

# NEUROLOGICAL SIGNS AND OPERATIVE INDICATION BY AGENESIS OF THE PERISYLVIAN REGION

A STUDY OF 13 OPERATED CASES

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The first signs of Sylvian Depression in embryogenesis may be detected at the end of the second intrauterine month and become more apparent as a Sylvian groove at the end of the third month. The temporal operculum grows more effectively in the anterior two thirds and reaches the Sylvian fissure in the fourth month. At the same time the fronto-parietal operculum becomes evident and gradually extends backwards to meet the anterior part of the temporal operculum. These changes occur in the later half of the fifth month. Because the growth of the temporal operculum is more intense than that of the fronto-parietal one, it follows that, when the opercula meet in the sixth intrauterine month, there is more of the Sylvian area covered by the temporal than by the fronto-parietal operculum. (Fig. 1)

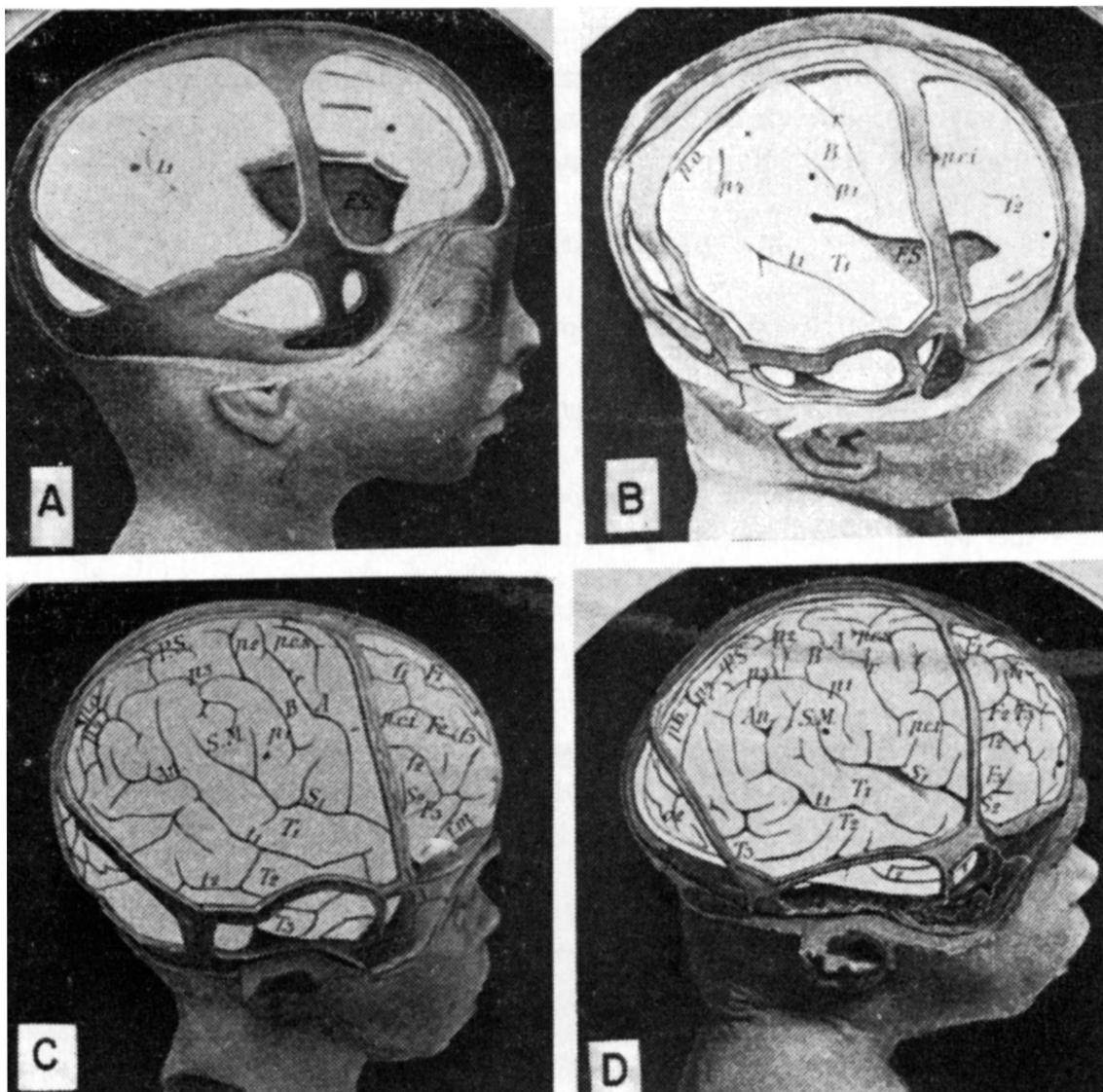
The present syndrome (Agenesis of the perisylvian region) is considered to be a disturbance of this cerebral embryogenesis and becomes evident during the last three months of foetal life. Arachnoid cysts are to be found in the hypoplastic region, sometimes with space occupying character. 70 cases of agenesis of the fronto-temporal area with diagnosis was mostly made during operation. The patients were operated under the tentative diagnosis of a space occupying lesion like subdural haematoma, external hydrocephalus, porencephaly, intracerebral haematoma or tumor.

