

Supratentorial extracerebral cysts in infants and children¹

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SYNOPSIS Twelve cases of supratentorial extracerebral cysts in infants and children are reported. Eight were located in the Sylvian fissure, two in the interhemispheric fissure, and two over the convexity of the cerebral hemispheres. Irrespective of their precise location these cysts, in their common, uncomplicated form, give rise to a clinical syndrome different from that recorded in older patients, with a symmetrical macrocrania of a severe degree unassociated with any neurological signs or abnormalities in psychomotor development. Extensive unilateral transillumination of the skull is common (six cases). These features, in association with specific angiographic and pneumoencephalographic findings, make a preoperative diagnosis possible. Extracerebral cysts (either arachnoidal or histologically more complex) should be distinguished from intracerebral cavities which may closely mimic them, even at surgery. The natural history of infantile cysts is studied and serial head-measurements (pre- and postoperative) are presented in five cases. Insufficient knowledge of the spontaneous course and incidence of complications prevents definite statements on the necessity and type of therapy.

Intracranial, extracerebral cysts in the supratentorial compartment of the skull—often referred to as arachnoid cysts—presenting as space-occupying processes, have long been recognized, both in adults and in children. Few reports, however, deal specifically with the peculiar features of supratentorial cysts of infants and children. In this paper we wish to report our experience with 12 cases of supratentorial cysts in children less than 15 years of age and to comment upon the clinical presentation, diagnosis, natural history, and treatment of these lesions.

METHOD

SUBJECTS Our cases have been collected over a 20 year period (1953-72) in a department of paediatric neurology (Hôpital Saint Vincent de Paul in Paris) and a paediatric neurosurgical unit (Hôpital des Enfants-Malades, Paris). All patients presenting with evidence of a large, fluid containing process, enclosed by thin membranes and located outside the brain parenchyma, above the tentorium cerebelli, have been selected. Suprasellar cysts (Harrison, 1971)

and cysts of the incisura tentorii projecting into the supratentorial compartment of the skull (Katagiri, 1960; Alvord and Marcuse, 1966) have been excluded as they give rise to an entirely different clinical picture. Only those lesions large enough to produce symptoms and signs by virtue of their volume were considered. No effort was made to distinguish completely closed cavities—properly named cysts—from cavities communicating through a small orifice with the subarachnoid space, more correctly called 'pouches', as such a distinction is clinically impossible. Nor did we try to separate histologically verified arachnoid cysts from grossly similar lesions with a different histological picture, as the clinical presentation is identical.

The diagnosis was confirmed by surgery in 10 cases and at postmortem examination in one. The 12th patient was, in all respects, so similar to the others that a clinical diagnosis was deemed warranted. The cases have been divided into three groups on the basis of their anatomical location: (1) cysts of the Sylvian fissure (eight cases) which spread this cleft widely open and cause marked foreshortening of the adjacent temporal lobe as well as elevation of the inferior aspect of the frontal lobe, thus exposing the insula; (2) parasagittal cysts (two cases) located within the interhemispheric fissure, between the inner aspect of the frontal and parietal lobes, which are pushed lateralwards and downwards, and the falx

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TABLE
SUMMARY OF CASES

Obs.	Sex	Age	Localization	Initial complaint	Signs + Symptoms			
					Neurol.	Eye	Convulsions	Transillumination
1. M.R.	M	3 a 6 m	Parasagittal L	Large, asymmetrical head since first months	N1 DQ=97	N	0	+
2. S.E.	M	2 a 8 m	Parasagittal R	Large head since birth	N1 DQ=N	N	0	+
3. V.C.	M	10 m	Sylvian L	Generalized seizures since 4 m	N1	N	+ general	ND
4. T.F.	F	2 a 4 m	Sylvian L	Fortuitously discovered 'lump'	N1 DQ=N	N	0	ND
5. S.J.F.	M	8 a	Sylvian R	Convulsions since age 4 yr, large head since birth (> 3 SD)	N1 IQ=N	N	+	ND
6. V.T.	M	4 a 2 m	Sylvian L	Large head since birth (> 3 SD)	N1 IQ=N	N	0	ND
7. B.O.	M	1 a 2 m	Sylvian R	Large head since birth	N1 DQ=N	N	0	+
8. N.G.	M	5 a 6 m	Sylvian L	Post-traumatic raised intracranial pressure	N1	+	0	ND
9. P.J.L.	M	8 a 4 m	Sylvian R	Raised intracranial pressure	N1	+	0	ND
10. L.V.	M	7 m	Sylvian L	Large head from birth (> 4 SD)	N1	N	0	+
11. A.J.C.	M	5 d	Convexity occipital L	Large head (> 3 SD)	Hypotonia	N	0	+
12. G. Val	F	1 a	Convexity parietal L	Large head asymmetrical	Hypertonia	N	0	+

ND: not done. R: right. L: left.

* See text.

