Congenital Anomaly of Gall Bladder Assessed Through Magnetic Resonance Cholangiography in Relation to Its Clinical Relevance

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Abstract

Objective: The overall incidence of Phrygian cap is about 04%. It is an anomaly of gall bladder found congenitally. It can be a misguided pathological diagnosis often seen mimicking a liver mass on imaging of the hepato-biliary tree. Although Phrygian cap is a congenital anomaly, it does not show any pathological significance, In light of this condition, the purpose of this research was to find out the congenital anomalies of the gallbladder through magnetic resonance cholangiopancreatography (MRCP) along with its clinical significance.

Methods: A total of 377 patients were selected with a number of 192 females and 185 males, with ages ranging from 16-90 years, with a mean age of 48 years, who had undergone MRCP for diagnosed stone in gallbladder or bile duct on clinical basis, carcinoma of pancreas and inflammatory changes of pancreatic and / or bile ducts. The patient's imaging was done using 1.5-T superconductive magnet which was four-channel phase-arrayed body coil and breath-holding technique was utilized, using multi-sliced T2-weighting half-Fourier acquisition single-shot turbo spin echo (HASTE), MIP reconstruction, and also single-shot T2-weighted turbo-spin-echo sequence rapid acquisition with relaxation enhancement (RARE) having varying thickness of slices.

Results: The MRCP imaging results demonstrated extra hepatic biliary tract anomalies in 94 out of the 377 patients which represented a frequency of 24.93%. However, a Phrygian cap was found to be present in 6 patients with the frequency of 1.6% with 4 females and 2 males.

Conclusion: From the results that we achieved, it was made quite apparent that congenital anomalies can be classified as a complex spectrum of variations that have a propensity to occur on a regular basis, emphasizing their value for attention in clinical as well as surgical settings along with the need to be readily identifiable with MRCP.

Keywords: Gallbladder, congenital anomalies, magnetic resonance imaging, cholangiopancreatography, magnetic resonance.

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Introduction

In studying the various characteristics and congenital anomaly of an organ through ultrasonography, gall bladder is one such organ, which can be easily visualized. Any anomaly might affect in addition to location of gall bladder, number, form or different size. This is a part of the extra hepatic biliary apparatus, being responsible for the storage of bile that is synthesized by the liver[1]. First removal of gallbladder (cholecystectomy) was performed in 1882 following which anomalies and variation in biliary tract were identified[1] such as, variations in the shape, position and numbers of gallbladder. Fifteen percent to 20% of patients will have altered anatomy during development showed variations in bile duct and anomalies in biliary appa-

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ratus. One of the most dangerous situations is the variations in cystic duct with the accompanying cystic artery, choledochal cyst and Phrygian cap[2]. Choledochal cysts are linked to anomalous union of pancreatic-biliary duct, in which pancreatic duct and common bile duct form a union outside duodenal wall, forming a long common channel having a length of above 15 mm. Biliary tree's anatomy remains one of most variant areas of the body. Gallbladder is an organ shaped like a pear that is affixed to the under surface of segments IVB and V of the liver. It has no capsule. There is a slight out pouching of the distal gallbladder called Hartman's Pouch, which tapers distally to the cystic duct, which contains valves of Heister. The joining of both cystic and bile duct occurs at the confluence of common hepatic (proximal) and common bile (distal) ducts. The ampulla of Vaterin duodenum receives the emptying of common bile duct. Bile acid's flow into duodenum is controlled by sphincter of Oddi. The proximal common hepatic duct branches into both right as well as left hepatic radicals in liver. These radicals eventually branch into smaller intrahepatic ducts. There may be small ducts that traverse uninterrupted towards gallbladder from the bed of gallbladder in the liver, called the ducts of Luschka that may result in postoperative bile leaks if not identified and addressed at the time of surgery, which can lead to post-operative complications. The cystic artery supplies blood to gall bladder, which usually is the branch of right hepatic arterythat in turn branches from common hepatic artery. There is no formal venous structure associated directly with the gallbladder. A commonly used landmark is the triangle of Calot. There are two definitions of this anatomical landmark. The original description uses cystic artery, cystic duct, and liver. The more commonly used description of Calot's triangle is common hepatic duct,cystic duct and undersurface of liver. A latter description helps in identifying cystic artery, which is presentinside the triangle beneath the lymph node of Calot. The portal vein lies just below the common bile duct. The surgeon must always be aware of the high incidence of the diversity of this area of the body, and there, in fact, is no such thing as standard biliary anatomy. There are many anatomical variances such as choledulco cysts, fusiform gallbladders, accessory ducts, intrahepatic gallbladders, and Phrygian cap[3,4]. During development, gallbladder sometimes shows rare anomalies known as a Phrygian cap that is the folding of over its fundus part[5,6] with 4% of prevalence rate. The term "Phrygian cap" was used when cap was worn out by Hellenic language slaves for indication of freedom / head garment well worn by residents of geographical area (present Turkey) in 1200 - 700 before Christ. In 1935, Boyden EA early defined the features of Phrygian cap[7]. It mightpretend asliver massat the time of imaging of hepato-biliary treethat suggests a tumour is present. Even duplication of gall bladder can be suspected[8]. Although Phrygian cap itself contains no significance pathologically but this uncommon anomaly can play an etiologic role in gallstones, cholecystitis, and pancreatitis. For this reason, it is vital to perform techniques of imaging properly in order to distinguish a Phrygian cap from other diagnosis. Being familiar with imaging appearances of Phrygian cap with regards to its anatomy and variants can aid in properly interpretation and accurately diagnosis it.

