Dandy Walker malformation

Dandy-Walker malformation (DWM) is the most common human cerebellar malformation, characterized by hypoplasia of the cerebellar vermis, cystic dilation of the fourth ventricle, and an enlarged posterior fossa with upward displacement of the lateral sinuses, tentorium, and torcular.

Pathogenesis

Although its pathogenesis is not completely understood, there are several genetic loci related to DWM as well as syndromic malformations and congenital infections. Dandy-Walker malformation is associated with other central nervous system abnormalities, including dysgenesis of corpus callosum, ectopic brain tissue, holoprosencephaly, and neural tube defects.

The Dandy-Walker variant is a milder form of the Dandy-Walker complex and is characterized by normal-sized posterior fossa, mild vermian hypoplasia, and a cystic lesion that communicates with the fourth ventricle. This syndrome has been described in association with schizophrenia, obsessive-compulsive disorder, manic episode, psychosis (delusional type), and recurrent catatonia.

Diagnosis

Hydrocephalus plays an important role in the development of symptoms and neurological outcome in patients with DWM, and the aim of surgical treatment is usually the control of hydrocephalus and the posterior fossa cyst. Imaging modalities, especially magnetic resonance imaging, are crucial for the diagnosis of DWM and distinguishing this disorder from other cystic posterior fossa lesions. Persistent Blake's cyst is seen as a retrocerebellar fluid collection with cerebrospinal fluid signal intensity and a median line communication with the fourth ventricle, commonly associated with hydrocephalus.

Mega cisterna magna presents as an extraaxial fluid collection posteroinferior to an intact cerebellum. Retrocerebellar arachnoid cysts frequently compress the cerebellar hemispheres and the fourth ventricle. Patients with DWM show an enlarged posterior fossa filled with a cystic structure that communicates freely with the fourth ventricle and hypoplastic vermis. Comprehension of hindbrain embryology is of utmost importance for understanding the cerebellar malformations, including DWM, and other related entities.

Outcome

Infants antenatally diagnosed with DWC had worse perinatal and short-term neonatal outcomes than those with mega-cisterna magna (MCM) or persistent Blake's pouch (PBP). Those with associated anomalies had uniformly poorer outcomes than those with isolated anomalies.


3) Ghali R, Reidy K, Fink AM, Palma-Dias R. Perinatal and short-term neonatal outcomes of posterior