Radiological signs			Special investigations	Histology	Treatment	Course
Plain films	Angiograms	Pneumoencephalograms				
Enlarged L hemi-cranium, thin calvarium	Typical*	R lateral ventricle shifted to R, L not demonstrated	EEG: hypo-activity frontal R	ND	Uncapping	Smooth. At 9 yr 4 m head > 4 SD
Enlarged R hemi-cranium, spread sutures, lesser wing elevated	Typical*	ND		ND	Uncapping	Smooth
	ND	Normal ventricular system	EEG: generalized slow waves and spike waves	ND	Puncture (burr hole)	Smooth (not followed-up)
Enlarged L temporal fossa with thin bone	Typical*	ND	EEG: hypo-activity L temporal	ND	Uncapping	Smooth, normal at 10 yr
Enlarged R hemi-cranium, lesser wing elevated	Typical*	Ventricular system shifted to L, temporal horn elevated	EEG: hypo-activity R temporal generalized slow waves	ND	Uncapping	Smooth. Seizures disappeared 5 yr later EEG normalized
Enlarged L hemi-cranium, lesser wing elevated, spread sutures	Typical*	Ventricular system shifted to R, cyst visualized	EEG: normal, brain scan: slight L hyper-activity	ND	Uncapping opening into the basal cisterns	Complicated. Subcutaneous effusion chronic. At 8 yr 10 m head > 3 SD
Enlarged R hemi-cranium, lesser wing elevated, spread sutures	ND	Ventricular system shifted to L	EEG: hypo-activity fronto-temporal R	Normal arachnoid	Uncapping	Complicated. Subdural bleeding.* Repeated seizures 18 m after operation
Enlarged L temporal fossa, acutely spread coronal suture	Typical* vessels distant from inner table	ND	EEG: bilateral slow waves	Thickened arachnoid	(1) Subdural haematoma (burr hole). (2) uncapping cyst	Sudden death 12 hr after 2nd operation. No bleeding in cyst. Persistent subdural haematoma
Enlarged R temporal fossa, lesser wing elevated	Typical, gap between cortical vessels and bone	ND	EEG: normal	ND	(1) Subdural haematoma (burr hole). (2) puncture of cyst	Repeated punctures for 1 m, then smooth
Enlarged skull slightly asymmetrical, sutures spread	Typical*	Ventricular system shifted to R, L temporal horn elevated	EEG: hypo-activity fronto-temporal L, brain scan: hypoactivity. Cisternography*		Not operated	2 yr: head > 5 SD neurologically normal
Enlarged L hemi-cranium (posterior part) spread sutures	ND	L lateral ventricle pushed forwards			Not operated	Sudden death during attempted ventriculography
Enlarged L hemi-cranium	Space occupying process L parietal, avascular	Ventricular system shifted to R and pushed down	EEG: normal, brain scan: mild hyperactivity	Cylindrical epithelium with glandular formations. Glia and connective tissue	Opening into L lateral ventricle	3 yr: head > 1 SD above mean IQ = 102, no neurological signs

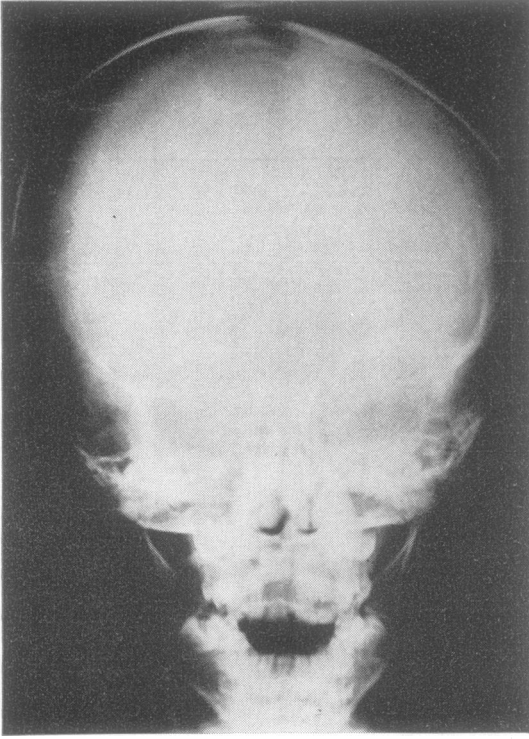


FIG. 1 Plain film of the skull demonstrating generalized enlargement of a whole hemicranium with thinning of bone and chronically spread sutures.

cerebri; (3) cysts of the convexity of the hemispheres (two cases), embedded within the substance of the parietal or occipital lobes, but clearly extraparenchymal as shown by the presence of cerebral cortex underneath the inner wall of the cyst.