Before the syndrome of agenesis of the perisylvian region had been completely understood, the following synonyms had been used: cystic pseudotumor cerebri, meningitis serosa circumscripta, relapsing juvenile subdural haematoma, subarachnoid pouch, cerebral arachnoid cyst, chronic subarachnoid cyst and temporal lobe agenesis syndrome.

The outward appearance of the patients is characterized by face and skull asymmetries with depression or elevation of the eyebrow; sometimes other

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*Fig. 1 — A) The temporo-fronto-parietal region at the 4th intrauterine month; B) at the 5th intrauterine month; C) at the birth; D) at the 6th month of life.*

congenital anomalies may occur like broad oculo-facial malformations. Other frequent symptoms are: skull and face asymmetries, localized bulging of the head, enlargement of the skull, exophthalmus, strabismus, hydrocephalic findings, convulsive seizures, retarded psychomotor development. In some cases the transillumination may be positive like in cases of external hydrocephalus. The EEG may show focal abnormalities over the region of the lesion like slow waves or low voltage. In patients with increased intracranial pressure papilledema and unspecific neurological signs may be found.

The typical radiological signs are: rounded and enlarged cranial vault, thinned cranial bone over the lesion, sagittal sinus groove displacement, elevated sphenoid ridge, depressed and elongated floor of the middle fossa. Carotid angiography shows non specific signs like a great space occupying lesion, the temporal area deficient of vessels and sometimes hydrocephalic findings. The pneumoencephalography or ventriculography demonstrates a dilatation, deformity or displacement of the ventricles. Sometimes, as a result of the brain-stem

shifting, an obstruction of aqueduct occurs and leads to hydrocephalus occlusus. The computerized tomography demonstrates a hypodensive region over the perisylvian area (Fig. 2). In cases with space-occupying arachnoid cyst of subdural haematoma the CAT-scan diagnosis may be difficult.

MATERIAL AND METHODS

The summary of the clinical signs in our 13 operated patients demonstrates that:

1. As in the literature (relation 4:1 or 5:1) the male sex dominates in our cases (11:2);
2. The perisylvian agenesis was always left sided, except in 2 cases;
3. There is a high incidence of left handed or ambidextrous patients (Table 1);
4. The arachnoid cysts were in 7 cases localized in the temporal region, in 5 cases fronto-temporal and in one case the whole fronto-temporo-parietal region was taken from a huge arachnoid cyst of 280 ml volume;
5. Five patients decompensated after light trauma, four of them with subdural haematoma. The rest showed increased frequency of convulsive seizures,

