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# Type I Schizencephaly: CT and Neuropathologic Findings

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**Type I schizencephaly, which is less commonly recognized than the type II variety, consists of a "fused" cleft in the cerebral mantle. Its CT appearance is significantly different from the type II variety, and it may be confused with postdevelopmentally acquired lesions. The CT appearance of eight cases (seven girls and one boy, aged 10 months to 18 years) of type I schizencephaly was correlated with clinical findings and previously recorded neuropathologic features. Presenting symptoms included developmental delay, seizures, and hemiparesis. CT findings in each case showed a unilateral, cortically lined cleft in the parasylvian region extending from the hemispheric surface to the ventricle. Micropolygyria was present in the cortex lining the cleft, and in two of eight patients was also visible elsewhere in the cerebral cortex. Those with associated microcephaly (four of eight patients) had significantly more severe neurologic deficits than did those without microcephaly. CT findings in type I schizencephaly are characteristic and show excellent correlation with neuropathologic specimens. The appearance of these lesions can be explained on the basis of a disorder of neuronal migration in early gestation and they should not be confused with postdevelopmentally acquired lesions.**

In 1946, Yakovlev and Wadsworth [1, 2] described the pathologic findings in a number of patients with clefts in the cerebral wall, which they termed schizencephaly. They divided the patients into two groups: those in whom the walls of the cleft were fused (type I) and those in whom the walls of the cleft were separated, with coexistent "hydrocephalus" (type II). Previous reports describing CT findings in patients with schizencephaly have primarily dealt with the type II variety [3-5]. The purpose of this paper is to describe the CT findings in the less commonly recognized type I variety and to correlate them with the neuropathologic and clinical findings in order to gain a further understanding of this disorder.

## Subjects and Methods

Eight patients with type I schizencephaly were scanned during the past 2 years as part of their neurologic evaluation. The patients were from a total of approximately 5,000 pediatric patients who had head CT scans during that period. A ninth patient was evaluated elsewhere and the postmortem specimen was referred to us for neuropathologic consultation. CT scans were obtained on a GE 9800 scanner in the axial plane with 10-mm contiguous slices before and after administration of IV contrast material (2 ml/kg).

## Results

The clinical and CT findings for each patient are presented in Table 1. These lesions appear as a cortically lined cleft extending through the full thickness of the cerebral mantle (Figs. 1 and 2). The cortex lining the cleft is of normal density but the cortex is thicker than normal in other areas (Figs. 1 and 2). At the ventricular end of the cleft a small triangular diverticulum may be present and gray matter may

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TABLE 1: Clinical and CT Findings in Type I Schizencephaly

Case No.	Age Gender	Clinical		CT				
		Presenting Symptom	Other Findings	Cleft Location	Ventricular Diverticulum	Symmetry	Ventricular Enlargement	Associated Anomalies
1	12 y M	Seizure	Left hemiparesis	Right parietal	Yes	Unilateral	None	None
2	18 y F	Seizure	Left hemiparesis	Right parietal	Yes	Unilateral	Minimal	None
3	8 y F	Left-hand weakness	None	Right parietal	No	Unilateral	Minimal	None
4	2 y F	Developmental delay	Left hemiparesis	Right parietal	No	Unilateral	Minimal	Microcephaly
5	10 mo F	Left hemiparesis	Developmental delay	Right parietal	Yes	Unilateral	Minimal	Microcephaly
6	2½ y F	Seizures	Left hemiparesis, developmental delay	Right parietal	No	Unilateral	Minimal	Microcephaly, micropolygyria
7	18 mo F	Left hemiparesis	Developmental delay	Right parietal	No	Unilateral	Minimal	Microcephaly, micropolygyria
8	5½ y F	Right hemiparesis	None	Left parietal	Yes	Unilateral	None	None

Note.—y = years; m = months.

