Index Case: Teenage Girl with Abdominal Cyst

You are consulted by the emergency department to evaluate an unexpected finding on the abdominal ultrasound of a previously healthy 14-year-old girl who presented with a 2-day history of lower abdominal pain. The patient’s abdominal exam had revealed low right lower quadrant tenderness to palpation without rebound. She was afebrile and her vital signs were within normal limits. A complete blood count, urine pregnancy test, and urinalysis were normal.

Ultrasound demonstrated a normal appendix and a 3-cm hemorrhagic right ovarian cyst, which was felt to account for her symptoms. However, it also showed a large cystic mass in the left upper quadrant measuring approximately 9 cm in greatest dimension, as shown in Figure 44.1.

Q1: What are the potential etiologies of this cystic mass?

This is an incidentally discovered left upper quadrant cystic mass, unlikely related to the patient’s acute symptoms. Cystic masses in the left upper abdomen in this age group may arise from the stomach (duplication, gastric teratoma), the left lobe of the liver (simple hepatic cyst, hydatid cyst, liver abscess), the spleen (epidermoid cyst, posttraumatic pseudocyst, hydatid cyst, splenic abscess), the tail of the pancreas (cystic neoplasm, pseudocyst), or the diaphragm (epidermoid cyst, bronchopulmonary foregut malformation). Lymphatic malformations may also be seen and can arise from any of these structures.

The radiologist has the impression that the mass arises from the spleen but is unable to confirm this finding.

Q2: Would you proceed with additional investigations?

A chest x-ray would help rule out a thoracic location of the mass. Cross-sectional imaging of the abdomen will likely reveal the source of the cystic mass and further characterize it. An alpha-fetoprotein level should be checked as a potential marker for a malignant cystic teratoma.

The chest x-ray and alpha-fetoprotein levels are normal. CT scan images are shown in Figure 44.2. A single cyst, measuring $9.9 \times 9.8 \times 9.2$ cm, is found in the upper pole of the spleen with a claw sign in the splenic parenchyma. A pressure effect on the stomach is seen.
On obtaining further history from the patient, she reports that she has experienced early satiety and lost 3 kg over the past 6 months. She also experiences episodes of left upper quadrant pain approximately once a week.

Q3: Should this incidentally discovered cyst be treated?
Yes. Splenic cysts smaller than 5 cm may be observed, but larger cysts should be treated. As in this case, many patients with large cysts will have subtle symptoms such as intermittent pain and early satiety. Large cysts are also at risk of hemorrhage, infection, and rupture.

Q4: What are the treatment options, and what will you recommend?
A variety of treatments have been described for splenic cysts. These include percutaneous injection and sclerosis, marsupialization, cyst excision, and open or laparoscopic partial splenectomy. The most definitive procedure associated with the lowest chance of cyst persistence or recurrence is partial splenectomy with a small rim of adjacent normal spleen. Fortunately, these cysts typically arise from the upper or lower poles. A partial splenectomy, even when performed for very large cysts, will typically result in preservation of most of the spleen. The procedure yields itself nicely to a laparoscopic approach, which minimizes the morbidity of an open operation.

Q5: How will you prepare the patient for the surgical procedure?
Although a partial splenectomy is planned, the patient should be prepared for the possibility of a total splenectomy. This involves vaccination against the three bacterial species that are responsible for most cases of postsplenectomy sepsis, namely pneumococcus, meningococcus, and Haemophilus species. Current recommendations include polysaccharide pneumococcal, conjugate Haemophilus influenzae type b, and polysaccharide meningococcal vaccinations. Vaccinations should be completed at least 2 weeks before the operation. A type and cross-match for blood should also be sent, in case transfusion is required for a hemorrhagic complication.

The findings on laparoscopy are shown in Figure 44.3. A laparoscopic partial splenectomy is completed and shown in Video 44.1.
Q6: The pathologist characterizes the lesion as an epidermoid cyst. What are the histologic features that result in this diagnosis?

