Dandy-Walker Spectrum: Degree of vermian dysplasia (not “cyst” size) determines prognosis

Dandy-Walker Spectrum features:
- Vermian dysplasia or hypoplasia
- 4th ventricle dilated or encysted
- Vermian fusion anomalies
- Cisterna magna enlarged
- Cerebellar and labyrinthine anomalies often coexist

Dandy-Walker Malformation PLUS
- Cerebellopontine hypoplasia...
- Poor outcome expected

Dandy-Walker Malformation MINUS
- Less vermian dysplasia...
- Even better outcome

Dandy-Walker "variant"
- This fetus (a) had other congenital anomalies = Wolf-Hirschhorn syndrome
- These 2 (b,c) had no other anomalies ... neuropathology and autopsy MRI were normal

Pseudo Dandy-Walker variant
- US scan plane too coronal gives false appearance

Mega cisterna magna: vermian fully formed
- Fastigium & lobulation normal
- IV ventricle “closed”

Vermis fully formed but small
- IV ventricle “closed”

Arachnoid cyst: vermian fully formed
- Fastigium & lobulation abnormal due to extrinsic compression

Vermian fusion anomalies
- Joubert's syndrome
- Rhombencephalosynapsis

Note: Rhombic lip forms cerebellum and cochlear nucleus, rhombencephalon induces otocyst. Cerebellum and labyrinthine anomalies often coexist.