Section of Neurology

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Stenosis of the Aqueduct of Sylvius

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FOR nearly a century it has been known that hydrocephalus is occasionally associated with occlusion of the aqueduct of Sylvius. But it is only since we have had a more complete knowledge of the circulation of the cerebrospinal fluid that the strategic importance of the aqueduct has been appreciated. Here is a small tube, in the adult human brain some 20 mm. long by 2–3 mm. in diameter, which is asked to convey all the cerebrospinal fluid secreted in the lateral and 3rd ventricles down to the outlets in the 4th ventricle. A minute lesion which in other parts of the brain might cause no trouble at all may narrow the aqueduct so that the flow of the cerebrospinal fluid is obstructed, and hydrocephalus results.

In general, such lesions may be either neoplastic or non-neoplastic. The first group includes tumours of the quadrigeminal plate and other parts of the mid-brain which may compress the aqueduct and produce interesting neurological abnormalities ; but, as they are generally infiltrating tumours in an inaccessible situation, treatment at best affords only temporary relief of symptoms, and if we could be sure of the diagnosis, it may be asked whether this treatment should ever be surgical. Nevertheless, this group deserves consideration in the differential diagnosis from the more benign lesions which may produce a very similar clinical state, but which have considerably better prospects of cure.

In the non-neoplastic group, the commonest pathological process seems to have been described first by Spiller in 1902. A boy of 19 who had suffered from periodic headache since childhood was admitted to hospital with symptoms of increased intracranial pressure and signs which pointed to the diagnosis of cerebellar tumour. He died suddenly, and examination of the brain showed that the aqueduct was almost completely obliterated, but to the naked eye the structure of the mid-brain otherwise appeared normal and no evidence of tumour was seen. Histological examination showed that the stenosis had been effected by a proliferation of the normal subependymal glia around the aqueduct. In 1936, Stookey and Scarff collected from the literature 16 cases of non-neoplastic stenosis ; in the 12 of these in which an adequate histological examination was made, this proliferation of subependymal glia appeared to be the underlying pathology. These subependymal glial cells (and especially their fibrillar elements) seem to proliferate, and may cause a true stricture of the aqueduct from without ; or the proliferation may break through the ependyma to form tufts within the lumen which may be valve-like or membranous ; or there may be a more or less regular spread of the glial processes within the aqueduct, encroaching on its lumen from within. In yet another variety, the aqueduct may be split up into a number of tiny channels which may be almost invisible to the naked

eye ; the aqueduct becomes a swamp through which the cerebrospinal fluid seeps, but with considerable difficulty.

This paper concerns 18 cases of non-neoplastic stenosis of the aqueduct which have been observed in Professor Cairns' Clinic. This diagnosis cannot be established until the mid-brain is examined histologically after death, and unfortunately six of the cases in this series have been so confirmed. But these six cases, and a study of other recorded verified cases, have given sufficient information about the clinical state and the ventriculographic appearances to warrant the diagnosis in the unconfirmed cases.

Juvenile Type

The clinical picture is not uniform, but the following case is fairly characteristic :—

G. O'S, aged 10, was admitted to the Radcliffe Infirmary on August 25, 1939, referred by Mr. John Kelly. He was born at full time, and apart from having a big appetite and being very plump, was, according to his parents, a perfectly normal child until three years before admission, when he became clumsy with his hands, and very apt to spill things at the table. His hands were shaky, and about eight months before admission this tremor of the hands became so marked that he was treated in hospital for two months as a case of chorea. He did not improve, however, and one month before admission he began to suffer from severe headache and occasional bouts of vomiting. It was these symptoms which led to his admission to the Radcliffe Infirmary.

On examination he was a fat child, 4 ft. 10 in. tall, weighing 7 st. 2 lb. He had a large head, 57 cm. circumference, with a cracked-pot note on percussion, and a blowing systolic murmur was audible all over the skull. He was cheerful and co-operative, but had occasional bouts of headache which seemed to distress him considerably. There was no dysarthria. He had bilateral papilloedema, 3 D, but the visual acuity was normal and there was no defect in the visual fields. The pupils and external ocular movements were perfectly normal, and the functions subserved by the other cranial nerves were likewise normal. In the limbs there was no definite dystonia or weakness, but there was a vibratory tremor of the outstretched limbs, rather coarser than that seen in hyperthyroidism, and exaggerated on purposive movements. He was a little unsteady on his feet, but was able to walk without support. There was no sensory disturbance. X-rays of the skull showed general enlargement, convolutional thinning, and separation of the sutures ; erosion of the sella was so marked as to raise the question of the lesion being in the immediate neighbourhood of the sella. The cerebrospinal fluid was normal.

