



## Bouveret's Syndrome: The rarest obstructing gallstone

Joshua Bramson<sup>a,\*</sup>, Arthur Topilow<sup>a</sup>, Ronald Matteotti<sup>b</sup>

<sup>a</sup> Department of Medicine, Jersey Shore University Medical Center, NJ, United States

<sup>b</sup> Department of Surgery, Jersey Shore University Medical Center, NJ, United States



### ABSTRACT

Bouveret's Syndrome is the rarest form of gallstone ileus. It is characterized by the passage of a large gallstone through a cholecystoduodenal fistula resulting in obstruction of the proximal bowel. The rarity, and non-specific presentation of Bouveret's Syndrome, make it a frequently overlooked diagnosis, with a significant associated mortality. We report the case of an 86 year old male presenting to the Emergency Department with nausea, vomiting, and constipation. Diagnosis of Bouveret's Syndrome was made by CT scan of the abdomen demonstrating a large stone in the proximal duodenum. After failed attempts at endoscopic removal of the gallstone, the patient was taken for surgical enterotomy and foreign body retrieval. Bouveret's syndrome is a rare entity with limited evidence to favor one approach to management over another. In this case, we review the diagnostic and therapeutic options for this rare form of small bowel obstruction.

### Introduction

Gallstone ileus complicates approximately 0.3–0.5% of all cases of cholelithiasis cases. Bouveret syndrome is an unusual cause of gallstone ileus, accounting for approximately 1–3% of all cases [1]. Bouveret's Syndrome generally presents in elderly patients (> 75 years old), with a female to male predominance (1.86:1) [2]. There is considerable morbidity and mortality associated with condition; with an estimated mortality of 12% in a 2009 review of literature [3]. This is due in large part to the relative rarity of this condition, and the ease at which the diagnosis may be overlooked, as well as the advanced age of the typical patient. Generally, endoscopic retrieval of the stone is recommended as the first line of therapy due to a relatively higher risk associated with surgical intervention (surgical mortality 19–24%) [3]. Nevertheless, surgical intervention is frequently necessary, as endoscopic intervention has a high rate of failure (approximately 77%) [4]. To date, there is no consensus recommendation regarding the preferred surgical approach in these cases.

### Case report

An 85 year old Caucasian male presented to the Emergency Department with complaints of right upper quadrant abdominal pain, nausea, vomiting, and constipation. The patient had a medical history significant for chronic kidney disease, coronary artery disease, atrial fibrillation, hypertension, type II diabetes mellitus, and benign prostate hypertrophy. A CT scan of the abdomen was performed without IV

contrast due to the patient's history of renal insufficiency. Imaging revealed a distended gall bladder with diffuse wall thickening and marked inflammatory changes. Additionally, there was noted to be an area of discontinuity of the gallbladder wall with an adjacent stone, suggestive of erosion of the gallstone into the proximal duodenum [Image 1]. Imaging also revealed a focal fluid collection (3.4 × 2.7 cm) along the medial aspect of the gallbladder, thought to represent a localized abscess. The patient was afebrile, and did not display signs of systemic infection or evidence of sepsis (Sequential Organ Failure Assessment score of 0 on admission). Initial management included placement of a cholecystostomy tube for drainage of the abscess and cultures of the fluid were sent for analysis. Laboratory data at the time was as follows: bilirubin 1.1 mg/dL, alkaline phosphatase 123 iU/L, AST 22 iU/L, ALT 16 iU/L, WBC 8300. Abscess cultures grew *E. coli* and *Candida Albicans* and patient received therapy with ampicillin/sulbactam and metronidazole, followed by ceftriaxone and fluconazole according to sensitivities.

