Diaphragmatic Eventration

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20.1 Introduction

In general, diaphragmatic eventration (DE) is defined as abnormal elevation of all (or a portion of) an attenuated (but otherwise intact) diaphragmatic leaf [1]. The term "eventration" was used first by Becklard in 1829 (through Petit probably described the condition in 1970). Bingham described plication of the diaphragm in 1954 [2, 3]. Based on etiopathogenesis, DE may be classified as "congenital" or "acquired" even though the clinical features and principles of management are similar for both forms.

20.2 Pathogenesis

Congenital DE is a developmental abnormality characterized by muscular hypoplasia of the diaphragm. It is often confused with congenital diaphragmatic hernia (CDH) with a hernial sac. However, unlike the uniform contour in an eventration, the margins of the normal diaphragm should be apparent in a CDH with a hernial sac. In addition, unlike CDH, pulmonary hypoplasia is uncommon. Congenital DE shows a male predominance and involves usually affects the left side of the diaphragm. It has been reported as a single entity and in association with other malformations.

Acquired DE is caused by injury to the phrenic nerve with resultant paralysis and elevation of the entire diaphragm. Hence, acquired DE is often termed "paralytic". The phrenic nerve may be damaged at birth, by trauma, or during intrathoracic surgery [4, 5]. Involvement of the phrenic nerve at birth is associated with obstetric brachial plexus injury (OBPI). The incidence of OBPI is about 1.51 per 1,000 live births in the USA [6], and reports vary from 0.38 [7] to 5.8 [8] per 1,000 live births worldwide.

Risk factors for injury include shoulder dystocia, macrosomia (defined as birth weight >4,500 g), instrument-assisted delivery, and downward traction of the fetal head [9]. The most common cause of diaphragmatic paralysis (DP) in children is injury to the phrenic nerve during thoracic surgery. Cardiac surgery involves procedures in which DP can be a complication: DP occurs in 0.3–13% of cardiac operations. The superficial course of the left phrenic nerve renders it prone to injury during cardiac surgery, although the right phrenic nerve is also occasionally damaged (possibly as a result of traction). High incidences of DP have been noted after procedures

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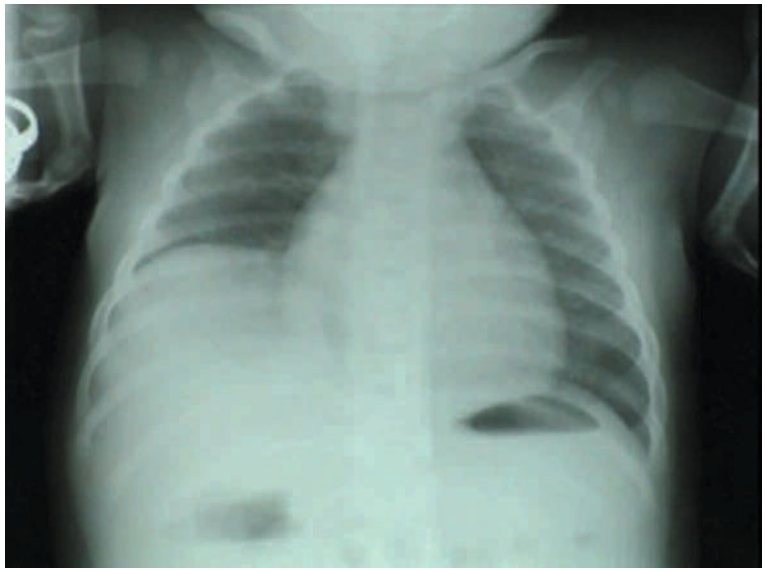

Fig. 20.1 Chest radiograph showing right diaphragmatic eventration

such as the arterial switch, Fontan procedure, and Blalock–Taussig shunt. Phrenic-nerve injuries are more likely to occur during revision procedures [10-12]. Diaphragmatic palsy may be a feature of injury to the spinal cord [13]. This may be traumatic, as a result of compression from tumors, or after infarction or transverse myelitis. Spinal-cord injury at or above the C3–C5 level is likely to involve the phrenic nerves and cause total or partial paralysis of the diaphragm. Spinal-cord injuries are caused by motor vehicle accidents (47% of cases), accidental falls (24%), firearm injuries (14%) and sports injuries (9%) [9].

20.3 Clinical Features

Clinical features range from asymptomatic to severe respiratory distress. Patients may present with pneumonia, bronchitis, bronchiectasis, atelectasis, tachypnea, dyspnea, or cyanosis. Severe hypoxia may require intubation and ventilatory support. Occasionally, patients present later in childhood with vomiting or epigastric discomfort: these symptoms are due to the change of the esophagogastric angle [14].

20.4 Diagnosis

The diagnosis is usually made on frontal and lateral radiographs of the chest, which show an elevated diaphragm with a smooth, unbroken outline (Fig. 20.1). This is a simple and non-invasive investigation, but is not sufficient for the diagnosis of DE or for differentiating DE from a hernia because it does not provide information about diaphragmatic function.

