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Bilateral diaphragmatic paresis following pediatric liver transplantation

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Abstract

Background: Although diaphragm paresis or paralysis is fairly common following cardiac procedures; it is a less common complication following liver transplantation. Unilateral diaphragm paresis, usually right sided, has been described following liver transplantation in adults and has been rarely described in children.

Purpose: Diaphragmatic injury following LT is often unrecognized and is typically unilateral, involving the right hemidiaphragm. Bilateral diaphragm dysfunction following liver transplantation in children is a rare complication.

Methods: This is a case report of bilateral diaphragm paresis in a young child following a repeat liver transplantation.

Conclusion: Bilateral diaphragm paresis following liver transplantation in children is rare and spontaneous resolution is possible. A conservative approach with noninvasive ventilation as a first line treatment to allow the diaphragm to regain function should be considered.

Keywords

Respiratory medicine, gastroenterology/hepatology, liver, transplantation, diaphragm, paresis

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Introduction

Liver transplantation (LT) has become a widespread form of therapy for the treatment of end-stage liver disease in adults and children. Pulmonary complications have been reported as an important cause of postoperative morbidity and mortality in LT recipients.[1] Diaphragm paresis (DP) is an uncommon and often unrecognized complication following LT. Right DP has been reported relatively more frequently as a complication of LT.[2] Bilateral diaphragm injury after LT is much less frequent.

Case presentation

Patient is a 22-month-old female with a history of LT at 11 months of age in China after a failed Kasai procedure for biliary atresia presented with recurrent cholangitis. She was adopted at 18 months of age and came to the United States at that time. The initial transplantation was complicated by hepatic artery thrombosis and poly-microbial multidrug-resistant recurrent ascending cholangitis with *Klebsiella pneumoniae, Acinetobacter baumannii*, ESBL (extended spectrum beta lactamase)-producing *Escherichia coli*, and ampicillin-resistant *Enterococcus*. The patient was diagnosed with liver cirrhosis based on liver biopsy and a decision for repeat transplantation was made.

The repeat LT procedure at 22 months of age was complicated by the presence of dense adhesions and significant blood loss. The patient received fresh frozen plasma, platelets, and packed red blood cell transfusions during the procedure and postoperatively. Approximately 2 h were spent obtaining hemostasis with cautery. She was transferred to the pediatric intensive care unit (PICU) on mechanical ventilation for postoperative care.

Physical examination in the PICU revealed a sedated infant on multiple pressors. Her temperature was 38°C, pulse of 129, respiratory rate of 30, blood pressure of 82/46 mmHg, and pulse oximetry of 91% on mechanical ventilation and supplemental oxygen. Physical examination at that time was notable for equal breath sounds and a postoperative abdomen with some oozing of frank blood along the midline incision. On the

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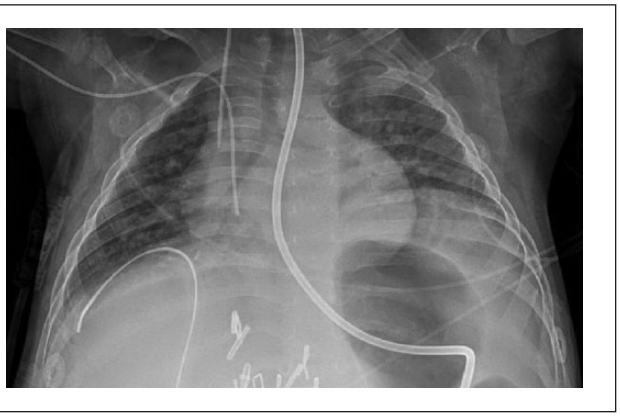

Figure 1. Chest X-ray showing bilateral decreased lung volumes with asymmetric elevation of left diaphragm.

