Ehlers Danlos Syndrome should not be considered an absolute contraindication to Bariatric surgery – a case report and review of the literature

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Introduction

Ehlers-Danlos syndrome (EDS):

- Inherited connective tissue disorders caused by defects in collagen synthesis.
- Significant genetic heterogeneity and phenotypic variability
- Prevalence thought to be 1:5000, though the true prevalence is likely higher [1, 2].

Classification: [3].

- Classical EDS (cEDS): joint hypermobility, skin hyperextensibility, recurrent abdominal hernias, atrophic scarring, pelvic floor prolapse, musculoskeletal pain, recurrent joint dislocations and skin fragility
- Hypermobile EDS (hEDS): similar to cEDS but with less skin fragility and more issues with mitral valve prolapse and aortic root dilatation.
- Vascular EDS (vEDS): rare but more serious; arterial rupture, spontaneous colon perforation, uterine rupture, easy bruising, thin, translucent skin, and spontaneous pneumothorax [2, 3].
- 10 other subtypes

Obesity:

- Prevalence in US adults: 42.2% [4].
- Common comorbidities: type 2 diabetes, high blood pressure, heart disease and joint disease.
- Bariatric surgery provides significant weight loss, substantial improvement in comorbidities [5-8].
- EDS patients have a higher risk of surgical morbidity: wound dehiscence, anastomotic leak, bleeding, and hernia formation [9-11].

Case Report

We present a 35-year-old woman with EDS (subtype unknown) and a body mass index (BMI) of 67 kg/m^2 who was referred for bariatric surgery. Her comorbidities include obstructive sleep apnea, polycystic ovarian syndrome, and hypertension. Regarding her EDS, she suffers from a chronically dislocated shoulder, subluxed ribs, and hyperextensible joints. Several of the patient's family members also have been diagnosed with EDS, but she has no personal or family history of vascular abnormalities or spontaneous colon perforations. She subsequently underwent a laparoscopic Roux-en-Y gastric bypass. An upper gastrointestinal study on post-operative day one was negative and she was discharged on post-operative day two. She did require readmission 3 weeks post-operatively for dehydration. At her one year follow up she had achieved a total body weight loss of 30% (an excess body weight loss of 49%), bringing her BMI down to 46 kg/m^2 .

Publication	Year	EDS Subtype	Surgery Type	Complication
Belluzi et al.	2020	Hypermobile	LSG	None
Gerogiannis et al.	2020	Vascular	RYGB	Gastric diverticulum, gastro-esophageal junction stenosis, hiatal hernia
Hakky et al.	2019	Vascular	RYGB	Hiatal hernia, anastomotic leak
Herdes et al.	2019	Hypermobile	LSG	Sleeve stenosis
		Hypermobile	LSG	None
Thornopoulos et al.	2020	Unknown	RYGB	Internal hernia
Verdure et al.	2020	Hypermobile	RYGB	None
		Hypermobile	LSG	None
		Hypermobile	LSG	None
		Classical	LSG	None
		Hypermobile	LSG	None
		Hypermobile	RYBG	None
		Vascular	RYGB	Significant bleeding requiring reoperation

LSG: laparoscopic sleeve gastrectomy. RYGB: Roux-en-Y gastric bypass

Discussion

- Available literature regarding gastrointestinal surgery in EDS patients has focused on urgent and emergent abdominal surgery, especially spontaneous colon perforations [9, 10, 18-22].
- Increased risk of perioperative complications due to impaired healing and increased fragility, regardless of EDS subtype [10, 11, 19].
- Minimize peri-operative morbidity when possible [23].:
  - ✓ Careful attention to patient positioning can help prevent joint dislocations and skin injury
  - ✓ Laparoscopic surgery minimizes incision size thereby decreasing hernia risk
  - ✓ Close skin in multiple layers and using skin glue or steristrips to support the closure
- Surgery selection
  - ✓ Shorter operative time and less tissue handling with LSG
  - ✓ LSG avoids an enteric anastomosis and the potential for anastomotic leak
  - ✓ Long gastric staple line in LSG still poses the risk of leakage.
  - ✓ Staple line reinforcement or oversewing the long staple line to mitigate risk of leak in LSG
- Several publications discuss the significant morbidity, and mortality associated with non-elective endoscopy and abdominal surgery in patients with vEDS [9, 18, 20, 22, 24-26].

Conclusions

- EDS patients undergoing surgery are at increased risk of wound dehiscence, hernia formation, anastomotic breakdown, and bleeding due to their inherent defective collagen synthesis.
- The terminal nature of untreated morbid obesity makes the need to perform metabolic surgery a more pressing issue.
- Individualized pre-operative preparation should be undertaken to provide the appropriate treatment to improve overall health.
- Given this significant increase in peri-operative risk with vEDS, serious consideration should be given to non-operative options for managing all conditions including obesity prior to pursuing elective surgery.

