We report the case of a 33-year-old primipara with a normal and completely investigated pregnancy (first and second trimester screening).

Ultrasound examination at 26 weeks’ gestation revealed the appearance of a translucent, round-shaped in axial section, well-defined image localized on median line, beneath tentorium, cranial to cerebellar vermis who was pushed in the caudal direction. MRI exam. denied the presence of any cystic structure with described localization, denied the presence of any focal lesion, specifying the slightly diffuse hypoplastic appearance of cerebral parenchyma, above and below the tentorium.

Follow-up 3D ultrasound examination at 31 weeks’ gestation has demonstrated that the 2/3 cranial vermis was replaced by a translucent structure with the same features as described above. The lesion was accompanied by significant hypoplasia of cerebellum (cerebellar transverse diameter under 2-nd percentile) and enlarged cisterna magna. Both hemispheres were with regular contour aspect and homogeneous hypoechogetic content.

Ultrasound evaluation revealed at 35 weeks the appearance of a significant Blake pouch cyst, mega cisterna magna (13 mm) and hydramnios (amniotic index 30 cm) which required amniocentesis for maintaining the amniotic index under 20 cm.

Any other structural anomaly or vascular abnormality was not found either in second trimester or in 3-rd trimester of gestation.

Cesarean section was performed at 38 weeks’ gestation. Postnatal MRI was not performed yet, but the infant had unremarkable neurological development at 1 month of extrauterine life. Parents refused genetic studies.