Surgical Failures: Is It the Surgeon or the Patient? The All too Often Missed Diagnosis of Ehlers-Danlos Syndrome

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Source: The American Surgeon, Volume 79, Number 6, June 2013 , pp. 608-613(6)
Publisher: Southeastern Surgical Congress

Abstract:
Ehlers-Danlos syndrome (EDS) is a closely related group of disorders caused by a heritable defect in collagen synthesis, which leads to marked healing difficulties. It has been estimated to occur in between one in 2500 and one in 5000 individuals but likely occurs more frequently than reported. EDS has probably been seen by all general surgeons several times over the course of a career. The purpose of this report is to describe the findings that should raise the index of suspicion, to aid in the diagnosis, and to characterize the general surgical procedures seen in patients with EDS by reviewing a single surgeon's experience in managing such patients with a review of the literature. Recommendations for treatment are given. A retrospective review of the experience of a single surgeon of 25 procedures in 15 patients with EDS is being reported. This is believed to be the largest series by one surgeon as yet reported. There was a wide variety of procedures performed, including ventral hernia repair (n = 6), inguinal hernia repair (n = 4), colectomy (n = 3), anal fistula (n = 3), and one each of an exploratory laparotomy, an appendectomy, a closure of a dehiscence, a Hickman catheter placement, an open lysis of adhesions for small bowel obstruction (SBO), a laparoscopic lysis of adhesions for SBO, an open cholecystectomy, a laparoscopic cholecystectomy, and an excision of a round ligament endometrioma. There was only one death, which was in a patient with Type IV EDS who was the first patient in this series. He presented with a spontaneous sigmoid perforation treated by Hartmann procedure and went on to develop two more colon perforations and to die of sepsis. The morbidity included only two recurrent ventral hernias, a wound dehiscence, a wound hematoma, and recurrence of the anal fistula. Although patients with EDS pose significant healing problems, successful general surgical procedures can be performed in most patients. Among other recommendations, total avoidance of colon anastomoses and colostomies in favor of total abdominal colectomy and ileostomy and routine closure of the abdominal wall with mesh or retention sutures is advocated. Making the diagnosis is the key to having successful outcomes. Further recommendations on avoiding operation and on the conduct of the operation, if needed, are given.

Document Type: Research article
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Publication date: 2013-06-01