Sylvian aqueduct syndrome and global rostral midbrain dysfunction associated with shunt malfunction

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Object. This study is a retrospective analysis of clinical data obtained in 28 patients affected by obstructive hydrocephalus who presented with signs of midbrain dysfunction during episodes of shunt malfunction.

Methods. All patients presented with an upward gaze palsy, sometimes associated with other signs of oculomotor dysfunction. In seven cases the ocular signs remained isolated and resolved rapidly after shunt revision. In 21 cases the ocular signs were variably associated with other clinical manifestations such as pyramidal and extrapyramidal deficits, memory disturbances, mutism, or alterations in consciousness. Resolution of these symptoms after shunt revision was usually slow. In four cases a transient paradoxical aggravation was observed at the time of shunt revision. In 11 cases ventriculocisternostomy allowed resolution of the symptoms and withdrawal of the shunt.

Simultaneous supratentorial and infratentorial intracranial pressure recordings performed in seven of the patients showed a pressure gradient between the supratentorial and infratentorial compartments with a higher supratentorial pressure before shunt revision. Inversion of this pressure gradient was observed after shunt revision and resolution of the gradient was observed in one case after third ventriculostomy. In six recent cases, a focal midbrain hyperintensity was evidenced on T₂-weighted magnetic resonance imaging sequences at the time of shunt malfunction. This rapidly resolved after the patient underwent third ventriculostomy.

It is probable that in obstructive hydrocephalus at the time of shunt malfunction, the development of a transtentorial pressure gradient could initially induce a functional impairment of the upper midbrain, inducing upward gaze palsy. The persistence of the gradient could lead to a global dysfunction of the upper midbrain.

Conclusions. Third ventriculostomy contributes to equalization of cerebrospinal fluid pressure across the tentorium by restoring free communication between the infratentorial and supratentorial compartments, resulting in resolution of the patient's clinical symptoms.

Key Words * aqueductal stenosis * hydrocephalus * Parinaud's sign * sylvian aqueduct syndrome * shunt complication * third ventriculostomy

In a few patients with shunted hydrocephalus, usually related to aqueductal stenosis, shunt malfunction can be revealed by a sylvian aqueduct syndrome (Parinaud's syndrome or pretectal syndrome) characterized by vertical gaze restriction, abnormal pupillary reaction, upper lid retraction, and convergence-retraction eye movements. Paralysis of convergence and skew deviation may also be seen.[2,61] Rarely, this syndrome can be observed as a presenting symptom in active hydrocephalus;[10] however, according to some authors,[27] it is probable that the very common "setting sun" sign in neonatal hydrocephalus is caused by the same mechanism.

In some cases the sylvian aqueduct syndrome observed during shunt malfunction can be associated with a more complex clinical picture, suggesting a global rostral midbrain dysfunction, according to the definition proposed by Barrer, et al.[3] The evolution of this rare global rostral midbrain dysfunction is progressive and can be life threatening in the absence of appropriate treatment. Despite several hypotheses, the pathophysiological mechanism of this syndrome remains obscure.

At Hôpital Necker-Enfants Malades, during the last 25 years we have observed 28 patients suffering from sylvian aqueduct syndrome or global rostral midbrain dysfunction at the time of shunt blockage. The goal of this report is a retrospective analysis of the clinical data obtained in these patients.

CLINICAL MATERIAL AND METHODS

All charts of patients affected by congenital or acquired aqueductal stenosis, who were treated in our department during the last 25 years were retrospectively reviewed to identify the neurological symptoms revealing a shunt failure. According to their clinical histories, 28 patients had presented with several episodes of Parinaud's syndrome, sylvian aqueduct syndrome, or global rostral midbrain dysfunction during shunt malfunction. All ventriculography, computerized tomography (CT), and magnetic resonance (MR) imaging studies obtained in these patients were analyzed and the patients' charts were carefully reviewed to define the clinical features of midbrain dysfunction during episodes of shunt failure.

