In and so they are usually not

In 1856, Mickel described the first congenital solitary nonparasitic hepatic cyst in anadult.(22, 35) Between 1856 and 1995, more than 400 caseshave been reported in the pediatric literature, among which only 5% involveinfants less than 2 years of age.

(22, 35) True nonparasitic congenital hepatic cystshave a reported overall prevalence ranging between 0. 1% and 2. 5%,(18, 36) although noninvasive imaging has shown thatasymptomatic hepatic cysts are likely more common in the general populationthan originally believed.(35, 37) Calculation of the precise frequency ofcongenital hepatic cysts in the pediatric population is difficult, because thelesions rarely produce symptoms and so they are usually not detected untiladulthood after they become symptomatic. Furthermore, only 10%–40% ofcongenital cysts are estimated to become symptomatic.

(35, 38) Quiescent lesions are usually discovered as anincidental radiographic, intraoperative, or autopsy finding. In a retrospectivereview at Boston Children’s Hospital, Boston, MA, USA, in which 30 cases ofcongenital nonparasitic hepatic cysts were reviewed in children less than 8years of age, only 40% of children were symptomatic from abdominal distensionwhereas 60% of lesions were incidental findings at autopsy.(18, 35) When detected(35) in adulthood, hepatic cysts are mainly foundbetween 20 and 50 years of age with a median age at presentation of 48 years.(39, 40) The female-to-male ratio is 5: 1.(18, 35)ClassificationThe termhepatic cyst commonly refers to solitary nonparasitic cysts of the liver, whichknown as simple cysts.

(1, 2)However, many other cystic lesions might be distinguished from true simplecysts. Cystic lesions of the liver include Simple cysts, multiple cysts arisingin the setting of polycystic liver disease (PCLD), parasitic or hydatid(echinococcal) cysts, cystic tumors and abscesses. Nonparasitic liver cysts areclassified as solitary cyst or multiple cysts. Although multiple cysts may beassociated with congenital polycystic disease of the kidney or dilatation ofthe bile ducts (Caroli’s disease), SLCs never accompany other organ cysts.(3, 4, 32) Solitaryliver cyst is a benign liver lesion.

It is seen four times more common infemales.(3, 4, 32, 41) Further, it can be classified as simple solitary liver cyst (SLC) and solitaryintrahepatic biliary cyst (SIBC).(4, 32, 42) Theetiology of SLC is not known. SLC has been hypothesized that obstruction ofaberrant bile ducts from infection or as a result of a vascular disruptionduring the fetal period can be a cause.

(32, 41-44)Alternatively, overdeveloped intralobular bile ducts may become dilated withtime and result in an Solitary liver cyst.(32, 45) However, none of these etiologies can explainthe Solitary intrahepatic biliary cyst in our patient. Although, the cyst wasconfirmed to be a true biliary cyst located close to the main intrahepatic bileducts. It has small canalicular connections with the main intrahepatic bileducts. Simple cysts reported for most of the prenatally detected and postnatallydiagnosed solitary lesions.

They may be underrepresented in this series becausesome asymptomatic simple cysts would not have been referred to our unit. Fromaberrant bile ducts, simple cysts are generally thought to be developed,(23-25) but anorigin from intrahepatic peribiliary glands has also been suggested.(26,

27)Hepaticcysts may be classified broadly into two major categories – congenital oracquired.

(35, 46, 47) whichcan also be categorized on the basis of history, symptoms, and radiographicappearance. Congenital liver cysts arise from abnormal development of theintra- or extrahepatic biliary tree. They are solitary simple or multiplepolycystic.(35, 48) Unlikepolycystic liver disease, simple hepatic cysts do not have a genetic basis, andthere is no association with renal, splenic, pancreatic, or lung cysts.(18, 35) Acquired liver cysts are parasitic(infectious) or nonparasitic (post-traumatic and neoplastic).

(35, 46) Parasitic cysts are usually hydatid cysts whichresults from infection with Echinococcus granulosus. Most acquired nonparasitichepatic cysts are post-traumatic pseudocysts.(35, 46) as shown in table 4. Table4. Solitary liver cysts in children Congenital Acquired Simple Parasitic (hydatid) Mesenchymal hamartoma Posttraumatic Intrahepatic choledochal cyst Neoplastic: cystadenoma, sarcoma, teratoma Ciliated hepatic foregut cyst Biliary cysts (biliary atresia post Kasai, traumatic) Epidermoid cyst Abscess (eg, pyogenic, amoebic) Lymphangioma Smallasymptomatic liver cysts are considered benign, but they should be followed forgrowth and the possibility of malignant degeneration.

