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Mini-symposium: Chest Wall Disease

Pulmonary Complications of Abdominal Wall Defects

Howard B. Panitch *

Professor of Pediatrics, Perelman School of Medicine at The University of Pennsylvania, Division of Pulmonary Medicine, The Children's Hospital of Philadelphia

EDUCATIONAL AIMS

1) To understand the contribution of the ventral abdominal wall to inspiration and exhalation
2) To recognize the pulmonary complications of giant abdominal wall defects in neonates
3) To recognize the influence of the abdominal contents and the ventral abdominal wall on formation of the thoracic cage

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SUMMARY

The abdominal wall is an integral component of the chest wall. Defects in the ventral abdominal wall alter respiratory mechanics and can impair diaphragm function. Congenital abdominal wall defects also are associated with abnormalities in lung growth and development that lead to pulmonary hypoplasia, pulmonary hypertension, and alterations in thoracic cage formation. Although infants with ventral abdominal wall defects can experience life-threatening pulmonary complications, older children typically experience a more benign respiratory course. Studies of lung and chest wall function in older children and adolescents with congenital abdominal wall defects are few; such investigations could provide strategies for improved respiratory performance, avoidance of respiratory morbidity, and enhanced exercise ability for these children.

INTRODUCTION

The abdominal wall, rib cage and intercostal muscles comprise the chest wall. The back, lower rib cage, pelvis and iliac crests constrain movement of the lateral and posterior abdomen, so that the ventral abdominal wall is the only freely moving part of the abdominal compartment. Its motion in relationship to that of the thorax lends important insight into the mechanics of the chest wall and respiratory pump. Alterations in the characteristics of the abdominal wall affect overall respiratory pump function. In addition, congenital malformations of the ventral abdominal wall can be associated with pulmonary hypoplasia and neonatal respiratory distress. This review will focus on select congenital lesions that alter the properties of the ventral abdominal wall and thus respiratory pump function.

EMBRYOLOGY OF THE VENTRAL ABDOMINAL WALL

During the third and fourth weeks of gestation, the embryo undergoes gastrulation, becoming a trilaminar structure including ectoderm, mesoderm and endoderm. At the same time, the head and tail of the embryo bend ventrally towards each other and cause the embryo to assume the fetal position [1]. A second body folding occurs from the sides towards the middle, involving structures called the lateral body folds. The lateral body folds form from an area of the mesoderm called the lateral plate mesoderm and include the overlying ectoderm [1]. They move ventrally and fuse in the midline by the end of the fourth week of gestation. The processes that lead to the ventral movement of the lateral body folds and their fusion with each other in the midline are poorly understood, but likely involve some combination of cell proliferation, cell migration, production of extracellular matrix materials,

* 3501 Civic Center Boulevard, Philadelphia, PA 19104. Tel.: +215 590 3749; fax: +215 590 3500.
E-mail address: panitch@email.chop.edu.

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apoptosis and specialized cell to cell contacts [1–3]. Several genes have been implicated in the process of ventral abdominal wall formation [4,5], but it is likely that additional genes are responsible for proper formation of the ventral body wall. It has been hypothesized that defects in the movement of one or both of the lateral body folds to meet their contralateral counterpart result in such defects as ectopia cordis, gastroschisis, and bladder exstrophy [3,6].

The midgut begins to grow rapidly during the 6th week of gestation, resulting in a physiologic herniation of the intestines through the umbilical ring. The midgut then rotates and returns to the abdominal compartment by the 10th week of gestation. If the intestine fails to return to the abdominal compartment, a small hernia into the umbilicus can occur, resulting in a small omphalocele, with minimal widening of the umbilical ring [7]. The embryological events that cause large omphaloceles are unclear: some authors propose that an abnormality in body folding in the cephalic region results in an epigastric omphalocele as seen in pentalogy of Cantrell, while a caudal folding abnormality causes a low or hypogastric omphalocele often in association with bladder or cloacal exstrophy, and a lateral body wall folding problem results in a mid-abdominal defect [5,7]. Others, however, point to the fact that the organs within an omphalocele are covered by a sac that includes amnion along with Wharton's jelly and peritoneum, so that an omphalocele does not represent a body closure defect and some other mechanism for its creation must exist [3].

Even when no defect in body wall closure exists, additional abnormalities in ventral abdominal wall formation can take place. Between 6 and 10 weeks of gestation, muscles of the ventral abdominal wall form. Absence or marked diminution of abdominal wall musculature can occur, often in association with urinary tract abnormalities that lead to obstruction of the bladder and cryptorchidism in males. This triad of abnormalities, known as prune belly syndrome, triad syndrome or Eagle Barrett syndrome [8], has been ascribed to two possible mechanisms: the first involves a defect in intermediate and lateral plate mesoderm resulting in maldevelopment of both abdominal wall and urinary tract musculature [9]. The second mechanism invokes blockage of egress of urine from the bladder *in utero*, which then results in marked distension of the bladder, ureter and kidneys. The distended bladder compresses the abdominal wall and causes atrophy of the muscles, either by direct pressure or by interruption of blood flow [8].

