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CLINICAL CASE REPORT

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Thoracoscopic Approach for Repair of Diaphragmatic Hernia Occurring After Pediatric Liver Transplant

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Abstract: Diaphragmatic hernias (DH) occurring after pediatric liver transplantation (LT) are rare. However, such complications have been previously reported in the literature and treatment has always been surgical repair via laparotomy. We report our experience of minimally invasive thoracoscopic approach for repair of DH occurring after LT in pediatric recipients.

From April 2010 to December 2014, 7 cases of DH were identified in pediatric LT recipient in Samsung Medical Center. Thoracoscopic repair was attempted in 3 patients. Patients' medical records were retrospectively reviewed.

Case 1 was a 12-month-old boy, having received deceased donor LT for biliary atresia (BA) 5 months ago. He presented with dyspnea and left-sided DH was detected. Thoracoscopic repair was successfully done and the boy was discharged at postoperative day 7. Case 2 was a 13-month-old boy, having received deceased donor LT for BA 2 months ago. He presented with vomiting and right-sided DH was detected. Thoracoscopic repair was done along with primary repair of herniated small bowel that was perforated while attempting reduction into the peritoneal cavity. The boy recovered from the surgery without complications and was discharged on the 10th postoperative day. Case 3 was a 43-month-old girl, having received deceased donor LT for Alagille syndrome 28 months ago. She was diagnosed with right-sided DH during steroid pulse therapy for acute rejection. Thoracoscopic repair was attempted but a segment of necrotic bowel was noticed along with bile colored pleural effusion and severe adhesion in the thoracic cavity. She received DH repair with small bowel resection and anastomosis via laparotomy.

Thoracoscopic repair was attempted in 3 cases of DH occurring after LT in pediatric recipients. With experience and expertise in pediatric minimally invasive surgery, thoracoscopic approach is feasible in this rare population of patients.

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Abbreviations: BA = biliary atresia, CDH = congenital diaphragmatic hernia, DH = diaphragmatic hernia, ER = emergency room, GRWR = graft-recipient weight ratio, LLS = left lateral section, LT = liver transplant.

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INTRODUCTION

Liver transplant (LT) is a life-saving treatment for patients with end-stage liver disease from various etiologies. Survival data reported from analyses of large registries have shown excellent long-term graft and patient survival for pediatric LT.[1,2] Pediatric LT is a complex surgical procedure and may lead to various surgical and nonsurgical complications in the posttransplant period. Diaphragmatic hernia (DH) is a rare complication occurring in LT recipients. Less than 30 cases of DH after LT have been reported in the literature to date, usually as case series of single-center experiences.[3–8] Potential factors contributing to the development of DH after LT are increased intraabdominal pressure with a thin or weakened diaphragm.[6] A small infant receiving a large-for-size left lateral section (LLS) graft would be the typical description of a case with DH occurrence after LT.

Surgical repair by way of primary closure or patch closure is the treatment of choice for DH occurring in children of all ages. The conventional approach for DH repair has been by open laparotomy or thoracotomy. Minimally invasive method for DH repair has recently been successfully attempted in small children, as well as neonates.[9–11] Recent reports have suggested that thoracoscopic repair of DH may be associated with decreased morbidity compared to open repair.[12] The surgical approach for DH after LT described in the literature was limited to laparotomy or thoracotomy. In this report, we present 3 cases of DH occurring in pediatric LT recipients that were managed with a minimally invasive thoracoscopic approach.

METHODS

From June 1996 to December 2014, 217 cases of pediatric LT were performed at Samsung Medical Center (Seoul, Korea). A retrospective medical record review was conducted of all cases and 7 cases of DH occurring in the posttransplant period were identified (7/217, 3.2% incidence). Four earlier cases were previously reported.[6] Here, we describe 3 recent cases which were managed by a minimally invasive thoracoscopic approach.

The surgical procedure for thoracoscopic DH repair is done as follows. After general anesthesia, the patient is placed in supine position with a roll inserted beneath the lower part of the ipsilateral thorax to elevate the affected diaphragm. A 5 mm trocar (camera port) is inserted in the 4th intercostal space on the anterior axillary line. Carbon dioxide instillation is initiated at 3 to 5 L/minute and up to a pressure of 5 mm Hg. Two additional 3 mm trocars are inserted on either side of the camera port for use as working ports. Gentle manipulation of the bowel is done to reduce the herniated organs back into the abdominal cavity and interrupted sutures using 3-0 or 4-0 silk are applied to repair the defect in the diaphragm. Chest tubes are not routinely inserted. All cases were performed by a single surgeon (JMS) experienced in pediatric thoracoscopic surgery.

