being unable, for valid reasons, to attend at the appointed place, the Local Authority shall arrange for a certifying ophthalmic surgeon to visit such a case and pay expenses. (5) Every certificate should be made out on a form prescribed by the Ministry of Health (copy attached). (6) The information so obtained should be collected and studied with a view to prevention of blindness.

There is a short paragraph on the importance of the examination of children of school age who show serious visual deterioration, and a discussion on the definition of blindness. In this the Committee suggest three groups: Group 1, vision below 3/60 Snellen; group 2, vision of 6/60 Snellen or better; group 3, intermediate degrees of visual acuity. The condition of the visual field is strongly emphasized: hemianopia of any kind with tolerably good form vision (6/18 or better) should not be certified; and finally of paramount importance is the age at which blindness occurred.

Among the appendices will be found the report of the Council of British Ophthalmologists which has formed the basis on which this report has been built.

A word of praise is due to the Secretary, Miss Cracknall, for her share in the preparation of this report.

We consider that this report should be in the hands of every practising ophthalmic surgeon in the country; it can be obtained at a cost of 1s. from the Secretary, Prevention of Blindness Committee, 66, Victoria Street, S.W.1.

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**ABSTRACTS**

**GENERAL MEDICINE**


This comprehensive and important work extending to some 228 pages appears in *Hereditas*, a periodical published by the Mendelian Society in Lund. In the introduction Sjögren gives an account of the work of previous authors on the subject of juvenile amaurotic idiocy and a description of the disease based largely on a review of the subject by Schob (1924). The author's own material was obtained from certain institutions for the blind in Sweden and it is surprising how many cases of juvenile amaurotic idiocy he found, as
hitherto no case of this disease had been published in Sweden. In
the Värd institution for the blind in Lund, he found 29 cases which
showed the disease from the earliest to the latest stages. These he
personally examined. Syphilis was always excluded. The negative
result, together with the general examination, and ophthalmoscopic
examinations by Doz. B. Rosengren with their surprising uniformity,
made, he considered, the diagnosis certain. A histological examina-
tion in 18 of these cases was made by Prof. Einas SJövall which in
all cases confirmed the clinical diagnosis. From records at the same
institution he found nine more cases. From Drottning Sophias
Stiftelse in Vänersborg he obtained 34 cases; one of these was
personally examined. Tomteboda cases numbered eight and Jäxjö
cases five; of these SJögren examined four. Cases that were not in
any of the above institutions numbered 30; in four of these the author
made a clinical examination. Five cases were from colleagues. The
diagnosis in one case was made in the psychiatric clinic at Stockholm
and confirmed anatomically by Prof. E. SJövall.

The clinical analysis is based principally on the 39 cases of
juvenile amaurotic idiocy investigated by the author, who had the
opportunity of following personally the majority of these cases for
from two to three years. The children develop normally until the
fifth to eighth year when the disease begins with marked diminution
of vision which in the course of one to two years leads to blindness,
only the power to count fingers or perception of light remains. In
the early stages the optic disc is yellowish grey with narrow
vessels. The thin vessels are very characteristic and are present
before vision is lost. In these early cases pathological pigmentation
is not as a rule present. The changes are symmetrical. In some-
what more advanced stages there are found at the periphery—and
this is by far the most frequent type of degenerative change—small,
round, closely placed and partly confluent spots with badly defined
dges, with sparsely scattered pigment granules, as well as retinal
atrophy of the papilla; in other cases the ophthalmoscopic
appearance is exactly that of retinitis pigmentosa, although differ-
ing from typical retinitis pigmentosa by its early onset and
rapid progress. Still later bone-corpuscle-like pigmentation
appears at the periphery of the retina and in the most advanced
cases may cover the fundus completely. At this stage pos-
terior cortical cataract sometimes appears. Pupils in the early
stage have good reactions, with on-coming amaurosis they become
fixed. In seven cases in the earliest stage no ophthalmoscopic
changes were seen though there was amaurosis or marked
diminution of vision. All these cases (except Berta J., who was
only once examined) at later stages showed ophthalmoscopic
changes of characteristic type. SJögren then refers to particulars
of seven cases that seemed suitable to throw light on whether the
conception of "familial maculo-cerebral degeneration" advanced by certain authors (Batten, 1903; Oatman, 1911; Stargardt, 1913, 1917) is justified. All of these seven cases have shown in the earliest stage degenerative macular changes and five of them on subsequent ophthalmoscopic examinations have shown the characteristic ophthalmoscopic picture of juvenile amaurotic idiocy. As regards the onset of blindness the author's material shows that when the disease comes on in two or more siblings, as a rule it is at the same age and likewise of the same type.

Psychic symptoms appear about the same time as the onset of blindness though occasionally later. There is a slow progressive failure of all psychic functions. Epileptic attacks may set in early, but usually they occur at later stages. Disturbance of speech is noticed about the same time as psychic deterioration begins. It is very constantly found two years after the onset of blindness. It is characterized by a peculiar stuttering, often explosive articulation, with reiteration of words or syllables—at this stage dementia of imbecile type occurs. The disease has now so progressed that the children can no longer be instructed in the blind school.

An interesting account is given of the neurological symptoms. This is accompanied by photographs to show characteristic attitudes and disturbances of gait at different stages in subjects of juvenile amaurotic idiocy. There is disturbance of gait in form of marche à petits pas, often combined with a Démarche trépidante. As a rule, signs of a pyramidal path lesion have not been found. Phenomena like astasia abasia, as well as balance disturbance as in cerebellar ataxy, occur in later stages. A crouching attitude with head and upper part of body bent is manifest. When these patients try to walk it is often observed that they tramp on the same spot before progress is made. In the last stages they sit where they have been placed, without interest in anything, with dull fixed mask-like expression—speech has gone—death occurs often from intercurrent disease. The age at death is shown in Table V. In Groups I and II it is about 17 years and in Group III 18 years.

