retraction of the wound, hypotony, sensitiveness either spontaneously or to light touch, and failing vision.

The object now is to apply with discretion the histological facts to the procedures for the removal of the exciting eye. Whenever enucleation is performed as a preventive measure the ordinary method of cutting the nerve on a level with the outer surface of the sclera may be employed with safety. Once signs of sympathetic ophthalmia appear the only hope of saving the uninjured eye lies in the immediate removal of the source from whence came the invasion and from which fresh virus would constantly be sent forth. Before operating it is possible to gain valuable information from examination of the sympathising eye as to the state of the infiltration in the injured eye and its appendages. If central synechia of the iris can be partially broken up and kept under control it is likely that the infiltration is in the first stage in the injured eye or at least is still intra-ocular. Should the iris be swollen and discoloured or, most important of all, should the pupillary zone be hopelessly bound down, these are signs that the inflammation has reached its acme in the exciting eye and that the extra-ocular structures are infected. The difficulty of removing an acutely irritated and painful globe surrounded by orbital cellulitis seems to account for the many instances in sympathetic ophthalmia in which the optic nerve is cut within the scleral canal. A long strip of the inferior oblique muscle should be excised and the optic nerve and the retrobulbar tissues around it should be removed almost to the apex of the muscle funnel. The wound should be left open to drain. If there remain in the exciting eye some degree of vision it is axiomatic to postpone enucleation as the benefit of the operation is always problematical and there is no justification for sacrificing even the faintest vision.

LINDAU'S DISEASE—PROGRESSION IN AFFECTED FAMILY

BY

ALEXANDER E. MACDONALD
TORONTO, CANADA

It is unfortunate that Lindau's disease has been inextricably associated with allied conditions in so many published reports of the clinical findings and pathological records. It is a rare condition, and to substantiate the diagnosis it seems reasonable to require that the presence of multiple haemangioendothelioma be verified, for at
least one member, with a similar tumour in another member of the family. It seems certain that this will happen more frequently during the life of the patient, as the advances made in surgery of the brain indicate that earlier diagnosis is probable.

We have had the opportunity to follow the course of Lindau's disease in a family where three of the four members of a sibship has each undergone the successful removal of cerebellar tumours, which on pathological examination proved to be haemangioendothelioma.

The death of sibling two, a female, occurred 3½ years after the cerebellar tumour was removed from sibling four, the youngest and only male in the family. All four members have had eye involvement, and sibling two had a cataract, in a glaucomatous eye which after extraction of the cataract developed pain so severe that enucleation resulted. The early pathological report showed that a retinal detachment due to intra-ocular haemorrhage was present, and it was only after sibling one had a cerebellar tumour removed and verified as a haemangioendothelioma that a further search was made for the presence of a tumour in the enucleated eye of sibling two. A haemangioendothelioma was found when further sections were made and the findings reported in the Transactions of the American Ophthalmological Society, Vol. 37, 236-250, 1939, and Archives of Ophthalmology 23: 564-576, 1940. The interest in this remarkable family, which produced for several years three verified living cases of Lindau's disease, may justify the following brief review of the present status of the various members and the clinical report of sibling IV who previously was reported as being healthy.

**Review**

In 1912 the mother of this family, at the age of 22 years, had a painful eye enucleated, according to Dr. Wilber Fraser of Ottawa who found the record in Dr. Minnes' files. The globe was opened and a complete cup-like calcification of the choroid was found, but no microscopic sections were made. The family state that her death resulted after an operation for a brain tumour in 1918, but no record of this could be found at the hospital in Ottawa.

**Sibling II**: In 1934, while bending over, suddenly lost the sight of her left eye, due to an intra-ocular haemorrhage. On February 24, 1936, at the age of 23 years, at a second stage cerebellar exposure a vascular tumour the size of a walnut, which pressed on the medulla, was found in the lower hemisphere. The upper part of the spinal cord was cystic.

Roentgen therapy was administered to the region, seventeen
PLATE I.

Fundus O.D. of Sibling I before radon treatment; showing detachment about peripheral tumour and dilated vessels.

PLATE II.

Fundus O.D. of Sibling IV before radiation. The capillary angiom covers the upper part of the disc and extends into the retina above.
times, and she was re-admitted for vomiting caused by this treatment.

March 30, 1937: Left cataract extraction.

May 31, 1937: Enucleation L.E. blind and painful due to secondary glaucoma. The pathological report of this eye at first showed that the detachment of the retina was due to intra-ocular haemorrhage. A re-examination was made in April, 1939, and a haemangioendothelioma was found and reported.