Fluoroscopy is the main dynamic investigation which can assess the motion of the diaphragm dome, yielding real-time information. The lateral projection is often employed because it allows simultaneous views of both hemidiaphragms. A paralyzed hemidiaphragm will move paradoxically cephalad during inspiration. Assessment of diaphragmatic excursion during quiet respiration may be inconclusive. The investigation is best undertaken by asking the child to inhale as fully as possible and then to exhale as fully as possible or, alternatively, asking the child to sniff [15]. This requires considerable compliance and is applicable only for older children. False-negative results are common in ventilated children be-

cause positive end-expiratory pressure tends to flatten the diaphragm. If the investigation is conclusive under these conditions, the ventilator must be disconnected temporarily.

The limited use of ultrasonography for the diagnosis of DE is because the magnitude of movement of the dome of the diaphragm cannot be determined accurately. However, ultrasound has several advantages: avoidance of ionizing radiation; portability; non-invasive nature; and minimal requirement of cooperation from the patient. Hence, several authors have investigated its use in DE. The thickness of the diaphragm at the end of expiration has been measured along with changes in thickness during inspiration using ultrasound. Changes in thickness during inspiration are proportional to diaphragm shortening in adults and infants, whereas the thickness of the diaphragm measured at the end of expiration is proportional to diaphragm strength. With diaphragmatic paralysis, the diaphragm does not thicken during inspiration [16, 17].

Other modalities include: pneumo-peritoneography; contrast peritoneography: transcutaneous electrical stimulation of the phrenic nerve: intragastric and intraesophageal manometric catheters for measuring transdiaphragmatic pressure changes; radioisotope imaging; and computed tomography. However, these are used mainly in adults because of the difficulty in compliance with infants [14, 18].

20.5 Management

Treatment of DE is dependent upon the presence or absence of symptoms. Children (and slightly older subjects) with asymptomatic DE in children may be amenable to conservative treatment. Symptomatic DE in children (congenital or acquired), especially patients in respiratory distress, requires prompt surgery but only once physical status has stabilized. Stabilization can be through: endotracheal intubation and ventilation with humidified oxygen to minimize diaphragmatic movement; a nasogastric tube to decompress the stomach; and intravenous fluids. The "gold standard" surgery is plication of the diaphragm, for which several methods have been described. This type of surgery can be undertaken by open or minimally invasive methods [10, 19].

One particular scenario is the child with phrenic-nerve palsy after cardiac surgery. The indications for plication in this case are controversial. Phrenic-nerve palsy after cardiac surgery causes significant morbidity. These children require prolonged mechanical ventilation and are more likely to develop respiratory infections. These sequelae are considerably more common in the presence of bilateral palsies. Historically, several authors argued that most phrenic palsies resolve within a few weeks of surgery (presumably because of neuropraxia). Hence, waiting 2–3 weeks after surgery before considering placation was not uncommon [20, 21-33]. However, it is now widely accepted that there is little merit in delaying plication for these children because plication avoids lengthy periods of mechanical ventilation [10].

20.5.1 Surgical Management

Plication of the diaphragm is carried out under general anesthesia. The subject is positioned supine on a warm blanket. Several approaches have been described. Open surgery allows an approach to the diaphragm through the abdomen or thorax. Several authors have argued that the right diaphragm should be approached by thoracic access through a posterolateral incision *via* the sixth space to avoid the liver [19]. An abdominal approach through a subcostal incision is the favored approach for the left diaphragm. Open bilateral plication should be done through the abdomen because this approach allows good visualization of the entire diaphragm from front to back as well as easier mobilization of abdominal contents [10]. The aim of the procedure should be to flatten the diaphragm to a physiological position. Plication of the diaphragm is carried out using non-absorbable sutures and avoiding injury to the

