 6/6.$$

Corneal opacities seen with naked eye. With the ophthalmoscope and +20.0 D. lens, the opacities are seen to be disc shaped but mostly irregular outline as though some of the discs had joined. All are far forward in the cornea. Surface of cornea smooth. Sensation normal.

16. MARY JANE L., age 16 years. VI (42). She had not previously had her eyes examined and had no complaint about her eyes, or vision. She has had no serious illnesses, and is healthy in every way. On naked eye examination her eyes look normal.

$$\text{R.V. : } 6/18 \text{ with } \frac{+1.0 \text{ D. sph.}}{-1.5 \text{ D. cyl. } 180^\circ + 0.75 \text{ D. sph.}} = 6/9 \text{ and J.I.}$$

$$\text{L.V. : } 6/9 \text{ with } \frac{+0.75 \text{ D. sph.}}{-1.25 \text{ D. cyl. } 180^\circ} = 6/6 \text{ mostly and J.I.}$$

The pupils are large, equal and react normally to light and near vision. The corneal sensation is normal and the surface is regular. There are numerous small grey and brownish spots confined to central area of cornea of various sizes, a few comma shaped, a few ring shaped, and a few have a fine thread-like process running

in a horizontal direction. There is no reticulation. The areas between the opacities are quite transparent. All the opacities are far forward in the substance of the cornea. The peripheral zone is quite clear.

With the ophthalmoscope and +20 D. lens the opacities are shown up as black spots against the red reflex of the fairly large pupil, and the shape of the opacities can be very accurately observed. The fundi are normal.

17. ALEXANDER C., age 47 years, seaman. VI (52).

He has been a healthy man all his life. No previous examination of his eyes made. He has never had glasses, but now has some difficulty with his near work.

Vision of both eyes 6/9, no refractive error, with +1.5 D. sph. J.I. Pupils active; corneal sensation normal; cornea smooth.

Grey spots in the central region of both corneae, spots fairly large but well spaced and clear cornea between spots. Peripheral zone clear. Fundi normal.

18. JOHN C., age 33 years, fisherman. VI (55). Eyes not previously examined. Has had transient attacks of inflammation, felt as though sand was in eyes. Thinks he can see perfectly. Is married but has no family.

With naked eye his eyes appear normal. Corneal sensation normal.

—0.25 D. sph.

R.V.: 6/18 with $\frac{\text{—0.75 D. cyl. 90}^\circ}{\text{—0.25 D. sph.}} = 6/6.$

L.V.: 6/18 with $\frac{\text{—0.75 D. cyl. 60}^\circ}{\text{—0.25 D. sph.}} = 6/9. \text{ J.I.}$

Fundi normal. Corneal opacities are small spots only, well spaced. Killed by enemy action.

19. ARCHIBALD MCC., age 21 years. VI (61). Says right eye has been dim as long as he can remember. No attacks of redness or pain. Eyes quiet. Pupils equal and active, no squint of right.

R.V.: 6/60. Refraction mixed astigmatism, not improved with lenses. L.V.: 6/6.

Opacities on both corneae consist of very fine dots, circles and a few fine horizontal lines. No fundus abnormality seen. His brother, James, had been operated upon for congenital ptosis six years previously. He has 6 D. of myopia, vision with glasses 6/12. Both corneae clear.

Another brother, Wm. George, said his left eye has always been dim.

R.V.: 6/6. L.V.: 1/60, refraction 16 D. of myopia, but no improvement with glasses. The mother's eyes were examined and it was found—R.V.: 6/60 with +3.0 D. sph. 6/6. L.V.: 6/60, mixed astigmatism, not improved with lenses.

20. GEORGE P. MCC., age 17 years. VI (67). He has not had his eyes previously examined. Says he sees well and has had no attacks of pain or redness of his eyes.

Eyes quiet; macroscopically corneae appear normal. Deepish anterior chambers. Blue-grey iris. Pupils equal and active.

