17th World Congress in Fetal Medicine

A case of caroli disease and concominant autosomal ressesive polycytic kidney disease

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Objective

Caroli disease is a rare congenital disorder characterized by multifocal, segmental dilatation of large intrahepatic bile ducts, mostly the left segment. Rarely it could be associated with autosomal recessive polycystic kidney disease (ARPKD). However it has not been clarified whether all cases with Caroli disease are related to ARDPK.

Methods

We present a case of a 24-year-old nullipara at 18 weeks of gestation with Caroli disease and concomitant ARPDK.

Results

Ultrasound examination revealed dilated intrahepatic bile ducts, polycystic kidneys, invisible bladder and anhidramnios. MRI was essential to the prenatal diagnosis of Caroli disease and all the ultrasound findings were confirmed by MRI.

Conclusion

To our knowledge this is the first case diagnosed prenatally so early in the pregnancy.