Differential diagnosis. Pathognomonic parenchymatous changes make the diagnosis clear in cases where the brain has been examined anatomically. In clinical differential diagnosis, chiefly of interest are juvenile general paralysis, some forms of congenital cerebral syphilis and cerebro-spinal affections with optic atrophy or choroid-retinitis, as well as certain rare forms of familial juvenile diffuse brain sclerosis.

In the part of this work devoted to heredity the material has been worked out and analysed in the same thorough manner as in the clinical part. For hereditary statistical analysis the author's own material consists of 59 families, belonging to 50 stocks with 115 cases of juvenile amaurotic idiocy. Early in the investigation it was
observed regarding geographical position that these stocks showed a
distinct tendency to be collected at centres. Further the tendency
to migrate was strikingly insignificant. Consanguineous marriages
in the parents of cases of juvenile amaurotic idiocy have been worked
out as well as their frequency compared with those occurring in
the general population. In a summary of the nearer degrees of
consanguineous marriages, namely marriage between (1) 1st cousins,
(2) 1st cousins once removed, (3) 2nd cousins, the following results
are given. For Group I, 25, 7±7, 4 per cent. Group II, 27, 5±6,
3 per cent. Group III, 25, 4±5, 7 per cent. The corresponding
numbers for the general population are 2:3 per cent. (Lenz), 1:29±0,
0:09 per cent. (Wulz), 9:6±1:4 per cent. (Spindler). This marked
increase in the percentage of consanguineous marriage is the most
important criterion for a recessive inheritance. As Lenz (1919) has
shown, the inbreeding frequency of parents of recessive-homozygotes
must be all the more frequent the rarer the disease is in the population.
From the analysis in Table XVIII, the 59 families are seen
to be at 23 centres that are distributed in south and middle Sweden.
The centres consist of country parishes that are often relatively
isolated. With few exceptions, as regards the ancestors of the
parents, we have to do with a purely country population that
consists mostly of farmers and farm workers.

Sex proportion: Of the 115 cases with juvenile amaurotic idiocy
61 are males and 54 females. The sex proportion in Sweden at the
age of seven years (1911-1920) is 104 boys and 100 girls.

There were five twin births. Of these 10 twins, two have juvenile
amaurotic idiocy. The twin sibling of both died at a very early age.
Four are normal and two have not yet reached the age at which
juvenile amaurotic idiocy becomes manifest.

A summary of the hereditary analysis shows that juvenile
amaurotic idiocy follows with a high degree of probability the
course of a recessive monohybrid inheritance. The disease from
the hereditary aspect is quite distinct from the infantile form.
The analysis of the ascendants and the probable heterozygote
lineage has shown a distinct tendency to accumulation in certain
defined areas in different parts of the country. Approximate
calculation of heterozygote frequency in the country has given for
this disease a value of about 1 per cent.

From the year 1917 a continuous marked diminution of the
frequency of juvenile amaurotic idiocy in Sweden has been
shown. The natural explanation of this may be a diminished
frequency of consanguineous marriages from an increased migratory
tendency as well as from possibly diminished birth rate.

Statistical investigations have been carried out in certain categories
of relatives of juvenile amaurotic idiots, namely siblings, step-
brothers and sisters, parents and parents' siblings. For the last
category, which represents the largest material, the following numbers have been obtained of probability of disease (Group III). For juvenile amaurotic idiocy: 0.49±0.34 per cent. For epilepsy 1.05±0.52 per cent. For dementia praecox 1.46±0.65 per cent. For "oligophrenie" 1.50±0.61 per cent. The numbers have, in comparison with those of the German research institute for psychiatry in Munich for the general population, been found high, but on the other hand opposed to this no statistical significant difference could be found in regard to a single disease.

Sjögren's work is well illustrated by numerous figures, pedigree charts, tables, and some photographs, and there is appended a bibliography. The author of this valuable and very interesting article, which will appeal equally to neurologists and ophthalmologists, thus briefly gives his principal results.

1. It has been shown that juvenile amaurotic idiocy in Sweden appears relatively frequently in proportion to its extraordinary rarity in foreign literature.

2. It was established that the ophthalmoscopic changes as well as the development of the neurological symptoms showed marked similarity and constancy in their progressive course. Special attention has been drawn to the importance of symptoms of extra-pyramidal motor character, motor disturbance, disturbance of gait in the form of a marche à petits pas, often with Démarche trépidante with in addition crouching attitude, hypertonus of preponderating rigid type. The typical course of development and symptoms of the disease have been described in form of a classification. The disease shows in its typical manifestations so characteristic a picture that as a rule a certain clinical diagnosis may be made, even though it appears singly in a family.

3. Juvenile amaurotic idiocy follows with a high degree of probability the course of a recessive and monohybrid inheritance.

4. The disease in hereditary respects is quite different from the infantile form.

5. The ancestors who are probably heterozygotes show a distinct tendency to accumulation in several defined areas in different parts of the country.

6. The statistical investigations in regard to the inheritance of dementia praecox, "oligophrenie," and epilepsy amongst the siblings of the parents, have given a remarkably high figure for disease probability, without, however, significant differences being obtained regarding the individual diseases found in the general population.

A. F. MacCallan.