R.V.: 6/9 not improved. L.V.: 6/6 mostly, not improved. Fundi normal.

In pupillary region of both corneae there are numerous grey opacities, mostly small discs but some circles with clear centres. All are discrete and are situated well forward in the cornea.

21. INA M., age 31 years, domestic servant. VI (70). *Complaint*: Eyes sore, not seeing so well both far and near.

Duration: Has had attacks of sore lids 2-3 times a year ever since she left school. Eyes very red and painful two months ago.

Past History: Took fits at age 15-16 and took one per month till last year, more since then. Bronchitis a year ago. No trouble with menstruation. Bowels regular. No cough.

Ophthalmological Examination: Borders of lids red, no ciliary injection. In the central region of both corneae are grey spots, some very small and some larger. None can be seen with the naked eye. All are in the substance of the cornea. No irregularity of corneal surface. Corneal sensation normal. The majority of the spots are circular dots, but some are ring-shaped and there are a few fine horizontal lines. No criss-cross formation. More numerous on L. than on R.

Vision—R. and L., 6/12 and J.I. Gutt. atrop. sulph. 1 per cent. to both eyes twice weekly. Cod-liver oil by mouth.

October 22. Eye red, lids swollen, pupils dilated. (Atropine irritation.) Atropine discontinued. Admitted into Eye Ward, Aberdeen Royal Infirmary.

General Examination: Chest normal. Abdomen normal. Heart normal. B.P. 126/80. Knee jerks and ankle jerks present. Urine—albumen—negative. Sugar—negative. X-ray of chest and sinuses "calcified scars at R. hilus, no other evidence of infection. Paranasal sinuses clear. W.R.—negative.

Slit-lamp Examination: Anterior and posterior surfaces of corneae clear. All spots lie between Descemet's and Bowman's membranes. The spots occupy the central area of the cornea covering about a diameter of 5 mm. The spots vary in size but there appear to be two distinct varieties, (a) small circular spots with regular edges, and (b) larger circular spots with crenated edges, giving them an icing sugar appearance. There are a few horizontal fine lines.

Vision—R. and L., 6/6 partly. Discharged December 31, 1937. Advised to take two capsules of Halibut Liver oil daily.

March 20, 1940. Says she is not seeing so well. Since last seen in 1937 she had one attack of redness and pain. The pain was very severe and she had to go to bed. She said that the taking of the Halibut liver oil capsules kept her from taking the attacks, as she stopped the capsules about two months before her eyes became sore and red.

The corneal opacities can now be seen with the naked eye. Slight ciliary injection in the palpebral fissure of both eyes. Vision—R. and L., 6/9. The spots are more numerous and larger than when last examined.

February 14, 1942. Opacities seen easily with naked eye and irregular shaped clumps and some circles. Vision—R., 6/9, L., 6/6. No more attacks of redness.

22. Mrs. R., age 29 years. VI (72). She has had several attacks of pain and redness of her eyes similar to the attacks suffered by her sister Ina. She has never had her eyes previously examined and sees well. No opacities seen with naked eye.

R.V.: 6/12 with +0.5 D. cyl. ax. 60°, 6/6. L.V.: 6/6p. with +0.5 D. cyl. ax. 90°, 6/6.

With the ophthalmoscope typical opacities seen in the central region of each cornea, mostly discs but a few circles. Opacities well separated. No other abnormality seen.

23. ALEXANDER C., age 12 years. VII (35). This boy had no complaint about his eyes. No attacks of pain or redness.

R.V.: 6/6. L.V.: 6/6 no hm. Naked eye appearance normal.

With transmitted light and binocular loupe no opacities seen. With the luminous ophthalmoscope and +20 D. lens there are visible minute black dots in the central region. The spots are well separated and number about 20 on the right eye and 30 on the left.