Repeat CT scan showed migration of the large gallstone to the fourth portion of the duodenum [Image 2]. The patient was felt to have a high risk for obstruction. Taking into consideration the patient's age and comorbidities, a decision was made to attempt endoscopic removal of the large gall stone. During the procedure retained food products were observed in the stomach, and a gaping fistulous tract was seen between the gallbladder and duodenal lumen. A large gallstone was found impacted in the distal 3<sup>rd</sup> portion of the duodenum with inflammation and ulceration surrounding the gallstone. Multiple attempts were made to remove the gallstone, including use of a Roth net

\* Correspondence to: Department of Medicine, Jersey Shore University Medical Center, 1945 Route 33, Neptune, NJ 07753  
E-mail address: [joshua.bramson@hackensackmeridian.org](mailto:joshua.bramson@hackensackmeridian.org) (J. Bramson).

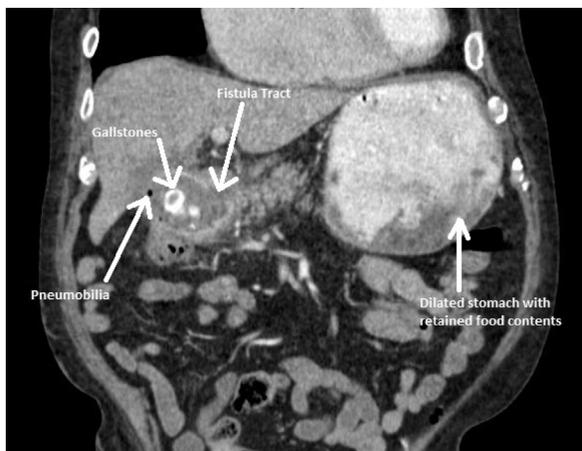


Image 1. Initial imaging revealed dilated stomach with large radiopaque gallstones.



Image 3. Gallstone (6.5 × 4.5 × 4.0 cm).

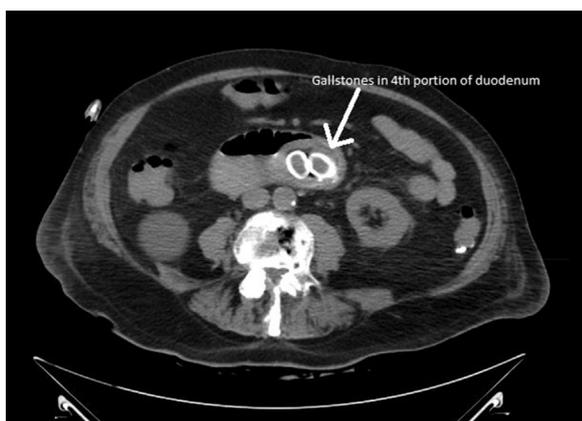


Image 2. Repeat CT demonstrating migration of gallstone to fourth portion of duodenum.

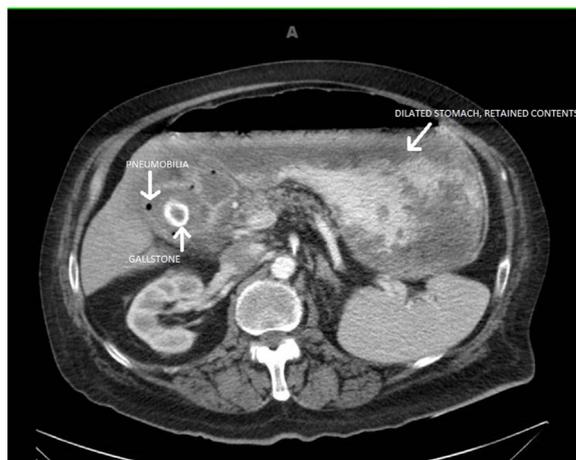


Image 4. Rigler's Triad of dilated stomach, pneumobilia, and a radiopaque shadow in the duodenum. In combination with demonstration of stone migration (Image 2) this represents Riglers tetrad.



Video 1. Surgical enterotomy with extraction of large obstructing gallstone. Supplementary material related to this article can be found online at <http://dx.doi.org/10.1016/j.nhccr.2017.08.004>.

retriever and large snare, as well as attempts to cut directly into the stone with a needle knife. Due to the rotation, and sheer size of the stone, these attempts were unsuccessful, and the decision was made to proceed to surgical removal of the stone [Video 1].