References

1. Beighton, P., et al., *Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. Ehlers-Danlos National Foundation (USA) and Ehlers-Danlos Support Group (UK).* Am J Med Genet, 1998. **77**(1): p. 31-7.
2. *National Organization of Rare Diseases: Ehlers Danlos Syndromes.* [cited 2021 August 17]; Available from: https://rarediseases.org/rare-diseases/ehlers-danlos-syndrome/.
3. Malfait, F., et al., *The 2017 international classification of the Ehlers-Danlos syndromes.* Am J Med Genet C Semin Med Genet, 2017. **175**(1): p. 8-26.
4. Craig M. Hales, M.D.C., Cheryl D. Fryar, Cynthia L. Ogden, *Prevalence of Obesity and Severe Obesity Among Adults: United States, 2017–2018.* NCHS Data Brief No. 360, 2020.
5. Chang, S.-H., et al., *The Effectiveness and Risks of Bariatric Surgery: An Updated Systematic Review and Meta-analysis, 2003-2012.* JAMA Surgery, 2014. **149**(3): p. 275-287.
6. Padwal, R., et al., *Bariatric surgery: a systematic review and network meta-analysis of randomized trials.* Obes Rev, 2011. **12**(8): p. 602-21.
7. McTigue, K.M., et al., *Screening and interventions for obesity in adults: summary of the evidence for the U.S. Preventive Services Task Force.* Ann Intern Med, 2003. **139**(11): p. 933-49.
8. Christou, N.V., et al., *Surgery decreases long-term mortality, morbidity, and health care use in morbidly obese patients.* Ann Surg, 2004. **240**(3): p. 416-23; discussion 423-4.
9. Maltz, S.B., et al., *Surgical complications of Ehlers-Danlos syndrome type IV: case report and review of the literature.* J Trauma, 2001. **51**(2): p. 387-90.
10. Kulas Saborg, M.L., et al., *Increased Need for Gastrointestinal Surgery and Increased Risk of Surgery-Related Complications in Patients with Ehlers-Danlos Syndrome: A Systematic Review.* Digestive Surgery, 2017. **34**(2): p. 161-170.
11. Fogel, S., *Surgical Failures: Is It the Surgeon or the Patient? The All too Often Missed Diagnosis of Ehlers-Danlos Syndrome.* The American Surgeon, 2013. **79**(6): p. 608-613.
12. Belluzzi, A. and M. Foletto. *Sleeve Gastrectomy in a Patient with Obesity and Ehlers-Danlos Syndrome: A Case Report.* 2020.
13. Gerogiannis IN, G.I., Dempster NJ, Gillies RS, Sgromo B, *Obesity surgery and Ehlers–Danlos syndrome: challenges and considerations based on a case report.* The Annals of The Royal College of Surgeons of England, 2020. **102**(1): p. e7-e11.
14. Hakky, S. and N. Petrou, *When all the gastric bypass goes north : The emergency management of an acute complex para-oesophageal hernia post Roux-en-Y gastric bypass.* Surgery for Obesity and Related Diseases, 2019. **15**(10): p. S87.
15. Herdes, R.E., et al., *Experience With Vertical Sleeve Gastrectomy in Adolescent and Young Adult Ehlers-Danlos Syndrome Patients: a Case Series and Review of the Literature.* Obes Surg, 2021.
16. Thomopoulos, T., et al., *Management of a Complicated Internal Herniation After Roux-en-Y Gastric Bypass in a 28-Week Pregnant Woman.* Obes Surg, 2020. **30**(12): p. 5177-5178.
17. Verdure, L., et al., *Bariatric Surgery is feasible in patients with Ehlers-Danlos Syndrome.* Surg Obes Relat Dis, 2020. **16**(9): p. 1328-1331.
18. Adham, S., et al., *Natural History and Surgical Management of Colonic Perforations in Vascular Ehlers-Danlos Syndrome: A Retrospective Review.* Dis Colon Rectum, 2019. **62**(7): p. 859-866.
19. Burcharth, J. and J. Rosenberg, *Gastrointestinal surgery and related complications in patients with Ehlers-Danlos syndrome: a systematic review.* Dig Surg, 2012. **29**(4): p. 349-57.
20. El Masri, H., et al., *Bowel perforation in type IV vascular Ehlers-Danlos syndrome. A systematic review.* Tech Coloproctol, 2018. **22**(5): p. 333-341.
21. Freeman, R.K., J. Swegle, and M.J. Sise, *The surgical complications of Ehlers-Danlos syndrome.* Am Surg, 1996. **62**(10): p. 869-73.
22. Speake, D., et al., *Management of colonic complications of type IV Ehlers-Danlos syndrome: a systematic review and evidence-based management strategy.* Colorectal Dis, 2020. **22**(2): p. 129-135.
23. Wiesmann, T., et al., *Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s).* Orphanet J Rare Dis, 2014. **9**: p. 109.
24. Beridze, N. and W.H. Frishman, *Vascular Ehlers-Danlos syndrome: pathophysiology, diagnosis, and prevention and treatment of its complications.* Cardiol Rev, 2012. **20**(1): p. 4-7.
25. da Rocha de Sousa, F., et al., *Surgical management of vascular Ehlers-Danlos syndrome and its challenges: a case report.* Swiss Med Wkly, 2020. **150**: p. w20379.
26. Omori, H., et al., *Sigmoid colon perforation induced by the vascular type of Ehlers-Danlos syndrome: report of a case.* Surg Today, 2011. **41**(5): p. 733-6.