HELEN C., age 18 years (1943). VII (42). Eyes examined at my request, no complaint. No serious illnesses. Vision = 6/6, no refractive error. Macroscopically eyes appear normal.

Fine grey spots in central region of both corneae all round and well spaced. Periphery of cornea clear, sensation normal; surface of cornea smooth.

Details of Pedigree

I have been able to trace the pedigree to two brothers, Alexander (Councillor) I(2) and James (Snappy) I(3).

Alexander was twice married. By his first wife he had Alexander II(1), (who died without issue) and James II(2). By his second wife he had eight of a family. The members suffering from corneal degeneration are descendants of James II(2). I have not made a detailed examination of the descendants of the second family, but have examined about twelve members and they are all negative. All information received about this branch of the family makes it unlikely that there are any affected. It would appear therefore that the disease was transmitted through Councillor's first wife I(1).

ESSENTIAL PARTICULARS ABOUT AFFECTED MEMBERS

Pedigree No.	Sex	Age	Occupation	Uncorrected Vision		Refractive Error	Corrected Vision		Attacks of pain	Colour of Iris	Blood Group	Any Other Defect
				R.	L.		R.	L.				
IV V	M.	90	Fisherman	1/60	3/60	H.	Not improved	Yes	Blue	A.2.M.	Tobacco Amblyopia	
	M.	62	Baker	5/60	4/60	H.	Not improved	Yes	Blue	A.1.M.N.	Conjunctivitis	
	F.	68	Housework	6/60	5/60	H.	6/60	6/60	Yes	Blue	O.N.	None
	M.	67	Fisherman	6/12	6/12	M. Astig.	6/9	6/9	No	Blue	A.2.B.M.	None
	M.	62	Fisherman	6/60	6/18	M.	6/12	6/6	No	Blue	B.M.	None
	M.	48	Cooper	1/60	1/60	M.	6/60	6/12	Yes	Blue-grey	A.2.B.M.N.	None
	M.	69	Fisherman	6/60	6/36	M. Astig.	6/12	6/12	Yes	Grey	—	Trigeminal Neuralgia
	M.	67	Fisherman	6/60	2/60	M. Astig.	6/12	6/36	No	Blue	A.1.M.	None
	M.	63	Fisherman	4/60	2/60	M. Astig.	6/12	6/18	Yes	Blue	A.1.M.N.	Arthritis
	F.	58	Housework	2/60	2/60	M.	6/36	6/36	Yes	Blue	O.M.N.	None
VI	M.	22 (1906)	Banker	6/6p	6/60	Mix. Astig.	6/6	6/12p	Yes	Blue	—	None
	F.	50	Music Teacher	6/12	6/18	H. Astig. M. Astig.	6/9m.	6/12m	Yes	Greenish	O.N.	None
	F.	40	Housework	6/6p	6/9p	H. Astig.	6/6m.	6/9-	Yes	Blue	—	None
	F.	38	Housework	6/12	6/9p	M. Astig.	6/9	6/9m.	No	Blue	—	None
	F.	36	Housework	6/9p	6/12	M. Astig.	6/6p	6/6	No	Blue	—	None
	F.	16	Shop Assist	6/18	6/9	Mix. Astig.	6/9	6/6m	No	Blue	O.M.N.	None
	M.	47	Seaman	6/6	6/6	E.	6/6	6/6	No	Blue	—	None
	M.	33	Fisherman	6/18	6/18	M. Astig.	6/6	6/9	Yes	Blue	O.M.N.	None
	M.	21	Grocer	5/60	6/6	M. Astig.	Not improved	6/9	No	Blue	O.M.N.	None
	M.	17	Baker	6/9	6/6m	H.	Not improved	6/9	No	Blue-grey	B.M.	None
VII	F.	31	Domestic	6/18	6/12	H. Astig.	6/9	6/6	Yes	Blue	A.1.M.N.	Fits
	F.	27	Housework	6/12	6/6p	H. Astig.	6/6	6/6	Yes	Blue	O.M.	None
	M.	12	School Boy	6/6	6/6	E.	6/6	6/6	No	Greenish	—	None
	F.	18	Shop Assist.	6/6	6/6	E.	6/6	6/6	No	Blue	—	None

