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An evaluation of the surgical treatment of intracranial arachnoid cysts in children*,**

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Abstract. In the period 1976–1987, the number of intracranial arachnoid cysts treated at our institute was 60: sylvian, 29; midline supratentorial, 13; subtentorial, 18. The diagnosis was mainly made by means of the results of a combination of CT, dynamic cisternography, and ventriculography. Based on an analysis of the preoperative investigations and operative results, an attempt was made to determine the appropriate treatment more precisely in cysts at different locations. The direct microsurgical approach with membrane excision was mainly used in combination with a preliminary VA shunt to treat hydrocephalus. The direct approach was supplemented with secondary cavity shunting in 5 cases. In more than half of the patients we used membrane excision alone (mainly children with sylvian cysts). In suprasellar cysts we consider the subfrontal approach to be more appropriate than the transventricular one. We restricted the use of primary cyst shunting as an alternative treatment to only 3 infants, with huge cysts. The follow-up reveals that 82.7% of the cases were favorably affected to varying degrees.

Key words: Primary intracranial arachnoid cysts – Children – CSF dynamics – Surgical indications – Operation – Results.

In 1958, Starkman et al. [17] demonstrated that intracranial arachnoid cysts (ICAC) are a separate nosological entity, postulating that they arise from a focal derangement of the leptomeninges. This has subsequently been confirmed by others [6, 12]. The advent of the CT scan in the past 15 years has increased the number of cases detected [3]. Despite the voluminous current literature regarding ICAC, many unsolved problems persist concerning their pathogenetic peculiarities and the proper surgical treatment. The surgical problems amount, in general, to a reasonable definition of the surgical indications and especially to the optimum surgical method. To date, the questions under discussion have evolved around the most reasonable approach – a craniotomy with membrane excision [1, 5, 10], primary cyst shunting [7, 16, 18], or a combination [14]. Some of the details about the surgical technique in different cyst locations and in accompanying hydrocephalus are still not clear. Some authors [8] recommend more flexibility in choosing the surgical tactics. Our department is the biggest pediatric neurosurgical referral center in the country. In the last decade we have had to deal with an increasing number of children with ICAC. In view of the controversies mentioned we tried to analyse our surgical experiences.

Materials and methods

In the period 1976–1987 there were 60 cases with ICAC in the pediatric division of the Department of Neurosurgery, Medical Academy, Sofia. Besides the routine clinical investigations, the diagnostic workup included: pneumoencephalography (7 cases), angiography (45 cases), CT scan (53 cases), radionuclide (^{169}Yb-DTPA or ^{111}In-CaDTPA) cisternography (27 cases), metrizamide CT ventriculography (10 cases), and metrizamide CT cisternography (2 patients). Direct and indirect CT volume measurements [19] were performed in 10 patients with sylvian cysts.

Two patients died before the surgical treatment and the cysts were verified at necropsy. Of the remaining 58 patients, 32 underwent craniotomy alone, 16 had craniotomy and VA shunting, 5 craniotomy with additional shunting of the cavity (in 4 the shunt was placed during a second operation), 3 had primary cyst shunting, and another 3 had ventricular shunting alone (refused further treatment). Only true arachnoid cysts were considered in this series. The follow-up periods varied from 1 to 11 years (median follow-up was 3 years and 5 months).

Results of case analysis

Our ICAC cases amounted to 5.8% of all intracranial space-occupying lesions in the pediatric age group for the same

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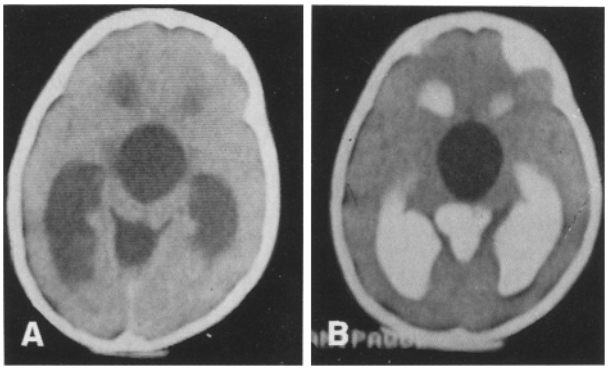