August 10, 1939: A cystic cerebellar haemangioendothelioma was removed and verified by pathological examination. Following this she remained well and carried on her work as a nurse until headaches and unsteadiness of gait developed early in 1947.

August 5, 1947: At a re-exploration of the cerebellum two large tumours were found; one was the size of a hen's egg, the smaller in the roof of the fourth ventricle. Both were removed, and it was found that the tumour infiltrated the muscles of the neck.

August 8, 1947: Following the operation she developed signs of medullary failure, was unable to swallow, and died. The autopsy revealed multiple cysts of the pancreas, kidneys, chronic interstitial pancreatitis and terminal broncho-pneumonia.

Sibling I: Female, aged 29 years.

May 6, 1938: A haemangioendothelioma, verified by pathological examination, was removed from the left cerebellum. This firm tumour was the size of a walnut. It was attached to the dura, lateral sinus and tentorium by large thin-walled vessels, which contained arterio-venous blood. For three years she had noticed a loss of vision in the right eye. The fundus showed central degenerative changes and her visual field showed a lower nasal defect which extended to 40 degrees from central fixation. With a widely dilated pupil, a whitish mass with a haemorrhage on its surface was seen well out to the periphery. The upper temporal vessels were greatly dilated (see plate I).

May 28, 1939: Four radon seeds of 0.7 millicuries each were placed subconjunctivally on the sclera, opposite the site of the tumour and removed seven days later. The seeds were tied separately to a silk thread and this part of the suture was dipped in thin pyroxylin solution. Moderate reaction followed at first, but for many months a depression at the treated area remained on the globe. Possibly one-half the dosage would have been sufficient to destroy the retinal tumour.

March 30, 1942: She complained of staggering, weakness of left arm and difficulty in controlling her speech. At this time RV=C.f. at 2 m and LV=6/18. The right eye was divergent 40 degrees.

April 24, 1947: A tumour the size of a walnut was removed from approximately the same area as at the previous operation. Dr. Eric
Linell reported the tissue as being extremely cellular. The cells were uniform in size with fusiform nuclei and a moderate amount of cytoplasm. The cell boundaries were indistinct. Some large blood channels were present but the Mallory stain showed capillary-like channels in large numbers. No mitotic figures could be seen with the cresyl violet stain.

August 6, 1947: The family stated that she still complained of unsteadiness of gait and deafness in the left ear, but she was able to do her housework.

Sibling III: A female, was re-examined on August 6, 1947. The naevus on the limbus of the left eye was less distinct than when seen in 1938. She looked well and stated that her health had been good.

Sibling IV: A synopsis of his voluminous history follows: Flt. Sgt. W. R., a white male, aged 27 years, began to complain of headaches, which were worse when he was in the prone position and during the morning. They were severe and gradually became worse. In July, 1942, he overturned a plane while landing it, and a careful physical examination followed. His history was referred for advice to Dr. K. G. McKenzie, who recalled the family history and asked for his admission to Christie Street Military Hospital, as from the record the presence of a brain tumour was considered possible.

January 28, 1943: The vision in each eye was normal with correction. The fields of vision showed only a slight enlargement of the blind spot in the right eye and the fundi were reported as being normal, but one week later it was stated that the right disc showed blurring on the upper and nasal borders with a moderate physiological cup. The left disc showed a slight haze at the upper border. Horizontal nystagmus was present, bilateral, more marked on looking at the right. The gag reflex was absent, the electrocardiograph negative. The impression was that the relative hypertension 154/114, nystagmus and absent gag reflex might be due to hyperinsulinism, an adrenal or intracerebral tumour.

February 2, 1943: Dr. McKenzie, in view of the above examination, combined with the family history, injected the ventricles and found the left lateral hemisphere under pressure, the whole ventricular system was enlarged and the picture suggested the presence of cerebellar angioma. He was transferred to the Toronto General Hospital for the operation.

February 27, 1943: A vascular tumour the size of a walnut was found in the angle between the right tonsil and the cerebellar hemisphere which shelled out by finger dissection. See Figs. 1 and 2. The post-operative treatment required a re-opening of the wound to remove an extra-dural blood-clot that developed within
24 hours, but by March 5, 1943, he was well enough to be transferred to his military hospital. At this time he had a marked horizontal nystagmus, dizziness and bilateral 6th nerve paralysis with right sided inco-ordination, but five days later he was able to sit up. One week later he could walk with help.