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- Unaffected male
 ○ Unaffected female
 ● Affected (examined by ophthalmologist)
 ⊗ Probably affected (not examined by ophthalmologist)
 + Deceased
 × Examined by ophthalmologist
 ⊗ Congenital cataract
 ○—○ Twins
 ○ Blood grouped
 ⊙ Pigmented iris

No one is now alive who remembers James II(2), so that it is impossible to know the condition of his eyes. Several people remember about the members of the next generation. Santy III(1) certainly had bad eyes and often suffered from attacks of inflammation, as a grandson remembers that he bathed his eyes frequently. His brother III(2) went to Canada and worked for the Hudson Bay Company. There is no evidence that there was anything wrong with his eyes. Katie, his sister, III(3) had poor eyesight and was unmarried. Santy III(1) had six sons and two daughters.

The oldest son, James IV(1) had apparently normal eyes and all descendants examined are clear of the disease. His daughter V(6) married a man who suffers from congenital cataract. Three of their children also suffer from this disease and one, VI(19) is in a Mental Hospital. No other abnormality has been found in any other member of this branch of the family.

Barbara IV(2) died at the age of 87 years, and was said to have had good eyesight.

George IV(3) was drowned at sea at the age of 49 years, and no one now living ever heard that he complained about his eyes. In all probability, however, he had corneal dystrophy as several of his descendants are affected:—V(20), VI(36), VI(37), VI(38), and VII(35).

I have been able to examine all the living members of George's family IV(3) who are in this country. Several other members are alive, but live in the U.S.A. and in Canada. I feel confident that some of them will have corneal dystrophy.

Jean IV(4) had an illegitimate daughter V(23) who is still alive and has corneal degeneration. Dr. Usher examined this daughter in 1902 and diagnosed corneal degeneration, but the name not being the same he was unable to place her in the same family as the only other case of corneal degeneration he had a record of V(36). This illegitimate daughter is a cousin of this other case. Jean IV(4) married and had three sons and three daughters. Her husband is still alive and is said to be aged 102 years. V(23) as already stated has corneal dystrophy. She married and had six of a family two of whom VI(52) and VI(55) have corneal dystrophy. The daughter of VI(52) is also affected VII(42). VI(55) was married but has no family and was killed by enemy action. V(24) is affected. He is married, but has no family. V(25) is dead. She was married but had no family and there is no record about her eyes. Margaret V(26) is also dead and there is no information about her eyes. She has one daughter VI(56), who has not corneal degeneration, but has one blue iris and one brown. William V(27) is affected. He is married and has 5 sons. One son is affected, but I have been unable to examine 2 members

of this family, so that in all probability there will be more affected. Another hereditary disease comes into this branch of the family, namely anisometropia, one eye being highly myopic. This is transmitted through the mother who is the wife of V(27). VI(57) is affected in this way and also VI(59) who has in addition congenital ptosis. V(28) is clear and also all the members of her family examined. John V(29) is affected and also his only son VI(67).

Andrew IV(5) according to his daughter had bad eyes, and from the description given had corneal dystrophy. He married a sister of IV(4)'s husband and had seven sons and three daughters. The oldest son John V(30) is affected. He was unmarried. This member has since died and I was fortunate to get his eyes for histological examination.

Andrew V(31) is affected. He had two sons. One died young and the other died at the age of 22 years.. No examination of the eyes of either was made at any time.

Barbara V(32) died at the age of 18 years and she had at no time complained about her eyes.

Alexander V(33), is affected, married, but has no family.

Mary Jane V(34) is married and has three of a family. This member has the disease in its most advanced form of any member examined. Her two daughters are affected, and it was one daughter VI(70) in whom the disease was first discovered.

