Aqueduct stenosis

Case review and discussion

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SUMMARY Twenty-seven cases of hydrocephalus associated with aqueduct stenosis are reviewed, and a further nine cases discussed in which hydrocephalus was present and the aqueduct was stenosed but some additional feature was present. This was either a meningocele or an encephalocoele, or else the aqueduct was not completely obstructed radiologically at the initial examination. The ratio of the peripheral measurement from theinion to the nasion to the distance between theinion and the posterior lip of the foramen magnum is presented for each case with an outline of the ventricles. The cases behave as would be expected if the aqueduct was being blocked by the lateral compression of the mid-brain between the enlarged lateral ventricles. On reviewing these cases and other evidence it is suggested that non-tumourous aqueduct stenosis is more likely to be the result of hydrocephalus than the initial cause. The response to treatment is reviewed and a high relapse rate noted. It is suggested that assessment of the extracerebral pathways may be advisable before undertaking third ventriculostomy or ventriculo-cisternostomy.

Dandy (1920, 1945) stated that stenosis of the aqueduct of Sylvius was the most common cause of congenital hydrocephalus; this pathological relationship had been suggested by Schlapp and Gere in 1917, and has been largely accepted subsequently (Sheldon *et al.*, 1930; Parker and Kernohan, 1933; Pennybacker, 1940; Bickers and Adams, 1949; Russell, 1949; Beckett *et al.*, 1950; Petit-Dutaillis *et al.*, 1950; Paine and McKissock, 1955; Greenwood and Hickey, 1956; Johnson, 1958, 1968; Edwards, 1961; Scarff, 1963; Elvidge, 1966; Nag and Falconer, 1966; Schechter and Zingesser, 1967; Johnson and Johnson, 1968; Milhorat, 1972; Crosby *et al.*, 1973; Jansen, 1975; Little *et al.*, 1975).

Williams (1973) has questioned this relationship between hydrocephalus and aqueduct stenosis, and proposed on theoretical grounds that non-tumourous aqueduct stenosis could best be understood as the result rather than the cause of hydrocephalus. This paper reviews cases admitted to The Midland Centre for Neurosurgery and Neurology between 1964 and 1974. The material

has been analysed retrospectively for evidence on whether the stenosis was the cause or the result of the hydrocephalus.

Case material

The selection of cases was difficult because there is no acceptable definition of aqueduct stenosis. For example, one of the most common varieties of hydrocephalus in association with narrowing of the aqueduct occurs in association with defects in the coverings of the neuraxis, both spina bifida and encephalocoeles. These cases were numerous but their inclusion was of little value because they were not always investigated by ventriculography. Many had been treated on the basis that investigation would not alter the choice of treatment. Additionally, the discussion of such cases is complicated by the widespread idea that such an association may be the result of teratogenic influences acting throughout the neuraxis. Many other reviews have excluded such patients. Other cases which gave rise to difficulty were those in which the aqueduct was narrow but not occluded. This is rather an artificial division and depends partly upon the contrast medium since complete arrest of a

water soluble medium is very uncommon. It was possible to demonstrate a hold-up in many patients who later proved to have only a delay in the passage of contrast medium. If pictures were taken after some days, even iophendylate (Myodil) was found to have passed through the aqueducts of some cases which had initially seemed blocked.

The criteria for inclusion in our initial group, therefore, included freedom from defects of

closure of the neuraxis and obstruction at the aqueduct as demonstrated at initial ventriculography.

Table 1 summarises the data of the 27 initial cases (1 to 27). The ages when they presented to us ranged from 5 weeks to 43 years. Table 2 gives the age of presentation of cases in this series compared with those in some others. The male to female ratio was 14 : 13.