Posttransplant management of LT recipients was done according to the in-house protocol at our transplant center. A

bolus of methylprednisolone was given during the transplant procedure before reperfusion of the graft and tapered off within 4 weeks. Maintenance immunosuppression was tacrolimus monotherapy.

This study was approved by the Institutional Review Board at Samsung Medical Center (SMC 2015-07-103). The Board waived the requirement for written informed consent.

RESULTS

Patient characteristics and clinical course before and after DH repair are outlined in Table 1.

Case 1 was a 12-month-old boy. He had received deceased donor liver transplant (DDLT) with a LLS graft for biliary atresia (BA) at 7 months of age. His posttransplant course was uneventful except for an incidence of asymptomatic cytomegalovirus antigenemia that was treated preemptively with intravenous ganciclovir. He presented to the emergency room (ER) with recurrent vomiting. Chest X-ray taken in the ER revealed an abnormal air density in the left thorax with displacement of the mediastinal structures to the right side (Figure 1A). Left-sided DH was suspected and the patient was taken to the operating room. Thoracoscopic exploration of the left thoracic cavity was done and herniation of the stomach through a 4 cm defect in the mid portion of the left diaphragm was noticed (Figure 1B). The stomach was reduced into the abdomen and the defect was closed with interrupted sutures. Feeding was initiated on the following day and the patient was discharged without complications on postoperative day 7. The patient has been followed for 42 months since thoracoscopic DH repair without recurrence.

Case 2 was a 13-month-old boy. He had received DDLT with a LLS graft for BA at 11 months of age. He presented to the ER with vomiting and decreased activity. Chest X-ray taken in the ER revealed a subtle upward bulging of the right diaphragm with abnormal-appearing air density in the right thorax (Figure 2A). DH was suspected and confirmed with thoracoabdominal ultrasound revealing a right diaphragmatic defect with herniated bowel loops. Upon insertion of the thoracoscope into the right thoracic cavity, large amount of serous pleural effusion was observed. Loops of small bowel were herniated through a 2 cm-sized defect in the right diaphragm. The herniated bowel was reduced into the abdomen after laterally extending the defect by 2 cm (Figure 2B). The incarcerated bowel loops appeared slightly congested at first, but regained its normal color after reduction. Iatrogenic injury causing a small perforation in the herniated bowel was made during manipulation of the bowel with thoracoscopic instruments. The site of perforation was closed with intracorporeal interrupted sutures. The defect was closed with interrupted sutures and a 10 Fr chest tube was inserted. Feeding was initiated on the following day and the chest tube was removed on postoperative day 4. The patient was discharged without complications on postoperative day 9. The patient has been followed for 5 months since thoracoscopic DH repair without recurrence.

Case 3 was a 43-month-old girl who had received DDLT with a LLS graft for Alagille syndrome at 15 months of age. She was admitted for evaluation of elevated liver enzymes and acute cellular rejection was diagnosed by liver biopsy. On the 5th day of steroid pulse therapy she developed high fever and shortness of breath. Chest X-ray and chest computed tomography were taken and massive right pleural effusion with a bowel-like mass bulging through a defect in the right diaphragm was seen (Figure 3). DH was suspected and emergency operation by thoracoscopic approach was undertaken. The right thoracic cavity was filled with bile-colored fluid and severe adhesion was present. Segmental resection and anastomosis of the perforated bowel seemed necessary and the procedure was converted to laparotomy via a previous right subcostal incision. The site of the perforation was on the mid-portion of the Roux-en-Y limb. Segmental resection and hand-sewn end-to-end anastomosis was done. The diaphragmatic defect was closed with interrupted sutures. Feeding was initiated on the 3rd day following surgery. Large amount of pleural effusion developed which was drained with a percutaneously inserted catheter. The patient was discharged on postoperative day 25. The patient has been followed for 8 months without recurrence.