(35, 37)PathophysiologyCongenitalliver cysts arise from the biliary tree and more specifically, they arebelieved to originate from enlargement of congenital aberrant biliary ducts. Congenitalliver cysts are usually lined with biliary columnar epithelium, but there maybe present of squamous or cuboidal cells.(35, 46) In spite of their biliary origin, only 25% ofcysts have a

connection with the biliary tree,(35, 46) which suggest that the underlying etiology maybe progressive dilatation of biliary microhamartomas which is secondary tofailure of normal connections with the biliary tree. Differentiation betweenlesions that communicate with the biliary system and those that do not isessential for deciding appropriate management. Although the exact etiology maynot be clear, the overall pathogenesis involves a congenital malformation withaccompanying inflammation and fluid accumulation from secreting biliaryepithelium.(18, 35) Therefore, destruction of the epithelium isthe basis for treating these lesions. The most common anatomic location issegment 5 of the right hepatic lobe,(35, 49) with right lobe lesions reported in the literaturetwice as long as left.

(18, 35) Unilocular cysts are much more common thanmultilocular cysts. The multilocular type is extremely rare in children.(18, 35) Cysts may be solitary, multiple, or diffusethroughout the liver (polycystic disease). Despite, they usually contain clearfluid with variable amounts of protein, cholesterol, blood cells, mucin, andepithelial cells.

(35, 49) ClinicalpresentationSolitaryliver cysts SLCs are typically asymptomatic and their symptoms do notmanifest until patients are in their forties or fifties.(3, 4) The anteroinferior segment of the right hepaticlobe is the most common site (3, 4) of SLC.(3, 4, 41) The pathophysiologyof simple liver cysts is mostly related to the continuous secretion of fluid byepithelial cells. In spite of this, only 10%–15% of liver cysts cause symptoms.

(35, 38) Most lesions are not symptomatic anddiscovered incidentally in adulthood. Abdominal discomfort and right upperquadrant pain are

commonly the first presenting symptoms in adults, butsymptoms can stay inactive as a consequence of the low internal pressure of thecysts.(35, 49) In thepediatric population, a hepatic cyst may be first discovered on prenatalimaging. In children, progressive abdominal distension is the most commonpresenting sign.(18, 22, 35, 39, 50)Associated symptoms are related to the space occupying effect of the lesion onadjacent viscera and include upper abdominal and right shoulder pain, nausea, vomiting, gastroesophageal reflux, early hungry, difficulty in breathing, and respiratorydistress.(35, 50, 51)Pressure on the vertebral column may cause lumbar pain, and pressure on thebladder may cause urinary frequency.

(35, 38) Most cysts do not occupy biliary or vascularstructures, but they may cause obstruction or compression atrophy.(35, 52) Infrequently, the cyst exerts pressure on theliver parenchyma or porta hepatis,(35, 53) which results in obstructive jaundice,(35, 37, 38, 46)cholangitis,(35, 36) or portal hypertension.(35, 38) Except for patients who present withobstructive jaundice, liver function tests are usually within normal limit.(35, 37) Liver failure has been reported, but it isextremely rare.(35, 53) Urgent or emergent surgical resection israrely necessary, and it is limited to acute complications, which includesinfection, bile leak, rupture, peritonitis, intra-cystic hemorrhage and torsion.(18, 35, 38, 52) An SLCis usually unilocular and lined by cuboidal or columnar epithelium with no connectionto the biliary tree.(4, 41) The outer layer of the cyst wall is composedof collagen, muscle fibers, bile ducts, and liver cells, which are separatedfrom the epithelial lining by vascular tissue.(4, 43) The cystic fluid may be clear, brown, oroccasionally bilious.

(4, 43)Thedifferential diagnosis included a mesenteric cyst, lymphatic malformation, hepatic malignancies, biliary atresia, choledochal cyst, or liver cyst. Accurate diagnosis is made primarily by radiographic studies often requiringhistopathological verification. On ultrasonography, a SLC appears as ananechoic unilocular fluid-filled space with a posterior acoustic enhancement.

(3, 4) SLC typically cannot be differentiated fromcystic malignant tumors on radiological studies; tissue diagnosis is necessary, even in asymptomatic patients.(3, 4) Histological criteria for an SLC include anouter layer of thin dense fibrous tissue, an inner epithelial lining (cuboidalor columnar), and lack of mesenchymal stroma or cellular atypia(4, 54) as in our case. Perioperativecholangiography should be performed in all cases to detect connections between theSLC and the biliary tree.(4, 41) The communication between the cyst and thebiliary tree was seen on cholangiography as in our case. The postoperativecourse may be complicated by infection, cholangitis, and recurrence.

