

CONGENITAL AGENESIS OF GALLBLADDER- A HOSPITAL-BASED STUDYPurujit Choudhury¹, Abhijit Sarma²¹Associate Professor, Department of Surgery, Gauhati Medical College, Assam, India.²Associate Professor, Department of Surgery, Gauhati Medical College, Assam, India.**ABSTRACT****BACKGROUND**

Embryologically, gallbladder is developed from pars cystic of ventral duodenal bud. During its rotation in clockwise direction towards dorsal bud, it fails to give rise to develop gallbladder or its different parts. But it is a very rare anomaly. Incidence is 1 in 1000. Gallbladder agenesis is an isolated finding in more than two-thirds (70%) of people. The person with isolated gallbladder agenesis is healthy. No treatment is needed and the prognosis is excellent.

The aim of this study is to describe the clinical profile of patients with agenesis of gall bladder.

MATERIALS AND METHODS

It is a descriptive study. Twenty two patients of agenesis of gallbladder admitted at GMC between 2009 - 2017 were studied in a descriptive way that was carried out at Gauhati Medical College with the help of much skilled and experienced surgeons in laparoscopic surgical procedure. This type of congenital anomalies of biliary tree and very particularly absent or agenesis of gallbladder can be managed by the aid of laparoscopic procedure itself without any difficulties, which is the accepted procedure of choice in comparison to laparotomy. But laparotomy was usually used to adopt in the past and by inexperienced surgeon in this field and the center where laparoscopic and laparoscopic ultrasound are not at all available. By accepting this procedure of laparoscopy, morbidity and different probable complications following open procedure can be minimised considerably. Most of the patients had typical right hypochondriac pain, which was colicky and referred to right shoulder and back, off and on vomiting, nausea and flatulent dyspepsia. The clinical features suggestive of gallstone diseases which were further confirmed by transabdominal ultrasonography and depicted very small contracted gallbladder with few stones having its posterior acoustic shadow.

RESULTS

Gallbladder was found to be absent during initial diagnostic laparoscopic assessment of intraperitoneal organs in all the patients resulting in conversion to exploratory laparotomy in five patients. Gaining much confidence on laparoscopic surgery, the other patients of congenital agenesis of gallbladder were able to diagnose exclusively by conventional laparoscopic means to have the developmental defect like agenesis of gallbladder. But with much experience and skill in hand, instead of conversion it was able to do by laparoscopic only and further confirmed by post-operative imaging evaluation.

CONCLUSION

Congenital absent or developmental defect like agenesis of gall bladder is very uncommon. With much confidence in laparoscopic surgery, the condition no longer becomes mandatory for the procedure to be converted to laparotomy for finalising its diagnosis. All the patients showed sign of improvement after surgery.

KEY WORDS

Congenital Gall Bladder Agenesis, Laparoscopic Cholecystectomy, Laparotomy.

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BACKGROUND

Developmental abnormalities of gallbladder like absence of gallbladder or congenital defect of biliary tree, mostly agenesis of the gallbladder is much uncommon congenital maldevelopment during the process of embryogenesis of gallbladder from ventral duodenal bud. This but gives rise to pars cystic and pars hepatica and it is the rotator which moves towards dorsal but to develop pancreas. Almost all cases are isolated and hence their descriptions in literature are not conclusive. About 50% of those patients present with

gallstone diseases. Preoperative diagnosis is difficult and mostly confirmed per-operatively during CBD extirpation and dissection of complete periductal fibroareolar tissue to make it complete isolation and inspecting possible sites of ectopic gallbladder. Earlier majority of agenesis can be easily diagnosed by exploration. But, now-a-days those patients can easily be diagnosed by the advancement of laparoscopic surgery and application by expert in this field. Twenty-two patients were examined at our Medical College and found to have gall stones. But per-operatively even after thorough inspection, all the patients in this study did not have gallbladder at all.

Objective

To describe the clinical profile of the patients with agenesis of gall bladder.

MATERIALS AND METHODS

It is a descriptive study. Twenty-two patients admitted at Gauhati Medical College were taken for a study in a

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descriptive way from 2009 - 2017. Twelve male patients and ten female patients with an average age of 34 years (range 18-46 years) were the sample strength in this present study. For first five patients, laparotomy was executed after a diagnostic laparoscopy to make the diagnosis final. In this study other patients were diagnosed by the way of laparoscopic procedure only. Post-operative radiology was the investigation adopted to confirm the diagnosis.

Pre-operatively, all the patients in this study were advised to get the ultrasonography done and ultrasound of the abdomen revealed gallbladder calculi with acoustic shadow. Per-operative laparoscopic ultrasound analysis depicted dense fibrous tissue in the gallbladder fossa. CBD was found to be dilated, but echo free. Preoperative MRCP or ERCP were not done, because they did not present past history of obstructive jaundice. Post-operatively, CECT/ MRCP was advised to finalise the diagnosis of absent of gallbladder. CECT clearly ascertained absent of GB in the post-operative period. Post-operative MRCP depicts absent of GB with absent of different component of biliary apparatus.