Fig. 1 a, b. A suprasellar cyst associated with acute hydrocephalus: **a** native CT scan; **b** CT ventriculography with metrizamide

period. These cases occur 6.5 times less frequently than brain tumors, but in the age group under 3 this ratio tends to unity (1.2:1). The patients were between 40 days and 13 years old with a distinct predominance of males over females (2:1). More than 68% of the cases were diagnosed at or before 3 years of age. In two-thirds of the cases internal hydrocephalus was present. The location of 60 ICAC is shown in Table 1.

The cases with midline and subtentorial cysts were predominantly infants; all showed non-specific signs of progressive hydrocephalus syndrome and one-third were in poor preoperative condition (signs of herniation) (Table 2). The progression of the symptoms in sylvian cysts can be both sudden (in 7 patients with hemorrhagic complications after minor head trauma) or slow (in 15 patients). The remaining 7 patients demonstrated a non-progressive oligosymptomatic clinical course (chronic headaches and/or seizures); this group is worthy of mention with regard to the definition of surgical indications, especially in the case of a vague dislocation effect on CT and the suspicion of brain agenesis.

Following the van der Meche and Braakman technique [19], we calculated the cyst and brain hemisphere volumes in ten different types of sylvian cysts. Comparison of both hemispheres (Student's *t*-test for paired observations) showed an insignificant difference ($P > 0.01$) between the two sides (mean value of the affected side $98.6\% \pm 4.45\%$). The results from the indirect measurements obtained by calculating the skull "radii" were similar. The "radius" on the cyst side was always greater and the difference between both radii was directly proportional to the cyst size, i.e., the increased hemicranium on the affected side is at the expense of the cyst volume. This strongly suggests that sylvian cysts have a primary space-occupying character and an absence of brain tissue deficiency in the affected hemisphere.

The insufficient diagnostic value of angiography and CT in suprasellar and subtentorial cysts prompted us to use metrizamide CT ventriculography. This was the diagnostic method of choice for differentiating between a suprasellar CSF cyst and an extremely dilated III ventricle (Fig. 1), and between a large retrocerebellar cyst and a Dandy-Walker cyst. The ventriculography demonstrated lack of communication with a blocked III ventricle in four suprasellar cysts; in six investigated subtentorial cysts, there were three isolated cysts with a blocked IV ventricle, two with cyst filling 1 h after contrast application, and one cyst communicated freely with the IV ventricle. The combination of CT ventriculography with dynamic cisternography allowed us to draw some conclusions about the site of CSF pathway occlusion and the relationship between the cyst and CSF spaces.

There were three types of pathological cisternographic findings (Table 3): (1) cysts with early entrance and delayed clearance (suprasellar and subtentorial); (2) cysts with late

Table 1. Location of 60 intracranial arachnoid cysts

Location	Number (%)
Sylvian	29 (48.3%)
Fissura Sylvii	24
Brain convexity	4
Temporomedial	1
Midline	13 (21.7%)
Suprasellar	11
Quadrigeminal	2
Subtentorial	18 (30.0%)
Retrocerebellar	12
Supracerebellar	2
Laterocerebellar	2
Cisterna magna-IV ventricle	2
Total	60 (100%)

Table 2. More frequently observed symptoms and signs in 60 children with ICAC

Symptoms and signs	Sylvian cysts	Midline cysts	Subtentorial cysts	All locations
Headache, vomiting and/or drowsiness	18/29	13/13	14/18	45/60
Increases head circumference	11/29	12/13	14/18	37/60
Psychomotor retardation	12/29	10/13	10/18	32/60
Local skull bulging	23/29	–	5/18	28/60
Focal signs related to the cyst	11/29	1/13	8/18	20/60
Papilledema	7/29	2/13	6/18	15/60
Signs of herniation	3/29	1/13	10/18	14/60
Seizures	5/29	1/13	1/18	7/60

