Eur. J. Anat. 19 (3): 257-268 (2015)

Characterization of kyphoscoliosis and associated giant hiatal hernia in a 97-year-old female cadaver

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SUMMARY

The purpose of this investigation was to characterize severe kyphosis and scoliosis (kyphoscoliosis) and giant hiatal hernia (HH) in a 97-year-old female cadaver. Kyphosis is a ventral curvature of the thoracic spine that exceeds 50°. Scoliosis (lateral spinal curvature) is usually combined with vertebral rotation. HH is a hernia in which part of the gastrointestinal (GI) tract protrudes through the esophageal opening of the diaphragm. Although kyphoscoliosis has been suggested as a causative factor in the development of HH, scarce published data exist. If true, then this is clinically important in the evaluation and treatment of patients that present with spinal deformities and GI symptoms. Gross anatomical dissection was done. Vertebral deformities and displacement of structures were visualized with digital radiologic imaging using full-body x-ray films, and high-resolution CT and MRI Scans. Image analysis, multi-planar reformatting, and 3D- reconstruction were done on radiographic series. The *Cobb* and *Aaro-Dahlborn Methods* were used to determine the degree of spinal curvature and vertebral rotation, respectively. To examine the possible relationship between kyphoscoliosis and HH, intra-abdominal volume (IAV) was measured and compared to the IAV of unaffected cadavers. The heart was displaced superior and to the left with the apex touching the thoracic cage, whereas the aorta was 7.3 cm to the right of midline. The stomach was completely within the mediastinum. Thoracic dextroscoliosis, lumbar levoscoliosis and thoracic kyphosis had Cobb Angles of 45°, 34° and 78°, respectively. All thoracic and lumbar vertebrae were left-rotated; maximum rotations were T12 (18°) and L5 (29°). IAV was 4224 cm^3, and that of unaffected females ranged from 4449 to 7927 cm^3. This study provides insight into the relationship between kyphoscoliosis 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.

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Key words: Hiatal hernia – Scoliosis – Kyphosis – Kyphoscoliosis

Abbreviations:
anterior (A); anterior-posterior (AP); ascending aorta (Aa); axial (AX); computed tomography (CT); coronal (COR); degenerative disc disease (DDD); descending aorta (Da); diaphragm (DPH); empty space (ES); esophagus (E); foot or inferior (F); gastrointestinal (GI); hiatal hernia (HH); hiatal hernias (HHs); intervertebral (IV); intra-abdominal volume (IAV); left (L); left upper quadrant (LUQ); liver (Lv); magnetic resonance imaging (MRI); multi-planar reformatting (MPR); posterior (P); right (R); right upper quadrant (RUQ); spleen (Sp); stomach (S); three-dimensional (3D); weighted (Wtd).

INTRODUCTION

Kyphoscoliosis is defined as a deformity of the vertebral column represented by a combi-

Submitted: 23 March, 2014. *Accepted:* 4 April, 2013.

nation 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. The thoracic region of the vertebral column is normally curved. If this curvature exceeds 45°, then it is considered abnormal or kyphotic (Ferreira-Alves et al., 1995; Schuchert, et al., 2011). Kyphosis in the thoracic or lumbar spine is classified into three, age-associated categories: congenital; during life (postural Type A) and late in life (postural Type B). Congenital kyphosis is the least common type of abnormal kyphosis. This is caused by an abnormal development of the vertebrae prior to birth. This can lead to several of the vertebrae growing together (i.e., fusing) in kyphosis. During life, several events can distort the spinal column, and this was first described by Scheuerann (Scheuermann, 1921). Scheuermann's kyphosis also is first noticed during adolescence. This type of kyphosis is the result of a structural deformity of the vertebrae, and can be further subdivided into thoracic (or typical); thoracolumbar and lumbar (or reduced lordosis). It is more common to develop scoliosis (kyphoscoliosis) with Scheuermann's kyphosis than with the other types of kyphosis (McMaster and Singh, 1999). The diagnosis requires X-rays to show a wedge of at least 5 degrees at the front of at least three neighboring vertebral bodies. The reason for this abnormal wedging of the vertebrae is not understood. Other conditions that can do this include cancer, tuberculosis and some forms of arthritis. Vertebrae can also fracture anteriorly secondary to rapid deceleration injuries such as in motor vehicle crashes when the victim is not wearing a seat belt. Lastly, postural kyphosis is the most common type of kyphosis. This is more common in girls than in boys and is typically first noticed during adolescence (Tribus, 1998). It is caused by poor posture and a weakening of the muscles and ligaments in the back (i.e., paraspinous muscles). Typically, vertebrae are shaped normally in postural kyphosis. It is often slow to develop and usually does not become progressively worse with time.

