Thoracoschisis: case report and review of the literature

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Introduction **Thoracoschisis is a rare congenital malformation characterized by herniation of intra-abdominal contents through a thoracic wall defect. There have been six previously reported cases. We describe our novel approach incorporating closure of the chest wall defect with temporary abdominal wall expansion utilizing a silastic pouch.**

Case report **A male child born at 29 weeks' gestation was transferred to our institution for the management of a right anterior chest wall defect with herniation of intra-abdominal contents through this defect. The patient was taken to the operating room for reduction of the herniated viscera from the right chest wall defect into the abdomen utilizing a spring-loaded silastic pouch to cover the abdominal viscera.**

Discussion **The cause of thoracoschisis is unclear. Multiple mechanisms have been proposed for the development of thoracoschisis, including amnionic rupture, vascular injury, and embryologic maldevelopment. In previously reported cases, a majority of patients had associated limb abnormalities. It has been proposed that this association between extremity agenesis/deformity and chest wall defects is related to the limb–body wall complex. In addition, most of the cases reported also had an accompanying diaphragmatic defect, allowing the abdominal viscera to enter the chest and then herniate through the thoracic defect.**

Conclusion **Overall, thoracoschisis is a very rare congenital abnormality characterized by a chest wall defect with herniation of intra-abdominal organs through this defect. Previously, only six cases have been reported, most of which had an associated limb anomaly or diaphragmatic hernia. *Ann Pediatr Surg* 11:143–146 © 2015 Annals of Pediatric Surgery.**

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Introduction

Thoracoschisis is a very rare congenital anomaly characterized by the evisceration of intra-abdominal organs through a thoracic wall defect. Currently, there are only six previously reported cases (Table 1). In most of the reported cases, a limb abnormality accompanies the chest wall defect. Multiple etiologies have been proposed as potential causes of this anomaly, including vascular abnormalities, amnion rupture, and fetal maldevelopment [5]. These cases pose many surgical and anesthetic challenges.

Case report

A male child, weighing 1680 g, was born at 29 weeks' gestation to a 23-year-old mother by spontaneous preterm labor. The patient had received minimal prenatal care starting only 1 week before birth. He was intubated immediately after birth for hypoxia and respiratory distress and transferred to our institution for the management of a right anterior chest wall defect with herniation of intra-abdominal contents through this defect (Fig. 1). A right radial arterial line and peripheral intravascular lines were placed in the neonatal intensive care unit for close hemodynamic monitoring and fluid administration. He was placed on a conventional ventilator and despite aggressive ventilator settings had a preoperative arterial blood gas of pH 7.16, $PaCO_2$ 70, PaO_2 50, bicarbonate 25, base deficit −5, and oxygen saturations in the low 90 s. Chest radiograph was obtained and indicated 11 ribs bilaterally and a mild elevation of the right hemidiaphragm. A transthoracic echocardiogram was also performed showing a patent foramen ovale, small patent ductus arteriosus with no evidence of pulmonary hypertension, normal cardiac anatomy, and normal biventricular function.

He was then taken to the operating room the same day for exploration and reduction of herniated viscera. Anesthesia was induced and maintained with fentanyl, rocuronium, and sevoflurane. A left femoral central line was placed for possible administration of vasoactive agents. Ventilation and oxygenation proved to be very difficult secondary to pulmonary hypoplasia and atalectasis from compression. Because of the small size of the infant and because surgical manipulation in the right chest led to decreases in tidal volume, manual ventilation was required throughout the operation.

Upon inspection of the deformity, it appeared to be emerging through the right chest wall at the level of the fifth through the eighth ribs. The defect measured 4×3 cm. Herniated through this defect was a large amount of small bowel, duodenum, and most of the liver. In multiple locations, the herniated viscera adhered to the rim of the chest wall defect. The abdominal midline was opened from the xiphoid to an infraumbilical location. Upon entering the peritoneal cavity, it was

