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R E Ramirez

AJNR Am J Neuroradiol 1984, 5 (6) 830-831

<http://www.ajnr.org/content/5/6/830.citation>

This information is current as
of August 9, 2025.

Sonographic Recognition of Lissencephaly (Agyria)

Rosa E. Ramirez¹

Lissencephaly ("smooth brain") or agyria is a rare congenital malformation resulting from an arrest of the development of the human brain during the third to fourth month of gestation [1]. Before this stage of fetal cerebral development, the absence of gyri is normal [2]. The radiologic findings on plain skull radiography, pneumoencephalography, cerebral arteriography, and, more recently, cranial computed tomography, have been well documented [1, 3–5]. The real-time cranial sonographic findings in an infant with lissencephaly are presented. These have not been described previously, but are characteristic and may prove to be diagnostic.

Case Report

A 5½-month-old baby girl had been delivered by a 30-year-old gravida 2, para 1 mother at 37 weeks of gestation after a normal pregnancy. Apgar scores were 8 at 1 min and 9 at 5 min. The initial physical examination revealed a 2.35 kg hypotonic infant with epicanthic skin folds, small philtrum, micrognathia, puffiness of hands and feet, and a head circumference of 32.5 cm. The nursery course was complicated by hypoglycemia and polycythemia, requiring a partial exchange transfusion. After discharge, the infant showed delay in developmental milestones. She was hospitalized at age 3½ months for respiratory distress associated with an upper respiratory infection, which eventually required mechanical ventilation. Further complications included gastroesophageal reflux, documented by radionuclide scan, and the development of tonic clonic seizures. She was transferred to our institution at age 4½ months in moderately severe respiratory distress and with the clinical appearance of decerebration. Her head circumference was 42 cm. The hospital course was complicated by right middle- and lower-lobe pneumonia and intermittent seizure activity despite therapeutic levels of anticonvulsants. She died 1 month later after an episode of massive aspiration of formula.

Cranial sonography at 3 months of age, performed on an ATL 860 real-time sonographic unit using a 5.0 MHz transducer, showed a smooth brain surface with deep interhemispheric fissure; increased prominence of the subarachnoid space; the presence of three small midline calcifications, two of which were located in the roof of the cavum septi pellucidi; and uniformly dilated ventricles (figs. 1A–1E). The sylvian fissure and cingulate gyri, structures readily identified in normal patients, were absent (fig. 1C). A small area of increased

echogenicity was noted in the region of the right basal ganglia (figs. 1C and 1D), which corresponded to a focus of calcifications seen later on microscopic examination of the brain at autopsy.

Cranial computed tomographic findings correlated well with the sonographic findings, showing a small brain with smooth surface, widened subarachnoid space, uniformly enlarged ventricles, lack of insular opercularization, and three small midline calcifications, two of which were located in the roof of the cavum septi pellucidi (fig. 1F).

Discussion

The persistence of agyria beyond the fourth month of gestation indicates an arrest in the development of the brain. This includes a lack of the normal migration of the neurons from the periventricular germinal matrix to the periphery of the cerebral hemispheres to form the cortex. There is also lack of involution of the paraphysis, a neurosecretory structure located in the septum pellucidum, which normally involutes and disappears late in the third month of fetal life. The small, round, midline calcification described in many patients with agyria (and noted also in our patient) is located in a vestige of this paraphysis [1]. The etiology for this arrested development in the fourth month of gestation is unknown, although several reports of this condition occurring in pairs of siblings may indicate a hereditary basis [3].

The real-time cranial sonographic findings in our case raise the possibility of early diagnosis of this condition by noninvasive techniques. In the past, agyria has been diagnosed mainly at autopsy and, more recently, before death, by computed tomography. However, on the basis of the findings in our patient, it seems likely that sonography also can be used to delineate the problem. This is important because real-time sonography is often the first screening method for evaluation of neonatal intracranial disease. Further experience may demonstrate the feasibility of in utero diagnosis of this condition by sonography.

I conclude that the sonographic findings, when combined with the clinical features, are sufficient for the specific diagnosis of neonatal lissencephaly.

Received April 26, 1983; accepted after revision July 25, 1983.

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¹ Department of Radiology, Brooke Army Medical Center, Fort Sam Houston, TX 78234. Address reprint requests to Chief, Department of Radiology.

AJNR 5:830–831, November/December 1984 0195–6108/84/0506–0830 \$00.00 © American Roentgen Ray Society

