

Regression and complications of z-score based giant aneurysms in a Dutch cohort of Kawasaki disease patients

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Introduction

Kawasaki disease (KD) is a pediatric vasculitis. Its main complication is the development of coronary artery aneurysms (CAA), with giant CAA at the end of the spectrum. We evaluated regression and event-free rates in a non-Asian cohort of patients with giant CAA using the current z-scores adjusted for body-surface area instead of absolute diameters.

Methods

KD patients with giant CAA (z-score ≥ 10) visiting our outpatient clinic between January 1999 and September 2015 were included. Patient characteristics and clinical details were extracted from medical records. Regression was defined as all coronary arteries having a z-score of ≤ 3 . A major adverse event was defined as cardiac death, myocardial infarction, cardiogenic shock or any coronary intervention. Regression-free and event-free rates were calculated using the Kaplan-Meier method.

Results

We included 52 patients with giant CAA of which 45 had been monitored since the acute phase. The 1-year, 2-year and 5-year regression-free rates were 0.86, 0.78 and 0.65, respectively. The 5-year, 10-year and 15-year event-free rates were 0.79, 0.75 and 0.65, respectively. Four children whose CAA would not have been classified as 'giant' based on absolute diameters instead of z-scores, had experienced an event during follow-up.

Conclusions

We found a high percentage of children in whom the lumen of giant CAA completely normalized. Four children not classified as 'giant' based on absolute diameters with z-scores of ≥ 10 , experienced a cardiac event. Hence, the use of z-scores seems to be justified.