remain obscure it is a matter for comfort that it lies within our power to recognize that some of the more alarming episodes of medical practice need not, in fact, occasion any fear of an unhappy ending.

REFERENCES

- ¹ Gowers, Sir William: *The Borderland of Epilepsy*, 1907.
- ² Wilks, Sir Samuel: *Lectures on Diseases of the Nervous System*, 1883.
- ³ Ryle, J. A.: *Guy's Hospital Reports*, 1925, lxxviii, 371.
- ⁴ Idem: *Lancet*, 1931, i, 737.

THE MENACE OF HEREDITARY BLINDNESS

BY

J. MYLES BICKERTON, M.A., B.CH.CANTAB., F.R.C.S.

SENIOR OPHTHALMIC SURGEON, KING'S COLLEGE HOSPITAL; OPHTHALMIC SURGEON AND DEAN, ROYAL EYE HOSPITAL; CONSULTANT OPHTHALMIC SURGEON TO ST. OLAVE'S AND LEWISHAM HOSPITAL

The total world figures relating to blindness must be of staggering dimensions. In China one disease (trachoma) has been estimated to have blinded one million in both eyes, three to four millions in one eye, and to have gravely injured twenty millions—this out of a population of, say, 400 millions. In the United Kingdom the registered blind were reported as follows:

TABLE I.

Year	England and Wales	Scotland	Total
1919	25,840	—	—
1929	52,727	—	—
1930	56,853	8,516	65,369
1932	62,488	? 10,000	? 72,727
War blind	—	—	2,000

About 8,000 of these blind are also mentally defective, and many are deaf.

There has been an increase, therefore, of 35,000-odd in the registered blind in the last twelve years, while in the year 1929-30 the increase was 4,126, or over eleven a day. The total figure, however, is by no means a true indication of our blind population, for the following groups of persons are largely excluded, and may increase it to 250,000 blind persons.

1. Almost all blind persons of the well-to-do classes.
2. Infants below school age (many die blind).
3. Those who become blind after leaving school at 14 or later, and are not registered until reaching blind pension age at 50. Many of these refuse to be certified, regarding it as rather a disgrace.
4. Those who become blind after 65 and are not registered. They obtain the old-age pension, which is the same in amount as the blind pension—namely, 10s. weekly. This is a very large group, and is likely to grow as the average length of life increases.

5. Those blind persons who are eligible for the blind pension—that is, they are between the ages of 50 and 65—but who do not apply for it because they have never heard about it.

On one random afternoon at hospital recently I saw ten persons who, although blind, were not registered, or not likely to be registered, as matters stand to-day.

The certification of the blind, however, is done in a very haphazard way in this country, and the real causation of blindness is not truly shown, so that the figures in Table II are only approximate. A child, I should explain, is considered blind if it cannot read school books with safety to the eyes; while the criterion for an adult is the capacity to do work for which eyesight is necessary.

TABLE II.—Rough Figures of Causation of Blindness in the United Kingdom*

Cause	Per cent.	Largely avoidable per cent.
Trachoma	2	2
Purulent conjunctivitis	2	1
Ophthalmia neonatorum	5	5
Accident	5	2
Congenital defects	6	6
Senile degeneration	10	—
Glaucoma	10	1
Myopia	14	14
Syphilis	15	10
Senile cataract	15	1
Others... ..	16	2
	100	44

The 44 per cent. in column 2 may represent 70 per cent. of blind years lived. Those blind in one eye may number about one million.

Under 1 Year of Age*

Cause	Per cent.	Category
Accident	2	Neglect
Syphilis	5	Hereditry
Purulent conjunctivitis	8	Dirt
Congenital defects... ..	30	Hereditry
Ophthalmia neonatorum	50	Dirt
Others	5	—
	100	

(21 per cent. of the blind are blinded in the first year of life.)

A glance at Table II will show that blindness in the more active years of life is very largely avoidable, while much of it is due either to definitely dysgenic births or to the unrestricted production of children in poor or bad circumstances. In this table I have given the groups of the blind and the percentage in each group which could, in my opinion, be greatly reduced by certain preventive measures, since for practical purposes bad genes and certain types of bad environment may be included together as causes of blindness.

