**Is There A Role For Corticosteroids In The Treatment Of Kawasaki Disease?**

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May 31, 2016

Kawasaki Disease (KD) is a vasculitis which predominantly affects children. While the cause of KD is poorly understood Coronary Artery Abnormalities (CAA) can occur and are the most feared sequelae of the disease. CAA development can start to manifest within 7-9 days of the commencement of fever. It is estimated that 15-20% of children with KD are refractory to their first treatment with IVIG or have recrudescent fever requiring a “Rescue Therapy”. Patients who do not respond to their first treatment with IVIG or have recrudescent fevers are considered to be at a higher risk for CAAs. Initial therapy for patients with KD is Intravenous Immunoglobulin (IVIG). If fevers persist beyond 24 hours after initial IVIG treatment or fevers become recrudescent then a second medication is administered (IVIG, corticosteroids, cytotoxic agents, infliximab, or plasma exchange). In America retreatment with IVIG is the accepted practice as per The American Heart Association recommendations stated in 2004.

Corticosteroids have been useful in treatment in other vasculitides but due to one study showing that a prolonged course of steroids as a monotherapy for KD had a high incidence of CAA (Newburger 2004), corticosteroids have not been used routinely in KD. Other studies since have shown that corticosteroid may have a therapeutic role in high risk patients. In patients with high risk KD does the addition of corticosteroids to IVIG as first line or second line therapy in comparison with IVIG monotherapy decrease the risk of CAAs?

A literature search was conducted on PubMed to help answer the aforementioned question. Keywords included “Kawasaki Disease” AND “IVIG” AND “Corticosteroids”. Three relevant articles were found that further discuss the potential role of corticosteroids in KD. Kobayashi et al. (2013) was a multicenter, unmatched, retrospective cohort study which showed that prednisolone along with a taper combined with IVIG as rescue therapy had lower odds of requiring additional rescue therapy after treatment in comparison to IVIG monotherapy. It also showed a lower odds of developing CAAs when compared to IVIG alone. The limitations of this study included that it was completed on a solely Japanese population and that due to it being a retrospective study residual confounding factors may have been present obscuring the validity of the study. Additionally Z-scores for CAAs were not used and instead CAAs may have been underestimated as a Japanese criteria was used in place. Ogata et al. (2012) was a multicenter prospective randomized powered pilot study which used the Egami Scoring system to determine responsiveness to initial treatment with IVIG. This study suggests that IVIG in combination with intravenous methylprednisolone (IVMP) as a single pulse is a safe and effective first line therapy to be used in patients considered high risk (noted as having a score of ≥3). Additionally it suggested that the addition of IVMP to IVIG treatment decreased the need for additional rescue therapy and decreased the risk of CAA formation. Again, this study was comprised of a single ethnic patient population in Japan. Additionally, while the Egami Scoring is more sensitive and specific in the Japanese population it has been shown to have a lower sensitivity and specificity in a study completed in an America. Additional studies in a multiracial population are warranted to see if these results can be applied to all pediatric populations with KD or to see if other ethnicities/races should stratified differently. Kobayashi et al. (2012) was a prospective, randomized, multicenter, open label trial with blinded endpoints which suggested that combination treatment with IVIG and IV Corticosteroids with a taper as initial therapy may decrease the need for further rescue therapies, may improve inflammatory markers, may decrease the total days of fever and ultimately may decreased CAAs. Again, this study was completed in Japan with an exclusively Japanese population using the Egami Scoring system. Additionally studies showing the safety of prolonged steroid treatment with taper would be warranted.

In conclusion, these studies are suggestive that there is likely a role for corticosteroids in high risk or refractory patients with KD. Additional multi-centered, multiracial, randomized, controlled, double blinded studies with varying steroid treatment regimens (both as primary and rescue therapy) would be useful to assess optimal timing and dosing of corticosteroids that would improve overall outcome in CAA incidence and persistence. Further, a scoring system that could be applied to a multiracial population would improve assessment of patients who are likely to be refractory to first line treatment and may benefit from corticosteroids. Being that a single pulse of steroids has been shown to be relatively safe and overall has the potential for cardiac protective effects, a single pulse of steroids in high risk groups may be considered during initial or subsequent therapy as an adjunct to IVIG.

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