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Diaphragmatic nerve palsy in young children following liver transplantation

Successful treatment by plication of the diaphragm

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Abstract Diaphragmatic paralysis was identified in four children after liver transplantation. All presented with persistent right upper lobe atelectasis, pleural effusion and recurrent respiratory infections and could not be weaned from mechanical ventilatory support. Fluoroscopy and real-time ultrasound confirmed paradoxical right diaphragmatic movements. Diaphragmatic plication was undertaken and enabled rapid and sustained weaning from respiratory support in all four cases. Vascular clamping of the suprahepatic vena cava seems to be the cause. Diaphragmatic plication allows optimal recruitment of the respiratory muscles with a favourable impact on lung mechanics and gas exchange.

Key words Liver transplantation, child, diaphragmatic nerve palsy · Diaphragma, nerve palsy, liver transplantation · Nerve palsy, diaphragma, liver transplantation · Child, diaphragmatic nerve palsy, liver transplantation

Introduction

Injury to the phrenic nerve may occur at any level of its course, from the spinal cord to the diaphragm [7, 12]. Diaphragmatic dysfunction can occur as a consequence of dissection, stretch, contusion, thermal injury or hypothermic damage due to topical nerve cooling [4, 20]. Orthotopic liver transplantation may be complicated by a degree of phrenic nerve injury owing to its close anatomical proximity to the inferior vena cava [6, 11]. Invariably, the right phrenic nerve is affected because of the close relationship with the diaphragm and suprahepatic inferior vena cava, and it is injured by a high application of the caval clamp. Nevertheless, few patients develop respiratory failure [6, 14]. In children, particularly infants, diaphragmatic paralysis is associated more commonly with postoperative problems because they lack the compensatory mechanisms that adults have to counteract the paradoxical movement of the hemidiaphragm (strong intercostal muscles, stable mediastinum, upright course of the ribs) [10, 16].

The two treatment options are mechanical ventilation until the diaphragm recovers or early surgical plication. Mechanical ventilation may be prolonged and can lead to significant complications, and recovery cannot be predicted with accuracy. Early diaphragmatic plication is the best option because it allows rapid weaning from mechanical ventilation [2, 15]. We present our experience with diaphragmatic plication in four children following liver transplantation.

Materials and methods

Case 1

A 14-month-old female underwent a left lobe transplant following previous Kasai porto-enterostomy for biliary atresia. There were dense adhesions that had to be divided. Otherwise, the operative and early postoperative course was uneventful. The child was extubated on the 3rd postoperative day but rapidly became hypoxemic and tachypnoeic and had to be re-intubated and reventilated. The next day the child underwent laparotomy for a perforation of the

ileum, which was oversewn. Five days later, the child underwent a second laparotomy for re-perforation and the distal ileum was exteriorised. A subsequent attempt to extubate the child failed, with development of right upper lobe atelectasis, pleural effusion and persistent respiratory infection. The clinical suspicion of right hemidiaphragmatic paralysis was subsequently confirmed by fluoroscopy and real-time ultrasound, and a transabdominal diaphragmatic plication was performed on the 35th postoperative day. The child was extubated the following day and there were no further respiratory complications.

Case 2

A 10-month-old female underwent whole orthotopic liver transplantation after previous Kasai porto-enterostomy for biliary atresia. Dissection was difficult due to dense adhesions and varices. Postoperative graft function was excellent. However, a repeated trial of extubation failed and she developed atelectasis of the right upper lobe, right pleural effusion and a persistent respiratory infection. Right diaphragmatic paralysis was confirmed by fluroscopy and ultrasound. Transabdominal diaphragmatic plication was performed 11 days post-transplant and the child was extubated at 48 h without sequelae.

Case 3

A 34-month-old female underwent whole orthotopic liver transplantation for haemangio-endothelioma. The operation and early postoperative course, including graft function, was satisfactory. The child failed two trials of extubation and developed right upper lobe atelectasis and a pleural effusion. Right diaphragmatic paralysis was confirmed by ultrasound and a transabdominal diaphragmatic plication was performed 3 days post-transplant. The child was extubated 24 h later without further respiratory complications.

Case 4

A 54-month-old male with Alagille's syndrome who was initially transplanted with a left lateral segment was retransplanted for acute intractable rejection at 20 days with a second left lateral segment graft. After extubation his respiratory function gradually deteriorated and he was intubated and reventilated. Right hemidiaphragmatic paralysis was identified by ultrasound and a transabdominal diaphragramatic plication was performed 11 days after the second transplant. He was extubated on return from theatre but developed right upper lobe collapse and respiratory infection that required mechanical ventilation for a further 38 days before successful weaning.