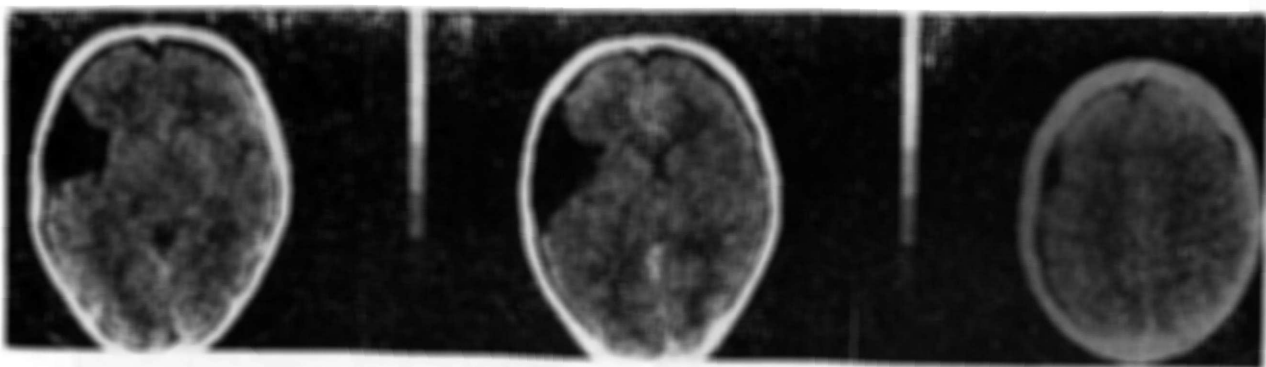


Fig. 2 — Typical computer-tomography findings by agenesis of the perisylvian region.

Age at operation		12	5	6	44	33	2	12	7	42	9	15	3	53	S(13)
Cause of admission	Seizures, EEG-Deterioration				X	X	X	X		X		X		X	7
	Decompensation after light trauma	X	X	X						X	X	X			6
	Neurological signs	X			X	X	X		X	X	X	X			8
	Papilledema	X	X	X							X				4
	Hydrocephalic signs		X	X	X								X		4
Localization of arachnoid cysts	frontal		X					X	X	X		X		X	6
	temporal	X	X	X	X	X	X	X	X	X	X	X	X	X	13
	parietal							X							1
Subdural hematoma		X		X						X		X			4

Table 1 — Age, sex, side of the agenesis and handedness in 13 cases with perisylvian agenesis.

Age at operation		12	5	6	44	33	2	12	7	42	9	15	3	.53	S = 13
Sex	male	X	X	X		X	X	X	X	X	X	X	X		11
	female				X									X	2
Side of Agenesis	right	X												X	2
	left		X	X	X	X	X	X	X	X	X	X	X		11
Handedness	right	X			X	X			X	X		X		X	7
	left		X				X								2
	ambidextrous			X				X			X		X		4

Table 2 — Clinical signs and operative findings in 13 cases with perisylvian agenesis.

