Case Report

Prenatal Diagnosis of Phrygian Cap Gallbladder: Two- and Three-dimensional Ultrasound, Magnetic Resonance Imaging, and Three-dimensional Reconstructions

Maria Rosa Pereira Martins[1], Juliana Assis Alves[1], Mayra Martins Melo[1], João Victor Jacomele Caldas[1], Edward Araujo Júnior[1]*, Patrícia Soares Oliveira Szejnfeld[2], Pedro Teixeira Castro[3], Heron Werner[3], Liliam Cristine Rolo[1]

[1]Department of Obstetrics, Paulista School of Medicine, Federal University of São Paulo (EPM-UNIFESP), São Paulo, Brazil, [2]Department of Imaging Diagnostic, Paulista School of Medicine, Federal University of São Paulo (EPM-UNIFESP), São Paulo, Brazil, [3]Department of Fetal Medicine, Biodesign Laboratory DASA/PUC, Rio de Janeiro, Brazil

Abstract

A congenital anomaly of the gallbladder called Phrygian cap occurs when an enlarged gallbladder presents a folded shape in the fundus along the body of the organ. The incidence in adults is about 4%. The diagnosis is usually incidental during abdominal imaging for other causes, but prenatal diagnosis of this condition is rare. In this report, two cases of prenatal diagnosis of Phrygian cap were performed using two-(2D) and three-dimensional (3D), magnetic resonance imaging, and 3D reconstructions.

Keywords: Magnetic resonance imaging, Phrygian cap gallbladder, prenatal diagnosis, ultrasound

Introduction

A congenital anomaly of the gallbladder called Phrygian cap occurs when a fold forms in the fundus of the gallbladder. This form of gallbladder is named after the bonnet of a similar shape worn by the ancient Phrygians who lived in Minor Asia.[1] The incidence of this finding in children and adults is about 4%.[2] The diagnosis is usually incidental during abdominal imaging for other causes, but fetal diagnosis is rare.[3-5] The distended and folded form of the gallbladder may cause biliary stasis and simulate abnormal findings as a mass in the liver on hepatobiliary examination, but it has no pathologic significance and is usually asymptomatic.[6]

In this article, we describe two cases of prenatal diagnosis of Phrygian cap gallbladder by two-(2D) and three-dimensional (3D), magnetic resonance imaging (MRI), and 3D reconstructions.

Received: 01-09-2023 **Accepted:** 11-10-2023 **Available Online:** 26-02-2024

Video available on: https://journals.lww.com/jmut

Access this article online

Quick Response Code:

Website: https://journals.lww.com/jmut

DOI: 10.4103/jmu.jmu_109_23

Case Reports

Case 1

A 29-year-old multiparous woman (two previous cesarean sections) was referred to the Department of Obstetrics, Paulista School of Medicine, Federal University of São Paulo at 25 weeks' gestation by prenatal 2D ultrasound diagnosis of a serpentiform image in the topography of the gallbladder.

In the first obstetric ultrasound performed at the service, the following findings were observed: serpentiform hypoechoic image of 28 mm in hepatic topography, suggestive of choledochal stenosis, and liver dimensions close to the upper limit of normality, with a diameter of 37 mm in coronal section.

During the follow-up, a change in the characteristics of the gallbladder was observed on 2D ultrasound, which was confirmed by 3D ultrasound in the rendering mode using

Address for correspondence: Prof. Edward Araujo Júnior, Rua Belchior de Azevedo, 156 Apto. 111 Torre Vitoria, São Paulo, Brazil. E-mail: araujojred@terra.com.br

How to cite this article: Martins MR, Alves JA, Melo MM, Caldas JV, Araujo Júnior E, Szejnfeld PS, *et al*. Prenatal diagnosis of Phrygian cap gallbladder: Two- and three-dimensional ultrasound, magnetic resonance imaging, and three-dimensional reconstructions. J Med Ultrasound 2025;33:73-6.

