

**THE SIGNIFICANCE OF HEREDITY IN
OPHTHALMOLOGY. PRELIMINARY SURVEY OF
HEREDITARY EYE DISEASES IN TASMANIA***

BY

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(Continued)

It is not within the scope of this paper to, attempt to expound the problems of Mendelian characters in regard to ophthalmology, but rather to decide whether or not hereditary eye disease in Tasmania conforms to these established laws. On the whole I must admit that they do, and this is only to be expected in view of the fact that most of the original members of these pedigrees were born within the British Isles.

Since 1900, Mendel's laws have become widely appreciated, so that patients affected with hereditary diseases are now constantly seeking advice from their medical practitioners. Thus it behoves all medical men, and especially ophthalmologists to be conversant with the various types of transmission, and I therefore propose to give a brief survey of the behaviour of Mendelian and sex-linked characteristics, for the guidance of those who may not be acquainted with these facts.

Dominant Transmission

- (1) If both parents are affected—
 - (a) $DD \times D(R)$. All children affected.
 - (b) $DD \times DD$. All children affected.
 - (c) $D(R) \times D(R)$. $3/4$ children affected, $1/4$ normal.
- (2) If one parent is affected—
 - (a) $DD \times RR$. All children affected.
 - (b) $D(R) \times RR$. $1/2$ children affected, $1/2$ normal.

Recessive Transmission

- (1) If both parents are affected—

$RR \times RR$. All children are affected.
- (2) If one parent is affected—
 - (a) $DD \times RR$. All children normal.
 - (b) $D(R) \times RR$. $1/2$ children affected, $1/2$ normal.

* Continued from p. 43.

Sex Linkage Transmission

The disease may be dominant, or recessive (generally recessive).

A. If the disease is recessive—

1. The males are affected more frequently than the females.

2. Females are of three kinds.

(a) Completely normal and not a carrier. XX .

(b) Completely normal but a carrier. XX .

(c) Affected. XX .

3. Males of two kinds.

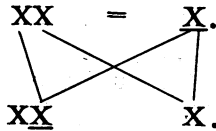
(a) Completely normal. X .

(b) Affected. X .

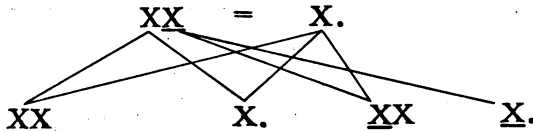
Note.—Males cannot be carriers in the sense of (2b).

Examples of recessive sex linkage.

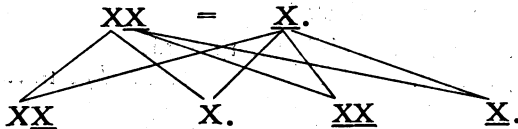
1. A female of class (2a) married to an affected male (3b) will produce normal children. All the daughters will be carriers (2b), and will transmit the disease to half their sons.



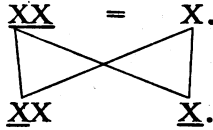
2. A carrier female (2b), married to a normal male will transmit the disease to half the sons. Half the daughters will be carriers (2b).



3. A carrier female (2b) married to an affected male (3b) will have half the daughters affected (2c), and the other half carriers (2b). Half the sons will be affected (3b), and half will be normal.



4. An affected female (2c) married to a normal male (3a) will hand on the disease to all the sons (3b), and all the daughters will be carriers (2b), and will hand on the disease to half the sons.



B. If the disease is dominant.

Females.

- (a) Diseased. XX.
- (b) Diseased. XX
- (c) Normal. XX.

Males.

- (a) Diseased. X.
- (b) Normal. X.

Later when I comment on each individual pedigree which I am reporting, I shall endeavour to define the mode of transmission, but as this is only a preliminary report, and in consequence many of the pedigrees short, this will not always be possible. With regard to the brevity of some pedigrees, I am encouraged in presenting them by the fact that the Bureau of Human Heredity (115, Gower Street, London), appears to be as interested in shorter, as in longer ones, and appeals for unselected family trees (*Brit. Med. Jl.*, 1936). My fifty-three pedigrees are quite unselected.

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Astigmatism

Unfortunately in order to keep the diseases in alphabetical line I must commence with the least satisfactory of the hereditary diseases. Nor am I helped by the literature, which is extraordinarily scanty.

Astigmatism, according to Duke Elder (1934) and Ruggles Gates (1929) is undoubtedly hereditary in many pedigrees, and

is usually dominant in transmission. They state that not only is the degree of astigmatism maintained, but also the axis in most pedigrees. I have not been able to confirm the former assertion, but in pedigree 21, 8 of the 13 members of a myopic family have their astigmatism against the rule; and in pedigrees 20 and 25 every examined member has his astigmatism with the rule. But this cannot be followed in every case. The mode of transmission also is certainly recessive in some strains (pedigrees 3 and 5), but in others more data are needed to form an opinion.

With astigmatism manifesting itself in at least 3/4 of ophthalmic cases, the surgeon ultimately regards it as a necessary evil and gives it scant consideration. Nevertheless I feel if ophthalmologists working in confined areas (as I do) would pay some attention to the hereditary aspect and report their findings, that they would be agreeably surprised to find that astigmatism is not as boring as they had thought. I certainly intend to follow this aspect of heredity more closely.

As I have found some difficulty in separating the astigmatic pedigrees from those of myopia, I would suggest that these two diseases be read in conjunction rather than treating them as two separate entities as I have done. On the other hand as each characteristic has a separate gene it might have been expedient to have had separate pedigrees for each characteristic.

Comments on Pedigrees

My interest in hereditary astigmatism arose through pedigrees 1 and 2.

In *Pedigree No. 1*, Case No. I/1 was referred to me for compensation purposes. He had high mixed astigmatism with the rule, and with correction saw only 6/24 in the better eye. He attributed his defective sight to the accident, and I to inherited astigmatism. The dispute was still unsettled when his son II/2 reported with defective sight due to high mixed astigmatism with the rule, of a very similar nature to the father's. The dispute accordingly ceased.