(4, 41)TreatmentoptionsTreatmentof congenital solitary liver cyst is indicated for symptoms including abdominalpain, nausea, vomiting, obstruction, and complications like perforation, secondary infection, internal bleeding, torsion, and to rule out malignancy. Nonsurgical and surgical options are available. Nonsurgical options includeserial observation, and aspiration with or without sclerosis. Surgical optionsinclude enucleation that is cystectomy, fenestration that is unroofing, internal drainage, hepatic resection, and, rarely, hepatic transplantation. Celebi et al.

performed a 10-year retrospective review of congenital livercysts in their institution.(35, 55) Of the 67 pediatric cases diagnosed prenatallyor postnatally, in which, 10 underwent open surgery with the main indicationbeing a complication of the cyst or increasing size of the cyst, causingsymptoms such as abdominal pain due to mass effect. Four patients underwentopen fenestration, five underwent total enucleation, and one underwent apartial hepatectomy. Intraoperative cholangiogram was performed in cases where therewas preoperative evidence of biliary communication.(35, 55)NonsurgicaltherapyLivercysts were conventionally diagnosed when they were large enough to becomeapparent as an abdominal mass, or as an incidental finding during laparotomy orautopsy. Extensive use of improved imaging has led to increased detection ofincidental lesions.

(18, 35) The patients which are asymptomatic with smalllesions do not necessarily require treatment, as the possibility of complicationsis believed to be lower than the risk associated with treatment.(35, 53) Although considered benign, untreated cysts mightbe followed with a serial ultrasound or CT scan as consequence of the small riskof malignant transformation.(35, 37) Treatment of a liver cyst requires excision ofthe fluidsecreting epithelium. For this reason, simple percutaneous needleaspiration is almost universally ineffective and is associated with 100%recurrence.

(35, 38, 40, 53, 56) As aresult, simple aspiration has been accepted as definitive therapy. It is onlyappropriate in select circumstances as a temporizing measure to relieve severeor life-threatening symptoms, for example of, respiratory distress in a newborn, or when definitive treatment is

associated with high operative risk. Several reportsof sclerotherapy in the adult population have shown variable results,(35, 38) despite the fact that aspiration withsclerosis yields better results than aspiration alone.(35, 48)Sclerotherapy is less invasive and is associated with fewer complications thansurgical intervention, but it is associated with a higher recurrence rate. (35, 53, 56) In astudy of 23 adults with symptomatic nonparasitic hepatic cysts, 100% (6/6) ofthose who underwent sclerotherapy had a recurrence of their cyst, whereas 25%(2/8) of the patients' status postlaparoscopic fenestration had a recurrence.

(35, 57) There are few reports of congenital livercysts treated with sclerotherapy in the pediatric population.(35, 58) Intracystic ethanol injection was used in atleast one newborn infant with a rapidly enlarging liver cyst, with no evidenceof recurrence at 7-year follow-up.(35, 59) Caution must be used, as commonly usedsclerosing agents (e. g.

, ethanol) can cause alcohol intoxication andirreversible sclerosing cholangitis.(35, 48)SurgicaltherapySurgicalintervention is retained for significant abdominal or respiratory symptoms dueto cyst growth, acute complications, or neoplastic growth. Surgical options consistof fenestration, enucleation, internal drainage, resection, and rarely, livertransplantation. The basic principles are to completely remove or destroy thecyst epithelium, and to provide a mechanism for drainage of the cyst fluid. Ifthe cyst communicates with the biliary tree, an internal drainage procedure isnecessary. This procedure is also known as cystgastrostomy or cystjejunostomy.

If the cyst does not communicate with the biliary system, free drainage intothe peritoneal cavity is acceptable, because the fluid is reabsorbed by theperitoneum. Total excision should be attempted for easily accessible solitaryliver cysts (SLCs) and enucleating may also be chosen for small cysts. (4, 43) When total excision is not possible, partialexcision has been recommended.(4, 41) As much of the cyst wall as possible should beexcised, taking care not to damage vital structures.(4, 43) Internal drainage of the cyst viacystoenterostomy or Roux-en-Y hepaticojejunostomy has been recommended when thebiliary system drains into the cyst and bile drainage into the enteric circulationcannot be obtained.(4, 41) Biliary reconstruction is required in severalclinical situations, including resection of biliary malignancies, benignbiliary strictures, intraoperative injury, and liver transplantation. Bilioenteric anastomosis using the Roux-en-Y jejunum limb is a well-establishedapproach.

