Absent gallbladder at prenatal imaging does not always predict biliary atresia - a case report.

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Introduction

In the course of availability of prenatal high-definition ultrasound and magnetic resonance imaging (MRI) extrahepatic biliary tract pathologies come into focus as they are diagnosed more often antenatal. A predictive sign to raise suspicion of a neonatal cholestatic disease such as a biliary atresia is the prenatal non-visualization of the gallbladder.

Biliary atresia is a diagnosis of exclusion which can be evaluated through clinical signs and symptoms, laboratory studies, imaging tests such as abdominal ultrasound or hepatobiliary scan and cholangiogram or liver biopsy. Although prenatal diagnosis of biliary atresia is reported in literature, our case states that the prediction, early recognition and diagnosis of biliary atresia is not as simple as it may seem.

Case description

A 32-year old primipara underwent an ultrasound at the second trimenon which failed to identify a gallbladder. Two following prenatal MRI *(Figure 1)* underlined the absence of a gallbladder. Thus, a biliary atresia was highly suspected and further postnatal diagnostic steps were addressed.

Due to non-visualization of the fetal gallblader and the suspicion of biliary atresia the pregnancy was considered a high-risk pregnancy and the mother was referred to our center and to our spezialized department of feto-maternal medicine to have paediatric surgery and neonatal intensive care units nearby. After an inconspicuous course of pregnancy a healthy female neonate was born at the 41+0 week of pregnancy.

Abdominal ultrasound *(Figure 2, A-B)* at the first day of life revealed a 4mm gallbladder rudiment and magnetic resonance cholangiopancreatography *(Figure 2, C-F)* two weeks later showed a dysplastic gallbladder without any signs of a hepatobiliary excretion pathology.

Subsequent postnatal clinical examination and laboratory parameters were always within normal ranges (*Table 1*). The patient still remains under regular clinical observation and sonographic surveillance and did not show any signs of a neonatal cholestatic pathology till this date.

Discussion

Seven studies with a total of 280 cases report a non- visualization of the fetal gallblader. In 20% of the cases the non-visualization of the fetal gallbladder is associated with additional structural anomalies which increase the risk of chromosomal anomalies, biliary atresia and cystic fibrosis to a 9-fold, 6-fold and 16-fold higher risk.

Overall, in cases of isolated non-visualization of the fetal gallblader (170 cases) the gallbladder was later visualized in pregnancy or after birth in 70.4% of the cases, while the incidence of biliary atresia, gallbladder agenesis, cystic fibrosis and chromosomal anomaly was reported with 4.8%, 25.2%, 3.1% and 1.9%. **[1,2]**

Although, the non-visualization of the fetal gallbladder raises suspicion of a biliary atresia, the gallbladder is often visualized later in pregnancy or after birth. Therefore, our case states that the absence of the fetal gallbladder in prenatal imaging may not always be clinically relevant. If the diagnosis of biliary atresia was suspected during pregnancy due to prenatal high-definition imaging physicians should express their concerns and further therapeutic steps such as Kasai's portoenterostomy or liver transplantation cautiously until the diagnosis is more assessable postnatally.

References

1. Di Pasquo, E., et al. "Outcome of non-visualization of fetal gallbladder on second-trimester ultrasound: cohort study and systematic review of literature." Ultrasound in Obstetrics & Gynecology 54.5 (2019): 582-588.
2. Shen, O., et al. "Absent gallbladder on fetal ultrasound: prenatal findings and postnatal outcome." Ultrasound in obstetrics & gynecology 37.6 (2011): 673-677.
3. Chalouhi, G. E., et al. "Prenatal non-visualization of fetal gallbladder: beware of biliary atresia!." Ultrasound in Obstetrics & Gynecology 38.2 (2011): 237-238.

Figures & Tables

	2nd day of life	3 weeks after birth	4 months after birth	1 year after birth	range
total bilirubin (mg/dL)	7.35	3.47	-	0.77	0.2-1.3
conjugated bilirubin (mg/dL)	0.00	0.00	-	0.00	<0.3
unconjugated bilirubin (mg/dL)	7.21	3.15	-	*0.53*	*0.0-1.1*
direct bilirubin (mg/dL)	0,14	0.32	-	0.24	0.0-0.4
gamma-glutamyl transpeptidase (GGTP) (U/L)	76	83	20	*<10*	*10-54*
akaline phosphatase (U/L)	76	248	170	*153*	*131-476*
cholinesterase (kU/L)	4.99	7.71	7.79	*6.89*	*4.6-10.4*
glutamic pyruvate transaminase (GPT) (U/L)	26	65	80	*24*	*0-52*
glutamic oxaloacetic transaminase (GOT) (U/L)	67	70	82	*64*	*0-77*

Table 1: Laboratory tests in order to assess hepatic function and rule out other etiologies for cholestasis were done. Table 1 shows the relevant cholestatic parameters found in our case.

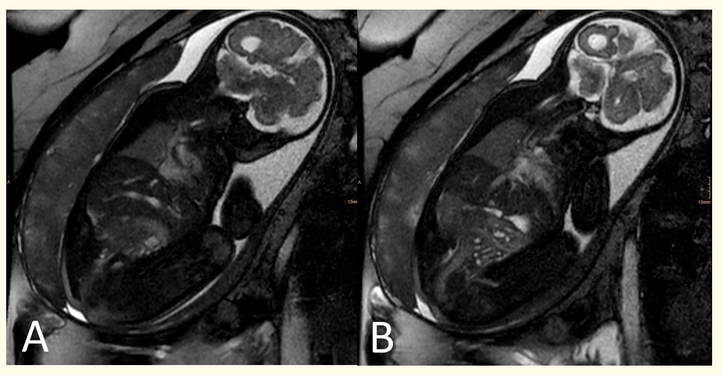

Figure 1: The performed prenatal MRI at the 27th (Figure A) and 32nd (Figure B) week of pregnancy showed no sign of a fetal gallblader.