In patients having pancreato-biliary tract disorders, multiple techniques (radiological) are used in practice for visualization, these include ultrasonography, conventional cholangiography, biliary scintigraphy, endoscopic retrograde cholangiopancreatography (ERCP) and per-cutaneous trans-hepatic cholangiography[9]. Magnetic resonance cholangiopancreatography (MRCP) is a recent and constantly evolving imaging technique that studies the bile and pancreatic duct, with accuracy reaching 90-95%. It has been preferred over ERCP and other imaging techniques, and has been considered the ideal modality to study the pancreatobiliary tract anatomy, as it is non-invasive, ionizing radiation free, does not require anaesthesia. For example, the common and potentially severe complications of ERCP due to its invasiveness and their high risk of mortality and morbidity as pancreatitis, haemorrhage, bowel perforation, and infection are not encountered using

MRCP; Due to these reasons in this study we relied on MRCP images to demonstrate the anatomic variations and anomalies of the biliary and pancreatic system. For detailed visualization of pancreato-biliary tract as well as hepato-biliary tract systems, a special imaging through magnetic resonance, MRCP is used. In order to help physicians and surgeons to help understand both hepatic and pancreato-biliary tract's physiology as well as pathology for diagnosing and treating medical diseases, MRI (Magnetic resonance imaging) which is a non-invasive radiological test is done. To assess whether pancreato-biliary tract is patent, it is vital for post-operative patients that have gone through pancreato-biliary tract's surgery, since complications after surgery are commonly observed. Due to its non-invasiveness, MRCP has achieved a pivotal part in identifying pancreato-biliary tract's ductal systems. MRCP give a precise as well as crystal clear visualization of pancreato-biliary tract for finding various anomalies present in pancreato-biliary tract along in bile duct variations, to identify and diagnose a variety of differentiating disorders[11]. It is importance to identify, diagnose and treat any congenital or con-current anomaly of pancreato-biliary junction since it might increase risking of cancer formation because of long-term exposure of its epithelium to pancreatic enzymes.

Pancreatic and hepatic parenchymal changes can also be identified with the help of MRCP. It is usually very safe, well-tolerated and more importantly, non-invasive as well as having the benefit of being cost-effective. MR examinations aredone using 1.5 TESLA systems (Siemens, Avanto, Erlangen and Signa). Normal physiology of bile and pancreatic ducts are also be seen by MRCP. Compared with other techniques, however, MRCP is an advance test. Software mostly used is single shot faster spinning echo (SSFSE) and T2 weighing image or single- and/ or multisession via torso array coil. Coronal section identifies or visualizes the pancreato-biliary tract while axil section visualizes the bile and pancreatic ducts[12]. Through single-breath holding post 4-6 hours fasting for promoting filling of gall bladder, all sequences are acquired. In about 15 to 20 minutes, the whole examination is complete.

The purpose of this research is to prove to be a useful addition to the research data of our country benefiting for clinical health professionals and it will also help in the evaluation of variation and anomalies, which are related with pancreatobiliary tract. This will ultimately help in decreasing the incidence of surgical complications during the procedure.

