Table of Contents

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Marfan Syndrome and Abdominal Aortic Aneurysm Surgical Repair Outcomes: A Systematic Review

Meghan Smith, Mitchka Mohammadi, Majdeddin MohammedAli, Cheyaanthan Haran, Ishaq Wadiwala, Andrew Hamilton, Sherene Shalhub

Oregon Health and Science University, Portland Oregon. MD Candidate

Keywords

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Abstract

Background

Marfan Syndrome is a genetic condition defined by autosomal dominant variations in the FBN1 gene resulting in changes to the cardiovascular, ocular, and skeletal systems. While a commonly explored feature of Marfan Syndrome is dilation of the aortic root, abdominal aortic aneurysm (AAA) and thoracoabdominal aneurysm (TAAA) are less studied syndrome sequelae. This systematic review provides a summary of the literature detailing surgical outcomes for patients with Marfan Syndrome undergoing AAA or TAAA repair.

Methods

Researchers performed a systematic literature review using Ovid MEDLINE. Search terms were formatted as key words and MeSH terms. The search was limited to studies that were published after 1945. In addition to literature pulled through MEDLINE, researchers also included studies identified through reference mining and investigator recommendation.

Results

Of the 2,881 studies identified using the search strategy, investigators included 248 studies in this systematic review. Included studies contained information about patients with a clinical or genetic diagnosis of Marfan undergoing abdominal surgical intervention for AAA or TAAA. 59 studies contained relevant aggregate outcome data for 6,146 procedures. 189 studies contained individual outcome data for 348 patients undergoing 317 open repairs, 49 endovascular procedures, and 4 hybrid operations. There were 8 intraoperative deaths and 14 deaths within 30 days. 12.2% of the endovascular repairs required redo open operation, and 22.4% had documented endoleak. 16.6% of patients

went on to need future iliac or proximal aortic operations related to their condition. 4.9% of patients had genetically confirmed FBN1 variation and the average age of patients undergoing abdominal surgery was 37.45 years

Conclusions

The available evidence suggests that patients with Marfan undergoing surgical TAAA or AAA repair have good survival and low complication rates. These findings are complicated by a lack of genetic FBN1 variant confirmation in most patients studied.