Abbreviations

ACS	Acute coronary syndrome
ACS--FS	Taiwan Acute Coronary Syndrome Full Spectrum Registry
CABG	Coronary artery bypass grafting
DCF	Data collection form
ECG	Electrocardiography
GRACE	Global Registry of Acute Coronary Events
ICF	Informed consent form
IRB	Institutional Review Board
MI	Myocardial Infarction
NHIRD	National Health Insurance Research Database
NSTE--ACS	Non--ST--segment elevation acute coronary syndrome
PCI	Percutaneous coronary intervention
STEMI	ST--segment elevation myocardial infarction
T--ACCORD	Taiwan Acute Coronary Syndrome Descriptive
T--FORMOSA	TSOC--Fully Organized Registry for the Management Of Symptomatic ACS Study
TIMI	Thrombolysis In Myocardial Infarction
TSOC	Taiwan Society of Cardiology
TSOC ACS--DM	Acute Coronary Syndrome--Diabetes Mellitus Registry of the Taiwan Society of Cardiolog

the inversion mode postprocessing. At the 32nd week of gestation, fetal MRI was performed and demonstrated a topical gallbladder, with adequate repletion by content of high signal on T1-weighted, hyposignal on T2-weighted stem and intermediate signal on T2-weighted TrueFISP, highlighting variant morphology, being more elongated in its anteroposterior axis, besides the presence of fold in the fundic portion, and denoting gallbladder in Phrygian cap [Figure 1].

At 38 weeks and 4 days, a cesarean section was performed due to fetal tachycardia. A male newborn was delivered with Apgar scores of 8 and 9 for the 1st and 5th min, respectively, and birth weight of 3730 g. A postnatal abdominal ultrasound confirmed the intrauterine diagnosis. Both were discharged uneventfully to 3 days after delivery.

Case 2

A 31-year-old primiparous pregnant woman was regularly followed during pregnancy without complications, and at 27 weeks' gestation, a hypoechoic, regular, and elongated image were observed in the location of the gallbladder. The patient was referred to the Department of Fetal Medicine, Biodesign Laboratory DASA/PUC, and MRI (Magneton Avanto; Siemens. Erlanger, Germany) performed at 30 weeks gestation showed the cystic image with iso-hyperintense signal on T2-weighted sequences and confirmed the diagnosis of an elongated gallbladder.

T2-weighted TRUFI sequences were captured, and using the software 3D Slicer (v. 4.11; Birmingham, UK), 3D images were produced, demonstrating the elongated gallbladder and its proportion with the liver and stomach [Figure 2 and Video S1].

After spontaneous labor at 37 weeks gestation, a male newborn was delivered vaginally with a height of 50 cm and a birth weight of 3395 g. Both mother and newborn were discharged after 2 days. In the 2nd week of life, an abdominal ultrasound was performed and showed an elongated and curved gallbladder with normal function after breastfeeding, confirming the diagnosis of Phrygian cap gallbladder [Figure 3].

Discussion

The gallbladder is a diverticulum that is part of the extrahepatic biliary tree and is located on the visceral surface of the right lobe of the liver.[5] The gallbladder and cystic duct originate from the cranial bud of the hepatic diverticulum and develop in the anterior wall of the primitive midgut during the 4th week of embryogenesis. The pyriform morphology is acquired at the 12th week of gestation.[4]

In cases of obstructive biliary tract malformations, the mechanism that develops their phenotypic morphology is based on two main theories: the newest theory – postulates that pancreatic proteolytic enzymes cause epithelial and mural destruction, the oldest theory – postulates that the increased pressure inside the "obstructed" (or developing) biliary segment can increase the intraluminal pressure and dilate this segment.[7] In a recent case of prenatal diagnosis of a choledochal cyst, the longitudinal growth of an obstructed biliary tract of the fetus was demonstrated, forming a pear-shaped cyst and demonstrating the longitudinal development of the biliary tract.[8]