This daughter as already stated is unmarried. The other daughter is affected and has two children, but as yet there is no sign of the disease in either, but both children have the typical blue eyes of this pedigree.

The son is married and has one child, but both are clear of the disease.

William V(46), was accidentally drowned in the harbour at the age of 17 years. He had defective vision at school, being so bad that he had to be led home by someone else. He may have had corneal dystrophy, but in addition he must have had some other eye disease, probably congenital cataract as there was no history of any inflammatory trouble.

James V(36) was a twin. He was affected. He was examined and diagnosed by Dr. Usher in 1906. He had one son and one grandson, both being clear of the disease. This member died abroad a few years ago.

George V(37) is his twin brother and is alive and is unaffected, and also all the members of his family.

Ellen V(38) was married and had nine of a family. She died at the age of 42. Her eyes were never examined and there is no evidence that she had the disease. All her descendants who have been examined are found to be clear.

Archibald V(39) died at the age of three. His eyes had never been examined,

William IV(6). There is no evidence that this member suffered from corneal dystrophy. He had four of a family, but only one is now alive V(40), he is unmarried and he has not the disease.

Descendants of another brother V(42) are also clear of the disease.

Alexander IV(7) was affected. He was the oldest living member of the pedigree alive when this investigation commenced, he has since died at the age of 91 years. He had two sons and one daughter. The daughter is affected and is unmarried. The other two sons are unaffected, and also all the descendants of V(44).

John IV(8) was accidentally drowned in the harbour. According to his niece his eyesight was defective, so he could have had corneal dystrophy.

Analysis of Affected Cases

Sex.—Males 14, females 10. Males predominate, but in some pedigrees females outnumber the males.

Age.—The youngest affected is a boy aged 12 years, and the oldest was aged 90 years when examined; 8 of the affected were over 60 when examined.

Occupation.—The majority are engaged in occupations which do not require perfect vision. One is a music teacher, but as corrected vision is still 6/9, she has no difficulty with her work.

Vision and refractive errors.—I consider it necessary to analyse these two together, as the nature of the refractive error exerts a considerable influence upon the visual acuity. Very few of those affected complained of having poor vision; in fact a considerable number asserted that their vision was perfect. Most had a refractive error and several had myopia of fairly high degree, so that even without any corneal defect, it was not surprising that the visual acuity was not normal after correction. Fourteen suffered from myopia or myopic astigmatism. I am inclined to think that the low myopia and astigmatism were due in some cases to the corneal lesion, but in others, that the myopia was due also to heredity unconnected with the corneal disease. In support of this view those with high myopia were confined to two families and all had myopic crescents, and V(29) had also other myopic fundus changes. VI(61) had one defective eye which was unconnected with the disease, but was due to another congenital defect, the mother and two sons all having marked anisometropia.

With these exceptions the vision of both eyes differs very little from each other. When we come to analyse to what extent vision is actually affected by the corneal lesion, we find that up to the age

VITAL STATISTICS

Generation	Males	Females	Members with no Offspring	Number of individuals in each family		Age at Death	Members still Alive	Un-natural Deaths
III	2	1	2	8		Over 70	None	—
IV	6	2	2	12, 9, 7, 10, 4, 3		79, 87, 51, 85, 89, 76, 91, 35	None	Drowned 2
V	27	18	20	2, 8, 9, 2, 3, 2, 2, 5, 8, 5, 3, 4, 3, 6, 1, 5, 5, 1, 2, 3, 1, 3, 9, 2, 6		1, 1, 28, 26, 37, 42, 19, 1, 2, 4, 33, 41, 39, 73, 18, 17, 52, 42, 3, 54, 33, 37, 32	22	Drowned 4 Suicide 1 Burned 1
VI	45 Sex Unknown 7	48	65	1, 2, 3, 2, 1, 2, 1, 1, 2, 2, 1, 1, 4, 1, 5, 4, 3, 1, 4, 1, 1, 3, 2, 2, 2, 2, 1, 2, 1, 6, 6, 3, 1, 5, 8		23, 14, 34, 1, 24, 31, 25, 1, 1, 35, 1, 25, 29, 36, 26, 15	84	Enemy Action 2 Drowned 2 Fall from Height 1
VII	43	44	87	—		2, 6	85	Homicide 1