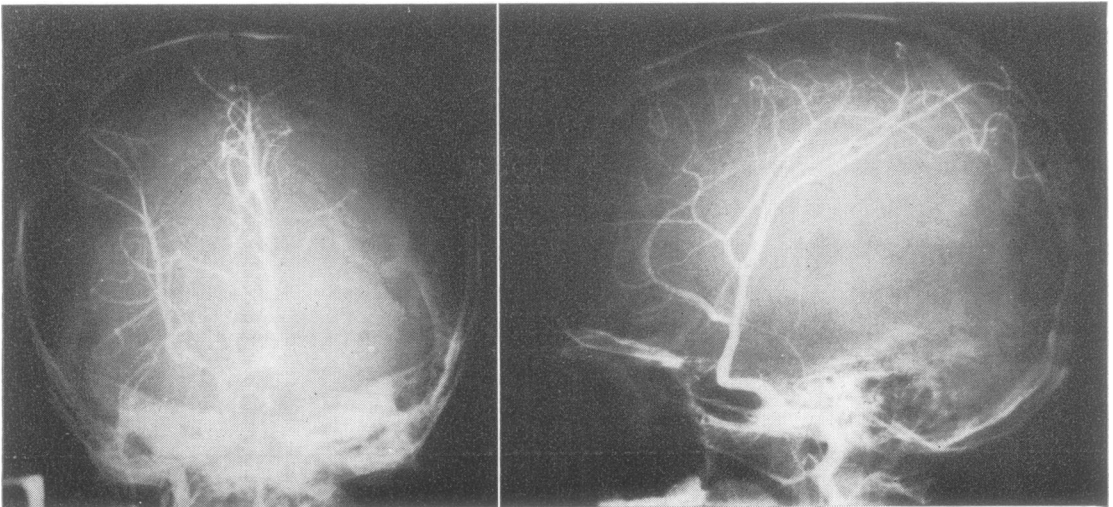
RESULTS

SYMPTOMS AND SIGNS The main characteristics of our patients are shown in the Table.

The age of affected children ranged from five days to 8 years 11 months. The two patients with cysts of the convexity were diagnosed very early (seven days and 1 month), whereas other cases were referred between 7 months and 8 years 11 months. Boys outnumbered girls (10 to two).

Whatever the location of the cyst, the commonest presenting complaint was an enlarged head (more than 2 SD) present in eight cases and, in all cases, dating back to birth or early infancy. In fact, most of the children were referred for evaluation of suspected hydrocephalus. Other presenting complaints included epileptic fits in two cases, symptoms of acutely raised intracranial pressure in two (both complicated with subdural bleeding), and localized bulging of the skull discovered fortuitously in a child of 2 years 8 months.

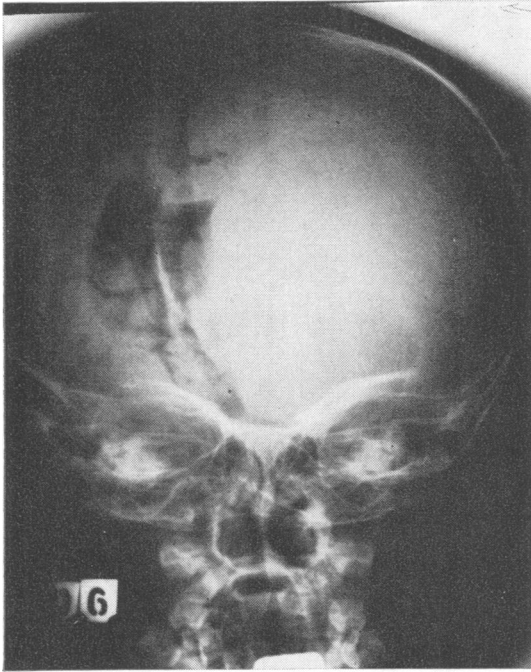
On clinical examination, macrocephaly of a



(a)

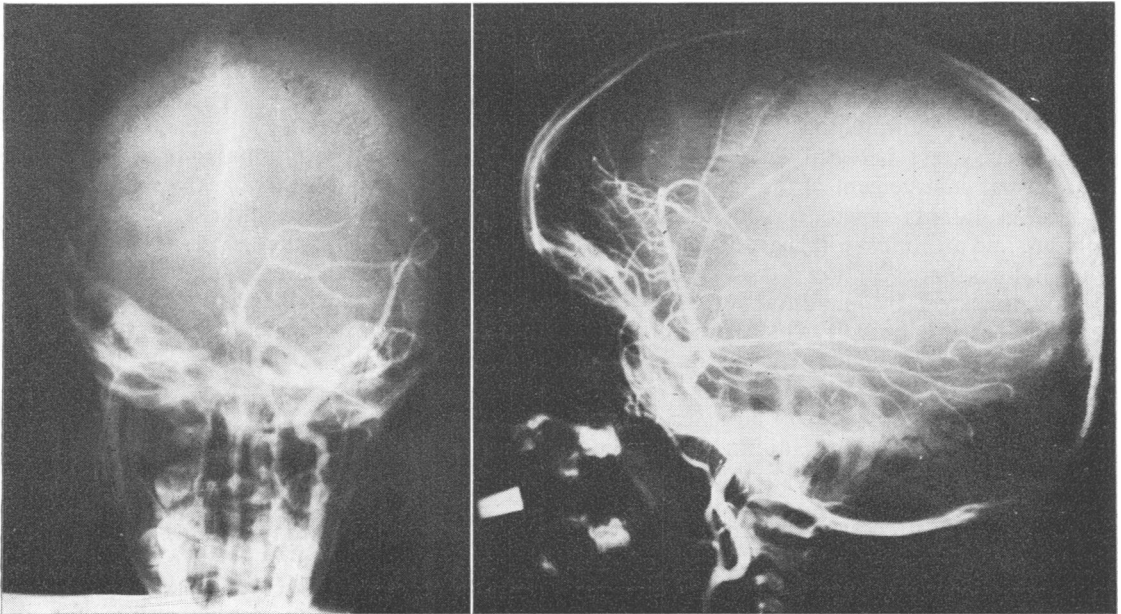
(b)

FIG. 2 Sylvian fissure cyst. Angiography. (a) Almost vertical take-off of middle cerebral artery with shift of the anterior cerebral artery towards the normal side (AP projection). (b) Elevation of the middle cerebral artery (lateral projection).



severe degree (more than 3 or 4 SD) was noticed in seven cases. Asymmetry, with disproportionate enlargement of one side of the cranial vault, was clinically evident in five patients. Localized bulging of the petrous squama without significant head enlargement was present in two cases. Interestingly, transillumination of the skull was positive over the lesion in the six cases in which it was looked for. Diffusion of the light spread over a considerable area, often the whole hemicranium. Localizing neurological signs were remarkably lacking in all cases, even when a complicating bleeding had taken place. Fundal examination showed bilateral papilloedema in the complicated cases only. Mental development was normal in all children. Preoperative electroencephalograms in 10 patients showed reduction of electrical activity over the lesion in five and paroxysmal activity superimposed on a slow background rhythm in two, both with seizures.

