The Isolated Mildly Enlarged Cisterna Magna in the Third Trimester

Much Ado About Nothing?

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Case—A 30-year-old woman, gravida 1, presents at 32 weeks for sonography to assess fetal growth. She is otherwise healthy, and her pregnancy has been uncomplicated. Her 18-week anatomy scan results were normal. Her serum screen for aneuploidy was low risk. On this sonographic examination, the fetus is found to have normal growth and fluid. The anatomic survey results are normal aside from a cisterna magna that measures 11 mm. The remainder of the intracranial anatomy is normal. How do you counsel her?

With advances in fetal sonography, variations in fetal anatomy and structure are being identified. In particular, posterior fossa abnormalities pose a challenging diagnostic and management dilemma, as they are often difficult to interpret, and the neonatal outcome is difficult to accurately predict. Outcomes for neonates with posterior fossa abnormalities diagnosed antenatally can range from normal to severe developmental disabilities.

The cisterna magna contains cerebrospinal fluid and is located behind the cerebellum in the posterior fossa of the brain. It communicates with the fourth ventricle via the foramina of Magendie and Luschka. Measurement of the anteroposterior diameter of the cisterna magna can be useful in diagnosing posterior fossa abnormalities.

An enlarged cisterna magna has been defined as measuring greater than 10 mm.1 The optimal time to assess the posterior fossa for diagnosis of abnormalities has been debated in the literature. Embryologically, complete posterior fossa development is thought to occur between 18 and 22 weeks. Therefore, caution in making a diagnosis of cerebellar dysgenesis before 18 weeks is warranted.2 However, false-positive findings have also been reported at later gestational ages, and some authors have recommended that the diagnosis of the different forms of vermian hypoplasia should not be made before 24 weeks’ gestation.3 If a posterior fossa abnormality is suspected, further characterization by follow-up sonography is usu-
In a similar study, Haimovici et al. found all of the neonates to have normal outcomes. In an additional series of 13 patients with an isolated enlarged cisterna magna, Long et al. found only 1 patient who showed signs of developmental motor delays (poor feeding and delayed walking at 29 months). The size of the cisterna magna in this case was not reported. The remaining 12 patients had normal neurodevelopmental outcomes. Dror et al. compared 29 children with a large cisterna magna identified in utero to 35 children with normal fetal sonographic findings. Overall, performance on 2 developmental scales was within normal limits for both groups. These findings suggest that an isolated enlarged cisterna magna is associated with a normal neonatal outcome, although these studies were limited by small sample sizes.

There has also been evidence to suggest that the size of the cisterna magna can vary due to gestational age. As gestational age increases, the average size of the cisterna magna on sonography also increases. Twickler et al. used MRI to measure the cisterna magna in 60 patients with and without central nervous system abnormalities. Their data show that as gestational age increases, the upper limit of normal for cisterna magna measurements also increases to greater than 10 mm. At 40 weeks, the 95% confidence interval approaches 15 mm. This variation of cisterna magna size by advancing gestational age has been confirmed in similar studies. The available evidence suggests that an isolated mildly enlarged cisterna magna with otherwise normal intracranial anatomy, specifically a normal cerebellum and vermis, is associated with normal outcomes.

An enlarged cisterna magna can also be seen in association with the Dandy-Walker malformation, which includes cystic dilatation of the fourth ventricle and agenesis or hypoplasia of the cerebellar vermis. Other malformations may also be present. The Dandy-Walker malformation is believed to occur during embryogenesis. It can occur sporadically but has been associated with a number of genetic disorders and exposures, including alcohol, diabetes, warfarin, rubella, toxoplasmosis, and isoretinoin. An enlarged cisterna magna in the presence of a Dandy-Walker malformation has also been associated with chromosomal abnormalities such as trisomy 13 and 18. In addition, it has also been reported in cases of trisomy 21.

Although an enlarged cisterna magna as part of the Dandy-Walker malformation is associated with poor outcomes, the importance and clinical outcomes of an isolated mildly enlarged cisterna magna, in the absence of other congenital, chromosomal, or structural abnormalities, is not well defined. This can make management and counseling challenging in this patient population.

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third trimester. Therefore, the smallest anteroposterior diameter in the axial plane from the cerebellar vermis to the inner edge of the occipital bone should be obtained, and in suspected abnormal cases, the cisterna magna should be measured in the midsagittal plane.

The size of the cisterna magna also appears to differ based on fetal sex. When stratified by sex, male fetuses have been shown to have a substantially higher rate of isolated enlarged cisternae magnae compared to female fetuses. In addition, the mean measurements of the cisterna magna appear to be larger for males. This finding is similar to those regarding lateral ventricle size, which have been shown to be larger in male fetuses.

On the basis of the available data, we believe that an isolated mildly enlarged cisterna magna in the range of 10 to 12 mm without other intracranial abnormalities in the third trimester appears to represent a normal variant and seems unlikely to be associated with long-term neurologic sequelae. Ongoing study will help better define the prognosis for an isolated enlarged cisterna magna in the third trimester; future analysis may contradict the reassuring results seen thus far. For now, however, on the basis of best available current evidence, an isolated mildly enlarged cisterna magna with otherwise normal intracranial anatomy should be considered a normal variant.

In cases such as those presented earlier, we think that it is reasonable to counsel women with a low-risk serum screen for aneuploidy that the finding of an isolated mildly enlarged cisterna magna with otherwise normal intracranial anatomy, especially in the third trimester, is usually a normal variant and therefore does not require further workup. This approach will hopefully alleviate the anxiety that accompanies a formerly ambiguous diagnosis.

References