of 50, corrected vision is in no instance less than 6/12. V(34) has the most dense corneal opacities of all those affected, and she also had 10 D. of myopia, her corrected vision is however, 6/36, and her age is 58. The visual acuity does not however suffer an irresistible diminution on the advance of age, as 4 affected have still 6/12 vision or better when well over 60 years of age. IV(7) had tobacco amblyopia and refused to give up his tobacco.

Only one case could have been certified blind—a male over 90 years of age. All those of military age would actually pass grade one with the exception of VI(61) who had a defective eye from another cause. All the others are able to carry on their usual occupation, including the baker V(20) whose vision is only 5/60. In no case, therefore, is any affected individual a burden upon the State or relatives. Bucklers (1938) states that compulsory sterilization of those affected with granular corneal dystrophy is not justified upon the grounds of defective vision, but upon the fact that it is a dominant disease, the transmission of which should be prevented by sterilization.

Differential Diagnosis

As the disease is most likely to be encountered in a routine ophthalmological examination, the eyes will be white. The condition is unlikely to be confused with any acute inflammatory disease such as corneal ulceration or superficial punctate keratitis, but as no staining takes place with fluorescein any loss of superficial epithelium can soon be ascertained.

The size and distribution of the opacities are very similar to "K.P." so the condition could easily be mistaken for cyclitis. When examined with the slit-lamp, however, the opacities of granular corneal dystrophy will be found to be in the substance of the cornea.

While examining an old granite polisher recently, I was struck with the similarity of the small, numerous, corneal nebulae that he had, to the appearance of hereditary corneal dystrophy. The lesions, however, are all on the surface, but the size and distribution of the opacities are exceedingly alike.

Salzmann's nodular dystrophy is often uni-lateral, not familial and the nodules are larger and fewer than granular dystrophy.

Any vascularization of the cornea or the remains of blood vessels indicates that the condition is not granular corneal dystrophy, but does not exclude the possibility of granular corneal dystrophy plus, say, interstitial keratitis.

To summarize, the lesion is not granular corneal dystrophy if it is:—

1. Uni-lateral.
2. Shows signs of vascularization.
3. Opacities visible on naked eye examination in a patient aged under 30 years.
4. Absence of a similiar condition in both parents.

Pathology

I do not think it possible to find another tissue in the body which can permit a more exact examination during life of any pathological lesion that may be present than the living cornea. The cornea has no counterpart in any other tissue in the body in respect of its combination of the five following characteristics:—

1. Optical homogeneity.
2. Regularity of surface.
3. Avascularity.
4. Mode of nutrition.
5. Clinical accessibility.

I consider that some of these factors have an influence on the pathology of corneal dystrophy.

As there is not an exact counterpart in the body it is not surprising that one encounters a disease confined solely to this specialized tissue. Arguing along similar lines one cannot fail to come to the conclusion that the lesion is in some way connected with the specialized mechanism of this tissue. The only other tissues in the body that are at all similar are articular cartilage, it being avascular, but comparatively non-sensitive and opaque, and the crystalline lens it being avascular, transparent, but insensitive. If lesions similar to the corneal lesions of granular corneal dystrophy were present in articular cartilage they could not be seen during life, but they would cause a defect similar to arthritis. Similar lesions in the crystalline lens would be called cataract. They can be examined during life with equal facility to the corneal lesions, and cause the same defect to the patient. The lens opacities, however, are absolutely painless, while the corneal opacities are the direct cause of painful attacks, or the opacities are the result of the condition which accompanies the pain. As the cornea is transparent the opacities can be examined during life under high magnification. The only advantage gained by histological examination is that the tissues can be stained by the various stains and the nature of the opaque substance ascertained. The opacities have been so examined by several observers and all agree that the substance is of a hyaline nature. That the opacities are confined to the axial region of the cornea is, in my opinion, a fundamental

factor in the pathology. It has been stated by many careful investigators that the opacities are independent of the corneal nerves, so the only other factors that can be considered as causing the lesion are an error of nutrition or an error of metabolism.