When viewed posteriorly, the spine usually appears perfectly straight. Scoliosis is usually associated with a rotation of the vertebrae. When grouped by age, scoliosis usually is categorized into three age groups: infantile scoliosis (from birth to 3-years-old); juvenile scoliosis (from 3 to 9-years-old) and adolescent scoliosis (from 10 to 18-years-old). In addition, scoliotic curves are often described based on the direction and location of the curve. Some common terms used to describe scoliosis are: Dextroscoliosis (i.e., describes a spinal curve to the right), Levoscoliosis (i.e., describes a spinal curve to the left), and thoracic, lumbar scoliosis or thoracolumbar scoliosis, based upon respective location of curvature(s). Classification consists of curve type, and lumbar and thoracic spine modifiers (Lenke et al., 2001). In addition, all three regions of the radiographic coronal and sagittal planes, the proximal thoracic, main thoracic, and thoracolumbar/lumbar, are designated as either the major curve (i.e., largest Cobb measurement) or minor curves, with the minor curves separated into structural and nonstructural types. In most cases, a small degree of lateral 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).

Kyphoscoliosis is associated with restrictive impairment of pulmonary function, chronic pain and decreased vitality (Mitiek and Andrada, 2010; Naoum, 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). Modern diagnostic technologies and treatment protocols have increased the understanding of esophageal disease and the foregut. However, the nature of the relationship between the gut tube and the vertebral column is not adequately understood. More recent studies have illustrated a link between spinal disease and hiatal hernias (HHs) (Yang et al., 2011). A hiatal hernia (HH) is an anatomical abnormality in which part (or all) of the stomach protrudes up through the esophageal hiatus of the diaphragm into the thorax. This type of hernia can be congenital or acquired over time. While a clinical association between kyphoscoliosis and HH formation is strongly suggested, there are scarce published data available (Comte, 1953; Kahl and Koch, 1965). Comte was the first to describe the association between HH and kyphoscoliosis (Comte, 1953). Expanding upon this work, Galvala and Matejcic hypothesized that kyphoscoliosis may represent a causative factor in the development of HHs (Gavala and Zarabini, 1961; Matejcic, 1967). They suggest that axial deviation of the spinal canal at the level of the hiatus can lead to distortion of the hiatal sling mechanism, thus promoting gastrointestinal (GI) reflux and hiatal herniation (Gavala et al., 1961; Matejcic, 1967). Other suggested risk factors include decreased intra- abdominal volume and increased intra-abdominal pressures seen in patients with kyphoscoliosis (Schuchert et al., 2011).

Because of the diversity of symptoms and conditions presented by patients with spinal deformities, it is important that anatomists and clinicians understand the characteristics of kyphoscoliosis, as well as the factors and mechanisms that give rise to kyhposcoliosis and associated conditions. In this study, we report on a case of severe kyphoscoliosis and giant HH discovered during the course of anatomic dissection and radiographic study at the Indiana University School of Medicine - Northwest (Gary, IN). High-resolution CT and MRI technologies are used to further characterize the relationship between kyphoscoliosis and HH and to illustrate the significance of decreased abdominal volume resulting from kyphoscoliosis and its effects on formation of HH, and further differentiate this presentation from the typical report. Findings are discussed relative to the current literature.

MATERIALS AND METHODS

Cadaveric specimen

This study was conducted on a 97-year-old, Caucasian female cadaver as part of the International Human Cadaver Prosection Program and the formal course of human gross anatomy, both at the Indiana University School of Medicine - Northwest Campus (IUSM-NW; Gary, IN). Informed consent and a secondary medical history were obtained from the family of the anatomical donor. All federal and state guidelines were followed regarding the use and care of cadaveric materials, as well as all regulations set forth by the State of Indiana Anatomical Education Program.

Gross examination and photography

Detailed physical examination was performed utilizing a “donor report”, which is similar to an autopsy report, where gross observations and quantitative data were collected (Talarico, 2010, 2013). Digital photography of the external features and thoracic viscera was done using a NIKON D3100 SLR Camera (B&H Foto & Electronic Corporation, NY) equipped with an 18-55 mm VR NIKKOR Macro lens and a Nikon 40 mm f/2.8G AF-S DX NIKKOR 2200 VR Micro lens.

X-ray film imaging

Plain x-ray imaging was done in the radiology suite located on the second floor of the Dunes Medical Professional Building of the IUSM-NW. The following plain films were obtained: (1) anterior-posterior (AP) chest; (2) AP abdominopelvic; (3) upper extremity (pectoral girdle, brachium, antebrachium and carpus/manus); (4) lower extremity (pelvic girdle, thigh, leg and foot); (5) AP skull and lateral (Lat) skull.

Advanced medical imaging

Full-body, high-resolution CT and MRI imaging was done at Methodist Hospitals Southlake Campus (Merrillville, IN) using a 64-slice CT scanner (*General Electric* Lightspeed® capable of 3-dimensional (3D) reconstruction, and an MRI scanner (*General Electric* HIGH Speed MRI). Coronal (frontal), axial (transverse) and sagittal (median) views were generated both digitally and on film. Additional MRI scans included: (a) MRI of the brain including T1-weighted (Wtd) axial and sagittal; T2-Wtd axial, axial diffusion, and FLAIR axial scans; (b) MRI of the abdomen and pelvis to include T1- and T2-Wtd sequences in coronal and axial planes; (c) MRI of the knees, hips, and shoulders to consist of T1-, T2-Wtd, and STIR images in at least two planes; (d) MRI of the entire spine including T1- and T2-Wtd sagittal images.

