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CEREBRAL MALFORMATIONS

MIDBRAIN AND HINDBRAIN INVOLVEMENT IN LISSENCEPHALY

Involvement of the midbrain and hindbrain (MHB) in the various groups of lissencephalies was examined in an MRI study of 111 patients (aged 1 day to 32 years; mean 5 years 4 months) studied at University of California San Francisco, and centers in France, Belgium, and Turkey. The three major groups of lissencephaly (cLIS or LIS type 1; vLIS or variant LIS; and CBSC, cobblestone complex or LIS type 2) showed significant differences in the appearance and severity of associated MHB malformations; the least severe MHB malformations occurred with cLIS and the most severe with CBSC lissencephaly. The extent of cerebral lissencephaly was significantly correlated with the severity of MHB abnormalities (P= 0.0029). Based on the data obtained here and that in the literature, a new classification of lissencephalies is proposed: *Classic LIS* (LIS1, LIS1 mossaicism, DCX (XLIS); *Variant LIS* (ARX, RELN, VLDLR, ND1, ND2, ND3, TL-LIS; *Cobblestone complex* (FCMD-Fukuyama type, WWS, MEB; and related MD syndromes (CMD merosin deficiency). (Jissendi-Tchofo P, Kara S, Barkovich AJ. Midbrain-hindbrain involvement in lissencephalies. **Neurology** Feb 3, 2009;72:410-418). (Respond and reprints: Dr Patrice Jissendi-Tchofo, Radiology Department, Erasme Hospital, Route de Lennik 808, B-1070 Brussels, Belgium. E-mail: jisendi@gmail.com).

COMMENT. Lissencephaly or smooth brain is characterized by a paucity of gyri, ranging from complete agyria to localized pachygyria. It is usually classified in 2 groups, classic (cLIS or lissencephaly type 1), and cobblestone complex (CBSC, lissencephaly type 2). Five genes are identified as causing cLIS, and numerous genes are associated with CBSC, also called dystroglycanopathies. Most patients with CBSC have congenital muscular dystrophies with CNS involvement, including Fukuyama CMD, and Walker-Warburg

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syndrome. Relatively few investigators had stressed the importance of mesencephalic and rhrombencephalic involvement in association with cerebral cortical dysgenesis until the work of Sarnat, colleagues and others in the 1990s. A classification of cerebral malformations proposed by Sarnat et al in 2004 also stressed the inclusion of thalamic, brainstem and cerebellar malformations in association with lissencephaly and holoprosencephaly. In an editorial, Dr Sarnat reviews the progress of our understanding of the genetic programming of neural tube development and the need for research on a genetic mechanism for the association of forebrain and hindbrain malformations in the lissencephalies. (Sarnat HB. Cortical malformations. Looking behind the cortex. **Neurology** 2009;72:394-395).

SURGICAL OUTCOME IN FOCAL CORTICAL DYSPLASIA

The predictors of surgical outcome and relevance of pathological severity were determined in 166 consecutive patients with intractable epilepsy and focal cortical dysplasias treated surgically at Konkuk University Medical Center, and National University Hospital, Seoul, Korea. Poor surgical outcome was associated with incomplete resection of epileptogenic area, mild pathologic features, and secondary tonic-clonic seizures. Patients with severe pathologic features had MRI abnormalities. MRI findings, EEG, PET and ictal SPECT were not associated with surgical outcomes. (Kim DW, Lee SK, Chu K, et al. Predictors of surgical outcome and pathologic considerations in focal cortical dysplasia. **Neurology** Jan 20, 2009;72:211-216). (Respond and Reprints: Dr Sang Kun Lee, Department of Neurology, Seoul National University Hospital 28, Chongno Ku, Seoul ,110-744, Korea. E-mail: sangunlee@dreamwiz.com).

COMMENT. Patients with focal cortical dysplasia and intractable epilepsy are at risk of a poor surgical outcome, when associated with incomplete resection, mild pathologic features, or secondary tonic clonic seizures. Incomplete resection of focal cortical dysplasia was the main predictor of poor postsurgical outcome in 149 pediatric patients operated at the Miami Children's Hospital (Krsek P et al. **Neurology** 2009;72:217-223).

In practice, a negative MRI does not exclude a subtle cortical dysplasia that may underly refractory seizures. Newer imaging techniques may uncover small dysplasias amenable to treatment in specialized epilepsy and surgical centers. (Mathern GW. **Neurology** 2009;72:206-207).

VASCULAR DISORDERS

INTRACRANIAL ARTERIOPATHY AND ISCHEMIC STROKE

Repeated vascular imaging findings and clinical charts of 79 children with anterior circulation arterial ischemic stroke (AIS) and unilateral intracranial arteriopathy of the internal carotid bifurcation were studied at the University Medical Center, Utrecht, The Netherlands, and other centers in France, UK, and Canada. The characteristics of 5 (6%) patients with progressive and 74 (94%) with transient cerebral arteriopathy (TCA) were compared after a median follow-up of 1.4 years. Most infarcts were localized in the basal ganglia. Follow-up vascular imaging showed complete normalization in 23% of TCA