

# Kyphoscoliosis and hiatal hernia: anatomical analysis stimulates new clinical interest

Ernest F. Talarico, Jr. and Alexander C. Vlahu

*Anatomy & Cell Biology, Indiana University School of Medicine-Northwest, Gary, Indiana, USA*

Prior research in our laboratories (Talarico and Vlahu, 2015) qualitatively and quantitatively documented the relationship between kyphoscoliosis and hiatal hernia (HH). Briefly, a 97-year-old female anatomical donor who died secondary to aspiration pneumonia was radiographed using plain x-ray, high-resolution computed tomography (CT) and magnetic resonance imaging (MRI), and later dissected. In life, she suffered from chronic back pain, arthritis, and gastroesophageal reflux disease (GERD), as well as long-term complaints of abdominal pain and dyspnea (or feelings of pressure on her chest). The underlying cause of these complaints was never discovered. She was diagnosed with kyphosis and scoliosis, but because of her advanced age she was never treated. Dissection and radiography determined that this patient suffered from severe kyphoscoliosis with Cobb Angels of 45° (thoracic dextroscoliosis), 34° (lumbar levoscoliosis), 78° (thoracic kyphosis), respectively. Furthermore, all vertebrae were rotated to the left of mid-line; and ranging from 4° at T1 to 29° at L5. The greatest degrees of rotation were found in vertebrae T12 and L5, and were determined to be 18° and 29°, respectively. The patient's heart was enlarged and displaced to the left of mid-line, with its apex touching the thoracic wall, and the aorta was enlarged and shifted 7.3 cm to the right of mid-line and along the anterior and right-lateral border of the scoliotic thoracic curvature. The left lung was compressed by the shifted heart. A giant HH was observed with the stomach (cardiac, fundus, body and pylorus) found to be

inside the mediastinum, and the esophageal hiatus of the diaphragm was dilated, with the most proximal part of the duodenum near the pyloric sphincter seen within the hiatus. Intra-abdominal volume (IAV) was 17% to 53% less than that of studied females that were unaffected from scoliosis, kyphosis and HH. We suggest that reduced IAV caused by kyphoscoliosis may contribute to the development and progression of paraesophageal hernias in patients with laxity of the diaphragmatic hiatal musculature, and that clinicians need to be aware of this association in the evaluation and work-up of patients that present with gastrointestinal (GI) complaints and spinal deformities.

Recently, we examined a more severe case of HH in a 66-year-old female cadaver with neoplasm of the brain diagnosed as the cause of death. There was no evidence of metastasis to the spinal column or other organs. CT and MRI studies (Fig. 1) revealed a Type IV, giant HH with the stomach, proximal two-thirds of the duodenum and the head of the pancreas, pushed superiorly into the mediastinum and posterior to the heart. Kyphoscoliosis was measured with Cobb Angels of 61° (thoracic dextroscoliosis), 39° (lumbar levoscoliosis), 37° (thoracic kyphosis), respectively. Vertebrae were rotated to the left and right of mid-line in the thoracic and lumbar regions of the spinal column. The degree of rotation ranged from 13° left at T1 to 28° left at T3; 19° right at T4 to 31° right at T7 and 26° right at T12; 5° right at L1; and 17° left L2 to 23° left at L5. IAV was determined to be 63% less than

**Corresponding author:** Ernest F. Talarico, Jr. Anatomy & Cell Biology, Indiana University School of Medicine-Northwest, Dunes Medical Professional Building, Room 3028A, 3400 Broadway, Gary, Indiana 46408-1197, USA. Phone: 219-981-4356; Fax: 219-980-6566. E-mail: etalaric@iun.edu

**Abbreviations:** Computed tomography (CT); Ehlers-Danlos Syndrome (EDS); Gastroesophageal reflux disease (GERD); Hiatal hernia (HH); Intra-abdominal Volume (IAV); Magnetic reso-

