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# OTOGENIC INTRACRANIAL HYPERTENSION

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INCLUDED amongst the intracranial complications of otitis media are many and varied clinical and pathological conditions; amongst these is a well-defined syndrome for which the name, " otitic hydrocephalus ", was first applied by Symonds in 1931.

Quinke, in 1896, described a syndrome, in which the signs of increased intracranial pressure were associated with the finding of clear cerebrospinal fluid on lumbar puncture. He was of the belief that the excess of cerebrospinal fluid was due to hypersecretion from the choroid plexus and referred to the condition as " serous meningitis " and described otitis media as one of the predisposing factors.

Other terms, which have been coined to describe conditions of this type were "cerebral pseudo-abscess ", " meningismus ", " serous ependymitis", "arachnoiditis", "hypertensive meningeal hydrops" and "toxic hydrocephalus ".

Symonds described several cases of this nature, and all of them occurred in patients suffering from ear disease. Without the aid of encephalography, he assumed that hydrocephalus was present, and he termed the condition " otitic hydrocephalus ".

Although its association with otitis media is best known, it is probable that other sources of sepsis, such as sinusitis, tonsillitis, and, more rarely, infectious fevers may be followed by a similar condition. In other cases, no focus of infection is demonstrable. It is therefore apparent that the syndrome may be caused by a variety of conditions, and in this respect the term " otitic ", may be misleading. The term, "hydrocephalus ", is not justified, because encephalographic examination shows that the ventricles are not enlarged, and, in fact, may be even smaller than usual. In this

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paper, however, no further attention will be paid to the nasopharyngeal or other conditions, which may have a causal relationship to it, and attention will be entirely devoted to the intracranial hypertension of otogenic origin. The condition will be referred to as " otogenic intracranial hypertension ", which appears to be a more adequate name.

**Clinical Manifestations**

The syndrome most often affects children and young adults, and is more common in the female sex. It may complicate acute or chronic otitis media and most patients show signs of mastoiditis. It occurs most frequently in right-sided otitis media.

There is increased intracranial pressure, with its associated clinical symptoms, such as headache, which may be slight or severe, nausea and vomiting, and swelling of the optic discs.

The most characteristic symptom is a clearly visible papillœdema usually connected with small hæmorrhages and occasionally with patches of exudate. The optic discs often show as much as 5-6 diopters of swelling. Visual fields show a tendency to generalized constriction. Nystagmus may be present, and occasionally the patient complains of giddiness. The sixth cranial nerve becomes frequently paretic, either on the side of the affected ear, or on the contralateral side, and in rare cases both abducens nerves may show paresis. The temperature may be elevated in the initial stages, but usually the pyrexia is of short duration.

The general condition of the patient is surprisingly good. The patient is usually physically fit and mentally alert, but dulling of the intellect may occur with the higher degrees of papillœdema.

Lumbar puncture is important in establishing the diagnosis. The pressure is high, usually exceeding 300 mm. $H_2O$, and it may be as high as 950 mm. $H_2O$, as in one of the cases recorded. Bacteriologically and biochemically the cerebrospinal fluid is normal. A large amount of cerebrospinal fluid is obtained on lumbar puncture. Another frequent observation is that, when trephination is performed, the exposed portion of the arachnoid membrane is found to be distended with fluid. If a canula is introduced into the lateral ventricle, a copious flow of cerebrospinal fluid occurs, which led to the erroneous assumption that hydrocephalus is present, before ventriculographic examinations were carried out.

The examination of the C.N.S. is singularly negative, with absence of any localizing signs, apart from the previously mentioned papillœdema and sixth nerve palsy, which leads to diplopia, usually only of a transient nature.

The condition usually clears up after weeks or months as a result of appropriate treatment, but if treatment is not carried out, or delayed too long, optic atrophy and blindness may ensue.



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# Otogenic Intracranial Hypertension

## Differential Diagnosis

Papillœdema and headache developing in a patient, who had recent middle-ear disease tends to make one suspect the development of such a complication as brain abscess. However, there are certain clinical and biochemical features, which make it possible to differentiate between otogenic intracranial hypertension and a localized affection of the brain, even in its initial stages. In the otogenic syndrome, the most characteristic feature is clearly visible papillœdema usually associated with small hæmorrhages, the optic discs often showing 5-6 diopters of swelling. In the case of a brain abscess, the disc swelling is not so pronounced and cannot be observed before the abscess has reached an advanced stage. At that time other neurological signs are usually present and help in establishing the diagnosis. In otogenic intracranial hypertension the general condition of the patient is surprisingly good, between the attacks of vomiting and nausea. In a brain abscess the patient's general condition is usually much more affected, and there is as a rule an increase in the pressure of the cerebrospinal fluid, but never so pronounced as in otogenic intracranial hypertension, when the pressure usually exceeds 300 mm. $H_2O$. In brain abscess one can often observe an increase in cells and a slight excess of protein in the cerebrospinal fluid.

A diagnostic difficulty may arise in cases, where development of the intracranial hypertension is preceded by a phase of meningitis with changes in the cerebrospinal fluid. In such an event, a correct diagnosis cannot be made, until, after repeated lumbar punctures, the fluid returns to normal.