The diagnosis rested between stricture of the aqueduct, suprasellar tumour, and a high vermis tumour. A ventriculogram showed gross internal hydrocephalus, a dilated 3rd ventricle, and the shadow of the aqueduct ending just beyond the 3rd ventricle. This was additional evidence for the diagnosis of stenosis of the aqueduct. At operation, a fistula was made between the 3rd ventricle and the subarachnoid space by splitting the lamina terminalis.

The post-operative convalescence was interesting in that the child developed a peculiar mutism for two to three days ; his physiological state was perfectly satisfactory, and he was alert enough to co-operate in quite a complete neurological examination. But he would not talk—until on the morning of the third day he suddenly started talking quite normally. This mutism has been observed in two other cases in which this operation has been done.

He was discharged from hospital three weeks after operation. The bone-flap was flush with the skull, the papilloedema had subsided, and the spinal fluid pressure was 70 mm. cerebrospinal fluid. The hydrocephalus thus appeared to have been relieved by the operation. He was free from headache, but the tremor of the hands and the unsteadiness of gait persisted.

He was readmitted to hospital five weeks later because the headache had recurred, and he had become very drowsy and incontinent of urine. On examination, two months after operation, the decompression was bulging, and he was clearly suffering from progressive hydrocephalus. The signs were much as on the previous admission, but in addition there were slight weakness, moderate ataxy, and dysdiadokokinesis down the left side of the body. It was suggested that these left-sided signs were due to a subdural collection of cerebrospinal fluid over the right hemisphere, but a further ventriculogram showed only severe hydrocephalus and no subdural collection. As we felt that we might be missing a cerebellar tumour, a posterior fossa exploration was made, but the

cerebellum appeared to be perfectly normal. The child derived no benefit from this additional decompression, and a week later the fistula in the 3rd ventricle was re-explored. It was found to be still patent but the hole was smaller than when it had been first made. It was enlarged to a diameter of about 1 cm. The effect of this operation on the intracranial pressure was apparently satisfactory as the bone-flap was sunken in, and the ventricular and spinal fluid pressures remained low. But the child died three weeks after this last operation from a staphylococcal infection which was probably acquired at the second operation (cerebellar decompression).

At autopsy the hole in the lamina terminalis was widely patent, and although the right Sylvian cistern was obliterated by operative adhesions, the left one and the cisterna basalis were ballooned out, suggesting that the fistula was effective. To the naked eye, the aqueduct appeared to be almost completely obliterated. Histologically this obliteration was shown to be due to proliferation of the subependymal glia.

This case illustrates what may be termed the juvenile type of the affection in which the hydrocephalus develops while the skull is still soft enough to stretch before the expanding ventricles. They are plump children with large heads. Although it is usually the recent onset of headaches and visual failure which brings them for treatment, the long duration of the lesion is betrayed by the large head and by the history of tremor of the hands, clumsiness or a little unsteadiness of gait, sometimes for years before the onset of pressure symptoms. With the papillœdema there may be secondary optic atrophy with corresponding affection of visual acuity. In one case it was difficult to say whether the atrophy was of the primary or secondary type, and there were bilateral central scotomata, presumably due to compression of the optic nerves by the dilated 3rd ventricle. Of oculomotor signs there may be limitation of upward movement of the eyeballs, squint, nystagmus, and sluggish reactions or immobility of the pupils, but three of the six verified cases have shown no abnormalities of the pupils or external ocular movements. Other cranial nerve palsies are not common. In the limbs a very common and characteristic feature is the fine vibratory tremor, especially of the hands, which was described in the case above ; and this may be the first or only abnormality of motor function. Hypotonicity and diminution of tendon reflexes may occur, or there may be spasticity, exaggerated tendon reflexes, and extensor plantar responses. The motor signs are usually bilateral, although they may be more marked on one side. Sensory abnormalities are rare. Incontinence of urine is common and may occur early in the course of the affection, before any signs or symptoms of increased intracranial pressure. Genital development is commonly delayed. One of these cases was associated with von Recklinghausen's neurofibromatosis and one with syringomyelia.