Image analysis

Processing of images, creations of 3D-reconstructions, and quantitative image analysis were done using Konicia PDI Viewer 1.00 V1.0R0.00 (*KONICA Minolta*, Ramsey, NJ) and TDK CDRS Dashboard V1.0.0.5 (*TDK Medical*, Minneapolis, MN) for digital x-ray films; eFILM™ Lite™ Viewer 3.0 (*Merge Healthcare*, Chicago, IL) for radiographic series from CT-Scans; and Philips iSite Viewer (*Philips iSite*, Amsterdam, Netherlands) for radiographic series from MRI Scans. Additional image analysis, reformatting, and three- dimensional (3D) recontructions were done using PACSGEAR (*Perceptive Software*, Pleasanton, CA) on radiographic series from CT Scans and MRI Scans.

Software (above) used the *Cobb Method* (Kittleson and Lim, 1970) to determine the degree of curvature of the spine. Briefly, this measurement is made by drawing a line perpendicular to a second line drawn across the superior endplate of the upper-end (most tilted) vertebra and the inferior endplate of the lower-end vertebra; the angle formed by the intersection of the two perpendicular lines is defined as the *Cobb Angle*, which is the measure of the magnitude of the curve.

The vertebral rotation was measured in the cadaver utilizing the *Arso-Dahlborn Method* (Arso et al., 1978; Lam et al., 2008) applied to CT images. The measurement was made by first drawing a line joining the anterior midline of the body to the central aspect of the vertebral foramen. Then, a second line is drawn which runs through the midline of the vertebral body. The degree of rotation is the angle between these two lines.

Intra-abdominal volume (IAV) was defined as the area under the diaphragm (including the liver), posterior to the border of the rectus muscles, anterior to and excluding the retroperitoneal viscera and vasculature down to the plane of superior pelvic aperture (Moore et al., 2014). Using the method validated by *Agnew et al.* (Agnew et al., 2010), the IAV in this kyphoscoliosis-affected cadaver was compared to IAV of 13 female donors, ranging in age from 51 to 90-years, showing no evidence of grossly visible or radiographically measurable kyphoscoliosis, kyphosis or scoliosis. Briefly, IAV is calculated by taking three distance measurements within the defined parameters. In the axial plane at level T12, the left-to-right (along the mid-coronal plane) and anterior-to-posterior (along the mid-sagittal plane) distance measurements were made, while the superior-to-inferior distance measurements were made in the coronal plane. These three measurements approximate the area to resemble a rectangle and the equation for the volume of a rectangle is used to calculate the IAV. *Example*:

Intra-Abdominal Volume (IAV) = $AP\ x\ LR\ x\ SI$

AP = anterior-posterior distance; LR = left-right distance; SI = superior-inferior distance

IAV = $90.6\ mm\ x\ 244.3\ mm\ x\ 253.2\ mm = 5604222.456\ mm^3$

Conversion to cm^3:

$5604222.5\ mm^3\ x\ (0.001\ cm^3\ /\ 1\ mm^3) = 5604\ cm^3$

$1\ mm^3 = 0.001 cm^3$

HISTORY

AP = anterior-posterior distance; LR = left-right distance; SI = superior-inferior distance

The patient is a 97-year-old, white female that died in 2013 secondary to aspiration pneumonia. She suffered from chronic back pain, arthritis, and heartburn/acid reflux, as well as long-term complaints of abdominal pain and breathing difficulty (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. Earlier, the patient did have surgery with implants on her feet secondary to rheumatoid deformity, as well as bilateral knee replacements secondary to osteoarthritis. For almost 3 years prior to her death, she was mostly wheelchair-bound due to unsteady gait, but did occasionally ambulate a few steps with the assistance of a walker. She had a history of hypertension and

