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Review

Advances in the surgical approach to congenital diaphragmatic hernia

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SUMMARY

Congenital diaphragmatic hernia is a birth defect that affects about one in 2500 live births. Although the overall survival has improved over the last several decades thanks to advancements in postnatal resuscitation and intensive care treatment, morbidity and mortality remain high. The surgical management of these infants is far from being standardized, and many aspects are still disputed among experts. The timing of surgical repair remains controversial and the indications for the ideal time for surgery have not been validated. The main novelty in the surgical treatment is related to the use of minimally invasive techniques, although these have been associated with intraoperative blood gas disturbances and higher recurrence rates. Herein, we report and comment on the main controversies of postnatal CDH repair in this rapidly evolving field.

1. Introduction

The surgical management of congenital diaphragmatic hernia (CDH) has improved enormously from the time when Greenwald and Steiner in 1929 wrote: “For the patient in whom the hernia makes its appearance at birth, little or nothing can be done from a surgical standpoint” [1]. Although it has been said that surgical repair of the actual defect is often the most straightforward aspect in the care of these infants [2], CDH still represents a challenging surgical condition with many unsolved dilemmas.

Surgical CDH repair can be performed by the traditional open approach (either laparotomy or thoracotomy), or by minimally invasive surgery (either laparoscopy or thoracoscopy). However, each of these techniques has its own advantages and disadvantages, and the debate among experts continues.

When the diaphragmatic defect is too big to be closed primarily, a patch repair is performed. A long-standing controversy concerns the type of patch material – whether natural or synthetic, absorbable or non-absorbable – as these have an influence on CDH recurrence. Several patches have been reported in the literature, but the ideal material has not been identified yet. Moreover, in specific circumstances muscle flaps have been employed to close the defect, again with different approaches reported by different institutions.

The purpose of this paper is to review the main aspects of postnatal CDH repair, such as timing of surgery, the different surgical approaches and patch repair, and comment on different surgical practices reported in the literature. This article focuses on the repair of the diaphragmatic defect in infants with CDH without discussing related issues such as gastroesophageal reflux or chest wall deformities.

2. Timing of surgery

The timing of surgery seems not to generate significant controversy: repair of the defect usually occurs once the neonate with CDH is physiologically stable. However, for decades the classical teaching was the opposite, as eminent surgeons such as Robert E. Gross supported the concept of “immediate repair after birth” in the belief that reduction of the abdominal contents from the chest would relieve the compression of the lungs and achieve higher survival rates [3]. Over the years, better understanding of CDH pathophysiology has shown that most infants operated on day 1 of life would go through a brief postoperative honeymoon period characterized by adequate gas exchange, but then followed by progressive respiratory deterioration, establishment of pulmonary hypertension, right to left shunting, hypoxemia, and potentially death due to respiratory failure [4]. Therefore, nowadays the infant born with CDH undergoes a period of preoperative medical cardiorespiratory stabilization, and delayed surgical repair is carried out once the overall clinical conditions are stable. This approach is supported more by common sense than by evidence. On the one hand, no study in the literature has ever shown a detrimental effect of delayed surgery awaiting stabilization. On the

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other hand, some studies, including a Cochrane review of the literature, have not demonstrated that delayed (when stabilized) as compared with immediate (within 24 h of birth) CDH surgical repair has a significant effect on survival [5,6]. A recent study from the CDH Study Group on prospectively collected data from 1385 infants with CDH showed that, when adjusted for severity of illness, delay in repair did not predict mortality or need for postoperative extracorporeal membrane oxygenation (ECMO) [7]. It is possible that the most severe cases have been operated late and that many institutions contributing to the above registry were repairing CDH only after the infant was stable, thus limiting the possibility of detecting an advantage from delayed repair. Therefore, the question of whether timing of surgery matters remains apparently unanswered. The authors' practice is in favor of delayed repair after the child has reached clinical stability.

3. Surgical approaches

The diaphragm of infants with CDH can be repaired by various surgical approaches: trans-abdominal or trans-thoracic open techniques, as well as laparoscopic or thoracoscopic minimally invasive techniques. None of these approaches has shown an absolute superiority over the others, and advantages and disadvantages of each of them are reported in Table 1.

3.1. Laparotomy

The classical surgical method for CDH repair is through a laparotomy. This entails supine positioning of the patient followed by a subcostal or upper transverse muscle-cutting incision on the same side of the hernia. The peritoneal cavity is explored and contents of the hernia are then gently reduced in the abdomen. The diaphragmatic defect is identified and repaired using interrupted simple sutures of a non-absorbable suture material.