Table 3. Radionuclide cisternography findings in 27 ICAC

Finding	Sylvian cysts (13)	Midline cysts (7)	Subtentorial cysts (7)
Cysts			
– Non-communicating	5	–	–
– Early entrance	1	7	7
– Late entrance	7	–	–
– Delayed clearance	8	7	7
Ventricular reflux	1	2	3
Blocked lateral CSF pathway	10	–	–
Abnormal convexital stasis	8	5	4

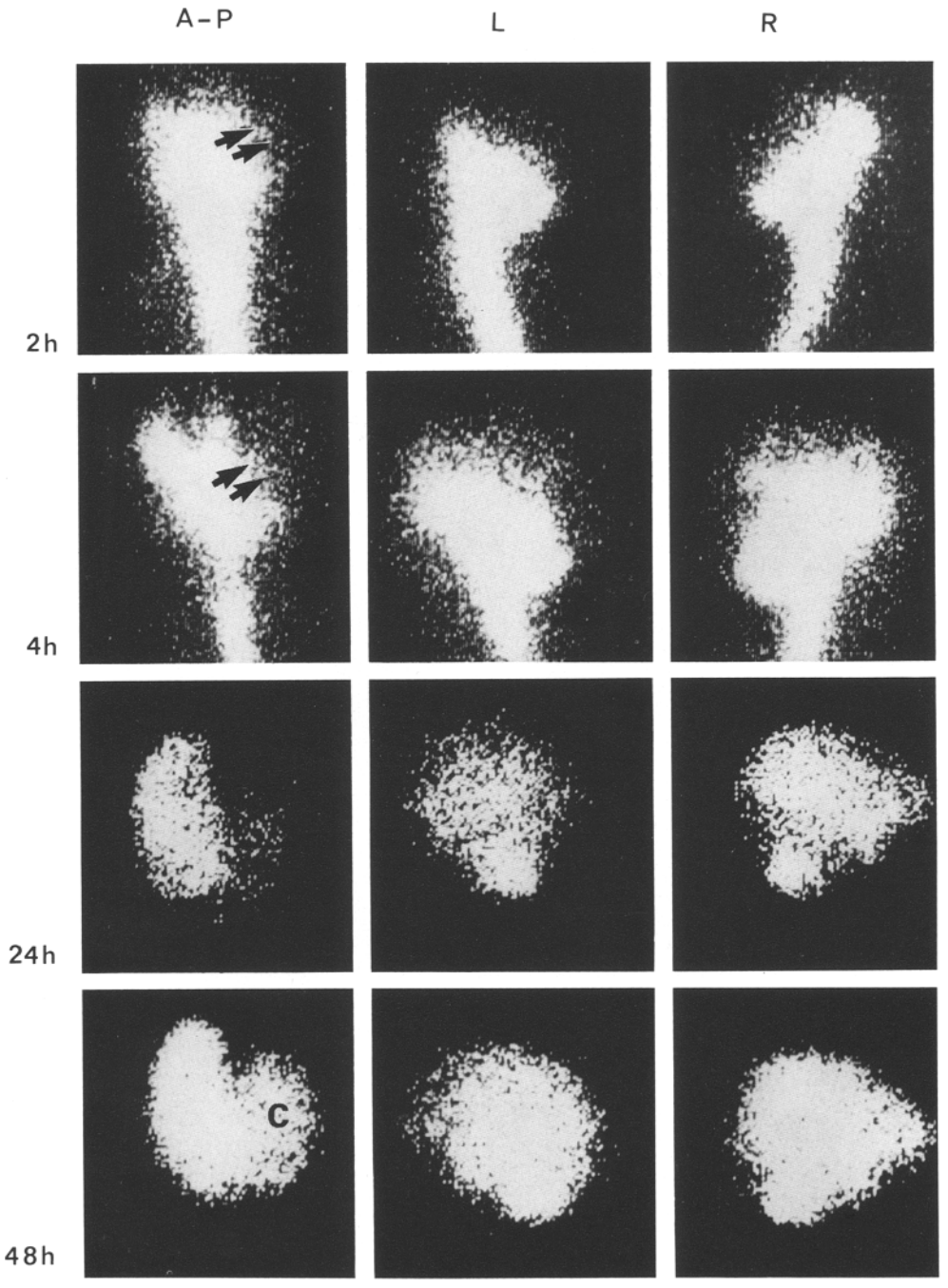

Fig. 2. Radionuclide cisternography in a case with left sylvian cyst. Note the blocked left lateral CSF pathway (*arrows*) and the late entrance of the tracer into the cyst (*c*)