Trachoma (2 per cent.).—This disease does not occur in clean households (see China, Egypt, etc.).

Purulent Conjunctivitis (2 per cent.).—This condition rarely occurs in clean households, and in my opinion 1 per cent. is entirely avoidable (only one child in five is born into a clean home in England).

Ophthalmia Neonatorum (5 per cent.).—This disease, due to gonococcal infection at birth, could and should be entirely wiped out by the prevention of unsuitable births.

Accidents (5 per cent.).—A fair number of these occur to infants, a fact which has considerable relevance to the limitation of births. A woman explained in a South London court, for instance, that she was the only person alive and responsible for sixty-eight children (three marriages). There was recently reported a family in Buckinghamshire of thirty-nine children, and one woman, newly married, had nine children in two and a half years (three sets of triplets). How can she manage with a husband earning £3 a week?

Syphilis (15 per cent.).—It is probable that 10 of the 15 per cent. syphilitic blind owe their condition to congenital syphilis, which might be called a "pseudo-heredity" since it never goes beyond the second generation, and which could be entirely eliminated by preventing syphilitic women from having children. Naturally, if a woman has been shown (by the Kahn, Wassermann tests, etc.) to be syphilitic, she should avoid conception at all costs, unless, and until, she has been completely cured. If, however, her condition is not discovered until after conception, the problem becomes more difficult; but treatment during pregnancy will always obtain a non-syphilitic child if the treatment is thorough and commenced about the fourth month. If treatment cannot be thorough the choice is between the birth of a congenital syphilitic and inducing an abortion, and I, for my part—

* Column 1 is modified from Harman, and the other figures are my own estimates.

though I know many will not agree with me—am strongly inclined to the latter. Congenital syphilis, which shows itself in the eyes anywhere between the ages of 2 and 35 (usually between 5 and 25), is a slow and terribly distressing affliction to patient, parents, and even doctors. From two to ten years' treatment is often necessary before even a quiescent stage is reached, and it may cause blindness in three different ways: by interstitial keratitis (or inflammation) of the cornea; by a widespread choroiditis with destruction of the retina; or by juvenile optic atrophy. Needless to say, a congenital syphilitic is often ament, and is never really healthy. In my opinion, therefore, Wassermann (or other) tests should be compulsory in appropriate cases, and a positive indication should be followed by treatment, contraception, sterilization, or abortion, according to the circumstances. I have said "compulsory," but I believe that if the women of England knew that congenital syphilis, with all its terrible concomitants, could be entirely eliminated, they would welcome the appropriate routine from the medical profession. As it is, large numbers of syphilitics marry and produce defective children. At King's College Hospital last year 34 per cent. of the new syphilis cases—mostly eye cases—were congenital.

Of greater importance and interest are those types of blindness which are strictly hereditary—congenital defects 6 per cent., glaucoma 1 per cent., extreme myopia 14 per cent., senile cataract 1 per cent., other types 2 per cent.—a total of 24 per cent., or practically one-quarter of all forms of blindness. As a prelude to discussing them I should like to repeat the platitude, too often forgotten, that normal sight (emmetropia) is also hereditary, a great number of genes being jointly at work in its production. The various departures from normality are due to the aberration of one or more members of this gene-complex.

GENETIC TYPES OF BLINDNESS

Heredity is the study of differences or variations, the laws of which show that we may all differ in three ways: (1) by modification (environment); (2) by combination in accordance with Mendel's laws; (3) by mutation, the production of new hereditary types. In certain simple forms of life 10 per cent. of the offspring are mutations; and according to Darwin's law of natural selection, the best or hardiest of these varieties survive. Thus variation has a beneficial effect on the community if the law of natural selection is operative. The trouble in England now is that we have dysgenic selection. Mutations act very well plus eugenic selection. They act fairly well plus natural selection. They act badly minus natural selection, and very badly plus dysgenic selection. We must get back to natural selection first, and then to eugenic selection. This conclusion is logically irrefutable. Education and environment have remarkable effects upon the individual during his lifetime, but they do not influence his hereditary equipment.

We know more about the hereditary diseases of the eye than about those of any other organ, and for the good reason that, being the most important and complicated of our sense organs, its slightest defects cause marked disturbances of function.