Statistical Analysis

The data was entered in Microsoft Excel 2007 and analysed using SPSS version 16.0 software. The descriptive statistical methods like mean, standard deviation, frequencies and proportion were used. Chi-square test was used for comparison of categorical variables. A 'p' value of less than 0.05 was taken as significant.

RESULTS

In this descriptive study of 22 patients, twenty patients (91%) were symptomatic for gallstone diseases. Sex incidence was found to be almost equal (M: F= 6: 5). Male is 55% and female is 45% of total number of cases in this descriptive type of study.

On laparoscopy, the gallbladder was absent in all seven patients leading to conversion to laparotomy in the initial five patients. With increasing experience, the next two patients were diagnosed conclusively by laparoscopy to have agenesis of gallbladder without the need for conversion which was further confirmed by post-operative imaging studies.

In our institution, six out of seven patients that we have encountered were symptomatic for cholelithiasis. We have found an almost equal sex incidence in our patients (M:F=4:3).

Preoperative ultrasound suggested cholelithiasis with contracted gallbladder in all our patients. None of our patients had abnormal Liver Function Tests (LFT) or a history suggestive of CBD calculi. Preoperative ductal imaging with an MRCP or ERCP was therefore not performed in any of our patients.

Complete dissection of the entire biliary system along with exploration of the possible ectopic sites of gallbladder was in the past mandatory to conclusively diagnose agenesis.[1,2,3,4] In our initial five patients, conversion to laparotomy was needed for complete exploration. In the subsequent two patients, we were able to perform a thorough exploration laparoscopically. Postoperative radiological studies were performed to document the rare abnormality and also to rule out CBD calculi. A CT scan was done in the initial six patients and an MRCP scan was done in the last patient that confirmed agenesis of the gallbladder in all these

patients. All the patients on follow-up continue to be asymptomatic.

Gallbladder was found to be absent during initial diagnostic laparoscopic assessment of intraperitoneal organs in all the patients resulting in conversion to exploratory laparotomy in the five patients. The number of patients in this study to be converted was only 5 patients out of 22. Total % of conversion is 23 only. Gaining much confidence on laparoscopic surgery, the other patients of congenital agenesis of gallbladder were able to diagnose exclusively by conventional laparoscopic means to have the developmental defect like agenesis of gallbladder. But with much experience and skill in hand, instead of conversion we were able to do surgery by conventional laparoscopic procedure only and further confirmed by post-operative imaging evaluation. All patients remained asymptomatic in post-operative follow-up period.

Complications	Number of Male Patients (N= 22)	Number of Female Patients (N= 13)	P value
Fibrosed gallbladder with stone	6(8.8%)	4(8.1%)	0.514
Laparotomy adopted	2(1.1%)	1(5%)	0.028
Asymptomatic	4(9%)	3(4.5%)	0.767
CT scan	6(8.8%)	3(4.5%)	0.079
Fibrosed GB containing multiple small calculi	4(9%)	2(4.9%)	0.081

Table 1. Detailed Clinico-radiological Evaluation of the Patients

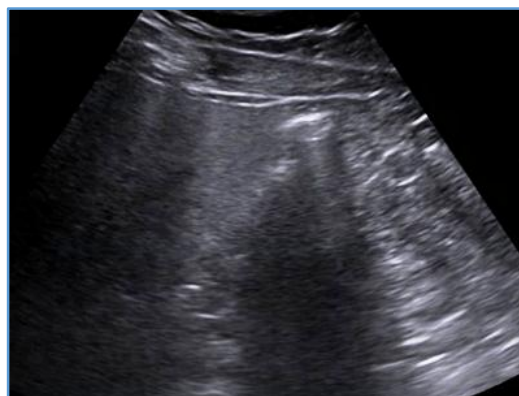


Figure 1



Figure 2

DISCUSSION

Though congenital abnormalities in the form of ductal and vascular is the rule not the exception in relation to stone disease, yet congenital defect like agenesis is very very rare as far as gallbladder is concerned. Though exact incidence is not mentioned in different small study, yet gallbladder agenesis without other parts of biliary apparatus occurs in 0.02 - 0.04% in average^[2] and this commonly appear in association to other congenital or developmental abnormalities like congenital heart diseases and lung diseases. Among PDA fibrotic disease and contraction of lung and coarctation of aorta are the most common.

Much less cases of this type have been (300 - 500 cases) [1] reported in literature. Bennion et al highlighted four separate groups of patients with absent gallbladder. 55% of patients showed symptoms of gallstone diseases. About 90 - 95% patients presented with right upper abdominal pain, flatulent dyspepsia (38%), nausea and vomiting (77%), intolerance to fatty food cannot be tolerated and appearance of jaundice. The aetiology of these symptoms are difficult to ascertain. Probable cause might be in favour of biliary dyskinesia and results in high pressure in the distal CBD which is the sphincter of Oddi region. Common hepatic duct may be dilated in these patients and act as a bile reservoir/storage. Biliary akinesia or dyskinesia results in cholestasis and might lead to infection which subsequently precipitate CBD stone formation. 33% of patients are symptom-free and the other 17 - 19% associated with many developmental defect might impact on survival of life.^[2,5]

On reviewing different studies, it was found that most of these patients complained of symptoms suggestive of gallstone disease. It is obvious by our study that twenty patients among total twenty-two in the study presented with typical symptoms of gallstone diseases. But the sample size is too small that further randomised and multicentre study might bring consensus conclusion that we might consider. Patients diagnosed presented the incidence in females as slightly higher than males that different studies opined (3: 1). Though, the proportions are said to be equal between the sexes^[6,7] as reported by autopsy. In our small series too, we found an almost equal male: female ratio (M: F= 6: 5).