In *Pedigree No. 2*, cases III/2 and 3, who were sisters, had high mixed astigmatism of almost identical type and strength in the four eyes. A year later the mother (II/13) called complaining of bad headaches which physicians had failed to cure. She had high astigmatism in both eyes, but especially the right. Her right refraction was simple oblique myopia, and her left simple oblique hypermetropic astigmatism. I asked her why, in view of her daughter's defect, she had not attributed her headaches to defective sight. She admitted that she had no idea defective sight was hereditary. I then made further searches and during the past

two years have collected three more pedigrees as well as those recorded under myopia.

Pedigree No. 3.—As only II/2 has been examined, it is difficult to comment with authenticity, but II/2, who is a man of wide knowledge and experience, assures me that his mother and elder sister are similarly affected. In that case the disease is most likely recessive in transmission, and I advised him to watch the eyes of his two sons.

Pedigree No. 4.—This pedigree, in part, will be discussed under nystagmus. II/3, has compound myopic astigmatism with the rule, and her left eye is amblyopic with 45° of divergence.

II/4. Has complicated cataracts from iridocyclitis.

III/8. Has mixed astigmatism against the rule.

III/60. Has left divergent concomitant strabismus, with partial amblyopia. She is emmetropic.

III/63. Has left divergent concomitant strabismus with complete amblyopia, and high oblique compound myopic astigmatism.

IV/3. Has high oblique astigmatism.

III/60 and 63 are interesting because the paternal parent belonged to the astigmatic portion of the pedigree, and the mother to the nystagmus portion. The result was squint with amblyopia—such amblyopia occurring in two distant relatives, II/3 and IV/22 (one on each branch of the family), without nystagmus, and in four other cousins (on the maternal side) with nystagmus. As members of this pedigree are constantly reporting, I may later have more definite comments to offer.

Pedigree No. 5.—This is of special interest as a member of the first family, in which at least four members have compound myopic astigmatism, married a member of the second family, of which at least three members have compound hypermetropic astigmatism. The result has been five sibs—three of which are affected with refractive errors, one with myopia in the right eye, and compound hypermetropic astigmatism in the left.

Another with pure hypermetropia, and the third with gross mixed astigmatism.

I/2. High compound myopic astigmatism with the rule and fundus changes.

I/3. Low compound myopic astigmatism with the rule.

II/3. Myopic astigmatism—no details.

II/6. Mixed astigmatism against the rule.

II/9. High compound hypermetropic astigmatism against the rule.

II/11. Low compound hypermetropic astigmatism—oblique.

II/15. High compound hypermetropic astigmatism against the rule.

III/2. Right eye, low myopia. Left eye, low hypermetropic astigmatism against the rule.

III/3. Moderate hypermetropia.

III/5. High mixed astigmatism.

The disease definitely appears to be recessive, but more than one gene is certainly involved.

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Congenital Cataract

According to Macklin (1927) it is estimated in Canada that 13 per cent. of the pupils in institutions for the blind are suffering from hereditary cataract. The proportion in Tasmania is much higher than this; 3 of the 6 blind pupils in the Tasmanian Institution for the Blind are suffering from cataract. On the other hand, Best (1934) considers that 13·7 per cent. of blindness in U.S.A. is due to cataract, but in Tasmania I find 20·45 per cent. of the blind of whom reliable clinical information is obtainable suffering from this disease. This figure appears to me to be astounding especially in view of the fact that of all hereditary eye diseases causing blindness, cataract should be the most responsive to treatment. I can account for this high percentage in Tasmania in the following manner:—

(1) Late operative interference. This is exemplified in pedigree 7, where one father (IV/14) refused to allow his children to be operated on until they had developed nystagmus and strabismus with amblyopia. In other cases operation has not been considered, due to ignorance or isolation of the parents.

(2) Incomplete operative interference. This is exemplified by pedigree 6 where III/5 had a right needling done many years ago, but did not report for glasses, which eventually gave him 6/12 partly and J. 4. Fortunately he has submitted his daughter aged 1½ years for operation, and the visual result should be good.

(3) Conservative Medical Advice. The unsatisfactory results that have sometimes followed discission of the crystalline lens has often deterred ophthalmologists from advising operative interference.

(4) The Commonwealth Government does not insist on applicants for blind pensions undergoing expert examination and treatment before being accepted. It is left to the applicant's choice between blindness with pension, and sight without pension, and unfortunately, many choose the former course. This undoubtedly should be remedied at once.

Sex Incidence

In my four pedigrees (Nos. 6, 7, 8 and 9), there are 13 affected males and 18 affected females, giving a percentage of 41.9 and 58.1 respectively. In pedigree 7, there are five members whose sex has not definitely been determined. Taking the pedigrees reported by Bishop Harman (1909), Macklin (1927), Galloway (1930), Khan (1926), Knapp (1926) and Dunforth (1914), I have found the sex incidence as 55 males and 63 females, giving a percentage of 46.6 and 53.4 respectively, which conforms with the predominance of females shown above.

Age of Onset

Loy (1936) quotes Nettleship's six laws pertaining to the occurrence of cataracts in females, and the 5th law states that—"the age of onset is approximately the same in persons of the same generation." In my three pedigrees, so far as I am able to ascertain, every case was apparent from infancy, with the exception of III/23 of pedigree 8, where the opacities are only partially developed at puberty.

Type of Opacity

According to Clapp (1934) congenital cataracts may take 10 partial forms, and of course one complete form, but nowhere in his book can be found a discussion on the hereditary nature of these opacities.

My longest pedigree, No. 7, exhibited zonular opacities almost uniformly with occasional dislocation of the lens, and in one case at least the lens had been practically absorbed. In pedigree No. 6 the two cases examined were of coralliform type, while pedigree 8 exhibited no uniformity in the two cases.

I have used the word "zonular" in reference to pedigree 7 intentionally, as I feel the word "lamellar" should be left for those cataracts following infantile tetany.