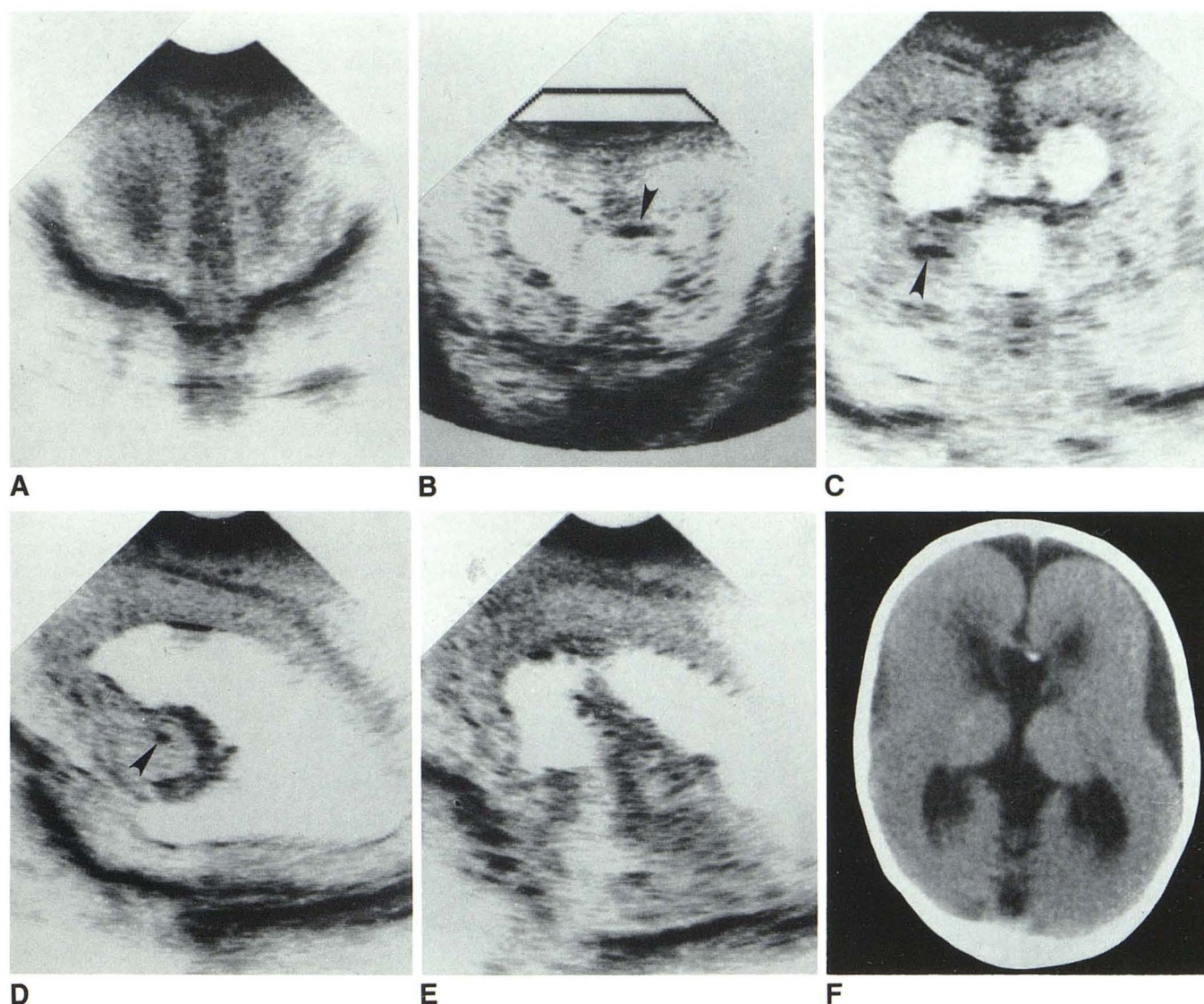


Fig. 1.—A–C, Real-time cranial sonograms (ATL 860, 5.0 MHz transducer), coronal plane, at 3 months of age. Section through frontal lobes (A) shows smooth brain with deep interhemispheric fissure and widened subarachnoid space. B, Midline calcification in roof of cavum septi pellucidi (arrowhead); frontal horns of lateral ventricles are dilated. Section through third ventricle (C) shows cavum septi pellucidi between lateral ventricles; dilatation of third and lateral ventricles. Cingulate gyri and sylvian fissure are absent. Echogenicity in right basal ganglia (arrowhead) corresponded to microscopic calcifications

found at autopsy. D, Right parasagittal section. Focus of calcification (arrowhead) in right basal ganglia, absence of cingulate gyri, increased subarachnoid space, and ventricular dilatation. E, Midsagittal section. Midline calcifications; dilated fourth ventricle. F, Cranial computed tomogram at 3 months of age. Axial section at level of basal ganglia shows absence of sylvian fissure, midline calcification in roof of cavum septi pellucidi, and faint area of increased attenuation in right basal ganglia.

REFERENCES

1. Wesenberg RL, Juhl JH, Daube JR. Radiological findings in lissencephaly (congenital agyria). *Radiology* **1966**;87:436–445
2. Chi JG, Dooling EC, Gilles FH. Gyral development of the human brain. *Ann Neurol* **1977**;1:86–93
3. Garcia CA, Dunn D, Trevor R. The lissencephaly (agyria) syndrome in siblings. *Arch Neurol* **1978**;35:608–611
4. Ohno K, Enomoto T, Imamoto J, Takeshita K, Arima M. Lissencephaly (agyria) on computed tomography. *J Comput Assist Tomogr* **1979**;3:92–95
5. Miller JQ. Lissencephaly in 2 siblings. *Neurology* **1963**;13:841–850