third postoperative day (POD), the patient was extubated but required continued respiratory support with noninvasive ventilation (NIV) in the form of bi-level positive airway pressure (BiPAP) via nasal mask. She remained relatively stable and tolerated weaning until POD 7 when she began experiencing increased respiratory distress. A chest X-ray (CXR) revealed left basilar atelectasis with asymmetric elevation of the left hemidiaphragm (Figure 1). There was suspicion for left hemidiaphragm paralysis; however, fluoroscopy of the diaphragms revealed bilateral diaphragmatic paresis (DP) as no movement of either diaphragm was detected. A preop chest CT was reviewed; the lung parenchyma seemed intact. Spirometry was not performed because patient couldn't tolerate this procedure. In addition to ventilatory support, the patient also received treatment with nebulized ipratropium and 3% hypertonic saline and also received manual chest physiotherapy during the hospitalization.

After a prolonged PICU stay and substantial clinical improvement, the patient was transitioned to high-flow nasal cannula (HFNC) while awake and BiPAP while asleep at POD 27 and was transferred to the general pediatrics floor. Eventually, she was weaned to room air while awake and BiPAP while sleeping on POD 49 and was discharged without home nursing on POD 52. Her NIV settings at discharge included spontaneous/timed AVAPS (average volume assured pressure support) mode: EPAP (expiratory positive airway pressure) 6 cm water, IPAP (inspiratory positive airway pressure) range 12–20 cm water, tidal volume of 8 cc/kg, rate 24, and FiO_2 0.21. Venous pCO_2 prior to discharge was in the 40–50 mmHg range.

On follow-up 3 months after the surgery, the patient has had no respiratory complications using NIV overnight and was found to have normal oximetry and capnography prompting discontinuation of NIV. Repeat fluoroscopy demonstrated improvement in the DP with normal movement of the left hemidiaphragm and minimal movement of the right hemidiaphragm.

Discussion

While DP is a well-known complication following operations for congenital heart disease with an incidence of 0.3%–12.8%, it is an uncommon and often unrecognized complication following LT operations.[3] DP can occur as a consequence of disruption of phrenic nerve integrity. The injury can occur at any level of its course; high spinal cord injuries (at cervical C1 or C2) result in diaphragmatic paralysis, whereas diaphragmatic function is partially preserved with midcervical lesions (at C3 through C5).[4] Operative trauma to the phrenic nerve may occur directly during dissection, be related to thermal injury (iced saline or cautery current), or occur during placement of central lines.[5] DP may complicate postoperative recovery following LT and prolong the hospital course.

Dysfunction of the diaphragm ranges from a partial loss of the ability to generate pressure (weakness) to a complete loss of diaphragmatic function (paralysis).[4] We preferred the term DP in this article since it is not possible to predict the course of recovery at presentation. Right DP is a relatively more frequent complication of LT compared to left DP. In a retrospective study in adults, right phrenic nerve dysfunction ranging from DP to abnormal nerve conduction were present in up to 80% of cases, and complete right diaphragm paralysis was present in 38% of adult patients following LT.[2] In another study, right diaphragm paralysis was detected in 21% of adult patients and right DP was detected in 23% of patients.[6] In a recent pediatric review of LT, 16 patients developed right diaphragm paralysis out of 151 transplantations performed (10.5%).[1] In all three studies, there was no left DP detected. To our knowledge, this is the first reported pediatric case of bilateral DP following LT.

The right phrenic nerve enters anterolateral to the aortic hiatus and is typically the nerve affected during LT procedures because of its close relationship with the suprahepatic inferior vena cava and the diaphragm, which subsequently results in unilateral right paresis. The mechanisms responsible for DP after LT include indirect injury resulting from sustained use of diathermy during liver mobilization or direct injury from the use of a suprahepatic caval clamp. It was also reported that repeat transplantation is a risk factor for development of right DP.[7] Left phrenic nerve injury was reported in a patient with situs inversus who underwent LT.[8] Given the extensive use of cautery for hemostasis and clamps in this patient, it is likely that this may have contributed to our patient's bilateral DP. We suspect that the cause of DP in our case is related to bilateral phrenic nerve damage from stretching during the procedure.

