

BACKGROUND

- Ehlers-Danlos Syndrome (EDS) is a heterogenous group of heritable connective tissue disorders.
- Hypermobile EDS (hEDS) is the most common.
- Bloating, distention, change in bowel habits and abdominal pain and delayed gastric emptying are very common in EDS patients.
- The pathophysiology of GI symptoms in EDS patients remains unknown but visceroptosis, defined as prolapse of abdominal organs below their natural position, has been proposed as a cause of GI symptoms in EDS.

AIM

- To define the normal dynamic movement of the viscera during supine and upright radiographs of the abdomen in healthy subjects.
- To assess the prevalence of visceroptosis in hEDS patients.

METHODS

- Patients with hEDS fulfilling Rome IV criteria for irritable bowel syndrome were included. Patients with previous abdominal surgeries were excluded.
- Following ingestion of 16 oz barium, the passage of contrast was serially followed through the small bowel until it reached the colon. At that point, supine and upright radiographs of the abdomen were obtained.
- Measurements were calculated on supine and fully upright positions with respect to a reference line drawn across the top of the iliac crests. Dynamic measurements included: the lowest point of stomach, bottom of small bowel column in the pelvis, inferior tip of the liver, and top of jejunal column. (Figures 1) Correction for patients' height was made by normalizing data to the height of T12 vertebral body.
- hEDS patients with visceroptosis were defined if any of their measurements exceeded two standard deviations above the mean established in the healthy control cohort (Table 1).

