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Schizencephaly: Rare Cerebral Malformation Demonstrated by Sonography

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Schizencephaly is a rare, severe congenital cystic malformation of brain. We describe the sonographic demonstration of schizencephaly in a premature infant with comparative autopsy description and contrast it to porencephaly.

Case Report

An 820 g, 28 week, small-for-gestational-age boy was born after an uneventful but shortened pregnancy. At birth, the circumference of the infant's head (23 cm) was well below the third percentile. Transillumination of the skull was increased over the parietotemporal areas bilaterally. Initially, he had minimal spontaneous movements, little response to stimulation, absent suck reflex, minimal Moro response, and absent plantar reflexes. Subsequently, spontaneous jittery movements of all extremities developed. The baby died at 2 days of age from respiratory distress syndrome.

Sonography of the infant's head was performed at 11 hr of age with a real-time unit, using a 7.5 MHz transducer focused at 3 cm. Sonograms were obtained using the anterior fontanelle as an acoustic window.

Coronal and parasagittal sonograms showed enlargement of both lateral ventricles and absence of the septum pellucidum (fig. 1A). More posteriorly, large bilateral clefts in the parietotemporal cerebral mantle were demonstrated (fig. 1C). The lateral ventricles were contiguous with these clefts, and choroid plexus extended into the left cleft (figs. 1C and 2C).

At autopsy, gross examination of the brain revealed large, bilateral, slightly asymmetric clefts, each extending about 2 cm along the midlateral surfaces of the cerebral hemispheres in a nearly horizontal orientation (fig. 2). The cortical mantle appeared to roll in at the clefts, which were filled with straw-colored cerebrospinal fluid contained by a clear, delicate pia-arachnoid membrane. On coronal section, it was evident that the clefts and both lateral ventricles were contiguous as had been shown on sonography (figs. 1B and 1D).

Microscopically, the brain was severely malformed. Cortical layering was indistinct, and there was extensive polymicrogyria. The cleft walls were lined by a thick band of neural cell matrix that was contiguous with the subependymal germinal cell masses. There were numerous disorganized masses of germinal cells within the hemispheric central white matter. The hippocampus, caudate, thalamic, hypothalamic and midbrain nuclei all had anomalous neural organization, and the corticospinal system was nearly absent.

Discussion

Porencephaly was first proposed by Heschl [1] to describe congenital defects in the cerebral mantle that were associated with other developmental anomalies and sometimes with hydrocephalus. However, it was not certain whether such porencephalic defects represented anomalies of development or infarcts superimposed on normal development. This issue was addressed by Yakovlev and Wadsworth [2, 3], who described five patients with congenital clefts in the cerebral mantle associated with polymicrogyria and abnormalities of migration of germinal cells. They argued that the abnormalities are anomalies of development involving varying degrees of cerebral agenesis, and probably beginning during the fifth or sixth fetal week. Yakovlev and Wadsworth further postulated that clefts form because paraventricular germinal cells fail to migrate to the cortical mantle. The clefts are characteristically bilateral and nearly symmetric and are most likely to form along an axis of normal future fissure development, particularly the sylvian fissure (fig. 2). The dysgenetic walls may resist ventricular pressure poorly and consequently yield, creating gaps in the clefts. Yakovlev and Wadsworth called this severe developmental lesion *schizencephaly* and differentiated it from encephaloclastic or destructive porencephalic lesions.

Posthemorrhagic porencephalic cysts [4] as well as congenital intracranial cystic lesions [5] have been demonstrated with sonography. Angiographic and pneumographic demonstration of a case of schizencephaly has been reported [6]. However, we found no previous description of schizencephaly with sonographic and autopsy correlation. The bilaterality, extent, and symmetry of the lesion as noted on sonography suggested the diagnosis. The severe histologic disorganization of the brain seen at pathology was not demonstrated sonographically per se.

Patients with schizencephaly may live for a varying number of years and typically have profound mental retardation, spastic diplegia, cortical blindness, and seizures [7]. In contrast, patients with porencephalic cysts after trauma, circulatory

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Fig. 1.—Coronal slices of brain specimen viewed from behind with approximately corresponding sonograms. **A** and **B** are more anterior than **C** and **D**. Planes of sonograms are between true coronal and axial planes. Left cleft (*short straight arrows*) is larger and extends further posteriorly than right cleft (*curved arrows*). Choroid plexus extends into left cleft (*long straight arrow*). Lateral ventricles are contiguous with clefts, and septum pellucidum is absent. R = right.

