

Giorgi C. The cerebellum contributes to higher functions during development. Evidence from a series of children surgically treated for posterior fossa tumours. Brain May 2000;123:1051-1061). Impairments of auditory sequential memory and language processing occurred with right cerebellar hemisphere tumors, whereas deficits in spatial and visual sequential memory were associated with left cerebellar tumors. Vermis lesions resulted in post-surgical mutism and behavioral disorders, including autism.

Cerebral white matter lesions and cognitive dysfunction. An MRI study in elderly subjects at the Erasmus University Medical School, Rotterdam, showed that the more severe periventricular white matter lesions affected speed of cognitive processes more than global cognitive and memory tasks. (de Groot JC, de Leeuw F-E, Oudkerk M et al. Ann Neurol Feb 2000;47:145-151).

The genetic basis of cognition was reviewed from the Institute of Molecular Medicine, John Radcliffe Hospital, Oxford, UK. (Flint J. Brain Nov 1999;122:2015-2031). Genetic approaches are limited to exploring neuronal function. Genetic mutations with a cognitive and behavioral phenotype are characterized by specific effects, but their delineation is difficult to determine in the mentally retarded. How a specific gene determines cognitive function is poorly understood.

DEVELOPMENTAL DISORDERS

REVISED DIAGNOSTIC CRITERIA OF TUBEROUS SCLEROSIS

A consensus conference sponsored by the National Institutes of Health and the National Tuberous Sclerosis (TS) Association in July 1998 provided a revised list of diagnostic criteria of TS. The major and minor features are as follows:

Major features: facial angiofibromas, ungual fibroma, >3 hypomelanotic macules, shagreen patch, retinal hamartomas, cortical tuber, subependymal astrocytoma, cardiac rhabdomyoma, lymphangiomyomatosis, renal angiomyolipoma.

Minor features: dental enamel pits, rectal polyps, bone cysts, white matter migration tracts, gingival fibromas, nonrenal hamartoma, retinal achromic patch, confetti skin lesions, multiple renal cysts.

Definite TS Complex: 2 major or 1 major with 2 minor features.

Probable TSC: 1 major and 1 minor feature.

Possible TSC: 1 major feature or 2 or more minor features.

The diagnosis of TSC is frequently made on dermatological evidence, and examination with UV light is used in screening asymptomatic relatives. Genetic criteria are not included and genetic testing is unavailable. Two genes account for TSC, *TSC1* found on chromosome 9, and *TSC2* on chromosome 16, both transmitted as autosomal dominants. Affected children of asymptomatic parents are explained by germline mosaicism. Sporadic cases make up two thirds of TSC patients, and 75% are caused by *TSC2* mutations. (Hyman MH, Whittemore VH. National Institutes of Health Consensus Conference: Tuberous Sclerosis Complex. Arch Neurol May 2000;57:662-665). (Respond: Mark H Hyman MD, University of California, Los Angeles, 11980 San Vicente Blvd, Suite 102, Los Angeles, CA 90049).

COMMENT. Dermatological features are important among the diagnostic criteria for tuberous sclerosis complex. Four major and 1 minor required criteria are dermatologic. Renal and retinal hamartomas are also major diagnostic features. Genetic criteria are not included in the list of diagnostic features.