Case 3 Discussion

Diagnosis: Central Nervous System Disorder Manifested as Developmental Arrest or Loss of Skills

The question facing the primary care pediatrician in this case is the extent to which the growth and developmental findings are consistent with a preterm, very low birth weight (VLBW) infant or whether they signal some other problem. Despite the neonatal course and difficulty in obtaining a complete history, the significant observations suggesting the focus of the diagnostic examination are the actual loss of developmental skills with a plateauing of physical growth. Although intraventricular hemorrhage in a VLBW infant is associated with increased risk of developmental delay associated with cerebral palsy, increasing evidence indicates that VLBW infants experiencing relatively uncomplicated courses that include intraventricular hemorrhage without parenchymal damage are not likely to be delayed. Prior maternal concerns in this case may be difficult to interpret in the absence of a clearer idea of personal and cultural expectations. In any event, in the prior visit at 16 months, direct observation indicated an infant with a physical growth pattern seen in VLBW children and gross motor skills within normal for corrected age. Even among delayed preterm children, however, continued acquisition of developmental skills and growth along a percentile established during the second year should be expected, although the rate of development may be slower than in children without delay.

The loss of milestones, with the need for support in standing (previously done independently), suggests one of the inherited metabolic disorders affecting the nervous system. More detailed neurologic examination may be helpful but not necessarily diagnostic. Although many metabolic disorders are associated with hypotonicity and decreased tendon reflexes, VLBW children also may exhibit tone problems, including hypotonia, and other neurologic findings, such as spasticity, nystagmus, alterations in deep tendon reflexes, and ataxia with various forms of cerebral palsy. Although cerebral palsy is the more common problem among premature infants, the pediatrician should remember that neurologic abnormalities also might occur with central nervous system tumors irrespective of duration of gestation. Other findings, such as altered facies, visceromegaly, or ocular findings (corneal clouding, cataracts, or cherry-red spots), or a suggestive family history (consanguinity, prior unexplained fetal or infant loss) point to the inherited disorders. Confirmation of the diagnosis of a hereditary disease requires appropriate biochemical assay of urine or tissue.

This particular child was diagnosed as having metachromatic leukodystrophy. The actual regression noted in his case is more suggestive of an inherited disorder than it is of cerebral palsy. (*Marie C. McCormick, MD, ScD, Brigham & Women's Hospital, Boston, MA*)

PIR Quiz-CME Credit

The American Academy of Pediatrics is accredited by the Accreditation Council for Continuing Medical Education to sponsor continuing medical education for physicians. As an organization accredited for continuing medical education, completion of the PIR Quiz meets the criteria for 2 hours of credit, per issue, of the American Academy of Pediatrics' PREP Education Award.

The American Academy of Pediatrics designates this continuing medical education activity for 2 credit hours, per issue, in Category 1 of the Physician's Recognition Award of the American Medical Association.

This program has been reviewed and is acceptable for 2 Prescribed hours per issue by the American Academy of Family Physicians. (Terms of approval: Beginning date Janaury 1992. Enduring Materials are approved for 1 year, with option to request renewal. For specific information, please consult the AAFP Office of Continuing Medical Education.)

The questions for the PIR quiz are located at the end of each article in this issue. Each question has a SINGLE BEST ANSWER. To obtain credit, record your answers on the PIR Quiz Card found in the January issue, and return the card to the Academy. (PREP group participants will receive the PIR Quiz Card and Self-Assessment Credit Reply Sheet under separate cover.) To receive CME credit on the 1992 annual credit summary, you must be enrolled in PREP or subscribe to *Pediatrics in Review* and return the PIR Quiz Card by February 28, 1993. PIR Quiz Cards received after this deadline will be recorded in the year it is received; with cards from the 1992 PIR journals, accepted through December 31, 1994.

The PIR quiz card is bound into the January issue. Complete the quizzes in each issue and send it to: American Academy of Pediatrics, PREP Office, PO Box 927, Elk Grove Village, IL 60009-0927.

The correct answers to the questions in this issue appear on the inside front cover.

American Academy of Pediatrics

Continuing Education Programs

Annual Meetings

San Francisco, California
October 10–15, 1992

Washington, DC
October 30–November 4, 1993

Spring Sessions

New York, New York
April 11–16, 1992

Chicago, Illinois
March 20–25, 1993

Continuing Medical Education Courses

Clinical Advances in Pediatrics
Santa Fe, New Mexico
May 8–10, 1992

Pediatric Advances
Hilton Head, South Carolina
May 29–31, 1992
Memorial Day Weekend

Clinical Pediatrics
Washington, DC
June 19–21, 1992

Pediatrics for the Practitioner
Seattle, Washington
September 4–6, 1992

Advances in Pediatrics
Newport, Rhode Island
October 2–4, 1992

Pediatric Update II
Williamsburg, Virginia
December 11–13, 1992

Current Concepts in Pediatrics
Vail, Colorado
January 7–10, 1993

Pediatrics 1993
Maui, Hawaii
March 5–7, 1993

To those enrolled in PREP (Pediatrics Review and Education Program), these programs feature subject matter coordinated with the PREP curriculum. Credits earned in these courses may be applied toward the PREP Education Award available to Fellows and Candidate Fellows of the Academy.

For further information contact:
CME Registration
American Academy of Pediatrics
PO Box 927
Elk Grove Village, IL 60009-0927
800/433-9016,
Outside the US and Canada: 708/228-5005