It is now known that gaseous exchange can take place through the healthy cornea, oxygen passing inwards and carbon dioxide passing outwards. To effect such an exchange it is necessary that the cornea be adequately lubricated with tears, and that the surface epithelium and endothelium be in a healthy state. It has been established that the surface epithelium is under nervous control, an alteration in its permeability taking place if the 5th nerve be cut.

Many investigators have stated that the sensitivity of the cornea in hereditary corneal dystrophy has been found to be lowered. Doyne and Stephenson (1905). Personally I did not find this so, and am of the opinion that this is not a contributing factor to the lesion. As the gaseous exchange would be equal over the whole area of the cornea, any error in this mechanism would not produce a lesion in the axial region but would be evenly distributed over the whole cornea.

The cornea receives its nutrition from the vascular loops at the limbus, lymph permeating centrally between the layers of the corneal lamellae. Any error in this circulation of lymph would affect the area furthest from the supply, namely, the central region of the cornea. Weight is given to this argument by the observation of several observers (Bucklers) who state that considerable improvement followed an intercurrent inflammation with vascularization of the cornea.

Treatment

It is unfortunately true that hereditary eye diseases are not amenable to any form of curative treatment. Surgical interference in such diseases as congenital cataract certainly improves vision, but it does not cure the cataract.

When it is remembered that a hereditary defect must be present in the germ cell, the defect developing as part of the organism, and the pathology of the defect being in most instances unknown, any form of treatment must of necessity be of an empirical nature. It will be interesting and also instructive if we review the various therapeutic measures that have been tried by various ophthalmologists. Many of the remedies are obviously empirical, but several indicate that an endeavour was made to treat the cause of the disease, while others again were devoted to the treatment of the local corneal opacities. No differentiation is made in the treatment of the three different types of dystrophy.

General Treatment

The term "nodule" given to the disease no doubt influenced several observers towards thinking that tuberculosis was the cause, and also the fact that most eye diseases of unknown aetiology were by continental ophthalmologists believed to be mainly due to tuberculosis.

Injections of tuberculin were given by Car (1927) Maxwell (1919). As the opacities are somewhat similar to the quiescent appearance of interstitial keratitis, anti-tuberculous treatment was given.

Iodine and Mercury—Fehr (1904).

Pot. Iod.—Fehr (1904), Satanowsky (1932), Schieck, Zentmayer and Rush (1926).

Neo-Salvarsan—Car (1927):

Local Treatment

Chemotherapy.—Previous to 1890 Iodine was the favourite drug employed in the clearing of corneal opacities. This drug would then naturally be tried by the early investigators of this disease. Haab (1899) says that iodoform ointment was tried by Horner and that Wehrli dusted iodoform powder on to the scraped corneal region. Wustefeld inserted a tablet of iodoform into a previously prepared corneal pouch.

To cause an increased flow of lymph, dionin was used by Bartels (1930), Car (1927), Chou (1932), Jacqueau (1909), Puscariu (1913), Rollet (1933) and others.

Paderstein (1913) tried the effect of jequirital.

Sub-conjunctival injections of saline and sublimate were recommended by Bartels (1930), Car (1927), Puscariu (1913) and Satanowsky (1932).