Another condition, which has to be taken into consideration, is a tumour of the posterior fossa. The differentiation from a tumour of the cerebellum or fourth ventricle may be impossible on purely clinical grounds. However, careful attention to certain details in the history may be helpful in those cases, where physical signs are scarce. As a rule, a tumour in this situation shows slow progression. Headache, for example, due to a cerebellar tumour, may at first be intermittent, increasing in severity as the weeks pass, contrasting with the initial severe headache in otogenic intracranial hypertension, which diminishes, or even disappears after the first two or three weeks. Again, double vision, when it occurs in otogenic hypertension, does so within the first three weeks as a rule, whereas its appearance in a cerebellar tumour is usually later. In the case of a cerebellar tumour, repeated examinations will usually reveal some evidence of inco-ordination in the limbs or walking, which is absent in intracranial hypertension due to ear disease.

Ventriculography is used for the purpose of differential diagnosis in order to exclude an abscess, tumour, or other space-occupying lesions within the skull. Characteristic results of ventriculography are : Normal ventricular system, as far as size, position and shape are concerned, and a



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certain difficulty in filling the ventricles, because the gas streams into the subarachnoid space and out into the convex sulci. If these are the findings on ventriculography, then, in association with the clinical symptoms and signs, the diagnosis of otogenic intracranial hypertension appears to be justified.

Ventriculography not only permits of establishing the correct diagnosis,but also allows one to proceed with the treatment of repeated lumbar punctures, which can now be carried out with impunity, without fear of producing a pressure cone, as may be the case if there is a localized space-occupying lesion.

### Ætiology and Pathology

The pathologic-anatomical base for the conception of otogenic intracranial hypertension is not known, and the ætiology is very uncertain.

Dandy reports twenty-two cases with increased intracranial pressure and normal spinal fluid. Only one had chronic otitis media ; the others gave no history and showed no sign of affection of the ear.

Many authors regard the cause for this condition to be increased secretion from the choroid plexus or diminished absorption of cerebrospinal fluid, or possibly a combination of those two factors. Other authors, amongst them Warrington, maintain that the ætiological factor could be an inflammation somewhere else in the organism and that the affection of the meninges is only secondary.

The name, " serous meningitis ", is not adequate, as it is not an inflammatory disease. At a post-mortem examination of a patient, treated by Davidoff and Dyke, where death occurred from an intercurrent disease, no signs of inflammatory changes could be observed in the pericellular and perivascular parts of the brain.

The possibility of an obstructive internal hydrocephalus from inflammatory swelling or exudate was considered, but was thought to be unlikely owing to the large volume of cerebrospinal fluid obtained on repeated lumbar puncture. Moreover, in this syndrome, the residual pressure of cerebrospinal fluid after withdrawal of 15-20 c.c. is relatively high, a point which is against the theory of obstructive hydrocephalus.

A feature which has engaged the attention in the condition of otitic intracranial hypertension is the large proportion of cases in which there has been an associated thrombosis of the lateral sinus on the side of the affected ear. It has been suggested that thrombosis of the lateral sinus may in itself be responsible for the increased intracranial pressure. Against this, however, the argument was put forward that the lateral sinus is very often thrombosed without producing intracranial hypertension. The incidence of lateral sinus thrombosis is, however, so high in this condition, that it must have some important bearing on the problem of ætiology.



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## Otogenic Intracranial Hypertension

The next possibility to be considered is that of a thrombophlebitis beginning in the lateral sinus and extending by mural spread, but not necessarily with obstruction, up to the confluens sinuum, with extension to and thrombosis of the straight sinus. Resulting from this, engorgement of the choroid plexus may occur with subsequent overproduction of cerebrospinal fluid. This hypothesis is invalidated by the experimental observations of Bedford, who found it impossible in the dog or monkey by occlusion of the Great Vein of Galen to produce this hypertensive state.

The remaining possibility is, that, with or without obstruction of the lateral sinus, a mural thrombophlebitis may spread up into the superior longitudinal sinus and interfere with the absorption of cerebrospinal fluid through the arachnoid villi.

Two questions arise: can retrograde thrombophlebitis extend from the lateral sinus into the superior longitudinal sinus, and, secondly, is it not possible, that thrombosis of the superior longitudinal sinus may produce only the symptoms of intracranial hypertension and none others? Both these questions can be answered in the affirmative.

Nielsen and Courville report post-mortem observations in forty-three cases of lateral sinus thrombosis associated with otitis media. In six of these, the thrombosis had extended into the superior longitudinal sinus.

Another case is reported, in which the patient had a right-sided otitis media with thrombophlebitis of the lateral sinus. The patient developed all the signs and symptoms of intracranial hypertension. When the patient died from cardiac failure due to mitral stenosis, a post-mortem examination was carried out, and adherent thrombi were found extending from the right sigmoid sinus up to the torcular and thence along the superior longitudinal sinus to include its middle third. At no point could complete occlusion of any of the sinuses be demonstrated. Microscopic examination showed organization and canalization of the thrombi, which occupied about half the lumen of each sinus.

Freckner in one of his cases, suspecting lateral sinus thrombosis with retrograde spread, attempted a sinogram by the injection of perabrodil into the superior longitudinal sinus through a drill hole 5 cm. above the external occipital protuberance, following a technique employed by him since 1934. In this instance, although X-rays showed his canula to be correctly inserted into the sinus, he could obtain no blood. Subsequently an opaque injection into the internal carotid artery enabled him to obtain a phlebogram, which showed obstruction of the superior longitudinal sinus from approximately the level of the parietal eminence back to the torcular.