Type of Inheritance

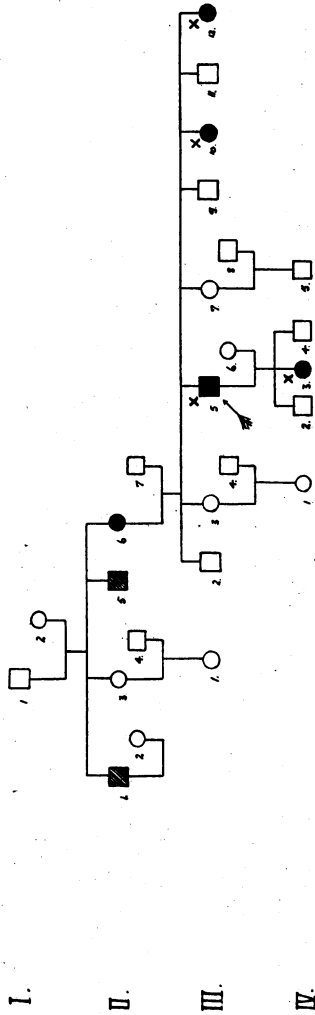
In pedigree 7 the defect is possibly dominant in transmission, while in the other three pedigrees the number of affected individuals is too small to make even a conjecture.

Associated Defects

Feeble-mindedness.—There is no evidence of this in any of the four pedigrees—a small number of the affected individuals are somewhat retarded mentally and temperamental occasionally, but in no case could I classify them as feeble-minded.

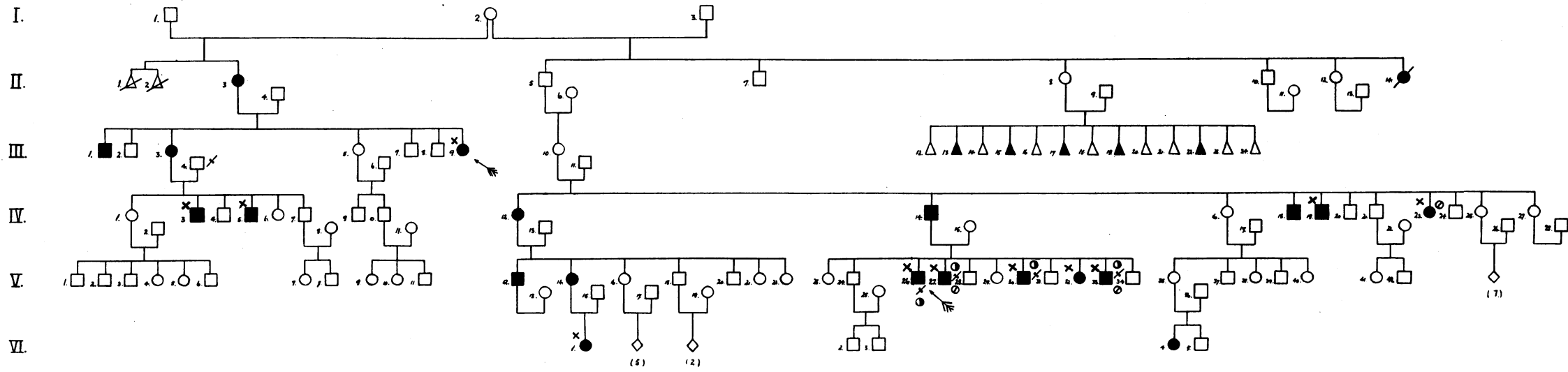
No. 6. CONGENITAL CATARACT.

- - Affected Males.
- - Affected Females.
- ▨ - No History
- X - Examined.
- ✂ - Propositus.



No. 7. CATARACT.

- - AFFECTED MALES.
- - AFFECTED FEMALES.
- × - STRABISMUS - CONVERGENT.
- ⊙ - DILATED LENS.
- ⊙ - NYSTAGMUS.
- △ - SEX UNKNOWN.
- △ - DIED IN INFANCY.
- × - EXAMINED.
- ↘ - PROPOSITUS.



Dystrophia Myotonica.—The one pedigree (No. 10) of this disease is described separately.

Endocrine Disturbances.—There is no evidence of these in Tasmanian pedigrees.

Treatment

Operative interference in the form of repeated needling has been singularly successful in these Tasmanians, with one exception. When I say successful I refer to the anatomical, but not the functional results. All the poor functional results were due to late operation, and this stresses the fact that early operation is essential if good visual acuity is to be expected. To obtain this the public must be educated and its confidence gained.

Comments on Pedigrees

Pedigree No. 6.—This is an example of coralliform congenital cataract.

No. II/6, has cataracts but has not yet been examined.

No. III/5, aged 26 years, has had his right cataract needled after puberty, with a very fair visual result; while his left eye exhibits a typical coralliform cataract.

No. III/10 and 12—sisters of the above—have been examined by an ophthalmic surgeon, and certified for pension purposes as blind from congenital cataract.

No. IV/3, had typical coralliform cataract in a very advanced state right and left, with nystagmus at 13 months. Both cataracts have been needled, but it is too early yet to forecast the visual result.

Pedigree No. 7.—This pedigree is one of zonular cataract. III/9 is the eldest member examined, and also one of the propositi. She was examined at the age of 48 years. 25 years previously her right lens had been removed at operation, with good operative but poor visual results, the latter due to amblyopia. In her left eye besides her cataract she has gross keratoconus.

No. IV/5, has been examined for pension purposes, and certified blind from congenital cataracts.

No. IV/23, had opaque lenses right and left when examined by me at the age of 42 years. Both irides were tremulous, and the left lens obviously dislocated. There was marked nystagmus. I extracted the right lens by the intracapsular method of Sinclair, and although the anatomical result is good, acquired amblyopia prevents a good visual one. Nevertheless the vision is materially improved by the operation.

No. V/26, aged 23 years, has had both zonular cataracts needled by me, and his squint straightened. But nystagmus and amblyopia—both congenital, and ex anopsia prohibit a good visual result, yet he is considerably improved by the operative interference, and can use a typewriter with ease.

No. V/27, aged 21 years. Has also congenital cataracts, nystagmus, and squint. Both lenses are markedly dislocated, so that through the aphakic gaps with correction he is able to read 5/60 with both eyes together. In view of the post-operative visual result of his brothers, I feel that no operative interference would improve the present visual acuity.

No. V/30, aged 16 years. Had dense zonular cataracts right and left, with right convergent concomitant strabismus, and gross nystagmus. He has had both lenses needled, and his squint straightened by me three years ago; and is now pursuing a High School education. His visual result is poor due to amblyopia.