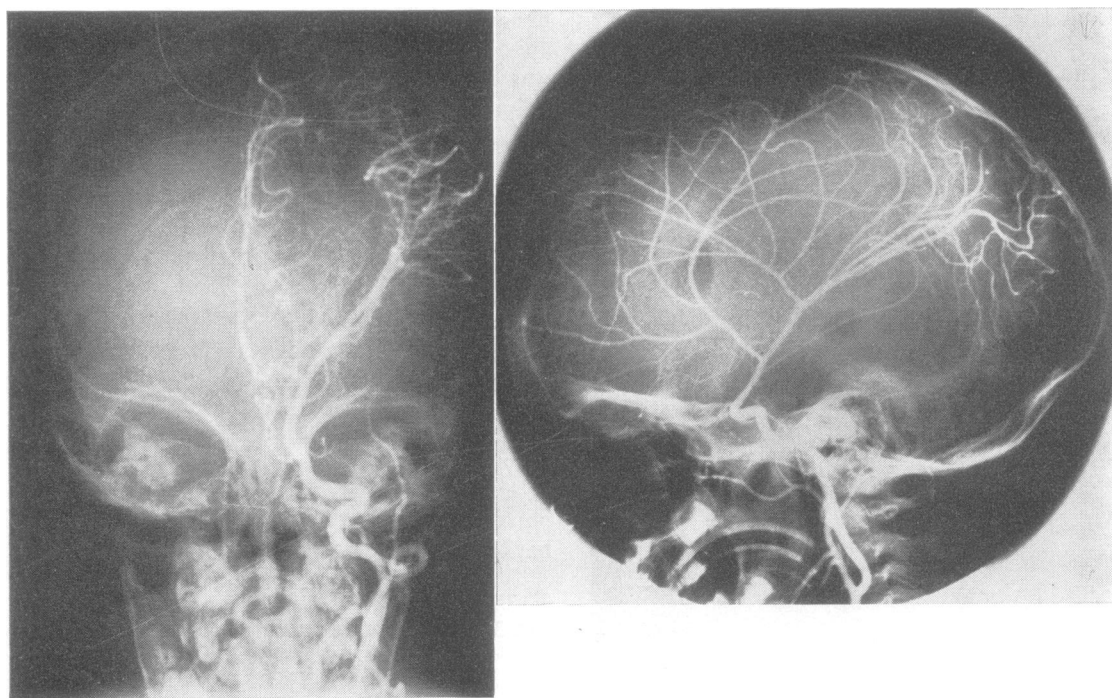
FIG. 3 Giant Sylvian fissure cyst. Pneumoencephalography. The ventricular system is shifted away from the cyst.



(a)

(b)

FIG. 4 Parasagittal cyst. Depression of the initial segment of the middle cerebral artery. (a) AP projection. (b) Lateral projection.



(a)

(b)

FIG. 5 Cystic dilatation of the temporal horn mimicking an arachnoidal cyst. Note the wide sweeping of the anterior cerebral artery on the lateral projection, reflecting generalized dilatation of the lateral ventricle.

In summary, supratentorial cysts offer a remarkable contrast between an easily detected deformity or enlargement of the skull, especially marked in infants where it simulates hydrocephalus, and an almost total lack of neurological abnormalities. An exception is the Sylvian fissure cyst complicated by subdural or intracystic bleeding (cases 8 and 9) which presents with clinical features of acutely raised intracranial pressure. The location of the lesion may be difficult to determine in the case of large lesions and is best approached by radiological studies, which, in every case, are necessary to confirm the diagnosis.

RADIOLOGICAL FEATURES Plain films of the skull showed a thin, outward bulging, petrous squama with elevation of the lesser wing of the sphenoid bone and forwards projection of the greater wing in only three cases. In the remaining patients, plain films were much less suggestive, as

they demonstrated only asymmetrical enlargement of the vault, with thinning of bone, absence of normal convolutional markings on the inner table, and widely spread sutures with long digitations (Fig. 1). Elevation of the lesser sphenoid wing was noted, however, in all Sylvian cysts but one. Contrast studies, on the other hand, permitted precise localization of the lesion in every case. In Sylvian cysts the almost vertical origin of the middle cerebral artery in the anteroposterior projection and its elevation in the lateral projection were highly suggestive (Fig. 2). This appearance, however, is not specific. When combined with a normal-sized ventricular system and caudalwards and inwards dislocation of the temporal horn on pneumoencephalography, it is quite characteristic. It is of note that giant Sylvian cysts in infants may produce a tremendous shift of the ventricular system towards the unaffected side (Fig. 3), as was observed in four cases.

