Case No. 10

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This 7-1/2 year old girl was born at term with Apgars of 7^1, and 8^5. Shortly after birth, a diagnosis of congenital heart disease was made: a double outlet right ventricle with a ventricular septal defect. Pulmonary artery banding was performed at six weeks of age: shortly after surgery generalized seizures developed. An EEG after the onset of seizures showed a grade I generalized dysrhythmia better developed over the left hemisphere.

Seizures were well controlled with phenobarbital, until 20 months of age when she developed status epilepticus. Seizures were more or less continuous for almost one day before they were controlled by high doses of phenobarbital and Dilantin. The seizures were of a generalized, grand mal type although with accentuation on the left side of the body. The status was followed by generalized flaccidity and unresponsiveness for one week. Consciousness returned, but a left-sided hemiparesis remained.

A CT scan shortly after the status revealed a diffuse decrease in the density of the right cerebral hemisphere, with no midline shift. A CT scan one week later showed slight atrophy of the right hemisphere. An EEG showed generalized slowing of electrical activity over the right hemisphere, but no epileptiform activity.

In subsequent years, there was a residual left-sided spastic hemiplegia more severe in the upper extremity. She was able to walk. She was maintained on phenobartital and Dilantin, and she had only occasional, minor seizures predominantly affecting the left arm and leg. There was developmental delay of moderate severity. Clinical microcephaly became evident by the age of 5 years.

Despite attempted definitive correction of the congenital heart defect at 4 years of age, an inoperable aortic subvalvular stenosis remained, and there was gradual deterioration of cardiac function: death resulted from heart failure at the age of 7-1/2 years.

At autopsy, the brain weighed 535 grams: its gross appearance is illustrated by the kodachrome slide. The H&E stained section from the right frontal lobe, and a portion of the head of the caudate is representative of the pattern of injury throughout the right hemisphere. The left hemisphere was mostly unremarkable, except for a localized area of neuronal loss and gliosis which affected the border of the parietal and occipital lobes.

Diagnosis: Cerebral hemiatrophy, with crossed atrophy of cerebellar hemisphere

For discssion: Pathogenesis