



Published in final edited form as:

Anat Histol Embryol. 2016 June ; 45(3): 246–248. doi:10.1111/ahel.12200.

Bochdalek Congenital Diaphragmatic Hernia in an Adult Sheep

R. D. Williams¹, M. G. Katz^{1,*}, A. S. Fargnoli¹, A. P. Kendle¹, K. L. Mihalko², and C. R. Bridges¹

¹Sanger Heart & Vascular Institute, Carolinas HealthCare System, Charlotte, NC, USA

²Cannon Research Center, Comparative Medicine, Carolinas HealthCare System, Charlotte, NC, USA

Summary

Congenital diaphragmatic hernia (CDH) is a rare condition. The aetiology of CDH is often unclear. In our case, a hollow mass was noted on MRI. Cardiac ejection fraction was diminished (47.0%) compared to 60.5% (average of 10 other normal animals, $P < 0.05$). The final diagnosis of congenital diaphragmatic hernia (Bochdalek type) was made when the sheep underwent surgery. The hernia was right-sided and contained the abomasum. Lung biopsy demonstrated incomplete development with a low number of bronchopulmonary segments and vessels. The likely cause of this hernia was genetic malformation.

Introduction

Congenital diaphragmatic hernia (CDH) is a rare condition that occurs in <1 in 5000 live human births (Gallot et al., 2007). Incomplete embryonic fusion between the lateral and posterior components of pleuroperitoneal membranes leads to hernia development (Doyle and Lally, 2004). The aetiology of CDH is unclear, although up to 15% are due to chromosomal abnormalities (Bedoyan et al., 2004). A review of the literature produced no reports of congenital Bochdalek hernias in large animals, although another type of congenital diaphragmatic hernia associated with an anterior defect, Morgagni, has been noted in dogs (Schmiedt et al., 2009) and horses (Pauwels et al., 2007). Anatomically, there is a marked difference between the two types: Morgagni hernias occur retrosternally and are more easily diagnosed, often manifesting visually as a protrusion from the upper medial abdomen, whereas Bochdalek hernias occur posterolaterally and are much more difficult to diagnose. The pathophysiology of the two conditions also differs.

Materials and Methods

All animal care and procedures were performed in compliance with the National Institutes of Health and guidelines established by the Carolinas Medical Center Institutional Animal Care and Use Committees. A 1-year-old male Dorsett sheep weighing 43 kg with no history of trauma or previous surgery was accepted to our facility for research study. At baseline, all

*Correspondence: Tel.: 704 6140540; Fax.: 704 355-0540; michael.katz@carolinashealthcare.org.

body systems and tests were normal as assessed by physical examination, complete blood count, chemistry panel and parasites.

Results

Hemodynamic measurements obtained by magnetic resonance tomography (MRI), and Millar catheter demonstrated diminished cardiac function. Left ventricular ejection fraction (EF) was 47.0%, well below the average baseline EF measured in 10 normal sheep in our laboratory of $60.5 \pm 1\%$ ($P < 0.05$). End-systolic volume was 61.0 ml versus 36.9 ml, and end-diastolic volume was 115.0 ml versus 92.7 ml compared to normal animals (both $P < 0.05$). A non-specific hollow oblong mass approximately 12 cm long and 8 cm in diameter was identified in the right postero-inferolateral region of the thoracic cavity. Our initial diagnosis was that of a cyst deemed to most likely be benign given that there were no signs of inflammation or malignant tumour processes. One week later, the animal received gene therapy during cardiac surgery. Sedation, anaesthesia and post-operative care were administered as previously described (Pritchette et al., 2013). On examination, it was found that a portion of the abomasum (fundus and body) protruded through a congenital diaphragmatic hernia in the Bochdalek foramen of the right side, causing slight lateral displacement of the heart. The caudal lobe of the right lung was also found to be hypoplastic (Fig. 1a,b). An echocardiogram revealed no other congenital malformations of the heart. The surgeon and attending veterinarian elected not to surgically repair the hernia defect, and the animal was closed normally. Post-operatively, the animal required more than usual extended ventilation assistance. After that the animal was returned to the colony and received typical post-operative care. Accordingly, the animal's diet was supplemented with high calorie nutrition. Six weeks after surgery, the animal's weight began to decrease and the decision was made to euthanize the animal under anaesthesia via sodium pentobarbital injection.