In six cases supratentorial intracranial pressure (ICP) was recorded with the aid of a coronal extradural sensor (Plastimed, Saint-Leu La Forêt, France) filled with saline and connected to a transducer (either an 800 Bentley Trantec [Kontron S.A. Trappes, France] or an HP 1280 [Hewlett-Packard, Orsay, France]). In seven cases, supratentorial and infratentorial ICPs were recorded simultaneously before and after shunt revision by using a coronal supratentorial extradural sensor and an occipital infratentorial extradural sensor. Informed consent was obtained from the parents of the patients before implantation of the infratentorial extradural sensor. The supratentorial sensor was placed 2 cm anterior to the right coronal suture and 3 cm lateral to the midline. The infratentorial sensor was placed 3 cm below the right transverse sinus and 3 cm lateral to the midline. After the patient had been placed in a strictly horizontal left-lateral position, so that the two sensors were on the same horizontal line, level with the right atrium, the sensors were calibrated to zero at atmospheric pressure. Ten minutes later the sensors were again calibrated to zero, after drift and fluid temperature had been stabilized. The ICP recording was then started with the patient in the lateral position and continued for 24 hours. The zero level was controlled at the end of recording.

RESULTS

The causes of hydrocephalus included malformative aqueductal stenosis in 17 cases, meningitis in four cases, intraventricular hemorrhage in two cases, tectal tumor in one case, posterior fossa tumor in two cases, vein of Galen malformation in one case, and toxoplasmosis in one case. The age of the patients at

the time of cerebrospinal fluid (CSF) shunt implantation ranged from 1 month to 14 years (mean 57 months; median 28 months). None of the patients underwent third ventriculostomy as a first procedure, as has been customary for the treatment of aqueductal stenosis in our department since 1970. The reasons for not performing third ventriculostomy were the lack of ventriculographic criteria[24] for a safe procedure (12 cases) at the beginning of our experience, communicating hydrocephalus at the time of diagnosis (seven cases), the first procedure having been performed in another hospital (four cases), technical reasons (two cases), presence of an optic glioma (one case), and surgeon's choice (two cases). Each patient underwent at least one shunt revision (range 1-18 revisions; mean 4.4 revisions).

The neuroradiological examinations (13 ventriculography and 15 CT studies) at the time of the first diagnosis of hydrocephalus revealed a triventricular dilation with aqueductal stenosis and axial enlargement of the third ventricle in 20 cases and a communicating hydrocephalus with quadriventricular dilation in eight cases with posthemorrhagic, postmeningitic, or posttumoral hydrocephalus. In 20 cases, baseline CT scans were obtained during follow-up evaluation in the absence of clinical signs of shunt failure. These revealed slit ventricles in 14 cases, normal ventricles in five cases, and dilated ventricles in only one case. At the time of shunt failure, all patients exhibited a triventricular dilation with significant axial enlargement of the third ventricle. In the six cases treated in recent years, sagittal T₂-weighted MR imaging sequences performed during shunt malfunction revealed anatomical modifications (Fig. 1) and focal hyperintensity of the midbrain.

Fig. 1. Case 18. Left: Sagittal T₂-weighted MR image (TR 5000 msec, TE 125 msec, 512 X 256 matrix, two excitations) obtained 48 hours after the patient underwent shunt revision. At the time the MR image was obtained, the patient presented with sylvian aqueduct syndrome and coma. Note the deformation of the floor of the third ventricle, bulging into the interpeduncular cistern, and the midbrain deformation. The whole midbrain and the periaqueductal region present an abnormal hyperintense signal. Right: Sagittal T₂-weighted MR image (TR 4000 msec, TE 119 msec, 256 X 192 matrix, two excitations) obtained 3 days after the patient underwent endoscopic third ventriculostomy that allowed rapid resolution of the clinical signs. The anatomical deformation has completely resolved, the midbrain hyperintensity has disappeared, and the CSF flow artifact is visible through the floor of the third ventricle.

These abnormalities resolved in all cases within a few hours following third ventriculostomy (Figs. 1-3). In two patients (Cases 26 and 27) who were surgically treated for posterior fossa tumors, in whom the obstruction was located in the lower portion of the fourth ventricle, the T₂ hyperintensity was diffuse to the

whole floor of the fourth ventricle above the obstruction (Fig. 3).