None of these cases showed any of the symptoms commonly associated in the mind of the clinician with this state, for instance, unilateral or bilateral convulsions, hemiplegia or diplegia.



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One comes to the conclusion, therefore, that the superior longitudinal sinus may be thrombosed without causing any serious cerebral damage. Probably the rate of onset is an important factor. If the progress from the lateral sinus is slow, an adequate collateral circulation may be established for the venous drainage of the cerebral cortex, especially if the anterior part of the sinus remains patent. Under such circumstances, the only clinical symptoms may be those due to failure of absorption of the cerebrospinal fluid with resulting hypertension intracranially.

If this syndrome is the result of lateral sinus thrombosis spreading upwards into the superior longitudinal sinus, then this complication ought to occur more frequently with infection of the right ear, than the left, for the superior longitudinal sinus is more frequently continuous with the right transverse sinus (6 : 4 or 7 : 3). It appears, therefore, that the chances of a right, as opposed to a left, lateral sinus thrombosis extending into the superior longitudinal sinus should be at a proportion of 6 : 4 or 7 : 3. This is actually the case, but the number of cases is too small to be of statistical value. Nevertheless, the figures are very suggestive.

It seems, therefore, that this clinical syndrome is symptomatic of superior longitudinal sinus thrombosis and does not necessarily result from an otitic condition. One case, for instance, was described, in which a sub-mandibular adenitis led to severe headaches, bilateral papillœdema, with normal cell count and normal protein content in the cerebrospinal fluid. It was thought that the adenitis had led to thrombosis of the jugular bulb with retrograde spread to the superior longitudinal sinus. An abscess in the vicinity of the tonsil is said to be an important factor in the production of a phlebitis of the internal jugular vein with possible retrograde spread.

Gardner also arrived at the conclusion that the syndrome is due to sinus thrombosis, but it appears to him that the increased pressure is due to intracranial venous engorgement, occasioned by the interference with the outflow of venous blood from the cranial cavity.

Additional evidence that interference with the return of venous blood from the cranial cavity can cause increased intracranial pressure is the occasional occurrence of choked discs after ligation of one, or even more so after ligation of both, internal jugular veins, or after accidental laceration and packing of the lateral sinus during mastoidectomy. Two of the cases to be described seem to support this theory, whereas the third does not exclude the possibility. A more detailed discussion on the possibility of intracranial venous engorgement is to follow.

### Prognosis

The prognosis is favourable, but it depends on how early treatment is given. All cases usually recover, but blindness is the great danger.



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The pressing need in this type of case is early diagnosis and, as neurological signs are scanty, this depends on the employment of ophthalmoscopy in all cases of ear disease that show any signs of intracranial disturbance. It is possible for the condition to be overlooked through neglect of ophthalmoscopy, which will lead to the omission of the required treatment.

Even if the headaches and vomiting are severe in the early stages, they are always better at the time when the patient is becoming blind. Treatment, therefore, should be entirely governed by the state of the eyes.

It may take a considerable time, months or even several years, till the intracranial balance, which depends on the development of an efficient collateral circulation is re-established.

**Treatment**

There are cases reported, in which vision returned to normal without treatment. In other cases, although the other symptoms clear up, vision may remain seriously impaired.

It is known that repeated lumbar puncture alone can lead to complete recovery. Dehydration therapy may be employed. The patient's consumption of liquids is reduced and ammonium chloride or a similar therapeutic agent administered. The treatment of the infective focus, in this case the ear, must be carried out. The most rational treatment consists of a decompression operation. After this, the patients usually recover. The important thing is to begin treatment at an early stage, so that the changes in the optic disc are not allowed to go on for too long. It is a sure method of preserving vision. It should be carried out in every case, in which papillœdema becomes at all severe, or in which there is any sign of lowering of visual acuity.

In a series of cases, reported by Davidoff and Dyke, thirteen were treated by sub-temporal and one by occipital decompression. The symptoms cleared up and there was improvement or arrest of the failing vision. Final disappearance of the papillœdema required from six months to several years.

In one case, reported by Cairns, in which the patient was practically blind, vision returned in a most dramatic manner after a decompression operation had been carried out. The operation, however, cannot be expected to effect a return of vision, after blindness has been present for some weeks.

In those cases where there is a doubt as to the diagnosis, ventriculography should be carried out, and if the ventricles are found to be of normal size, the treatment by repeated lumbar puncture can be initiated.



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# S. Horowitz

CASE I.

Miss A. McF., aged 17. This patient was admitted to the Infirmary on the 7.2.48.

*History.*

About one year prior to her admission to the Glasgow Royal Infirmary, this patient had a right-sided mastoidectomy performed in another hospital. No details of the operation were available. There was slight tenderness along the post-auricular scar, which, according to the patient, had been present since her operation in 1947.

Her left ear had been discharging at intervals since childhood, but two days prior to her admission, she had developed severe pain in the left ear, headaches, mainly in the temporal and occipital region, and had two " attacks of shivering."

*Aural Examination* (*L*).

There was purulent discharge. After mopping the meatus dry, a thickened drum, with a posterior perforation, was seen, through which pulsating discharge was noted. There was post-auricular œdema and tenderness over the mastoid process, particularly over the tip.

Rinne negative. Weber not lateralized. Whisper voice heard at a distance of 3 inches.

*Aural Examination* (*R*).

Tender post-auricular scar. No discharge. Healed cortical mastoidectomy.

Rinne negative. Whisper voice heard at a distance of 8 inches.

