Journal of Cardiovascular Magnetic Resonance

BioMed Central

Meeting abstract

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1039 Prevalence of cardiovascular manifestations in patients with Marfan syndrome: a cardiovascular magnetic resonance study

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from 11th Annual SCMR Scientific Sessions
Los Angeles, CA, USA. 1–3 February 2008

Published: 22 October 2008

Journal of Cardiovascular Magnetic Resonance 2008, **10**(Suppl 1):A164 doi:10.1186/1532-429X-10-S1-A164

This abstract is available from: http://jcmr-online.com/content/10/S1/A164

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Introduction

Marfan syndrome (MS) is a rare connective tissue disease (incidence 1/10,000 persons) caused by mutations of the fibrillin-1 gene. Although it can affect several systems, the cardiovascular system is the major source of morbidity and mortality. As a result, Cardiovascular Magnetic Resonance (CMR) can play an important role in identifying and evaluating cardiovascular manifestations in this population.

Methods

The study population consisted of 120 consecutive Marfan patients referred to our centre between January 2003 and June 2007 including 77 males (64%) and 43 females (36%), with a mean age of 34.9 years. We evaluated thoracic aortic dimensions at different segments, and assessed for structural abnormalities of the aorta, aortic arch branches, and main pulmonary artery. Aortic and mitral valves anatomy and function were also evaluated.

In one study both valves were not evaluated due to incomplete study caused by claustrophobia, while in another study the mitral valve was not assessed due to technical artifact.

Although it was not the main objective of this study, assessment of chest wall deformities and pulmonary abnormalities was also performed.

Results

The aortic root was dilated in 69 patients (57.5%), average diameter 44.1 cm, the ascending aorta was dilated in 23 patients (19.2%) average diameter 32.7 cm, the aortic arch was dilated in 19 patients (15.8%), average diameter 23.7 cm, and the descending aorta was dilated in 18 patients (15.0%), average diameter 23.8 cm. The arch vessels were dilated or aneurysmatic in 9 patients (7.5%), and the abdominal aorta was involved in 9 patients (7.5%). The main pulmonary artery was dilated in 16 patients (13.3%). Aortic dissection was noted in 9 patients (7.5%): one acute type A dissection, 7 chronic type A dissections, and 1 chronic type B dissection. Intramural haematomas were seen in 2 patients.

There was aortic regurgitation in 50 out of the 101 patients with native aortic valve (49.5%), which was moderate or severe in 14 patients (13.9%). The native aortic valve was bicuspid in 4 patients. Of the 116 patients with identifiable native mitral valve, 43 patients (37.1%) met criteria for mitral valve prolapse. Mitral regurgitation was identified in 22 patients (19.0%), which was moderate or severe in 8 patients (6.9%).

Fifty three patients (44.2%) had pectus carinatum and 13 patients (10.8%) had pectus excavatum. Scoliosis was present in half of the patients, being moderate or severe in 32 patients (26.7%). Finally, apical bullae were detected in 4 patients and a pneumothorax was detected in one patient.

Conclusion

Due to its wide field of view and excellent image quality, CMR has become an important tool in screening and evaluating patients with MS. This study shows a high preva-

lence of aortic complications in this group of patients. Valvular involvement was also frequent, warranting regular follow-up in this population. In addition, there was a high prevalence of chest deformities in this group of patients which may be helpful to clarify the diagnosis of this syndrome in patients with a borderline phenotype. The prevalence of chest wall deformities in this population highlights the importance of CMR in assessing the cardiovascular system of these patients in whom echocardiography is likely to be difficult.

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