DISCUSSION

In this case series, we report 3 cases of DH occurring in pediatric LT recipients that were managed by thoracoscopic

TABLE 1. Patient Characteristics and Clinical Course

	Case 1	Case 2	Case 3
Sex	Male	Male	Female
Age, months	12	13	43
Body weight, kg	8.7	8.4	13.7
Liver transplant characteristics			
Donor type	Deceased	Deceased	Deceased
Graft type	Split LLS	Split LLS	Split LLS
GRWR, %	4.6	4.8	3.26
Etiology	Biliary atresia	Biliary atresia	Alagille syndrome
Diaphragmatic hernia characteristics			
Left/right	Left	Right	Right
Time from LT, months	5	2	28
Herniated organ	Stomach	Small intestine	Small intestine
Surgical approach	Thoracoscopy	Thoracoscopy	Thoracoscopy, laparotomy
Postoperative LOS, days	7	9	25

GRWR = graft-recipient weight ratio, LLS = left lateral section, LOS = length of stay, LT = liver transplant.

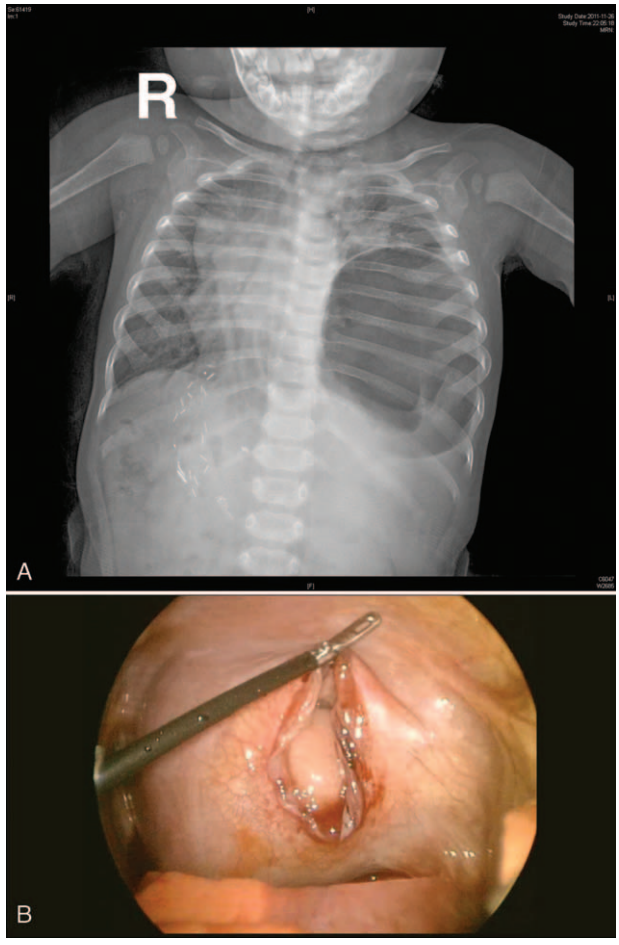

FIGURE 1. (A) Chest X-ray showing a large abnormal air density in the left lower thorax and (B) thoracoscopic view of the left diaphragmatic defect with the stomach reduced into the peritoneal cavity in Case 1.

