

children in an urban population. The majority of juvenile delinquents have had conduct disorders at age 7. The main cause of conduct problems is linked to harsh, inconsistent parenting, but hyperactivity (ADHD) and learning difficulties contribute. Programs to improve parenting and school interventions to reduce antisocial behavior may be effective in children under 10 years, but are less successful in adolescence.

Fetal cocaine exposure and brain abnormalities, one presumptive cause of ADHD, was studied by cranial ultrasound of 134 cocaine-exposed and 132 control newborns, at the University of Florida, Gainesville, FL. (Behnke M, Eyerl FD, Conlon M, et al. J Pediatr Feb 1998;132:291-294). Subependymal, third ventricle, and choroid plexus cysts, and enlarged ventricles were identified in 17 cocaine-exposed infants and in 10 controls, but the difference was not significant. The incidence of cerebral abnormalities was lower than that previously reported. Temporal lobe arachnoid cysts have been linked to some cases of ADHD (Millichap JG. TLAC/ADD syndrome. Neurology May 1997;48:1435-1439).

VASCULAR DISORDERS

HEREDITARY HEMORRHAGIC TELANGIECTASIA

Hereditary Hemorrhagic Telangiectasia (HHT), also known as Osler-Weber-Rendu disease, was the subject of an NIH workshop, organized by the National Heart, Lung, and Blood Institute, on July 10-11, 1997. Victor McKusick (Johns Hopkins University) opened the conference, reviewing historical aspects with emphasis on genetics. HHT is an autosomal dominant disorder characterized by focal abnormalities in vascular architecture, and manifested by bleeding in specific organs, especially brain, lungs, nose, and gastrointestinal tract. Lesions are direct arteriovenous connections without a capillary bed, and range from smaller, cutaneous and mucocutaneous telangiectases to larger, arteriovenous malformations. Symptoms range from recurrent epistaxis to cerebral hemorrhage in children and congestive heart failure in adults. The prevalence is 1:10,000 rather than 1:100,000, as previously quoted. There is little data on the age of onset, the natural course of the vascular lesions, and on clinical manifestations and treatment. HHT is primarily a vascular disorder, although sometimes classified as a neurocutaneous syndrome. Two distinct genes with mutations identified in HHT are expressed in endothelial cells. One called endoglin is located on chromosome 9q33-q34 and is associated with HHT type 1, the most severe variety. Thrombosis may be a modifying factor in neurological sequelae secondary to pulmonary and cerebral arteriovenous malformations. Genetic variation in other coagulation factors might contribute to the variable expressivity of clinical phenotypes of HHT. Factor V Leiden, a common polymorphism, is a potential modifier of the HHT prognosis. Phospholipid therapy was suggested in the therapy of bleeding, especially epistaxis, which is often a chronic oozing. (Marchuk DA, Guttmacher AE, Penner JA, Ganguly P. Report on the workshop on hereditary hemorrhagic telangiectasia, July 10-11, 1997. Am J Med Genet March 1998;76:269-273). (Respond: Douglas A Marchuk, Department of Genetics, Duke University Medical Center, Box 3175, Durham, NC 21170).

COMMENT. Discussants of neurological complications of HHT were sadly lacking in this otherwise excellent report. Osler-Weber-Rendu disease presents with telangiectases on the tongue, face, mucous membranes, liver, and brain, recurrent epistaxes, intracranial hemorrhage, and bleeding in the lungs and gastrointestinal tract. The diagnosis should be considered in children with intracranial hemorrhage and mucocutaneous telangiectases.