Fig. 2. Case 6. Left: Sagittal T₂-weighted MR image (TR 4000 msec, TE 120 msec, 256 X 192 matrix, three excitations) obtained at the time of shunt malfunction 3 hours before the patient underwent endoscopic third ventriculostomy. Note the floor of the third ventricle, bulging into the interpeduncular cistern, and the ballooning of the suprapineal recess, bulging into the quadrigeminal cistern. Mild deformation of the midbrain is visible with abnormal hyperintensity. At the time of the MR imaging study, the patient presented upward gaze palsy and pyramidal signs. Right: Sagittal T₂-weighted MR image (TR 4000 msec, TE 114 msec, 256 X 192 matrix) obtained 48 hours after the patient underwent endoscopic third ventriculostomy. The anatomical deformations (left) have resolved and the midbrain hyperintensity has disappeared. Note the CSF flow artifact through the third ventriculostomy. The patient's clinical signs had completely resolved.

Fig. 3. Case 27. Axial T₂-weighted images obtained in a patient who had undergone surgery for a medulloblastoma in another country and was admitted for shunt malfunction revealed by sylvian aqueduct syndrome and akinetic mutism. Left: Image (TR 4500 msec, TE 119 msec, 512 X 256 matrix, two excitations) obtained through the upper pons revealing dilation of the lower aqueduct and abnormal hyperintensity of the tegmentum at the pontine level. Right: Image (TR 4000 msec, TE 119 msec, 512 X 256 matrix, two excitations) obtained 18 hours

after the patient underwent endoscopic third ventriculostomy at the same level as that shown on the left. Note the resolution of brainstem hyperintensity, the reduction in aqueduct size, and the flow artifact in the prepontine cistern.

Clinical Findings at Shunt Malfunction

In all patients a paralysis of upward conjugate gaze was observed; in most cases this was associated with paralysis of convergence (19 of 28 cases). Collier's sign was observed in 19 patients, pupillary reflex to light and convergence were sluggish or abnormal in eight patients, ocular pursuit was abolished in 10 patients, lateral nystagmus was observed in one patient, and convergence nystagmus in one patient (Table 1). The incidence of retraction-convergence nystagmus is probably underestimated because health care trainees do not routinely look for it in patients admitted for shunt malfunction. In all cases the ocular signs resolved completely after shunt revision with variable delays (mean 24 days; median 5 days; range 1-120 days).

TABLE 1 OCULAR FINDINGS IN 28 PATIENTS WITH OBSTRUCTIVE HYDROCEPHALUS AND SHUNT MALFUNCTION*					
Case No.	Upward Gaze/ Convergence Paralysis†	Collier's Sign	Retraction or Lateral Nystagmus	Pupil Abnormalities	No Ocular Pursuit
1	+	+	—	+	—
2	+	—	—	+	+
3	+	—	—	—	+
4	+	—	—	—	—
5	+	—	—	+	—
6	+	—	—	—	—
7	+	+	—	—	—
8	+	+	—	—	—
9	+	+	—	—	—
10	+	+	—	—	—
11	+	+	—	+	+
12	+	+	—	+	+
13	+	—	—	—	—
14	+	+	—	+	+
15	+	+	+	—	—
16	+	—	—	—	—
17	+	—	—	—	—
18	+	—	—	—	—
19	+	+	+	—	+
20	+	+	—	—	—
21	+	+	—	—	—
22	+	+	—	—	—
23	+	+	—	+	+
24	+	+	—	—	—
25	+	+	—	—	—
26	+	+	—	—	+
27	+	+	—	—	+
28	+	+	—	+	+
* + = present; — = absent.					
† Upward gaze paralysis with or without paralysis of convergence.					

Gaze paralysis and oculomotor dysfunction were isolated in seven cases and associated with other clinical signs in 21 cases (Table 2). Extrapyrarnidal signs were observed in eight cases (bradykinesia in one case, speech difficulty in four cases, abnormal movements in two cases, and tremor in five cases), pyramidal signs in nine cases (hyperreflexia in six cases, hypertonia in six cases, upper limb hyperextension in four

cases, and decerebrate posturing in three cases), akinetic mutism in nine cases, memory disturbances in five cases, and progressive alterations in consciousness leading to deep coma in 16 cases.

