

Pathophysiology, Clinical Features and Associated Pathology in the Posterior Fossa Anomalies:

Chiari type I malformation

-Pathophysiology

Cerebellar tonsils displaced into cervical spinal canal, small posterior fossa

-Clinical features

Usually presents in late teens or adult years; wide variety of neurologic symptoms caused by the upper cervical canal compression

-Associated Pathology

Syringomyelia, syringobulbia, scoliosis, skeletal anomalies

Chiari type II malformation

-Pathophysiology

Cerebellar vermis and brain stem displaced into cervical spinal canal

-Clinical features

Presents at birth or early infancy; lower brain stem and cranial nerves dysfunction; could be medical emergency

-Associated Pathology

Myelomeningocele and other lumbosacral neural tube closure defects, hydrocephalus, syringomyelia

Chiari type III malformation (very rare)

-Pathophysiology

Cerebellum displaced into large occipital encephalocele

-Clinical features Respiratory and swallowing disorders, cranial nerves deficits, dystonias; often fatal

-Associated Pathology

Corpus callosum agenesis, tentorium dysplasia, midbrain deformities

Dandy-Walker malformation

-Pathophysiology

Cyst-like dilation of the fourth ventricle, enlarged posterior fossa, hypoplasia and anterior rotation of cerebellar vermis

-Clinical features Very heterogeneous in presentation, depending on associated pathology; ataxia, brain stem dysfunction, mental retardation (varies), hydrocephalus;

-Associated Pathology

Corpus callosum agenesis, brain stem anomalies, hydrocephalus

Jourbet's syndrome (extremely rare)

-Pathophysiology

Cerebellar vermis aplasia

-Clinical features

Motor hypotonia, ataxia, behavioral delay

-Associated Pathology

Occipital meningocele, scoliosis, hydrocephalus, hepatic fibrosis

Cerebellar disruptions (very rare)

-Pathophysiology

Cerebellar tissue loss

-Clinical features

Motor deficits, mental retardation, often early death

Pontocerebellar hypoplasia (very rare)

-Pathophysiology

Pontine hypoplasia, cerebellar hypoplasia

-Clinical features

Severe developmental disorders, seizures, often early death

Rhombencephalosynapsis (extremely rare)

-Pathophysiology

Cerebellar hemispheres fusion, vermis agenesis, fusion of dentate nuclei and superior cerebellar peduncles

-Clinical features

Variable presentation; mental retardation, epilepsy, spasticity common

-Associated Pathology

Hydrocephalus, ventriculomegaly