Subjects and Methods

This was an observational, prospective,cross-sectional study. This study was done in the radiological department of AKUH and DUHS in eight months from May 2012 to December 2012. Total 377 no of patients was selected from the radiological department of AKUH and DUHS. Before examining the patients, they were booked. A 6-8 hours fasting period for patients was preceded before examination, the hospital provided loose gowns to them before examination. Every patient was sought for written as well as informed consent. Procedure was brief before examination and described in detail. Machine's tunnel, sound was explained along with breath-holding technique. For communicating with technologist, call bell was given to each patient. 20 minutes prior to examination, around 350-500 ml of pine-apple juice was provided to the patients that were given as an oral contrast (negative) before using MRI scanner, MRCP was carried out with imaging gained through the usage of 1.5 TESLA (Magnetom, Vision, Erlangen, Siemens and Germany). The images were obtained on 1.5 TESLA via Siemens. A large circular magnet surrounded around 1.5 meters long tunnel of MRI scanner. Through a sliding couch, patients were taken in towards scanner. All around, behind and on the body part being examined, a receiver was placed in order to help detect tiny signals that the patient's body was emitting. Mobility and silence of patients were maintained during each image that was being scanned. In approximately 15-20 minutes, the whole process of examination was completed. During the whole procedure, with the help of

a monitor present in control room, the participant and radiographer both sat down to observe the proceedings of the examination. Through statistician's and epidemiologist's help, Open Epi 3.01 calculator calculated the required sample size with the prevalence kept at 43% of anatomic variability as well as congenital anomalies. Keeping an error margin of 5%, significance level at 5% and confidence interval at 95%, sample size was calculated to be 377. Therefore, the sample size in the study was of 377 patients.

Inclusion criteria was patients having been previously diagnosed as a case of pancreato-biliary disease through ultrasonography and other techniques, post-pubertal patients of either gender, specifically on imaging from MRCP were enrolled and which had clear anatomical views were included in the study.

Exclusion criteria included all the patients that had underwent MRCP however the imaging was not up to the mark such as due to technical issues, demented patients, claustrophobic, those that could not hold their breathe for above 30 seconds and those patients who had a non-MRI compatible implants, pace makers or mechanical prosthesis.

Results

Phrygian cap was observed in about 4 females from 196 with 2% frequency. Phrygian cap was also observed in about 2 males from 181 in totalwith a 1.9% frequency. It was observed in about 1.6%. A significant p-value of <0.001 was observed in patients with Phrygian cap as compared to those without Phrygian cap. (Fig 1, 2 and Table 1.).

Table 1. Shows frequency of distribution of Phrygian cap among adult male and females through MRCP

PHRYGIAN CAP	Female	Male	Total	p-value
Present	4	2	6	
	2.0%	1.1%	1.6%	
Absent	192	179	371	<0.001*
	98.0%	98.9%	98.4%	
Total	196	181	377	

(P value at <0.001* is significant)

Discussion

Given the wide availability and frequency of radiological techniques, radiologists are now aware of malformations. Cases of double gallbladder and cystic-duct duplication (with gall congenital anomaly) exist (Boyden, 1935), and no rare anomaly has been reported in searching of studies of phrygian cap in a human subject. At the time fundus of gall bladder has flexed on its body, a "Phrygian cap" variation is formed. Concerning the bending, shape and folding of the gall bladder on its body, this may easily be identified by ultrasonography. Therefore, radiologist should at all times notify physician regarding existing of an aberrant Phrygian cap if any exists[13].

At 4th gestational week, primitive midgut's ventral wall gives rise to two buds consisting of hepatic diverticulum which gives origination to liver, biliary tract and partially to pancreas. Liver and extra-hepatic-biliary tree is formed from the cranial segment of the diverticulum, while a superior and inferior bud is formed from the caudal segment of the diverticulum. Cystic duct and gall bladder are grown from the former bud. The latter bud gives rise to the ventral portion of the pancreas. Although biliary tract is thoroughly formed and recognized by ending of 5th week of gestation, however, gall bladder keeps remaining solid up and until around 12th gestational week. Regarding congenital anomaly of the biliary tract, folding of gall bladder is a commonly anticipated anomaly; the terminology used is "Phrygian cap" that describes that the fundus of gall bladder is found to be folded. In 1935, Boyden was the first