The patient was very pale and appeared gravely ill. The tongue was dry and coated. T. 101·8 ; P. 126 ; R. 26.

There was marked nuchal rigidity. Kernig was present on both sides, but more marked on the right side. The knee and ankle jerks were very brisk; the Babinski reflex was plantar on the left side and equivocal on the right side. Examination of the eyes, including ophthalmoscopic examination, revealed no abormality. The central nervous system showed no other abnormality. The cardiovascular, respiratory and genito-urinary systems were normal.

The diagnosis of chronic middle-ear suppuration with acute exacerbation of the left ear, complicated by meningitis, was made. After giving the patient a pre-operative dose of 1,000,000 units of penicillin, a radical mastoidectomy was performed.

*Operation Findings.*

The mastoid contained large cells, which extended to the tip and behind the lateral sinus. Some of the cells contained pus. The lateral sinus was exposed in a small area and the sinus wall found to be normal. The tegmen was eroded, and was removed. The middle fossa dura was congested, and one area was covered with granulations. The wound was left open and packed with a gauze strip, soaked in hypertonic saline solution.

Penicillin (500,000 units daily) and Sulphadiazine (2 gm. followed by 1 gm. four/hourly) were commenced.

The T.P.R. came down to normal within the next two days and remained so until 13.2.48. Progress was satisfactory ; wound clean and granulating.

Sulphadiazine administration was discontinued, but penicillin was continued.



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14.2.48. T. 100·8 ; P. 125 ; R. 22. No complaints.

15.2.48. Patient complains of pain in the right ear. Some tenderness on pressure over mastoid, particularly over scar line. 16.2.48. Tender red swellings over the radial third of the dorsa of both hands. Lymphangitis of left forearm. The left supratrochlear lymph gland enlarged and tender. No affection of axillary lymph glands. Penicillin was continued and sulphadiazine was commenced again.

The swellings were thought to be due to embolic spread from infected lateral sinus.

17.2.48. Ligation of left internal jugular vein. The lateral sinus was exposed. The wall of the sinus was thickened, but on opening the sinus, no thrombus was found. The proximal and distal ends of the exposed sinus were packed.

Blood culture: "Staphylococcus albus—coagulase negative—probably due to contamination."

18.2.48. T.P.R. normal. Patient feels much better. R.B.C. normal. W.B.C. 10,900/c.mm.

25.2.48. Swellings over both elbows and over lower lumbar vertebrae. Urinary output had fallen to 25 ounces in twenty-four hours. Sulphadiazine was discontinued.

T. 100·6 ; P. 120 ; R. 20.

26.2.48. Blister on left forearm. Redness and swelling over left eyelid, tender swellings over anterior aspect of both knee joints, both index fingers and left thumb.

27.2.48. Swelling and tenderness over glabellar region. Swellings over knee joints less marked. Rigor.

T. 101·8 ; P. 136 ; R. 26.

Because of increased tenderness over right mastoid area and the fact that the spread of emboli after ligation of the left internal jugular vein did not subside, it was decided to re-open the right mastoid.

*Operation Findings.*

Fibrous tissue and periosteum were removed and the mastoid antrum was re-opened. The antrum contained cholesteatoma. The tip was necrotic and was removed. There were large cells behind the lateral sinus, some of them containing beads of pus. The sinus plate was removed and the sinus wall exposed, which was thickened. The sinus was opened. A small thrombus was found adherent to the wall, which was removed. Free bleeding was obtained from both ends of the opened lateral sinus. Both ends of the exposed sinus were packed, thus controlling the bleeding. Wound was left open and packed.

28.2.48. T.P.R. normal. One pint of blood was transfused to the patient.

29.2.48. Patient does not complain. All swellings have disappeared.

12.3.48. Patient well, and remained so for the next ten days.

13.3.48. Patient complains of intermittent diplopia. Very irritable and slightly confused. Some nuchal rigidity. Tender swellings reappearing on forearms and knees.

*Eye Examination.*

Fundi normal. Some inco-ordination of the external eye muscles but no paresis. Visual acuity and fields normal.



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Blood culture : sterile.

Mastoid wound on left side very clean and closing in. It was decided to explore the mastoid wound on the right side again because of the meningeal signs and to ligate the right internal jugular vein, because of the possibility that the embolic spread may have come from that side.

L.P. was performed : Cerebrospinal fluid normal.

*14.3.48. Operation.*

Middle and posterior fossa exposed. Dura was covered with granulations. Right internal jugular and common facial veins were ligated. One pint of blood and one pint of 10 per cent. glucose-saline were given.

16.3.48. Patient complains of headache, nausea, blurred vision and intermittent diplopia.

Ophthalmoscopic Examination : gross papillœdema of both eyes, greater however, on right side (5 diopters). Small hæmorrhages and exudates on both fundi. Paresis of right external rectus muscle.

The diagnosis of otogenic intracranial hypertension due to ligation of both internal jugular veins was made.

20.3.48. Ophthalmoscopic findings: no change since the previous examination.

24.3.48. T. 103° F. ; P. 130 ; R. 26. Wound on right side clean and granulating well. Wound on left side almost healed.

26.3.48. Tender swellings over forehead, and fingers of left hand.

28.3.48. More tender swellings on forearm. Examination of heart : normal size, systolic bruits at aortic and pulmonic areas. The diagnosis of endocarditis was made. Vision is becoming more blurred.