No. V/32 aged 12 years. Had only thick capsules right and left, although she had had no previous operations. A capsulotomy was done on each eye by me four years ago, and with correction she is able to see 6/36 right and left, and J. 10 right, and J. 16 left with bifocals.

No. V/33 aged 7 years, also had dislocated zonular cataracts, nystagmus and concomitant strabismus. Three years ago both lenses were needled, and his squint straightened by me, but the visual improvement is only slight owing to amblyopia.

I think V/32 certainly indicates that had these 5 sibs been operated on in infancy, the visual results would have been most satisfactory.

Pedigree No. 8.—No. III/36, aged 11 years, has had cataracts since birth, and was operated on when 5 months of age in both eyes, without result; and is now an inmate of a blind institution, with gross nystagmus, shrunken lenses, and secondary glaucoma.

No. III/23, aged 14 years, is placed in this order as he was examined a considerable time after III/36. He complained of defective sight and had marked anterior and early posterior cortical opacities in both eyes, but the vision with correction in each eye is still 6/12. I had been at a loss to account for the condition of III/36 until III/23 appeared.

Pedigree No. 9.—No. II/1, is a blind pensioner, certified by an ophthalmic surgeon as having congenital cataracts. The type is not stated, but his vision is less than 6/60 in each eye.

No. III/1, aged 19 years, has been examined by me, and has gross post-cortical lens opacities, so that right and left vision is reduced to hand movements. To date she has refused all operative interference.

Cataract with Dystrophia Myotonica

In 1911 Greenfield reported the first cases of cataract associated with dystrophia myotonica, and in 1924 Adie pointed out that cataract occurred alone in many individuals in a dystrophia myotonica pedigree. Vogt in 1921, and Goulden in 1928 gave a lengthy description of the slit-lamp appearance, so typical of the disease, and many cases of successful extraction have been reported (Souter, 1933). Rouquier and Chatain (1935) consider the neurological lesion to be in the diencephalon and the mesencephalon, but no satisfactory explanation has been given for the accompanying cataracts.

Comment on Pedigree No. 10.—I have but one pedigree (No. 10), to report, and that one exhibits cataract only in father I/1, and two children, II/8 and II/15, and dystrophia myotonica in the same two sibs. I can make little comment on I/1 who has not been examined by me, but has had a recent cataract extraction. II/8 had her left cataract extracted four years ago at the age of 37 years, and her right lens exhibits an advanced, but typical stage of myotonic cataract. She has obvious signs of muscle wasting and the typical myotonic grip. Her brother, II/15, conformed physically to an historic case reported by Caughey (1933), viz. :—“ He is short, bald, speaking through his nose, looking forty at the age of twenty-five.” As a matter of fact he was 29 when he came under observation, and had typical posterior star-shaped lens opacities in the cortex of both lenses. He had muscular weakness, and myotonic grip.

I needled the left cataract, and at present he has 6/4 and J. 1 vision in that eye, with aphakic correction.

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Chronic Glaucoma in Adults

Increased intra-ocular tension both in its acute and chronic form is relatively rare in Tasmania, and especially in its acute form. When chronic glaucoma does occur in the residents, it is surprising to find that a very marked proportion are not native born Australians. Out of 4,880 private case records during the past 5½ years (*i.e.*, January, 1931, to July, 1936), I can find the following cases of increased tension :—

Buphthalmos	1
Acute Glaucoma	5
Chronic Secondary Glaucoma	10
Chronic Primary Glaucoma	44

If an analysis is made of my 4,880 cases, then the percentage of chronic primary glaucoma amongst these cases, is 0.9 per cent., which compares favourably with the statistics quoted by Julia Bell (1932) which ranged from 0.4 per cent. in Germany to 2.73 per cent. in Italy; and estimates from almost a million European cases make an average percentage of 0.97 per cent.

Of these 44 cases of chronic primary glaucoma, 30 were native born Australians, and 14 were born in the British Isles, which in percentages is 68.2 per cent. and 31.8 per cent. respectively.

Now, according to the 1933 Census, 95 per cent. of the population of Tasmania were born within the Commonwealth, and 5 per cent. outside the Commonwealth, so that one would have expected to find the percentage of glaucoma identical. Presuming 5 per cent. of the population are not native born, then 244 of the above 4,880 cases should have been born outside Australia, and 4,636 of the cases born within Australia. Of these approximately 244 cases, no less than 14 have glaucoma—a surprisingly high percentage of 5.73, while of the 4,636 Australian born cases, only 30 have glaucoma—a surprisingly low percentage of 0.65. I am unable to account for these figures, and intend reviewing them again at a later date, in order to confirm the startling disproportions. If we conclude that “worry” is a predisposing cause of glaucoma, then a faint ray of truth may be revealed.

The recent emigrants have undoubtedly been subjected to far more anxiety than their Tasmanian brothers and sisters, who by reason of the prolonged isolation of Tasmania, and the isolated position of their residences were prohibited reasonable contact with the outside world until 1925. Since the advent of radio in 1925, and the world-wide telephone communication in 1936, this isolation to a great extent has been banished, and it may be interesting to see if the glaucoma figures will rise in the next half of this century (See Table VI).

Hereditary Influences

Of my 44 cases of chronic primary glaucoma, only one hereditary pedigree (No. 13) of 2 cases can be found. The two cases of pedigree 14 are amongst these 44 cases, but this is purely familial in incidence, but I think worth quoting. Pedigree 11 was given me by a patient who had not glaucoma herself, while No. 12 was kindly supplied by Dr. Counsell. So that amongst my own patients the hereditary factor is extremely low. Bell (1932) complains of the paucity of reliable records of hereditary glaucoma, and finds only 3 pedigrees reported by Nettleship, and one by Usher, and was able to collect only 68 reported pedigrees in the literature.

Sex Incidence of Hereditary Glaucoma

Taking the 3 hereditary pedigrees (Nos. 11, 12 and 13), I find in a total of 8 cases there are 5 males (62.5 per cent.) and 3 females (37.5 per cent.) in all. Whereas Bell's figures (1932) are 99 males to 97 females. Zorab (1932) on the other hand in surveying 6 pedigrees found 25 males and 10 females affected; while Frank Kamenetzki (1925) recorded a pedigree in which every affected member was a male.

