

2017

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Recommended Citation

Hatton, G., & Bartell, N. (2017). Management and Complications of a Late-Presenting Congenital Diaphragmatic Hernia. *Quill & Scope*, 9 (1). Retrieved from

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Management and Complications of a Late-Presenting Congenital Diaphragmatic Hernia

Gabrielle Hatton and Nicholas Bartell

A Bochdalek hernia, otherwise known as a congenital diaphragmatic hernia (CDH), is a congenital defect resulting from the failure of the posterolateral diaphragmatic folds to fuse in utero. The incidence has been reported as high as 1 in 2000, with many cases being diagnosed in utero in the United States (1). Patients not diagnosed during the prenatal period typically present with life-threatening cardiorespiratory distress soon after birth. Rarely, patients can remain undiagnosed until adulthood.

An adult with a congenital diaphragmatic hernia can present in a number of different manners ranging from asymptomatic to unstable, making the diagnosis a difficult task. An understanding of the possible presenting features, and long-term complications of congenital diaphragmatic hernias are important for the clinician presented with a patient with dyspnea or abdominal pain when other more common causes have been ruled out. Late-presenting congenital diaphragmatic hernias most frequently cause dyspnea, recurrent pulmonary infections including pneumonia, pleural effusion, chest pain, postprandial fullness, or abdominal pain. Pulmonary hypoplasia and pulmonary hypertension will be reviewed in light of the possibility that they may be the presenting features of a CDH. This then may make surgical repair, even in an asymptomatic patient, a more viable and necessary option.

PULMONARY HYPOPLASIA

The direct causes of pulmonary hypoplasia in diaphragmatic hernias are still unknown. Reductions in the size and function of the pulmonary parenchyma are not simply the result of mass effect, as there is considerable disruption in pulmonary and cardiac development before abdominal viscera have herniated. (2)

Pulmonary hypoplasia is the most feared complication of any diaphragmatic hernia in the pediatric population as it confers the highest risk of mortality. (3) However, since symptoms of pulmonary hypoplasia are generally seen early in life, an adult with an asymptomatic CDH is unlikely to have symptomatic pulmonary hypoplasia. Despite this, particular attention should be paid to the patient's lung function postoperatively, as there is potential for reductions in lung function as a result of surgical intervention.

PULMONARY HYPERTENSION

Cases of adults with congenital diaphragmatic hernias presenting with pulmonary hypertension have been reported. (4) Pulmonary hypertension would be a strong indication for immediate surgical repair in order to prevent right

heart or pulmonary failure. In these cases, postoperative follow up is even more important as effects on the pulmonary vasculature can persist even after the hernia has been repaired. These cases illustrate why diaphragmatic hernia should be included in the differential diagnosis for pulmonary hypertension.

In a case reviewed by Dobarro and colleagues, an 82 year old woman presented with an 85% oxygen saturation with no other complaints. X-ray showed a large diaphragmatic hernia, electrocardiogram revealed signs of right bundle branch block and right heart strain, and echocardiography showed increased right heart size with normal left heart parameters. The patient refused surgical intervention and was discharged on oxygen therapy. Currently there is no research linking surgical repair of CDH in asymptomatic adults to a decrease in mortality, as it is such a rare condition. However, cases such as those reviewed by Dobarro and colleagues give us a glimpse of the possible natural history of the disease; suggesting that surgical intervention in an asymptomatic patient is the proper course of action.

Pulmonary hypertension is associated with significant morbidity and mortality as it eventually can result in cor pulmonale. A number of modalities are utilized in the diagnosis of pulmonary hypertension including: chest radiographs, electrocardiogram, echocardiogram, and pulmonary function testing. A chest radiograph in pure pulmonary hypertension will show increased lung vessel markings, and may show evidence of right-sided hypertrophy of the heart. In the case of a diaphragmatic hernia, abdominal contents may also be seen in the thorax. Electrocardiogram will show evidence of right heart strain including right axis deviation, or right bundle branch block. (5) Echocardiogram may show a number of signs of right heart failure including, pressure overload, bulging of the septum into the left ventricle, and late closure of the pulmonary valve. As heart failure progresses, echocardiography will also show right-sided dilatation and loss of structural integrity. (6)

SURGICAL REPAIR

Once diagnosed, surgical repair is strongly recommended to avoid life-threatening complications such as cor pulmonale, gastrointestinal strangulation, hemorrhage, or visceral perforation into the peritoneal or thoracic cavities. Recent data have revealed more favorable outcomes in patients undergoing laparoscopic repair than with repair by more invasive laparotomy or thoracotomy. (8) Due to the rarity of this presentation, there are no long-term outcome stud-

ies for patients undergoing surgical repair of a congenital diaphragmatic hernia. The following discussion summarizes case reports and series in current literature.

