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Hydrocephalus in Dandy Walker malformation

Ibrahim Shoukry[1], Hadeer Abdelghaffar[2], Marian Yousry[1], Tarek Farid[3], Heba Alawady[2]
[1] *Cairo University, Egypt*
[2] *Fayoum University, Egypt*
[3] *National Research Centre, Giza, Egypt*

Introduction: Dandy Walker malformation consists of cystic expansion of the fourth ventricle in the posterior fossa and midline cerebellar hypoplasia resulting from developmental failure of the roof of the fourth ventricle during embryogenesis. Dandy walker variant is part of the Dandy Walker complex, characterized by partial agenesis of the vermis resulting in communication between the fourth ventricle and cisterna magna.Our aim is to correlate between size of head circumference and associated hydrocephalus and other structural anomalies in Dandy Walker complex.

Methods: Sixteen cases with Dandy Walker complex were retrospectively reviewed, eleven males and five females with age range from 4 months to 3 years. Neurological assessment and MRI brain were done for all case; digital EEG was done if seizures reported and CSF flowmetry when hydrocephalus was present.

Result: Thirteen cases were offspring of first degree consanguinity (81%). Eleven cases had psychomotor retardation (68%) with hypotonia in four cases, ataxia in three, spasticity in two and dystonia in one case. Microcephaly (below third centile) was present in seven cases (43.7%). Abnormal EEG was reported in five cases. MRI showed Dandy Walker malformation in twelve cases (75%) and Dandy Walker variant in four cases (25%). Five cases had hydrocephalus (31%) and three had agenesis of corpus callosum (18.7%)

Conclusion: In Dandy Walker complex there is a high incidence of consanguineous marriage denoting an underlying genetic etiology. Whereas fourth ventricle dilatation is a common finding in Dandy Walker malformation, hydrocephalus of the whole ventricular system is not commonly associated. Head circumference is microcephalic in most cases particularly with agenesis of corpus callosum. Macrocepaly occurs when true hydrocephalic changes are seen and confirmed by CSF flowmetry studies.