In the case of right-sided CDH with an intrathoracic liver, division of the falciform ligament and rotation of the liver around the hepatic vein axis aids repair and avoids the risk of liver and vessel damage [8]. Reduction of the liver may be challenging [9].

A hernial sac is present in about 20% of patients and it seems to be associated with better fetal lung growth and clinical postoperative outcomes [10,11]. If present, the sac should be excised to avoid leaving a space-occupying lesion in the chest and to ensure proper healing of the defect [12].

Table 1
Theoretical advantages and disadvantages of the various surgical approaches to congenital diaphragmatic hernia (CDH) repair.

Surgery	Advantages	Disadvantages
Laparotomy	• most commonly used • good visualization • diagnosis ± correction of malrotation • inspection of reduced organs • excision of sac if present	• cosmesis • risk of intestinal adhesive obstruction • postoperative pain
Thoracotomy	• main indication for right CDH • useful in hernia recurrence	• postoperative pain • no bowel inspection • difficult reduction
Thoracoscopy	• cosmesis • pain control • less risk of adhesions	• learning curve • longer operating time • higher recurrence rate • intraoperative acidosis and hypercapnia
Laparoscopy	• cosmesis • pain control • less risk of adhesions	• longer operating time • learning curve • less widely used • difficult visualization • difficult organ reduction

A long-standing controversy of CDH surgery is whether to look for and eventually correct an associated intestinal malrotation. Most infants with CDH are considered to have a degree of intestinal rotation abnormality, ranging from non-rotation (first stage rotation anomaly) to complete malrotation of the bowel [4]. In a retrospective study, Rescorla et al. reported that 2.9% of patients with CDH not treated for malrotation presented with midgut volvulus [13]. Interestingly, the presence of intestinal malrotation has been noticed also in the nitrofen rat model of CDH [14]. It has been suggested that the abnormal positioning and fixation of the bowel in infants with CDH is due to the large communication between the abdomen and the thorax and the abnormal positioning of the surrounding structures, such as the liver [14–16].

Some surgeons favor the laparotomy approach as this allows inspection of the bowel for rotation anomalies, which if found can be corrected with Ladd's procedure.

3.2. Thoracotomy

The trans-thoracic approach was initially promoted by Everett Koop in 1952, who considered it advantageous over laparotomy given the rapidity of the technique in emptying the thoracic cavity, especially in the respiratory-distressed child and the better visualization of the diaphragm for its repair [17]. However, the following year, Gross firmly disputed the trans-thoracic approach [3], and confined its potential advantages to the same selected cases that are nowadays advocated by some authors such as diaphragmatic hernia recurrence or adult diaphragmatic hernia [18–21].

3.3. Laparoscopy

The laparoscopic approach to CDH repair found its prominent role in cases of Morgagni's hernias, with the first adult patient reported in 1992 [22], and the first pediatric case in 1995 [23]. Unlike patients with Bochdalek's hernia, those with Morgagni's hernias are clinically more stable, more likely to present at a later age and could be asymptomatic, making them the best candidates for minimally invasive procedures [4].

Given that Morgagni's hernias can present with a bilateral defect in up to 50% of cases [24], laparoscopy is more advantageous than thoracoscopy in these patients [4]. Various case series have reported the feasibility and safety of a laparoscopic-assisted repair of Morgagni's hernia, with the usually quoted advantages of minimally invasive procedures over open surgery: quicker recovery, less postoperative pain, and better cosmesis [24–27]. Interestingly, Garriboli et al. reported a high recurrence rate (42%) in their series of 12 infants who underwent laparoscopic-assisted repair of Morgagni's hernia without patch [24]. The authors concluded that laparoscopic repair of Morgagni's hernia should avoid tension of the closure, and that a prosthetic patch repair should be considered more often [24].

Recently, reports of single port laparoscopic Morgagni's hernia repair have been described in the literature [28,29].

The use of laparoscopy for the repair of Bochdalek's hernias is sporadic. In a study from the Congenital Diaphragmatic Hernia Study Group, where 4390 infants with Bochdalek's hernia from 93 international institutions underwent operative repair over a 15-year period (1995–2010), laparoscopy was performed in 0.6% of infants versus the 2.8% of thoracoscopic repairs [30].

3.4. Thoracoscopy

In 2001, Becmeur et al. reported the first use of thoracoscopy for the repair of CDH in three infants aged between 8 and 19