In parasagittal cysts, the angiogram showed a shift of the anterior cerebral artery away from the affected side, while the middle cerebral artery was displaced outwards and its initial segment depressed. A large avascular area was thus created between the two vessels, which, on pneumoencephalography, was shown not to be due to ventricular enlargement (Fig. 4).

Cysts of the convexity gave rise to the picture of an avascular mass lesion in the appropriate area. The mass was shown to be located outside the brain parenchyma by the course of the cortical vessels which curved at a sharp angle at the border of the lesion.

Information provided by contrast studies should be integrated with the results of other methods, to allow for a correct diagnosis. When a typical radiological picture is combined with a positive transillumination, with a silent area on isotopic scan, and with a negative neurological examination, the diagnosis is reasonably assured. In one of our patients (case 10), isotopic cisternography with Yb-DTPA evidenced a total lack of penetration of the tracer into the ipsilateral Sylvian fissure. This technique may thus be of some diagnostic help in addition to its theoretical interest.

OPERATIVE FINDINGS, TREATMENT, AND COURSE

Ten children were operated upon. One (case 10) was left alone in spite of a huge lesion and is doing well except for the large head at 2 years 2 months. One infant died suddenly at 11 days of age during attempted ventriculography. In eight patients operation was decided, despite the lack of symptoms, on account of the large head and the assumed danger of complications. Post-operative course was smooth in six of them. Two children were apparently improved (progressive disappearance of seizures in case 5 and normalization of head circumference in case 12). The remaining four were unchanged. Subdural bleeding developed postoperatively in one patient who finally recovered after a stormy course. A complicated course was observed in another patient, with issue of fluid through the cutaneous scar, necessitating reoperation. In two cases (cases 8 and 9) the basis for operation was subdural haematoma on the same side as a Sylvian cyst, giving rise to raised intracranial pressure. The postoperative course was complicated in both,

one of the patients dying 12 hours after surgery.

Operation consisted in uncapping the cyst in seven cases (with, in one case, an attempt to open it into the optic cistern along the middle cerebral artery), simple puncture of the cyst through a burr-hole in two cases, and opening of the cyst into the lateral ventricle in one.

Histological examination of the cyst membrane, performed in only four cases, revealed simple arachnoid mater in two Sylvian fissure lesions. The two cysts of the convexity had more complex walls, featuring glial and ependymal cells. In one case (case 12) cylindrical epithelium with a few glandular formations was also present, an unequivocal evidence of a dysembryoplastic origin.

COMMENTS

The problem of supratentorial non-tumoural clear fluid-containing cystic cavities has given rise to much confusion. These constitute a heterogeneous group of space-occupying lesions, variously described as benign brain cysts (Handa and Bucy, 1956; Törma and Heiskanen, 1961), cystic hygromas, pseudoporencephalies, or closed porencephalies (Drew and Grant, 1948). More recently, it has become evident that at least part of these cavities belong to a well-defined anatomoclinical entity, usually designated as middle cranial fossa or Sylvian fissure arachnoidal cyst (Starkman *et al.*, 1958; Tiberin and Gruszkiewicz, 1961), but otherwise referred to as middle fossa external hydrocephalus or the temporal lobe agenesis syndrome (Robinson, 1955, 1958, 1964). Whatever the differences in opinion concerning the exact location and nature and mechanism of the 'pouches' or cysts, their clinical and radiological presentation is remarkably uniform. The patients, usually older children or young adults, have discovered fortuitously a localized temporal bulging on one side of the head. There are no neurological signs, except when bleeding within the cyst (Oliver, 1958; Robinson, 1964; Pichard, 1968) or in the subdural space (Robinson, 1958, 1964; Huber, 1961; Pichard, 1968) has taken place. Plain skull radiographs and neuroradiological examinations allow an unequivocal diagnosis (Childe, 1953; Robinson, 1964; Berkmen *et al.*, 1969). More rarely, cysts with a similar gross appearance have

been reported in other locations. A number of these have also been asymptomatic, but others have produced, at various ages, clinical symptoms and signs, most often those of a space-occupying lesion. The most common location has been the interhemispheric fissure (Okonek, 1938; Zehnder, 1938; Van Den Herrewegen, 1954; Roger *et al.*, 1964; Jakubiak *et al.*, 1968; Bhandari, 1972), but a few cases of cysts overlying the convexity of the hemispheres are on record (Bernard *et al.*, 1966; Wilson and Bertran, 1966; Berkmen *et al.*, 1969; Ghatak and Mushrush, 1971; Ryvicker and Leeds, 1973). The clinical and radiological features of such lesions are less stereotyped than those of Sylvian fissure cysts but are nevertheless very suggestive and have been reviewed recently (Bhandari, 1972).