Table 1 Summary of data on 27 patients with aqueduct stenosis

Case	Duration of illness and symptoms	Initial treatment	Post-operative results	Late results
1	2 mo increasing head size	SHV	Vomiting, visual impairment, and right hemiparesis lead to revision	Psychomotor retardation
2	3 mo increasing head size	SHV	Drowsiness and vomiting	Multiple revisions required
3	3 mo increasing head size	SHV	Persistent valve problems	Satisfactory after revisions
4	2 mo increasing head size	SHV	Satisfactory	Satisfactory until death
5	1 mo increasing head size	SHV	Satisfactory. Aqueduct became patent	Partial blindness and squint
6	3 mo increasing head size	SHV	Satisfactory	Slight mental retardation
7	3 mo increasing head size	3V	Continued difficulties	Satisfactory
8	3 mo increasing head size in infancy. Visual deterioration at age 6 yr	T	Satisfactory. Aqueduct became patent	Shunt infection leading to death
9	3 wk headaches, vomiting, papilloedema	T	Satisfactory	Visual acuity impairment
10	1 mo headache, vomiting, optic atrophy	T	Satisfactory	Satisfactory
11	3 yr ataxia. Tremor, spastic gait	T	Post-op. AEG. Air entered lateral ventricle through reopened aqueduct. Acute relapse at 12 years	Mental retardation
12	4 mo dysarthria. Left hemiparesis	T	Increasing spasticity left side. Re-operation	Blind when he awoke after SHV operation. Residual spastic gait. Small subdural haematoma at 13 years
13	6 mo diplopia. Ataxia	Cannulation of the aqueduct	Satisfactory	Residual hemiparesis
14	Papilloedema	T	Satisfactory	Satisfactory
15	Headaches 6 mo. Precocious puberty	T	Re-operation required	Satisfactory
16	7 mo tremor, headaches, papilloedema	3V	Initially well; after 2 years, headaches, vomiting, ataxia and disturbance of eye movements	Spontaneous CSF rhinorrhoea, meningitis
17	Clumsiness of hands	3V	Well for 3 years	Died
18	Failing vision	3V	Well for 3 years	Repeated surgery for relapses lead to satisfactory result apart from left hemiparesis
19	Papilloedema	T	Satisfactory	Satisfactory
20	1 mo headaches, visual impairment. Ataxia. Papilloedema	T	Satisfactory	Satisfactory
21	Birth injury, mentally retarded	T	Satisfactory	Satisfactory
22	9 mo ataxia, headaches, epilepsy	T	Satisfactory	Satisfactory
23	Epilepsy for 6 mo	T	Satisfactory	Satisfactory
24	Meningitis aged 2 yr	Theca laparostomy	Satisfactory	Satisfactory
25	SHV after relapse	3V	Satisfactory	Satisfactory
26	Hydrocephalus in infancy which spontaneously arrested. 3 mo headaches, epilepsy. Papilloedema.	T	Satisfactory for 3 months then drowsiness and papilloedema recurred. Needed revision	Well for 6 years. Headaches and ataxia required revision of Torkildsen shunt
27	7 mo epilepsy. Ataxia	—	Died	Torkildsen shunt
28	10 yr recurrent headaches. Papilloedema	3V	Persistent headaches, vomiting. Satisfactory after valve	Died 6 months after operation, cause unknown, (tumour not excluded)
29	4 wk headaches. Papilloedema	3V	Satisfactory	Satisfactory
30	5 mo headaches. Episodes of unconsciousness in one of which he was admitted	Ventricular drainage SHV	Satisfactory	Satisfactory
31	Headaches. 4 yr. Papilloedema	3V	Well for 3 mo. Then recurrence of headaches and papilloedema	Torkildsen shunt was followed by right hemiplegia

SHV = Spitz-Holter valve; 3V = Third ventriculostomy; T = Torkildsen's operation; TL = Theca-laparostomy.

Table 2 Age at presentation in present series and in other studies

Studies	Age at presentation (years)										
	0-	5-	10-	15-	20-	25-	30-	35-	40-	45-	50+
Dandy (1920)	15			1							
Parker and Kernohan (1933)	1	1	1	2			1				
Beckett <i>et al.</i> (1950)	3	1		1	1	1			1		3
Petit-Dutaillis <i>et al.</i> (1950)		6	4	4		9					
Paine and McKissock (1955)		9		8		5		2		1	
Nag and Falconer (1966)		1		1	2	2	1	1		1	
Schechter and Zingesser (1967)	22	10	10	10		17		6		8	5
Nöetzel (1970)	4		1	1							
Harrison <i>et al.</i> (1974)*						20		10		7	8
Little <i>et al.</i> (1975)†				2	6	3	2	2		1	1
McMillan and Williams (present ser.)	9	6	5	2	1	1	1	1	1		

* Age at diagnosis in selected adult cases, some of which may also appear in Paine and McKissock's and Schechter and Zingesser's reviews.

† Adult cases.