The patient underwent a diagnostic laparoscopy revealing an inflammatory mass in the area of the distal duodenum, followed by a partial open, subtotal cholecystectomy, and foreign body retrieval via enterotomy. After retrieval of the 6.5 × 4.5 × 4.0 cm ovoid gallstone [Image 3], a primary repair of the cholecystoduodenal fistula was performed with reinforcement of the repair via a falciform ligament vascular pedicle flap and omental patch. Portions of the gallbladder which were sent for pathological review showed evidence of chronic inflammation without malignant or pre-malignant transformation. The patient tolerated the procedure well, and has not experienced any significant complications of his surgery at 6 months post-operatively.

### Discussion

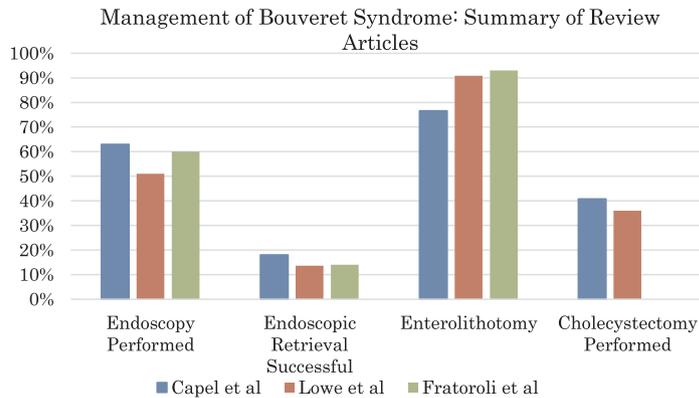
The first description of duodenal obstruction from gallstone passage through cholecystoenteric fistula was reported in 1841, observed in post-mortem autopsy [4]. In 1896, French physician Leon August Bouveret was the first to make the diagnosis pre-operatively [4]. Inflammation of the gallbladder leads to local adhesion between and inflamed gallbladder and the duodenum. In the presence of a large gallstone, there is local necrosis and deterioration of the bowel wall with passage of the stone into the enteric lumen. Larger stones have the potential to lead to mechanical bowel obstruction after passage in the small bowel. The duodenum is the least common location for this to occur. The majority of obstructing gallstones are found at the narrowing of the ileocecal junction where the ileocecal valve separates the small and large intestine. This accounts for 60–70% of cases of gallstone ileus [5]. Stones that bypass this area typically pass through the gastrointestinal tract uneventfully. However, very large stones, carry a higher risk of more proximal obstruction. Because of the relative rarity of the diagnosis and the often times non-specific symptoms, the diagnosis may be overlooked. However, with the wide availability of imaging, early diagnosis should be more likely.

The diagnosis can be made via abdominal plain film, ultrasound, and/or CT imaging. However, 15–25% of gallstones are isoattenuating and therefore not well visualized on CT scan [6]. In these cases, MRCP can be useful, but differentiation of solid structures from air can be a challenging limitation [5]. The classic presentation on imaging is Rigler's triad, which is the presence of a dilated stomach, pneumobilia, and a radiopaque

**Table 1**

Summary of review articles describing the management of Bouveret Syndrome (Al-Habbal Y et al, World journal of Gastroenterology. 2017 January 27; 9(1): 25–36.).

Reference	Year	Articles Reviewed	# Of Cases	Endoscopy Performed	Endoscopic Retrieval Successful	Enterolithotomy	Cholecystectomy Performed
Capel et al	2006	111	128	63%	18%	76.60%	40.80%
Lowe et al	2005	39	44	51%	13.60%	90.90%	36%
Fratoroli et al	1997	79	79	60%	14%	93%	Not Reported