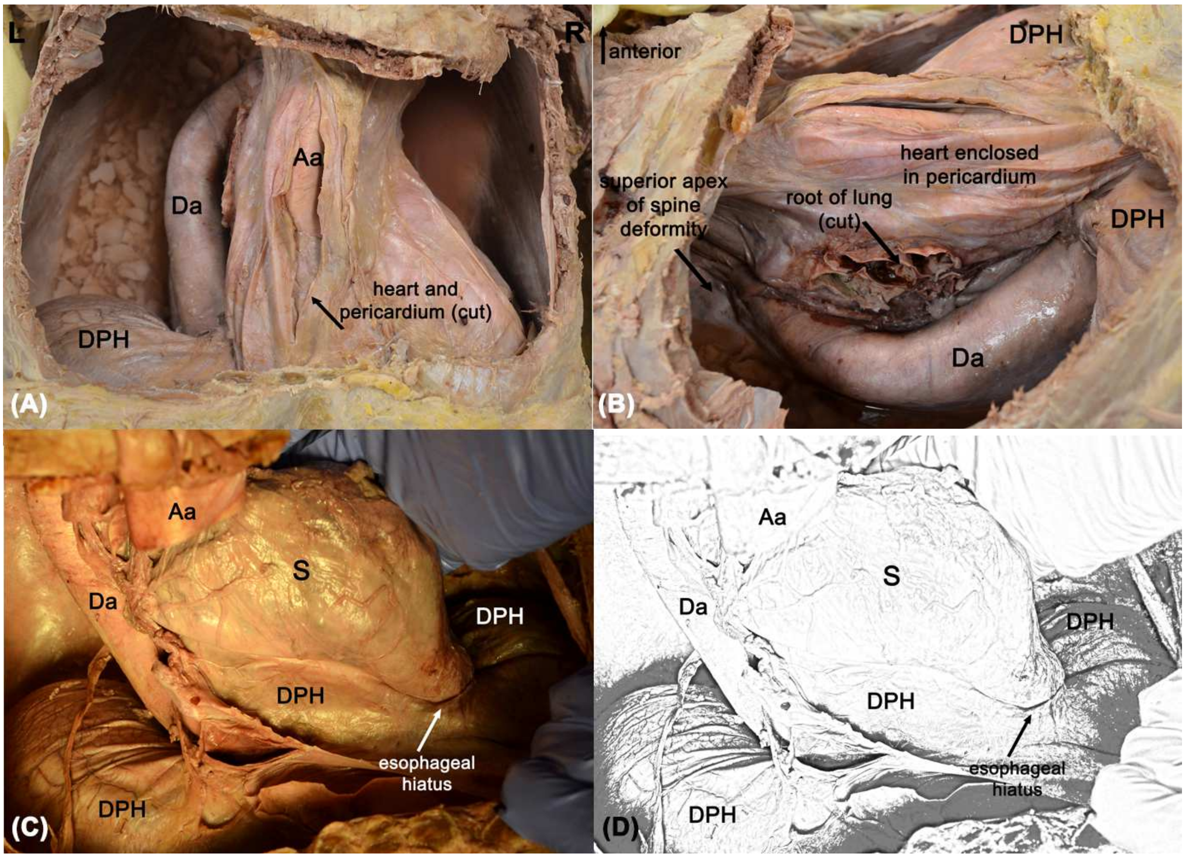

Fig. 1. Gross Anatomy of the Thorax. **(A)** Anterior view of mediastinal structures showing extensive right, lateral deviation of the descending, thoracic, aorta (Da) and slight right lateral deviation of the ascending aorta (Aa) and enlarged heart. The dome of the diaphragm (DPH) over the liver is also observed. **(B)** Right lateral view showing marked deviation and exaggerated right convex curvature of the Da. The superior apex defined by the Cobb Method of the spine deformity is seen in this view. **(C)** Right oblique view of the mediastinum with the heart and pericardium removed reveals the stomach (S) fundus, body and pyloric regions inside the thorax. The cut ascending aorta (Aa) is seen, as well as the Da and DPH. A dilated esophageal hiatus is seen (white arrow) at the origin of herniation. **(D)** Black-and-White diagrammatic representation of "C" for clarity. [Left (L) and right (R) markers in panel "A" apply to panels "C" and "D".]

cataract removal. History was also notable for occasional consumption of alcohol, conservative diet, use of calcium supplements, but no known use of recreational drugs or tobacco. There were no known allergies. Neuropsychiatric health was unremarkable; she was oriented to date, time and place, and had a positive life outlook.

RESULTS

Gross examination

A 97-year-old female cadaver was examined during the course of training in the Summer 2014 International Human Cadaver Prosection Program and the formal gross anatomy course at the IUSM-NW (Talarico, 2010). Gross examination revealed lateral curvatures in both the thoracic (right curvature) and lumbar (left curvature) regions of the vertebral as well as an anterior rounding (humpback) of the thoracic spine. Dissection of the thorax revealed an enlarged heart shifted to the left of midline with its apex touching the thoracic wall, and an enlarged aorta shifted 7.3 cm to the right of midline and along the anterior and right- lateral border of the scoliotic thoracic curvature (Fig. 1A and B). The left lung was compressed by the shifted heart. After removal of the heart, the stomach (cardiac, fundus, body and pylorus) was found to be inside the mediastinum (Fig. 1C and D). The esophageal hiatus of the diaphragm was observed to be dilated. This was confirmed by dissection of the abdominal cavity and GI tract; where the most proximal part of the duodenum near the pyloric sphincter was seen within the hiatus, and the region of the epigastrum where the stomach would normally be located was empty. The head of the pancreas had adhesions near to the duodenum and was shifted superiorly near to the hiatus. The remaining pancreas and spleen were in their usual anatomical locations. Osteoarthritis with osteophyte formation was grossly observed along the spine. The hands and feet bilaterally demonstrated the characteristic deformities of rheumatoid arthritis. The metacarpophalangeal joints were enlarged and the digits were shifted laterally from midline. The feet were even more so deformed with phalanges shifted laterally from midline and almost perpendicular to the metatarsal bones. There were surgical scars over the dorsum of the feet from prior implant surgery, as well as scars over the anterior knee articulations secondary to bilateral total knee arthroplasty.