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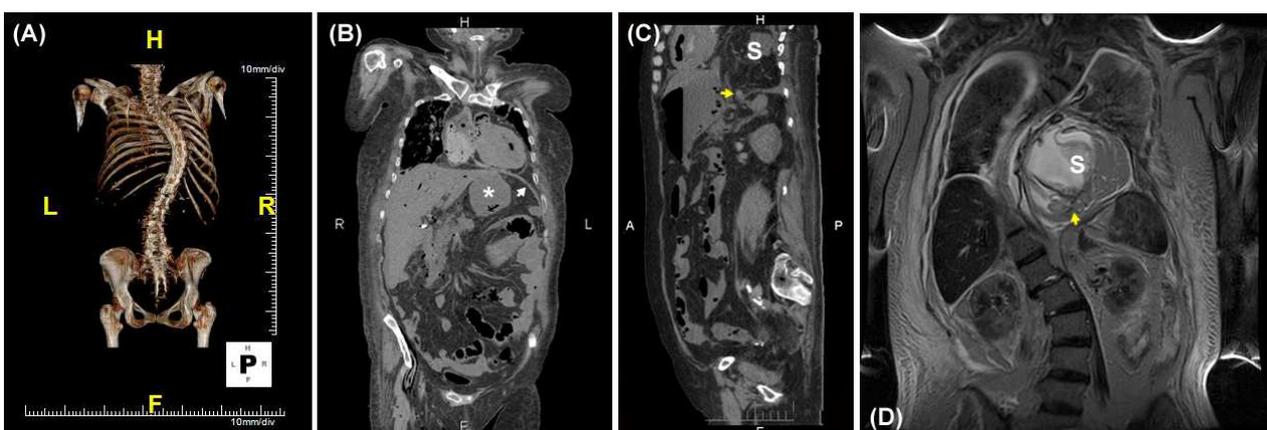
that of the same unaffected cadavers in the initial study.

Kyphoscoliosis is defined as a deformity of the vertebral column represented by a combination of a posterior-convex curvature (i.e., kyphosis) in the spine from the sagittal view and a left or right, or left and right (i.e., side-to-side) lateral curvature (i.e., scoliosis) in the spine from the posterior view (Ferreira-Alves et al., 1995; Schuchert, et al., 2011). In most cases, a small degree of curvature does not cause any medical problems. However, larger curves can cause postural imbalance and lead to muscle fatigue and pain. More severe scoliosis can interfere with breathing and lead to arthritis of the spine (i.e., spondylosis). Still further, kyphoscoliosis is associated with restrictive impairment of pulmonary function, chronic pain and decreased vitality (Mitiek and Andrade, 2010; Naoum et al., 2011). It is most commonly identified in women (Schuchert et al., 2011). The prevalence of kyphoscoliosis increases with age, ranging from 2% of the adolescent population to as high as 15% among the elderly population while also being more prevalent in women (Lonstein, 2006). Thus, it is reasonable that with the increase in World population aging, particularly in Western countries, and where the aging population is comprised of more women than men, that the frequency (or incidence) of kyphoscoliosis would increase.

A HH can be defined as the protrusion of an organ, typically the stomach, through the esophageal hiatus in the diaphragm. Risk factors for HH include age over 50-years, obesity, and smoking. HH is classified into 4 types (Mitiek and Andrade, 2010; Landreneau et al., 2005): Type I – sliding hernia; Type II – paraesophageal hernia; Type III – mixed sliding and paraesophageal hernia; and Type IV – herniation of additional organ (i.e., co-

lon, omentum, pancreas, spleen). Literature survey shows that there is no uniform definition of giant HH: some authors define it as a herniation of more than 30% of the stomach, whereas others describe it as a herniation of more than 50% (Talarico and Vlahu, 2015). Thus, any HH with more than 50% of the stomach contained in the thorax should be considered a giant HH. Although the size of a HH bears a small relationship to symptoms (Br Med J, 1976), larger HHs have their own particular problems in addition to the same mechanical problems of hernias found elsewhere in the body, such as gastric ulcers within the herniated loculus, bleeding and carcinomas (MacArthur and Wright, 2005; Holt, 1968; Windsor and Collis, 1967). Large HHs are associated with significant morbidity and mortality (Naoum et al., 2011; Sihvo et al., 2009; Skinner and Belsey, 1967). Life-threatening complications can include mechanical obstruction, incarceration, and compression of the left atrium and pulmonary veins (Mitiek and Andrade, 2010; Naoum et al., 2011). Symptoms usually reported by patients include heartburn (i.e., GERD), dysphagia, postprandial pain, vomiting, dyspnea and decreased exercise capacity (Naoum et al., 2011; Mitiek and Andrade, 2010; Pierre et al., 2007; Gordon et al., 2004). Even further, 17% of patients with giant HH, when defined as greater than 75% of stomach herniated, treated medically exhibit progressive symptoms without gastric strangulation, and 4% of patients die from aspiration pneumonia (Allen et al., 1993; Mitiek and Andrade, 2010).

