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Coronary computed tomography angiography and echocardiography in children with homozygous familial hypercholesterolemia

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Introduction
Homozygous familial hypercholesterolemia (hoFH) is a rare genetic disease, hallmarked by a lifelong exposure to very high levels of low-density lipoprotein cholesterol (LDL-C). Untreated, patients can experience their first cardiovascular event in the first decade of life. Lipoprotein apheresis (LA) may delay this process, but the required intensity of this invasive therapy is under debate. Early detection and monitoring of subclinical atherosclerosis in these patients is therefore extremely important. We compared the diagnostic yield of low dose coronary computed tomography angiography (cCTA) compared to echocardiography in assessing subclinical atherosclerosis.

Methods
For this single-center cross-sectional study, we included all pediatric hoFH patients that were treated with LA in Amsterdam UMC. All had undergone both echocardiography and cCTA.

Results
Six hoFH patients were included. Median ages at diagnosis, onset of LA and cardiovascular assessment (cCTA and echocardiography) were 2.6, 6.5, 10.8 and 11.1, respectively. Echocardiography revealed no signs of atherosclerosis in any of the six patients. In two patients, mild dilatation of the cardiac chambers was detected and two patients showed signs of mitral or aortic insufficiency. On cCTA, however, non-calcified plaques without stenosis were detected in four patients. In two patients calcified coronary plaques were found at the ostia of the right coronary artery (RCA) or the left main coronary artery (LMCA). Aortic root calcifications were found in two patients

Conclusion
In hoFH children, low dose cCTA is superior to echocardiography for the detection of subclinical coronary and aortic root atherosclerosis and should therefore be part of the cardiovascular monitoring in these children, on top of routine echocardiography.