shadow in the duodenum representing ectopic gallstones [7]. [Image 4] Serial imaging with demonstration of the stone migrating over time comprises Rigler’s tetrad [1] [Images 1, 2]. The gold standard of diagnosis is esophago-gastric duodenoscopy with direct visualization of the stone. This offers both diagnostic and potentially therapeutic utility. Multiple endoscopic techniques have been evaluated including lithotripsy, endoscopic retrieval basket, and snaring of stone. However, in up to 40% of cases, final diagnosis is not confirmed until surgery [7]. A January 2017 review of literature from 1990 to 2016 found that endoscopic therapy was tried in a total of 160 cases, and was successful in 46 cases [7]. This estimate of a 29% success rate is greater than estimates found in the majority of previous literature which suggested a > 90% failure rate [4]. This supports the role of endoscopic intervention as a primary intervention in cases of Bouveret’s syndrome. Still, the majority of patients are not amenable to endoscopic intervention and, as in our case, ultimately laparoscopy or laparotomy is required.

There remains considerable debate regarding the optimal surgical approach to these patients. The majority of cases involving some method of enterotomy with stone retrieval. The decision to proceed with cholecystectomy during the initial procedure is somewhat more controversial, with some sources recommending a staged intervention due to the relatively increased mortality observed in combined procedures. In a comprehensive review, cholecystectomy was performed in about 35% of cases [3, 7]. [Table 1] The benefit of performing cholecystectomy stems from the theoretical reduction in the risk of developing gall bladder cancer as a result of prolonged exposure to gastric secretions and subsequent chronic inflammation. To date, there is no consensus about the best surgical approach, and the majority of relevant data is gathered from case reports.

**Conclusion**

Bouveret’s Syndrome is a rare disorder presenting diagnostic and therapeutic challenges. While generally endoscopic intervention is supported by literature as the first step in intervention, it is frequently unsuccessful, and surgical intervention is necessary. The optimal surgical approach remains undefined. Further investigation is warranted to

determine the long term risk of gallbladder cancer in patients who have not undergone cholecystectomy, though the role of inflammation as the primary risk factor for development of gallbladder malignancy intuitively supports this as a reasonable preventative intervention. Whether the cholecystectomy should be performed as a primary intervention or as a subsequent surgical procedure is less clear. We feel that several factors, including the skill and experience of the surgeon, the patient’s comorbid conditions, and the degree of local inflammation are significant factors in this decision. In this case the surgeon chose to perform an immediate subtotal cholecystectomy. Due to the low incidence of the condition, early suspicion and diagnosis is the optimal treatment approach.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Competing interest declaration**

We have no conflicts of interest to declare.

**References**

- [1] R.Q. Ghazi, Sohail Bakkar, Khaled Jadallah, Bouveret’s Syndrome: an overlooked diagnosis, *J. Int. Surg.* 99 (2014) 819–823.
- [2] S. Mitchell, M.D. Cappell PhD, FACP and Michael Davis DO, *Am. J. Gastroenterol.* 101 (2006) 2139–2146, <http://dx.doi.org/10.1111/j.1572-0241.2006.00645.x>.
- [3] A.S. Lowe, S. Stephenson, J. Kay, L. Kay, Duodenal obstruction by gallstones, *Endoscopy.* 37 (1) (2005) 82–87, <http://dx.doi.org/10.1055/s-2004-826100>.
- [4] Y. Al-Habbal, M. Ng, D. Bird, T. McQuillan, H. Al-Khaffaf, Uncommon presentation of a common disease – Bouveret’s Syndrome: a case report and systematic review of literature, *World J. Gastroenterol.* 9 (1) (2017) 25–36.
- [5] R.E. Lawther, T. Diamond, Bouveret’s Syndrome: gallstone ileus causing gastric outlet obstruction, *Ulst. Med. J.* 69 (1) (2000) 69–70.
- [6] D. Tuney, C. Cimsit, Bouveret’s Syndrome: ct findings, *Eur. Radiol.* 10 (11) (2000) 1711–1712.
- [7] Felix Nickel, Matthias M. Müller-Eschner, Jackson Chu, Hendrik von Tengg-Kobligk, Beat P. Müller-Stich, Bouveret’s Syndrome: presentation of two cases with review of the literature and development of a surgical treatment strategy, *BMC Surg.* (2013), <http://dx.doi.org/10.1186/1471-2482-13-33>.