Several lumbar punctures were carried out on the seven following days. The pressure was increased (up to 380 mm. $H_2O$), but the cell, chloride and protein estimations were normal. No further complaint of headaches.

6.4.48. Patient was transferred to the Neuro-surgical Unit.

7.4.48. Ventriculography was carried out. The cerebrospinal fluid was normal. Ventriculograms showed no abnormality, thus the possibility of a unilateral space-occupying lesion was excluded.

8.4.48. Lumbar puncture : xanthocromic fluid, otherwise normal.

10.4.48. Lumbar puncture : cerebrospinal fluid normal. Pressure 400 mm. $H_2O$.

12.4.48. Systolic bruits at the aortic and pulmonic areas very marked. Painful swellings over hands and chest wall.

Blood culture and sub-culture were sterile.

Urine : A few stellar phosphates observed. Culture gave a very scanty growth of coliform bacilli.

Penicillin (500,000 units daily) being given.

13.4.48. Lumbar puncture : cerebrospinal fluid normal.

14.4.48. Lumbar puncture : cerebrospinal fluid normal. Fluid xanthochromic.

*Eye Examination.*

Ocular movements full. Some jerky nystagmoid movements, which are not maintained.



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Ophthalmoscopic Examination : discs hyperæmic and swollen.

Margins almost completely obliterated. Several hæmorrhages around the discs. In the papulo-macular area of both eyes are several small punctate spots of exudate, which have not yet taken on the shape of a macular star.

Visual fields full.

Ear examination : left ear healed and dry. Right ear slightly moist.

15.4.48. Lumbar puncture : cerebrospinal fluid normal.

16.4.48. Lumbar puncture : cerebrospinal fluid normal. Pressure 420 mm. $H_2O$. Fluid xanthochromic.

17.4.48. Painful swellings on back of hands have disappeared. Feeling well. On daily L.P. Pressure still increased. Pencillin continued.

19.4.48. L.P. : cerebrospinal fluid normal. Progress satisfactory.

7.5.48. Ophthalmoscopic Examination : nasal margins, upper and lower poles of discs still fuzzy. There is kinking of both arteries and veins close to the discs. There is no apparent fresh hæmorrhage and no visible exudate.

10.5.48. There is no measurable swelling of the discs now.

11.5.48. There is still a slight discharge from the right ear. Patient well.

17.5.48. There is a painful swelling on the middle of the forehead. It is soft, but not fluctuant and just merges into the neighbouring tissue. No discolouration. Slight tenderness over spleen, but no enlargement. Swelling of hands still present.

18.5.48. R.B.C. 4,180,000. Blood culture sterile.

Hb. 95 per cent. Sahli = 12·3 gm. per cent. C.I. = 1·1.

21.5.48. X-ray of hands : " no joint or bone abnormality."

X-ray of chest : " the heart shadow is normally placed and not enlarged. There is some prominence of the pulmonary conus, but no abnormality otherwise. No pulmonary disease."

22.5.48. W.B.C. 7,400.

29.5.48. Swellings on forehead and dorsum of hands have now subsided and she feels much better.

B.S.R. 4 mm./hr.

16.6.48. Right ear almost dry.

21.6.48. Discs flat. General condition good. T.P.R. normal.

26.6.48. L.P. : cerebrospinal fluid normal. Pressure raised only slightly.

10.7.48. Patient transferred to Medical Ward for treatment of the heart condition.

The skin lesions were considered to be the result of emboli thrown into the general circulation from one or the other, or possibly both, internal jugular veins. In the same manner, the endorcard must have become the victim of embolic infection. Once the internal jugular vein had been ligated, no further spread seemed to have been possible from this source, and embolic spread from the endocardial vegetations must be assumed.

Case II.

Mrs. J. McPh., aged 27 years.

This patient was admitted to Stobhill Hospital, Glasgow, on 6.6.46.

The patient's left ear had been discharging at intervals for the last four years.



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# S. Horowitz

*Aural Examination* (*L*).

There was foul smelling pus in the meatus and after syringing and mopping the ear dry a thickened tympanic membrane with attic perforation was seen.

Rinne negative. Weber was lateralized to left.

Whisper voice was heard at a distance of 4 inches.

Examination of the eyes, C.N.S. and the other systems revealed no abnormality.

The diagnosis of chronic mastoiditis with attic perforation was made and operation was decided upon.

7.6.46. *Operation Findings.*

Mastoid antrum contained pus and cholesteatomatous masses. There was caries throughout. Sinus plate was necrotic and sinus wall was covered with cholesteatoma.

The lateral sinus was situated rather anteriorly. During the operation the sinus was opened accidentally. The bleeding from it was controlled by packing the sinus proximally and distally. The wound was left open and packing was inserted. The radical type of mastoidectomy was performed.

16.6.46. Patient progressing well, but complains of slight headache.

19.6.46. T.P.R. normal. Wound granulating well.

Occasional headache for the last few days.

21.6.46. Mastoid wound stitched.

23.6.46. Patient complains of diplopia and headaches.

*Eye Examination.*

Paresis of right external rectus muscle. Nystagmus present in all directions.

Ophthalmoscopic examination : bilateral papillœdema present, more severe in right eye (4 diopters). Small hæmorrhages.

27.6.46. Lumbar puncture was carried out. Cerebrospinal fluid clear. All constituents within normal limits, but the pressure is greatly increased (480 mm. $H_2O$). No organisms. W.B.C. 11,000/c.mm.