Because the group with an external defect and those with aqueducts which were narrowed but not occluded present features of interest, nine additional cases are described (a to i), five with spina bifida, two with encephalocoeles, and two with patent but stenosed aqueducts—one of whom had a post-meningitic hydrocephalus and an incomplete aqueduct septum. Outlines of ventriculograms are given (Fig. 2) but the age of onset and clinical features have not been reported.

AETIOLOGY

No aetiological factors were found. Factors of possible significance include birth injury and anoxia (case 10), and prematurity and jaundice (case 3). Enquiries into the antenatal history revealed no evidence of rubella or other viral infections, and no evidence of toxoplasma infection has been obtained from either parents or children. Enquiry has revealed no association with mumps in any case, either in the parents or in the children before the manifestation of the hydrocephalus. Case i and case 21 had septic meningitis but both were atypical.

SYMPTOMS AND SIGNS

There were no clinical features to enable the condition to be recognised with certainty. Most patients presented with symptoms and signs of raised intracranial pressure without focal signs. A few cases presented features suggestive of a focal cerebral lesion: case 11 had a spastic ataxic gait, case 12 presented with left hemiparesis, cases 19, 20, and 23 with epilepsy, and case 14 with precocious puberty. Major features are shown in Table 1.

INVESTIGATIONS

Evidence of raised intracranial pressure was a

consistent finding on plain skull radiography. Patients with aqueduct stenosis often have a low ion, and shallowness of the posterior fossa was noted in this series. Schechter and Zingesser (1967) defined the peripheral ratio, in which the numerator was the peripheral distance between the ion and the nasion, and the denominator the distance between the ion and the posterior lip of the foramen magnum. They compared the ratio found in cases of aqueduct stenosis with a group of normal cases. The ratio for their normal group was 2.8 : 1. They did not detail their methods of measurement nor the types of cases measured. The ratio calculated for 100 normal adult skull radiographs at this unit was 6.5 : 1 (standard deviation 1.04), a considerable divergence from Schechter and Zingesser's result which we are unable to explain. Our results of the ratio in the initial cases of aqueduct stenosis are given in Fig. 1 alongside the outlines of the ventriculograms (mean 8.56) and Fig. 2 which gives the same information for the additional cases (mean 13.24).

CONTRAST STUDIES

In four patients presenting with focal neurological signs (cases 17, 18, 20, and 25) carotid angiography was performed, and in one case vertebral angiography; hydrocephalus was the only abnormality found. Ventriculography was performed in all cases. Air was used in 22 cases, iophendylate (Myodil, Pantopaque) in four, and meglumine iocarmate (Dimer X) in one. In cases 6, 7, 11, 15, and 23 pneumoencephalography was used after ventriculography, allowing the fourth ventricle to be outlined. Figures 1 and 2 give tracings of the ventricular outlines for all cases. Ventricular enlargement was evident in all except case 21. Diverticula can be seen extending posteriorly from the third ventricle, sometimes bulging into the

Fig. 1 Outlines of ventriculograms of 27 patients with aqueduct stenosis. Small numerals indicate the peripheral ratio. (cf. Investigations)

Fig. 2 Outlines of ventriculograms of nine additional patients. Key as in Fig. 1.

posterior fossa or posterior hypothalamus as in case 14. Case 6 seemed to have a cavum vergae, and case g had a greatly enlarged suprapineal recess with a forward extension which may have been within the tela chorioidea. Sometimes there was an appearance suggesting channelling alongside the aqueduct, and occasionally the site of the original aqueduct was indeterminable. In many cases there was bulging of the third ventricle anteriorly into the sella turcica.

Treatment and results

Figure 3 illustrates the times of treatment for the

27 cases. The initial form of treatment chosen in one case was aqueduct cannulation. The cannula was placed relatively easily during posterior fossa exploration. Six patients were treated initially by third ventriculostomy, four of whom needed re-operation. Of the eight patients treated initially with a Spitz-Holter valve, four required revision and one was removed without having to be replaced. Of the 11 patients initially treated with ventriculo-cisternostomy shunting, as described by Torkildsen, three required further operations. The results are summarised in Table 1. Seventeen cases were regarded as satisfactory at most recent examination. Five patients died, cases 4, 7, and 16 from

Fig. 3 Time of treatment (27 patients).

meningitis, case 24 before treatment, and case 25 of an unknown cause. Three patients showed mental retardation, two had poor vision, one was ataxic, another spastic, and one suffered from chronic epilepsy. Three cases were of particular interest.