So that there appears to be no doubt that males are more prone to be affected than females, when the disease is hereditary. Conversely, of the 42 non-hereditary cases seen by me since 1930, 25 were females (59.5 per cent.), and 17 males (40.5 per cent.), which gives a greater predominance of females, and conforms to Rimpler's statistics—quoted by Frank Kamenetzki—of 2,021 cases of glaucoma in which 1,150 (56.9 per cent.) were females, and 871 (43.1 per cent.) were males.

Age of Onset

I can give no useful information as to this. Pedigree 11, which was collected long before I anticipated writing this paper, might have helped considerably, but these details are wanting unfortunately, and quite possibly the propositus would not have been able to supply them. In my own pedigree (No. 13) one patient was 88, and his niece 37; while in the familial pedigree (No. 45) the two cases examined were 86 and 84 respectively.

Mode of Transmission

Although Macklin (1927) considers the transmission of hereditary glaucoma dominant, yet my few pedigrees prove that this is not always so by any means, and even so great an authority as Nettleship did not feel disposed to commit himself on the mode of transmission.

Frank Kamenetzki (1925) reports one pedigree in which all the afflicted members are males, and the mode of transmission was a recessive sex-linked one.

Concurrent Anomalies

In the very interesting pedigree of Frank Kamenetzki (1925) the individuals affected with glaucoma had an accompanying atrophy of the iris stroma. Bell (1932) states, "So far as I can judge, cases of hereditary glaucoma appear in otherwise healthy stock, and very few examples of associated hereditary diseases or anomalies are found amongst the affected individuals." My few cases exhibit no peculiar anomalies.

Treatment

There appears to be little variation from the orthodox treatment in the hereditary cases. Frank Kamenetzki (1925) found iridectomies difficult in his cases owing to the iris atrophy, while James (1927) reports satisfactory results from trephining. Personally I have found no indication in my few cases to depart from routine procedures.

Comment on Pedigrees

Pedigree No. 11.—As mentioned before this pedigree was obtained roughly, long before I intended to write this monograph, and therefore is not very helpful. The propositus III/5, called to be examined on account of the predominance of glaucoma in her family. She herself was unaffected, and as the other members of her family live, or lived in the north of the island, I have been unable to examine them to date.

Pedigree No. 12.—Was kindly supplied to me by Dr. Counsell. I/1 is dead, while II/1, at the age of 70 has had to have one eye trephined.

Pedigree No. 13.—III/3, aged 88 years, is under treatment by me for cavernous atrophy of the optic discs, and raised tension—central vision has been lost in one eye, but the fields of vision are normal to 1° white. His niece IV/17 has advanced glaucoma right and left at the age of 37 years. Four other members of the family examined are free from this defect, but all four of these exhibited gross astigmatism in both eyes.

Pedigree No. 14. are 3 sibs. II/4 is dead, but his 2 sisters (II/1 and 3) at the age of 89 and 86 respectively, have advanced chronic glaucoma in both eyes, and they did not report for treatment until aged 86 and 84 respectively. One significant fact about these old ladies is that when each reported (not under miotics)

the tension in both eyes was 20 mm. Hg. (Schiötz), *i.e.*, the four eyes had similar normal tension, and yet the four fields were greatly, and typically constricted; and the optic discs grossly cupped.

TABLE VI
*Incidence of Hereditary Glaucoma in Tasmania
from January 1, 1930, to July 1, 1936.*

No. of cases examined - - - - -	4880
Chronic Primary Glaucoma - - - - -	44= 0.9 per cent.
Born in Australia - - - - -	30=68.2 per cent.
Born outside Australia - - - - -	14=31.8 per cent.
Males - - - - -	18=40.9 per cent.
Females - - - - -	26=59.1 per cent.
Hereditary cases amongst above - - - - -	2
Males - - - - -	1
Females - - - - -	1
Non-hereditary cases amongst above - - - - -	42
Males - - - - -	17=40.5 per cent.
Females - - - - -	25=59.5 per cent.
No. of hereditary cases reported in Tasmania - - - - -	8
Males - - - - -	5=62.5 per cent.
Females - - - - -	3=37.5 per cent.
Percentage of native born Australians in popu- lation - - - - -	=95 per cent.
Percentage of non native born Australians in population - - - - -	= 5 per cent.
Approximate number of my 4880 cases born in Australia - - - - -	=4636
Approximate number of my 4880 cases born outside Australia - - - - -	= 244
Percentage of glaucoma amongst 4636 assumed Australian born cases - - - - -	=0.65 per cent.
Percentage of glaucoma amongst 244 assumed non-Australian born cases - - - - -	=5.75 per cent.

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Keratoconus

Although keratoconus is an hereditary disease, yet, as in retinitis pigmentosa, many sporadic cases, in which there appears to be no hereditary influence whatsoever are found in routine work. In my 4,880 private cases (mentioned under glaucoma and pterygium) I find 19 cases in all of keratoconus—making a case incidence of 0·39 per cent. Of these 19 cases, only one can definitely be said to be hereditary (pedigree 16), while three others are strongly familial in incidence (pedigree 15). Of these 19 cases, 18 came from Southern Tasmania where the proportion of city and country dwellers is equal. Yet of these 18 patients no less than 14 came from the country districts, where good food and sunlight exist in abundance.

Associated Defects

Fleischer's Ring.—This was present in only one of the 38 eyes examined and was not accompanied by opacity of the cornea.

Apical Corneal Opacities.—These were present in 6 of the 38 eyes, and in 2 eyes they received surgical treatment.

Mode of Transmission

According to Franceschetti (1930), keratoconus is transmitted both as a dominant and recessive. In my one pedigree (No. 24) we must undoubtedly presume that the mode of transmission was recessive.

Sex Incidence

Of these 19 patients, 12 were females, and 7 males, while the one hereditary pedigree (No. 16) shows only females affected. As I am unable to obtain a copy of Jaonsch and Stahli's articles, I cannot compare and contrast these figures with statistics from other countries.

Concurrent Diseases

Mongolian Idiocy.—Two of these 19 patients were Mongolian idiots, with advanced conical cornea in 3 eyes, which needed and received operative interference.

Thyroid Dysfunction.—Of the 19 patients, 10 were generally examined. Three showed no trace of endocrine dysfunction, three females were subthyroid, and 4 hyperthyroid. Knapp (1924) found low basal metabolic rates in 2 out of 6 patients.