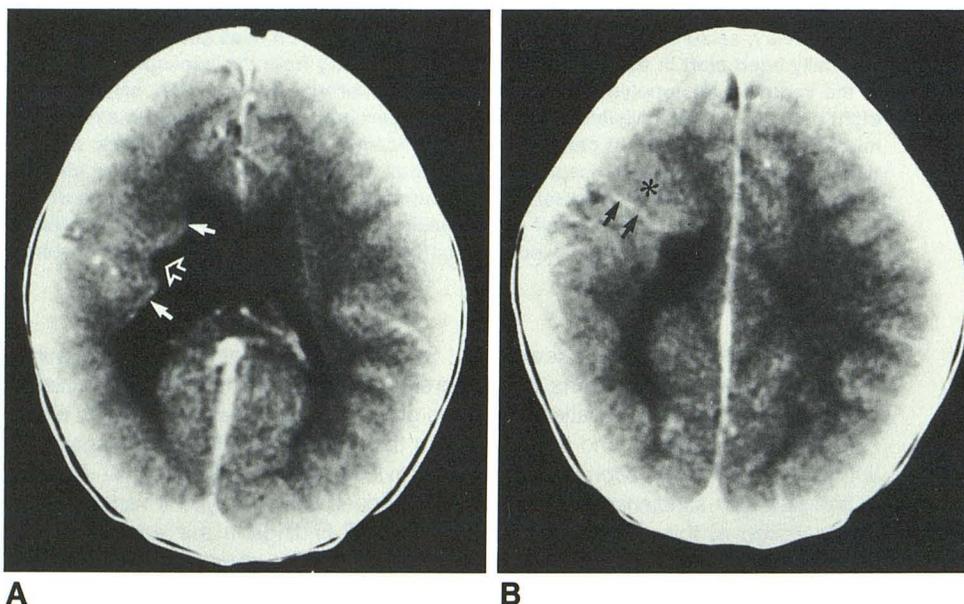


Fig. 1.—A and B, Type I schizencephaly. Postcontrast CT scans show abnormal cleft traversing full thickness of cerebral mantle in right parasyllian region. A small ventricular diverticulum is associated with the cleft (open arrow, A) and gray matter extends along ependymal surface for a short distance (closed arrows, A). A vessel is present in subarachnoid space of cleft (arrows, B) and the cortex lining cleft (asterisk, B) is thicker than cortex in other areas.

extend along the ependymal margin of the ventricle for a short distance (Fig. 1). None of the lesions showed abnormal contrast enhancement. The neuropathologic specimen, which was not from one of our eight cases, had a cortically lined cleft in the left parasyllian region that extended to the lateral ventricle (Fig. 3).

### Discussion

In previous reports of schizencephaly, most patients have had bilateral open clefts and only a small number of cases with unilateral fused clefts has been reported [3–5].

The fact that we have seen eight such cases in the past 2

years suggests that this variation may be more common than previously thought. There are two reasons why this is so: first, because of different CT findings from the type II variety; and second, because of confusion with other lesions, such as prior infarction.

Three findings indicate that this is a developmental anomaly rather than a postdevelopmental destructive process. First, the clefts are lined by cortex, which is not a feature of destructive lesions. The fact that the cortex lining the clefts is often thicker than normal is probably the result of micropolygyria, which is known to occur in the clefts and in other areas of cortex in patients with this disorder [1, 5] (Figs. 1 and 2). Second, the triangular configuration of the ventricle adjacent to the cleft is not generally associated with ex vacuo

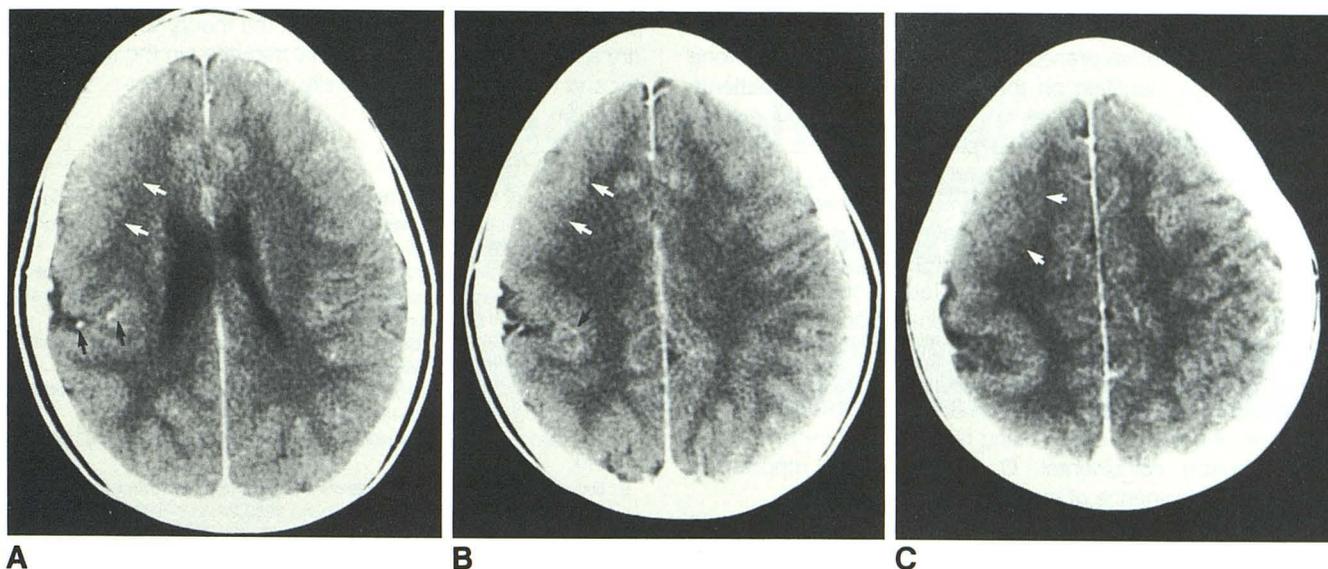


Fig. 2.—A-C, Type I schizencephaly. Postcontrast scans show abnormal cleft in posterior right parasyllian region. Note abnormal thickness and lack of sulcation of cortex anterior to cleft representing micropolygyria (white arrows). A vessel is present within subarachnoid space of cleft (black arrows).

