Surgical Conditions of the Diaphragm: Posterior Diaphragmatic Hernias in Infants

Priscilla P.L. Chiu, MD, PhD, Jacob C. Langer, MD*

KEYWORDS

- Congenital diaphragmatic hernia
- Extracorporeal membrane oxygenation
- Gentle ventilation • Pulmonary hypoplasia
- Pulmonary hypertension • Morbidity

Congenital diaphragmatic hernia (CDH) is a defect of the diaphragm associated with herniation of abdominal viscera into the chest cavity. Related conditions of the diaphragm that mimic CDH but are not associated with diaphragmatic defects include diaphragmatic eventration and phrenic nerve palsy. These two conditions result in a mechanically impaired "high-riding" diaphragm without visceral herniation. The locations of the defect in CDH include posterolateral defect (foramen of Bochdalek), anterior/retrosternal defect (foramen of Morgagni), crural defect (paraesophageal hernia), and diaphragmatic agenesis (**Fig. 1**). Of these, Bochdalek CDH and agenesis are the most common and are associated with the greatest complexity and mortality risk.[1] In this article, we focus on the diagnosis and management of these two related entities.

EMBRYOLOGY

The diaphragm starts to form during the fourth week of embryological development from four components of mesodermal structures. Lateral extension of the septum transversum from the midline joining to the lateral margins of the pleuroperitoneal fold results in the formation of the membranous (central tendon) and muscular diaphragm (**Fig. 2**). The esophageal mesentery extends into the pleuroperitoneal fold to form the crural and dorsal portions of the diaphragm. Abdominal wall-derived mesenchyme migrates inward to fuse with the other components. Finally, lung expansion is thought to promote the fusion of all layers, resulting in a single fibromuscular tissue that physically separates the thoracic and abdominal cavities by the end of the 12th week.[2]

Diaphragmatic defects are thought to result from the failure of one or more of these layers to fuse with each other. In the case of Bochdalek hernias, abnormal development of the pleuroperitoneal fold (muscular diaphragm) is thought to result in the typical posterolateral defect as demonstrated through animal models of CDH.[3] Because the right pleuroperitoneal fold is thought to close first, left-sided Bochdalek hernias occur more frequently than right-sided CDH. The most severe CDH cases include complete diaphragmatic agenesis (ie, the complete absence of a muscular diaphragm) and bilateral CDH. The embryological defects associated with the development of anterior or foramen of Morgagni hernias

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Division of Pediatric General and Thoracic Surgery, The Hospital for Sick Children, 555 University Avenue, Toronto, ON, Canada M5G 1X8
* Corresponding author.
E-mail address: jacob.langer@sickkids.ca (J.C. Langer).

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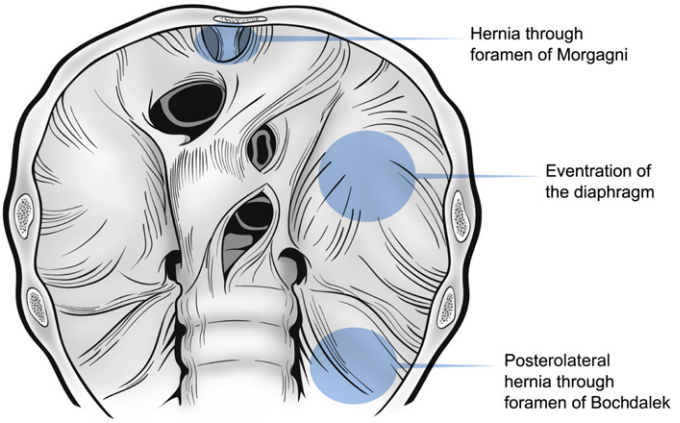

Fig. 1. Types of CDH.

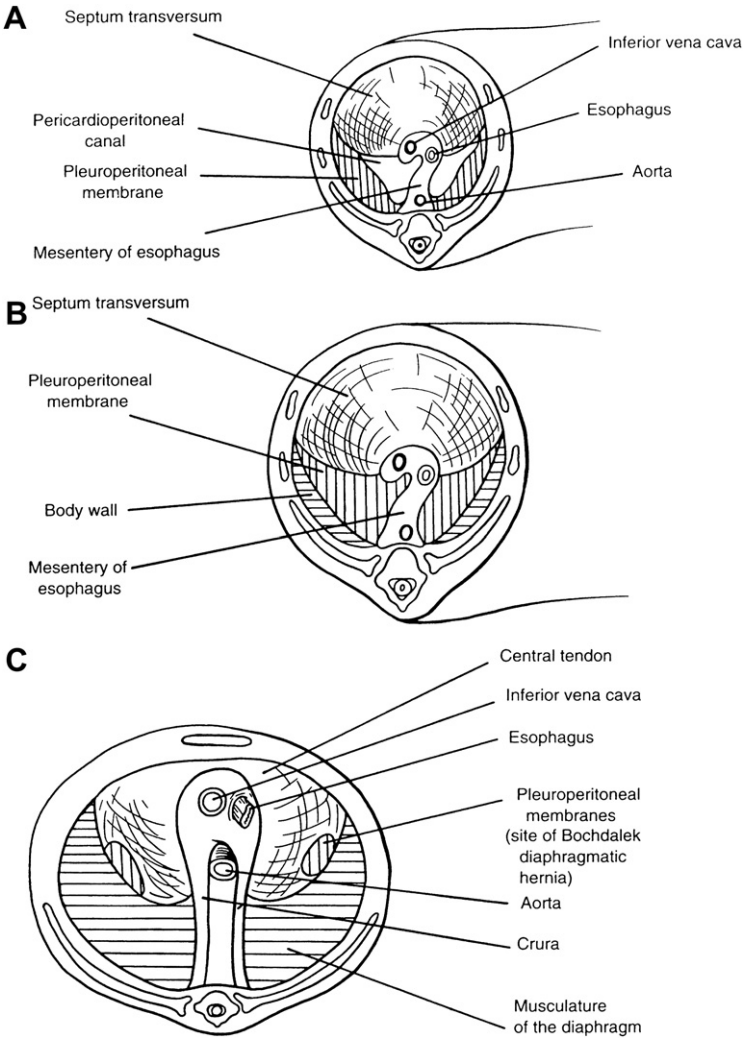

Fig. 2. Embryology of the diaphragm. (*From* Langer JC. Normal fetal development. In: Oldham KT, Colombani PM, Foglia RP, editors. Surgery of infants and children. Philadelphia: Lippincott-Raven Publishers; 1997. p. 44; with permission.)