PREOPERATIVE PLANNING

There is a broad spectrum of symptoms on diagnosis of a late-presenting CDH from asymptomatic to sepsis and shock. Adult patients presenting with gastrointestinal strangulation, perforation, or hemorrhage secondary to a CDH should be emergently explored by midline laparotomy. Damage control surgery (DCS) may be employed if viscera repair is prolonged or patient is hemodynamically unstable. The abdomen may be left open and the diaphragmatic defect may be repaired after adequate resuscitation.

With increased use of X ray and CT imaging, CDH are increasingly found incidentally in asymptomatic patients. Front and lateral chest X rays are recommended followed by spiral thoracic CT to confirm diagnosis. Late-presenting patients with mild or absent symptoms are often best suited for a laparoscopic, thoracoscopic, or robotic repair. Minimally invasive techniques are associated with shorter recovery time, faster return to a full diet, faster return to presurgical activity, and preferable cosmesis. (9)

Medical optimization is vital for the success of a congenital hernia repair. While early-presenting patients often have coinciding congenital defects, late-presenting patients have coinciding congenital defects in less than 10% of reported cases. (10) Even so, cardiopulmonary stability should be assessed preoperatively. Pulmonary function tests should be performed and if functional limitations are present, preoperative workup may include an echocardiogram to look for congenital heart disease or pulmonary hypertension.

THORACOSCOPIC APPROACH

Thoracoscopic repair of late-presenting CDH has been widely reported and proponents cite its increased accessibility to the classic posterolateral location of the defect and the ability for thoracic adhesiolysis as reasons for technique preference. (11, 12)

Patients should be prepared for surgery via left thoracoscopic approach. One lung ventilation should be employed using a double-lumen trachea tube and the patient should be placed in a right lateral decubitus position. The first trocar for the thoracoscope is placed at the midaxillary line, often through the seventh intercostal space. Upon examination of the thoracic cavity, second and third working trocars can be placed over the anterior and posterior axillary lines. At this point the patient may be placed in reverse Trendelenburg position. Gas pressure may be increased at this point to induce an artificial pneumothorax while carefully monitoring the patient's cardiopulmonary status. Safe reduction of hernia contents may be completed. Up to 20% of CDHs are associated with a hernia sac, which should be carefully excised. (13) Thoracoscope may be in-

serted through hernia defect to examine reduced organs for bleeding or bowel torsion. If organs cannot be confidently examined, laparoscopic port(s) may be placed for more appropriate visualization.

Repair of the diaphragm may be primary or with mesh, depending on the size of the defect. If the diaphragm edges can be approximated without tension, permanent sutures may be used to close the defect primarily. If tension-free approximation is not possible, mesh of the surgeon's preference should be used.

Disadvantages to a thoracoscopic repair include difficult visualization of reduced abdominal contents and difficulty with organ manipulation.

LAPAROSCOPIC APPROACH

Laparoscopic repair will allow better examination of reduced hernia contents if strangulation is suspected. Special consideration for intraoperative tension pneumothorax should be discussed among the surgical and anesthesia teams prior to insufflation and a chest tube should be prepared preoperatively.

Patients should be placed in supine position with general intubation anesthesia. The first trocar for the laparoscope should be placed supra- or sub-umbilical. Upon insufflation, the abdominal cavity should be inspected. Working trocars can then be placed at the surgeon's preference. Suggested port placement includes left subcostal at the midaxillary line, subxyphoid, and at the left anterior axillary line. (14) Hernia contents should be reduced with adhesiolysis as needed. The hernia sac should be excised, if present. Upon reduction of the hernia contents, pneumothorax may develop secondary to CO₂ insufflation. This may be corrected by placing a suction tube into the thorax through the hernia defect or by inserting a chest tube. Reduced hernia contents should be inspected for bleeding or bowel torsion. Repair of the diaphragm should proceed as previously described.

ROBOTIC APPROACH

Successful robotic repairs have been reported in both adults and children. (15, 16) Both transthoracic and transabdominal approaches have been successful. This approach is limited by surgeon experience and robot availability.

POSTOPERATIVE CARE

Patients with an uncomplicated operative course may be observed overnight. CXR should be monitored for pneumothorax or pleural effusion, which are among the most common postoperative complications. Activity and diet may be advanced as tolerated with consideration of bowel injury. Recurrence rates are more likely to follow large defect repair and repair with mesh as observed in pediatric follow up reports. (17)

CONCLUSIONS

A late-presenting congenital diaphragmatic hernia may cause cardiorespiratory distress, strangulation, perforation, or hemorrhage of herniated abdominal organs and thus should be surgically repaired. Minimally invasive techniques result in better patient outcomes as well improved visualization for the surgeon. Long-term studies are needed to evaluate preferred approach and hernia defect closure technique. Long-term complication and recurrence studies are also warranted. Patients undergoing repair by laparoscopy, thoracoscopy, or robotically have favorable overall outcomes.

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