Calomel powder was dusted on by Fehr (1904), Puscariu (1913). *Guttae.*

Zinc Sulph.—Groenouw (1898), Rollet (1933). Boric Acid—Gunn (1902). Silver nitrate—Pascheff (1926). Pot. Iod.—Tritscheller (1921), Salzmann (1926). Resorcin—Schmoll (1932).

Ointments.

Hg. Oxidi Flav.—Fehr (1904), Gunn (1902), Nemeth (1935). White precipitate—Fehr (1904). Hg. Oxidi Rub.—Satanowsky (1932). Ichthyol—Nemeth (1935). Boric—Gutzeit (1929). Ethyl morphine hydrochloride and mercury each 3 per cent.—Lowenstein (1918-29).

Physical Therapy.—Most ophthalmologists gave in addition to the drops or ointment heat in some form or another, such as poultices, hot bathing, electric heater or diathermy. Fehr, however, prescribed cold compresses.

U.V. light was given by Evans (1930) and X-rays was given by Nemeth (1935) and Rollet (1933). Mercury—Car (1927), Fehr (1904), Rollet (1933). As the opacities were in the nature of deposits and might be due to errors of metabolism, anti-gout treatment was given. Colchicum—Jacqueau (1909). Intra muscular injections of fibrolysin—Satanowsky (1932).

Glandular Therapy.

Thyroid extract—Masuda (1935), Solarikova (1931), Sallzmann (1926), Zentmayer and Rush (1926). Ovarian extracts—Lowenstein (1918-29), Sommer (1925). X-rays to ovaries—Car (1927). Vitamin A—Bucklers (1937). The patients received benefit generally from the vitamin A (*e.g.*, they felt better in health). Protein shock—Gutzeit (1929). Iron preparations—Evans (1930), Gutzeit (1929). Liver preparations—Bartels (1930).

Surgical Treatment

Superficial scraping was undertaken partly for diagnostic purposes and partly as therapeutic measures by Bartels (1930), Bietti (1934), Dimmer (1899), Deutschmann (1908), Fehr (1904), Groenouw (1898), Pascheff (1926), Uhthoff (1915), Schmoll (1932), Sommer (1923), Stein (1932) and Wehrli (1907).

Corneal translation—E. Fuchs (1926), A. Fuchs (1925), Friede (1936), Franceschetti and Kiewe (1936), and Hilgartner and Henry (1937).

Mucous membrane graft from the lip was performed by Bartels (1930).

Paracentesis—Wehrli (1907), Bartels (1930).

Cauterization—Deutschmann (1908).

Optical Iridectomy—Bietti (1934), Clausen (1911), Lowenstein (1929), Koerber (1902) and others.

Sympathectomy—Gală (1929).

Contact glasses—Stein (1932).

Bucklers (1938) examined several patients who had an optical iridectomy, done some time previously, and he came to the conclusion that not only was vision not improved, but that the patients were made more uncomfortable by dazzling and coloured vision.

Personally I feel that in the treatment of the granular type of corneal dystrophy a large number of the treatments tried were unjustified. The progress of the disease was not brought to a standstill with certainty with any, and far less was the disease made to regress. In certain cases the attempted cure was more harmful than the disease.

I recommend that any refractive error be corrected with glasses, and the refraction periodically checked as it appears to alter. As the attacks of pain appear to come on by a lowering of the general condition, the general health should be kept at as high a level as possible with careful attention to diet supplemented by the addition of vitamin A and B periodically.

Of all the affected people that I have examined, one only would benefit by surgical interference. This is V(34), for whom an optical iridectomy might with benefit be performed upon one eye. By dilating her pupils with homatropine distance vision was not improved, but near vision was improved from J.16 to J.2. I intended to keep one pupil dilated with atropine, but atropine irritation supervened after one application of the drug. The daughter of this woman also took atropine irritation after being two weeks on a 1 per cent. solution. I wondered if this was a coincidence, or was actually due in some way to the corneal condition. V(20) was not benefited by having a dilated pupil, so an optical iridectomy in his case is not contemplated.

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