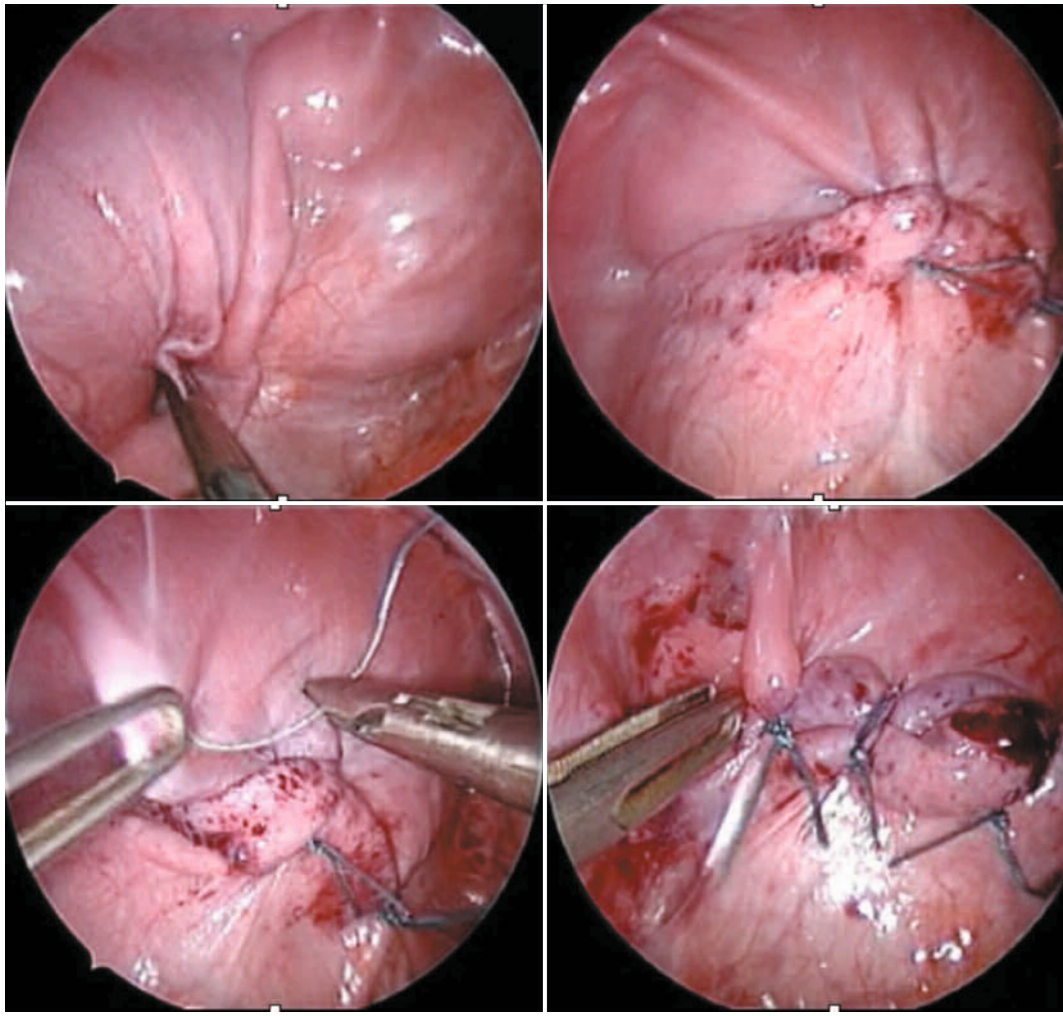

Fig. 20.2 Plication of a diaphragmatic eventration using a non-absorbable suture

phrenic nerve and structures on the blind side of the diaphragm. In cases of complete eventration, the diaphragm may be strengthened by a muscle flap or prosthetic patch [19].

In the last few years, minimally invasive methods of plication have become increasingly popular. The transthoracic approach allows plication of the diaphragm to be carried out readily. The procedure can be done in an identical fashion to open plication and the view is excellent, with magnification helping to avoid branches of the phrenic nerve. Infusion of CO_2 is used for enhancement of intrathoracic visualization. There is also the added benefit of the diaphragm being pushed down due to the infused pressure used for pneumothorax. This pressure enlarges the involved pleural space and aids the plication of the hemidiaphragm. Plicating sutures are placed and tied thoracoscopically (Fig. 20.2). Thoracoscopic plication using a knifeless Endo-GIA™ stapler has been described in adults [19, 34-36].

20.5.2 Postoperative Care

After transfer to the Intensive Care Unit, the infant is kept warm, and given maintenance of

intravenous fluids. Vital signs are monitored closely with regular analyses of blood gases and measurement of pre-ductal and post-ductal oxygenation. Ventilatory support is continued with the aim of maintaining pre-ductal partial pressure of oxygen (PO_2) at approximately 80–100 mmHg. The intrathoracic air pocket usually reabsorbs, but evidence of increasing air and fluid with mediastinal shift necessitates insertion of a chest drain. However, if intraoperative complications and pulmonary hypoplasia are absent, the patient can be extubated 24 h after surgery [19].

20.5.3 Complications

There are three main potential complications of diaphragmatic plication:

- recurrent eventration is rare except in children with neuromuscular disorders (for whom the procedure is of no benefit anyway);
- persistent respiratory failure after plication is the result of parenchymal lung disease, and results in death;
- gastrointestinal morbidity is largely due to gastroesophageal reflux (which is common), but there is a small incidence of adhesional obstruction. Failure to thrive may require nutritional support *via* gastrostomy, and many patients require fundoplication for intractable reflux [10].

20.6 Conclusions

Rapid identification and treatment of patients with symptomatic DE can improve outcome and reduce or avoid lengthy periods of mechanical ventilation. The treatment for symptomatic patients is surgery. Plication can be carried out equally well through the chest or abdomen, and the results of endoscopic surgery are equivalent to those of open surgery. The prognosis of patients in the absence of pulmonary hypoplasia is usually excellent.

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