Congenital Diaphragmatic Hernia

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19.1 Introduction

Congenital diaphragmatic hernia (CDH) is a defect of development of the pericardioperitoneal canals. In general, it occurs at 6–7 weeks of gestation. It is responsible for herniation of the abdominal organs into the thoracic cavity. CDH is delimited anteriorly by the diaphragm, sideways by the diaphragmatic pillars and posteriorly by the costal margin. CDH causes lung compression on the side of the diaphragmatic defect, precluding normal development of the lung ("lung hypoplasia") [1, 2]. CDH has an incidence of 1 case per 3,000–5,000 in live births with a male predominance. CDH is not a hereditary disease. CDH is on the left side in 86% of cases (Figs 19.1 and 19.2).

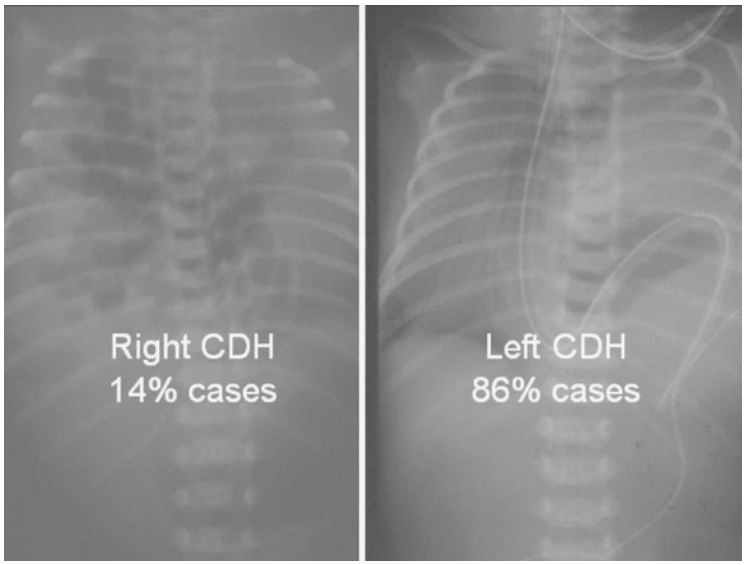

Fig. 19.1 Radiograph of the thorax showing right and left CDH

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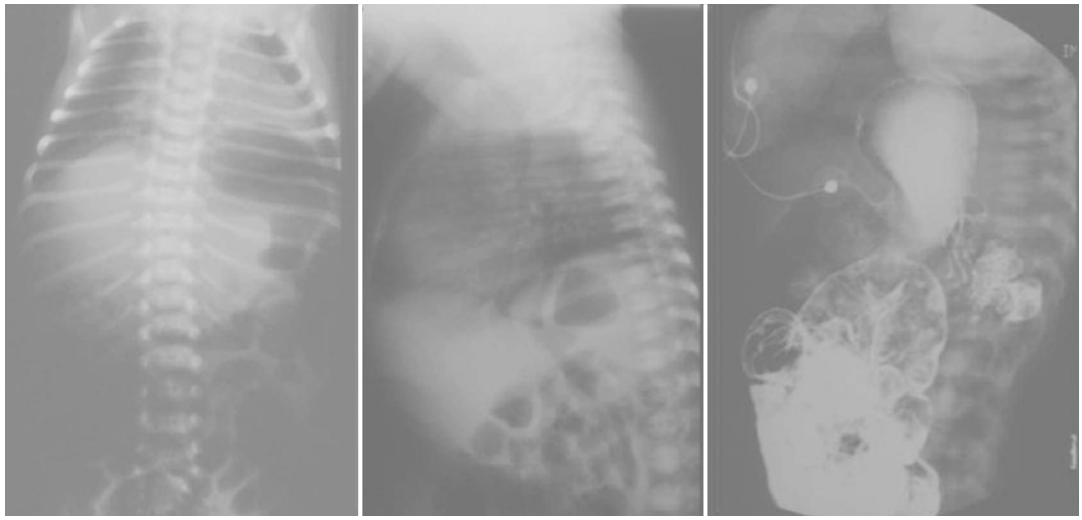

Fig. 19.2 Contrast studies showing left CDH

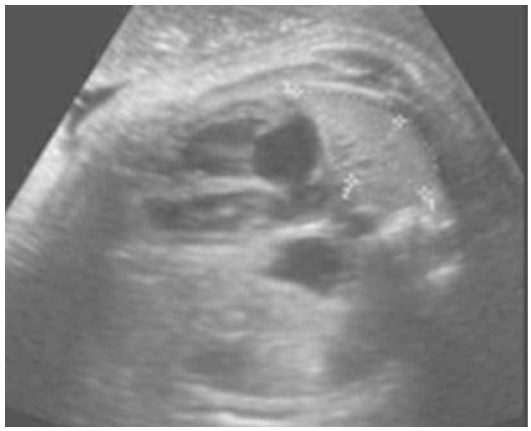

Fig. 19.3 Prenatal evaluation using ultrasound. Lung hypoplasia can be verified by calculating the lung-to-head ratio

