Dandy-Walker Malformation

A Dandy-Walker malformation results from the 4th ventricle failing to close. This leads to hypoplasia of the cerebellar vermis as well as cyst development in the 4th ventricle (Dandy-Walker cyst or “Blake’s pouch”). Blockage of cerebrospinal fluid outflow from the cyst can result in noncommunicating hydrocephalus. Dandy-Walker malformations often cause several neurological abnormalities such as spina bifida.



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Characteristics

Cerebellar Vermis Hypoplasia

Silver-cerebellum-bell with Worm and Hippo-plates

The cerebellar vermis is an area located along the anteromedial aspect of the cerebellum that connects the two cerebellar hemispheres and interacts with the cerebral cortex and the spinal cord. The vermis is involved in regulating body posture and motion. In Dandy-Walker malformation, there is hypoplasia or agenesis of the cerebellar vermis which leads to cystic enlargement of the 4th ventricle.

Cystic Enlargement of 4th Ventricle

Enlarged Sisters 4-fork Vent

The 4th ventricle is a cerebrospinal fluid-filled, diamond-shaped sac located at the base of the skull. In Dandy-Walker malformation, the 4th ventricle fails to close during embryonic development which results in cerebellar vermis hypoplasia and cystic enlargement of the 4th ventricle.

Associations

Noncommunicating Hydrocephalus

Nun-communicating Hydras-in-head

Hydrocephalus is a disorder in which an excessive amount of cerebrospinal fluid (CSF) accumulates within the cerebral ventricles and/or subarachnoid spaces, resulting in ventricular dilation and increased intracranial pressure. In Dandy-Walker malformation, there is obstruction of the passage of CSF from the ventricles to the subarachnoid space leading to hydrocephalus. This is thought to be due to the failure of fourth ventricle foramina fenestration which normally allows CSF to pass from the 4th ventricle to the subarachnoid space.

Spina Bifida

Spine Buddha

Central nervous system disorders related to Dandy-Walker malformation include neural tube defects (NTD) such as spina bifida (refer to Picmonic on Spina Bifida Assessment).

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