



Case Studies from the Child Development Center

A teenager with hemiplegia and learning disability

Mark Simms, MD, medical director, Child Development Center, Children's Hospital of Wisconsin; professor, Pediatrics, Medical College of Wisconsin.

BL was a 13-year 5-month-old boy referred to the Child Development Center for evaluation of learning difficulty. He attended a regular eighth grade class in the local middle school and received help for reading and math.

When tested by our psychologist, BL had a normal full-scale IQ of 85 on the Wechsler Intelligence Scale for Children (WISC-IV). His verbal comprehension index was 88, perceptual reasoning index was 75, working memory index was 83, and processing speed index was 77. Academically, BL was reading at a mid-fifth grade level. He read slowly and inaccurately at times. Frequent spelling errors also were noted. Math was at an early fifth grade level.

Physical examination was within normal limits except for mild left sided hemiplegic posturing and a slight degree of underdevelopment of the left arm. There was full range of motion in the left arm and some tightness of the heel cord on the left. Deep tendon reflexes were increased on the left and normal on the right. A positive Babinski reflex was present on the left.

Mother's pregnancy with BL was unremarkable except for elevated blood pressure in the last trimester. Fetal movement patterns were reported to be normal. Birth weight was 6 lb. 7 oz. Apgars were 7 and 8 after a normal spontaneous vaginal delivery. He was discharged from the nursery on the second day of life. At his 6-week well-child visit, BL was noted to have weakness of the left arm and leg. At 3 months of age, a CT of the head showed a wedge-shaped defect of the right cerebral cortex in the distribution of the middle cerebral artery. Testing for coagulation disorders was negative. At that time, his mother was told BL likely suffered an intrauterine stroke as a complication of her SLE condition even though it had been in remission during pregnancy.

The past medical history was unremarkable. BL never had experienced a seizure. He took no chronic medications and was never hospitalized overnight. An orthopedic surgeon followed him for a tight left heel cord and he wore an AFO. Family history was negative for learning disorders or mental retardation.

Despite BL's obvious motor impairment, early cognitive and language milestones were normal. He started saying single words by 15 months, walked alone by 19 months and spoke in sentences by age 3 years. He attended a community preschool program and received occupational and physical therapy through the local Birth-to-Three Program and Early Childhood Programs. He was well behaved and got along well with other children. Difficulty learning the alphabet and decoding written words was noted during kindergarten. He required only minimal assistance for most activities of daily living.

Because unilateral focal brain injuries usually do not result in learning disabilities, BL's clinical picture suggested a bilateral brain disorder. I ordered an MRI scan of the brain, which showed unilateral Type II schizencephaly on the right, and bilateral pachygyria. No other brain abnormalities were noted.

Discussion: Schizencephaly is a rare congenital brain anomaly with a prevalence of approximately 1.5/100,000. The lesion consists of a cleft in the cerebral hemisphere extending from the pial surface to the lateral ventricle. Approximately two-thirds (63 percent) are unilateral. Schizencephaly may appear as an isolated malformation or associated with other brain anomalies, such as agenesis of the corpus callosum and septum pellucidum, septo-optic dysplasia and hydrocephalus. Clefts typically develop in the region of the Rolandic and Sylvian fissures and involve the frontal cortex. The walls of the clefts are lined with thickened cortex consisting of pachygyria or polymicrogyria and also may contain large neuronal heterotopias. Schizencephaly results from a disruption or disorder in neuronal migration in the first trimester of fetal brain development. The cause likely is multifactorial. Sporadic and familial cases of Type II schizencephaly have been associated with mutations in the homeobox gene *EMX2* which is expressed in the neuroblasts of the ventricular zone. This gene is involved in the structural patterning of the developing forebrain. The lesions also have been noted following intrauterine CMV infection. Vascular disruption involving thrombotic occlusion in the distribution of the middle cerebral artery has been proposed as a possible mechanism for schizencephaly, however maternal and/or infant

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thrombophilia is more likely to result in porencephaly, a cystic lesion of the brain parenchyma. However, as noted by Fernandez-Bouzas et al (2006), "When portions of the brain are injured or do not form, the blood supply is markedly reduced, and the blood flow is diminished...for this reason it is not possible to conclude which anomaly was first: failure in brain development or absence of the artery."

Neonatal strokes are more common than schizencephaly (incidence of approximately 1/4,000). In most cases, the child appears normal at birth but develops tonic-clonic seizures in the first 4 days of life. Risk factors are found in approximately half of children with neonatal stroke, including birth asphyxia, maternal diabetes, perinatal infection, maternal drug abuse, congenital heart malformations and sickle cell disease. Conditions that result in a hypercoagulable state, such as Factor V Leiden mutation, protein C or protein S deficiency and antiphospholipid syndromes. Maternal SLE is associated with the presence of antiphospholipid antibodies that may result in arterial and venous thromboses and fetal loss. Pregnancies resulting in surviving infants frequently are complicated by preeclampsia, IUGR due to placental insufficiency and preterm delivery. Neonatal lupus erythematosus is a rare complication and is associated with congenital heart block, cardiomyopathy, cutaneous lupus lesions, hepatobiliary disease and thrombocytopenia.

The absence of neonatal seizures and the presence of long-term cognitive problems suggested another diagnosis. In

this case, the MRI was able to better define the lesion and offered an alternative explanation for the child's problems.

I met with BL's parents to review the MRI findings and discuss the diagnosis of schizencephaly. His mother began to cry when I explained that her son's problem was due to a rare disorder of early brain formation and not the result of a neonatal stroke or a complication of her SLE disease. Since his birth, she had carried a strong feeling of guilt that her own medical problem had caused her son to be physically disabled.

References available upon request.

International Adoption Clinic overview

The International Adoption Clinic at Children's Hospital of Wisconsin, based in the Child Development Center, offers services to families adopting children from outside the United States. We are one of a handful of international adoption clinics based in an academic medical center. This clinic provides three services: 1) preadoption medical record review for prospective adoptive families; 2) medical evaluations of newly arrived children; and 3) post-adoption evaluations of children from international settings when there is a concern about a health, behavioral or developmental problem. All services are coordinated with the child's primary care physician.

For further information, or to make an appointment for a child, please contact Liz Schaefer, MSW, (414) 266-2945 or e-mail: eschaefer@chw.org.

Case Studies from the Child Development Center is a limited edition newsletter to help inform referring physicians and other professionals on the depth and breadth of pediatric communication and behavioral issues diagnosed and treated in the Child Development Center at Children's Hospital of Wisconsin.

It is written by Child Development Center staff and produced by Children's Hospital of Wisconsin in January, March, May, July, September and November.

It also is available online at www.chw.org, Child Development Center, Related Links.

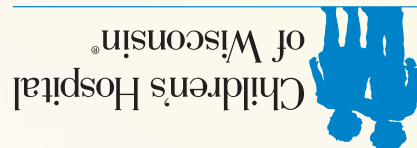
Questions and suggestions can be forwarded to:

Mark Simms, MD, Child Development Center, MS744,
Children's Hospital of Wisconsin, PO Box 1997, Milwaukee, WI 53201-1997, or call (414) 266-2928.

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Milwaukee, WI 53201-1997
PO Box 1997
Child Development Center, MS 744

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