COMMENT. After a detailed history and examination, a consensus-based staged approach to the evaluation of the child with global developmental delay is suggested. The timing of this evaluation is often a problem, a subject that needs further study. State-based newborn screening programs will identify some metabolic disorders shortly after birth. All states screen for phenylketonuria and congenital hypothyroidism, and most screen for sickle cell disease and galactosemia. (Pediatrics 2000;106:383-427). Thirty two states require universal newborn hearing screening. All children with GDD should have auditory and visual testing. Based on diagnostic yield, the MRI (nonenhanced) had the highest yield (55%), and metabolic screening the lowest (1%). The Committee emphasizes that the report is meant as an educational service, and is not meant to exclude alternative individualized evaluations of GDD.

EARLY DIAGNOSIS OF FRAGILE X SYNDROME

Surveys from 274 families with at least one child with fragile X syndrome (FXS) were used to determine factors associated with the discovery of the diagnosis in a study at the University of North Carolina, Chapel Hill, NC. The average age at first concern was 15.6 months, professional confirmation was at 25.9 months, entry into early intervention or special services was at 32 months, the FXS test was ordered at 56.2 months, and the diagnosis was made at 60 months. Variability of the timing of these steps in diagnosis was considerable; the average age of diagnosis ranged from 6 months to 30 years. Children born later than 1990 were identified much earlier; for boys, the average age at diagnosis was 31.5 months. Girls were identified with FXS about 6 months later than boys. Many families had additional children with FXS before becoming aware of increased risk. Parents of children with FXS perceive the discovery of the diagnosis to take too long, leading to delays in interventional services, including counseling. Future solutions to the delay in diagnosis may include universal newborn screening. (Bailey DB Jr, Skinner D, Sparkman KL. Pediatrics February 2003;111:407-416). (Reprints: Donald B Bailey Jr, PhD, Frank Porter Graham Child Development Institute, CB #8180, University of North Carolina, Chapel Hill, NC 27599).

COMMENT. The authors predict that the diagnosis of FXS is a challenge to current criteria for newborn screening candidates. Despite the growing emphasis on early diagnosis of mental retardation syndromes, most children with disabilities are not identified at birth. Greater attention to parental concerns, and regular developmental screening might enhance the earlier diagnosis of children with disabilities.

ANATOMICAL CORRELATES OF DYSLEXIA

The relation between measurements of the posterior temporal lobe, inferior frontal gyrus, cerebellum and whole brain, determined by MRI, and measures of reading, spelling, verbal intelligence and language skills was studied at the University of Florida, Gainesville, FL. Dyslexic children (14 males, 4 females) and controls (19 males, 13 females) in grades 4-6 were selected from a family genetics study. Dyslexics had specific deficits in word reading relative to the population mean and verbal IQ. Dyslexics had

significantly smaller right anterior lobes of the cerebellum, pars triangularis bilaterally, and brain volume. Measures of the right cerebellar anterior lobe and the pars triangularis correctly classified 72% of the dyslexics and 88% of controls. These neuroanatomical measures were significantly correlated with reading, spelling and language measures related to dyslexia. Anomalies in a cerebellar-frontal circuit are proposed as a neuroanatomical basis for dyslexia. (Eckert MA, Leonard CM, Richards TL et al. Anatomical correlates of dyslexia: frontal and cerebellar findings. Brain February 2003;126:482-494). (Respond: Mark A Eckert, PO Box 100244, Department of Neuroscience, University of Florida McKnight Brain Institute, Gainesville, FL 32610).

COMMENT. Anatomical deficits in a frontal-cerebellar system may lead to dyslexia. Measures of the right cerebellar anterior lobe and inferior frontal gyrus may be used to predict reading skills. Children with right cerebellar tumors have poor verbal and naming performance compared to patients with left-sided tumors (Scott et al, 2001; cited by Eckert et al).

VASCULAR DISORDERS

RISK FACTORS FOR ARTERIAL ISCHEMIC STROKE

Risk factors in 212 children (54% male; median age 5 years) presenting with a first arterial ischemic stroke (AIS) over 22 years were studied at Great Ormond Street Hospital for Children, London, UK. Patients were grouped as 1) symptomatic AIS (97), at risk because of a preceding medical diagnosis; and 2) previously healthy (115). Cerebral arterial imaging in 185 (87%) (including 115 previously healthy patients) was abnormal in 79%. Echocardiography in 104 previously healthy patients was abnormal in only 8. A comparison of the prevalence of trauma, infection, fever, varicella zoster within previous year, and hypertension within the 2 groups showed 2 significant differences: 1) trauma within previous 2 weeks was more common in the previously healthy group (23/115 cf 2/97 symptomatic); and 2) previous v. zoster was more likely in previously healthy group (68/105 cf 31/71 symptomatic). Cerebral arterial abnormalities, previous v. zoster infection, preceding trauma, recent infection, and anemia are common findings in children with AIS. Previously undetected structural cardiac abnormalities are rare. A previous medical diagnosis was identified in approximately one half. Hypertension was recognized in approximately 50%; anemia in 40%; and elevation of total plasma homocysteine or homozygosity for the MTHFR mutation in 21%. Prothrombotic screening is commonly negative. (Ganesan V, Prengler M, McShane MA et al. Investigation of risk factors in children with arterial ischemic stroke. Ann Neurol February 2003;53:167-173). (Respond: Dr V Ganesan, Lecturer in Paediatric Neurology, Neurosciences Unit, Institute of Child Health (UCL), The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. Risk factors and precipitating triggers for arterial ischemic stroke in children may be identified by clinical history and examination. Cerebral arterial imaging is usually abnormal, and some risk factors such as anemia and hyperhomocysteinemia (Hhcy) may be modifiable. Identification of all risk factors is important. Hhcy may be due to cystathionine b-synthase deficiency (Kelly PJ et al. Neurology Jan 2003;60:275-279).