The genetics of variations in normal eye colour are too well known—though not yet fully elucidated—for mention here, and the abnormal condition of albinism is also fairly widely recognized as a simple recessive. But the eye defects associated with albinism are not so generally realized, nor is it always understood that they are really but symptoms of a general deficiency. Practically all albinos are below the average in health and vitality, many are aments, and all, as a direct result of their condition, are photophobic, because their eyes lack the pigment to protect the sensitive retina. The conse-

quence is that they hang their heads, pucker their brows, and half close their eyelids to shut out the light; often, moreover, they suffer from nystagmus or myopia. The incidence in the population is 1 in 10,000, and 30 per cent. of it is due, like so many similar recessives, to consanguineous marriages.

CHART 1.—Albino (Tertsch).

The Myopia Group

The genetics of this varied group have not been fully disentangled, but one can say for certain that short sight never arises in the absence of a hereditary predisposition, though that predisposition may remain latent in good health or be accentuated by illness and physical disability—as the myopia increases the eye stretches until the retina is destroyed. Dysgenic births are the chief cause of severe myopia, and many high myopes go blind, while there are immense numbers whose very defective vision is practically equivalent to blindness, but who do not come within the scope of the Blind Persons Act.

Without intending to imply that it is a simple unit character, I must agree with Clausen, who considers that myopia tends to be recessive. Wilson, for instance, found that in 100 families with one myopic parent there were 200 myopic children and 250 non-myopic; while in ninety-one families with neither parent myopic only 100 children were myopes and 300 non-myopes. Therefore, although short-sighted offspring are often born of normal parents, when both parents are short-sighted their children invariably suffer from the same defect.

CHART 2.—Sex-linked myopia (Worth).

CHART 3.—Dominant myopia, with nystagmus (Vogt).

Wilson's figures are interesting. In 677 myopic children hereditary evidence was as follows: grandparents, 54; mothers, 369; fathers, 254; siblings, 164; uncles, aunts, 31; cousins, 9.

Father to son	...	...	...	...	79
Father to daughter	...	...	...	...	149
Mother to son	...	...	...	...	111
Mother to daughter	...	...	...	...	231

Hereditary evidence was found in 65 per cent. of low myopes, up to 3 dioptries; in 66 per cent. of medium myopes, 4 to 6 dioptries; in 67 per cent. of high myopes, 7 to 12 dioptries; and in 67 per cent. of very high myopes, over 12 dioptries. It was also found in 65 per cent. of

school children, 63 per cent. of home workers, 68 per cent. of factory workers, 60 per cent. of clerks, and 60 per cent. of labourers. The run of the figures in these last groups indicates independence of environment.

Reference must be made here to detachment of the retina, which is a fairly common sequel to high myopia. Bogatch reported a family of eleven high myopes, in seven of whom detachment of the retina supervened in one or both eyes. Retinal detachment is found as a dominant, recessive, or male sex-linked in different pedigrees.

In the same way astigmatism, long sight, and squint (which is often associated with long sight) may be inherited. Squint, from which more than 2 per cent. of school children suffer, is responsible for a large number of blind eyes.

CHART 4.—Long sight—internal squint (Clausen and Bauer).

CHART 5.—Hereditary nystagmus (Holm).

Microphthalmia, or very small eye, with anophthalmos (lack of an eye) as the extreme form, has been found as a dominant, recessive, or recessive sex-linked (Ash).

CHART 6.—Microphthalmia (Ash).

Retinitis Pigmentosa

Czellitzer says that nearly 4 per cent. of all the blind are due to hereditarily determined atrophy of the retina. Some 300 pedigrees are given in the *Treasury of Human Inheritance*. The condition begins in early youth and progresses to complete blindness sooner or later, often round the age of 40. As in other diseases, there are several forms, as shown by their different heredity, the commonest being recessive; 33 per cent. of those thus affected owe their condition to consanguinity. In some families the condition is linked with deafness.

CHART 7.—Retinitis pigmentosa (Sambuc).

CHART 8.—Retinitis pigmentosa (Mooren).

CHART 9.—Retinitis pigmentosa (Mann). Leber's optic atrophy.

