Dandy Walker syndrome

Dandy-Walker syndrome is a set of abnormalities of the posterior fossa including three modalities: classic Dandy Walker malformation, Dandy-Walker variant and mega cisterna magna.

Insults to developing cerebellar hemispheres and the fourth ventricle are believed to be the cause of malformation.

Hydrocephalus is variably associated with each of these, and DWC-associated hydrocephalus has mostly been treated by shunting, often with 2-compartment shunting

Endoscopic treatment of DWC-associated hydrocephalus should be strongly considered as the primary management in place of the historical standard of creating shunt dependence [1)].

Three cases of Dandy-Walker modalities are reported: one case of classic Dandy-Walker malformation, one case of Dandy-Walker variant, and one case of false Dandy-Walker. In the first two cases the patients underwent legal abortion, whereas in the last one a healthy male newborn was delivered in the week 38 of gestation.

Malformations in the posterior fossa, including Dandy-Walker syndrome, are still a challenge in prenatal diagnosis. Technical developments in imaging, such as in three-dimensional sonography and magnetic resonance, allow higher resolution and multiplanar images for an easier diagnose. There is a high rate of false positive, particularly before the 18th week of gestation. It is advisable not to establish a final diagnose before that week [2)].

Case report

Our patient was born from noncomplicated pregnancy, noncomplicated nontraumatic vaginal delivery at term, excellent Apgar scores, without peculiarities in clinical status. She was brest-fed by the 42nd hour of life when she had rightsided seizures during sleep that repeated for five times in next 24 hours. Brain Ultrasound (US) revealed clot in left lateral ventricle, slight dilatation of left ventricle, both sided periventricular echodensity, ischemia, slight enlargement of forth ventricle and a bit smaller cerebellum. There was no visible flow through left transverse, superior sagittal and straight sinus. Magnetic Resonance (MRI) confirmed the finding and showed thrombosis of left and right transverse venous sinuses and confluence of sinuses. Electroencephalogram (EEG) showed leftsided focal changes. The newborn was treated with phenobarbiton for 8 days and had no convulsions during that period. All coagulation parameters, homocistein, lipoproteins (a) and D-dimers were normal. There were no mutations on FV R506Q, PT 20210A, MTHFR 677C/T. No antiphospholipides were found. Heart US showed no structural anomalies. No other patology or risk factors were present at the time. Before discharge, US showed hydrocephalus. Flow in affected sinuses was visible with color Doppler. MRI showed recanalization of affected sinuses, also hydrocephalus and presentation of Dandy Walker On EEG there was borderline finding. Due to progression of hydrocephalus ventriculo-peritoneal shunt was placed. In age of 1 year EEG was slower for age but without focus. Neurological development was normal for age. The question is whether this child had intrauterine insult and inception of Dandy Walker with further postnatal progress of thrombosis and evolution to full picture of Dandy Walker with hydrocephalus OR thrombosis that led to development of hydrocephalus and Dandy Walker malformation in this child were accidental coexistance [3)].

1)
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2)
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3)
Gverić-Ahmetasević S, Colić A, Gverić T, Gasparović VE, Pavlisa G, Ozretić D. Coexistance of cerebral sinovenous thrombosis and Dandy Walker malformation in newborn. Coll Antropol. 2011 Jan;35 Suppl 1:303-7. PubMed PMID: 21648352.