Case 16 A 10½ year old boy presented with ataxia, and investigation showed gross hydrocephalus with obstruction of the aqueduct to iophendylate. Posterior fossa exploration was carried out

followed by an anterior third ventriculostomy. He was well for two years, and then relapsed with papilloedema and ocular incoordination. Ventriculography showed a gross posterior enlargement of the third ventricle with three diverticula, one to each side of the falx above the tentorium, and one excavating the region of the aqueduct. The site of the aqueduct obstruction had moved to 2 cm below the midpoint of a line joining the internal occipital protuberance to the clinoid

processes. After a Torkildsen shunt he lived for 10 months and then died of meningitis after developing spontaneous CSF rhinorrhoea.

Case 21 This female patient had a history of bacterial meningitis at age 2 years followed by hydrocephalus which had been treated by theca-laparostomy and then by a Spitz-Holter valve at the age of 9 years. She presented to us at age 17 years with acute onset of drowsiness, bilateral ptosis, and failure of upward gaze. Ventriculography with water soluble medium showed small ventricles with an occluded aqueduct and blockage of the valve system. All symptoms resolved after revision.

Case 24 A 32 year old woman gave a 10 year history of severe occipital pain which occurred in separate attacks and completely resolved between bouts. Three months earlier she had been admitted elsewhere with suspected meningitis, but after a normal lumbar puncture she had recovered and been discharged. At the time of admission she had been ill for two weeks. She was drowsy, with bilateral papilloedema and marked neck stiffness. Skull radiographs showed evidence of raised intracranial pressure, and pneumoventriculography showed a hold-up at the aqueduct with an irregular appearance.

Despite close observation she died while awaiting further investigation. Postmortem examination showed a benign stricture of the aqueduct with forking and complete obliteration of the channels. There were distended diverticula of the back end of the third ventricle which appeared as though they were attempting to dissect the peri-aqueduct area alongside and dorsal to the original aqueduct.

Discussion

There are several reasons to doubt the belief that, when the two conditions are present together, aqueduct stenosis has caused hydrocephalus.

One reason is exemplified by our difficulty in deciding which patients to include among the initial cases. The radiological difficulty parallels the problems which histologists have found in defining the limits of normal. Why is it difficult to decide which cases have aqueduct stenosis and which do not? It seems that although hydrocephalus is clearly present in all cases in which there is an association between the two conditions, aqueduct stenosis is not always present, or is present to a partial or imperfectly defined extent (Sheldon *et al.*, 1930; Russell, 1949; Woollam and Millen, 1953; Drachman and Richardson, 1961). If the aqueduct stenosis were the cause of the hydro-

cephalus then the latter should not occur until the stenosis was extremely severe. A CSF production of around 20 ml/hr should be able to escape along an aqueduct with the bore of a small hypodermic needle before the pressure rises significantly. Such an aqueduct would, however, arrest air or iophendylate for an appreciable time. The expected result would be that in many cases the stenosis would be radiologically complete and the hydrocephalus in doubt, instead of the other way around.

The second difficulty over selection of cases concerns whether or not to include cases of spina bifida or encephalocele. The deformities of the aqueduct, the forking and atresia, are no different histologically from those which occur in association with the other varieties of hydrocephalus (MacFarlane and Maloney, 1957). Although virtually all cases of spina bifida have hydrocephalus most of them do not have radiological blockage of the aqueduct (Emery, 1972). The aqueduct is, however, usually narrowed (Emery, 1972; MacFarlane and Maloney, 1957). It is clear, therefore, that aqueduct stenosis does not cause the hydrocephalus in such cases but that it is perfectly reasonable to consider the reverse relationship.

Case 21 presented at age 17 years with an aqueduct stenosis within the initial definition, although she had not had an aqueduct stenosis at first but a communicating hydrocephalus secondary to meningitis. Harrison *et al.* (1974) excluded such cases from their review of 55 cases of adult aqueduct stenosis, although we can see no reason to assume that the mechanisms of closure of the aqueduct should be so different after septic meningitis as to exclude them from our series. There is likely to be considerable gliosis in such cases, but that is clearly not solely responsible for the contraction of the ependymal surfaces in the present case. The theca-laparostomy followed by ventriculo-atrial shunt must be held partly responsible. The collapse of the ventricular walls is typical of that seen when the lowering of the intraventricular pressure has been excessive and the ependymal walls fuse together as they come into contact, a process sometimes referred to as 'seaming'. The aqueduct in such cases not uncommonly becomes blocked as the hydrocephalus is controlled by a shunt. Foltz and Shurtleff (1966) reported 27 such cases. They commented on the occurrences of 'forking' as the walls of the previously distended aqueduct collapse and apparently subsidiary channels become pinched off and scarred. Collapse after lowering of the intra-aqueductal pressure is not contrary to the notion that the aqueduct might in certain cases be com-