Treatment

Contact Lenses.—These were prescribed in one case, and 2 other cases had their visual acuity tested with them. In all three cases the result was highly satisfactory. But a problem, which needs further investigation and discussion, is the possibility of retarding the progress of the corneal bulging by ordering contact lenses early in the disease.

Miotics and Hormone Therapy.—Although I have given both these prolonged trial, I frankly admit that I have seen very little benefit derived from their use. In one case I thought "hormone" retarded the progress.

Surgical Interference.—This was necessary on 3 eyes, and the electric cautery was applied to the apex of the cones until perforation resulted, after the manner of Knapp (1929). In one case the anterior chamber took three weeks to reform but the ultimate results in all were satisfactory.

Comments on Pedigrees

Pedigree No. 15.—This is purely familial in origin, but it is worthy of record, and reports four members of a sibship of 11, with marked keratoconus. All four of the affected members and two of the unaffected members were examined by me.

II/2, aged 37 years, cornea normal.

II/6, aged 34 years. Gross keratoconus right and left without corneal scarring. Right and left vision improved to 6/12 with high oblique concave cylinders.

II/10. Gross keratoconus. No details.

II/11. Normal cornea.

II/12, aged 24 years. Early keratoconus right and left. Vision right and left improved to 6/12 with myopic correction.

II/15, aged 19 years. Moderate keratoconus right and left without scarring. Keratoconus more marked in right eye.

III/9. Has defective eye sight. Doubtful causation.

Pedigree No. 16.—III/2, aged 20 years. Was examined by me, and has gross keratoconus right and left, but no corneal opacities.

III/8. A cousin was examined by Dr. Homewood of Launceston, and found to have keratoconus also, but I have no details.

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Microphthalmos

Among the hereditary eye diseases microphthalmos stands prominent as a potent cause of blindness. Bell (1932) analysed the vision of 91 cases of microphthalmos and found total blindness in both eyes of 33 cases, and vision of 6/60 or less in both eyes of 25 cases; blindness in one eye of 8 cases; "very bad" bilateral vision in 10 cases, leaving only 15 cases with good vision in both eyes, and even 4 of these 15 have latterly developed cataracts. She also analysed the other ocular defects accompanying microphthalmos, and pointed out the predominance of iris and lens defects reported. Usher (1921) has especially drawn attention to accompanying corectopia and myopia when reporting a pedigree of 11 affected individuals in five generations. Bell was unable to elucidate any peculiarities about sex incidence, and inheritance of microphthalmos; but on the other hand Ash (1922) has reported a pedigree of 34 individuals in four generations, eleven of whom were affected with microphthalmos, and every affected one was a male. Like Leber's optic atrophy the disease in this pedigree was strictly sex linked, and was transmitted by unaffected females only. The significance of this pedigree has been elaborated by Macklin (1927). In her 41 pedigrees Bell records consanguinity only 5 times. Wolff (1930) on the other hand considers consanguinity precipitated his pedigree, in which 5 of 10 children were afflicted with microphthalmos. (I have made reference to a similar occurrence under Leber's Disease.)

Treatment

I have no comment to offer except that great cosmetic improvement resulted from ordering 10.0 sphere before the left eye of V/5, for which the parent was most grateful.

Comments on Pedigrees

Pedigree No. 17.—This was given me by the mother (IV/9) of my patient (V/4), who was intelligent and most co-operative. She went to great pains to obtain it, and I think I can vouch for her integrity. I have been unable to examine IV/32 and so will briefly describe V/4's condition observed at annual examinations over 4½ years.

Right Eye.—Normal size. Dense crescentic corneal opacity below, extending inwards from the limbus, pupil circular and not ectopic. Gross pseudoneuritis.

Right vision under mydriatic with

+ 2.25 D. sph.	
+ 3.25	= 6/6 in all and J. 1.
	105

Left Eye.—Very small. Dense crescentic corneal opacity below, extending inwards from the limbus. Corectopia downwards. Opaque lens dislocated upward, giving faint fundus reflex below. No fundus details with the pupil dilated.

Left vision.—Hand Movements.

V/4's only sister (V/5) has "port wine stain" of the face, but both her eyes are normal.

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Myopia

According to Best (1934) myopia is the cause of 0.3 per cent. of blindness in the U.S.A., but just how prevalent it is in the community is not stated. However, in my 4,880 private cases there were 641 cases of myopia and myopic astigmatism; which gives a case incidence of 13.13 per cent. In Counsell's and my analysis of the 170 blind in Tasmania we found that myopia would account for 4 out of 92 with reliable information, *i.e.*, 4.35 per cent., which compared with Best's figures above, is very high.

That myopia is undoubtedly hereditary is shown by my 9 pedigrees (Nos. 18 to 26), but some authors—Levinsohn (1934), and Franceschetti (1930)—seriously doubt the hereditary factor, while Roden (1933) tries to explain all cases on a mechanical or pathological basis. At least 4.5 per cent. of my 641 myopes gave an hereditary history.

Mode of Transmission

Holm (1925) considers the mode of transmission may be dominant, or recessive, and this is borne out by my 9 pedigrees, although most other authors consider it recessive only in transmission.

Sex Incidence

Of 48 affected persons mentioned in my 8 pedigrees, 21 were males, and 27 females; giving a sex incidence of 44.7 per cent., and 55.3 per cent. respectively. I am unable to find figures in the literature for comparison, but Wilson (1935) considers 2/3 of myopes are females.

Associated Defects

Night Blindness.—Although reported by Holm (1925), there is no evidence of it in my pedigrees.

Exophoria.—This was only exhibited to any degree in one of the 29 cases examined, and therefore cannot be an important factor in the production of hereditary myopia, as suggested by Pascal (1935).