The cerebrospinal fluid pressure is raised, but the fluid itself is normal. X-rays of the skull show the general enlargement and convolutional thinning, separation of the sutures and possibly some erosion of the sella. Thirteen of our cases were of this juvenile type.

Adult Type

There is another interesting group of five cases in which symptoms first appeared between the ages of 18 and 25. These patients all came for treatment because of symptoms of increased intracranial pressure. They all had papillœdema, and one of them had no other abnormal signs. In the others the most striking feature was a disturbance of the pupils and external ocular movements ; defective upward movement of the eyeballs, skew asymmetry on conjugate deviation, and pupils which were virtually fixed to flashlight, but reacted well to accommodation-convergence. The neurological examination otherwise was negative, but in two cases there was tremor of the hands as in the juvenile group. One of these patients also had Recklinghausen's neurofibromatosis. Ventriculograms showed dilated lateral and 3rd ventricles, with defective filling of the aqueduct, and this appearance, together with the oculomotor signs, led to the diagnosis of tumour of the pineal gland or vicinity. They were all operated on (and had subsequent deep X-ray therapy) ; and they are all alive and

well now, after periods of seven, six, four and a half, and three years. These results suggest that the lesion was not neoplastic and is more likely to have been a benign stenosis of the aqueduct.

The diagnosis still rests on accurate ventriculograms. This clinical picture of increased intracranial pressure with signs of hydrocephalus may suggest a suprasellar tumour, a pineal or mid-brain tumour, or a tumour in the posterior fossa, especially in the upper part of the vermis. The ventriculogram shows dilated lateral and 3rd ventricles and perhaps a funnel-shaped termination of the aqueduct shadow just distal to the 3rd ventricle, and this may be sufficient to exclude suprasellar and pineal tumours. But the same appearances may be seen in some cases of tumours of the cerebellum and mid-brain, and in the present state of radiographic technique it may be impossible to say more than that the obstruction is below the 3rd ventricle. With improvements in technique, however, we may expect to get the right answer in a greater proportion of cases. Lysholm and Twining have described the characteristic deformities of the 3rd ventricle and aqueduct seen in lesions which may be confused with non-neoplastic stenosis of the aqueduct.

Treatment.—Attempts have been made to dilate the narrowed aqueduct by the passage of a small catheter from the 4th ventricle up into the 3rd ventricle. This was a blind procedure, capable of doing a great deal of damage to important structures in the neighbourhood of the aqueduct, and in 1922 Dandy discarded it with the comment that it could not be permanently effective. But he described a new operation for the relief of the condition: he had punctured the "floor of the 3rd ventricle" (evidently the lamina terminalis) in six cases of stenosis of the aqueduct, to allow the cerebrospinal fluid to escape from the 3rd ventricle into the supratentorial subarachnoid space without having to traverse the narrowed aqueduct. He thought this might be effective in all forms of obstructive hydrocephalus in which the obstruction was distal to the 3rd ventricle, although he made no claims for the success of the method. I have not been able to discover his more mature views about the value of the procedure, nor what happened to the six cases.

In the following year, Mixter passed an operating ventriculoscope through the lateral ventricle into the 3rd ventricle via the foramen of Monro, and punctured the floor of the 3rd ventricle under direct vision. This case was apparently successful, but again the method was not exploited.

In 1936, Stookey and Scarff reported six cases of stenosis of the aqueduct in which they had punctured not only the lamina terminalis but also the floor of the 3rd ventricle, thus allowing a double exit into the capacious cisterna interpeduncularis (basalis) as well as into the cisterna chiasmatis. This procedure was successful in three out of four non-neoplastic cases, and in one case of mid-brain tumour there had been a comfortable survival for seven months until the date of their report.

Other procedures which have been tried are (1) opening of the 3rd ventricle by splitting the corpus callosum; (2) temporal decompression; (3) cerebellar decompression.

In this present series, the results of these various procedures can be seen from the following tables:—

TABLE I.—JUVENILE TYPE (13 CASES).