pressed by the ventricles. The mechanism of forking is similar in that it is due to the existence of a higher compressive force around the aqueduct than the force which can be exerted outwards by the pressure within the aqueduct plus the strength of the walls. In the usual case, with an active hydrocephalus, this difference seems likely to be accounted for by the small area of the walls of the aqueduct which allows the intra-aqueduct pressure to exert only a small force against that produced by the distension of the lateral ventricles outside the midbrain (Williams, 1971). After shunting, the pressure is reduced throughout the intra-ventricular pathways and the collapse of the aqueduct may result from the growth of the brain tissues. This mechanism will be exaggerated within a cranial cavity which remains small and, additionally, the same mechanism which produces 'seaming' of the ventricles is likely to be at work.

The formation of septa within the aqueduct is almost certainly due to distension above and below an area of stenosis which has been developed in the aqueduct at an earlier stage of the disease. In cases where ependymitis may be suspected, such as our case i which was post-meningitic, a degree of sub-ependymal gliosis may be present and contribute to the retention of a narrowed aqueduct even though the third ventricle dilates above it and the fourth ventricle below (Fig. 2).

The aetiology of hydrocephalus in human cases is almost always unknown. There are a number of factors known to produce the condition in association with aqueduct stenosis, but most workers have concentrated on the aqueduct and have directed little attention to the examination of the subarachnoid space. Hereditary hydrocephalus has been reported in humans as an X-linked recessive condition (Bickers and Adams, 1949; Edwards, 1961; Jansen, 1975). There seems to us to be no proven basis for the usual assumption that stenosis preceded hydrocephalus. Some studies in animals, in which the stages of the disease may be thoroughly studied, also suggest that hydrocephalus causes the stenosis. Borit and Sidman (1972) studied a mutant strain of mice in which there is an initial phase of communicating hydrocephalus, followed by a phase of superimposed aqueduct stenosis. In animals there are many known if not perfectly understood causes, which include deficiency states and toxic and infective agents. Millen and Woollam (1958) reported the two conditions in vitamin A deficient rabbits, and suggested that the stenosed aqueduct was secondary to the hydrocephalus. Kesterson and Carlton (1971) reported hydrocephalus and aqueduct stenosis produced in

mice by feeding cuprizone, a substituted hydrazine. They suggested that the stenosis developed as a result of pressure exerted by oedematous mesencephalic tissue. This seems plausible, and it also seems possible that lateral ventricular pressure from a developing hydrocephalus could be an additional or even the principal factor.

Johnson and Johnson (1968) described hydrocephalus and aqueduct stenosis in mice after intracerebral inoculation with a strain of mumps virus. All the mice showed aqueduct stenosis. These workers have also shown that hydrocephalus can result from infection and can be associated with aqueduct stenosis even when inflammation of the aqueduct has not been seen (Johnson, 1968). No comment was made upon the appearance of the arachnoid villi, and these studies did not exclude the possibility that the stenotic aqueduct was secondary to the hydrocephalus. The studies of Masters *et al.* (1977), who produced experimental viral hydrocephalus in mice, suggest that arachnoid blockage causes hydrocephalus and that aqueduct stenosis is secondary.

EFFECTS OF HYDROCEPHALUS

It is generally accepted that smallness of the posterior fossa and depression of the tentorium cerebelli are the result of hydrocephalus occurring in early life.

As hydrocephalus advances, the direction of movement of the under surface of the temporal lobe is downwards. With depression of the tentorium the upper brain stem becomes more vulnerable to compression. There is competition for space at the incisura, particularly by the enlarging lateral ventricles and the medial portions of the temporal lobes (Johnson, 1958), thereby producing a bilateral tentorial cone. If the aqueduct becomes closed, the pressure within its lower part may drop below that in the lateral ventricles. Such a differential in pressure would, of course, tend to close the aqueduct more tightly, thus providing a valve-like action. The effect of pulsation is, therefore, important. If pulses pass up the aqueduct, for instance after coughing or straining, there may be delay in their return downwards, and progression of the hydrocephalus and aqueduct stenosis may occur. Such valve-like action has been measured in children with spina bifida although pressure differentials in such cases are most likely to be greatest at the foramen magnum (Williams, 1975). The occurrence of such a valve-like action would thus explain the principal ages of onset, shortly after birth and rapid growth, as has been postulated elsewhere in respect of not only the aqueduct