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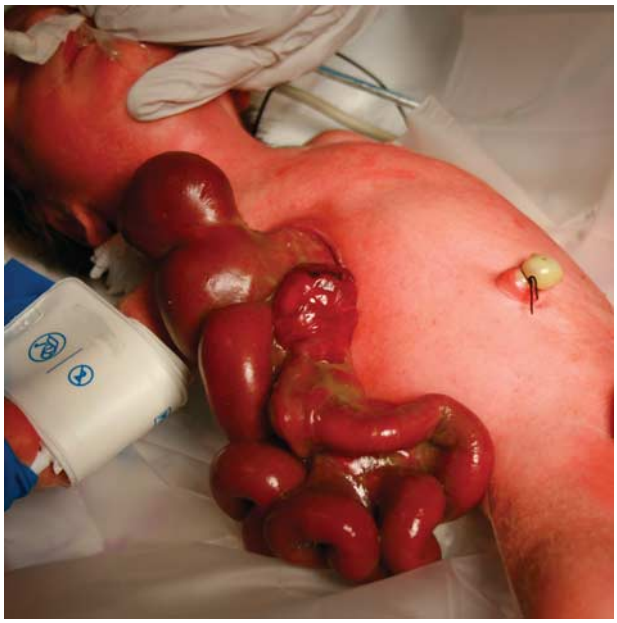
Table 1 All reported cases of thoracoscshisis

	Sex	Defect size (cm)	Defect location	Herniated organs	Diaphragmatic defect	Additional abnormality	Result
Davies *et al.* [1]	F	3	Left third intercostal	Left liver lobe, stomach, transverse colon, omentum	Left anterolateral	No left forearm, syndactyly	Alive
Bamforth *et al.* [2]	F	4 × 3	Left sixth rib	Left liver lobe	Left posterior	Left Poland anomaly, left scapula hypoplastic, forearm agenesis, ectopic pancreas	Alive
Derbent and Balci [3]	F	4 × 3	Right second to fourth ribs	Liver, intestines	Right anterolateral	NA	Intrauterine death
Biri *et al.* [4]	F	3 × 4	Left	Left liver lobe	Hiatal hernia	Forearm agenesis, ventricular septal defect	Died at birth
Karaman *et al.* [5]	M	2 × 2	Left eighth intercostal	Liver, transverse colon, omentum	No	Left diaphragm eventration, atrial septal defect, patent ductus arteriosus	Alive
This study	M	3 × 4	Right fifth to eighth ribs	Liver, intestines, omentum	No	Right fourth digit abnormality	Alive

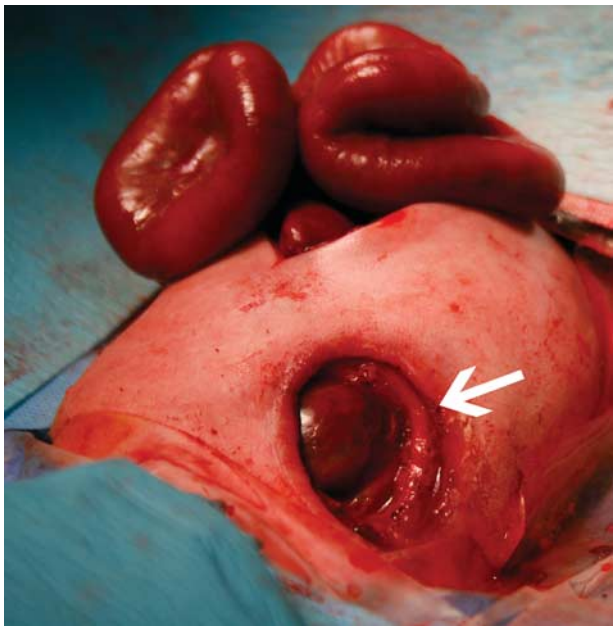
F, female; M, male; NA, not applicable.

Fig. 1



Preoperative photo showing a small bowel and liver herniated through the thoracic defect.