PHYSIOLOGICAL CONTRIBUTION OF THE ABDOMINAL WALL TO BREATHING

The abdominal wall plays important roles during inspiration, exhalation and airway clearance via coughing. During quiet breathing in an adult, the diaphragm runs parallel to the rib cage in a zone of apposition that includes approximately ¼ to 1/3 of the rib cage [10]. Thus, abdominal contents that are not compressible occupy a substantial portion of the lower rib cage. By virtue of this arrangement, contraction of the diaphragm not only lowers intrathoracic pressure, but it also increases intraabdominal pressure through the area of apposition (Figure 1). The incompressible abdominal viscera act as a fulcrum, resulting in expansion of the lower rib cage. Notably, experimental removal of abdominal viscera from the abdominal cavity causes the costal fibers of the rib cage to contract rather than expand the lower rib cage [11]. Lower rib cage motion in infants with lesions like giant omphalocele, in which the liver and other abdominal organs are displaced from the upper abdomen, has not been quantitatively studied to determine if a similar paradoxical motion of the lower rib cage exists during unassisted breathing and before reduction of viscera back into the abdominal cavity. Because the ventral abdominal wall has freedom to move, it is normally displaced outwards along with the lower rib cage. In contrast, in several patients with giant omphalocele, diaphragmatic contraction resulted in a cephalad rotation of the omphalocele rather than an outward movement of the abdominal wall [12], perhaps reflecting abnormal lower rib cage movement.

Compliance of the abdominal wall is an important factor in the determination of diaphragmatic motion during inspiration. For a given degree of neural activation of the diaphragm, the distance its dome will descend will depend upon the resistance to fiber shortening imposed by the stiffness of the abdominal wall and rib cage [10]. A poorly compliant abdominal compartment will limit diaphragm descent, and a non-compliant abdominal wall will restrict lower rib cage expansion (much like what happens after a large meal). On the other hand, a highly compliant abdominal wall, as seen in patients with prune belly syndrome, can lead to alterations in rib cage-abdominal wall relationships and diaphragmatic function in the upright position that resolve when subjects are supine [13]. Here, Ewig and coworkers postulated that the highly compliant abdominal wall led to loss of the fulcrum effect of abdominal contents on the lower rib cage, thereby limiting lower rib cage expansion and allowing the diaphragm muscle fibers to shorten excessively. These mechanical disadvantages in turn resulted in functional diaphragmatic weakness, a need for recruitment of accessory muscles of inspiration, and ultimately abdominal paradox (inward motion of the abdominal wall during inspiration) [13]. These findings all disappeared when the same subjects were studied in a supine position, and gravity caused abdominal viscera to exert cephalad pressure on the diaphragm thereby improving its length-tension relationships and increasing the area of apposition.

Under normal circumstances, the abdominal muscles are typically considered to be accessory muscles of exhalation.

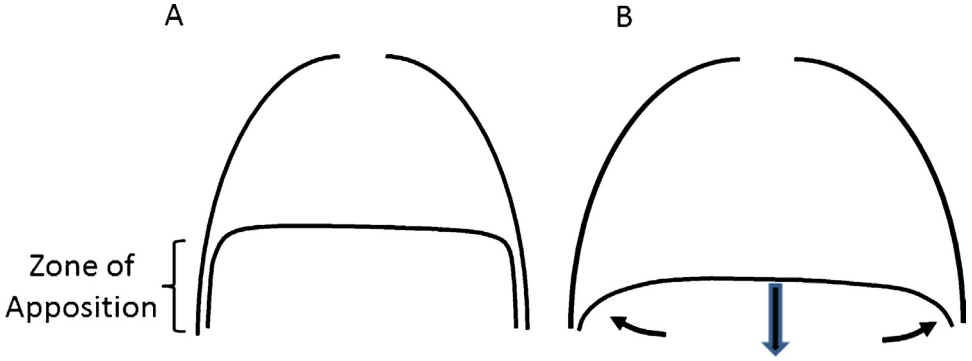

Figure 1. A) Diaphragm and chest wall at end-expiration. The zone of apposition is demarcated by the bracket; B) At end-inspiration, the diaphragm has descended, increasing pressure in the abdominal compartment. In turn, the abdominal contents act as a fulcrum to help elevate the lower rib cage.