

# Age Relation of Aortic Wall Compliance in Marfan syndrome: Evaluation with Velocity-Encoded MRI

J. J. Westenberg<sup>1</sup>, A. J. Scholte<sup>2</sup>, Z. Vaskova<sup>3</sup>, R. J. van der Geest<sup>1</sup>, M. Groenink<sup>4</sup>, G. Labadie<sup>1</sup>, P. J. van den Boogaard<sup>1</sup>, T. R. Radonic<sup>4</sup>, Y. Hilhorst-Hofstee<sup>5</sup>, L. J. Kroft<sup>1</sup>, A. de Roos<sup>1</sup>, and J. H. Reiber<sup>1</sup>

<sup>1</sup>Radiology, Leiden University Medical Center, Leiden, Zuid-Holland, Netherlands, <sup>2</sup>Cardiology, Leiden University Medical Center, <sup>3</sup>Radiology, St. Anne's University Hospital, Brno, Czech Republic, <sup>4</sup>Cardiology, Academic Medical Center, Amsterdam, Netherlands, <sup>5</sup>Clinical Genetics, Leiden University Medical Center

## Introduction

In patients with Marfan syndrome, aortic root dilatation due to reduced wall compliance often leads to dissection which is life threatening. Because wall compliance also decreases with age, the purpose of this study is to describe age-relation of wall compliance, expressed in Pulse Wave Velocity (PWV), Distensibility (Dist) and Stiffness Index (SI), with Velocity-Encoded (VE) MRI in Marfan syndrome.

## Methods

Twenty-five patients (mean age 36±14 years, range 18-63 years, 13 men) with Marfan syndrome and twenty-five age-matched healthy volunteers were examined on 1.5T Philips Achieva MRI (Philips Medical Systems, Best, The Netherlands). Through-plane VE MRI with  $V_{enc} = 150$  cm/s was performed perpendicular to the ascending and descending aorta at the level transecting the pulmonary trunk. A second acquisition ( $V_{enc} = 100$  cm/s) was performed at the abdominal aorta. PWV was determined for the aortic arch (AA), distal aorta (DA) and total aorta using the transit-time method described by Grotenhuis.<sup>1</sup> Brachial-cuff systolic and diastolic blood pressure (BP) was obtained. Distensibility (= luminal area change / (diastolic luminal area × pulse pressure)) and Stiffness Index (=  $\ln[(BP_{Systolic} / BP_{Diastolic}) / (\text{diameter change} / \text{diastolic diameter})]$ ) were determined at the ascending aorta in the gradient-echo magnitude images. PWV, Dist and SI were compared in Marfan and controls using paired t-tests. Age relation was determined by linear regression.

## Results

	PWV <sub>AA</sub> m/s	PWV <sub>DA</sub> m/s	PWV <sub>tot</sub> m/s	Dist mmHg <sup>-1</sup>	SI (-)
<b>Marfan</b>	5.6 ± 1.4	6.4 ± 2.4	5.9 ± 1.6	446 ± 262	3.0 ± 0.5
<b>Control</b>	4.8 ± 0.9	5.0 ± 1.5	4.9 ± 1.1	674 ± 426	2.7 ± 0.6
<b>p-value</b>	0.004	0.003	<0.001	0.02	0.07

Table 1. Aortic wall compliance in Marfan versus healthy controls.

		A ± Standard Error	B ± SE	Pearson R
PWV <sub>AA</sub>	Marfan	0.07±0.01 m·s <sup>-1</sup> ·year <sup>-1</sup> (p<0.001)	3.25±0.56 m/s (p<0.001)	0.69 (p<0.001)
	Control	0.03±0.01 m·s <sup>-1</sup> ·year <sup>-1</sup> (p=0.02)	3.70±0.46 m/s (p<0.001)	0.47 (p=0.02)
PWV <sub>DA</sub>	Marfan	0.08±0.03 m·s <sup>-1</sup> ·year <sup>-1</sup> (p<0.001)	3.52±1.22 m/s (p=0.008)	0.47 (p=0.02)
	Control	0.10±0.01 m·s <sup>-1</sup> ·year <sup>-1</sup> (p<0.001)	1.47±0.28 m/s (p<0.001)	0.94 (p<0.001)
PWV <sub>tot</sub>	Marfan	0.07±0.02 m·s <sup>-1</sup> ·year <sup>-1</sup> (p=0.001)	3.32±0.72 m/s (p<0.001)	0.63 (p=0.001)
	Control	0.07±0.01 m·s <sup>-1</sup> ·year <sup>-1</sup> (p<0.001)	2.32±0.23 m/s (p<0.001)	0.93 (p<0.001)
Dist	Marfan	-6.3±3.6 mmHg <sup>-1</sup> ·year <sup>-1</sup> (p=0.09)	676±139 mmHg <sup>-1</sup> (p<0.001)	0.35 (p=0.09)
	Control	-11.4±5.7 mmHg <sup>-1</sup> ·year <sup>-1</sup> (p=0.06)	1088±240 mmHg <sup>-1</sup> (p<0.001)	0.39 (p=0.06)
SI	Marfan	0.011±0.007 year <sup>-1</sup> (p=0.13)	2.5±0.3 (p<0.001)	0.31 (p=0.13)
	Control	0.019±0.008 year <sup>-1</sup> (p=0.03)	2.0±0.3 (p<0.001)	0.45 (p=0.03)

Table 2. Age relation of aortic wall compliance in Marfan versus healthy controls by linear regression  $A \times \text{age} + B$ .

In Marfan, PWV is generally increased at AA, DA and total aorta. Dist is decreased. Only the SI is not statistically significantly different. PWV in AA, DA and total aorta shows significant increase with age: 7cm/s to 10cm/s increase in PWV per year, except PWV<sub>AA</sub> in controls which shows only shows 3cm/s increase per year. Age relation of Dist was not statistically proven for Marfan or controls and SI increased with age only statistically significantly in controls and not in Marfan. SI and Dist rely on accurate assessment of aortic luminal area. Spatial resolution may not be adequate to determine significant differences between patients and controls. The higher increase in PWV per year in the aortic arch in Marfan compared to controls and compared to the distal part of the aorta may account for aortic root dilatation in patients with Marfan syndrome.

## Conclusions

Age-related increase in aortic wall compliance can be studied with Velocity-Encoded MRI. Patients with Marfan syndrome present with increased Pulse Wave Velocity and stiffness index and decreased distensibility. Age-related change in compliance is best expressed in increasing PWV.

## Acknowledgement

Funding for this study by the Netherlands Heart Foundation (Project 2006B138) is gratefully acknowledged.

## Reference

1. Grotenhuis HB et al. *J Magn Reson Imaging* 2009;30(3):521-6.