Fig. 2



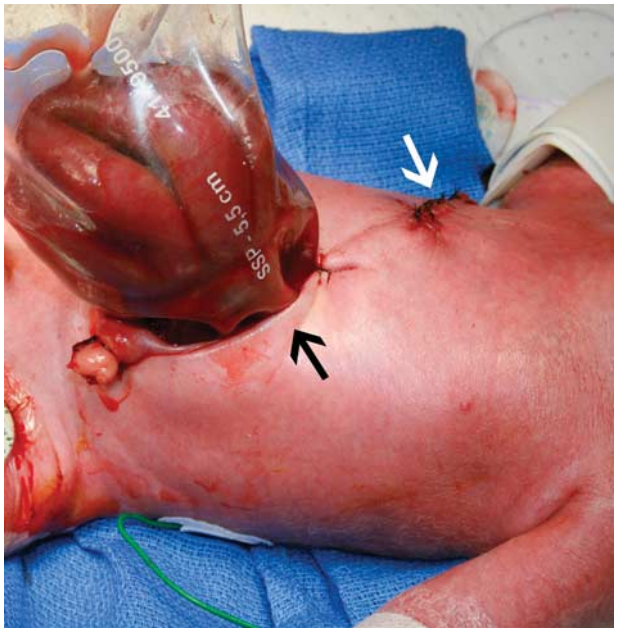
Chest wall defect after reduction of viscera into the abdomen (white arrow). Liver seen through the defect.

apparent that there was a connection between the abdomen and the right chest wall without disruption of the right diaphragm, which was located at a much higher than normal position. The lung could be seen through the thin diaphragm, but there was no opening into the pleural cavity. The diaphragm did not appear redundant or to require plication. At this point, the herniated bowel and liver were delivered from the right chest wall defect into the abdomen (Fig. 2). Following this, the respiratory and metabolic acidosis eventually improved.

Inspection of the bowel itself failed to show any evidence of atresia, ischemia, or necrosis. All other visible viscera appeared normal. Because of lack of mobility and adequate tissue, Surgisis biosynthetic mesh (Cook Medical, Bloomington, Indiana, USA) was utilized to close the deep portion of the chest wall defect. This was sutured circumferentially to the defect. Finally, the skin was sufficiently mobilized by raising skin flaps to allow primary closure over the mesh. A 5.5 cm diameter spring-loaded silastic pouch was utilized to cover the abdominal viscera. (Fig. 3)

At the end of the case, after a period of stability, the infant suddenly developed hypoxia and hypotension. Mainstem intubation, pneumothorax, right heart failure, anemia, acidosis, and hypothermia were all considered and a chest radiograph, transthoracic echocardiogram, and labwork were performed. It was believed that the sudden decompensation may possibly have been because of intermittent mainstem intubation with position changes in preparation for transport. The infant was transferred back to the NICU and was placed on a high-frequency oscillator for a few hours and then transitioned back to conventional mechanical ventilation. Following his initial operation, an investigation for additional congenital abnormalities, including abdominal and cranial untrasonography, indicated no malformations other than a right fourth digit anomaly. Because of the large amount

Fig. 3



Closed chest wall defect (white arrow) and silastic pouch covering abdominal viscera (black arrow).

of intestines contained within the silastic pouch and the small size of the patient, the abdominal viscera were subsequently reduced in a staged manner over the next several days at bedside in the NICU. On postoperative day 11, the final reduction and primary abdominal closure was completed in the operating room without difficulty. The patient required an extended period of time with ventilator support, likely because of underdevelopment of the right lung and altered chest wall dynamics, and ultimately required a tracheostomy. A gastrostomy tube was also placed for nutritional support. After several months of respiratory and nutritional support, the patient was eventually weaned from the ventilator, tolerating oral feedings, and discharged home.

A literature review was completed using Pubmed, Biosis, and additional sources using the following search terms: thoracoschisis, limb–body wall complex (LBWC), thoracoabdominal schisis, gastroschisis. All previous publications were in English.

Discussion

Thoracoschisis is a very rare congenital abnormality characterized by the herniation of intra-abdominal organs through a chest wall defect. Currently, only six cases have been reported. In previously reported cases, the thoracic defect was often associated with additional abnormalities, such as limb anomalies and diaphragmatic hernias.

The cause of thoracoschisis is unclear. Multiple mechanisms have been proposed, including amnionic rupture, vascular injury, and embryologic maldevelopment. Amnionic rupture may lead to thoracoschisis by interrupting normal morphogenesis or by deforming already developed fetal structures [6]. In contrast, vascular injury can cause structural abnormalities by damage to or interruption of fetal artery and vein development.