TABLE 2					
ASSOCIATED CLINICAL FEATURES IN 21 PATIENTS WITH OBSTRUCTIVE HYDROCEPHALUS AND SHUNT MALFUNCTION*					
Case No.	Extrapyr- amidal Signs	Memory Deficits	Pyramidal Signs	Mutism	Coma
1	+	-	+	+	+
2	-	-	-	+	+
5	+	-	-	+	-
8	-	-	+	-	+
9	-	-	+	-	+
10	+	-	-	-	-
11	-	-	-	+	+
13	-	-	+	-	+
14	+	+	+	+	+
15	-	-	-	-	+
16	+	-	-	-	-
18	-	-	-	-	+
19	-	-	+	+	+
20	+	-	-	-	-
21	-	+	+	-	+
22	-	-	-	-	+
23	-	+	-	-	+
24	+	-	+	-	+
26	-	+	-	+	+
27	-	-	-	+	-
28	+	+	+	+	+
* + = present; - = absent.					

In all cases the oculomotor disorders were the first signs to be noticed. Nine patients suffered several episodes of isolated and spontaneously regressive upward gaze palsy, although normal CT scans had been obtained in these patients. These episodes preceded the impending shunt failure by weeks or months. Shunt failure was revealed by ventricular dilation on CT scans, signs of intracranial hypertension, stable upward gaze palsy, and clinical deterioration that was usually progressive with the onset of pyramidal and extrapyramidal signs, akinetic mutism, and decreased consciousness. In two cases, memory impairment was observed several weeks before the diagnosis of shunt malfunction.

In the seven patients in whom the only signs of shunt malfunction were the intracranial hypertension syndrome and conjugate gaze palsy, the former resolved immediately after shunt revision, whereas the latter disappeared in less than 3 days (mean 1.6 days; range 1-3 days).

On the contrary, patients with more complex clinical features often did not demonstrate rapid improvement after shunt revision, leading to repeated ventricular tapping, ICP recordings, or subsequent shunt revisions.

In four cases clinical deterioration, manifested by onset of new symptoms, was observed after shunt revision. In these patients, the clinical deterioration was in contrast with radiological examinations that demonstrated decreasing ventricular size after revision and ICP recordings that showed normal or subnormal ICP levels.

Long-Term ICP Recordings

Supratentorial ICP recordings, performed in six patients during episodes of shunt dysfunction, showed high baseline ICP values in three patients (40-50 mm Hg) and normal baseline ICP values in three patients

(10-15 mm Hg). No correlation was found between ICP levels and the severity of the clinical condition. Simultaneous recordings of supratentorial and infratentorial pressures before shunt revision revealed in all cases a higher supratentorial pressure with a transtentorial pressure gradient ranging from 4 to 10 mm Hg (mean 6 mm Hg) (Figs. 4 and 5 upper).

Fig. 4. Case 14. Continuous simultaneous transtentorial pressure recordings obtained during shunt malfunction in a patient with a clinical picture of severe global rostral midbrain dysfunction. Intracranial pressure is higher in the supratentorial compartment (baseline 18 mm Hg) than in the infratentorial compartment (baseline 12 mm Hg).

Fig. 5. Case 21. Upper: Continuous simultaneous transtentorial pressure recordings obtained during shunt malfunction. The patient presented with Parinaud's syndrome, pyramidal signs, and memory deficits. Intracranial pressure is higher in the supratentorial compartment (baseline 19 mm Hg) than in the infratentorial compartment (baseline 14 mm Hg). Lower: Continuous simultaneous transtentorial pressure recordings obtained 6 days after the patient underwent shunt revision. There has been inversion of the pressure gradient with a higher infratentorial pressure (baseline 11 mm Hg) and lower supratentorial pressure (baseline 5 mm Hg). The patient exhibited complete clinical recovery, but Parinaud's syndrome persisted for 9 days after the shunt revision.