stricture but also the Arnold-Chiari malformation (Williams 1971, 1975). A valvular action could also explain the relapsing and remitting course of our case 21 who went through several critical periods of high pressure, the last of which was fatal. Similar periodicity was noted by Beckett *et al.* (1950) and Pennybacker (1940), and sudden unexpected death by Parker and Kernohan (1933). In the hydrocephalic process other cavities develop (Figs 1 and 2), and enlargement of the third ventricle results in extensions such as an enlarged suprapineal recess. These compete for space with the lateral ventricles and the temporal lobes to produce downward compression of the brain stem. All these factors, therefore, unite to embarrass the aqueduct further.

Localised neurological changes may occur from enlargement of the third ventricle in the pineal region. Precocious puberty may be associated with hydrocephalus and subarachnoid pouches in the posterior hypothalamus (Williams and Guthkelch, 1974), and in our case 14 there was an extension of the suprapineal recess in this situation (Fig. 1). Another effect is disturbance of eye movements, as in our cases 16 and 21 (Swash, 1974).

The susceptibility of the aqueduct to compression at the tentorium is undoubted. Ford and Spatz (1960) believed that relief of acute hydrocephalus as a complication of post-traumatic cerebral oedema and compression aided resolution of intracranial high pressure. In their cases the compression of the aqueduct was initiated by head injury with unilateral mass lesions but hydrocephalus appeared to be aggravating the problem.

RESPONSE TO TREATMENT

In the treatment of aqueduct stenosis, failure of all forms of internal shunting procedures is common (over 50% in this series). Most authorities agree that the major cause of failure is obstruction of the subarachnoid space (Dandy, 1920, 1945; Scarff, 1963; Paterson and Bergland, 1968).

There has been dispute as to whether the subarachnoid space is obliterated by pressure which is developed within the ventricles after formation, or whether the spaces never properly develop because of failure of cerebrospinal fluid to reach the subarachnoid space. Milhorat (1972) examined the subarachnoid space in 52 cases of congenital hydrocephalus and in no case was there congenital absence of the space. He also reported re-expansion of the subarachnoid space in two cases of congenital hydrocephalus treated by ventricular drainage. In adults, once failure of internal shunting has occurred, the usual procedure is the in-

sertion of a Spitz-Holter valve.

Third ventriculostomy, introduced by Dandy and later modified by Stookey and Scarff, remains a popular method of treatment. Scarff (1963) reviewed a total of 528 cases with an operative mortality of 15% and a 2.5% incidence of late complications. Other workers agree that late failures are common, with incompetence of the subarachnoid space being the presumed cause. The rate of failure of third ventriculostomy in Scarff's series is 85%. No cause for these failures was determined but it seems reasonable to suppose that if hydrocephalus from some other cause was responsible for the aqueduct stenosis, the original cause might still be incompletely resolved. The absorptive mechanisms thus may be inadequate to lower the pressure to a satisfactory extent.

Ventriculo-cisternostomy with a shunt (Torkildsen) is effective in aqueduct stenosis. Failures with this treatment are common, Torkildsen (1960) and Milhorat (1972) estimate a failure rate of 42%. In this series 10 cases were treated by ventriculo-cisternostomy, and three of these required revision. Ten cases are currently surviving, apparently using Torkildsen's shunts.

Aqueduct cannulation, tried but later abandoned by Dandy, has advocates. Greenwood and Hickey (1956) and Elvidge (1966) reported favourable results from this procedure at least in selected cases. Crosby *et al.* (1973) reported 30 hydrocephalic children with aqueduct obstruction treated by this method. They used a two shunt procedure to open the subarachnoid space before cannulation. The only patient in the present series where cannulation was adopted is alive and well (case 13).

Pennybacker (1940) claimed good results from external decompressions in six cases—five cerebellar and one temporal. One possibility after posterior fossa decompression is that posterior dislocation may occur, as in our case 16, and move a compressed aqueduct to a more protected situation beneath the tentorium. It is possible that any form of decompression may relieve the brain stem and the aqueduct. This suggests that external pressure is important in occluding the aqueduct rather than the constriction of the ependyma or of the immediately adjacent tissues. Our view is that improvement after decompression suggests that the lateral ventricles had been compressing the midbrain.