Technique of diaphragmatic plication

The previous transverse abdominal wound was reopened and the liver mobilised to allow access to the hemidiaphragm. Diaphragmatic plication was performed in successive layers using intermittent 2/0 or 3/0 Vicryl sutures beginning on the right part of the tendinous portion of the diaphragm and passing laterally. The plication was continued until the hemidiaphragm was flattened and firm to palpation.

Discussion

Right diaphragmatic injury following liver transplantation remains a relatively unrecognised complication. The incidence is unknown as many patients with incomplete palsy are able to compensate [6, 11]; however, it can produce respiratory insufficiency and persistent chest infections, particularly in young children [6, 11]. Retrospective studies reveal that phrenic nerve abnormalities ranging from abnormal nerve conduction to partial paralysis are present in up to 80 %, and complete paralysis is present in up to 30 %–40 %, of patients after liver transplantation [6, 14]. Resumption of normal nerve function depends on the severity and location of the injury and may take a year to recover in severe cases [9, 13]. The evaluation of diaphragmatic function by means of fluoroscopy and real-time ultrasound will confirm the diagnosis and help assess the severity of the injury. The diagnosis is invariably missed in patients who are ventilated using continuous positive pressure [6, 11]. The diagnosis should be excluded in any patient failing to extubate successfully despite having good gas exchange on mechanical ventilation.

The mechanisms responsible for diaphragmatic paralysis after liver transplantation include direct injury from the application of the suprahepatic caval clamp and indirect injury as a result of sustained use of diathermy during liver mobilisation [6, 11]. Retransplantation appears to be a risk factor, presumably because the caval clamp has to be placed higher and a larger cuff of diaphragm is included. Our experience underlines the importance of the level of cross clamping of the suprahepatic inferior vena cava. Diathermy use was not excessive in any of our cases. Argon coagulation is used for diaphragmatic bleeding in the majority of cases, particularly at retransplantation, and may help to avoid phrenic nerve injury. Multiple sutures in the bare area of the liver or internal jugular vein cannulation [7, 9] may injure the right phrenic nerve. Phrenic nerve injury following cardiothoracic surgery usually recovers spontaneously within 4–14 months but depends on the location and extent of the injury. The rate of regeneration of the nerve fibres is approximately 1 mm per day. Recovery of reversible phrenic nerve injury following liver transplantation should be more rapid than after thoracic operations; however, there is evidence to suggest that crush injury from a vascular clamp rarely resolves [3, 5].

The majority of patients with diaphragmatic paralysis do not develop respiratory failure, although a fall in FVC and FEV_1 of 25 % may occur [2] because the intercostal and accessory respiratory muscles help to compensate. It should be noted, however, that these muscles are inhibited during rapid eye movement sleep and some patients may develop noctural dyspnoea [8, 17]. The effects of diaphragmatic paralysis are much more marked in liver transplant recipients because they tend

to have significant muscle wasting and weakness of the accessory respiratory muscles [6, 11]. Postoperative gastrointestinal ileus and ascites exacerbate the respiratory embarrassment [1, 3, 5, 6]. Patients with unilateral phrenic nerve injury present with a restrictive ventilatory pattern with diminished VC, FVC, FEV_1, TLC and FRC [2, 8, 19]. The paradoxical movement of the diaphragm governs the degree of respiratory failure. Successful plication considerably improves these variables, expanding lung volume and abolishing the paradoxical diaphragmatic movement [5, 6, 19]. Maximal voluntary ventilation increases by as much as 55% [2, 16, 17] and there is improved blood gas exchange with increases in PaO_2 and PLCO (single-breath CO diffusing capacity) [17–19], reflecting resolution of atelectasis and improved ventilation-perfusion [1, 2].

We use the abdominal approach for diaphragmatic plication. This avoids a second wound and allows inspection of the abdomen and biopsy of the graft. The mobilisation of the graft and bowel is not usually difficult and remains our preferred approach. Others prefer a transthoracic approach to avoid entering an abdomen that may have been operated on several times and to avoid the placement of sutures that may further injure phrenic nerve branches or the right phrenic artery.

Early plication should be performed after confirming paradoxical diaphragmatic movement [21]. Further trials of extubation should not be undertaken. Clinical and physiological improvement is dramatic [3, 12, 16, 17]. Our first three cases were successfully extubated within 48 h, but the fourth had persistent sepsis associated with profound immunosuppression for persistent rejection. Failure to wean within 24–48 h of surgery is due to unsatisfactory plication or to a different cause of respiratory failure. Young children, particularly those undergoing retransplantation, appear to be at a higher risk, emphasising that care should be taken to include the smallest cuff of diaphragm possible on application of the suprahepatic caval clamp.

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