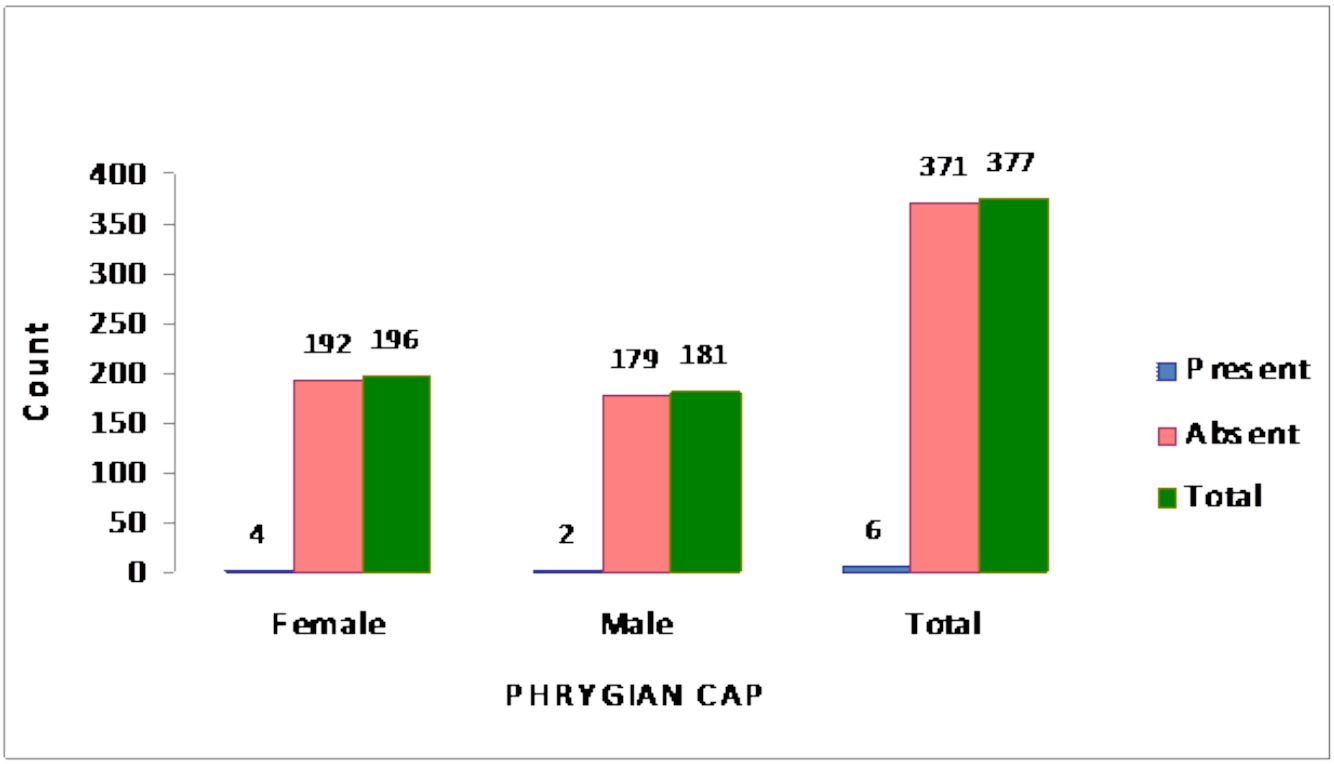

Fig 1. Depicting distribution of Phrygian cap frequencies in adult males and females through MRCP

Fig 2. Coronal MR Cholangiopancreatography (MRCP) shows the gallbladder (GB) having Phrygian Cap (PC). Common bile duct (CBD) and Cystic duct (CD) all marked and labeled

to describe it and was the one who termed it on a pre-historic cap (conical) having its top being pulled in forward direction, that the ancient Phrygia's citizen wore during the 12th century BC (Centre of present Turkey)[14]. The reported incidence of Phrygian cap is 4% normally being free of symptoms; however, it has the possibility of being mistakenly regarded as a lesion in the liver during imaging study. With the help of ultrasonography, CT, MRCP as well as in delaying scans of scintigraphy, Phrygian cap can be easily identified if it is kept in mind. Emptying of the gall bladder's "Phrygian cap" is found to be at normal rate. Removal of gall bladder ought to be done solely if deformity of gall bladder leads to symptoms such as pain in the right upper quadrant of the abdomen[15].