Medical imaging

Plain X-ray (AP chest and AP abdominopelvic) showed the scoliotic curvatures of the vertebral column in the thoracic and lumbar regions. Scoliosis in the thoracic region was a right curvature, and in the lumbar region was a left curvature (Fig. 2A and B). The vertebral bodies appeared to be rotated in both the thoracic and lumbar regions, and the ribs subsequentially displaced. The vertebral bodies showed osteophyte formation at multiple levels, and the intervertebral (IV) discs demonstrated narrowing at all levels, collectively evidence of osteoarthritis and degenerative disc disease (DDD). The border of the diaphragm was visible with liver and spleen located in the right upper quadrant (RUQ) and left upper quadrant (LUQ), respectively (Fig. 2A). However, a radiolucent area was observed where a radiopaque area would be expected representative of the stomach (Fig. 2A and B), as well as gas and feces in the colon. Calcification of the wall of the aorta was visible, as well as the lateral border of the thoracic (descending) aorta to the right of midline and following the right-lateral, scoliosis. [Findings on films of head and extremities are not relevant,

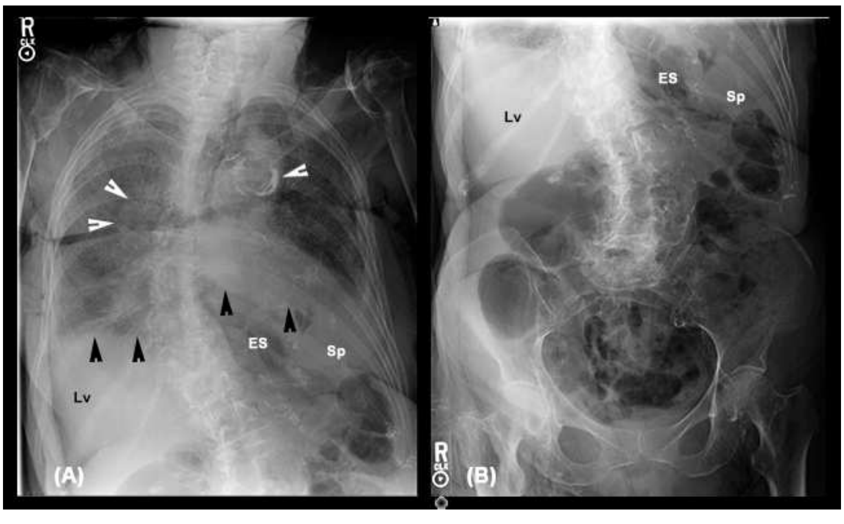

Fig. 2. Plain X-Rays of the Thorax and Abdomen. **(A)** AP flat-film of the thorax shows a calcified aortic knob and right-displaced thoracic aorta shadow (white arrows). The border of the diaphragm is also visible (black arrow). The spleen (Sp) can also be observed in the LUQ. Medial to the spleen, where a density is normally observed representing the stomach, is a radiolucent area (or empty space, ES). A large opacity in the RUQ is representative of the liver (Lv). Thoracic scoliosis is also seen. **(B)** This AP abdomino-pelvic film shows the Sp, Lv and ES, as well as sententious lumbar scoliosis. [Anterior-posterior (AP), right (R), right upper quadrant (RUQ), left upper quadrant (LUQ), spleen (Sp), empty space (ES), liver (Lv)].

