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Incidence of coronary aneurysms in children with Kawasaki disease after early treatment with IVIG and aspirin: Report of 55 cases
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Introduction.— Kawasaki disease (KD) is the most common cause of acquired pediatric heart disease. It is associated with coronary artery aneurysms in a variable percentage of cases, with an overall case-fatality rate of 0.5–2.8%. We describe the incidence of coronary artery aneurysms in 55 patients with Kawasaki disease after early treatment with IVIG and aspirin.

Methods.— We performed a single-centre retrospective review of 54 patients who met the diagnostic criteria for Kawasaki Disease between 2002 and 2012. Early treatment with IVIG (2 g/kg) and aspirin (initial dose of 100 mg/kg/day) was started as soon as the diagnosis was suspected, always, within the first 10 days of fever.

Results.— The review included 55 patients, 34 males and 21 females. 70% of cases occurred prior to the age of age 3, and 67% of those in the first 6 months of life, with a peak incidence of the disease occurring in the first six months of the year. Between the third and seventh day of the disease the patients presented the following symptoms: 100% fever, 98% erythema of the oral cavity, 94% conjunctivitis, 92% changes in the skin and mucous membranes, 75% cervical adenitis, and 72% desquamation of the skin of the fingertips. In 76% of the patients treatment with IVIG and aspirin was started between 3 and 7 days after onset of the clinical manifestations, and in the 14% of the patients between 7th and the 10th day of illness. Coronary artery aneurysms were documented by echocardiography in 9% of the patients. Other authors have reported a reduction of the incidence of coronary artery abnormalities from 25% to 3–5%, with early treatment. KD cases cluster in the first 6 months of the year suggest an infectious trigger for the disease. The disease is generally benign and self-limited.

Discussion.— In our study, coronary artery aneurysms were detected in 9% of the patients. Other authors have reported a reduction of the incidence of coronary artery aneurysms from 25% to 3–5%, with early treatment. KD cases cluster in the first 6 months of the year suggest an infectious trigger for the disease. The disease is generally benign and self-limited.

Conclusion.— Early treatment with IVIG and aspirin in patients with KD may decrease the incidence of coronary artery abnormalities.

Further reading

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Co-trimoxazole monotherapy as induction therapy for localized granulomatosis with polyangiitis
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Introduction.— In a number of patients with granulomatosis with polyangiitis (GPA), disease is limited to the upper and lower airways without systemic vasculitis and organ- or life-threatening symptoms. Case reports and small series suggest that these patients may benefit from treatment with co-trimoxazole monotherapy. However, safety and efficacy have never been evaluated in a large group of patients.

Methods.— We retrospectively evaluated safety and efficacy of induction therapy with co-trimoxazole monotherapy for patients with localized GPA treated at our center between 1989 and 2012.

Results.— Forty-nine patients, 20 males and 29 females, with a mean age of 49 years (SD 18) were treated with co-trimoxazole monotherapy 800/160 mg twice daily. Forty patients were newly diagnosed with GPA and nine had a localized relapse. Forty patients were ANCA positive. With co-trimoxazole monotherapy, 35 patients attained remission. Twenty of these patients did not develop a relapse during follow-up with a median disease free survival (DFS) of 146 months (IQR 66–154). In fifteen patients, a localized (n = 12) or systemic (n = 3) relapse developed after a median DFS of 22 months (7–27), see figure 1. Ten patients had to start additional immunosuppressive therapy for refractory disease with development of systemic disease in two. Four patients had to stop co-trimoxazole due to side-effects, primarily rash and nausea. Carriers of Staphylococcus aureus at diagnosis (n = 19) had a significant (P = 0.047) shorter DFS compared to negative patients.

Figure 1

Conclusion.— Co-trimoxazole mono-therapy induces remission in a substantial number of patients with localized disease. Therefore, co-trimoxazole can be considered as a first-line therapy in patients with localized GPA.

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Successful treatment of refractory cutaneous ulcer associated with polyarteritis nodosa with bosentan
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Introduction.— A cutaneous form of polyarteritis nodosa (PN), affecting predominantly the lower extremities, is distinguished from systemic PN