Optic Nerve Atrophy and Optic Neuritis

These frequently cause blindness, and the hereditary variety is well recognized by ophthalmologists, 1,000 pedigrees being available for study; 85 per cent. of those affected are males. The disease comes on at different ages, the average being about 20. Diminution of vision is the chief symptom, and often appears suddenly; there is a central scotoma, and after six months or so the disease remains stationary. The malady has been traced once through six generations, twice through five generations, and often through four generations. Leber's optic atrophy is a name for the sex-linked variety. It appears to be an abiotrophy, or lack of vital force, occurring at the prime of life.

CHART 10A.—Hereditary optic atrophy (Hawkes).

CHART 10B.—Hereditary optic atrophy (Bickerton, 908).

CHART 11.—Hereditary optic atrophy (Thomsen, 802).

CHART 12.—Hereditary optic atrophy (Norris, 750).

CHART 13A.—Hereditary optic atrophy (Norris, 881).

CHART 13B.—Hereditary optic atrophy (Hogg, 837).

Aniridia

All cases of aniridia (absence of the iris) that I have seen have been of blind standard. I particularly wish to call attention to the Risley pedigree (1915) of aniridia: one blind man with this affection had thirteen children similarly affected (100 per cent.), sixty-one grandchildren out of sixty-three, and thirty-nine great-grandchildren out of forty-two (Chart 14).

Congenital Cataracts and Senile Cataracts

These are usually dominant, and are very important; Macklin states that 13 per cent. of the pupils in blind schools are blind through cataract, and Hirst gives 10 per cent. in 1,300 blind persons (anophthalmia and microphthalmia being next) (Charts 15 to 17).

CHART 15A.—Lamellar cataract (Nettleship).

1
13 Affected children (all blind).
63—61 Defective (blind).
42—39 Defective (blind).
CHART 14.—Aniridia [absent iris] (Risley, 1915).

FIG. 15b.—Dominant night-blindness. Nougaret family. 2,116 persons; 135 affected (Nettleship).

CHART 16A.—Cataract (Loeb, 328).

CHART 16b.—Cataract (Fromaget, 290 L).

CHART 17a.—Cataract (Loeb).

CHART 17b.—Coralliform cataract (Nettleship).

Ectopia Lentis

Ectopia lentis (congenital displacement of the lens) has been traced as a dominant through six generations, though in some pedigrees it is recessive. Cameron's pedigree shows both lenses affected in each case.

CHART 18.—Hereditary dislocations of lens (both) (Cameron).

Glaucoma

In this group the hereditary factor is not always very marked, though congenital glaucoma does occur, and there is a probable hereditary predisposition to it in many other cases. This is known as hereditary anophylaxis, or abiosis—a lack of vital force, which causes the structure to die before its time. Many pedigrees suggest a dominant heredity, but the hereditary course is occasionally interrupted, and casual causes are often necessary to bring out the disease. It may arise early in life as a juvenile glaucoma.

CHART 19.—Inflammatory glaucoma (Howe).

CHART 20.—Juvenile glaucoma (Kamenetzki).

CHART 21.—Glaucoma (Bickerton, 1932).

Blue Sclerotics

This is a condition in which the coat of the eye is too thin or too transparent, accompanied by a failure in development of connective tissues. It is often associated with fragilitas ossium, or brittle bones, and with a type of deafness due to otosclerosis. The female is more likely both to show and to transmit it. Seventy-three pedigrees since 1903 show 463 defective individuals; it is usually dominant. Of adults with blue sclerotics 60 per cent. had liability to fractured bones, 60 per cent. had otosclerosis, and 44 per cent. had all three defects.

CHART 22.—Blue sclerotics. x = Fragilitas ossium (Burrows). Otosclerosis 40 per cent.

Glioma

This is a rare malignant growth, occurring in infants, of the nerve elements of the eye, which in some families shows a strong hereditary tendency, and is apparently recessive. It often affects both eyes (25 per cent.), and their early removal is necessary to save life. Some sufferers survive, marry, and often transmit the defect. Mohr considers the condition to be lethal in a homozygous condition. The death of a child with glioma of the retina is an appalling experience for parents and child, and may take as long as two years. The law which forces us to withhold euthanasia in these cases at the same time forces us to act in a barbarous and monstrously inhuman fashion, and is, in my opinion, indefensible.

CHART 23.—Glioma retinæ (Newton, 339).