Treatment

To me the main interest lies in treatment. We are all conversant with the necessity for adequate correction, and limited close work, and we are all aware of the disappointing results. As the theory of calcium deficiency has been advocated by many writers, I had the blood calcium in six progressive myopes examined, with only one subnormal result. It was then that my colleague, Dr. A. W. Shugg, suggested that perhaps progressive myopia was not a matter of calcium deficiency, but one of defective calcium utilisation. Acting on his advice, I have 22 patients on "radiostoleum" (vitamin A and D), and the first 2 which were progressing rapidly over an 18 months' period, ceased progress at once, and have remained stationary since. The other 20 cases have been under treatment for too short a period to comment upon, but I intend to report the results later. Walker (1932) reports favourable results from calcium and parathyroid; but admits failure from calcium therapy alone, while Law (1934) reported satisfactory results from calcium and parathyroid, and D. V. Giri who entered into the subsequent discussion suggested that vitamin D instead of parathyroid might aid the assimilation of calcium. This fact I am endeavouring to affirm, and to date the result has been more than encouraging.

Comments on Pedigrees

Pedigree No. 18.—I/1. Believed to be normal.

I/2. Not examined, but supposed to be myopic.

II/1, aged 14 years. Had more than 17 dioptres of myopia right and left, but was not refracted.

II/2, aged 12 years. High compound myopic astigmatism. Now on radiostoleum.

II/3. Not examined, and believed to be normal.

II/4, aged 7 years. Lamellar cataracts right and left, of the tetany type. Moderate simple myopic astigmatism.

This family is of extreme interest. I/1 was a lighthouse keeper on a most isolated and wind-swept island on the southern coast

of Tasmania. Fresh fruit and vegetables, also fresh milk were unprocurable for a greater part of the year, and so the children's calcium intake was very low. As the children arrived at school age they were tutored by correspondence course until considered sufficiently responsible to send to a city school. II/4 was still dwelling on the island. Apparently her calcium metabolism had been so deranged in early life that cataracts resulted, and I think the presence of these cataracts in conjunction with high myopia in her sister and brother, definitely point to a faulty calcium metabolism as an aetiological factor in myopia.

Pedigree No. 19.—III/4, aged 20 years. Compound myopic astigmatism of over 20 dioptres with definite choroidal stretching, and one haemorrhage.

III/5. Amblyopia ex anopsia as the result of squint in childhood. Eyes now straight by cover test.

III/7, aged 25 years. First cousin of III/4. Has low grade compound myopic astigmatism.

Pedigree No. 20.—I/1. Believed to be myopic, and certainly has defective sight.

II/1, aged 44 years. High compound myopic astigmatism, with the rule, with choroidal stretching.

II/3, aged 40 years. Right retinal detachment, cured by Larsson method. High compound myopic astigmatism with the rule, with albinism and nystagmus.

II/7, aged 34 years. Simple myopic astigmatism with the rule.

II/11, aged 24 years. High myopic astigmatism with the rule.

III/4. High mixed astigmatism with the rule.

It is interesting to note that the astigmatism in this pedigree is with the rule in every case.

Pedigree No. 21.—III/8, aged 85 years (1932). Simple myopic astigmatism against the rule—Ocular palsy—cavernous atrophy of discs.

III/10, aged 84 years (1935). High compound oblique myopic astigmatism in right eye. Gross lens and fundus changes in left eye.

III/12, aged 82 years (1936). Compound hypermetropic astigmatism with cavernous atrophy of both discs, accompanied by right central scotoma.

III/13, aged 47 years (1910). Myopic astigmatism against the rule. Wife of III/12.

IV/4. Hypermetropic astigmatism. Also epilepsy.

IV/6, aged 61 years (1936). Compound hypermetropic astigmatism with the rule.

IV/7, aged 55 years (1932). Wife of IV/6. Compound hypermetropic astigmatism.

IV/10, aged 57 years (1933). Compound myopic astigmatism against the rule.

IV/11, aged 58 years (1931). Hypermetropia. Wife of IV/10. Died from 8th nerve tumour.

IV/14, aged 51 years (1936). Oblique compound myopic astigmatism.

IV/18, aged 53 years (1934). High oblique compound myopic astigmatism.

IV/20, aged 22 years (1906). Compound myopic astigmatism against the rule. Died of pulmonary tuberculosis.

IV/22, aged 58 years (1935). Compound myopic astigmatism against the rule.

IV/23, aged 54 years (1935). Compound myopic astigmatism against the rule.

IV/24, aged 53 years (1935). Compound myopic astigmatism against the rule.

IV/27. Emmetropic.

IV/29. Emmetropic.

IV/32, aged 51 years (1935). Compound myopic astigmatism against the rule.

IV/33. Presenile cataracts. Wife of IV/32.

IV/34, aged 48 years (1934). Pure myopia.

IV/37, aged 45 years (1931). Oblique compound hypermetropic astigmatism.

IV/42, aged 48 years (1932). Oblique compound hypermetropic astigmatism.

V/9, aged 35 years (1936). Hypermetropic astigmatism with the rule.

V/21, aged 16 years (1933). Oblique myopic astigmatism.

V/25, aged 26 years (1935). Mixed astigmatism.

V/32, aged 17 years (1933). Myopic astigmatism against the rule.

V/42, aged 15 years (1931). Pure myopia.

This pedigree seems to conform to both dominant and recessive types of transmission, but as many remain to be examined, one cannot be too dogmatic. Of the 13 cases of astigmatism no less than 8 were against the rule, which is significant.

IV/34 and V/42 are interesting, as father and daughter were the only examples of pure myopia in the pedigree.

Pedigree No. 22.—III/1. Low compound myopic astigmatism.

IV/1. Not examined.

IV/2. Low compound myopic astigmatism. Refraction very similar to father—III/1.

IV/7. High compound myopic astigmatism.

Pedigree No. 23.—I/2. Not examined.

II/3. High compound myopic astigmatism, with old choroiditis and positive W. R.

II/7. Not examined.

III/2. Moderate, compound myopic astigmatism.

The myopia in this pedigree is certainly dominant. 1/2 transmitted it to half her children, and so did II/3.

Pedigree No. 24.—II/1 and 2. No details.

III/6, aged 17 years. High compound myopic astigmatism of over 20 dioptries.

III/7, aged 14 years. High compound myopic astigmatism of over 10 dioptries.

Pedigree No. 25.—I/1. Normal.

II/3. Compound myopic astigmatism with the rule.

II/4. Compound myopic astigmatism with the rule. Stammers.

II/7. Compound myopic astigmatism with the rule.