RADIOLOGIC ASSESSMENT

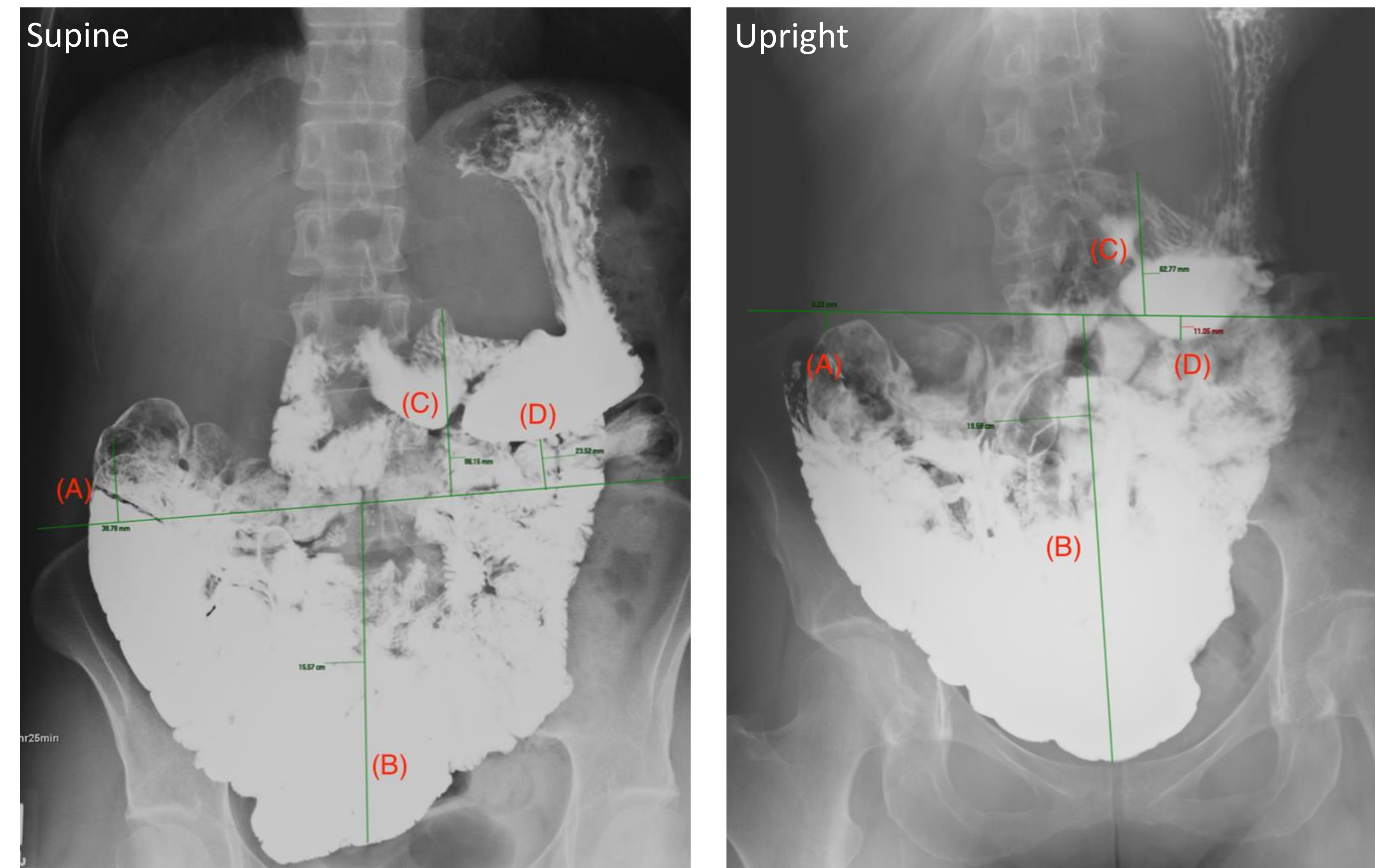


Figure 1. Baseline measurements for assessment of visceroptosis in supine position with respect to line drawn across iliac crest: (A) inferior tip of the liver, (B) bottom of small bowel column in the pelvis, (C) top of jejunal column, (D) lowest point of stomach.

RESULTS

- Eleven healthy subjects and nine patients with hEDS were enrolled (91% vs 100% Female, mean age 34 ± 13 vs 30 ± 8 years, mean Beighton score 1.8 ± 2.2 vs 7.4 ± 1.3). All hEDS subjects had abdominal pain and bloating. Constipation and mixed pattern were present in 78% and 22%, respectively.
- Only values for the lowest point of stomach and top of the jejunal column show altered distribution between healthy and EDS subjects.
- Three (33%) hEDS patients met the radiographic criteria for visceroptosis.
- Neither Beighton score, height, weight, or BMI correlated with radiographic evidence of visceroptosis.

CONCLUSIONS

- This is the first study to establish a normal range for the dynamic movement of the viscera during supine and upright radiographs of the abdomen.
- The prevalence of visceroptosis in hEDS patients with functional GI symptoms needs to be validated in larger cohorts.

REFERENCES:

- Reinstein, et al. *Visceroptosis of the Bowel in the Hypermobility type of Ehlers-Danlos Syndrome: presentation of a rare manifestation and review of the literature.* 2012. Eur J Med Genet.
- Zeitoun, et al. *Functional Digestive Symptoms and Quality of Life in Patients with Ehlers-Danlos Syndromes: Results of a National Cohort Study on 134 Patients.* 2013. Plos one.

Landmarks	Healthy subjects (n=11)		hEDS patients with GI symptoms (n=9)		Number of subjects with visceroptosis
	Centimeter drop in upright position (mean \pm std)	Normalized values based on T12 size (mean \pm std)	Centimeter drop in upright position (mean \pm std)	Normalized values based on T12 size (mean \pm std)	
Lowest point of stomach	8.99 \pm 3.13	2.92 \pm 0.97	9.89 \pm 7.22	3.25 \pm 2.28	3 (33%)
Top of jejunal column	1.96 \pm 1.89	0.64 \pm 0.59	2.19 \pm 1.70	0.74 \pm 0.58	0 (0%)
Bottom of small bowel column in the pelvis	4.28 \pm 2.09	1.47 \pm 0.87	3.60 \pm 2.78	1.19 \pm 0.89	0 (0%)
Inferior tip of the liver	5.99 \pm 2.02	1.99 \pm 0.73	4.07 \pm 2.91	1.34 \pm 0.90	2 (22%)

Table 1. Normative values for radiographic measurements of visceroptosis in healthy subjects along with values observed in hEDS patients.