Necropsy revealed a larger portion of the abomasum (fundus and body) and a proximal portion of the omasum in the thoracic cavity. The mediastinum was shifted to the left. Microscopic analysis of lung tissue samples revealed incomplete development of the right caudal lobe with abnormally small alveoli and decreased blood vessel density compared to the left lower lobe (Fig. 2a,b). The other organs were normal.

Discussion

This rare Bochdalek hernia, previously unreported in large animals, had significant effects on cardiac and pulmonary function. In our opinion, the underdevelopment of the pulmonary tree with a reduction in the number of lung vessels caused an increase in pulmonary vascular resistance and a predisposition for pulmonary hypertension with development of cardiac insufficiency. Abomasal protrusion through the hernia in this sheep reduced the cardiac preload resulting in a load-dependent reduction in cardiac function. The decision not to repair the hernia during the surgical procedure was made because it has been demonstrated that reduction in the hernia does not improve lung mechanics and may decrease the compliance of the chest wall in the post-operative period, which was contraindicated after cardiac surgery. Reduction in the viscera into abdominal cavity can also cause increased abdominal pressure that may be detrimental to the animal (Sakai et al., 1987). The animal

also received heparin as a normal part of the procedure, elevating the risk of abdominal bleeding. The pre-operative diagnosis of a Bochdalek hernia in adult human patients is challenging, and it is commonly misdiagnosed. Unlike infants, most adult patients are asymptomatic. The animal's history and physical examination in our case was not helpful in making the diagnosis. Only MRI scans and surgery allowed us to establish the correct diagnosis.

In summary, here we describe the incidence of cardiac insufficiency and lung hypoplasia caused by a congenital diaphragmatic hernia (Bochdalek type) in an adult sheep. Additional studies are necessary to obtain more information about the aetiology of this rare defect.

References

- Bedoyan JK, Blackwell SC, Treadwell MC, Johnson A, Klein MD. Congenital diaphragmatic hernia: associated anomalies and antenatal diagnosis. Outcome-related variables at two Detroit hospitals. *Pediatr Surg Int.* 2004; 20:170–176. [PubMed: 15064962]
- Doyle NM, Lally KP. The CDH Study Group and advances in the clinical care of the patient with congenital diaphragmatic hernia. *Semin Perinatol.* 2004; 28:174–184. [PubMed: 15283097]
- Gallot D, Boda C, Ughetto S, Perthus I, Robert-Gnansia E, Francannet C, Laurichesse-Delmas H, Jani J, Coste K, Deprest J, Labbe A, Sapin V, Lemery D. Prenatal detection and outcome of congenital diaphragmatic hernia: a French registry-based study. *Ultrasound Obstet Gynecol.* 2007; 29:276–283. [PubMed: 17177265]
- Pauwels FF, Hawkins JF, MacHarg MA, Rothenbuhler RD, Baird DK, Moulton JS. Congenital retrosternal (Morgagni) diaphragmatic hernias in three horses. *J Am Vet Med Assoc.* 2007; 231:427–432. [PubMed: 17669046]
- Pritchette LA, Carty AJ, Katz MG, Fagnoli AS, Kirsch J, Bridges CR. Swallow syncope after cardiac surgery in a sheep. *Vet Surg.* 2013; 42:898–899. [PubMed: 24033841]
- Sakai H, Tamura M, Hosokawa Y, Bryan AC, Barker GA, Bohn DJ. Effect of surgical repair on respiratory mechanics in congenital diaphragmatic hernia. *J Pediatr.* 1987; 111:432–438. [PubMed: 3625415]
- Schmiedt CW, Washabaugh KF, Rao DB, Stepien RL. Chylothorax associated with a congenital peritoneopericardial diaphragmatic hernia in a dog. *J Am Anim Hosp Assoc.* 2009; 45:134–137. [PubMed: 19411649]