II/8. Compound myopic astigmatism with the rule.

Examination of I/2 may be the clue to the mode of transmission.

Pedigree No. 26.—IV/2, aged 20 years. High myopia without astigmatism.

It is surprising that more members of this sibship do not as yet show myopia, in view of evidence of the disease in both paternal and maternal relatives.

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Nystagmus—Consanguinity

The spasmodic precipitation of hereditary eye disease had always remained a mystery to me, until I commenced to collect pedigrees for this paper. Then the pernicious influence of consanguinity became apparent, and will be referred to again under Leber's optic atrophy, and especially under retinitis pigmentosa.

In my pedigrees on nystagmus, consanguinity is evident both in pedigrees 4 and 30. It will be seen in pedigree 4 that in generation III two first cousins married in a family in which hereditary nystagmus was recessive, and as the result 3 of the 4

sibs of this union developed the disease in a gross form. In the case of pedigree 30, repeated consanguinity precipitated the first case of nystagmus in a perfectly healthy family. Many branches of this latter family are personally known and have been examined by me; and it is surprising, with the frequent inter-marrying which has taken place, that more members have not exhibited congenital defects in one form or another.

Mode of Transmission

Hereditary nystagmus may be transmitted in one of two ways :

- (a) Ambisexual group, where either parent transmits it to both sexes.
- (b) Recessive male limited group, transmitted by unaffected females to their male offsprings.

According to Knighton (1929) and Holm (1927) (b) method of transmission is rare. Pedigree No. 27 in which only males could be affected, will be watched with interest by me for evidence of sex-linkage.

Sex Incidence

In my 4 pedigrees, the sex incidence was 9 males to 3 females—*i.e.*, 75 per cent. males. I therefore took 5 available reported pedigrees, at random, and found that the total sex incidence in these was 42 males to 13 females, *i.e.*, 76.36 per cent. males. This is a very interesting comparison, and I think proves that the sex incidence in Tasmania conforms very closely to that in other parts of the world.

Concurrent Defects

Albinism.—Partial albinism is most apparent in nearly all cases of congenital nystagmus, and pedigrees 29 and 4 exhibit this tendency most markedly.

Head Nodding.—This has been reported by Cox (1936), and many other authors, but I have not found a case of this in my pedigrees. It must not however, be confused with lateral rotation of the head to one side, and both eyes to the other, which many patients take up in order to steady their eyes, and improve their visual acuity. Case III/81 of pedigree 4 had developed this habit to a marked degree. Nor must this head nodding be confused with spasmus nutans—a totally different clinical entity.

Astigmatism.—This is a most common feature as indicated by Holm (1927), and Niccol (1915), and its correction offers one hope

in the alleviation of this disease. Frank (1936) states it is the only treatment of any account.

Amblyopia.—This varies considerably in members of the same pedigree. Pedigree No. 27 has 3 affected members, I/1 and II/2 having 6/9 or better vision with correction; while II/1 would not improve above 6/24 in each eye. Similarly in pedigree 4, IV/18 would not improve above 6/60 right and left, while III/81 improved to 6/9 partly with astigmatic correction. It is interesting to note here, that Usher in 1912 (quoted by Niccol) reported a case of congenital nystagmus in which “a microscopic examination of the retina showed that there was no proper fovea and a few ganglion cells were present in this region. There was no evidence of lack of retinal pigment.”

Stammering.—Although none of the affected members of pedigree 4 was affected with stammering, yet two brothers of III/81 had this defect in a marked degree.

Treatment

Except for correction of refractive errors, and especially astigmatism, there is little treatment availing. But I must stress here, that careful retinoscopy combined with careful mydriatic and post-mydriatic subjective testing will frequently produce quite promising results, and due compensation for extra care taken. Orthoptic training which has been mooted on occasions, appears to hold out definite hope of improvement. III/81 has responded rapidly to it.

Comments on Pedigrees

Pedigree No. 27.—The father I/1 has high myopic astigmatism, the correction of which produces 6/6 in either eye. He also has 30° of alternating divergent concomitant strabismus, while both fundi are absolutely normal. His eldest son (II/1) aged 12 years, has hypermetropic astigmatism, but his vision will not improve above 6/24 in either eye. His brother (II/2) aged 10 years, on the other hand is practically emmetropic, and has 6/9 in each eye. Both sibs have normal fundi.

With regard to the nystagmus in this family, I/1 and II/2, who have good central vision, have only nystagmus when both eyes are deviated to one or the other side; while II/1 with poor central vision has his nystagmus most marked when looking straight ahead.

Pedigree No. 28.—Both affected members (I/2 and II/1) have fine horizontal nystagmus. While I/2, aged 36 years, has pure

myopia, and II/1, aged 9 years, has hypermetropic astigmatism—all four eyes correct to 6/9 all.

Pedigree No. 29.—The one examined member (IV/2), aged 10 years, has been under my observation for 6 years. He is partial albino, has gross mixed astigmatism, and with correction improves to 6/18. The nystagmus is more marked on looking laterally, and both maculae exhibit fine pigmentary stippling in contrast to the pale remainder of the retina.

No. 4 (See Astigmatism).—This complex pedigree has been referred to before under astigmatism. I shall here refer only to the right hand section of the pedigree. III/60 and 63 have divergent squints and amblyopia, but have been dealt with under astigmatism.

III/81, aged 13 years, the most recent member under observation, has marked nystagmus, with mixed astigmatism, the correction of which improves her vision from 6/24 to 6/12 each eye.

III/79 and 82. No nystagmus, but stammering.

IV/18, aged 19 years. Has high myopic astigmatism and nystagmus, with only 6/60 in each eye. His brother and sister, although not examined, I believe have grossly defective sight also.

IV/22 is interesting. Although he has no nystagmus, he has congenital amblyopia of his right eye, without any lateral deviation, and only 5 dioptries of vertical deviation.

IV/26. Has not been examined.

Pedigree No. 30.—This has been included to show how repeated consanguinity will precipitate congenital nystagmus. IV/2, aged 47 years, and V/1, aged 22 years, have been examined—the former is almost emmetropic and shows no nystagmus, while V/1, as well as the nystagmus, has high myopic astigmatism, and will not improve beyond 6/24 in either eye.

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(To be concluded)