It is well known that in a case with apparently complete blockage of the aqueduct on iophendylate studies the contrast medium can pass eventually through the aqueduct, indicating an incomplete

anatomical obstruction (Jakubowski and Jefferson, 1972). There is some evidence of opening up of the aqueduct after decompression. Our cases 5, 8, and 11 all showed patency of the aqueduct after treatment. During exploration of the posterior fossa, cerebrospinal fluid may run down the aqueduct. This occurred in three cases in this series (2, 11, and 18).

In infants and young children with congenital hydrocephalus the usual treatment is the insertion of a Spitz-Holter valve—internal shunting is rarely successful, probably because in infancy the condition is more severe. If the aqueduct stenosis is indeed secondary to the hydrocephalus, another factor may be that the absorptive defect has incompletely resolved. Whatever was responsible for the blockage of the absorptive mechanism may still be present and bypassing the aqueduct perhaps does not deal with the fundamental defect. The flattened, thinned, and relatively plastic cerebral mantle of the infant is liable to fill the skull completely and flatten the subarachnoid space, thus continuing the chain of events which leads to decompensation.

Conversely, in the older cases which must have been at some stage 'compensated' the absorptive capacity of the subarachnoid space has necessarily developed relatively well. The subarachnoid space is, therefore, not so easily obliterated and treatment by internal shunting may be successful.

All methods of internal shunting are prone to failure, and it would seem advisable to assess the patency and functional capability of the subarachnoid space by air and radio-isotope studies before embarking on surgery.

Conclusion

In a review of this nature, which is retrospective and where pathological evidence is lacking, it is not possible to produce convincing evidence for any particular hypothesis related to the nature of aqueduct stenosis. One feature is that the cases have markedly variable findings. There are no clear boundaries, no patterns of behaviour which can be predicted, and there is no evidence of aetiology. Thus it seems that aqueduct stenosis is not a disease entity. It seems sensible to regard it as a radiological finding or as an event in the progression of a whole series of diseases—that is different causes of hydrocephalus. There tend to be other manifestations, lowering of the inion and multiple internal diverticulae being the most marked. Based on the evidence presented we suggest the following sequence of events.

Congenital or early onset of hydrocephalus, accompanied by mild ventricular enlargement, may escape attention initially because it becomes compensated as the ventricular or the overall skull size increases. Even a compensated mild hydrocephalus may lead to depression of the tentorium, the mid-brain thereby becoming exposed to compression by the lateral ventricles which does not necessarily obstruct CSF and thus lead to high pressure of itself. It seems probable that this stage is usually intrauterine. Narrowing of the aqueduct may be present at this stage. As the child grows, further factors come into play. If a valve-like mechanism operates at the aqueduct, holding back fluid which is forced into the lateral and third ventricle during straining, pressure differentials may exist for a time at the aqueduct which increase the moulding of structures around the incisura. The onset of respiration and crying may thus cause decompensation at birth, and the increase in the size of the body relative to that of the head may be responsible for the later incidence during growth. Other factors, such as aging of brain tissue and changes in elasticity, are also probably relevant. Once the hydrocephalus starts to become decompensated several additional factors act, including progressive flattening of the subarachnoid space and further aqueduct compression due to the bilateral 'cone' around the brain stem. In addition, the cystic extensions of the third ventricle above the aqueduct, and tracking which takes place alongside the aqueduct (sometimes even into the posterior fossa) add to its compression.

Patients, therefore, present at a time when the state of the aqueduct becomes critical. That this is usually some time after the onset of the first pathological changes is evidenced by the relative lowering of the inion and the development of an increased peripheral ratio. Such manifestations are certainly always of long standing, even when the clinical illness may seem very acute. Cure of the symptoms may be affected after bypassing the aqueduct or by decompressing it, but the failure rate suggests that factors other than aqueduct compression may still be operative in many cases.

Two important conclusions are suggested by this study. The first is that, before any proof of the hypothesis advanced here can be accepted, further research should be carried out on the nature of aqueduct stenosis. The second, from a practical standpoint, is that the high failure rate of third ventriculostomy and ventriculo-cisternal shunts confirms the belief that, before any form of internal shunting is performed, the functional capacity of the subarachnoid space should be

assessed and factors leading to its subsequent incapacity evaluated.

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