The typical gross and histologic features of an epidermoid cyst of the spleen are evident in this case, as shown in Figure 44.4: The inner cystic surface shows coarse trabeculations and septations (Figure 44.4a). On low power, the cystic wall contains variable fibrous tissue (Figure 44.4b). The cyst is lined by stratified squamous epithelium (Figure 44.4c). Keratinization may be present but is not seen in this case. Residual white pulp (arrowhead) and red pulp (arrow) of the spleen are illustrated (Figure 44.4d).
Q7: What is the embryologic explanation of such a cyst?

The spleen arises from the foregut. Its primordium originates as a mesenchymal bulge in the dorsal mesogastrium between the stomach and pancreas, first observed at the 10 mm embryo stage. A true epithelium is found soon afterward, as sinusoids communicate with capillaries. Epithelial, also referred to as epidermoid, cysts of the spleen are thought to arise when surface mesothelium becomes incorporated into the splenic parenchyma.

The patient has an uneventful recovery and is discharged on the third postoperative day. Twelve months after surgery, she is found to have mild anemia (hemoglobin 107 g/liter) and mild thrombocytosis with a platelet count of 460,000/µL on a complete blood count performed by her pediatrician to assess several episodes of heavy menstrual bleeding. He suggests an assessment of splenic function to ensure that the residual splenic remnant has survived.

Q8: How can this be done?

A residual spleen after partial splenectomy can be assessed by Doppler ultrasound and a peripheral blood smear. The ultrasound will assess the splenic remnant for possible atrophy and document the arterial and venous blood flow. In the absence of splenic filtering function, the blood smear will show several findings not seen with normally functioning spleens. These include Howell–Jolly bodies (red blood cell nuclear remnants), Pappenheimer bodies (iron granules), and Heinz bodies (denatured hemoglobin). A liver-spleen nuclear scan can also be performed to detect splenic function, but is rarely necessary.

Ultrasound, shown in Figure 44.5, shows a normal-appearing spleen, 9.2 cm in longest dimension, with good arterial and venous blood flow and no cysts. The peripheral smear is normal. The patient’s appetite has returned to normal and she no longer has episodes of upper abdominal pain.

![Figure 44.5 Follow-up ultrasound](image)

Discussion

The spleen has three major functions: red blood cell maintenance, immunologic activity, and reservoir function. Red blood cells are removed by the spleen at the end of their life span. In the absence of a spleen, red blood cell remnants such as Howell–Jolly bodies (nuclear remnants), Heinz bodies (denatured hemoglobin), and Pappenheimer bodies (iron granules) can be found on a peripheral smear and are evidence of an asplenic state. The spleen functions immunologically in multiple capacities. It serves as a biologic filter for bacteria and particulate matter in the blood.
It is a source of opsonins, antibodies that bind to foreign microorganisms or cells, making them more susceptible to phagocytosis. Certain encapsulated bacteria, most notably *Streptococcus pneumoniae*, *Neisseria meningitides*, and *Haemophilus influenzae*, require opsonization for optimal clearance. The risk of infection by these bacteria, which are all major childhood pathogens, increases after splenectomy. The spleen also exerts a specific immune function through antigen processing in the white pulp, which is rich in macrophages and helper T cells. Finally, the spleen serves as a reservoir for platelets and factor VIII, which explains both thrombocytopenia in cases of splenomegaly and thrombocytosis following splenectomy.

There are essentially only two major indications for elective splenectomy in children: anatomic abnormalities, and hematologic diseases. Each condition within these two large categories requires the surgeon to carefully assess the patient’s condition and the severity of the disease. The decision on whether to perform splenectomy, the appropriateness of a total versus a partial splenectomy, and the choice of a laparoscopic versus an open technique then ensue.

The most common anatomic abnormality requiring treatment is an epidermoid splenic cyst, as presented in the index case. As with many similar lesions, the incidence of these cysts has risen sharply in the era of ultrasound imaging. Many cysts are found incidentally during ultrasound performed to assess another condition or abdominal pain. Cysts smaller than 5 cm are safe to observe. Most surgeons recommend treatment of larger cysts. Multiple treatment options are available as discussed in the index case, but several recent series, including a large, French, multicenter study, have established laparoscopic partial splenectomy as the procedure of choice. The procedure is associated with minimal morbidity when performed by experienced surgeons and virtually eliminates the risk of cyst recurrence. Sclerotherapy of splenic cysts is characterized by the need for multiple treatments and a high incidence of residual cysts. Epidermoid cysts are typically single, although they may contain septations and calcifications due to previous hemorrhage. Multiple simple cysts should raise the likelihood of an alternate diagnosis. Figure 44.6 shows the CT scan of a 12-year-old boy with such a finding. A presumptive diagnosis of lymphatic malformation of the spleen was made. The child was followed annually with ultrasound with no change in the cystic masses over a 6-year period. Splenic cysts, as well as renal cysts, can also be seen in the setting of pleuropulmonary blastoma.