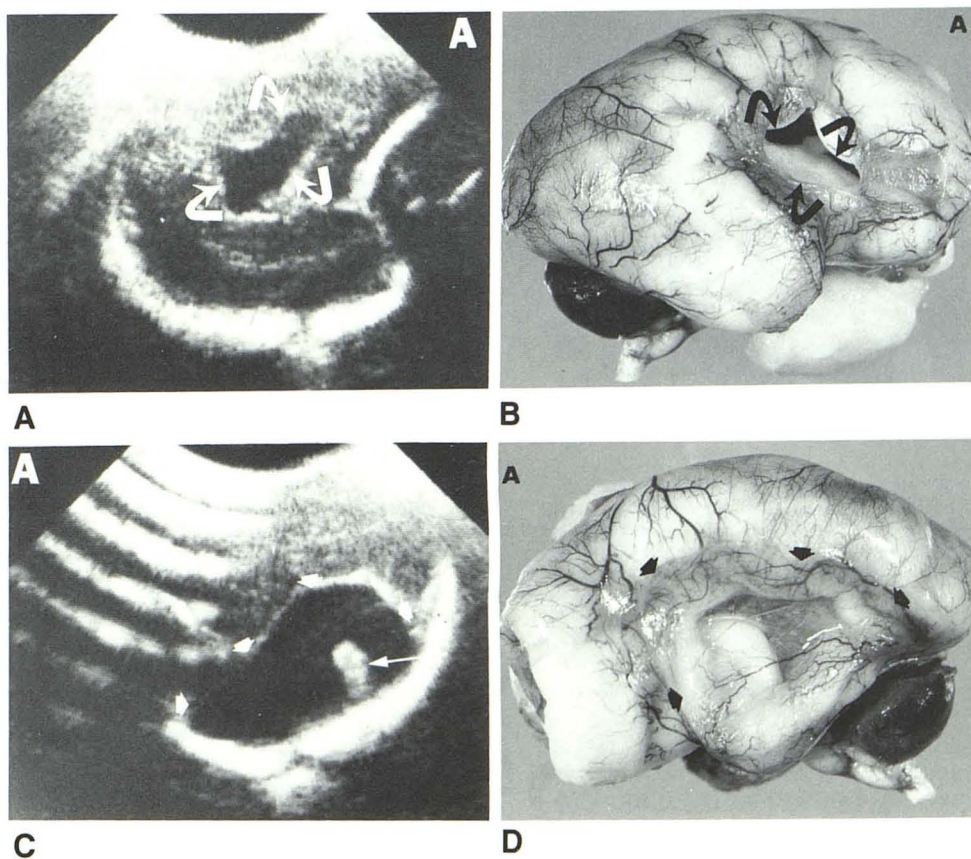
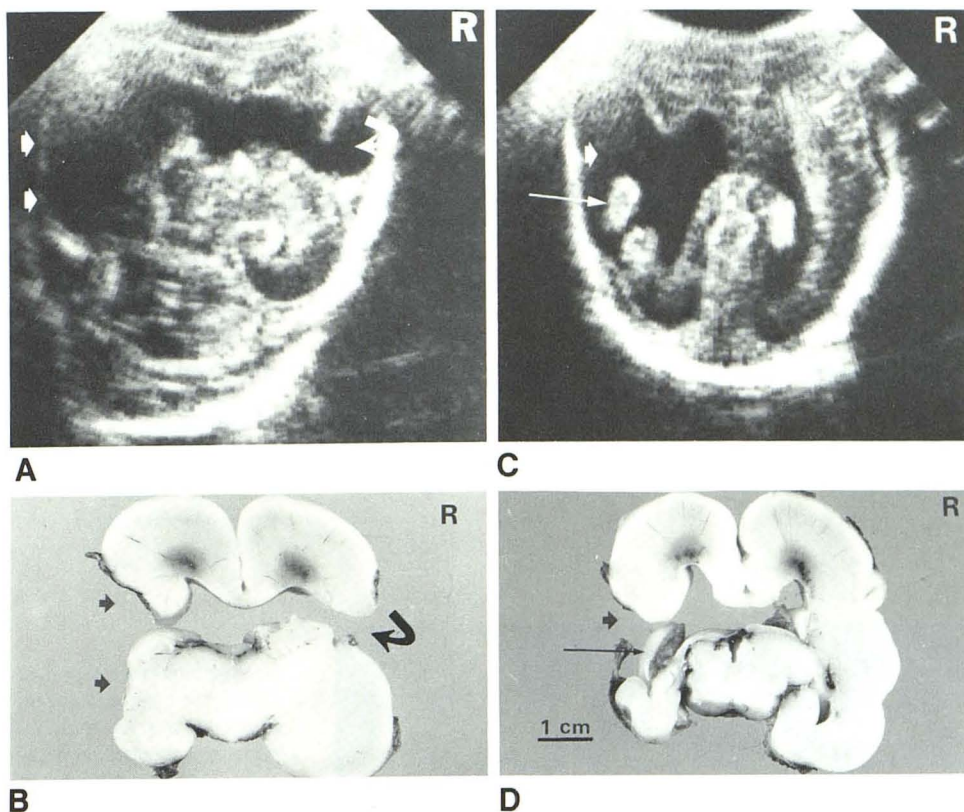


Fig. 2.—Lateral views of brain specimen with corresponding parasagittal sonograms (**A** and **B**, right; **C** and **D**, left). Large clefts along right (*curved arrows*) and left (*short straight arrows*) sylvian fissures. Choroid plexus extends into left cleft (*long straight arrow*). Pia-arachnoid membrane is opened on right (**B**) and still intact on left (**D**). A = anterior.

disturbance, or intraparenchymal hemorrhage during the neonatal or postnatal period have a relatively better prognosis. Schizencephaly is associated with severe neurologic disability, and recognition of this developmental anomaly is important for patient management.

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