Cysts presenting in this typical way, have been encountered in this series (cases 4 and 5). In a majority of our patients, however, particularly in infants and younger children the clinical and radiological features were quite different. The usual presenting complaint was an excessive size or rate of growth of the head and 10 of our 12 cases were referred for evaluation of suspected hydrocephalus. This agrees with the experience of Anderson and Landing (1966). Six of their eight patients had excessive head size. Macrocrania was also mentioned by Nagoulitch and Perovitch (1960), Roger *et al.* (1964), Vigouroux and Choux (1966), Bhandari (1972), in cysts of the Sylvian fissure, as well as in parasagittal cysts (Berkmen *et al.*, 1969) and in cysts of the convexity (Bernard *et al.*, 1966). Almost all reported macrocephalic cysts were in infants and young children and it seems worthwhile to delineate a giant variety of supratentorial cysts, usually recognized at an early age and which simulate hydrocephalus or chronic subdural haematoma. These infantile or giant cysts do not produce localizing neurological signs any more than the classical one. The positive transillumination of the skull over the lesion, which seems to be present in all infants and young children when it is looked for, is important and should not be misconstrued as a sign of subdural haematoma or hygroma. Pressure symptoms such as headache, tense fontanelle, vomiting, or lateral rectus palsy (Anderson and Landing, 1966; Bhandari 1972) may be slightly more common than in smaller lesions and seizures are

occasionally observed (Roger *et al.*, 1964; Baciocco *et al.* 1965; Vigouroux and Choux, 1966).

This clinical picture, dominated by macrocrania, may be seen whatever the location of the cyst. It seems, however, to be more frequent with parasagittal and convexity cysts, where it is the rule, than with Sylvian fissure cysts (nine of 42 cases under 15 years reviewed). Because of the huge size of the collections, it is often difficult to ascribe to them a precise location, since in many cases they spread out of the Sylvian or interhemispheric fissure to overlie a large part of the hemispherical convexity (Roger *et al.*, 1964). The radiological features of the giant infantile cysts differ largely from those of the classical form. There is usually no localized bulging of the temporal squama but rather expansion of the cranial vault over the whole middle fossa and, often, over the whole hemicranium, with thinning of bone and absence of the inner table digital markings (Fig. 1). In almost all patients signs of chronically raised intracranial pressure are evident, especially chronic spreading of the sutures with long digitations. Again, a precise localization is not obvious from plain films on account of the huge size of the collections. Contrast studies, however, still allow of a correct diagnosis. The main angiographic findings of arachnoidal cysts do not depend on the size of the collection and the characteristic vascular displacements are evident even with huge lesions (Fig. 2). Nonetheless, some angiographic features deserve comment. There is often a marked or even tremendous shift of the anterior cerebral artery away from the affected side in the absence of a complicating subdural haematoma, contrary to the statement of Huber (1961) and Robinson (1964). An extensive gap is common between the cortical arteries and the inner table of the skull, the result of the collection spreading up from the Sylvian fissure or hanging down out of the interhemispheric fissure. However, in our experience, there is always a zone, however narrow, where the vessels reach the inner table, except when there is a complicating subdural haematoma (our cases 8 and 9).

Pneumoencephalography similarly demonstrates the extreme shift of the ventricular system towards the healthy side. Contrary to what is observed in classical cysts, there may be an appreciable degree of ventricular dilatation in

giant lesions, as noted in our cases 1 and 6, and in one patient of Roger *et al.* (1964), possibly due to the distortion of cerebrospinal fluid channels.

The differential diagnosis of giant infantile supratentorial cysts is mainly that of a large head. The common types of hydrocephalus are easily excluded by clinical and radiological examination. On the other hand, several lesions which can produce, in infants, large asymmetrical head and extensive transillumination should be discussed. Chronic subdural hygromas are usually associated with neurological signs, mental retardation, and seizures. There is often a history of trauma to the head in the first months. This is in contrast with the normal mentality, lack of neurological defects, and of previous history of trauma which is the rule in arachnoid cysts. The angiographic picture is quite different. The same would apply to the peripheral sub-arachnoid collections of normal cerebrospinal fluid communicating with the ventricular system through a small opening of vascular origin described by Freeman and Gold (1964).

Intracerebral cysts or huge cystic dilatation of the temporal horn, usually accompanied by less marked dilatation of the rest of the ventricular system, has proved to be the most difficult diagnostic problem as it presented in four of our patients. In two, the bulging was mainly temporal or parietotemporal and the angiographic picture was identical with that of a Sylvian cyst, except for the wide sweeping of the anterior cerebral artery on the lateral projection, due to generalized ventricular dilatation (Fig. 5). In the remaining patients, the whole hemisphere was involved. In all patients, head circumference was between 2 and 5 standard deviations above the mean. Pathologically these lesions are either extreme dilatations of one part of the ventricle or porencephalic cysts communicating with the ventricle or, rarely, paraventricular diverticula associated with hydrocephalus.

In one of our infants the external wall of the lesion was reduced to a thin membrane and the surgeon thought he had entered an arachnoid cyst until he discovered the open temporal horn and choroid plexus. The mechanism by which such atrophic lesions produce an expansion of the skull remains obscure, but the fact has been already discussed (Pendergrass and Perryman,

1946; Childe, 1953; Huber, 1961), although a temporal location appears rare. Another possible mechanism for cystic expansion of the temporal horn is atresia of the lateral ventricle (Cloward, 1969). This abnormality was not encountered in our patients, however.

Whatever the mechanism, these intracerebral 'cysts' can be distinguished from extracerebral cysts by some clinical and radiological features which are quite unusual in the latter. These are (1) focal, neurological signs, mainly hemiparesis, hemiatrophy, or unilateral pyramidal tract signs; (2) mild to moderate mental retardation; (3) wide sweeping of the anterior cerebral artery in the lateral projection, reflecting dilatation of the lateral ventricle. Pneumoencephalography would then easily correct the diagnosis by showing the tremendous dilatation of the temporal horn, with less marked enlargement of the rest of the ventricle. This distinction is of practical prognostic and therapeutic importance.