![Figure 44.6 Splenic lymphatic malformation: CT scan](image)

Abscesses, hydatid cysts, and posttraumatic pseudocysts can also be found in the spleen. Although rare, mycobacterial and fungal abscesses have been reported in the spleens of immunocompromised children on chemotherapy. Splenectomy is performed when there is lack of clinical improvement or response to antibiotic therapy. Echinococcal cysts of the spleen are rare, even in endemic regions. The PAIR (puncture, aspiration, injection, re-aspiration) technique is highly successful in treating the disease and preserving the spleen. Splenectomy is reserved for hydatid cysts complicated by abscess or hemorrhage. Pseudocysts of the spleen are a rare complication of non-operative management of blunt splenic trauma, estimated to occur in less than 1% of all cases. Routine delayed imaging of the spleen after non-operative management is therefore not recommended. Spontaneous resolution of large pseudocysts is rare. Laparoscopic excision or marsupialization is the preferred treatment.
Wandering spleen is a rare anatomic abnormality that requires surgical intervention and sometimes splenectomy. In this condition, the spleen lacks the normal peritoneal ligamentous attachments to the stomach, colon, and retroperitoneum. It is therefore not fixed in the left upper quadrant and can travel through the abdominal cavity. Intermittent abdominal pain with a palpable discrete mass is the classic presentation. Torsion and infarction of the spleen can also occur, necessitating splenectomy. A viable spleen can be surgically immobilized in the left upper quadrant by a variety of techniques, including placement of the spleen in an extraperitoneal pocket, or splenopexy within a mesh basket. A rare manifestation of wandering spleen is chronic torsion of the spleen without infarction, resulting in presinusoidal portal hypertension. This in turn manifests as splenomegaly and hypersplenism, with mesenteric varices. I encountered such a case in a 17-year-old girl, in whom the diagnosis was made during a work-up for thrombocytopenia. She had complained of severe bouts of intermittent abdominal pain. Figure 44.7 shows the MRI appearance of the spleen and mesenteric varices, as well as the massively enlarged spleen with engorged hilar vessels on laparoscopy. The procedure was converted to open splenectomy due to bleeding from the hilar vessels. Splenectomy was curative. The thrombocytopenia resolved immediately after surgery, and the mesenteric varices resolved on MRI performed 6 months after operation. Preoperative embolization to decrease splenic vascularity should be considered in such cases.

Another rare anatomic abnormality is splenogonadal fusion, a condition in which the left gonad and the spleen are attached as a result of early fusion prior to testicular descent. In this condition, splenic tissue may be attached to the testicle and can be confused with a neoplasm. An accessory spleen may also be found along the course of testicular descent, separate from the testicle. Figure 44.8 shows a 1.5-cm accessory spleen lateral to the bladder at the left internal inguinal ring. The mass was detected on ultrasound performed to investigate macrohematuria in an 11-year-old boy. MRI demonstrated the mass clearly, but did not reveal its etiology. A liver-spleen nuclear medicine scan confirmed an accessory spleen, and the patient avoided surgical intervention.

