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Title: Preoperative left ventricular dysfunction is a risk factor for late death in paediatric patients after Rheumatic Heart Disease surgery for isolated aortic valve disease

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Background & Aims: For young people with rheumatic aortic valvular disease, evidence based indications for the threshold for cardiac surgical intervention remain sparse, with little known about mid or long term outcomes of left ventricular (LV) function.

Methods: Single institution retrospective cohort study of patients < 18 years with Rheumatic Heart Disease (RHD) who underwent surgical intervention for isolated rheumatic aortic valve disease between 2000 and 2019. Cohort of 38 patients 8-16 years old (median age 13 years), weight 27-157 kg (median 78kg); 95% Māori or Pacific Islander ethnicity. Pre-operative, 'intermediate' follow up (6-24 months post-operative) and 'late' follow up (> 2 years post operatively) data was collected. For those who underwent reoperation, late follow up data was immediately prior to their second surgery. LV function data were collected. Impaired LV function was defined as LV ejection fraction (LVEF) \leq 50% or LV fractional shortening (LVFS) \leq 25%.

Results: Initial operation was aortic valve repair in 6/38 (16%) and replacement in 32/38 (84%): homograft valve in 19/32 and prosthetic valve in 13/32. 30 day mortality was zero.

Preoperatively, 6/38 (16%) had impaired LV function. Intermediate follow up data was available for 30/38 (79%). There were 2 deaths and 2/38 (6%) had impaired LV function.

Late follow up data was available for 32 patients (84%), mean 6.6 years post initial surgery (range 2.5-15 years). There were 4 further deaths and 13/32 (40%) had impaired function. Of the overall cohort, 40 % (15/38) underwent reoperation (median time from initial surgery of 6 years (IQR 5.5-8.5years)), and of these 14/15 (93%) had initial aortic homograft replacement.

All-cause mortality was 16% (6/38), median age at death of 21 years (IQR 15-25), and median time from initial surgery of 6 years (IQR 1-10). Of those with normal pre-operative LV function, at late follow up none had died, but 37% (12/32) had impaired LV function and 3 were lost to follow up. Pre-operative LV dysfunction was associated with a 10-fold increase in the risk of death at late follow up (hazard ratio = 10.2, 95% CI: 1.7, 60.1 (p 0.01)), and this increased to 15-fold when adjusting for increasing BMI (hazard ratio 15.0, 95% CI: 1.7, 132 (p 0.01)).

Other major complications were common; rates of infective endocarditis were 2.4% per patient year of follow up. Thromboembolic events occurred at 3.8% per patient year of follow up, all after prosthetic replacement.

Conclusions: This study demonstrated that for paediatric patients undergoing surgical intervention for isolated rheumatic aortic valve disease preoperative impaired LV function is associated with an increased risk of late death. Moreover, that risk increases further when adjusting for BMI. Overall there was a high rate of reoperation for aortic homografts, high morbidity and moderate mortality in the follow up period.