I.—*Six cases proved by autopsy.*

	Treatment	Result
1 F. B.	Cerebellar decompression	Death on table due to wax embolus in lung
2 G.O'S.	Anterior 3rd ventriculostomy (Dandy); cerebellar decompression; exploration of 3rd ventriculostomy	Death from staphylococcal infection 3 weeks after last operation
3 M. D.	Double 3rd ventriculostomy (Stookey-Scarff)	Death 24 hours after operation (hyperthermia)
4 N. J.	Pineal exploration; splitting of corpus callosum	Death 1 month after operation
5 B. H.	No treatment	Death from infection 3 days after ventriculogram
6 L. M.	No treatment	Death 3 years after onset of symptoms

II.—*Six cases in which diagnosis was made by ventriculography.*

1	P. W.	Splitting of posterior part of corpus callosum	Temporary relief of symptoms. Death 10 months later. No autopsy
2	F. E.	Anterior 3rd ventriculostomy	*Satisfactory 1 year after operation
3	E. J.	Anterior 3rd ventriculostomy	Satisfactory 4 months after operation
4	B. C.	Anterior 3rd ventriculostomy	Satisfactory 2 years after operation
5	F. M.	Cerebellar decompression (+ X-ray therapy)	Satisfactory 3 years after operation
6	R. D.	Cerebellar decompression	Satisfactory 4 years after operation.

III.—*One case diagnosed on clinical evidence : no ventriculogram.*

1	O. R.	Cerebellar decompression	Satisfactory 3 years after operation

* In the results classified as " satisfactory ", the symptoms of increased intracranial pressure were relieved (without bulging of the decompression), and there was improvement in the neurological signs.

TABLE II.—ADULT TYPE (5 CASES).

Five cases in which ventriculogram showed obstruction below 3rd ventricle. The first four had deep X-ray therapy after operation as they were suspected of having pineal tumours.

		Treatment	Result
1	D. S.	Pineal exploration	Alive and well 7 years after operation
2	R. V.	Temporal decompression	Alive and well 6 years after operation
3	T. F.	Cerebellar decompression ineffective ; pineal exploration and splitting of corpus callosum	Alive and well 4 years after operation
4	R. I.	Cerebellar decompression	Alive and well 3 years after operation
5	G. B.	Cerebellar decompression	Recurrence of symptoms 3 months after operation. (Under observation at present)

The four patients who are alive and well are quite free of symptoms. No. 1 is serving as an A.R.P. warden, No. 2 is practising as a police court solicitor, No. 3 is a telephone operator, and No. 4 is serving in one of the armed forces.

TABLE III.—RESULTS OF VARIOUS PROCEDURES EMPLOYED IN TREATMENT OF STENOSIS OF AQUEDUCT.

I.—*Decompression operations.*			
A.	Temporal decompression (+ deep X-ray therapy)	Effective	1 case
B.	Cerebellar decompression	Effective	4 cases (2 + deep X-ray)
		Ineffective	3 cases
		Operative death	1 case
II.—*3rd ventricle operations.*			
A.	Pineal exploration and splitting of corpus callosum	Effective	2 cases (2 + deep X-ray)
		Ineffective	2 cases
B.	Anterior 3rd ventriculostomy (Dandy)	Effective	4 cases (? 3)
		Ineffective	(? 1)
C.	Double 3rd ventriculostomy (Stookey-Scarff)	Death after 24 hours	1 case

It is impossible to say from these results just what is the proper treatment of stenosis of the aqueduct. On physiological grounds, 3rd ventriculostomy seems to be a sound procedure. Splitting the lamina terminalis is an operation fraught with no great risks, and certainly in some cases it seems to be effective. The Stookey-Scarff operation is more dangerous, as the puncture of the floor of the ventricle usually has to be done blindly ; my single experience of it was not a happy one. Operations on the posterior part of the corpus callosum are more difficult and dangerous, and are apt to leave a permanent hemianopia.

A case can certainly be made for the external decompressions. Five of the best results in this series must be attributed to this method despite the assertion of

Stookey and Scarff that such operations are theoretically unsound. Indeed it is not easy to explain how these operations are effective, but that they may be so there can be no doubt. A cerebellar decompression will also occasionally disclose a tumour which would have been missed by a supratentorial operation. It may be that we should start by doing a cerebellar decompression ; if that is not effective, then a 3rd ventriculostomy. I would first split the lamina terminalis, and if that were not effective, puncture the floor of the ventricle at a subsequent operation. As we are dealing with an essentially benign condition, conservative operations carrying no great risks are to be preferred to more radical measures.

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