Antenatal diagnosis is possible in the second trimester of gestation using ultrasonography, which reveals abdominal organs (gut, spleen, stomach, liver) in the thorax; evaluation of lung hypoplasia is also possible (Fig. 19.3). The thorax and heart are shifted to the right because the left side is more affected [1, 2]. At birth, due to lung hypoplasia, babies can show acute respiratory distress. Sometimes CDH is in the first months or years of life. In these patients, CDH can present with incarceration or strangulation of the herniated organs [3].

The first reports regarding the thoracoscopic approach for the treatment of CDH were in patients with late-onset CDH. Recently, several reports focusing on neonatal video-assisted surgery have been published, but the benefits and risks of thoracoscopic and laparoscopic approaches are controversial. In selected patients, the thorascopic approach allows easy reduction of herniated viscera by low-pressure insufflation of carbon dioxide (CO_2) and a complete view of the diaphragmatic defect. Surgeons based at the Pediatric Surgery Department of Strasbourg University (Strasbourg, France) have used a thoracoscopic approach for CDH treatment since 1999, initially in patients with a late diagnosis of CDH and subsequently in neonates.

In this chapter we describe the surgical procedure and the potential advantages and disadvantages of this approach.

19.2 A Thoracoscopic Approach for the Video-assisted Treatment of CDH

19.2.1 Instrumentation

The thoracoscopic approach requires the placement of two 3-mm trocars and one 5-mm

trocar in a triangular fashion. The 5-mm trocar is used to introduce a 5-mm 0° telescope; the two 3-mm trocars are used for surgical repair. In general, 3-mm trocars are adequate for this procedure. Good positioning of the trocars is essential because the operative field is very small. Monopolar cautery can be used in most cases. Closure of the defect is made using interrupted non-absorbable sutures: Ethibond 2/0 or 3/0 is preferred depending on the age of the child. Most pediatric endosurgery sets should include instruments if conversion to open thoracotomy is required as well as a patch (Gore-Tex™ or Mersilene™) [4].

19.2.2 Preoperative Management

In general, extensive preoperative preparation is not necessary, but video-assisted repair should be undertaken only in selected patients [4–7]. Each surgeon must ascertain if thoracoscopy can be undertaken. The patient may have no signs of pulmonary hypertension and may be hemodynamically stable. Absolute contraindications to a thoracoscopic approach are:

- cardiopulmonary instability;
  - severe hemodynamic instability requiring multiple pressors;
  - persistent pulmonary hypertension;
  - severe lung hypoplasia with a high partial pressure of carbon dioxide (PCO_2);
- abnormalities associated with the cardiovascular or central nervous systems;
- large defect with herniation of the stomach and liver.

Candidates for video-assisted diaphragmatic repair are:

- newborns requiring oxygen administered through the nasal route;
- newborns with minimal or decreasing parameters of mechanical ventilation;
- infants with late-onset CDH;
- delayed/recurrent CDH;
- incarcerated diaphragmatic hernia.

General anesthesia is used. Unlike in adults in whom single-lung ventilation is achieved relatively easily, the induction of general anesthesia is more difficult in infants or small children. Infiltration of port sites with a local anesthetic, epidural analgesia and intercostal nerve block can be useful in the postoperative period to control pain [8].

19.2.3 Procedure

19.2.3.1 Patient Positioning

The patient is placed in the lateral decubitus position (slightly semi-prone) just as in a thoracotomy carried out in an adult (Fig. 19.4). The surgical team comprises one surgeon and one surgical assistant in front of the patient. The surgeons are positioned at the head of the patient. The scrub nurse is at the foot of patient, at the side of the surgeons. The monitor is in front of the surgeons, near to the legs of the patient. The screen is slightly oblique towards the patient so that the surgeon, di-