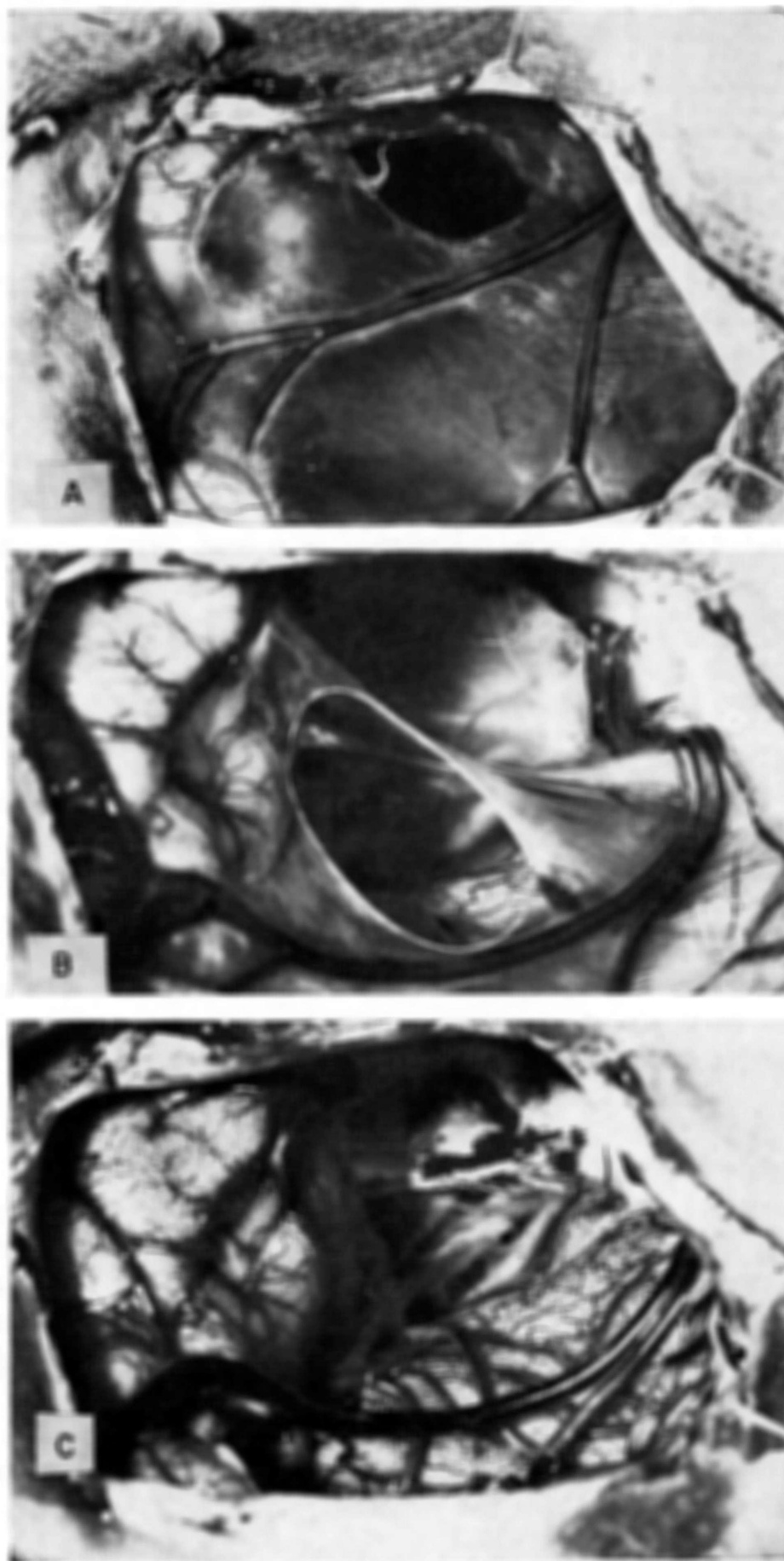
deterioration of EEG findings or general clinical signs of increased intracranial pressure (Table 2).

The psycho-neurological investigation (at the earliest 6 months after operation) showed that right handed patients have always pathological findings especially in visual constructive capacity, speech, reading, perimetry, audiogram and dichotic listening. The left handed are free of those symptoms. In a two years old boy with a preoperative right sided hemiparesis and post-operative neurological defects we assume a so called paretic left handedness. The operative treatment consists in resection of the cyst and cyst walls, communication to the basal subarachnoid space and occasionally implantation of Ommaya-reservoir and catheter in the cyst cavity (Fig. 3).

DISCUSSION

The syndrome of agenesis of the perisylvian region represents an embryogenetic hypoplasia of the whole perisylvian region, not only of the temporal lobe, concordant to the surface anatomy of cerebrum at the fifth to sixth month of foetal life. It is accompanied with space occupying arachnoid cysts. The left sided agenesis and the male sex are predominant. Many left handed or ambidextrous patients are to be found. If the patients become left handed or ambidextrous there are only slight or no neurological defects. Convulsive disorders are frequent. Cerebral decompensation or subdural haematoma after light trauma may occur and are not unusual.

In 6 from 13 of our patients, the indication for operation was post-traumatic decompensation and in 4 from these 6 accompanying subdural haematoma. In the remaining 7 cases the indication for operation was increased neurological disturbances such as in hemisymptomatic, aggravation of convulsive seizures and speech deficiency.



