Abstract:
Congenital absence of gall bladder is a very rare but well recognized congenital abnormality, the reported incidence ranging between 0.01 and 0.05. To our knowledge, this is a very rare case and all surgeons need to be aware of this unusual occurrence to prevent inadvertent injury to the dilated common hepatic duct. Laparoscopic cholecystectomy was planned on a suspected case of chronic cholecystitis and was found to have congenital absence of gall bladder and cystic duct. The diagnosis was confirmed by MRCP postoperatively. Standard investigations for chronic cholecystitis were misleading. Agenesis of gall bladder should be highly suspected whenever the gall bladder is not visualized on ultrasoundography or at laparoscopy done on misinterpreted ultrasound. If the gall bladder is not seen at laparoscopy, further procedure should be avoided and agenesis should be confirmed by a combination of imaging modalities namely CT scan, MRCP, laparoscopic or endoscopic ultrasound, and HIDA scan if available. If the gall bladder is not visualized at preoperative ultrasound, a combination of imaging modalities should be used for diagnosis without recourse to laparoscopy or laparotomy.

Keyword: congenital absence gall bladder

Case report
A 28-year-old male was referred to our surgical unit with right upper quadrant pain, which radiated to the back and precipitated by fatty meal also with pain in the right iliac fossa. General examination and abdominal examination were unremarkable except tenderness in R.I.F. Routine hematological and biochemical investigations were normal. Ultrasonography (USG) of the abdomen was reported to be showing posterior acoustic echoes arising from the region of the gall bladder bed consistent with gall bladder packed full of stones and inflamed appendix. The common hepatic duct was dilated and the lower end could not be visualized due to gaseous distention.
Introduction
Congenital absence of the gallbladder (CAGB) is a rare anatomical variation that can present a diagnostic and intraoperative dilemma to the surgeon. Most affected individuals remain asymptomatic for life. Some may present with right upper quadrant pain and dyspeptic symptoms that mimic gallbladder pathology. Routine investigations frequently fail to diagnose gallbladder agenesis, leading to operative intervention. We present herein our experience with one such case. We report a case of agenesis of the gallbladder in a 28-year-old male incorrectly diagnosed as chronic cholecystitis with cholelithiasis on preoperative ultrasound. Laparoscopic cholecystectomy with appendectomy was attempted based on history, clinical examination and sonographic findings. Laparoscopy failed to visualize gall bladder. (Fig. 3). Extensive and careful search did not reveal either ectopic gall bladder or any other abnormality in other upper abdominal viscera. Laposcopic appendectomy was done. No further procedure was attempted. The patient improved symptomatically following the laparoscopy. The diagnosis of agenesis was confirmed laparoscopically with laparoscopic exploration, and postoperative magnetic resonance cholangiopancreatogram (MRCP). Formal laparotomy was avoided.

Embryology
In 4th to 5th week of life, a 4-mm human embryo develops abud from the foregut that grows cephalad and the cranial portion becomes the liver and hepatic bile ducts. In the caudal portion of the growing bud, there develops a second bud, or diverticulum, and this is destined to become the gall bladder and cystic duct. Failure in organogenetic sequence at this point results in agenesis of gall bladder. Theories for this condition include failure of gall bladder analage from the hepatic diverticulum, or failure of recanalisation following the solid phase. Gall bladder agenesis can be part of multiple fetal anomalies and the association has been attributed to deranged development between the paired omphaloenteric and umbilical veins and the sinus venosus cordis; this results in disturbed positional development of the cells of the upper umbilical position [3,4].
Discussion
Lemary first reported agenesis of gall bladder in literature in 1701 [5,6]. The reported incidence in literature ranges between 0.01 and 0.05% [5]. The incidence is equal in both sexes from studies at autopsy where as 2-3 times more common in females in clinical cases [4]. Bennion et al described three categories of agenesis of gall bladder [7].

1. Multiple fetal anomalies (15-16%) - These patients invariably die in the perinatal period due to associated anomalies and agenesis of the gall bladder was only recognized at autopsy. The most frequently encountered malformations were cardiovascular, gastrointestinal, genitourinary, anterior abdominal wall, and central nervous system. In this group gall bladder agenesis is only a trivial anomaly.

2. Asymptomatic group (35%) - Agenesis of gall bladder was discovered either at autopsy, at laparotomy for unrelated diagnosis or by screening the family members of patients known to have agenesis of gall bladder. These patients do not have symptoms of biliary tract.

3. Symptomatic group (50%) - This major group present in 4th or 5th decades. This is usually an isolated anomaly. In symptomatic group, patients undergo surgery for right hypochondrial symptoms only to find no gall bladder at surgery. Common symptoms include chronic right upper quadrant pain (90%), dyspeptic (30%), nausea and vomiting (66%), fatty food intolerance (37%) and jaundice (35%) [8]. The possible mechanisms of symptoms include primary duct stone, biliary dyskinesia or non-biliary disorder. The usual preoperative investigations for biliary tract namely USG and oral cholecystography (OCG) are misleading. USG is highly operator dependant. Periportal tissues or sub hepatic peritoneal folds are usually focused and interpreted as thick, contracted, shrunken or scarred gall bladder [8]. Failure to see the gall bladder on scintigraphy and OCG is interpreted as nonfunctioning or diseased gall bladder [8]. Failure to outline the gall bladder at ERCP may be due to cystic duct block [5]. As a result preoperative diagnosis of agenesis of gall bladder is extremely difficult. Frey laid strict criteria for the diagnosis of agenesis of the gall bladder [9]. Intraoperatively, if the gall bladder is not visualized in its normal anatomical position, a thorough search should be carried out in the ectopic location namely intrahepatic, left sided, beneath the posterior inferior surface of liver, attached to the left lobe of liver, free floating with in the falciform ligament, between the leaves of lesser omentum, retroperitoneal, retrohepatic, retropancreatic, retroduodenal and in the anterior abdominal wall [6]. If gall bladder is not found in all these sites peroperative cholangiography is mandatory. CBD exploration should be carried out only if the cholangiogram shows calculi in CBD or CBD is dilated more than 20 mm.
Nothing should be done if no gall bladder is found in spite all these measures. Extensive dissection is required to fulfill Frey's criteria for agenesis of gall bladder. This amount of dissection is associated with complications that can be detrimental to the patient. It is a well known fact that agenesis of gall bladder is a harmless condition on its own. It is not worth performing a major operation just for the sake of diagnosis.

**Conclusion**
Recent literature suggests that when a case of agenesis of gall bladder is suspected on laparoscopy, further procedure should be abandoned and agenesis should be confirmed by diagnostic modalities such as CT scan of the abdomen, ERCP, MRCP, HIDA scan. If the agenesis of gallbladder is suspected preoperatively, it should be confirmed by MRCP and/or endoscopic ultrasound before embarking on laparotomy. Whenever available, laparoscopic ultrasound should be used to confirm the diagnosis of agenesis of the gall bladder, as it is an effective imaging modality [6]. Our case exactly fulfills these criteria as we have searched for the gall bladder in all possible sites and laparoscopic cholecystectomy abandoned with a provisional diagnosis of agenesis of gall bladder. Postoperative MRCP also confirmed the diagnosis. With a combination of these imaging modalities one should be able to diagnose this seemingly harmless condition with out recourse to laparotomy.

**References**