The association of exomphalos major, congenital liver cysts, and an accessory lobe of the liver is very rare. There are only two previously reported cases in the literature, both describing surgical excision of the accessory lobe with liver cysts during primary closure of the exomphalos defect. We report a case of this rare association, managed by delayed primary closure, where the cysts underwent spontaneous regression. This case, along with those previously reported, supports the etiopathogenesis theory of a malformative sequence of exomphalos and hepatic trauma within the sac of this rare association. Spontaneous regression of these cysts would favor a delayed primary closure in such cases.

**Keywords:** Accessory liver lobe, congenital cyst, exomphalos, primary closure

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### Case Report

**Surgical Dilemma: Exomphalos Major with Accessory Liver Lobe and Congenital Liver Cysts**

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**ABSTRACT**

The association of exomphalos major, congenital liver cysts, and an accessory lobe of the liver is very rare. There are only two previously reported cases in the literature, both describing surgical excision of the accessory lobe with liver cysts during primary closure of the exomphalos defect. We report a case of this rare association, managed by delayed primary closure, where the cysts underwent spontaneous regression. This case, along with those previously reported, supports the etiopathogenesis theory of a malformative sequence of exomphalos and hepatic trauma within the sac of this rare association. Spontaneous regression of these cysts would favor a delayed primary closure in such cases.

**KEYWORDS:** Accessory liver lobe, congenital cyst, exomphalos, primary closure

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**INTRODUCTION**

The incidence of exomphalos is 1 in 5000 live births and is defined as “major” when measuring >4 cm and/or with liver inside the sac. Exomphalos is associated with a high frequency of associated anomalies (80%), including chromosomal abnormalities (48%), congenital heart defects (28%), genitourinary malformations (20%), craniofacial anomalies (20%), and diaphragmatic anomalies (12%).[1] An abnormal liver anatomy has been reported with exomphalos and although rare is occasionally associated with different types of accessory lobes.[2] However, the association of an exomphalos major with congenital liver cysts in an accessory liver lobe is exceedingly rare. There are only two previously reported cases in the literature of exomphalos with accessory liver lobes similarly containing multiple cysts, found on antenatal ultrasound. In both cases, surgery was performed for excision of accessory hepatic lobe and cysts and closure of the exomphalos defect in the neonatal period.[3] This is the first such case to be reported, where delaying surgery for exomphalos allowed for spontaneous regression of the liver cysts, further aiding in an uncomplicated single-stage repair of exomphalos major defect.

**CASE REPORT**

A female term neonate, with an antenatal diagnosis of an exomphalos major defect and multiple liver cysts in the sac, was born by elective cesarean section. Following blood transfusion and stabilization, she was transferred to the pediatric intensive care unit for management of an iatrogenic birth injury of an umbilical cord bleed as a result of a tear at the attachment to the base of the exomphalos sac. A decision was made not to proceed with primary surgical repair of the exomphalos defect, as there was a risk of rupture to the liver cysts due to abdominal compartment syndrome and not being able to achieve a primary fascial closure given the size of the defect. The umbilical vessels were transfixed to stop the bleeding, and the edges of the ruptured sac were sutured to restore its integrity. The exomphalos defect which measured 5 cm, was managed with daily Flamazine ointment which resulted in escharification and epithelization forming a ventral hernia.

Ultrasound imaging at day 9 of life showed part of the liver contained within the sac along with multiple liver cysts, the largest of these measuring 6 cm × 3.7 cm. Stomach and kidneys were in an orthoptic position intra-abdominally [Figure 1a]. Both cranial and renal ultrasounds were reported as normal. Computed tomography imaging was performed at 3 months of age, showing multiple cysts measuring up to 6 cm × 3.3 cm within the liver, with no evidence of other associated congenital anomalies.

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tomography of the abdomen and pelvis on day 10 of life, demonstrated an exomphalos containing a loop of transverse colon, an accessory liver lobe and several liver cysts. The left portal vein and an accessory hepatic vein were seen to be crossing the defect to supply the herniated liver segment [Figure 1b].

