



GEN06 - Long-term implications of acute dissection as primary indication for aortic root replacement in Marfan syndrome patients

OBJECTIVE: To compare the long term clinical course of Marfan syndrome patients surviving emergency versus elective proximal aortic surgery.

ORGANIZATION

Lead Investigator: Howard Song, MD, PhD

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Funding Source: GenTAC

Samples • None

Data • Surgical and imaging data. Demographics and system review.

BACKGROUND AND RATIONALE

Patients with Marfan syndrome typically undergo aortic surveillance with serial imaging of the aortic root and are offered elective aortic root replacement when their aortic root diameter reaches an appropriate size in order to avoid life threatening presentation with aortic dissection and the high morbidity of emergency aortic surgery. While the immediate risk of aortic dissection and emergency repair for this patient group has been extensively studied, the long-term implications of failed aortic surveillance and emergency dissection repair are poorly defined.

DESIGN

- Hypothesis:*
- Marfan syndrome patients undergoing emergency aortic root replacement vs. elective aneurysm repair are disadvantaged by: lower incidence of valve sparing surgery; worse perioperative outcomes including length of stay, renal failure, prolonged ventilation, stroke, and need for transfusion; higher frequency of residual dissection involving the aortic arch and descending thoracic aorta following surgery; higher frequency of distal aortic aneurysms; greater need for follow up imaging; greater need for further procedures; Worse quality of life.
- Inclusion criteria:*
- Marfan subjects who have either undergone emergency aortic root replacement for acute dissection or prophylactic aneurysm repair.

CONCLUSIONS

- Results:*
- There are significant long term implications for Marfan syndrome patients who fail aortic surveillance and require emergency dissection repair with regard to status of the distal aorta, requirement for multiple procedures, and quality of life. These findings emphasize the importance of aortic surveillance and timely elective aortic root aneurysm repair for Marfan syndrome patients.
- Publication*
- Song HK, Kindem M, Bavaria JE, Dietz HC, Milewicz DM, Devereux RB, Eagle KA, Maslen CL, Kroner BL, Pyeritz RE, Holmes KW, Weinsaft JW, Menashe V, Ravekes W, Lemaire SA; Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions Consortium: Long-term implications of emergency versus elective proximal aortic surgery in patients with Marfan syndrome in the Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions Consortium Registry. J Thorac Cardiovasc Surg. 2012 Feb;143(2):282-6. Epub 2011 Nov 20.