Long-term follow-up of nine cases of ventriculocisternostomy for non-neoplastic aqueductal occlusion

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In 1938 a new surgical procedure (ventriculocisternostomy) was introduced by Torkildsen. Three patients with this condition were presented for whom this operation produced complete relief of increased intracranial pressure symptoms. Torkildsen later published additional papers and eventually a monograph (1947) on his experiences with this procedure. He concluded that ventriculocisternostomy is indicated in patients with inoperable obstruction of the foramina of Monro, the third ventricle, or the Sylvian aqueduct. He emphasized the fact that the procedure itself was basically palliative, relieving the pressure symptoms only, and that the ultimate fate of the patient depended on the nature of the pathology producing the internal hydrocephalus.

The Torkildsen procedure became a standard neurosurgical operation. J. C. White and one of the authors (J.J.M.) published the first results with this operation in the United States in 1942. Their patient had a benign aqueductal stenosis which should be permanently benefited by diversion of the ventricular fluid. Surprisingly, Torkildsen's own results in patients with this affliction did not come up to expectation.

MATERIAL

Our paper presents follow-up data for a period of eight to 27 years on nine patients with aqueductal stenosis, treated according to Torkildsen, including two patients reported in 1942.

These nine patients comprise a comparatively small group of more than 60 patients who were subjected to this operation since 1940 at the Massachusetts General Hospital. There were five males and four females, ranging in ages from 7 weeks to 56 years, their symptoms lasting from two weeks to seven years. Headache was the most prominent symptom. There were no lateralizing neurological signs. Intraventricular pressures were elevated in excess of 250 mm. of cerebrospinal fluid in all nine patients, and papilloedema was present in seven. The diagnosis was arrived at by contrast radiological

study. Following surgery, cerebrospinal fluid dynamics became normal with complete remission of symptoms. Details are summarized in the Table.

Characteristic x-ray findings were symmetrical enlargement of the lateral and third ventricles and non-filling of the aqueduct and fourth ventricle. The fourth ventricle was of normal size and in the normal position. With air brought in from below or intraventricular installation of Pantopaque, the aqueductal stenosis can be well demonstrated.

DISCUSSION

Torkildsen in his monograph (1947) commented: 'When in 1939 I published the results of my first four cases, I had the impression that the operation was not associated with noteworthy difficulties. When I published my first seven cases in 1941 I was also under the impression that the operation was combined with only minor difficulties, but further experience has shown that there are complications which ought to be mentioned.'

In his series then of 33 patients, eight had accumulated collections of cerebrospinal fluid under the scalp and, in half, the spinal fluid broke through the skin producing a fistula. Torkildsen felt this was due to the temporary persistence of increased intracranial pressure and, with repetitive lumbar taps and secondary suturing in one instance, the fistula healed subsequently without an instance of meningitis. In the larger series of Gröschel and Marguth (1963) of 155 cases, and of Lorenz (1966) of 142 cases, cerebrospinal fluid fistulas occurred in 23 and 26 patients, respectively. Lorenz commented that, if meningitis developed, in his experience it was resistant to therapy because the drain acted as a foreign body and maintained the infection. We have seen meningitis cleared on sulphonamide and penicillin therapy with a Torkildsen shunt in place. Thus it seems that it is not necessary in every instance to remove the shunt if meningitis develops.

From the experiences of these surgeons as well as those of our own, it is recommended that a tight closure of the dura, meticulous approximation of