NATURE AND MECHANISM The study of the giant infantile cysts adds little to our knowledge of the mechanism and origin of these lesions. In our cases, as well as in other published series, two main histological types were encountered; simple arachnoid cysts (Starkman *et al.*, 1958) and cysts with more complex walls featuring neuroglia, ependymal lining, and, sometimes, other tissues (Jakubiak *et al.*, 1968). All Sylvian fissure cysts studied have been of the simple arachnoid type (Oliver, 1958; Starkman *et al.*, 1958; Tiberin and Gruszkiewicz, 1961; Robinson, 1964; Gruszkiewicz and Peyser, 1965; Holst, 1965; Weinman, 1965; Wilson and Bertran, 1966; Jakubiak *et al.*, 1968; Pichard, 1968; Bhandari, 1972); so that the Sylvian cyst syndrome seems as well defined histologically as clinically. Cysts of the inter-hemispheric fissure and of the convexity, on the other hand, have been more variable, many of them containing an ependymal lining and neuroglial remnants (Okonek, 1938, our cases 11 and 12; Jakubiak *et al.*, 1968, cases 1 and 2; Berkmen *et al.*, 1969; Bhandari, 1972). A minority seem to belong to the simple arachnoid type (Van Den Herrewegen, 1954; Wilson and Bertran, 1966; Jakubiak *et al.*, 1968, case 3; Ghatak and Mushrush, 1971; Ryvicker and Leeds, 1973). The congenital origin of these lesions is well demonstrated

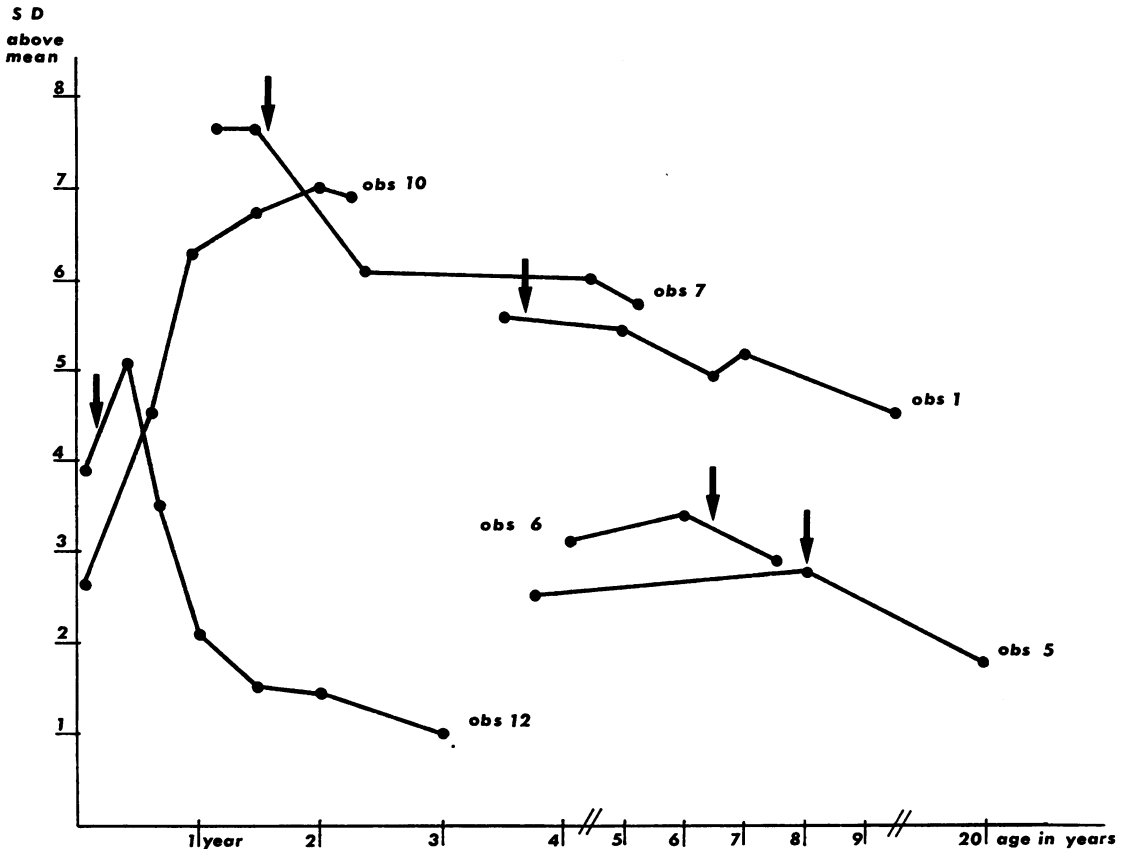


FIG. 6 Rate of growth of the head on serial measurements in five patients. Arrows indicate date of operation.

by the early onset of macrocephaly in many infantile cases. Their slowly growing character is attested by the radiological signs of long-standing intracranial pressure and by the tremendous displacement of brain and vascular structures which must have developed for months or years. There is no complete pathological documentation of a giant cyst, so that the precise relations of the cyst with the arachnoid and subarachnoid space are unknown. There is no doubt, however, that they behave as expansive lesions, due to some kind of a pressure mechanism. In no case did we find a history of trauma, either at birth or prenatally, a finding in contrast with that obtaining in subdural hygromas (Roger *et al.*, 1964).