CHART 24.—Glioma (Wilson, 349).

CHART 25.—Glioma (Thomson and Knapp, 335).

CHART 26A.—Glioma (Griffith, 333).

CHART 26B.—Glioma (Letchworth, 1898).

Maculo-Cerebral Degenerations

Several conditions might be described under this heading, including the rare disease of the central nervous system known as amaurotic family idiocy. Commencing at about 5 or 7 years of age, the children go blind within two years and mentally degenerate within the next two; death occurs about the age of 17 or 18. Amaurotic family dementia is a diffuse tapeto-retinal degeneration causing total, or nearly total, blindness, beginning at

birth or later, and frequently associated with cerebral degeneration producing idiocy. It is a not infrequent cause of congenital blindness.

CHARTS 27A and 27B.—Amaurosis sex-linked (Sedgwick).

CHART 28.—Amaurotic family idiocy—"Tay Sach" (King, Rus, Stewart).

Night- and Day-Blindness

Night-blindness is due to a defect of the rod vision (colourless vision). At dusk these cases are blind, though their sight is normal in good daylight. The famous Nougaret pedigree, by Nettleship, shows 135 affected persons out of 2,116 in nine generations (a dominant heredity). There are also a recessive sex-linked type, which is regularly associated with myopia, and one or more simple recessive types (Switzerland, Japan).

CHART 29.—Progressive night-blindness. Death sixteen months after complete blindness (Bordley).

CHART 30.—Stationary night-blindness (Sedan).

Day-blindness is a cone vision defect; such cases are colour-blind and also of "blind standard."

CHART 31.—Day-blind (Hessberg).

CHART 32.—Day-blind (Lutz).

POSSIBLE REMEDIES

The cost in misery and unhappiness of this state of affairs is appalling, and the cost in money (potential happiness) runs into millions. Are we justified in looking on complacency or are we blameworthy? Our aims are to prevent blindness by improving environment and heredity. Here are some of the means available.

1. Constructive birth control to avoid overcrowding and poverty.
2. Sterilization of mental defectives (and possibly certain criminals) as in America.
3. Review of the obsolete and ancient abortion laws, seventy years old. They are no longer effective, and are acting deleteriously and dysgenically. Abortion should act as a second line of defence to conception control, as in Russia.
4. Euthanasia for infants with gross defects to be available for parents who wish to make use of it.
5. Segregation of the mentally defective is difficult owing to the cost; some 300,000 are now at large.
6. The Catholics recommend control of marriage for the mentally defective, but this does not prevent illegitimacy.
7. The Wassermann reaction during or before pregnancy. This is done in Rumania before marriage.

In Mexico the State of Vera Cruz has put into effect laws for the legalization of birth control, sterilization of certain criminals and mental defectives, prevention of hereditary ailments where practicable, and the prevention of marriage by persons who are mentally deficient or physically or economically unfitted to have children. How long must we in England continue with our present inhuman methods?

THE CLASSIFICATION OF THE BODY CONSTITUENTS BY WATER CONTENT *

BY

HAROLD G. CLOSE, M.D.LOND.

ASSISTANT BACTERIOLOGIST AND LATE GRIFFITHS DEMONSTRATOR OF
PATHOLOGY, GUY'S HOSPITAL

For the sake of convenience it has been customary to describe the body by regions (anatomy), systems (physiology), or origin from embryonic layers (embryology), but for biochemistry there appears to be no such simple grouping. To remedy this deficiency a classification based on water content has been made and is presented here. Water deserves to be singled out for distinction because of its unique position in the economy of the body, both in the past and now. Besides being at one time the only medium in which life existed, it now forms the greater part of all living tissues, and plays an essential part in most, if not all, biochemical reactions. The data from which this classification was derived were obtained partly by estimations made on the chloride and water of tissues, and partly from the literature, and have been published elsewhere.¹ Unfortunately, there are still many gaps in our knowledge, but it is possible even now to see the outline of the pattern underlying the construction of tissues.

THE THREE TISSUE GROUPS

If the parts of the body are considered from the point of view of their water content they will be seen to fall naturally into three groups.

- A. The body fluids (extracellular).
- B. The nuclear or cellular tissues.
- C. The anuclear, "matrix," or supporting tissues.

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