The general patterns of the recordings were deeply altered and during sleep the pattern of paradoxical sleep was lost. Pressure waves were sometimes observed with "plateau" configuration. Details of the supratentorial and infratentorial pressure recordings are shown in Table 3.

Case No.	Preshunt Revision		External Ventricular Drainage		Post-shunt Revision		Post-ventriculocisternostomy	
	STP	ITP	STP	ITP	STP	ITP	STP	ITP
9	15	11	NP	NP	NP	NP	NP	NP
14	18	12	NP	NP	NP	NP	NP	NP
16	70	60	NP	NP	NP	NP	NP	NP
19	23	15	12	15	NP	NP	15	15
21	19	14	NP	NP	5	11	NP	NP
22	18	14	4	7	5	9	NP	NP
23	18	13	NP	NP	10	8	NP	NP

* ITP = infratentorial pressure; NP = not performed; STP = supratentorial pressure.

In three patients (Cases 21-23) simultaneous supratentorial and infratentorial ICP recordings were made several days after shunt revision (Fig. 5 lower), showing inversion of the pressure gradient with lower supratentorial pressure. Increases in this gradient were observed when the patients were placed in the sitting position; these were attributed to the "siphoning effect" of the well-functioning shunt (Fig. 6).

Fig. 6. Case 22. Upper Left: Continuous simultaneous transtentorial pressure recordings obtained during shunt malfunction. The patient was comatose with Parinaud's syndrome. Intracranial pressure is higher in the supratentorial compartment (baseline 18 mm Hg) than in the infratentorial compartment (baseline 14 mm Hg). Upper Right: Recordings obtained 7 days after the recordings shown in in upper left. External ventricular drainage (EVD) has been placed in the patient because of shunt infection diagnosed at the shunt revision performed immediately after the recordings shown in in upper left. When the external drain is open, supratentorial pressure is lower (baseline 4 mm Hg) than infratentorial pressure (baseline 7

mm Hg). When the external drain is closed, progressive inversion of the pressure gradient is observed (supratentorial pressure 18 mm Hg, infratentorial pressure 14 mm Hg). Lower Left: Recordings obtained 3 days after those shown in upper right. A new ventriculoperitoneal shunt has been implanted. Supratentorial pressure (5 mm Hg) is lower than infratentorial pressure (9 mm Hg). Lower Right: Recordings obtained on the same day as those shown in lower left. The pressure differential across the tentorium is increased when the patient is in the upright seated position. Parinaud's syndrome persisted for 3 months after shunt revision, but the patient did make a complete recovery.

Two patients (Cases 19 and 22) underwent simultaneous supra- and infratentorial ICP recordings after an external ventricular drainage system had been placed for shunt infection. The zero reference level of the external drain was placed at the level of the forehead in a patient who was lying down. Both cases showed lower supratentorial pressure when the external drain was open (12 mm Hg and 4 mm Hg, respectively) with a transtentorial gradient of 3 mm Hg in both cases. After the external drain was closed, an inversion of the pressure gradient was observed, with the supratentorial pressure becoming higher (23 mm Hg and 18 mm Hg, respectively) than the infratentorial pressure and with pressure gradients of 8 mm Hg and 4 mm Hg, respectively. (Fig. 6 upper right)

Third Ventriculostomy

In 11 cases third ventriculostomy was performed to remove an infected shunt (two cases) or to try to resolve the symptoms of the patients. The causes of the hydrocephalus were congenital aqueductal stenosis in five cases, posterior fossa tumor in two cases, vein of Galen malformation in one case, posthemorrhagic hydrocephalus in one case, toxoplasmosis in one case, and meningitis in one case. All patients had undergone surgery several times for shunt revision (mean four revisions; range 1-10 revisions). The mean interval between the first shunt placement and the third ventriculostomy was 80 months (range 18-197 months). In four cases the procedure was performed during ventriculography under radiographic control according to a technique already described;^[24,46] in seven cases it was performed under endoscopic control.