The diagnosis of intracranial hypertension was made. Daily lumbar puncture to be performed.

28.6.46. Lumbar puncture. Pressure 520 mm. $H_2O$. Cerebrospinal fluid normal.

29.6.46. Daily lumbar puncture continued.

15.7.46. Diplopia has disappeared. Cerebrospinal fluid has been normal throughout. Pressure still very high. Repeated ophthalmoscopic examinations showed no change.

19.7.46. Pain in right lower extremity, radiating down from hip to heel.

20.7.46. Similar pain in left lower extremity.

The pain suggests that arachnoid reaction around the cauda equina is taking place. Lumbar punctures were discontinued.

24.7.46. Patient was transferred to the Neuro-surgical Unit.

26.7.46. X-ray of skull negative. Headaches.

30.7.46. Lumbar puncture. Rather xanthochromic cerebrospinal fluid. Increased pressure. Protein 50 mg. per cent. ; cells 8/c.mm. ; Lange 1111000000 ; W.R. negative.



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*Eye Examination.*

Visual acuity normal. Pupils equal and react to light directly and consensually. Fields full on confrontation test. No diplopia elicited. Ocular movements full. Nystagmoid movements on looking to the right.

Ophthalmoscopic examination: discs of both eyes are swollen, the right more so than the left. Veins are full. No exudates or hæmorrhages.

31.7.46. In order to exclude a unilateral space-occupying lesion, it was decided to carry out a ventriculography.

Pressure was raised and cerebrospinal fluid was normal.

Ventriculograms: "Both lateral ventricles seem to be of normal size. There is a shift to the right which suggests a space-occupying lesion in the left hemisphere. There is not enough evidence on the present film to locate it accurately."

1.8.46. W.R. in blood and cerebrospinal fluid negative.

6.8.46. Findings on ophthalmoscopic examination unchanged. Because of the uncertainty of the diagnosis, it was decided to carry out a cysterno-encephalography.

*Cysterno-Encephalography.*

60 c.c. of cerebrospinal fluid were withdrawn and replaced by air. Encephalogram: no hydrocephalus, but the shift to the right is now scarcely perceptible.

The space-occupying lesion was therefore excluded.

To have daily lumbar puncture.

7.8.46. Xanthochromic fluid under pressure of 250 mm. $H_2O$ withdrawn on lumbar puncture.

18.8.46. Findings on eye examination unchanged. Had until to-day a daily lumbar puncture. Pressure has fallen to 255 mm. $H_2O$. To have lumbar puncture every second day.

20.8.46. Lumbar puncture: cerebrospinal fluid normal. Pressure 265 mm. $H_2O$.

26.8.46. The intracranial hypertension seems to have responded to lumbar puncture, which was discontinued five days ago. Patient is allowed up for a short time during the day.

27.6.46. Lumbar puncture and manometry. Clear fluid, pressure fluctuating from 210-220 mm. $H_2O$, cerebrospinal fluid normal.

4.9.46. Lumbar puncture: clear fluid. Pressure 250 mm. $H_2O$. Ophthalmoscopic examination: Papillœdema still present. Visual fields normal. Visual acuity slightly impaired.

6.9.46. Lumbar puncture: clear fluid: Pressure 300 mm. $H_2O$: cerebrospinal fluid normal. On daily lumbar puncture.

18.9.46. Lumbar puncture: pressure 310 mm. $H_2O$. Definite impairment of visual acuity. Papillœdema persisting. Complains of pain in her limbs, suggesting that arachnoid reaction round the cauda equina was taking place.

A subtemporal decompression was considered, but it was decided to wait another few days and await developments.

20.9.46. Eye examination showed no change. Intermittent pain both lower extremities.

24.9.46. Under avertin, endotracheal gas and oxygen, and local novocaine



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anæsthesia, a right subtemporal decompression was carried out. The dura was tense, but after some cerebrospinal fluid had escaped, the spongy, soft brain sank into the wound. The ventricle was tapped behind. It did not appear to be enlarged to any extent.

3.10.46. Patient much improved. Papillœdema subsiding.

7.10.46. Patient is well. Cupping of the discs has returned. Visual acuity is normal.

8.10.46. Cysternal manometry: cerebrospinal fluid normal; pressure 160 mm. $H_2O$.

9.10.46. Eyes normal. Her decompression was soft.

Discharged.

19.10.48. After two years, the patient was asked to return for re-examination.

Ear is healed, but she complains of occasional noises in it. Eyes are normal and the decompression is soft.

CASE III.

Miss E. B., aged 14. This patient was admitted to the Royal Infirmary on 13.11.47.

*History.*

Discharging ear (right) at the age of 3. No further trouble until four days prior to her admission, when she complained of earache on the right side and some discharge from the right ear.

*Aural Examination.*

Tympanic membrane: pinhead-sized perforation about the centre. Posterior part of drum bulging and red, rest of drum injected. Slight tenderness over mastoid antrum and tip of mastoid process.

Rinne negative. Weber lateralized to right side. Forced whisper heard at a distance of 2 inches.

The girl was pale and ill-looking. Her tongue was coated. T. 101°F.; P. 126; R. 26.

No abnormality was found on examination of the C.N.S. The diagnosis of chronic otitis media with acute exacerbation was made on the day of admission. Myringotomy was performed.

14.11.47 to 16.11.47, T.P.R. was normal.