April 7, 1943: No headache and no diplopia was present, but there was slight inco-ordination of both hands and feet. He was discharged to return for a check-up visit in six months.

April 9, 1947: He was re-admitted to Christie Street Hospital
-complaining of fatiguability, dysfunction of balance and slight loss of hearing of the right ear.

Laboratory examination.—Sedimentation rate 16 mm., urine acid, specific gravity 1021, negative for albumin and sugar, Kahn negative. Examination revealed no corneal reflex and cranial nerves essentially negative. Personal history: He had returned to work two months after his operation, as a checker in a lumber camp, but at the time of this admission he had been teaching in a school.

February 2, 1947: The fields of vision showed slight enlargement of the right blind spot. The pupils were equal in size, reacted to light and convergence, full ocular movements were present with no nystagmus. The disc of the right eye showed a reddish elevation over the upper half with new blood vessels. The superior temporal vein was engorged. Radiation was suggested for this developing angioma. He was seen also at this time by Dr. McKenzie, who found no evidence of a recurrence of the brain tumour.

June 10, 1947: See colour drawings of the right fundus showing angioma.


In the fundus of the right eye the media were clear. There was considerable connective tissue over the upper part of the disc with new loops of capillaries extending up into the retina. These were elevated about three dioptres. The mass was purplish-red and extended down over the cup. The fundus of the left eye essentially negative, only a slight crescent and clear, physiological cup. Partly on account of his obligation to teach in September and his good vision, he requested that the treatment be delayed. He stated that his four children: girl of 8, boy of 6, girl of 2 and a one-month old boy were well.

January 10, 1948: Fourteen treatments have been given to the area of the developing eye tumour. The vision R.V. =6/18, L.V. =6/12 and normal in each eye with correction.

The right fundus showed an area of grey oedema over the tumour mass which was seen with a plus four lens. Above the disc the capillary loops extended higher into the retina when compared with the drawings made in May, 1947.

DISCUSSION

The familial condition where multiple capillary haemangioendothelioma occur in the cerebellum, cerebrum, retina, spinal cord, kidneys, pancreas, epididymis, liver and bone, with occasionally hypernephroma, should be termed Lindau's disease.
A review of a family and the most recent case history in the only male has shown that the cerebellar tumour was verified for sibling I at the age of 29 years by pathological section and an eye tumour was treated by radon seeds. Sibling II—the cerebellar tumour was verified at operation at the age of 23 years and by pathological section at two subsequent operations. The eye tumour presented a similar pathological appearance. Sibling III had a naevus of the limbus of the left eye, which became less apparent after nine years. Sibling IV: The new case was verified by pathological examination at the age of 28 years and after extensive radiation an eye tumour, capillary in type, has slightly progressed in four years.

**Acknowledgment:** To Dr. K. G. McKenzie my thanks and congratulations for the surgical care and remarkable results that have meant so much to the members of this family. Dr. Eric Linell was responsible for the excellent pathological reports and photographs. Miss Wishart made coloured drawings, which add much to the written description. Also thanks to the Departments of Surgery, Neuro-Pathology, Veterans' Affairs, Ophthalmology, Art and Photography.

**METHODS OF INVESTIGATING EYE MOVEMENTS**

**By**

**H. HARTRIDGE and L. C. THOMSON**

**VISION RESEARCH UNIT, MEDICAL RESEARCH COUNCIL, INSTITUTE OF OPHTHALMOLOGY, LONDON**

Opinions differ widely at the present time with regard to the ability of a subject to perform accurate fixation, for whereas some hold that the eye is constantly making rapid oscillations, on the presence of which visual acuity for fine detail actually depends, others hold, on the contrary, that such oscillations are absent in normal subjects and that if these movements were present, far from aiding the perception of detail, they would have the effect of seriously interfering with it. Thus Hering (1899) thought that the irregularities at the contours between the images of objects which are produced by the retinal mosaic are smoothed by chance eye movements. Anderson & Weymouth (1923), Averill & Weymouth (1925), and Marshall & Talbot (1942) have held similar views, and they have extended the consideration to the cases of small differences of size and small defects in alignment. Wright (1942) claims that the eyes are continually performing "twittering" movements, and that in consequence the receptors are able to scan the retinal image in the same way that the bright spot in the cathode ray tube of a television receiver scans the fluorescent screen. He writes, "It is the dimensions of the cones rather than their number