TABLE

SUMMARY OF PRESENT SERIES

Case No.	Age-Sex (yr.)	Duration of Illness	Dominant Symptoms	Neurological Deficits	Air Study	Operations ¹	Lesion	Symptom Remission (yr.)	Outcome
5	F	2 wk.	Headache, 2 wk. emesis, 1 wk. stupor, 3 days	Normal, except for papilloedema	Ventriculogram enlarged lateral and third ventricle; no filling of aqueduct or fourth ventricle	Negative posterior fossa exploration and T.	Aqueductal atresia	Yes—11	Well, making good grades in school two and a half years later and has remained asymptomatic to date
7	M	2 wk.	Headache, 2 wk. emesis, 3 days	Truncal ataxia, papilloedema	Same as case 1	Negative posterior fossa exploration and T.	Probable aqueductal atresia	Yes—9	Well, to date
50	F	2 yr.	Headache and C.S.F. rhinorrhoea, 2 yr. emesis, ataxia, and mental confusion, 2 days	Stupor, truncal ataxia, bilateral extensor plantar reflex; no papilloedema	Pneumoencephalogram T. and ventriculogram demonstrated marked lateral and third ventricle enlargement, normal fourth ventricle, and occlusion of aqueduct	T.	Aqueductal atresia	Yes—1+	Loss of headache and ataxia after T., but intermittently persistent rhinorrhoea led to ventriculo-atrial shunt with low pressure valve 1 yr. later. At that time T. was patent and functional. Rhinorrhoea persisted and Pantopaque V. documented leak into petromastoid bone from temporal lobe. Leak ceased after Pantopaque study and has not recurred to date.
7 wk.	M	7 wk.	Abnormally enlarging head	No lateralizing defects; no papilloedema	Ventriculogram enlarged lateral and third ventricles; no filling of aqueduct and fourth ventricle	T.	Aqueductal atresia	Yes—8	Asymptomatic to date, presently attending teachers' college
39	F	1 mth.	Headache, blurred vision	Normal, except for papilloedema	Same as case 3	T.	Aqueductal atresia	Yes—10	Excellent recovery, asymptomatic to date
21	M	10 mth.	Headache, decreased vision	Normal, except for papilloedema	Same as case 1	Negative posterior fossa exploration and T.	Aqueductal atresia	Yes—27	Well, without neurological symptoms in subsequent 27 years to date
9	M	7 yr.	Mental retardation, headache, 1 yr.	Bilateral dysmetria, truncal ataxia, papilloedema	Same as case 1	T.	Aqueductal atresia	Yes—18	Teachers noted marked improvement in school work eight months later. Improvement in cortical function tests over the next several years but dull normal range. Unfortunate lesion not diagnosed sooner
51	M	6 mth.	Headache, ataxia	Truncal ataxia, papilloedema	Same as case 1	Negative posterior fossa exploration and T.	Aqueductal atresia	Yes—18	Well, without return of neurological symptoms in 18 years to date
56	F	10 mth.	Headache, decreased vision, ataxia, dysphagia	Normal, except for truncal ataxia and papilloedema	Same as case 1	Negative posterior fossa exploration and T.	Aqueductal atresia	Yes—12	Readmitted 12 years later with headache and left cerebral deficits. Lumbar pressure 150. Lung carcinoma diagnosed with left cerebral metastasis. Radiation given but slow decline until death 3 months later. No necropsy obtained

¹T. = Torkildsen shunt

the suboccipital incisions, and if pressures remain transiently elevated, frequent postoperative lumbar punctures and elevation of the head are useful measures in preventing a cerebrospinal fluid fistula. Gröschel and Marguth also suggest that a paramedian suboccipital incision, especially in children, is superior to a midline incision in preventing this complication. Other uncommon complications seen in the combined experience of these surgeons include epidural, subdural, and intracerebral haemorrhage, as well as intraventricular bleeding. These complaints are probably attributable to operative technique in some instances, as well as to the sudden relief of intracranial hypertension in others.

There were no operative complications in our series, and the shunt, utilizing a red rubber catheter, has remained functional in all patients, in one for 27 years. Torkildsen removed a red rubber catheter seven years after its insertion in one patient. It was patent without evidence of surrounding inflammatory reaction or deterioration. Thus, although plastic and silicone-coated tubes are available today, there is no indication that the standard red rubber catheter is not satisfactory.

Torkildsen lost four out of 13 patients within three days of surgery, in two instances due to 'malignant hyperthermia'. Late complications accounted for three more deaths, at 44 and 120 days and four years after surgery.

No late complications were observed in our patients. One patient (no. 4), although relieved of pressure symptoms, continued to experience intermittent cerebrospinal fluid rhinorrhoea. Even though intraventricular and lumbar pressures were normal,

a ventriculo-atrial shunt with a low pressure valve was done in hope of curing the rhinorrhoea. This was not successful. Subsequently the course of the leak was documented by Pantopaque ventriculography. Fluid traversed a fistulous path from the right temporal horn into the petromastoid bone, and by the eustachian tube into the nasopharynx. Rhinorrhoea ceased spontaneously after the Pantopaque study and has not recurred to date.

Our experiences indicate that the results of the Torkildsen shunt in non-neoplastic aqueductal stenosis should be gratifying. This is true for the permanent relief of pressure symptoms, as well as the absence of significant complications, especially when compared to newer shunt procedures which often require revisions.

SUMMARY

Nine cases of benign aqueductal stenosis treated with the Torkildsen operation of ventriculo-cisternostomy are presented. Pertinent clinical findings, long-term follow-up periods of eight to 27 years, and results of others with this operation are discussed.

REFERENCES

- Gröschel, D., and Marguth, F. (1963). Ergebnisse der Ventrikulozisternostomie nach Torkildsen. *Zbl. Neurochir.*, 23, 262-281.
- Lorenz, R. (1966). Komplikationen nach Torkildsen-Drainagen. *Acta neurochir. (Wien)*, 14, 246-253.
- Torkildsen, A. (1938). A new palliative operation in cases of inoperable occlusion of the Sylvian aqueduct; a preliminary report. *Forh. norske med. Selsk.*, pp. 102-108.
- (1947). *Ventriculocisternostomy*, Johan Grundt Tanum, Oslo, 240.
- White, J. C. and Michelsen, J. J. (1942). Treatment of obstructive hydrocephalus in adults. *Surg. Gynec. Obstet.*, 74, 99-109.