Synchronous splenectomy during cholecystectomy for hereditary spherocytosis: Is it really necessary?

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A B S T R A C T

Background/Purpose: Expert guidelines recommend performing synchronous splenectomy in patients with mild hereditary spherocytosis (HS) and symptoms of gallstone disease. This recommendation has not been widely explored in the literature. The aim of this study is to determine if our data support expert opinion and if different practice patterns should exist.

Methods: This is an IRB-approved retrospective study. All HS patients under 18 years of age who underwent cholecystectomy for symptomatic gallstones at a single institution between 1981 and 2009 were identified. Patients who underwent cholecystectomy without concurrent splenectomy were reviewed retrospectively for future need for splenectomy and evidence of recurrent gallstone disease.

Results: Of the 32 patients identified, 27 underwent synchronous splenectomy. The remaining 5 patients underwent cholecystectomy without splenectomy and had a mean age of 9.4 years. One of the 5 patients eventually required splenectomy for left upper quadrant pain. None of the remaining 4 required hospitalization for symptoms related to hemolysis or hepatobiliary disease. Median follow-up is 15.6 years.

Conclusion: The need for splenectomy in patients with mild HS and symptomatic cholelithiasis should be assessed on a case by case basis. Our recommendation is to not perform synchronous splenectomy in conjunction with cholecystectomy for these patients if no indication for splenectomy exists.

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Hereditary spherocytosis (HS) was first described in 1871 and is the most common form of congenital hemolysis among Caucasian and Hispanic populations with an incidence in the general population of approximately 1:5000 [1]. It follows an autosomal dominant mode of inheritance and is caused by defects of various erythrocyte membrane proteins including spectrin and ankyrin. The resulting impaired membrane integrity leads to both abnormal morphology and decreased red cell lifespan due to hemolysis [1].

Based on the extent of their disease, patients with HS are grouped into 3 categories: mild, moderate and severe. Clinical manifestations vary and depend upon the degree of hemolysis. Patients with mild disease are defined as having a hemoglobin level ranging from 11 to 15 g/dL, a reticulocyte count between 3% and 6% and a bilirubin level of 1–2 mg/dL [1]. These patients can be asymptomatic or experience only occasional jaundice. Moderate disease is characterized by symptomatic anemia which in nearly all cases will resolve following splenectomy. Patients with the severe form can experience profound anemia requiring frequent blood transfusions and typically remain anemic even after splenectomy. For patients with mild disease, splenectomy is generally not required. However, splenic sequestration and excessive hemolysis can lead to cholelithiasis in these children. Previous surgical management guidelines have been somewhat unclear, stating that for patients with mild disease, splenectomy “should probably not be performed” while also recommending that the spleen “always be removed” for patients requiring cholecystectomy for symptomatic cholelithiasis [1]. The reasoning behind the latter recommendation, we believe, is sound: there is a theoretical risk of intrahepatic choledocholithiasis after removing the gallbladder while leaving behind the spleen in patients with HS. The recommendation is based on expert opinion (level III evidence [2]) and there are no data in the literature to support this opinion. One model also suggests concurrent splenectomy in these patients, hypothesizing a 3-fold increased risk of gallstone formation as compared to the general population [3]. However, given the risks of splenectomy in children, and the advent of laparoscopic surgery, some have begun to question this practice, finding that many patients do not go on to require splenectomy [4]. In order to better characterize the risk of subsequent splenectomy or intrahepatic cholelithiasis, we report long term follow up of our experience with HS patients undergoing cholecystectomy without synchronous splenectomy.

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1. Patients and methods

This is an IRB-approved retrospective study of patients less than 18 years old who underwent cholecystectomy for symptomatic gallstones in the setting of HS at a single academic institution between January 1981 and December 2009. Synchronous splenectomy was defined as removing the spleen during a cholecystectomy, when no independent indication for a splenectomy existed. Of 32 patients identified with HS who underwent a cholecystectomy, 27 underwent synchronous splenectomy. The remaining 5 patients who underwent cholecystectomy without splenectomy were reviewed retrospectively for future need of splenectomy and evidence of intrahepatic and common bile duct stones or any hepatobiliary disease. Charts were reviewed for severity of HS, future splenectomy, peri-operative complications and subsequent hospitalizations. These 5 patients were also interviewed via telephone and asked a series of standardized questions to assess for potential complications of continued hemolysis and splenic sequestration. Our goal was to evaluate the eventual need for splenectomy as well as any evidence of choledocholithiasis and its complications.