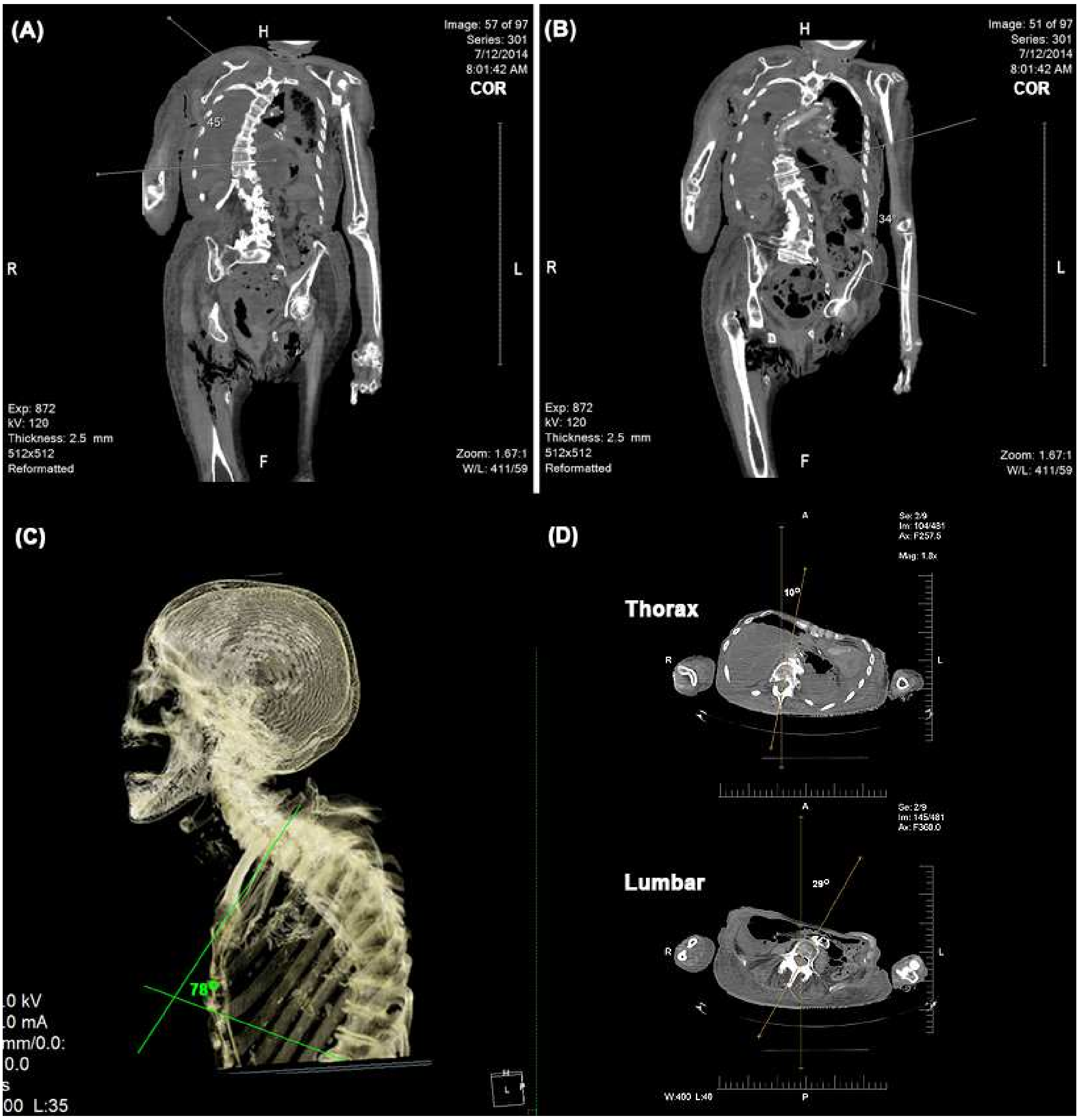

Fig. 3. Computed tomography of the vertebral column deformities**.** The *Cobb Angle* is used to measure and quantify the magnitude of spinal deformity. **(A)** This COR slice shows a right, thoracic scoliosis with a 45° Cobb Angel. **(B)** The second scoliotic curvature is a left, lumbar scoliosis with a 34° Cobb Angle. The right and left lateral curvatures of the vertebral column occur in the same plane (i.e., COR), but at different anterior-to-posterior slices (No. 57 and 51, respectively) secondary to rotation of vertebrae as part of this spinal deformity. **(C)** 3D CT-reconstruction was utilized to visualize and measure the Cobb Angle of the thoracic kyphosis, and was measured to be 78°. **(D)** These AX views document substantial left-rotation of vertebrae in the thoracic and lumbar regions of the spinal column with reference to the superior-to-inferior (i.e., longitudinal) axis. The upper panel shows a 10° rotation of vertebra T12, and the lower panel shows a 29° rotation of vertebra L5. [Left (L), right (R), anterior (A), posterior (P), head (H), foot (F)].

and thus not discussed here.]

CT scan and image analysis

Using a CT AX series, images of 0.25 mm slice thickness were generated of the entire cadaver in COR and SAG planes. Multi-planar reformatting (MPR) was used to view slices in different planes and for further analysis and measurements. The images obtained were used to measure the magnitude of scoliosis in the thoracic and lumbar spine as well as the kyphosis identified in the thoracic spine using the *Cobb Method* (Fig. 3A and B). The right thoracic scoliosis was measured to have a 45° Cobb Angle, and the left lateral lumbar curvature was measured to have a 34° Cobb Angle. In addition to these two scoliotic curvatures, the pelvis was shifted with the left ilium more inferior than the iliac crest on the right, and DDD (i.e., IV disc narrowing) and osteoarthritis (with osteophyte or bone spur formation) of the spine were observed. The calcified thoracic aorta (Fig. 2B) was meas-

Fig. 4. Coronal Magnetic Resonance Imaging (MRI) of patient. **(A)** A high-resolution T-2-weighted MRI scan on the posterior half of the thorax shows stomach with rugae and fluid contents (black asterisk) above the diaphragm. **(B)** A more anterior T-1-weighted image shows stomach with rugae (white asterisk) and a displaced thoracic aorta (black arrow), as well as liver (Lv) and spleen (Sp). The stomach no longer remains in the LUQ, or the umbilical, epigastric or left hypochondriac regions. **(C)** This T-1-weighed image through the mediastinum shows the displaced heart, stomach lumen (white asterisk), carina and left and right bronchi, and a greatly thickened wall of the aortic arch (white arrow).

ured to be 8 cm right-lateral to midline.