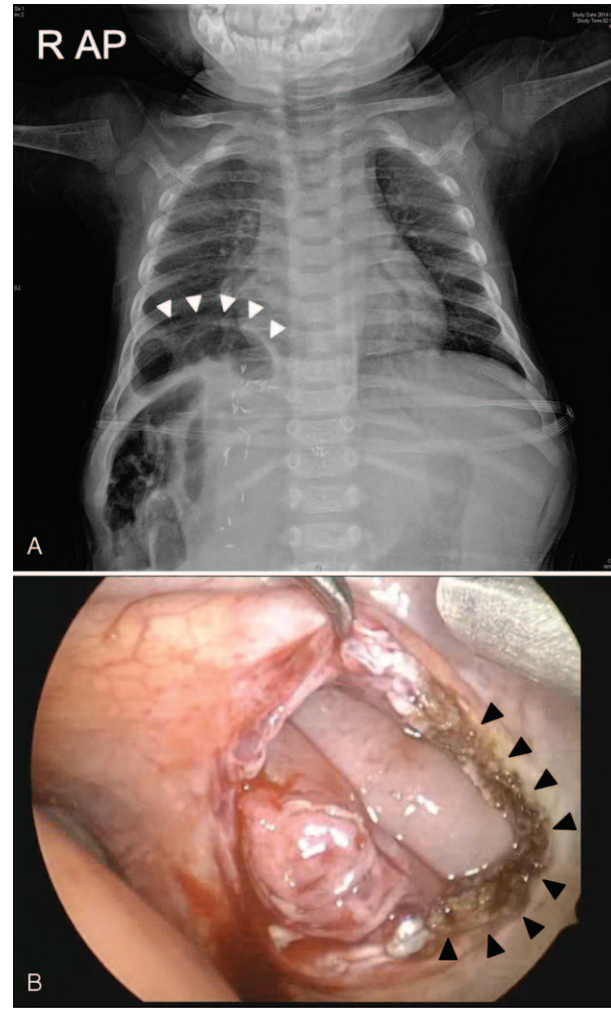

FIGURE 2. (A) Chest X-ray showing an upward bulging air density (white arrows) through the right diaphragm and (B) thoracoscopic view of the right diaphragmatic defect after reduction of the herniated small intestine in Case 2. Black arrows indicate the laterally extended portion of the diaphragmatic defect.

surgery. Two cases of DH were successfully repaired with this minimally invasive approach and 1 case required a combined laparotomy approach for small bowel anastomosis. To the best of our knowledge, this is the first report of thoracoscopic approach for the management of DH occurring in the post-transplant period in pediatric LT recipients.

Thoracoscopic repair of congenital diaphragmatic hernia (CDH) has been gaining interest as an effective method of minimally invasive surgery for this disease in newborns and older children.[9–11] However, there have not been any well-designed prospective trials comparing thoracoscopic DH repair versus the traditional open method by laparotomy and outcome measures such as hernia recurrences have varied between different studies.[13] We have found the thoracoscopic approach for DH in pediatric LT recipients to be feasible and possibly advantageous in some aspects. With thoracoscopic approach for DH, the surgeon is able to gain direct access to the diaphragmatic defect. The right upper abdomen of LLS graft recipients is usually packed with the Roux-en-Y jejunal limb and is the site of most severe postoperative adhesion. To approach the right diaphragm via the previous incision, a laborious and time-consuming adhesiolysis must be done which may lead to iatrogenic bowel injury. No previous surgical manipulation to the thoracic cavity has been done in most LT recipients, which would allow for safe and easy access to the diaphragm when approached thoracoscopically. Another point is that most DH occurring in pediatric LT recipients are small defects located posteromedially.[6] The most problematic cases in thoracoscopic repair of neonatal CDH are large defects located anteriorly. A large defect may exert tension on the sutured margins and lead to recurrence of DH.[14] Patch closure of large defects by thoracoscopy is associated with longer operative time and varying degrees of hernia recurrence compared to primary repair.[15,16] The diaphragmatic defect seen in LT recipients are usually small and may be approximated without tension by interrupted sutures.[3,4,6,7] The difficulty of this approach in smaller defects is in reducing the herniated bowel into the abdominal cavity. In this case, we have found it helpful to linearly extend the diaphragmatic defect for easier reduction. This technique, which we have applied in 2 of the 3 cases, allows for safe and effective reduction of the bowel with less risk of iatrogenic injury from sharp thoracoscopic instruments.