Hereditary spherocytosis is the most common hematologic disease requiring splenectomy in children. The disease is inherited, in an autosomal dominant pattern, in only 75% of patients. The remainder presents with sporadic mutations. The defect is most commonly in spectrin, a red blood cell membrane protein. The oxygen-carrying capacity of the cell is not affected. However, it is rendered spherical and poorly deformable, which results in splenic trapping and hemolysis. An increase in unconjugated bilirubin predisposes the patient to pigment gallstones. The disease manifests as a wide spectrum from mild to severe based on hemoglobin levels and requirement for transfusion, reticulocyte count, and bilirubin levels.
There are several controversies regarding the role of splenectomy in spherocytosis. Unfortunately, no widely agreed-upon guidelines exist as to which patients are candidates for splenectomy. The pattern of splenectomy is therefore heavily influenced by the philosophy of the treating hematology service, with some preferring relatively early splenectomy and others preferring to delay it as long as possible. There is agreement that patients with “severe” spherocytosis should undergo splenectomy. However, significant disagreement exists regarding the definition of “severe.” British guidelines consider severe spherocytosis to exist when the hemoglobin level is 60–80 g/L, the reticulocyte count is more than 10%, and the bilirubin level is more than 51 µmol/L. However, these three parameters do not always coexist. For example, it has been shown that many patients classified as “mild” based on hemoglobin levels have reticulocyte counts well above 10% and may see improvement in quality of life after splenectomy.

One of the arguments for delaying splenectomy, as well as for performing a partial splenectomy, is the risk of overwhelming postsplenectomy infection (OPSI). Estimates, calculated before vaccinations against encapsulated bacteria were routine, put the risk of this potentially lethal complication well below 10% over a lifetime. The actual risk at present is most likely in the 1%–2% range, given routine vaccinations with the polyvalent Streptococcus pneumoniae vaccine, in addition to the meningococcus and Haemophilus vaccines. A recent national series from Finland with excellent long-term follow-up demonstrated that OPSI was only seen in patients with an underlying immunodeficiency. Another concern regarding postsplenectomy portal vein thrombosis in children did not bear out in the same report. There have been a number of published retrospective cases series supporting the role of partial splenectomy in hereditary spherocytosis. The technique removes 85%–95% of the enlarged spleen, retaining a portion equivalent to 25%–30% of the volume of a normal spleen (typically the upper pole surviving on the short gastrics), the minimal amount thought to preserve splenic immune function. However, it is still not clear if there are any advantages to this approach. A recent North American multicenter registry report showed no discrete advantages to partial splenectomy. The hematologic response was more robust after total splenectomy. Reports have also emerged of recurrent anemia in patients with hereditary spherocytosis due to splenic regeneration or growth of accessory spleens left behind. Some patients have required completion splenectomy to control the disease. Finally, laparoscopic partial splenectomy is associated with longer operative time, higher blood loss, higher chance of converting to open surgery, and a longer length of stay than laparoscopic total splenectomy. Any candidate for a splenectomy for hereditary spherocytosis should undergo ultrasound of the biliary tract to screen for cholelithiasis. A concomitant cholecystectomy during splenectomy is recommended if stones are identified, even in asymptomatic patients, since there is no evidence that pigment gallstones resolve
after splenectomy. In cases of symptomatic cholelithiasis, a cholecystectomy should be performed, but there is no
consensus regarding the need for a concomitant splenectomy in this setting.

To balance these competing concepts, my approach to patients with spherocytosis is as follows. I work very closely
with our hematology colleagues and engage the patient and parents in shared decision-making. I do not perform
splenectomy before the age of 6 years, unless there are absolute indications, to further decrease the risk of OPSI.
Practically, the majority of our patients are teenagers who choose splenectomy due to severe splenomegaly and
chronic left upper quadrant discomfort. I prefer a laparoscopic total splenectomy, but I review the pros and cons of
a partial versus total splenectomy with the patient and parents, and come to a joint decision. A biliary ultrasound is
performed. If cholelithiasis is found, a concomitant laparoscopic cholecystectomy is performed. Video 44.2 shows
a laparoscopic total splenectomy and cholecystectomy in a 17-year-old boy with moderate disease, splenomegaly,
and cholelithiasis.

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Video 44.2 (https://youtu.be/zuC5LwbBino)
Laparoscopic splenectomy and cholecystectomy for spherocytosis.