Fig. 19.4 Patient positioning and placement of trocars

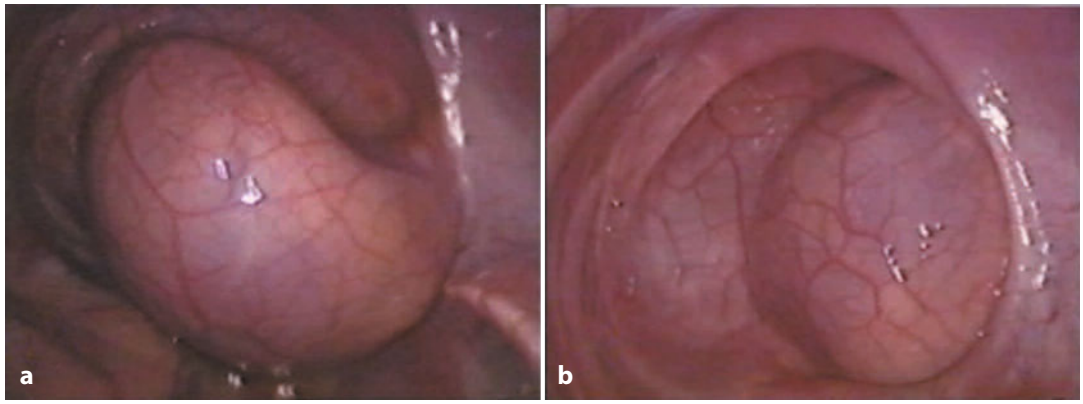

Fig. 19.5 CDH with hernial sac. Intrathoracic images show CDH with a sac before (**a**) and after (**b**) reduction by the induction of pneumothorax

aphragmatic defect and monitor are in the same direction.

19.2.3.2 Port Placement

The optic port is positioned below the tip of the scapula. The anterior working port is in the fifth intercostal space on the anterior axillary line. The posterior working port is in the fourth intercostal space between the optic port and spinal column. This last trocar must be positioned precisely because good mobility of trocars must be ensured [9–11].

19.2.3.3 Surgery

After introduction of the first trocar, intrathoracic insufflation with CO_2 is started to promote lung collapse. The intensity and duration of insufflation must be reduced in time to avoid the complications associated with abnormal intrathoracic insufflation of CO_2. Hence, low pressure (4–8 mmHg) and intermittent insufflation is usually appropriate. The first step is reduction of herniated viscera. Right-sided defects are, in general, are more difficult to repair.

If a hernial sac is present (found in approximately 20% of patients), intrathoracic insufflation (after waiting a few minutes) is sufficient to reduce the organs in the abdomen (Fig. 19.5). The sac can be excised using monopolar cautery without damaging the blood vessels and nerves of the diaphragm. Instead, the peritoneum can be entered to avoid leaving a loculated space-occupying lesion in the chest.

If a hernial sac is not present, the stomach and abdominal viscera must be reduced first. The spleen is the last organ to be repositioned. In this case, releasing posteriorly the pleuroperitoneal canal can be useful. Usually, the anterior rim of the diaphragm is evident. However, the posterior rim may not be immediately apparent and may require dissection for delineation. The defect is closed by interrupted non-absorbable sutures: Ethibond 2/0 or 3/0 is preferred depending on the age of the child (Fig. 19.6). If the defect cannot be repaired directly using sutures, prosthetic patches can be fixed by interrupted non-absorbable sutures and incorporated into adjacent tissue (Fig. 19.7). A chest drain (aspiration, 5–10 mmHg) can help lung expansion [9–11].

19.2.3.4 Limitations of the Thoracoscopic Approach

Thoracoscopy in newborns can be difficult: conversion to open thoracotomy may be indicated. There are some limitations of this approach that the surgeon must consider before surgery. These include: a restricted operative field; an unstable respiratory condition; early presentation; and inability to recognize malrotation of the abdominal viscera [9–11].

