Monozygotic twins discordant for type II congenital cystic adenomatoid malformation

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Objective
To report a case of monozygotic twins discordant for type II congenital cystic adenomatoid malformation (CCAM).

Methods
This is a descriptive study, based on a case report with literature review.

Results
The pregnant woman initially came to the evaluation at 23 weeks of pregnancy due to monochorionic and diamniotic twin pregnancy. First trimester screening had not been performed. The patient was a 31-year-old woman in her second pregnancy. Ultrasound examination performed at our hospital showed the presence of polyhydramnios and a lesion in the right lung with numerous small cysts inside in twin 2, i.e., a type II CCAM. Both fetuses had weights on the 50th percentile. Magnetic resonance imaging (MRI) revealed the presence of a multicystic image in the right lung of twin 2, predominantly of the lower lobe topography, measuring about 4 cm X 3.1 cm X 2.7 cm, apparently affecting over 50% of the lung volume. There was no significant deviation of the mediastinum. Echocardiography of both fetuses was normal. The following ultrasound examinations showed similar findings. There was no evidence of cardiac decompensation or hydrops in twin 2. Children were born by cesarean section at 36 weeks of gestation. Both were male. The first twin weighed 2,360 grams and had Apgar scores of 9 in the first and fifth minutes. The second twin presenting type II CCAM had similar measures and Apgar scores: he weighed 2,330 grams and presented Apgar scores of 8 and 9. He was clinically well and did not need to undergo surgery during his first weeks of life.

Conclusion
Monozygotic twins have an increased risk of structural defects compared with dizygotic twins. CCAM diagnosed during the prenatal period have a good prognosis except in children with large lesions and pulmonary hypoplasia. Large masses can cause mediastinal shift and lead to cardiovascular compromise resulting in fetal hydrops and death. MRI has also been helpful as an important complementation to the ultrasound examination, which can assist both in diagnosis but also in evaluation, for example, of the lesion extension.