(after 4 h) entrance and delayed clearance (sylvian cysts; Fig. 2); (3) non-communicating cysts (sylvian cysts; Fig. 3). The occlusive character of hydrocephalus was not clearly evident in all of the cases with suprasellar and subtentorial cysts. Elements of both occlusive and communicating hydrocephalus were found to be intermingled.

The direct operation consisted of wide resection of the outer membrane and opening the inner membrane of sylvian cysts to create a communication with the chiasmatic and interpeduncular cisterns (Fig. 4). The same wide opening to the surrounding CSF cisterns was attempted in other cyst locations (Fig. 5). In two patients with suprasellar cysts, we used a transventricular approach and in the remaining nine a subfrontal one. In the former, the cyst was resected in the part prolapsing through the dilated foramen of Monro. Biopsy material from this sector, as well as our sectional observations, indicated a two-layer structure. Over the cyst's dome, there was a neural layer consisting of compressed structural III ventricular elements. One of the patients deteriorated rapidly after the operation and died with signs of untreatable pulmonary edema; the second patient had a complicated postoperative period (transitory diencephalic sopor and paralytic ileus). All of the patients with a subfrontal operation tolerated the operation well.

The applied operative procedures and results are briefly outlined in Tables 4 and 5. To sum the data up, in more than

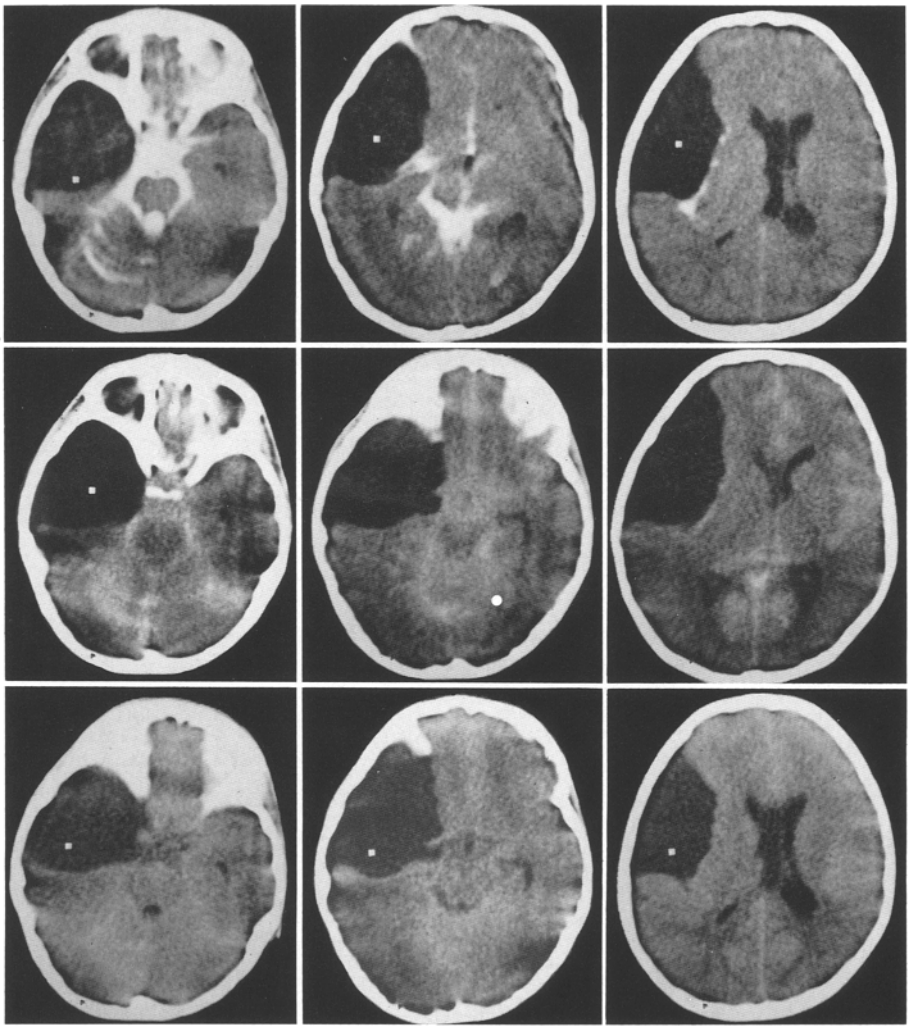

Fig. 3. CT cysternography with metrizamide in a non-communicating sylvian cyst

Table 4. Applied operative procedures and results

	Full recovery	Slight deficiency	Improvement	No change	Death	Total
Membrane excision	17	5	4	5	1	32
Membrane excision and VAA	8	4	1	1	1+1	16
Membrane excision and CAA	2	–	3	–	–	5
CAA	1	1	1	–	–	3
VAA	–	–	1	1	–	2
Total	28	10	10	7	2+1	58

Table 5. Results from the operative treatment of ICAC in children

Outcome	Number	(%)
Full recovery	28	(48.3)
Slight neurological deficiency	10	(17.2)
Improvements	10	(17.2)
No change	7	(12.1)
Deaths	2^{+1a}	$(3.5^{+1.7a})$
Total	58	(100)

[a] A patient with brain tumor accompanying the cyst

half of the cases we used only the direct approach; and these cases were mainly children with sylvian cysts. Three little babies with huge cysts were primarily treated with cyst shunting, which resulted with satisfactory control of their intracranial hypertension. Only two of five patients profited after combination treatment (secondary cavity shunting); these were cases with sylvian cysts. Initially, our treatment of cases with midline and subtentorial cysts involved attacking the cyst first and implanting a ventricular shunt only if the hydrocephalus was found to be unaffected. In the course