Because scoliosis and kyphosis occurred in the same thoracic region of the vertebral column, 3-dimentional CT reconstruction was applied to AX series to generate a complete construct of the kyphotic curve for image analysis (Fig. 3C). The thoracic kyphosis was measured to have 78° Cobb Angle.

Using *Arso-Dahlborn Method*, the degree of vertebral rotation was quantified. Rotation about the longitudinal axis (superior-inferior axis) was found and all thoracic and lumbar levels; at all levels was to the left and ranged 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 (Fig. 3D).

Distance measurements and calculated IAV are shown in Table 1. In the present anatomical donor, the IAV was 4224 cm^3, or 17% to 53% less than that of unaffected females whose IAV's ranged from 5064 cm^3 to 7927 cm^3. On average, the IAV of this kyphoscoliosis-affected donor was 2360 cm^3 or 29.8% less than that of unaffected cadavers examined here.

MRI Scan

A full body MRI was obtained using high-resolution 1.00 mm thick slices. MRI images were used to better illustrate the HH and the subsequent complications kyphoscoliosis imparts upon structures within the thorax and abdomen. T1- and T2-weighed images show the rugated stomach (with fluid in the T2-weighed image) above the diaphragm and within the mediastinum (Fig. 4A and B). There is also significant displacement of the thoracic aorta (Fig. 4B), and the heart (Fig. 4C) adjacent to the stomach.

DISCUSSION

This study has examined the relationship between giant HH and kyphoscoliosis in a 97-year-old female cadaver. Scoliosis was identified in both the thoracic (dextroscoliosis) and lumbar (levoscoliosis) spine and measured 45° and 34°, respectively. Kyphosis measured 78°. Scoliosis is characterized in adults with degrees of curvature greater than 10 degrees, whereas kyhposis is characterized in adults in whom the curvature exceeds 50°. Kyphosis can occur in the thoracic or lumbar spine, and can be classified into three, age -associated categories: congenital; during life (postural Type A) and late in life (postural Type B) (Schuchert 2009). Postural kyphosis is the most common type of kyphosis. This is more common in girls than in boys and is typically first noticed during adolescence (Tribus, 1998). It is caused by poor posture and a weakening of the muscles and ligaments in the back (paraspinous muscles; vertebrae are normally shaped; and it is often slow to develop and usually does not become progressively worse with time.

The cadaver was never diagnosed with kyphosis or scoliosis as a child and had medical history of osteoarthritis of the spine and bilateral deformities of the feet and hands secondary to rheumatoid arthritis. Grubb et al. (Grubb et al., 1998) and Aebi (Aebi, 2005) studied the relationship between DDD and spinal deformities. They found that patients that had DDD developed kyphosis and scoliosis. This supports the present study, where advanced DDD was identified via medical imaging. Further, the vertebral bodies in both the thoracic and lumbar regions were rotated turning the rib cage to the left along the longitudinal axis. The degree of rotation ranged from 4° at T1 to 29° at L5. These data and the patient's medical history suggest that this was a Type B kyphosis.

Identification of both kyphosis and scoliosis is called kyphoscoliosis and 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).

Gross anatomical dissection in the present specimen showed that the heart was displaced slightly superior and to the left. Removal of the heart revealed that the esophageal hiatus of the diaphragm was dilated, and that the most proximal part of the duodenum near the pyloric sphincter was within the hiatus, and the region of the epigastrum, observed at the time of abdominal dissection, where the stomach would normally be located was empty. The stomach was totally contained within the mediastinum. This condition known as 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., colon, 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 other describe it as a herniation of more than 50%. Thus, any hernia with more than 50% of the stomach contained in the thorax should be considered a giant HH. 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., gastroesophageal reflux disease), 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, 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). In the study presented

Table 1. Distance measurements and calculated abdominal volumes

Cadaver	Anterior-Posterior (mm)	Left-Right (mm)	Superior-Inferior (mm)	Intra-Abdominal Volume (cm^3)
1 (kyphoscoliosis)	66	248	258	4223
2 (unaffected)	104.7	217.4	222.5	5064
3 (unaffected)	102.4	251.4	231.8	5967
4 (unaffected)	142.8	249.2	224.1	7972
5 (unaffected)	114.7	245.3	261.6	7360
6 (unaffected)	93.2	228.3	246.6	5247
7 (unaffected)	125.3	238.6	263.3	7872
8 (unaffected)	112.3	246.2	245.3	6782
9 (unaffected)	106.8	257.6	269.2	7406
10 (unaffected)	116.3	233.8	247.6	6732
11 (unaffected)	84.3	232.3	268.8	5264
12 (unaffected)	98.6	252.3	255.8	6363
13 (unaffected)	104.3	243.6	266.3	6766
14 (unaffected)	101.3	253.4	264.8	6797

This table shows the values of distance measurements at vertebral level T12 and the calculated values for intra-abdominal volume (IAV) for the kyphoscoliosis-affected cadaver and 13 female anatomical donors, ranging in age from 51 to 90-years, showing no gross and radiographic evidence of spinal disease or hiatal hernia. Calculations were done using the *Agnew Method* (Agnew et al., 2010), here, the patient suffered from heartburn/acid reflux as well as long-term complaints of abdominal pain and feelings of pressure in her chest. Further, the cause of death was recorded to be aspiration pneumonia. With these findings and the image analysis done here, it is clear that the patient suffered from a giant HH that was never diagnosed.