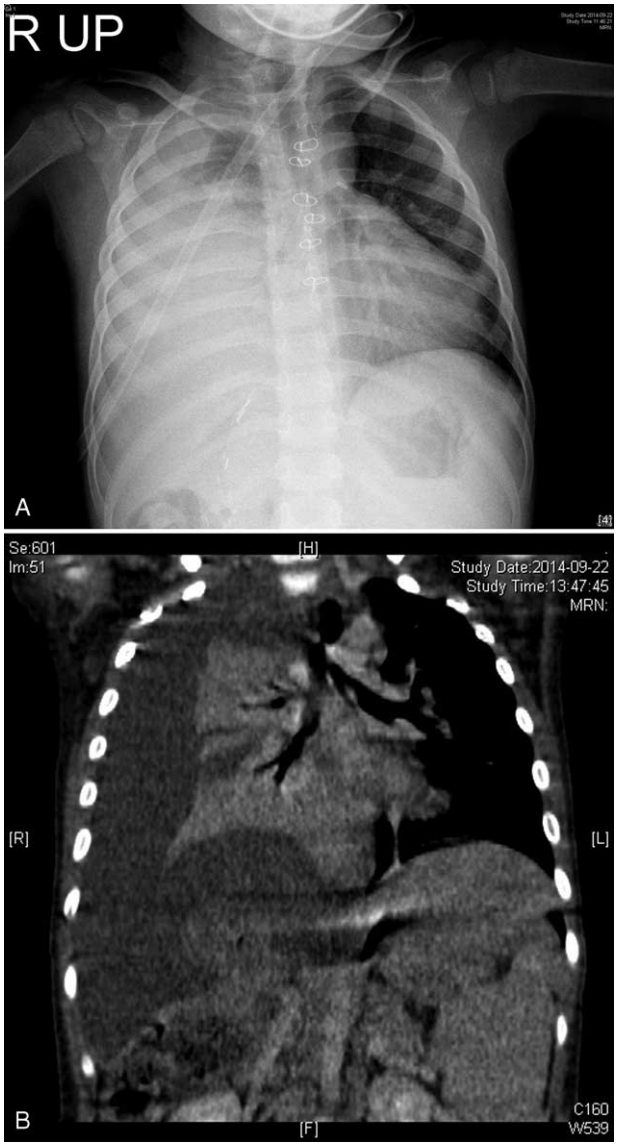

FIGURE 3. (A) Chest X-ray and (B) computed tomography of the patient (Case 3). There is massive pleural effusion in the right thorax.

The pathogenesis of DH after LT has been postulated to be direct surgical trauma, usually thermal injury from electrocautery to the bare area of the diaphragm.[3,6] There is usually extensive collateral vessel formation between the cirrhotic liver and the diaphragm of the pediatric LT recipient. Moreover, many pediatric LT recipients have had previous surgical procedures done which results in adhesion surrounding the liver. The extensive use of diathermy on the bare area of the diaphragm for hemostasis and the occasional iatrogenic injury to the diaphragm during hepatectomy may cause ischemia leading to latent necrosis and perforation of the diaphragm. Risk factors reported to be associated with DH in pediatric LT recipients include younger age at transplant, use of LLS grafts and high graft-recipient weight ratio (GRWR).[3,7] Cases 1 and 2 show typical features associated with DH after LT, with small infants receiving very large LLS grafts (GRWR > 4.5). Case 3 was

TABLE 2. Overview of Diaphragmatic Hernia Cases After Pediatric Liver Transplant in Our Center

	Patient Characteristics			Transplant Characteristics				Diaphragmatic Hernia Characteristics			
Case	Sex	Age (Months)	Weight (kg)	Donor Type	Graft Type	GRWR	Etiology	Location	Time From LT (Months)	Surgical Approach	
1	F	35	14.7	LD	LLS	1.6	AS	R	1	Laparotomy	Moon et al
2	F	6	8.5	LD	LLS	2.5	IHHE	R	7	Laparotomy	
3	M	74	19.4	LD	LLS	1.6	BA	R	2	Laparotomy	
4	F	7	7.4	LD	LLS	3.8	BA	R	3	Laparotomy	
5	M	12	8.7	DD	LLS	4.6	BA	L	5	Thoracoscopy	Present study
6	M	13	8.4	DD	LLS	4.8	BA	R	2	Thoracoscopy	
7	F	43	13.7	DD	LLS	3.3	Alagille syndrome	R	28	Thoracoscopy, laparotomy	

AS = angiosarcoma, BA = biliary atresia, DD = deceased donor, GRWR = graft-recipient weight ration, IHHE = infantile hepatic hemangioendothelioma, LD = living donor, LLS = left lateral section, LT = liver transplant.

older and the timing of DH occurrence was later in the post-transplant period compared to other cases in our center (Table 2). However, this patient had 5 episodes of acute rejection that required large doses of bolus steroid injections. Steroids and higher doses of other immunosuppressants are thought to be associated with impaired healing and may add to the risk of DH development.[3,5]

In conclusion, we report 3 cases of DH occurring in pediatric LT recipients that were managed by thoracoscopic surgery. In experienced hands, this technique can be safely applied in pediatric LT recipients presenting with DH.

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