By 4 months, the exomphalos major was fully epithelialized and a delayed primary closure was carried out in a single-stage procedure. Preoperative ultrasound showed marked reduction in size of herniated liver as well as resolution of the cysts. An upper midline incision was made over the epithelialized defect and the adherent part to the right lobe of the liver with the gallbladder was separated from the sac [Figure 2a]. A small accessory liver lobe was found to be adherent to the undersurface of the diaphragm with a feeding vessel from the liver which was transfixed and the accessory lobe was excised [Figure 2b]. A complete primary skin and fascial closure was achieved along with an umbilicoplasty. The baby made a good recovery from surgery and was discharged on postoperative day 5 on full oral feeds. She continues to thrive well at 9 months of age with no concerns, and her surgical repair is well healed with a good cosmetic outcome. Histological analysis of resected accessory lobe showed no evidence of any residual cyst. Postoperative follow-up imaging further demonstrated full resolution of all previously seen cysts.

**DISCUSSION**

Despite advances in medical and surgical therapies, management of exomphalos major can be challenging in the neonatal period. Conservative management of exomphalos major in the neonatal period is an established management strategy. The advantages of such an approach include avoidance of a major surgical procedure during the neonatal period, avoidance of silo construction and its attendant complications, and importantly allowing growth of the abdominal cavity to minimize the risk of abdominal compartment syndrome. However, rupture of the sac is considered to be a contraindication to continuation of conservative management because of the risks of desiccation, perforation, and infection of exposed intra-abdominal contents. Similar to previously published reports, the integrity of the ruptured sac was ingenuously restored by simple suturing which allowed for conservative management to be continued. Conservative management was continued to form a ventral hernia which was subsequently managed by a delayed single-stage repair. Surgical management either with a silo or primary closure in the neonatal period would have been challenging in our case, as there would be a risk of rupture of the cyst, abdominal compartment syndrome, and complications related to silo such as infections, tear or dislodgement of the silo, and fascial disruption, in addition to prolonged hospitalization, intensive monitoring, and parenteral nutrition.

Most of congenital liver cysts are usually considered simple hepatic cysts. Other congenital cystic lesions that can affect the liver in children are ciliated hepatic foregut cysts, cystic portion of a mesenchymal hamartoma, and cystic tumors such as mucinous cystadenomas or undifferentiated embryonal sarcomas. Multiple hepatic cysts can occur in polycystic liver disease. Exomphalos with associated malformations of an accessory liver lobe with cystic involvement is rare and unusual. Similar to our case, there are only two previously reported cases of exomphalos with multiple cysts in an accessory liver lobe, in the literature, both of which underwent surgical resection of the accessory liver lobe with the cyst on day 1 of life. These cysts were confirmed on histological analysis to be mesothelial inclusion cyst. In our case, the histology of the resected accessory lobe showed complete regression of the cyst with no residual cystic structure. The pathogenesis of these cysts is still hypothetical and has been addressed in relation to true splenic cysts. The plausible theory is that the hepatic cysts, similar to epithelial splenic cysts, could be derived from embryonic inclusions of pluripotent cells.
or from an abnormality in the coelomic organization. They could also be traumatic in origin, consecutive to a tear in the fibrous capsule, with secondary proliferation of the mesothelium. In our case, both theories could represent a plausible explanation for cystic lesions in a distorted accessory liver contained in a hernial sac. Our case, along with those previously reported, supports the etiopathogenesis theory of this rare association of malformative sequence leading to the development of an exomphalos or trauma endured by the hepatic parenchyma confined to the hernia sac.[3]

Management of exomphalos with rare anomalies such as an accessory lobe with multiple cysts can be challenging. In particular and unique to this report, we note a spontaneous regression of these congenital cysts, which would favor the decision for a delayed primary closure in such cases.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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