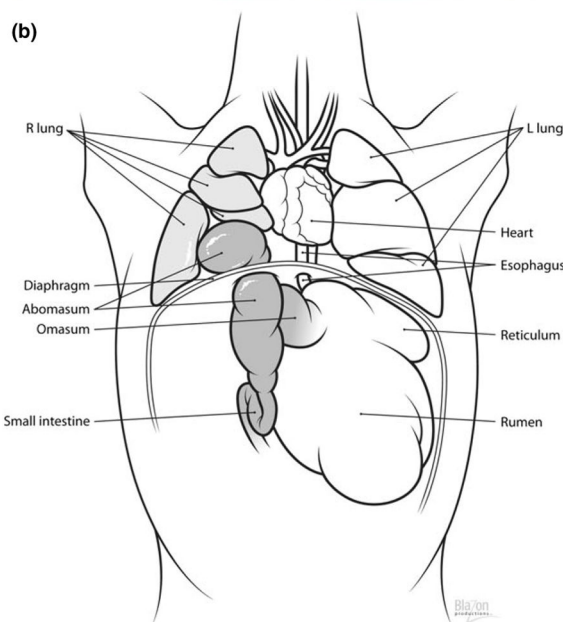
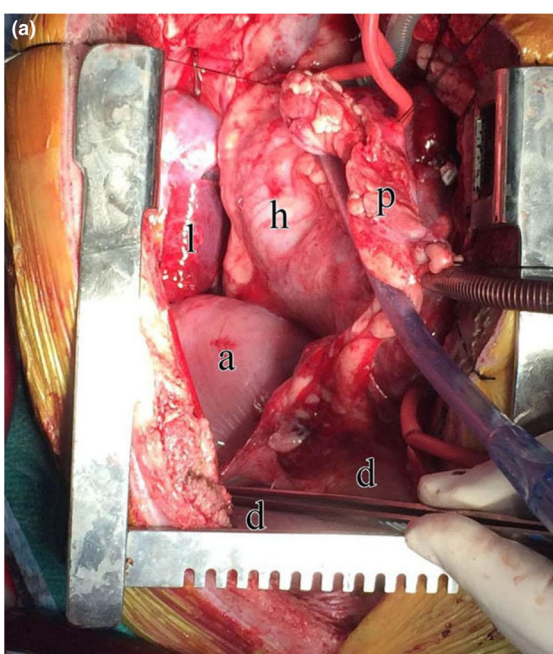


Fig. 1.

(a) Intra-operative view of thorax through median sternotomy during CPB. l – lung; h – heart; p – pericardium; a – abomasum; d – diaphragm. (b) Illustration of thoracic and gastrointestinal anatomy at the time of surgery.

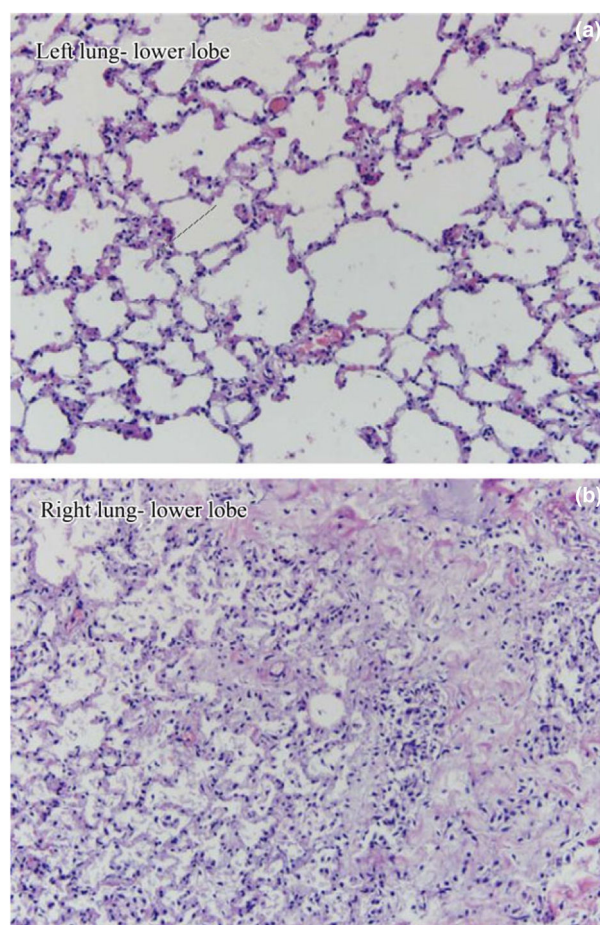


Fig. 2.

(a) Haematoxylin and eosin stained lung tissue from the caudal lobe of the left lung. (b) The hypoplastic caudal lobe of the right lung: incomplete development of the right caudal lobe with abnormally small alveoli and low blood vessel density.