The frequency of HH increases with age (Loffeld and Vand Der Putten, 2002; Gordon et al., 2004). Critically, understanding of esophageal disease and the foregut has evolved over recent decades. Modern diagnostic technologies and new management paradigms have provided progressive insights into the anatomy, physiology, and normal and abnormal function of the esophagus and stomach. Yet, the relationship between the gut tube and its close neighbor, the spine, is rarely discussed and likely underappreciated. While a clinical association between kyphoscoliosis and HH formation is strongly suggested, there are scarce published data available (Comte, 1953; Kahl and Koch, 1965). Comte was the first to describe the association of HH with kyphoscoliosis (Comte, 1953). Expanding upon this work, Galvala and Matejcic hypothesized that axial deviation of the spine may cause distortion of the hiatal sling leading to the development of HH (Gavala and Zarabini, 1961; Matejcic, 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).

In order to determine the effects kyphoscoliosis has on structures within the abdomen and to examine the possible relationship between kyphoscoliosis and HH, it was decided to study whether individuals with kyphoscoliosis have different abdominal volumes relative to those unaffected with scoliosis, kyphosis or kyphoscoliosis. Thirteen women unaffected with kyphoscoliosis, kyphosis or scoliosis, were measured and compared to the abdominal volume measured for the cadaver affected with kyphoscoliosis. All unaffected individuals exhibited IAV larger than that of the affected cadaver (Table 1). IAV of the affected cadaver was 4224 cm^3, and that of unaffected females ranged from 5064 to 7072 cm^3. These data suggested 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.

Bianchi et al. (Bianchi et al., 1960) hypothesized that diaphragmatic hernias were affected by the structural and static-dynamic forces of the spine and its abnormalities such that abdominal curvature of the spine such as kyphoscoliosis causes a stretching of the diaphragm. In the face of the data presented here, this seems reasonable. Specifically, the cura 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 gastroesophageal junction into the thorax. Further, decreased IAV in patients with kyphoscoliosos, may increase pressure on viscera and associated mesentery allowing displacement of the gastroesopheageal junction and formation of a HH. Intra-abdominal pressure becomes raised above normal, either intermittently, due to exertion or straining (i.e. Valsalva maneuver), or chronically due to obesity or external compression, and as a result the junction is pushed superiorly. This idea is supported by known mechanisms for inguinal and femoral hernias. In the case of indirect inguinal hernias, chronic or intermittently increased intra-abdominal pressure and muscle weakness result in the intestine herniating thorough the deep inguinal ring (a naturally occurring opening in the musculature of the abdominal wall) and into the inguinal canal (and scrotum). 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 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-abdoiminal pressure (De Luca et al., 2004). Finally, 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), and some studies report a depletion of elastic fibers in ligaments supporting of the gastro-esophageal sling (Curci et al., 2008).

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). Spinal deformities have long been hypothesized to be related to esophageal disease and HH (Polomsky et al., 2011; Comte, 1953). Further, the incidence of spinal deformities is also on the rise (Schuchert et al., 2011; Roman and Kahrilas, 2014). This study qualitatively and quantitatively documented the association between giant HH and kyphosocliosis in a cadaveric specimen, suggesting 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. Clinicians need to be aware of this association in the evaluation and work-up of patients that present with GI complaints and spinal deformities. Clinical evaluation should include a detailed history, barium esophagogram and upper endoscopy to assess presence and size of HH, and routine posteroanterior chest x-ray for 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 Kahailas, 2014).

ACKNOWLEDGEMENTS

The authors wish to express their sincere gratitude to Lydia Grady, who bequeathed her body for medical education and basic science research. Lydia Grady served as a U.S. Army Nurse in England, during World War II, where she treated and healed wounded men and women. After the War, she returned to Gary, IN, where for the remainder of her life she fought for the equal rights of African-Americans and other minorities. We also wish to thank Klari Neuwelt, daughter of Lydia Grady, for her assistance and cooperation with our investigation. As the daughter of Lydia Grady, Ms. Neuwelt helped provide important information regarding Ms. Grady’s medical history. We also express our sincere appreciation to Luis O. Marquez, former Director of Imaging Service, Methodist Hospitals South Lake Campus, and to Rhonda C. During, CT Technologist, Methodist Hospitals South Lake Campus, for their assistance in the development of radiographic materials for this manuscript.

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SUPPORT

Methodist Hospitals Southlake Campus (Merrillville, IN) Indiana University School of Medicine - Northwest (Gary, IN) Rocco Prosthetic and Orthotic Center (Cincinnati, OH) Mortech Manufacturing, Inc. (Azusa, CA).

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