Free discharge from the middle ear.

Tenderness over the mastoid process slightly diminished.

Patient's general condition improving. Patient on four-hourly douching with boracic lotion.

17.11.47. T. 101·2°; P. 130; R. 25.

18.11.47. T.P.R. normal.

19.11.47. T. 102·4°; P. 135; R. 30.

There was now increased tenderness over the mastoid process and a profuse discharge of creamy pus.

In view of those findings and the suspicion of lateral sinus thrombosis aroused by the intermittent temperature, mastoid exploration was decided upon.



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*Findings at Operation.*

Cellular mastoid. There was pus under tension in the antrum and it contained debris and carious bone. Extensive exenteration of the cells was carried out. The sinus plate was eroded and partly necrotic and was removed for a distance of about one inch.

The sinus wall was covered with granulations. The radical type of mastoidectomy was carried out. The wound was left open and a fine tube was stitched into the wound edge, through which penicillin was injected at four-hourly intervals, in order to irrigate the mastoid cavity. The patient was also put on systemic penicillin (250,000 units morning and evening).

21.11.47. T. 101·8°; P. 132; R. 26. First dressings carried out. Sinus wall discoloured with greyish-white necrotic patches on it. Temperature came down to 98·6°, in the evening.

22.11.47. Temperature rose to 104·6°. The diagnosis of lateral sinus thrombosis was made in view of the appearance of the sinus wall and the intermittent temperature.

*Operation.*

Sinus wall was slit. There was a small mural thrombus, which was removed and free bleeding was obtained from both the proximal and distal ends of the exposed sinus. Upper and lower end of the sinus were packed off. Internal jugular vein and common facial vein were ligated.

23.11.47. T. 99·4°; P. 121; R. 24.

24.11.47. T.P.R. normal.

Patient's condition improving daily.

6.12.47. Generalized headache, accentuated by severe attacks of paroxysmal pain in the temporo-parietal and retro-orbital region. Diplopia on looking straight forward and to the right.

*Examination of Eyes.*

Paresis of right VIth nerve. Visual acuity and visual fields normal. Fundus of right eye shows gross papillœdema (5 diopters). Fundus of left eye normal. Nystagmus to left. Slight photophobia. Rest of C.N.S. normal.

Provisional diagnosis of extradural abscess was made.

7.12.47. Findings are the same as on previous day, only the fundus of the left eye now shows even more papillœdema than the right and there are also small hæmorrhages present.

The diagnosis of brain abscess was now in doubt; because of uncertainty of the diagnosis ventriculography was carried out.

*Findings at Ventriculography.*

Cerebrospinal fluid clear and under enormous pressure (400 mm. $H_2O$). Biochemical and bacteriological examination of the cerebrospinal fluid revealed no abnormality.

Ventriculogram: ventricles were of normal width, no dislocation or local deformation of lateral or third ventricles.

Because of the normal cytology and the normal protein content of the cerebrospinal fluid and the normal appearance of the ventriculogram, the diagnosis of brain abscess had to be abandoned and the diagnosis of Gradenigo's Syndrome, complicated by otogenic intracranial hypertension, was made.



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8.12.47. It was decided to carry out daily lumbar punctures, in order to reduce the intracranial pressure. Mastoid wound clean.

11.12.47. Headache less severe. Papillœdema of both eyes persisting. VIth nerve palsy persists.

Lumbar puncture continued.

14.12.47. No change. Lumbar puncture continued.

21.12.47. Headaches still present, also VIth nerve palsy.

22.12.47. Mastoid wound clean and granulating well.

25.12.47. Area of skin where the lumbar punctures were carried out slightly inflamed and tender. Lumbar puncture therefore had to be discontinued.

Because of the persisting papillœdema, a subtemporal decompression was considered.

As her visual acuity and visual fields were unimpaired, it was decided to wait and to observe further developments carefully.

29.12.47. Headaches almost absent. VIth nerve palsy improving. Papillœdema of both eyes as before.

4.1.48. No headaches for last three days. VIth nerve palsy greatly improved. No change in papillœdema. Visual acuity and fields normal. Mastoid wound healed.

10.1.48. No headaches. Further improvement in VIth nerve palsy; sees double only occasionally. Papillœdema decreasing, cupping of discs returning. Hæmorrhages in left fundus disappeared.

18.1.48. Papillœdema still regressing.

27.1.48. Eye movements almost normal. Further improvement of papillœdema.

2.2.48. Still slight papillœdema. VIth nerve palsy completely disappeared. Discharged.

### Discussion and Comment

All the phenomena encountered in otogenic intracranial hypertension can be explained as being due to interference with the outflow of venous blood from the cranial cavity, without having to postulate increased production of cerebrospinal fluid due to engorgement of the choroid plexus, or diminished absorption of cerebrospinal fluid by blockage of the arachnoid villi, as set out previously.

It is said that a larger amount of cerebrospinal fluid is obtained on lumbar puncture, before the pressure returns to normal. This, however, does not necessarily mean that there is an increased quantity of cerebrospinal fluid. If one considers that the venous out-flow is interfered with and the cerebral veins are engorged, it is clear that the fluid removed is immediately replaced by an increased of venous blood within the already engorged cerebral veins. This engorgement forces a larger quantity of fluid through the needle than would be the case were the cerebral veins not engorged. A similar factor is operative in Queckenstedt's test, where compression of the jugular veins leads to an increase in pressure and rate of flow.



