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Dolichocephaly, variant of normal. Microcephaly, a diagnosis that is seldom confirmed postnatally

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Objective

To emphasize that, despite the apparent complexity of the case, dolichocephaly is a variant of normal and microcephaly is seldom confirmed postnatally.

Methods

We present a 37-year-old female, 18 weeks' pregnant when referred to our center. She has a history of mental retardation of unknown origin, as well as her mother and her sister. Her partner suffers from fetal alcohol syndrome and personality disorders. At 18 weeks of gestation, fetal brain biometries were below the expected and we observed dolichocephaly. Amniocentesis was performed. Genetics and TORCH serologies were normal. In following controls, dolichocephaly persisted, the head circumference remained between (1-2) SD. Encephalic structures were normal. At 25 weeks of gestation, we observed intrauterine growth restriction (estimated fetal weight <p3). Doppler was normal except of the index of uterine artery pulsatility > p95. She was referred to our reference hospital, where a series of head ultrasound scans, MRI, an echocardiography and fetal growth controls were performed. Head ultrasound was normal except marked dolicocephaly, head circumference between -1 and -2 SD, and suspected delay of sulcation. The fetal echocardiogra showed an increase in postvalvular flow and altered pulmonary venous return. The exam was not optimal due to the fetal position. A follow-up echocardiogram ruled out such findings. Fetal growth controls showed intrauterine growth restriction. Umbilical artery pulsatility index was > p95 and uterine artery pulsatility index > p95. The cervical length was shortened and she was admitted to hospital at 28 weeks of gestation due to an acute pyelonephritis. Chorioamnionitis was ruled out.

Results

She was admitted again at 33 weeks of gestation due to threatened preterm labor. The baby was born by forceps delivery due to fetal distress (intrapartum pH 7. 20). The birth weight was 1560 grams, the Apgar score was 9 and 10 at minute 1 and 5 of life, resepectively. The arterial cord pH was 7. 09 Physical exam showed: No microcephaly Normal birth weight Unilateral blepharophimosis that did not interfere vision Lone hemangioma on the shoulder Normal head brain ultrasound Normal echocardiogram Cognitive development is currently normal, although the girl's growth (weight, length and head circumference) remains in low percentiles (3, 3-10 and under 3, respectively). An underlying genetic problem has been ruled out.

Conclusion

The parents' background of mental disability and the early intrauterine growth restriction made us fear a bad prognosis in the newborn. Our key sign was always the existence of microcephaly (below -2DS) that, like in most cases, was not confirmed postnatally, and the dolichocephaly, which is a variant of normal. That is why the prognosis we gave to the future parents was uncertain.