(4, 60) Reconstruction using the jejunum allows forsafe and permissive anastomosis with infrequent postoperative bile leakage evenwhen the anastomoses involve small intrahepatic branches(4, 61) as in our case. Techniques for surgicaltherapy have largely been adopted from the adult population. In 1968, Lin et al. described the technique of wide fenestration for polycystic liverdisease in two pediatric and three adult patients.(35, 59) meanwhile that time, the technique has alsobeen applied successfully by many surgeons to treat simple liver cysts.(35, 37) Wide fenestration consists of creating a largeopening or window on the nonparenchymal side of the cyst to allow free drainageof fluid into the abdominal cavity as in our patient 3.

Although the cyst wallcontinues to secrete, the peritoneal lining reabsorbs the fluid.

Symptomaticrelief is achieved, because fluid is no longer accumulating within the enclosedspace of the cyst wall. Since fenestration does not involve dissection ofhepatic parenchyma, it is associated with lower morbidity than enucleation. Fenestrationhas a recurrence rate ranging from 0% to 38%.

(35, 38) Adding omentoplasty and oversewing the marginswith a running suture appear to reduce the recurrence rate to 0%–14%.(35, 38) Enucleation (complete cyst excision) involvescomplete removal of the cyst epithelial lining, often with a small rim of theliver, resulting in complete cure. Complete cyst excision is often consideredthe treatment of choice for congenital hepatic cysts found in adults.(35, 56) Recurrence after partial resection is relatedto the remaining epithelium within the cyst wall.

Even a small amount ofresidual epithelium can lead to recurrence. Although enucleation yields thelowest recurrence rate (4%), morbidity can be high, as the operation requires dissectionof liver parenchyma with the associated risks of hemorrhage and bile leak.(35, 62)In 1991, Z'Graggen reported the first laparoscopic fenestration of a solitarynonparasitic hepatic cyst in an adult patient.(35, 63) Since that time, others have reported use ofthe laparoscopic technique, but mainly in case reports or small series thatprovides little data on long-term outcome.(22, 35, 37, 50)Arditoet al. treated 47 adult patients diagnosed with nonparasitic hepatic cysts withlaparoscopic fenestration due to either a symptomatic presentation or

anuncertain diagnosis. None of the cases required conversion to open surgery.

Theoverall recurrence rate was 14. 9% (seven patients), yet only 4. 3% (twopatients) presented with recurrent symptoms that necessitated surgicalmanagement. There was no recorded postoperative mortality or morbidity, andfollow-up ranged from 24 to 142 months.(35, 64) Although no randomized controlled trials areavailable to make definitive recommendations on laparoscopic versus opentherapy, available data suggest that symptom relief and recurrence rate afterlaparoscopic fenestration are similar to the open approach, but with lower morbidityand mortality.

(35, 38) Also, laparoscopic fenestration has been shownto result in less pain, decreased length of hospital stay, and earlier returnto normal activity when compared with open fenestration.(35, 56) As with the open approach, recurrence has beenattributed to failure to ablate the cyst lining, and failure to resect enough cystwall to allow for adequate drainage.(35, 65) Although recurrence rates and long-termresults are not known, laparoscopic fenestration is considered the standard ofcare for solitary nonparasitic hepatic cysts located on the liver surface.(35, 38, 53) Thereare limited accounts of laparoscopy being used for treatment of hepatic cystsin pediatric patients, especially in neonates, due to the limited experience oflaparoscopic surgery in this patient population.

(35, 40) Nordin et al. reviewed all the case reports ofneonatal congenital hepatic cysts in the literature and found that five out ofthe eight cases were

managed laparoscopically, with two undergoing complete cystexcision and three undergoing fenestration. Of the reviewed cases, nonereported any postoperative complications or recurrence.(35, 66) Laparoscopic surgery is associated with decreasedbleeding, decreased risk of ileus, shorter hospital stay, early mobilization, andimproved cosmesis.(35, 62, 67, 68) Relativecontraindications to the laparoscopic approach are lesions located deep insidethe hepatic parenchyma and those associated with hilar structures, as theseincrease the risk of extensive hemorrhage or damage to bile ducts.

(35, 37, 48) Laparoscopyshould be reserved for lesions located in accessible areas, namely the anteriorand lateral segments.(35, 48) In thepediatric population, especially, one must consider the small working space inthe abdominal cavity and the technical challenges this can presentlaparoscopically. Open surgical management should be reserved for cysts thatare not accessible by laparoscopy.(35, 46) Hepatic resection is appropriate for giant cysts, complex recurrent cysts, polycystic disease, diffuse liver involvement, andmalignancy.(35, 46, 48) Hepatictransplantation is rarely performed for hepatic cyst disease, but it isreserved for giant cysts or polycystic disease in which resection wouldcompromise hepatic function.