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Another frequent observation is that on trephination the exposed portion of the arachnoid membrane is found to be distended with fluid. This has led to the assumption that external hydrocephalus was present. Encephalograms, however, which are essentially normal in these cases makes this assumption unlikely. It seems that the release of pressure at the site of operation produces local accumulation of fluid in the subarachnoid space of this area. This will take place, even when the amount of cerebrospinal fluid is normal in quantity, provided the fluid is able to circulate freely. The fluid will then follow the subarachnoid channels to the area of released tension, causing local distension of the subarachnoid spaces in this area.

Another fact is that on ventricular puncture a copious flow of cerebrospinal fluid is obtained. This has led to the belief that hydrocephalus is present. This belief is invalidated by encephalograms, which show the ventricles to be of normal size, or sometimes even smaller. If one considers, however, that the circulation of cerebrospinal fluid is free, it seems likely that the engorged cerebral veins force a greater amount of fluid to the point of least resistance. Therefore, towards the end of the drainage, the fluid that is escaping from the canula is really subarachnoid fluid, which has been forced into the ventricles as a result of the drainage. In these cases one cannot estimate the size of the ventricles from the amount of fluid obtained by ventricular puncture.

Occasionally, the fluid obtained on lumbar puncture is xanthochromic in appearance, which again speaks for vascular obstruction, with extravasation of blood into the arachnoid space, a phenomenon well known in Froin's syndrome.

If, as is not infrequently the case, there are anomalies in the veins of the skull and one of the lateral sinuses constitutes the chief emissary for the intracranial circulation, or the jugular vein on one side should be obstructed and the other be narrow or non-existent, or if both jugular veins—anatomically normal—should require ligation, then one can justify the opinion that it is possible for a thrombosis of this sinus alone to produce the picture of increased intracranial pressure.

In the three cases described, the symptoms appeared after ligation of one or both internal jugular veins. In the first case no symptoms of intracranial hypertension were present after ligation of the jugular vein on one side. The symptoms only appeared after ligation of the contralateral vein. It seems, therefore, that the contralateral internal jugular vein was capable of emptying the venous blood from the cranial cavity before its ligation. In the second case, the lateral sinus and internal jugular vein on the affected side may have been the main channel of drainage for the cranial cavity. The first two cases described showed the cerebrospinal fluid to be xanthochromic.

In the third case, which depicts also Gradenigo's Syndrome, there was



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probably a primary obstruction to the outflow of blood after jugular ligation. As in any other ligation of a blood vessel, a thrombus developed at that site, and the thrombus had the tendency to spread. The syndrome described by Gradenigo, was probably brought about by spread of the thrombus along the inferior petrosal sinus, which connects the lateral sinus to the cavernous sinus at the side of entry of the VIth nerve into the wall of this sinus. This will lead to congestion, œdema and possibly infection in the region of the petrous apex, thus affecting the VIth nerve and the Gasserian ganglion and producing the characteristic symptoms. In the first two cases described, there must also have been thrombosis at the site of ligation with consequent spread. One does not, however, require to postulate that this thrombotic spread must necessarily have led to either engorgement of the choroid plexus or to obstruction of the arachnoid villi with consequent increased intracranial pressure.

## Conclusion

Although at the present state of our knowledge the ætiology of otogenic intracranial hypertension and its mechanism of production must remain problematic, the three cases which have been described would seem to support the theory that this syndrome is most likely brought about by obstruction to the outflow of venous blood from the cranial cavity.

Most authors agree that the syndrome of increased intracranial pressure without localizing signs, occurring after an attack of otitis media, is usually due to sinus thrombosis. Some, however, are not convinced that the thrombus must necessarily extend into the sagittal sinus in order to produce increased intracranial pressure.

Since the total volume of the intracranial fluid is usually not increased and since encephalograms in these cases are essentially normal, there is obviously no hydrocephalus. One is inclined to attribute the intracranial hypertension simply to engorgement of the intracranial veins.

Although it is unlikely that every case of otogenic intracranial hypertension can be explained as the result of intracranial venous engorgement I think there are certain cases, possibly the majority of them, in which venous obstruction is the ætiological factor. It cannot be denied, however, that post-mortem examination in a number of these cases has demonstrated blockage of the arachnoid villi. In a number of cases, it is fair to assume that a combination of both factors must be operative in the production of otogenic intracranial hypertension.

Recovery from this state occurs when, by means of recanalization or the development of a collateral supply, the return of venous blood from the cranial cavity is once more adequate.

## Summary

A review is given on the syndrome of increased intracranial pressure without localizing signs.



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# Otogenic Intracranial Hypertension

It is suggested that the name "otogenic intracranial hypertension" is more adequate for this condition.

The Clinical Manifestations, Differential Diagnosis, Ætiology, Pathology, Prognosis and Treatment of this condition are discussed.

The value of a decompression operation is stressed, it being a sure method of preserving vision. It should be carried out in every case in which the papillœdema becomes at all severe, and in which there is any sign of lowering of visual acuity.

Three cases are described :—

The first had both internal jugular veins ligated, because of emboli producing skin lesions. The endocardium had also become infected by the emboli.

The second case was one of traumatic sinus thrombosis with jugular ligation.

The third case showed in addition to the intracranial hypertension, Gradenigo's Syndrome.

The theory of intracranial venous engorgement is discussed and the phenomena encountered in otogenic intracranial hypertension explained in the light of that hypothesis.

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