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Enzyme replacement therapy reverses endothelial dysfunction in Fabry disease
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Fabry’s disease (FD) is X-chromosomal-linked lysosomal storage disorder and this deficiency is associated with an abnormal accumulation of glycosphingolipids leading to pathologic organ damages. In cardiovascular system, it has been demonstrated FD patients exhibit a higher incidence of cardiovascular events, but there is few report demonstrating alteration of endothelial function in FD. Moreover, it is unknown regarding the effects of α-galactosidase A enzyme replacement therapy (ERT), a sole disease-specific treatment for FD, on endothelial function.
Methods: Endothelial function was compared by the reactive hyperemia peripheral arterial tonometry index (RHI) measured by the RH-PAT system using the Endo-PAT 2000; noninvasive and repeatable monitoring device. We measured endothelial function in 27 Japanese patients with FD (n=27; aged 43±15 year-old) and age-matched healthy subjects (n=20).
Results: The amplitude of the RH-PAT signal of the FD patient was smaller than the healthy control (RHI 1.6±0.4 for FD patients, 2.2±0.6 for healthy subjects; Fig 1). This trend of endothelial dysfunction was observed even in younger patients with FD (20-30 y/o) and those male FD patients exhibited worse than the age-matched female population (Fig 2). We further compared the effects of ERT on endothelial dysfunction observed in FD patients (n=6). RHI was found to be ameliorated after ERT (1.4±0.3 (before ERT) versus 2.0±0.6 (after ERT), n=6, P=0.028) in a short period after ERT (1.6±0.9 days).
Conclusion: The present study demonstrates endothelial dysfunction observed in Japanese FD patients. Endothelial dysfunction in FD was exhibited more specifically to the FD population of young and male. ERT had beneficial effect on the endothelial dysfunction observed in FD patients.

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Clinical feasibility of carotid plaque inflammation imaging by microwave radiometry
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Purpose: Carotid plaques exhibit thermal heterogeneity mainly attributed to increased density of inflammatory cells. Microwave radiometry (MR) is a new purpose: Carotid plaques exhibit thermal heterogeneity mainly attributed to increased density of inflammatory cells. Microwave radiometry (MR) is a new method specifically to the FD population of young and male. ERT had beneficial effect on the endothelial dysfunction observed in FD patients.

Fig. 1

Results: The amplitude of the RH-PAT signal of the FD patient was smaller than the healthy control (RHI 1.6±0.4 for FD patients, 2.2±0.6 for healthy subjects; Fig 1). This trend of endothelial dysfunction was observed even in younger patients with FD (20-30 y/o) and those male FD patients exhibited worse than the age-matched female population (Fig 2). We further compared the effects of ERT on endothelial dysfunction observed in FD patients (n=6). RHI was found to be ameliorated after ERT (1.4±0.3 (before ERT) versus 2.0±0.6 (after ERT), n=6, P=0.028) in a short period after ERT (1.6±0.9 days).
Conclusion: The present study demonstrates endothelial dysfunction observed in Japanese FD patients. Endothelial dysfunction in FD was exhibited more specifically to the FD population of young and male. ERT had beneficial effect on the endothelial dysfunction observed in FD patients.

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Computational fluid dynamics analysis of the relationship between small shear stress pattern and plaque distribution at the distal left main bifurcation area
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Purpose: Atherosclerosis in distal left main coronary artery (LMA) preferentially propagates to left anterior descending artery (LAD) rather than left circumflex artery (LCX). The mechanism, however, of it has not been well investigated.
Methods: We used computational fluid dynamics (CFD) to assess the relationship between wall shear stress (WSS) and plaque distribution at distal LMA bifurcation area (n=270 segments) in patients with chest pain. Multislice computed tomography was used for the initial screening test followed by coronary angiography and virtual histology intravascular ultrasound. Vascular numerical analysis, including WSS, was performed in 6 regions: distal LMA, proximal LAD, proximal LCX, outer wall of LMA to proximal LAD (zone 1), outer wall of LMCA to proximal LCX (zone 2), and the flow divider (carina, zone 3) (Figure).

Figure 1

Results: Peak and average WSS significantly decreased in zones 1 than zone 2 or zone 3 (2.05±2.16, 4.83±3.19, 8.91±7.59, Pa=0.001; 0.98±0.94, 1.73±1.33, 3.60±2.95, Pa=0.001, respectively). Peak and average WSS were also significantly decreased in distal LMA than proximal LAD or LCX (4.10±2.55, 5.71±4.79, 7.29±5.23, Pa=0.003; 1.41±1.00, 2.21±1.94, 2.84±2.11, P=0.002, Pa, respectively). Persistent lower peak WSS in zone 1 was observed irrespective of plaque type, bifurcation angle, and LCX vessel size (Figure).
Conclusions: Regardless of bifurcation angle degree, plaque type, and LCX vessel size, a significantly lower WSS pattern at zone 1 may contribute to the mechanism of predominant plaque distribution more often found originating in LMCA and distributing to proximal LAD rather than LCX.