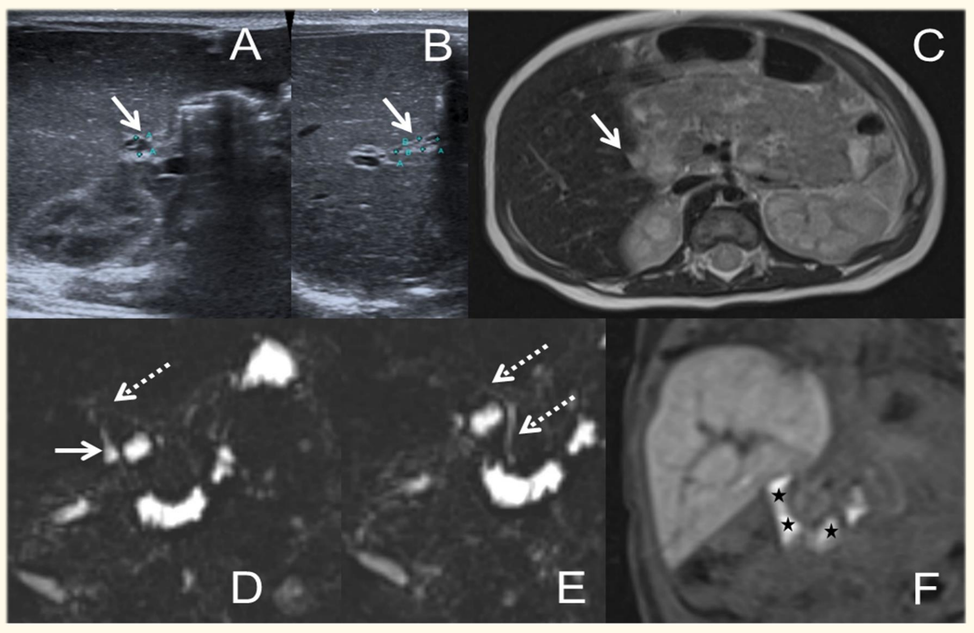

Figure 2, A – F: The 4mm gallbladder rudiment (white arrow) is shown in the abdominal ultrasound at the first day of life in the axial (Figure A) and sagittal plane (Figure B). A magnetic resonance cholangiopancreatography (MRCP) performed in august 2019 also showed the gallblader rudiment in the axial (Figure C) and coronal (Figure D, E) plane. The dotted arrow points to the ductus hepaticus communis (DHC). The MRCP showed an unrestricted hepatobiliary excretion and the contrast enhancement in the duodenum (Figure F, black stars).

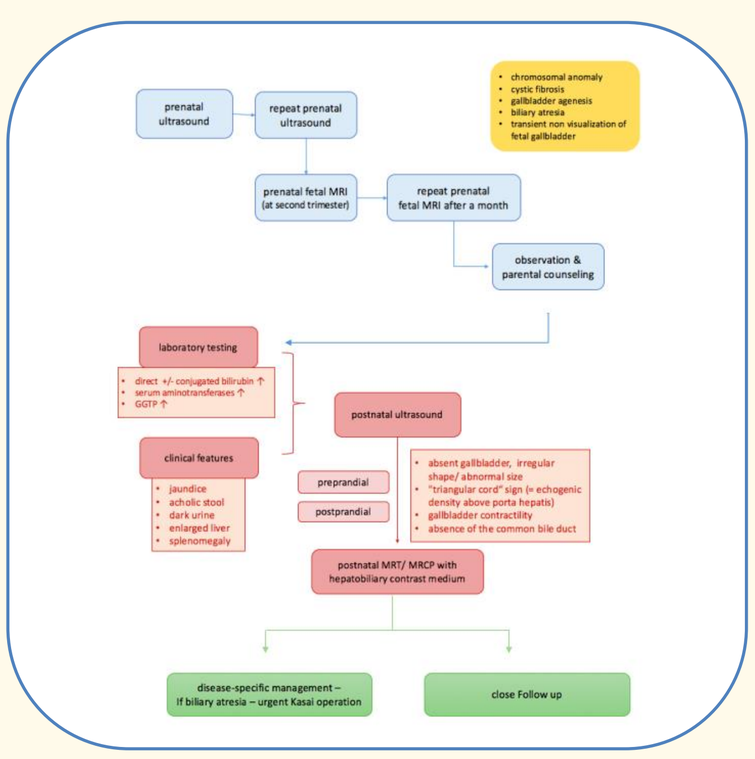

Figure 3: Proposed prenatal (blue) and postnatal (red) management after non-visualization of fetal gallbladder modified according to Chalouhi et al. **[3]**

Conclusions

Antenatal diagnosis of biliary atresia is reported in literature. However, sometimes the suspected cholestatic disease is not confirmed postnatally. Therefore, parental counseling based on prenatal imaging regarding further diagnostic, therapeutic steps and longterm outcome should be expressed cautiously by physicians.