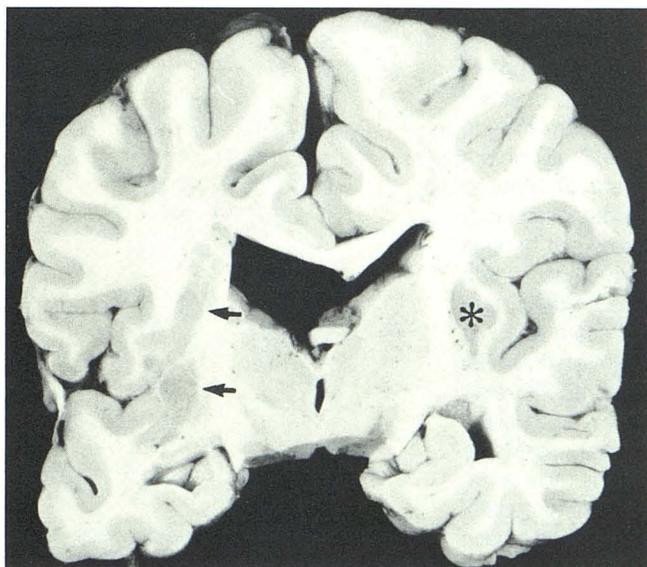


Fig. 3.—Neuropathologic specimen from a patient with type I schizencephaly. Section is parallel to orientation of cleft so anterior and posterior walls are not present on same section. Note abnormal gray matter of posterior wall extending from insular cortex to ventricular surface (arrows). An island of heterotopic gray matter is present in opposite hemisphere (asterisk).

ventricular dilatation. Third, a vessel is often seen extending between the cortical surfaces of the cleft. This probably represents a normal vessel in the subarachnoid space, which would not be present in an area of infarcted brain (Figs. 1 and 2).

Many theories have been proposed to explain the origin of

these lesions, although none has been proved. A temporary local impairment of the cerebral circulation may be responsible [6]. Regardless of the inciting event, the lesion appears to be the result of a focal area of abnormal migration of neuroblasts from the germinal matrix in the first trimester. Because of this defect the developing cortex anteriorly and posteriorly folds inward to form the cortically lined cleft (Figs. 1 and 2). The ventricular diverticulum is formed at the site of abnormal migration in the germinal matrix and represents the site where the pia and ependyma join to form the so-called pial-ependymal seam seen pathologically [1]. Whether the margins of the clefts are opposed, as in type I, or separated, as in type II, is probably related to the size of the area of abnormal migration from the germinal matrix, with larger areas of abnormal migration producing larger clefts. It has been proposed that the large ventricles seen in type II cases occur secondary to a lack of parenchymal development rather than from an obstruction of CSF circulation. This seems likely, because many patients with large ventricles and wide clefts are microcephalic [1].

Why the clefts occur most commonly in the parasyllian region is not clear; but if a circulatory disturbance is responsible, it may reflect the fact that the largest vascular territory (middle cerebral) is the most likely to be affected. It is remarkable that in seven of our eight patients, the cleft was on the right side. Right or left predominance has not been observed in other series, and is probably coincidental in our cases [3, 4].

The associated polymicrogyria is also a disorder of neuronal migration occurring in the first trimester. It may be a less severe manifestation of the same inciting event, as it most commonly occurs at the junction between normal cortex and the cortex lining the clefts, as well as in other areas in the

brain [6, 7] (Fig. 2). Pathologically, islands of heterotopic gray matter are also often present in this disorder, although none was positively identified on the scans of our eight patients (Fig. 3).

The clinical findings in these patients correlate with CT findings in that all the clefts involved the region of the central sulcus and each patient had a contralateral hemiparesis or monoparesis. The paresis in these patients is usually mild and correlates with the relatively small clefts as compared with the more severe type II defects. The four patients with associated microcephaly were developmentally delayed or mentally retarded, but those with a normal head size were not. Seizures were present in three of the eight cases but were not associated with any particular CT finding. The significance of seven of our eight cases being female is not certain and is also probably coincidental. One other series reported a marked predominance of males [3].

In summary, the CT appearance of fused, cortically lined clefts in the cerebral mantle is characteristic of type I schizencephaly, a disorder that may be more common than previously

thought. The findings also support the theory that the lesions are a result of abnormal neuronal migration in the first trimester and not postdevelopmentally acquired lesions.

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