**Fig. 3 — A) The arachnoidal cyst showing the exposed cerebral veins effecting basal drainage; B) and C) agenesis of the whole perisylvian region.**

Some cases of perisylvian agenesis may be accidentally detected by computerized tomography because of non specific symptom, such as headaches and hydrocephalic signs. We operated the cases which showed subarachnoidal cysts with space occupying character. If the subarachnoidal cyst has not the space occupying character, we recommend the regular neurological observation and computer tomographical check-up every 6 to 12 months.

RESUMO

*Sinais neurológicos e indicação operatória em casos de agenesia da região perisilviana: estudo de 13 casos operados.*

O artigo trata de 13 casos de agenesia da região perisilviana em pacientes de 2 a 53 anos. A aplasia foi encontrada com sua maior extensão no lobo temporal em 7 casos, na região fronto-temporal em 5 casos, e um caso abrangendo toda a região fronto-parieto-temporal, todos os casos acompanhados de cisto aracnóideo de caráter expansivo. Também foram encontrados aderências e distúrbios da circulação líquórica a nível das cisternas basais. Em todos os casos, com exceção de dois, localizava-se a aplasia do lado esquerdo; o sexo masculino é predominante (11 casos) atingindo o sexo feminino em nossa série em apenas dois casos. Os pacientes apresentam uma tendência a descompensação após traumatismos cranio-encefálicos leves, e em muitos casos foram encontrados hematomas subdurais ocasionados pela ruptura de veias cerebrais que se encontravam soltas sobre o cisto aracnóideo. Nos casos restantes a operação foi realizada por causa da existência de pressão intracraniana elevada, exacerbação de crises convulsivas ou por causa do caráter expansivo do cisto aracnóideo concomitante. Em pacientes com agenesia da região perisilviana porém com cistos aracnóideos que não apresentam caráter expansivo, não é indicado o tratamento cirúrgico e sim o acompanhamento neurológico e repetição da tomografia computadorizada em espaços de 6 a 12 meses.

ZUSAMMENFASSUNG

Es wird über 13 Fälle mit Aplasie der perisylvischen Region berichtet bei Patienten im Alter von 2 bis 53 Jahren. Die Aplasie betraf das temporale Operculum in 7 Fälle, das frontale Operculum in 5 Fälle und die ganze fronto-parieto-temporale Region in einem Fall. Sie wurde in allen Fällen von raumfordernden Arachnoidalzysten begleitet. Ebenfalls lagen Verwachsungen und Abflussbehinderungen der basalen Liquorräume vor. Zu 85% war die Aplasie linksseitig lokalisiert, ebenso häufig wurde das männliche Geschlecht betroffen. Die Patienten dekomensieren leicht nach einem Schädelhirntrauma, oft findet sich bei der Operation ein den aplastischen Bereich ausfüllendes subdurales Hämatom wegen Abrisses der frei liegenden basal drainierenden Hirnvenen. Bei den restlichen Fällen wurde die Operation wegen allgemeiner Hirndrucksymptomatik, Verschlimmerung eines bestehenden Anfallsleidens oder wegen des raumfordernen Charakters der begleitenden Arachnoidalzysten durchgeführt. Bei Patienten mit perisylvischer Aplasie ohne raumfordernde

Arachnoidalzysten wird eine Operation zunächst nicht durchgeführt, sondern es findet die regelmässige neurologische und computer-tomographische Kontrolle statt.

#### SUMMARY

The article reports on 13 cases of agenesis of the perisylvian region in patients aged 2 to 53 years. The agenesis occurred in the temporal operculum in 7 cases, in the frontal operculum in 5 cases and the whole fronto-parieto-temporal region in one case. The agenesis was always associated with space requiring arachnoidal cysts. Symphyses and obstruction of the flow of the basal CSF spaces were also seen. In 85% of the patients were males. The patients tend to decompensate after light cerebrocranial trauma, and in many cases surgery reveals a subdural haematoma which fills the aplastic region, due to detachment of the exposed cerebral veins effecting basal drainage. In the remaining cases, surgery was performed because of general signs of cerebral compression, exarcebation of an existing disease associated with attacks, or because of the space occupying character of the concomitant arachnoidal cysts. In patients with agenesis of the perisylvian region without space occupying arachnoidal cysts, surgery is not performed for the time being, instead, regular neurological and CAT-Scan control is effected.

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