Supratentorial and infratentorial pressures were recorded in one (Case 19) of the eight patients who were treated successfully by third ventriculostomy. The pressure gradient disappeared after the procedure, allowing equalization of infratentorial and supratentorial pressures.

No incidence of morbidity or mortality related to the technique was observed. Complete resolution of all symptoms was observed in all cases, with delays ranging from 1 week to 3 months (mean 3 weeks) after the ventriculostomy. In all cases the shunt was successfully removed and none of these patients has required reoperation after a mean follow-up duration of 81 months (range 4-189 months).

DISCUSSION

First described by Parinaud^[46] in 1883, the paralysis of vertical eye movements is presumed to represent a supranuclear paralysis of vertical gaze. It can result from unilateral^[6] or bilateral damage to nuclei within the midbrain tegmentum, including the rostral interstitial nucleus of the medial longitudinal fasciculus, the posterior commissure, the nuclei of the posterior commissure, the nucleus of Darkschewitsch, and the interstitial nucleus of Cajal.^[7,45,48] The most frequent type is paralysis of upward gaze; paralysis of downward gaze and that of both upward and downward gaze are less frequently observed.^[16,48,53,57]

The association of paralysis of upward gaze, also known as Parinaud's syndrome, with pupils that react

better to an accomodative stimulation than to a light stimulus, and with retraction-convergence nystagmus, lid retraction, and possible disjunctive eye position and abnormalities on convergence is also called Koerber-Salus-Elschnig or sylvian aqueduct syndrome.[10,15,27,35,36,44,52] It is induced by the involvement of the periaqueductal gray matter and ventral tegmentum[27] and has also been described as dorsal midbrain syndrome[13,14,55] or pretectal syndrome.[11,28]

During shunt malfunction in patients affected by aqueductal stenosis, this syndrome can be associated with more complex features such as parkinsonian syndrome,[3,5,8,21,56] akinetic mutism,[5,12,21,41] memory disturbances,[12,21] staring gaze as a consequence of paralysis of the reflexes of fixation and pursuit,[10] pyramidal signs,[3] and coma.[3,12,21] This clinical entity, well known in the neurosurgical literature, was defined by Barrer, et al.,[3] as global rostral midbrain dysfunction on the basis of two clinical observations. All the clinical signs can be explained by the progressive involvement of anatomical structures located in the upper midbrain (Fig. 7), such as the substantia nigra, the nigrostriatal and nigrocortical connections,[5] the Wernekink's decussation of the superior cerebellar peduncles, the mammillary bodies and mammillothalamic tract, the superior colliculus,[10] the cerebral peduncles,[3] and the dorsal raphe nuclei of the midbrain reticular formation.[20,22]

Fig. 7. Schematic representation of the anatomical structures of the midbrain and the upper

pons. Upper: Section obtained at the junction between the mesencephalon and diencephalon. 1 = pulvinar; 2 = red nucleus; 3 = substantia nigra; 4 = pineal gland; 5 = posterior commissure; 6 = pretectal area; 7 = posterior commissure nuclei; 8 = sylvian aqueduct; 9 = nucleus of Darkschewitsch; 10 = interstitial nucleus of Cajal. Center: Section obtained at the level of the superior colliculus. 2 = red nucleus; 3 = substantia nigra; 11 = dorsal tegmental decussation; 12 = tectospinal fibers; 13 = medial longitudinal fasciculus; 14 = ventral tegmental decussation. Lower: Section obtained at the level of the inferior colliculus. 3 = substantia nigra; 15 = periaqueductal gray matter; 16 = nucleus of the inferior colliculus; 17 = decussation of the superior cerebellar peduncles.