Splenectomy is much less frequent in patients with sickle cell disease. An autosplenectomy may have occurred
in many patients due to repeated splenic infarctions. The indication for a splenectomy is a splenic sequestration
crisis with severe anemia and potential hemodynamic instability. This complication of sickle cell disease is
associated with a significant risk of recurrence and high mortality. There is again some controversy regarding
whether splenectomy should be performed after the first sequestration crisis episode, or only if additional episodes
occur. However, since the function of the spleen is unlikely to be preserved in patients with sickle cell disease,
laparoscopic splenectomy after a single sequestration crisis is supported by most surgeons and hematologists.

Immune thrombocytopenic purpura (ITP) can present another indication for splenectomy in children. The
majority of children present with acute ITP following a viral illness or other trigger and improve spontaneously.
Chronic ITP is diagnosed when the thrombocytopenia persists for more than 6 months. In these cases, the
spleen exerts a dual pathologic role. The white pulp is the source of autoantibodies to the platelet membrane
glycoproteins, and the red pulp is the site of phagocytosis of autoantibody-coated platelets. First- and second-line
treatments for chronic ITP are medical, starting with steroids and immunoglobulins, and progressing to rituximab,
chemotherapeutic agents, and possibly romiplostim, a newer thrombopoietic agent. Surgery is reserved for patients
who do not respond to medical interventions or who recur after cessation of medical treatment. Traditionally,
patients who have shown a medical response were considered the most likely to respond to splenectomy. This has
recently been challenged, and splenectomy should essentially be offered to all patients with ITP who fail medical
management.

The surgical issues with ITP are quite different from those associated with spherocytosis. ITP spleens are of
normal size. The challenges are related to the coagulopathy and the need to remove all splenic tissue. Packed red
blood cells should be made available, and the patient may need to receive platelets at the induction of anesthesia.
An attempt should be made to control the splenic hilum early in the procedure if possible, to allow for elevation
of the platelet count as the rest of the procedure continues. A careful search should be made for accessory spleens.
These occur in approximately 20%–30% of all patients. Fortunately, about 75% will be found near the splenic
hilum as shown in Figure 44.9, taken during a laparoscopic splenectomy for ITP. However, a careful search for
accessory spleens should also ensue in the omentum, the lesser sac, the tail of the pancreas, and the retroperitoneal
space around the spleen. Recurrence of ITP after initial response to splenectomy should raise the suspicion for
missed accessory spleens, which may have enlarged after the splenectomy. I have encountered this situation in
two patients. Some publications have reported a higher incidence of missed accessory spleens after laparoscopy.
However, more recent experience has not supported this finding. Laparoscopic splenectomy is therefore an
excellent choice for the treatment of ITP and has been shown in some reports to be associated with less blood loss
than the open technique.
There is wide practice variation in the prescription of prophylactic antibiotics after splenectomy. Children younger than 5 years should receive antibiotics. Older patients are often prescribed antibiotics for 2 years or until they reach 18 years of age. Evidence for these approaches is weak. However, it is extremely important to educate the patient and parents regarding the symptoms and signs of OPSI and ask them to seek medical attention emergently for any febrile illness. Thrombocytosis after splenectomy is also common and can transiently exceed 1,000,000 platelets/µL. This is a benign condition, and aspirin prophylaxis is not indicated.

Finally, the spleen is rarely the site of solid tumors, either primary or metastatic. Some of these may be detected incidentally on imaging for other indications. The two most common primary malignancies of the spleen are lymphoma and angiosarcoma. Primary involvement of the spleen by lymphoma is quite rare, accounting for less than 1% of all lymphomas. Splenectomy is no longer indicated in lymphoma staging. Metastases to the spleen are also very rare and almost always accompany metastases to other organs. Primary sites include breast, lung, colon, ovary, and stomach.

Figure 44.10 shows a splenic Hodgkin lymphoma identified in a 17-year-old boy who received an ultrasound during work-up for acute abdominal pain. A 2.5-cm solid splenic mass was found. On physical examination, he was found to have enlarged left supraclavicular lymph nodes. Staging CT of the neck, chest, and abdomen showed extensive cervical and paratracheal adenopathy, as well as the splenic lesion demonstrated in Figure 44.10. A diagnosis of stage III Hodgkin lymphoma was made after biopsy of the cervical lymph node. The patient was successfully treated with chemotherapy and a splenectomy was not necessary.
Suggested Readings