Below right lobe of liver under lies the normal positioning of gall bladder, precisely in inter-lobar fissure's plane. The neck of gall bladder lies inside porta hepatis, which extends caudally towards inferior border of liver. In the majority of patients (70%), the main lobar fissure is visualized onultrasound on longitudinal (linear as well as hyper-echoic) views, that lie in between right portal vein cranially and gallbladder caudally. Transversely, the position of gall bladder is present at, either posteriorly to or a portion of it lays inside main lobar fissure, in-between the liver's right lobe and liver's left lobe within medical segment. Ectopic positioning of gall bladder is most commonly found at 4 sites namely liver's left lobe, intra-hepatic (within liver), transversely and retro placed either retro-peritoneal or retro-hepatic. In patients having a situs inversus totalis or not, visualization of gall bladder can be achieved below the liver's left lobe. If gall bladder's position is within the liver (intra-hepatic), it can present as a problem diagnostically since scintigraphy will show defective mass being present within the liver. A great deal of help can be achieved in such patients through an ultrasound[16]. With identification of cystic artery through the help of angiography, any ectopically positioned gall bladder can be found. The gall bladder's ectopic positioning is less commonly found at falciform ligament, supra-hepatic (above the liver's right lobe and under diaphragm), the abdominal wall, behind the pancreas, and others. If the shape of body of gall bladder is found to be folded or bent, an easy identification of it can be achieved through ultrasonography. If the fundus of gall bladder is seen to be folded over on to the gall bladder's body, a variation in the "Phrygian cap" is formed. Lumen of the gall bladder gets sub-divided partially through a fold of mucosa which is created during deformity of "Phrygian cap". This occurs in many of the cases. However, according to the finding of this study, orientation of the folds of the fundus was found to be in upward direction. At last, residing within gall bladder, either multiple or a solitary septa might be seen[17]. The reason for bile stasis and stone formation might be directed towards these septa.

Through vigilant examinations of gall bladder in patients, such disorders can easily be excluded. Extreme rarity is observed for a gall bladder having multiple septa, known as multi-septate gall bladder. If it is present, it is characterized with multiples of septa, which can be found in either the whole of gall bladder, or only in neck portion, or body of gall bladder or in fundic portion of gall bladder. It is thought to be the result of incomplete cavitation of bud of gallbladder. Frequencies of such an anomaly are found to be more among female patients and the presenting complain of patients is mostly pain in the right upper quadrant of the abdomen. The pain is found to be of colicky type that radiates towards the right shoulder's back. Pain is mostly always relieved after undergoing removal of the gall bladder. Ultrasonography of the septa of gall bladder are seen as an echogenic band having no acoustic shadow that crosses lumen of gall bladder and should always be separated from polyps. If present, the multi-septate gall bladder might be either found alone or in combination with some other anomaly as well such as a choledochal cyst of hypoplasia. As it has been stated above that the term "Phrygian cap" has been named so for describing gall bladder having a folded fundus.

In 1935 Boyden was the first to describe it and was the one who termed it on a pre-historic cap

(conical) having its top being pulled in forward direction, that the ancient Phrygia's citizen wore during the 12th century BC (Centre of present Turkey). The reported incidence of Phrygian cap is reported to be 4% and normally being free of symptoms, however it has the possibility of being mistakenly regarded as a lesion in the liver during imaging study. With the help of ultrasonography, CT, MRCP as well as in delaying scans of scintigraphy, Phrygian cap can be easily identified if it is kept in mind[18]. Emptying of the gall bladder's "Phrygian cap" is found to be at normal rate. Removal of gall bladder ought to be done solely if deformity of gall bladder leads to symptoms such as pain in the right upper quadrant of the abdomen. In our study the Phrygian cap's rate of prevalence reported was 1.6% that can be compared to a research which reported an incidence of 3.3%[19].

Conclusion

From the results that we achieved it was made quite apparent that congenital anomalies can classified as a complex spectrum of variations that have a propensity to occur on a regular basis. A Phrygian cap is most commonly observed congenital malformation of gall bladder which might be present as an incidental finding on imaging or at the evaluation of biliary colic. Once incidental, there seems to lie no indication for a gall bladder removal. A thorough pre-operative evaluation through MRCP is needed in order to confirm diagnosis and also to detect the related anomalies of biliary tree.

Conflict of Interests

Authors have no conflict of interests and received no grant/funding from any organization.

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