Phenotype of children with giant coronary artery aneurysms in Kawasaki Disease

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The UK Foundation for Kawasaki Disease

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BACKGROUND

Kawasaki disease (KD) is now the commonest acquired heart disease in the western world (8/100,000 children < 5 years per year in the UK).¹ Coronary artery aneurysms (CAA) occur in 19% of untreated patients, 1.6% of which are giant (GCAA, Z score ≥ 10).

Figure 1. Symptoms of Kawasaki Disease

METHODS

Our Heartsuite™ database from January 2000 to March 2018 revealed 7 children with GCAA, including their biomarkers, demographic data, urbanicity, and clinical outcome. Data is presented as median (range).

RESULTS

- At diagnosis, there were 4 boys, median age was 136 days (79-790) and weight 7 kg (5-12).
- All children had fewer than 4 clinical features, diagnosis made late at 13 days (7-26).
- Z score for right CA +13 (11-31), left main CA+14 (7-20), left anterior descending CA +15 (7-19), 5 having myocarditis or effusion.
- 5 patients received intravenous immunoglobulin (IVIG) and aspirin, and aspirin, 4 of which required additional IVIG, and 3 had infliximab or steroids.
- Outcome over 9 years (1-19) has shown no deaths, two with a current CAA Z score of <2.5

Figure 2². Annual incidence of Kawasaki disease by year onset.

CONCLUSION

Young infants of infants median 4 months old, with few features of KD is associated with giant CAA. In addition, CSF analysis can be confusing and analysis of blood shows those who are most likely to benefit from additional therapy. We await the long-term consequence of this important condition.

REFERENCES

Please scan the QR code for references.