737S

9:45 AM

Neurologic Status of Infants Born to HIV-1–Infected Mothers and Their Controls: A Prospective Study from Birth to 24 Months of Age

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Objective. Determine neurologic status (NS) of HIV-1–infected infants and controls from birth to 24 m; define early signs and subsequent patterns of HIV-1–related CNS disease progression; correlate NS with P-2 defining condition and AIDS.

Patients. Infants born to HIV-1 sero+ and sero− women. One hundred eighty-nine of 434 infants enrolled are ≥15 months of age and available for analysis: 20 HIV-1 infected, 63 seroreverters, 106 sero−.

Design. Serial exams assessing mental status (MS), activity, language, head circumference (HC), cranial nerves, motor, sensory, coordination, and developmental-maturational (DM) changes. Cross-sectional analyses (CSA) of NS at ages 3, 6, 9, 12, 18, and 24 months.

Results. 1. CSA: There was no statistical difference in NS between the sero−, seroreverters and HIV-1–infected children who did not have or go on to develop an AIDS-defining condition, but there was a difference in NS in infants who did have or develop AIDS. 2. The major domains affected were MS, activity, language, HC, hand use, motor and DM. 3. Early signs were head lag, axial hypotonia, ↓ vocalizations and ↓ activity. 4. There were two distinct patterns of progression to a) severe cognitive and motor deficits (spastic quadriparesis) (4 infants) or b) cognitive impairment, hypotonia and delayed motor development (5 infants).

Conclusions. HIV-1–related CNS disease occurs more frequently, but not exclusively, in infants who have or go on to develop AIDS. In infancy there are two distinct patterns of severity and progression of HIV-1–related CNS disease.

Study supported by NICHD.

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10:00 AM

Intravenous Gammaglobulin (IVGG) Therapy in Children with Severe Acute Inflammatory Demyelinating Polyneuropathy (AIDP)

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Objective. To compare outcome of children with severe AIDP (acute inflammatory demyelinating polyneuropathy) treated with high-dose IVGG (intravenous gammaglobulin) to those who received no therapy.

Background. In children with severe AIDP, neuromuscular paralysis needing artificial ventilation, residual deficits or prolonged recovery may occur. The effect of IVGG in this group versus natural recovery has not been prospectively studied.

Methods. Five children meeting criteria of AIDP functional grade 4 (i.e., bed or chairbound, or worse) were treated with IVGG infusions either as five doses each of 0.4 g/kg or a single dose of 2 g/kg. Recovery times, assessed by functional grading scales, were compared to untreated children who presented late, were ambulatory and were recovering from grade 4 illness.

Results. In the treatment group, mean age 5.5 years (range, 3 to 9.5 years), the first sign of recovery occurred within 2 days of initiating therapy. The mean time to an independent ability to walk (grade 2) was 10 and 38.6 days in the treated and untreated groups, respectively. Follow-up periods were 14.4 and 12.6 months (range, 5 to 26) in each group. In the untreated group, two patients had severe residual deficits.

Conclusions. We conclude that in children with severe AIDP, IVGG therapy shortens recovery time to independent walking and intensive care and hospitalization time.

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10:15 AM

Magnetic Resonance Spectroscopy in Niemann-Pick Disease Type C: Correlation with Diagnosis and Clinical Response to Treatment with Cholestyramine and Lovastatin

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Objective. To determine the short-term effects of cholesterol-lowering agents on the neurological features of early Niemann-Pick type C (NP-C) and to develop a noninvasive methodology of monitoring clinical response.

Background. NP-C is an autosomal recessive neurovisceral storage disorder that results from defective intracellular cholesterol esterification. Cholesterol-lowering agents have been shown to decrease tissue (hepatic) lipids in NP-C patients.

Design/Method. A 9-month-old boy who initially presented with neonatal jaundice and progressive hepatosplenomegaly was studied. Liver and bone marrow biopsy confirmed lipid storage and fibroblast culture documented defective cholesterol esterification. Central hypotonia and moderate developmental delay were evident. Water-suppressed proton magnetic resonance spectra from a supraventricular volume of central white and gray matter revealed an abnormal lipid signal consistent with an accumulation of cholesterol esters. The patient was treated with low-cholesterol diet and cholesterol-lowering agents (cholestyramine and lovastatin).

Results. Standardized neurodevelopmental assessments (Peabody and Griffith scales) at 13 months were normal. Repeat proton magnetic resonance spectra no longer detected the previously observed lipid resonance.

Conclusion. Early treatment of NP-C with low-cholesterol diet and cholesterol-lowering agents appears to have short-term beneficial effects. Magnetic resonance spectroscopy provides a noninvasive means of monitoring CNS response.