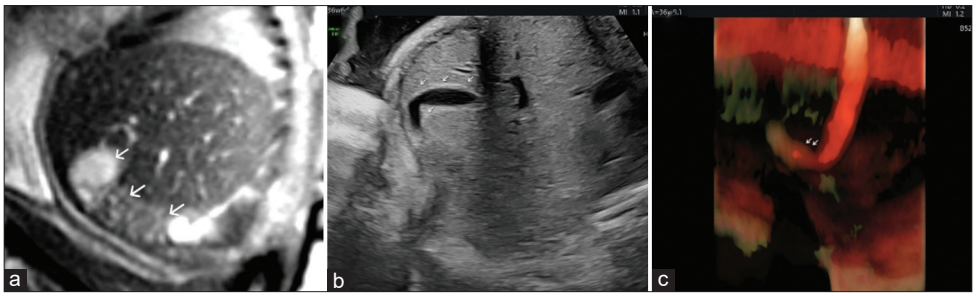

Figure 1: Case 1. (a) T2-weighted TrueFISP magnetic resonance image sagittal view at the 32nd gestational week showing a more elongated gallbladder than usual, with consequent elbowing of the fundic portion (arrows). (b) Two-dimensional ultrasound axial view at the 26th gestational week demonstrating a serpentiform hypoechoic image in hepatic topography, suggestive of choledochal stenosis (arrows). (c) Three-dimensional ultrasound at the 28th gestational week in the rendering mode using the inversion mode postprocessing showing the enlarged gallbladder, folded in its distal segment (arrows)

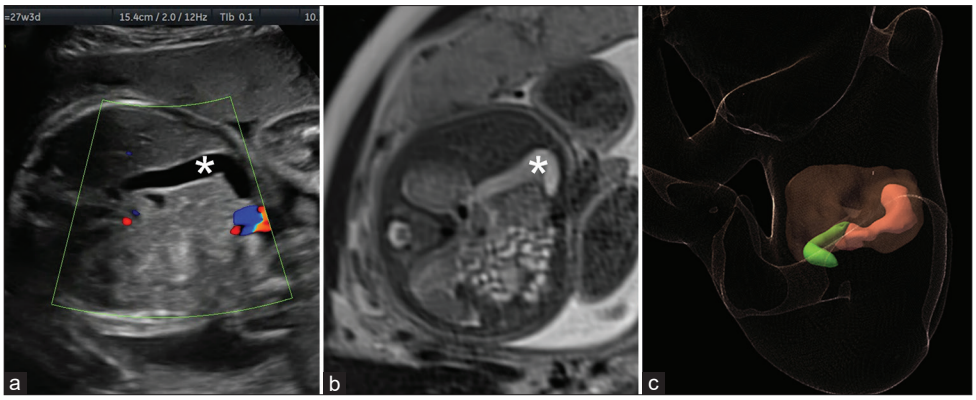

Figure 2: Case 2. (a) Axial view of two-dimensional ultrasound at the 27th gestational week demonstrating the enlarged gallbladder, folded in its distal segment (asterisk). (b) T2-weighted magnetic resonance image (MRI) axial view at the 30th gestational week demonstrating the isointense gallbladder, increased in length, and folding of its fundus (asterisk). (c) Three-dimensional reconstruction from MRI scan data. The elongated gallbladder (green) and its proportion with the liver (brown) and the stomach and duodenum (red)

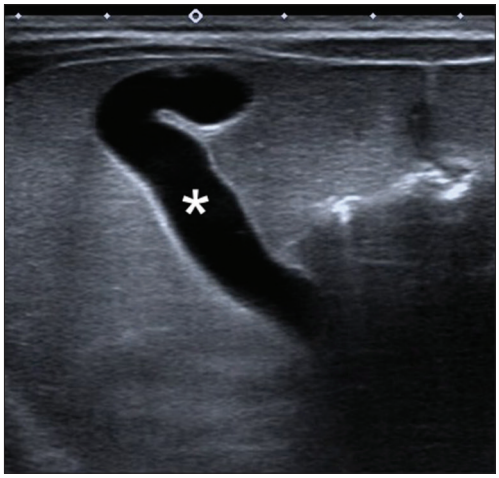

Figure 3: Case 2. Postnatal abdominal ultrasound demonstrating the Phrygian cap gallbladder (asterisk)

The congenital abnormal growth of the gallbladder called Phrygian cap affects 4% of the general population. This incidence may be underestimated because, in most cases, the diagnosis is incidental due to the lack of associated signs or symptoms.[1-4] The anomaly was first described by Boyden in 1935.[4,6] It is a fold between the body and the base of the gallbladder that loses its pyriform aspect and acquires the shape of a cap.[6] It became known as the Phrygian cap because it resembled the hood worn by ancient Greek slaves living in Phrygia (actually Turkey) as a symbol of liberation.[1,2]