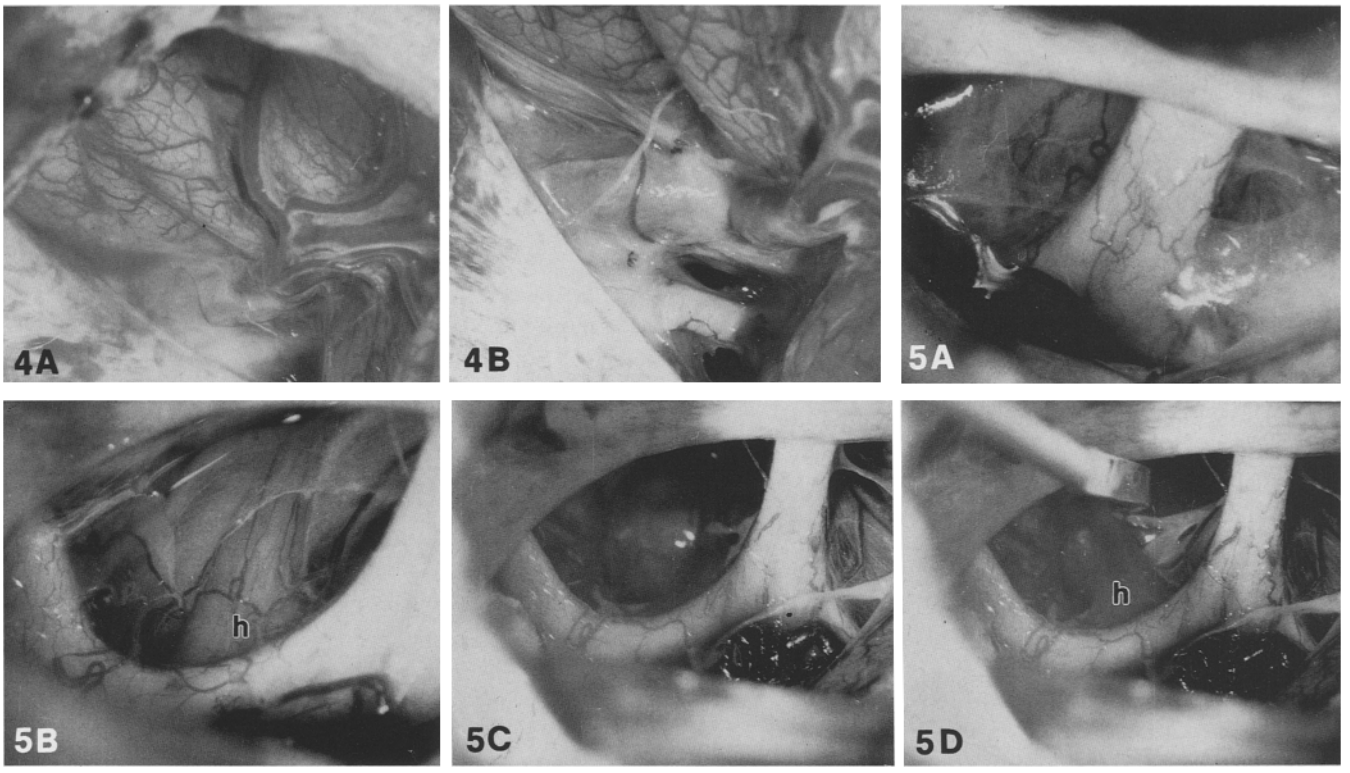

Fig. 4A, B. Intraoperative view into the thickened internal cyst membrane of a sylvian cyst, which separates the lumen from the basal cisterns: **A** before and **B** after creation of communication

Fig. 5A–D. Intraoperative photographs in suprasellar arachnoid cyst, which elevates and elongates the optic structures and the pituitary stalk (*h*): **A, B** and **C**, before cyst resection; **D**, membranes have been resected

Table 6. Combined surgical treatment of ICAC and results

Type of operation	Outcome	Full recovery	Slight deficiency	Improvement	No change	Deaths
Membrane excision						
Membrane excision and VAA						
Membrane excision and CAA						
CAA						
VAA						

of time, this treatment changed. We now consider that ventricular shunting should precede the craniotomy. As seen from Table 6, the improvement in results is related to shunting that precedes the craniotomy.

In 7 of 26 implanted shunts, different postoperative complications were seen; one patient with subdural bleeding after the craniotomy had to be reoperated upon. There were 3 postoperative deaths in this series, which (excluding the child with malignant brain tumor) gives a mortality of 3.5%. The clinical results do not differ essentially in cases with complete and incomplete reexpansion of the brain parenchyma, seen on postoperative CT follow-up.

Discussion

The results in pediatric ICAC series are known to be inferior to those in adults because of the prevalence of sylvian cysts, the fact that these cysts are more amenable to surgical treatment, and because of the more complicated surgical problems associated with hydrocephalus in young children. Over one-third of our cases with midline and subtentorial cysts