Pathophysiological Theories

Before the identification of the anatomical structure responsible for vertical gaze,[7,48] some authors proposed that cystic dilation of the suprapineal recess with herniation in the quadrigeminal cistern, frequently observed in obstructive hydrocephalus[32,37,43] and dilation of the upper portion of the aqueduct rostral to the obstruction,[29] could distort and compress the tectal plate where the center for vertical gaze was thought to be located.[9,18,38,39,49,54,61] Other theories included axial enlargement of the third ventricle leading to stretching of the posterior commissure,[12] axial enlargement of the third ventricle associated with distortion and caudal displacement of the mesencephalon,[3] dilation of the rostral portion of the aqueduct with distortion and stretching of the periaqueductal gray matter,[37] and gliosis of the periaqueductal gray matter.[37] Upward gaze palsy can also be observed in cases in which supratentorial space-occupying lesions induce posterior herniation of the temporo-occipital lobes. This can lead to compression and axial pear-shaped distortion of the dorsal midbrain with the onset of mesencephalic signs;[32] the same mechanism has been posited[12] to explain these symptoms during shunt malfunction. Most of these theories have had to be revised since the identification of the anatomical structure responsible for upward gaze, which is located in the periaqueductal gray matter ventral to the aqueduct, in the dorsal interstitial nucleus of the medial longitudinal fasciculus.[7,48]

The region of the upper brainstem as well as all structures located in or around the notch of the tentorium are known to be subject to significant anatomical modifications due to pressure variations across the tentorium;[4,9,18,23,29-33,37-39,42,43,49,58,60,62] this is typical in obstructive hydrocephalus due to aqueductal stenosis. All these changes can be explained by the existence of a long-standing pressure gradient across the tentorium, with higher pressure levels in the supratentorial compartment and lower pressure levels in the posterior fossa.[40,63]

After insertion of a shunt, these anatomical modifications resolve completely but may be overcorrected by continuous CSF drainage through the shunt.[17,30] At the time of shunt malfunction, the dilation of the supratentorial ventricular system produces the same type of anatomical distortions observed before shunt implantation, although to a lower degree.[12] The rapid onset of these distortions occurring during shunt blockage, with sudden stretching and compression of these vulnerable midline structures, could explain the observation that, in patients affected by triventricular hydrocephalus, upward gaze paralysis and other midbrain signs are more frequently observed at the time of a shunt malfunction than at the time of the first diagnosis.[1,26,37,59]

Transtentorial Pressure Gradient

In communicating hydrocephalus, the CSF hydrostatic pressure is equally distributed throughout all the compartments of the central nervous system.[40] There is no development of an abnormal stress within the brain parenchyma. Cerebrospinal fluid pressure can thus reach high levels, producing only symptoms of

intracranial hypertension without any other focal neurological sign. In contrast, when stress develops within the brain because of an expanding mass lesion, intracranial hypertension is accompanied by anatomical deformations and brain herniation.[64] Kaufmann and Clark[34] showed that a pressure differential of 10 mm Hg or more between the ventricles and the spinal subarachnoid spaces was necessary to produce fatal herniation in patients with head injury or intracranial space-occupying lesions.

Increased ICP in obstructive hydrocephalus is different from these two situations because a CSF pressure gradient exists in the absence of a space-occupying lesion, as demonstrated experimentally in dogs by Lim and coworkers[40] and clinically in a case reported by Chattha and Delong.[10] Nevertheless, this pressure gradient cannot exceed the critical value of 10 mm Hg, because the intraventricular pressure is partially transmitted through the brain parenchyma to the supratentorial subarachnoid spaces that communicate with the infratentorial compartment, thus contributing to infratentorial CSF pressure.[40] In fact, in all our cases in which supratentorial and infratentorial ICP recordings were performed (see Table 3), the pressure gradient range was between 4 and 10 mm Hg (mean 5.7 mm Hg) and never exceeded the critical level of 10 mm Hg described by Kaufmann and Clark,[34] even in cases of severe intracranial hypertension (Case 16), thus confirming the experimental data by Lim and coworkers.