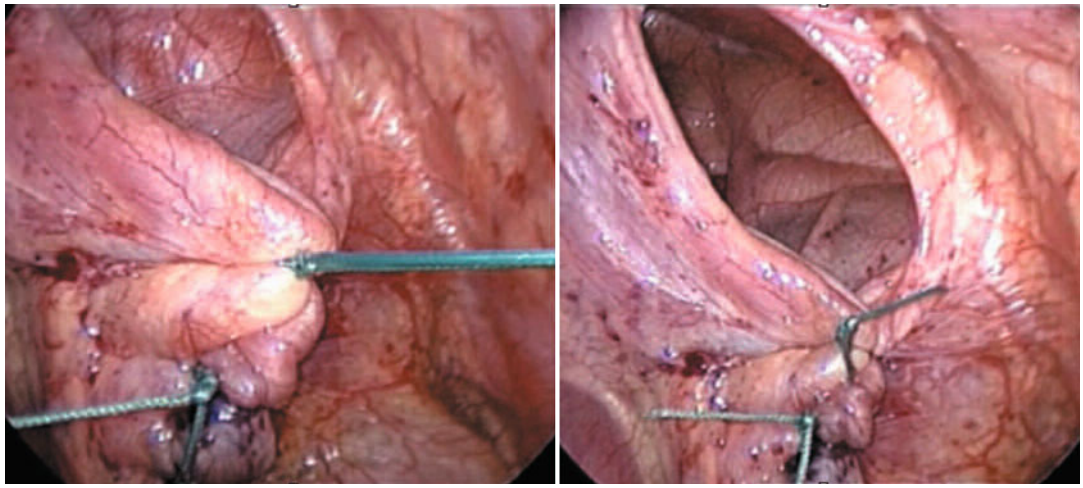

Fig. 19.6 Closure of the defect by suturing with a non-absorbable suture

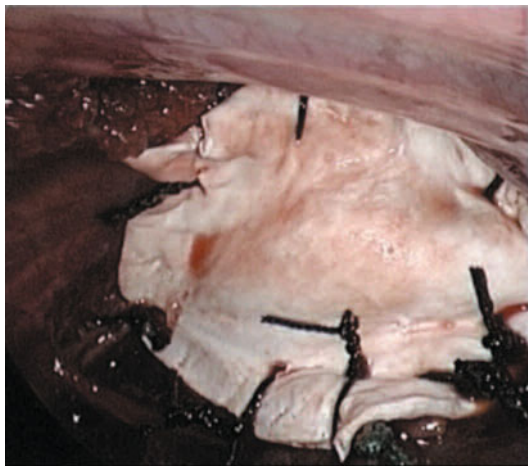

Fig. 19.7 Position of a prostethic plaque for a large CDH

19.2.4 Postoperative Management

The infant is transferred to the Intensive Care Unit. Vital signs are monitored and fluids given *via* the intravenous route. The postoperative course is dependent upon pulmonary hypoplasia and persistent pulmonary hypertension. These factors determine the ventilatory and hemodynamic situation. An epidural catheter is used to administer morphine and is removed on the second postoperative day. The chest drain is removed 24 h after surgery. Enteral nutrition is usually started on the first postoperative day.

19.2.5 Complications

Intraoperative complications related to thoracoscopic repair of a diaphragmatic hernia are usually rare. However, apart from the comments made above about the surgical procedure, the spleen must be handled very carefully, and CO_2 insufflation should be monitored very carefully because excess CO_2 can worsen respiratory status. Postoperative complications include recurrent herniation, obstruction of the small bowel, pleural effusion, chylothorax, and patch-associated problems such as infection [1, 2]. Scoliosis, asymmetry of the chest wall, and weakness in the shoulder girdle are linked with the position of non-absorbable plaques for CDH with a large defect (Fig. 19.8).

19.3 Conclusions

Video-assisted thoracoscopic surgery is a type of surgical approach for the correction of congenital CDH. During the past decade, employment of this type of surgery has increased dramatically. It is usually a simple procedure because CO_2 insufflation of the thorax allows good visualization of the defect and contributes to reduction of the viscera in the abdomen. The advantages of thoracoscopy for CDH include:

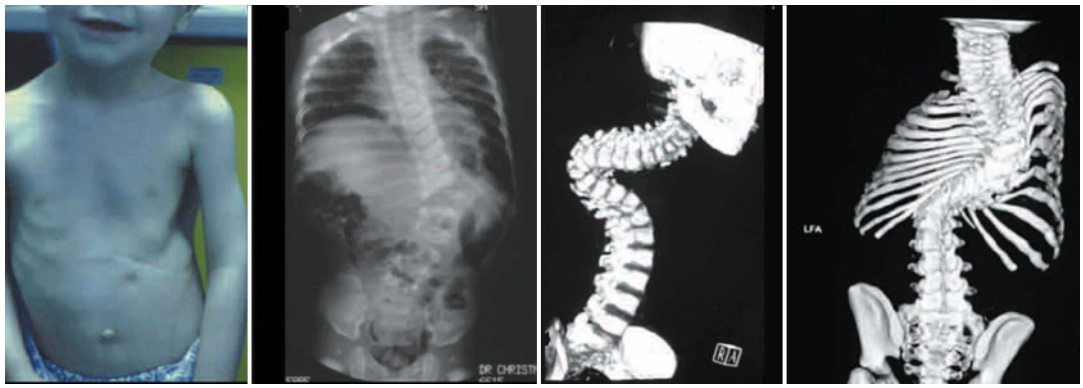

Fig. 19.8 Various images showing a patient with scoliosis after placement of a plaque for CDH

- smaller incisions in the chest that provide much better visualization and access to the anterior and posterior mediastinum;
- reduction of postoperative pain;
- more rapid postoperative recovery.

However, there are also some disadvantages:

- a relatively long learning curve for the surgeon;
- accurate selection of patients because CO_2 insufflation can worsen respiratory status, which is already unstable.

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