

Giant aneurysms: a gender-specific complication of Kawasaki disease?

Kuipers I.M. (2), Dietz S.M. (1), Tacke C.E.A. (1), Koole J.C.D. (1), Hutten B.A. (3), Kuijpers T.W. (1). Department of Pediatric Hematology, immunology and infectious diseases, Emma Children's Hospital, Academic Medical Centre (AMC), Amsterdam, The Netherlands (1); Department of Pediatric Cardiology, Emma Children's Hospital, AMC, Amsterdam, The Netherlands (2); Department of Clinical Epidemiology, Biostatistics and Bioinformatics, AMC, Amsterdam, the Netherlands (3).

Background: Kawasaki disease (KD) is a pediatric vasculitis of unknown origin. Its main complication is the development of coronary artery aneurysms (CAA) with giant CAA at the end of the spectrum.

Methods: In this cohort study, we evaluated the association between patient characteristics and the development of giant CAA based on z-scores. Multivariable, multinomial logistic regression analysis was used to identify variables associated with giant CAA.

Results: A total of 301 KD patients, comprising of 216 patients without enlargement, 45 with small-sized, 19 with medium-sized, and 21 with giant CAA with all echocardiographies at our center were retrospectively included. Remarkably, 95% of patients with giant CAA were boys. In addition to 'no/late intravenous immunoglobulin (IVIG) treatment', 'male gender' (OR 15.56, 95% CI 1.86-130.07), 'age <1 year' (OR 8.06, 95% CI 2.56-25.35), and 'IVIG re-treatment (6.38, 95% CI 1.86-21.88)' were significantly associated with an increased risk of giant CAA, with patients without enlargement as reference. Compared to patients medium-sized CAA, 'IVIG re-treatment' was significantly associated with giant CAA. The majority of giant CAA continued to increase in size during the first 40 days.

Conclusions: We identified risk factors associated with an increased risk of giant CAA. The difference in variables between the giant CAA group and the other CAA subgroups suggests a separation between patients with the treatment-resistant giant CAA and the other IVIG-responsive patients, in which gender may be factored as a most relevant genetic trait. The increase in size during the first 2 months indicates the need for repeated echocardiography.