2. Results

Of the 5 patients identified, all had mild HS symptoms. Their details are summarized in Table 1. In all cases, patients presented with only symptomatic cholelithiasis. Splenectomy was deferred due to absence of symptomatic anemia. None of the patients encountered post-operative complications. Patients were discharged from the hospital in an average of 1 day following their surgery. One patient underwent laparoscopic splenectomy at 10 years post-op for left upper quadrant pain secondary to splenomegaly. His post-splenectomy hospital course was uncomplicated and symptoms resolved post-operatively. None of the 5 patients remarked upon any signs or symptoms consistent with bile duct stones since their cholecystectomy and none had any complications from choledocholithiasis. Median follow-up was 15.6 years, range 11 to 19 years.

3. Discussion

Currently, there is no firm consensus surrounding the best surgical management for patients with HS who need a cholecystectomy, but have no independent indication for splenectomy [4]. In fact, recommendations overall favor performing a splenectomy in these situations. In the case of HS patients with mild disease and symptoms of gallstones, consideration must first be given to the risks and benefits most likely to be encountered by the patient. The most recent recommendations appropriately state that concurrent splenectomy is controversial while recognizing that splenectomy may also be associated with a decreased risk of common bile duct stones [5]. However, the absence of even a single case-report in the literature, including our series, would suggest that stone recurrence following cholecystectomy is exceedingly unlikely. The only other series to report on such patients, Alizai et al., describes 16 mild HS patients who underwent cholecystectomy without concurrent splenectomy with no evidence of stone recurrence at a median follow-up of 5 years.

Other reasons for concurrent splenectomy have included potential difficulty of reoperation, should the patient require splenectomy in the future. Prior to the development of minimally invasive techniques, open operations were performed which required larger incisions, necessitated longer hospital stays and were associated with increased morbidity versus laparoscopic procedures [6]. Open operations also elicit a greater intra-abdominal inflammatory response with adhesions that may complicate potential future operations. However, in a laparoscopic era where post-operative adhesions from a cholecystectomy are unlikely to prevent or complicate a future laparoscopic splenectomy, a more selective approach to splenectomy in these patients should be considered. Our review revealed that all mild HS patients who underwent cholecystectomy with concomitant splenectomy did so in an open fashion. This would suggest that the ability to perform laparoscopic procedures may have played a role in the decision not to perform synchronous splenectomy. Additionally, laparoscopic splenectomy has been associated with shorter hospital stays and significantly decreased post-operative pain when compared to open splenectomy [7]. Perhaps the benefits of minimally invasive surgery allow for staged procedures based on current indications for operation.

Post-splenectomy sepsis represents a small but real risk to this patient population. A review from 2001 that spanned 30 years and nearly 7000 post-splenectomy patients estimated an incidence of infection of 3.1% with a mortality of 1.3% in patients with a history of HS [8]. Other studies have reported infection and mortality rates as high as 6% and 3.3% respectively [9]. Perhaps avoiding splenectomy in this patient population would lessen the risk of post-splenectomy sepsis. Additionally, there is evidence to suggest an increased risk of pulmonary hypertension following splenectomy. Although this is more common in patients with a history of thalassemia, it has been reported in patients with HS [10].

In patients who do not undergo concurrent splenectomy, future need for splenectomy certainly exists. In the 2010 review by Alizai et al., 3 of the 16 mild HS patients eventually required splenectomy for hematological reasons. Just as with our patient, the splenectomies were uncomplicated and were all performed laparoscopically with hospital durations ranging from 1 to 3 days.

Our data in addition to Alizai’s demonstrate that patients with mild HS and symptomatic cholelithiasis and no other hematologic indication for splenectomy did not need a splenectomy in the future. Furthermore, the likelihood of developing bile duct stones is extraordinarily low, as none of the patients in either study developed symptoms related to bile duct stones after cholecystectomy alone. This case series is limited by its small numbers. Future efforts should surround the cultivation of additional long-term follow-up data surrounding this subset of patients in hopes of further characterizing the risk of future splenectomy and stone recurrence.

4. Conclusion

Given the long-term follow-up of 15.6 years demonstrating no signs or symptoms consistent with ductal or hepatic stone recurrence, our recommendation is to reconsider the prevalent expert opinion

### Table 1

<table>
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<tr>
<th>Case #</th>
<th>HS severity</th>
<th>Laparoscopic (Lap) vs. open</th>
<th>Indication</th>
<th>M/F</th>
<th>Age</th>
<th>Splenectomy</th>
<th>Stone recurrence</th>
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that patients with mild HS undergoing cholecystectomy should have synchronous splenectomy. Unlike stone recurrence, both future need for splenectomy and post-splenectomy sepsis represent real and documented risks in these patients. Furthermore, in the setting of increasingly refined minimally invasive techniques, we do not believe that splenectomy should be performed in conjunction with cholecystectomy in patients with mild HS, when no independent indication for splenectomy exists.

References