The malformation of the gallbladder alone has no clinical manifestations; however, it may cause cholestasis, which increases the risk of alithiasic or lithiatic cholecystitis.[3,6] In both reported cases, there were no associated signs or symptoms. The diagnosis was made incidentally during routine prenatal examinations for congenital malformations, in agreement with the findings in the literature.[3-6]

Phrygian cap gallbladder can be visualized by high-resolution 2D ultrasound, MRI, endoscopic retrograde cholangiopancreatography, cholangiography, and cholecystography; however, the MRI should be the first choice.[1,3] Prenatal diagnosis of this condition is rare. In a study that included 92 fetuses and evaluated the development and presence of morphologic changes in the gallbladder, only two fetuses presented a Phrygian cap.[5] In both described cases, 2D ultrasound was able to detect the fetal gallbladder anomaly during the prenatal period. MRI and 3D ultrasound were used to complement and clarify the diagnosis and for parental counseling.

The differential diagnosis should include the presence of a pouch or diverticulum, bilobed, septated, or hourglass gallbladder.[3] In some cases, the images may simulate the presence of a liver tumor.[4,5] In Case 1, the initial diagnosis was choledochal stenosis, which was ruled out a few weeks later in pregnancy. In cases of Phrygian cap gallbladder, exeresis is indicated only in the presence of symptoms or other pathologies.[3] The association with polyps larger than 10 mm or with gallbladder wall thickness >5 mm indicates prophylactic cholecystectomy due to malignant potential.[4]

In this article, we performed a 3D reconstruction of the fetal Phrygian cap gallbladder from MRI scan data, which clearly showed the spatial relationships between the gallbladder and the adjacent organs such as the stomach and liver. Previous case reports have shown the relationships between fetal abdominal structures (liver hemangioma and Caroli's disease) and adjacent organs, providing important information to the multidisciplinary team and parents.[9,10]

In summary, Phrygian cap gallbladder has a low incidence and is rarely described in prenatal diagnosis. It is a morphological alteration that increases the risk of biliary pathologies such as collections and cholelithiasis and should be part of their differential diagnosis. MRI is the main prenatal diagnosis method for this condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information

to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Hanif S. Phrygian cap gall bladder: A rare entity. J Surg Pak 2021;26:43-4.
2. Kannan NS, Kannan U, Babu CP. Congenital bilobed gallbladder with Phrygian cap presenting as calculus cholecystitis. J Clin Diagn Res 2014;8:ND05-6.
3. Kulkarni V, Ramteke H, Lamture Y, Gharde P, Nagtode T, Rewale V. A rare incidental finding of Phrygian cap in a case of pyloric perforation. Cureus 2022;14:e32451.
4. Rafailidis V, Varelas S, Kotsidis N, Rafailidis D. Two congenital anomalies in one: An ectopic gallbladder with Phrygian cap deformity. Case Rep Radiol 2014;2014:246476.
5. Kokilavani J, Indiran V. Phrygian cap. Abdom Radiol (NY) 2018;43:1264-5.
6. van Kamp MJ, Bouman DE, Steenvoorde P, Klaase JM. A Phrygian cap. Case Rep Gastroenterol 2013;7:347-51.
7. Davenport M, Basu R. Under pressure: Choledochal malformation manometry. J Pediatr Surg 2005;40:331-5.
8. Castro PT, Araujo Júnior E, Fazecas TM, Ribeiro G, Macedo N, Werner H. Choledochal cyst theories going pear-shaped? Evolution of choledochal cyst during intrauterine life in a case evaluated using magnetic resonance imaging and postnatal outcomes. J Obstet Gynaecol Res 2021;47:4456-60.
9. Castro P, Werner H, Matos AP, Peixoto-Filho FM, Andrade CV, Araujo E Jr. Caroli's syndrome evaluated by ultrasound and magnetic resonance imaging during pregnancy. Ultrasound Obstet Gynecol 2020;56:125-7.
10. Castro PT, Ribeiro G, Fazecas T, Peixoto-Filho FM, Araujo Júnior E, Werner H. Fetal giant liver hemangioma: Ultrasound, magnetic resonance imaging, three-dimensional reconstruction findings and perinatal outcomes of two cases. J Clin Ultrasound 2023;51:652-5.