

Two Studies and One Commentary on Kawasaki Disease and Its Treatment

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While we may not yet understand the pathophysiology underlying the development of Kawasaki disease, we are making progress in its treatment when it comes to lowering the risk of coronary artery aneurysms. Treatment with intravenous immunoglobulin (IVIG) has become the first line for Kawasaki disease, but what if IVIG does not work? This week, we release two studies looking at different adjunctive therapies to reduce the rate of or the worsening of coronary artery aneurysms once they occur. The

first of these studies, conducted by Dionne et al. ([10.1542/peds.2018-3341](#)) was a retrospective multicenter study of children detected with aneurysms at diagnosis and who were assigned to get IVIG or IVIG plus a corticosteroid or IVIG plus infliximab, a monoclonal anti-tumor necrosis factor antibody that serves as an anti-inflammatory agent. The results demonstrated that both adding corticosteroids or infliximab made a difference in the progression of coronary aneurysms in the 121 children studied compared to just IVIG treatment. The limitation of this study is that it was not randomized, nor prospective, but opens the door to such studies now being done as a result of wanting to determine the best intensification or adjunctive therapy needed to reduce the complications of this disease.

The second of these new Kawasaki studies opted to use not infliximab but etanercept as the adjunctive agent in a randomized controlled trial. Portman et al. ([10.1542/peds.2018-3675](#)) in a multicenter trial randomized patients to receive IVIG plus either etanercept or placebo. Etanercept is a TNF-alpha receptor antagonist binds to soluble and non-tissue associated TNF) making it preferable in preventing anti-drug antibodies to IVIG to form). The outcome was IVIG resistance as captured by persistence of symptoms and evidence of coronary artery aneurysm growth. The results showed that there was no statistical difference in lowering IVIG resistance in the etanercept group compared to placebo and that the size of identified coronary aneurysms was less in those with baseline dilation over time but really only in some subgroups such as those children over a year of age or of African American subjects. No adverse side effects were noted in either group of the 101 patients randomized.

So is etanercept the new preferred adjunctive therapy especially when there is no response to IVIG alone? We asked Kawasaki disease experts Drs David Burgner and Jane Newburger ([10.1542/peds.2019-0912](#)) to share their thoughts in an accompanying commentary. They note that this study may have been underpowered to show significant decreases in IVIG resistance and that it may be useful in certain subgroups but much larger samples are needed. There is certainly a need to continue to study adjunctive therapy for IVIG resistance and coronary artery dilation, and hopefully these two studies will form the basis upon which future larger randomized controlled trials are performed as a means of reducing the complications of Kawasaki disease.

- [Aspirin Dose and Prevention of Coronary Abnormalities in Kawasaki Disease](#)
- [Asymptomatic Kawasaki Disease in a 3-Month-Old Infant](#)
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