were in poor preoperative condition and two children died before treatment. Taking into account the risk of secondary bleeding after mild head trauma (in one-fourth of our sylvian cysts) and the high incidence of hydrocephalus in our series, we are of the opinion that the surgical indications should be expanded to almost every detected case in childhood. Comparison with other series with similar clinical material and surgical methods [5, 8–10] shows that our results tend to have optimum values.

The preoperative complex diagnosis is, in our experience, a very important step in the treatment and must solve two major problems: (1) finding the nosological characteristic of the cyst and its space-occupying effect and (2) making clear the real nature of disturbances in CSF-dynamics and the pathogenetic peculiarities of different cyst locations. It facilitates the differentiation of some sylvian cysts from compensatory CSF collections in destructive brain lesions. We found cisternography and ventriculography to be very useful in distinguishing suprasellar cysts from a dilated III ventricle, as well as huge retrocerebellar cysts from Dandy-Walker cysts. These methods enabled us to observe the relationships between the cyst and the CSF pathways more precisely. Such an approach in planning the surgical treatment reflects the current trends reported in the literature on ICAC [14, 15]. Stein et al. [18] pointed out the risks of brain dislocation after ventricular shunting in non-communicating cysts. They proposed the necessity for preoperative investigations of the CSF dynamics. Before proceeding with primary shunting in our three cases with huge cysts and hydrocephalus, we had to prove their communication with the surrounding CSF spaces.