Richards et al reported in his scientific forum reports that 27% of patients had jaundice and 18% had CBD calculus, they highlighted.^[8,9] In this present study, all the patients had normal liver function test on thorough biochemical analysis and no evidence of choledocholithiasis. This was determined by laparotomy in the initial five patients and confirmed by imaging by the other two in the post-operative period. All the patients in the present study had required pre- and post-operative investigations to exclude CBD stone for further management without any complications.

Per-operative diagnosis is much more important than pre-operative, because pre-operative is not easier to diagnose agenesis of this kind. Thick contracted gallbladder may be diagnosed in the Rouviere's sulcus as thick fold of peritoneum, which the radiologist gives impression by ultrasonography.^[7,9] Sonologist may give the diagnosis as calculi as seen by some echogenic shadow. Sometime, calcification in the site of porta may give rise to the mind of sinologist as being confused with calculi.

Though, ERCP is not the usual procedure pre-operatively, yet it is one of the best in this situation. In ERCP gallbladder

may not be visualised which bears different aetiology, but agenesis is the rarest explanation in this regard.^[8]

Frey opined the role of cholangiography in the diagnosis of agenesis and operative findings after complete dissection of CBD.^[3] Per-operative post-exploratory cholangiogram has much morbidity where a choledochoscope is mandatory. But most of the surgeons has the ability to ascertain possible sites of ectopic gallbladder laparoscopically only. But many also questioned. This might give rise to doubt in terms of conversion. This is the reason why role of intra-operative imaging like aid of ultrasonography comes in mind. There are different technologies of laparoscopic ultrasound and its probes are available and these may be fiberoptic malleable to rigid which make the laparoscopic identification of CBD, its branches and different tiny structures in different angle to become possible. For this reason, all the other than five patients could be managed only by means of laparoscope. These patients were confirmed by post-operative imaging.

Ectopic gallbladder missed on laparoscopic images might lead to an impression in the mind of surgeon to having some possibilities of future surgery. Despite these agenesis or absence of gallbladder, patients stand as negative result for exploratory laparotomy or laparoscopy. The patients by large remain symptom-free in their post-operative period. The reason of course is not known. The most possible reason in this regard is disappearance of adhesion in the hepatoduodenal ligament and right upper abdomen. Patients selected in this study remained asymptomatic. But long-term follow-up is to be advocated to know the recurrence and formation of CBD stone.

Due to advent of intraoperative laparoscopic ultrasound, agenesis can be diagnosed and managed by only laparoscopic mean, irrespective of complex and rarity of the condition, though laparotomy was the rule in the past in these circumstances that most of the authors now opined.^[10]

It is evident that biliary apparatus remains normal even when there is abnormal gallbladder like absent gall bladder as a part of developmental malformation. Bennion et al divided these patients into three groups: (i) Embryological malformation of foetus, (ii) Symptom free and (iii) With symptoms. It is about 13.1% and may rise to 35% if associated with biliary atresia, 30.6% and 56.7% respectively. Icterus, right hypochondriac pain etc. are the symptoms complained by symptomatic group. Pain is aggravated by fatty foods with flatulence and tendency of vomiting and occurrence of vomiting.

It is not ascertained which imaging procedure is the best to diagnose this defect of congenital origin. Due to failure to diagnose or misdiagnose, sometime adoption of surgical procedure is being unnecessary which causes enhanced morbidity. Study says that CT scan is the best to diagnose in pre-operative stage of those varieties of patients.^[11] But present study is having small sample to compare whether USG or MRCP are the best or effective.^[12] Malde algorithm is the specific way to say in this regard that has been reported by many series. MRCP, CT and ERCP are the investigation of acceptance in the respective manner if ultrasound fail to depict gallbladder which was the statement made by Malde.

CONCLUSION

Congenital defect of biliary tree and most commonly agenesis or absent gallbladder can easily be diagnosed in the per-

operative period. Agenesis is a very rare entity. More than half of the patients of congenital agenesis of gallbladder presented with symptoms that mimic gallstone diseases, yet gallbladder was not evident during per-operative assessment. Most of the patients of congenital agenesis of gallbladder remain symptom free in the post-operative period. After thorough assessment by laparoscopic means with a skill in hand and much experience in this field, these congenital conditions can be easily diagnosed by laparoscopic hands. Laparoscopy can reduce morbidity and mortality dramatically.

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