Because of this gradient the most important stress would be applied from within the third ventricle on anatomical structures located at the level of the tentorial hiatus that separate the third ventricular cavity from the infratentorial compartment. In fact, the floor of the third ventricle herniates downward into the interpeduncular cistern, the suprapineal recess herniates backward into the quadrigeminal cistern, and the midbrain is pushed downward and flattened. The more severe deformation is observed at the level of the periaqueductal region, with ballooning and funneling of the aqueduct above the obstruction, depression of its floor, severe compression of the periaqueductal gray matter ventral to the aqueduct, and stretching of the posterior commissure.[29,37] This selective deformation of the rostral aqueductal floor, where the rostral interstitial nucleus of the medial longitudinal fasciculus is located, is probably explained by the presence of crossing fibers of the Wernekinck's decussation, the ventral and dorsal tegmental decussation, and the supramammillary commissure. These crossing fibers are all located, in caudorostral order, ventral to the periaqueductal gray matter, and are probably more resistant to stretching and deformation than the complex of small, scattered nuclei of the periaqueductal gray matter located medial, dorsal, and rostral to the compact red nucleus. The focal hyperintensity of the midbrain shown on sagittal T₂-weighted MR imaging sequences demonstrates that focal, reversible damage occurs at this level during shunt malfunction. The rapid resolution of these images following third ventriculostomy could account for the absence of pathological lesions in the midbrain, as reported in some cases in the literature.[12]

Inversion of the Transtentorial Pressure Gradient

Following shunt revision or external ventricular drainage, inversion of the pressure gradient was observed (Figs. 5 lower and 6 upper right and lower left and right), because the infratentorial compartment continued to remain excluded from the supratentorial ventricular system drained from the shunt. In some cases, usually with a short clinical history, shunt revision allowed a rapid improvement in the clinical conditions; however, in most cases in which the diagnosis of shunt dysfunction was delayed because of unusual presentation, improvement was very slow.

In four cases a paradoxical aggravation following shunt revision was observed, and in all cases normal or low supratentorial ICP was found at continuous monitoring or ventricular tapping, and at further shunt revision the shunt was found to be functioning. Similar findings were described by some authors,[19,21,51] who observed a slow progressive deterioration in patients in whom shunts had been

placed for aqueductal stenosis, with the onset of upward gaze palsy, decreased consciousness, and coma some weeks or months following shunt implantation. In all cases repeated supratentorial ICP recordings showed low intraventricular pressure. It is possible that, in these cases, sudden pressure changes at the time of shunt revisions could induce even more functional impairment at the level of the periaqueductal gray matter, resulting in the dramatic clinical pictures observed in some of our patients, similar to the ones described by Galibert, et al.[19] In our experience, it is necessary to wait for progressive and slow improvement despite the very stormy clinical picture. Rekate[51] postulated that decreased brain turgor is at the origin of progressive ventricular dilation with low intraventricular pressure and showed that neck wrapping, by increasing venous pressure and brain turgor, could reestablish normal ventricular size, improving the patient's clinical condition.

Third ventriculostomy is the ideal solution to prevent this type of complication. Reddy and colleagues[50] have performed this procedure via subfrontal approach in five patients affected by slit ventricle syndrome in obstructive hydrocephalus. This procedure creates a communication between the third ventricle and the subarachnoid spaces, thus avoiding the onset of a dangerous transtentorial pressure gradient,[19] as demonstrated in Case 19 of our series. Endoscopic third ventriculostomy, when feasible, is also the best treatment for sylvian aqueduct syndrome or global rostral midbrain dysfunction during shunt malfunction in cases of obstructive hydrocephalus.

CONCLUSIONS

In obstructive hydrocephalus due to aqueductal stenosis, the lack of communication between the ventricular cavities across the tentorium creates a pressure differential between the supratentorial and infratentorial compartments. This is responsible for the significant anatomical distortion of structures located at the level of the tentorial hiatus that is well tolerated because of the slow progression of the aqueductal obstruction. Shunt placement resolves the situation by inverting the pressure differential and reestablishing a normal anatomy.

Shunt blockage can reproduce the original condition, but in a more acute way, inducing distortion and functional impairment of the periaqueductal region in the upper midbrain, where the center for upward gaze and midbrain reticular formation are located. This induces a sylvian aqueduct syndrome possibly associated with memory and consciousness deficits and/or pyramidal and extrapyramidal signs if the whole midbrain is involved.

Lack of improvement following shunt revision can be observed and could lead to repeated shunt revisions without resolution of the clinical picture. Third ventriculostomy allows rapid resolution of the clinical symptoms, reequilibrating pressure levels across the tentorium.

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