Like kyphoscoliosis, HHs are also more common in Western countries. The frequency of HH increases with age, from 10% in patients younger than 40 years to 70% in patients older than 70 years (Loffeld and Vand Der Putten, 2002; Gordon



**Fig. 1.** Medical imaging of a 66-year-old female cadaver with kyphoscoliosis and giant hiatal hernia. **(A)** High-resolution CT scans were graphically analyzed using TeraRecon<sup>®</sup> software generating high-resolution, 3-D skeletal reconstructions in the posterior view (shown here) showing dramatic scoliosis. **(B)** High-resolution CT in the coronal view showing left-displaced heart in the thoracic cavity; diaphragm (white arrow) and dilated colon with fecal matter (asterisk). **(C)** Sagittal CT scan of the patient demonstrating the stomach (S) and part of pancreas (yellow arrow) superior to the diaphragm. **(D)** High-resolution, T-2 weighted MRI in coronal view of the thorax (posterior to the heart) and superior abdomen showing the entire stomach (S) above the diaphragm and part of the pancreas (yellow arrow). Thoracic dextroscoliosis and lumbar levoscoliosis are also observed.

et al., 2004). Burkitt et al. suggest that the Western, fiber-depleted diet leads to a state of chronic constipation and straining during bowel movement, which could explain the higher incidence of this condition in Western countries. Paraesophageal hernias generally tend to enlarge with time, and sometimes the entire stomach is found within the chest. The risk of these hernias becoming incarcerated, leading to strangulation or perforation, is approximately 5%. This complication is potentially lethal, and surgical intervention is necessary. Because of the high mortality associated with this condition, elective repair often is advised wherever a paraesophageal hernia is found (Sihvo et al., 2009; Larusson et al., 2009).

The recent data and the historical information above suggest that anatomists and physicians alike have underappreciated the relationship between the gut tube and the vertebral column. Comte was the first to describe a possible association of HH with kyphoscoliosis (Comte, 1953). Gavala and Matejic hypothesized that axial deviation of the spine may cause distortion of the hiatal sling leading to the development of HH (Gavala and Zarabini, 1961; Matejic, 1967). Other suggested risk factors include decreased IAV and increased intra-abdominal pressures seen in patients with kyphoscoliosis (Schuchert et al., 2011). Still, a more recent study has documented improvement in reflux gastroesophagitis in patients with sliding HHs following correction of spinal thoracolumbar kyphoscoliosis; suggesting a link between primary spinal deformities and HHs (Yang et al., 2011). Critically, the data presented by Talarico and Vlahu (Talarico and Vlahu, 2015) and this more recent cadaveric examination *add measurable evidence* to support the hypothesis that decreased IAV associated with kyphoscoliosis can lead to HH.

The area of the esophageal hiatus of the diaphragm is a dynamic anatomical structure. The hiatus moves superior and inferior, as well as anterior and posterior, with each ventilatory cycle. It likely varies in size and shape during respiration and lateral flexion of the vertebral column (Moore et al., 2014). In addition, the stomach contracts approximately 3 times per minute, and the smooth muscle of the esophagus develops peristaltic waves that result in longitudinal and radial movements that are transmitted through the gastroesophageal junction. Anatomically, the crura of the diaphragm are musculotendinous bands that arise from the anterior surfaces of the L1-L3 vertebrae, IV discs, and the anterior longitudinal ligament. The right crus is larger and longer than the left crus, and splits into anterior and posterior bands forming the esophageal hiatus. Kyphoscoliosis by stretching the diaphragm may result in a loss of normal curvature and elasticity of the diaphragm leading to distortion, or widening, of the esophageal hiatus and reducing resistance of the gas-

troesophageal junction into the thorax. Decreased IAV in patients with kyphoscoliosis may increase pressure on viscera and associated mesentery, allowing displacement of the gastroesophageal junction and formation of a HH. Intra-abdominal pressure becomes raised above normal, either intermittently, due to exertion or straining (i.e. Valsalva maneuver; also increased in instances of constipation), or chronically, due to obesity or external compression, and as a result the junction is pushed superiorly and may change in shape and size.