Patients with unilateral DP are usually asymptomatic and this may be discovered as an incidental radiographic finding with an elevated hemidiaphragm. Often the first sign of DP following an operative procedure is the inability of the patient to be weaned off of ventilatory support. The most characteristic sign on physical examination is respiratory

distress and paradoxical breathing.[4] Bilateral phrenic nerve paralysis usually causes severe breathlessness and often requires mechanical ventilator support. Patients frequently have considerable dyspnea at rest, when supine, or with exertion. Patients who have bilateral DP are at an increased risk for hypoventilation during sleep.[4] Consequences of bilateral DP include infections of the lower respiratory tract as well as atelectasis. Young children and infants are especially intolerant of phrenic nerve injury due to their greater chest wall compliance, underdeveloped intercostal musculature, and mediastinal hypermobility.[5] The effects of DP are much more marked in LT recipients because they tend to have significant muscle wasting and weakness of the accessory respiratory muscles. Postoperative gastrointestinal ileus and ascites exacerbate the respiratory distress.[7]

Treatment options for postoperative DP include early diaphragmatic plication or mechanical ventilation allowing the diaphragm to potentially recover on its own. Plication of the diaphragm is a procedure in which the flaccid hemidiaphragm is made taut by oversewing the membranous central tendon and the muscular components of the diaphragm. The indications and timing for this procedure are not fully defined, given that most studies are retrospective and uncontrolled. Some recommend an early diaphragm plication in order to assure timely extubation and decrease the duration of hospitalization, particularly in the setting of cardiac surgery.[5] Although diaphragmatic plication can speed up the recovery process and can enable patients to no longer require ventilator support, it is important to be aware that over time it is possible for diaphragmatic function to resolve with NIV support. Watanabe et al., in a retrospective study of 125 children with DP following cardiac surgery, showed that 84% of patients had spontaneous resolution of DP within 5–55 days.[9] A recent article reviewed bilateral DP following cardiac surgery over a 10-year period and found that conservative management may be a reasonable approach as diaphragm recovery was relatively short, less than 7 weeks in all nine patients.[10] Although this study may not be comparable to DP following LT, treatment options should be individualized and be dependent on the severity of symptoms and duration of DP. It is also important to note that return to normal diaphragm function depends on the location and severity of the nerve injury and may take months to years to recover. A retrospective analysis of 72 children with phrenic nerve injury following pediatric cardiac surgery showed that plicated and nonplicated patients regained function at a similar frequency (60% and 54.8%, respectively). Plication status, age at diagnosis, and side of paralysis did not predict failure of recovery. Bilateral DP was seen in two patients and resolved in less than 60 days.[11] In another study involving diaphragm plication after repair of congenital heart defects in children, 16 of 17 plicated patients with unilateral DP demonstrated return of diaphragmatic function within 16 months.[5] Another study in adults showed complete recovery of right phrenic nerve conduction with recovery of diaphragm function taking up to 9 months.[12] Diaphragm plication is accepted as standard of treatment for children under 12 months of age; however, there are no studies showing the benefit of diaphragm plication following DP from LT surgery and the long-term outcome is unclear.[5] Plication is unlikely to be helpful in bilateral DP.[4]

Conclusion

In summary, diaphragmatic injury following LT is often unrecognized and is typically unilateral, involving the right hemidiaphragm. In this report, we present a case of bilateral diaphragm dysfunction which we believe is the first reported case of bilateral DP following LT. Treatment for DP needs to be individualized based on the severity of symptoms and clinical impairment. This report supports the potential for spontaneous resolution of bilateral DP following LT in children with a conservative approach with NIV as a first-line treatment to allow the diaphragm to regain function. Surgical interventions such as plication should be reserved for severe cases or in patients who do not improve over time.

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Ethical approval

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Informed consent

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