In this case, a localized vascular injury within the thoracic wall would lead to agenesis and thoracoschisis [7]. An additional theory suggests that a teratogen may alter the lateral body folds, causing a defect in the closure of the ventral body wall. A lack of adequate ventral advancement of these folds leads to the absence of fusion within the body wall. This theory also explains pectoral muscle defects and limb abnormalities observed in previously reported cases [2,3,8].

In the previously reported cases, a majority of patients had associated limb abnormalities. It has been proposed that this association between extremity agenesis/deformity and chest wall defect is related to the LBWC. LBWC defects are a form of amniotic band syndrome that consists of a thoracic or abdominal defect, herniation of internal organs through this defect, and associated anomalies of the extremities. This is caused by the formation of bands between the amniotic membrane and fetus, leading to neural tube defects, abdominal or thoracic wall defects, and extremity abnormalities [9]. This may explain some of the previously reported cases of thoracoschisis, but is unlikely to be the cause of our patient's chest wall defect as he had an isolated thoracoschisis without amniotic bands or severe limb abnormalities described previously in cases of LBWC.

Most of the reported cases of thoracoschisis also had an accompanying diaphragmatic defect, allowing the abdominal viscera to enter the chest and then herniate through the thoracic defect. It has been hypothesized that the primarily thoracic defect allowed the abdominal organs to herniate through the chest wall before diaphragm development was complete, resulting in a persistent diaphragmatic hernia [5]. In only one previously reported case and in our patient, the diaphragm was intact, but was located at a more superior position than normal. This higher position of the diaphragm is likely because of its formation above the herniated abdominal organs through the chest wall defect. In all of the patients who had an associated diaphragmatic hernia, the thoracic wall defect was at a higher position on the chest between the second and sixth ribs.

Because of the small overall size of our patient and the large volume of abdominal viscera herniated through the chest wall defect, we decided to proceed with a temporary abdominal closure using a silastic pouch with a staged abdominal closure. This novel technique has not been described previously for use in thoracoschisis patients and is similar to the silos used for gastroschisis. In addition, this allowed for decreased abdominal pressure and therefore decreased intrathoracic pressure to aid in improved ventilation. This was important in our patient as there was already some degree of pulmonary compromise and increased pressures would have only worsened this.

There are no cases in the literature describing the anesthetic care and the exact surgical technique required for repair of such cases. Extensive preoperative preparation with adequate vascular access and hemodynamic

monitoring was necessary not only in anticipation of suspected blood loss and fluid shifts but also for administration of vasoactive agents. Intraoperative anesthetic management in terms of oxygenation and ventilation in the setting of pulmonary hypoplasia, restrictive lung physiology, and atalectasis was challenging. This involved one anesthesiologist who focused solely on securing the endotracheal tube with one hand and manually ventilating with the other. The large amount of surgical manipulation in the setting of the small size of the infant toggled on the edge of mainstem intubation and complete extubation. In addition, aggressive resuscitation with blood products was necessary to keep up with blood loss and fluid shifts. The option for extracorpeal membrane oxygenation did not exist in this case because of the size and age of the infant. The use of a pulmonary vasodilator such as inhaled nitric oxide was considered, but deemed unlikely to be beneficial in the setting of a normal preoperative echocardiogram.

As with gastroschisis, thoracoschisis can be diagnosed prenatally by ultrasound, but given the rarity of thoracoschisis, it is likely to be misdiagnosed as either gastroschisis or omphalocele. Associated limb abnormalities may also be seen on ultrasound. Our patient received late prenatal care starting only 1 week before birth. The patient had been diagnosed with gastroschisis by ultrasound.

Conclusion

Overall, thoracoschisis is a very rare congenital abnormality characterized by a chest wall defect with herniation of intra-abdominal organs through this defect. Previously, only six cases have been reported, most of which had an associated limb anomaly or diaphragmatic hernia. The use of a temporary silastic pouch allows for a staged abdominal closure in which the viscera can be reduced over multiple days to reduce intra-abdominal and intrathoracic pressure.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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