The proposed mechanism for the development of HHs is similar to the mechanism for inguinal and femoral hernias. Chronic or intermittently increased intra-abdominal pressure and muscle weakness result in the intestine herniating through the deep inguinal ring and into the inguinal canal, or an indirect inguinal hernia. When the abdominal wall muscles are weak, as in the case of a direct inguinal hernia, increased pressure can result in a hernia that protrudes directly through the anterior abdominal wall within Hasselbach's triangle. Further, and more common in females, increased intra-abdominal pressure can cause the gut to protrude through the femoral canal inferior to the inguinal ligament, and below and lateral to the pubic tubercle. Additional support evidence comes from a study that showed evidence suggesting that the causative relationship between inguinal hernias found on clinical examination and HHs found on endoscopy in the same patients was increased intra-abdominal pressure (De Luca et al., 2004).

Other research indicates that differences in tissue composition may also play a role in predispositional weakening of the esophageal hiatus in HH. Studies show that a decrease in the collagen Type I-to-Type III ratio is associated with hernia formation (Brown et al., 2011). Curci's team (Curci et al., 2008) studied the periesophageal ligaments in patients with GERD and in patients diagnosed with GERD and HH. They found that the gastroesophageal sling in patients with GERD is characterized by prominent elastic fibers. In contrast, GERD with HH is associated with depletion of elastic fibers in two of three ligaments supporting the gastroesophageal junction. Thus, elastic fiber depletion in the periesophageal ligaments may provide a histologic, structural basis for the development of HH. Further, these differences in the extracellular matrix of tissues may also be related to kyphoscoliosis. Ehlers-Danlos Syndrome (EDS) is a group of disorders with disruption of the integrity of structural proteins in skin, ligaments, cartilage and blood vessels, leading to fragility of connective tissues secondary to abnormalities in collagen production. Generally, patients present with bruising and bleeding from the GI tract, dissecting aortic aneurysm, wide scars, laxity of joints, hyperelasticity of integument, and HH (Narayanan and Watkiss, 2009; Ignoto et al., 2006). EDS VI (also

known as kyphoscoliosis type) is a rare autosomal recessively inherited disease of connective tissue, where in addition to the usual signs and symptoms of EDS, patients also exhibit marked kyphoscoliosis. The underlying biochemical defect in EDS VI is a deficiency in lysyl hydroxylase (PLOD) activity resulting from mutations in the PLOD gene causing a low hydroxylysine content in various tissues. During collagen biosynthesis, specific hydroxylysine residues become glycosylated in the form of galactosyl- and glucosylgalactosyl-hydroxylysine. Furthermore, key glycosylated hydroxylysine residues,  $\alpha 1/2-87$ , are involved in covalent intermolecular cross-linking, and such cross-linking is crucial for the stability and mineralization of collagen (Terajima et al., 2014).

The incidence of HH has significantly increased over recent years, especially in our aging population (Gryglewski, 2014; Roman and Kahrilas, 2014; Loffeld and Vand Der Putten, 2002). Furthermore, the incidence of spinal deformities is also on the rise (Schuchert et al., 2011; Roman and Kahrilas, 2014). Current research investigations qualitatively and quantitatively document the association between giant HH and kyphoscoliosis in cadaveric specimens, suggesting that reduced IAV caused by kyphoscoliosis contributes to the development and progression of paraesophageal hernias in patients with laxity of the diaphragmatic hiatal musculature.

Physicians need to be aware of this association and vigilant in their evaluation and work-up of patients that present with GI complaints and spinal deformities – especially in the elderly. Clinical evaluation should include a robust medical history, barium esophagogram and upper endoscopy to assess presence and size of HH; and a routine posteroanterior chest x-ray should always be requested for determination of retrocardiac air bubble resulting from an air-fluid interface secondary to intrathoracic trapping of the stomach (Mitiek and Andrade, 2010; Naoum et al., 2011; Roman and Kahrilas, 2014). Anatomists need to be attentive in the study and documentation of cadavers affected by spinal deformities and HH. Even further, both physicians and anatomists must work together, taking the next steps in this causative investigation, perhaps by measuring tension in the diaphragmatic sling in both living patients and cadavers affected by kyphoscoliosis and HH, as well as using three-dimensional computer modeling to document and analyze the muscular anatomy of the gastroesophageal junction and progression of HHs (Yassi et al., 2009).

#### SUPPORT

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