Dandy Walker malformation and hypertrophic cardiomyopathy. *Unusual fatal association*

To the Editor

I would like to comment on the interesting case report by Kurdi et al[1] on the Dandy Walker malformation and hypertrophic cardiomyopathy.

First, antenatal care is the key strategy to lessen both maternal and neonatal morbidity and mortality. The ultrasonographic examination is necessary and accurate to delineate any potential fetal anomalies, including Dandy Walker malformation (DWM). In an American study,[2] the diagnostic accuracy of prenatal ultrasound with postnatal imaging and evaluation for associated fetal anomalies in Dandy Walker complex was carried out. Of liveborn cases, postnatal imaging confirmed prenatal intracranial findings in 50% of DW variants, and 100% of DWM, with additional CNS findings noted in 21% of all cases. Additional fetal anomalies were seen in 65% cases of DW variants, and 100% of cases of DWM. Documentation of DWM by ultrasonographic examination ultimately necessitates application of fetal echocardiography as associated cardiac anomalies were found in 41.7% of fetuses with DWM.[3] Fetal echocardiography allows accurate diagnosis of major heart abnormalities by 16-18 weeks gestation. The specificity and sensitivity of fetal echocardiography for cardiac abnormalities were found to be 98% and 42%. It has also a positive predictive value of 90% and a negative predictive value of 93%.[4] A multidisciplinary team approach including gynecologist-obstetrician, fetal ultrasonographer and echocardiographer, neonatologist, and pediatric surgeon, is, therefore, needed to successfully manage neonates with DWM.

Second, Kurdi et al[1] has substantially added a new associated cardiac anomaly to already well-known associations with DWM. The ongoing extended extracranial anomalies with DWM might indicate that it is a heterogenous constellation of intra and extracranial anomalies. Undetermined molecular mutations, physiologic immaturity, certain environmental factors, time over which polymorphism is initiated in early life, and other factors could proportionally interact to continuously expand the extracranial anomalies associated with DWM.

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Reply from the Author

We would like to thank Prof. Al-Mendalawi for his interest in our recently published rare fatal association of hypertrophic cardiomyopathy with DWM.[1] We appreciate his sharing his thoughts and experience in this field, which creates a rich scientific discussion forum. We agree that detailed antenatal ultrasonography can detect DWM; however, fetal echocardiography is not widely available in our region.[2,4] We propose routine echocardiography soon after birth in all cases to detect the rare cardiac defect, which can be initially asymptomatic. This is useful for counseling the families and providing accurate prognosis. Unfortunately, treatment options are limited and the prognosis remains universally guarded even with early detection.

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