NATURAL HISTORY AND TREATMENT The choice of treatment for supratentorial cysts is largely

dependent upon the natural history of these collections. Unfortunately, little is known about this, so that the comparative risks of various therapies are not readily assessed.

Supratentorial cysts can be tolerated indefinitely and several cases were incidentally found at necropsy (Davison and Friedman, 1946; Starkman *et al.*, 1958; Robinson, 1964), in subjects without any history of neurological disorder. Furthermore, several patients have been operated on solely on account of a fortuitously discovered skull deformity (our case 4; Bull, 1949; Childe, 1953). Finally, the reason for operation in many cases was the presence of fixed, minimal symptoms and signs, whose relation to the cyst may have been debatable. Thus, part of these collections, even large ones, may remain silent throughout life, but there is no

means of evaluating which proportion belongs in this category.

On the other hand, arachnoid cysts may produce severe symptoms, complications, and even death. Intracranial bleeding has been frequently recorded (Gruszkiewicz and Peyser, 1958; Robinson, 1958, 1964; Tiberin and Gruszkiewicz, 1961; Pichard, 1968) and is exemplified by our cases 8 and 9. It may be intracystic or subdural, sometimes on the contralateral side (Robinson, 1964), and gives rise to an acute neurological disorder. Even when no bleeding takes place, some cysts may produce symptomatic intracranial pressure, either in childhood (Okonek, 1938; Dott and Gillingham, 1958; Tiberin and Gruszkiewicz, 1961; Anderson and Landing, 1966; Bhandari, 1972) or much later (Handa and Bucy, 1956; Ghatak and Mushrush, 1971; Bhandari, 1972), or signs of a focal expanding lesion simulating brain tumour, especially in adults. Convulsive seizures may be related partly to the pressure effect of the cysts, as they may subside after operation (Roger *et al.*, 1964; Vigouroux and Choux, 1966; Bhandari, 1972). In infants, the effects of long-standing pressure with extreme dislocation of the skull contents, as observed in giant cysts, are potentially dangerous but, up to now, largely unknown. Again, it is not possible, from available data to determine the incidence of complicated cases or to know, beforehand, in which cases symptoms are to develop.

In view of the possibility of such serious accidents, most workers have felt that the mere presence of a cyst justifies operation and we adopted this attitude in 10 of our 12 cases. However, much more evidence is needed to consider this policy entirely warranted. We do not know, in fact, whether operation has any value in preventing intracranial bleeding. Nor do we have much information on the fate of the cystic cavities once opened or uncapped. Particularly little is known about giant infantile lesions in which the problem is further complicated by the disproportion between the size of the skull and that of the brain, with the resultant huge post-operative cavities. There is, in fact, some evidence that surgery may not be sufficient to cure these lesions. Bhandari (1972) has reported a case of fatal acute post-traumatic accident with a Sylvian cyst which had been excised 16 years

previously. Other cases of recurrence are on record after various durations (Handa and Bucy, 1956; Anderson and Landing, 1966). Radiological signs of a persisting pouch after excision of the external wall of the cyst have been observed (Tiberin and Gruszkiewicz, 1961; Robinson, 1964). Finally the rate of head growth in giant cysts has never been studied, either with or without surgery. We obtained serial measurements in five children (obs. 1, 6, 7, 10, 12). Figure 6 shows that in three cases the rate of growth was not clearly modified by operation. In case 12, in which a cyst of the convexity was made to communicate with the lateral ventricle, the rate of growth was definitely slowed down. Finally, in the one patient not operated on, the rate of head growth has been faster than in unoperated patients but at a much younger age. In view of these uncertainties, the risks of operating upon these lesions should be carefully weighed. At least four postoperative deaths are known (our case 8; Starkman *et al.*, 1958, case 1; Bernard *et al.*, 1966) and many more postoperative complications or sequelae have been recorded (Okonek, 1938; Robinson, 1964, case 4; Anderson and Landing, 1966, obs. 1, 7, and 8; Vigouroux and Choux, 1966, case 4, our cases 6 and 7; Pichard, 1968, case 2; Berkmen *et al.*, 1969). Infantile giant cysts may be particularly liable to such complications, as depletion of fluid and displacement of the brain in a greatly enlarged skull seem especially serious.

At the present time, surgery is certainly indicated in cysts complicated by haemorrhage, as well as in those cysts that, without bleeding, give rise to pressure or localized signs or symptoms. On the other hand, there is no definite evidence that operation would help asymptomatic patients, and it is thus better withheld in such cases (Robinson, 1964). Such children should be closely followed. Should operation become necessary on account of bleeding or pressure signs, it may possibly be limited to a minimum. Robinson (1964) states that a burr hole suffices to wash out the blood clots in the case of haematoma formation and Handa and Bucy (1956) have treated a probable arachnoidal cyst by simple aspiration repeated three times over a 20 year period. The experience, already cited, of re-growth of operated cysts, as well as the radiological evidence of a persisting pouch after

excision of the cyst wall (Oliver, 1958; Tiberin and Gruszkiewicz, 1961; Robinson, 1964), would seem to favour such a policy.

Because of the lack of definite evidence on many important points, a dogmatic statement on the indications and types of operation appears unjustified. Our personal policy, at present, is to refrain from operating upon asymptomatic cases.

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