Classification of the cysts in different cisternographic types was also done by other authors [13, 15]. We observed no cysts with early entrance and early clearance. Both cysts with early and delayed entrance of the tracer showed pathological stasis of the diagnostic agent. Like Sato et al. [14], we consider prolonged retention of the CSF tracer, which has penetrated the cyst, to be an indication of slowly increasing CSF volume. This results in progressive mechanical obliteration of the surrounding subarachnoid spaces and in a non-communicating cyst. There is still no reasonable explanation as to how this penetration occurs – through the thin, semipermeable membranes or through a small, "valvelike" orifice. Moreover, ultrastructural data for active secretional properties of the membranes cannot be found [12].

The decision for surgery is facilitated in types II and III cysts with brain dislocation on CT scan, irrespective of the clinical condition of the patient. In oligosymptomatic cysts with vague CT dislocation, we advocate operation if cisternography reveals one of the types mentioned. Based on interpretations of the data, we are of the opinion that the relationships between the suprasellar cysts (together with the basal cisterns) and the ventricles resemble two isolated CSF systems. Two pathogenetic mechanisms are probably involved here – obstruction of the basal CSF flow beyond the cyst, which acts as a subarachnoid diverticulum, and compression on the III ventricle. Thus, resection of the cyst membrane can both restore the CSF circulation in the rostral direction and relieve pressure on the III ventricle. Similar considerations are valid in subtentorial cysts, given that more varied pathogenetic relationships exist in some cases.

Di Rocco et al. [4] presumed that defective CSF absorption or development obliteration of the subarachnoid spaces in ICAC was a cause of hydrocephalus, whereas Galassi et al. [5] stressed the mechanical effect of the cyst. Taking into account our cisternographic findings, we agree with Binitie et al. [2] that the relationships between the cyst and accompanying hydrocephalus are more complex; both are likely to occur as a result of developmental damage to the subarachnoid spaces. This conclusion is supported by the observation that in some of the cases cyst resection does not affect the hydrocephalus. Because of uncertainty as to the exact pathogenesis, it seems more reasonable to attack both the cyst and hydrocephalus. The direct approach enabled us to remove the separating membranes, verify the lesion, and inspect the underlying brain. In sylvian cysts, this also has an important preventive value with respect to the hemorrhagic complications that such cysts tend to produce.

We consider primary cyst shunting to be a palliative procedure for it is directed solely at decompression as long as the shunt functions normally. Considering the shunt dependency for an indefinite period of time, the frequent shunt complications, the impossibility to verify the lesions, and the inapplicability when hemorrhage has occurred, we restricted the use of this procedure to an alternative operation. The additional implantation of a shunt into the cavity after the craniotomy, and resection only of the external membrane in sylvian cysts, has been used routinely by Sato et al. [14]. They maintain that this has contributed to the complete disappearance of the cyst and to brain reexpansion. We disagree that one should resect only the external cyst membrane. Since it is known that sylvian cysts are intraarachnoid in origin [12], we believe that one cannot expect restoration of the CSF flow rostral to the cyst without opening the inner membrane. Moreover, according to our CT follow-up, the clinical data do not differ essentially in cases with complete and incomplete brain reexpansion.

Some surgical considerations with regard to the different cyst locations should be stressed. In sylvian cysts the key element in surgery is opening the internal membrane; additional cavity shunting should be kept in mind in cases with unsatisfactory clinical and CT results during follow-up. Our unfavorable experience with the transventricular approach in suprasellar cysts with "en bloc" resection of the cyst dome resembles the recent observations of Lodrini et al. [8]. In agreement with Raimondi et al. [11], the subfrontal approach has turned out to be better tolerated. The manipula-

tions are limited to arachnoid membranes, the optic nerves are released from arachnoid adhesions, and the basal CSF circulation is restored. Membrane excision in subtentorial cysts permits restoration of the communication between the IV ventricle and the cisterna magna. In both suprasellar and subtentorial cysts, the results were better when ventricular shunting preceded the craniotomy.

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