Congenital Cardiology Solutions

SPECTRUM OF CONGENITAL HEART DISEASE AND OUTCOMES IN CHILDREN WITH TURNER SYNDROME: A SINGLE-CENTER EXPERIENCE

Poster Contributions
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Background: Turner syndrome (TS) is a common genetic abnormality that affects 1 in 2500. Despite the known association between TS and congenital heart disease (CHD), the outcomes following surgical repair have not been well described. We reviewed the spectrum of CHD associated with TS within our center and provide outcomes following aortic coarctation (CoA) repair or staged palliation of hypoplastic left heart syndrome (HLHS).

Methods: The records of patients diagnosed with TS at Children's Hospital of Wisconsin from 1999 - 2011 were reviewed. Those patients with TS and CoA or HLHS were then selected and compared against all non-TS patients undergoing like repairs.

Results: Of the 173 patients with TS, 77 (44.5%) were found to have CHD, with left sided obstructive lesions being the most common. However, the spectrum of CHD was wide and included systemic and pulmonary venous abnormalities as well as coronary arteries abnormalities. 21 TS patients were compared to all non-TS patients with isolated CoA undergoing repair during a similar time period. TS patients were statistically more likely to receive a subclavian flap repair, have a longer aortic cross clamp time and have a longer length of hospital stay following repair. Between age matched (age < 60 days) cohorts, the incidence of subclavian flap repair was not statistically different but both aortic cross clamp time and post-operative length of stay remained longer in the TS cohort. In long term follow up (8.8 +/- 9.1yrs) 17% of TS patients had residual hypertension, but no patient required re-intervention and there were no deaths. Finally, amongst 4 patients with HLHS, 3 died within the first year, compared to our previously published 93% survival for standard risk single ventricle patients.

Conclusions: We conclude that the spectrum of CHD seen with TS is not restricted to left sided obstructive heart lesions and included both arterial and venous abnormalities. Additionally, patients with TS undergoing surgical CoA repair may have a more challenging early peri-operative course, but their long term outcomes are similar to their non-TS peers. Finally, the combination of TS and HLHS remains a very challenging population with generally poor survival.