Hypoplastic Left Heart Syndrome: an audit of clinical outcomes in patients surviving beyond the age of 10 years

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Introduction: Hypoplastic left heart syndrome (HLHS) has seen dramatic shifts in management over the past two decades with increasing numbers of children surviving surgical palliation. However, little research has been conducted into the long-term outcome functional status of these individuals.

Methods: Audit of clinical outcome beyond the age of nine years at a single tertiary centre based on care record review.

Results: A total of 105 patients underwent staged Norwood palliation at our centre more than nine years ago (pre 2003) of whom 58 are still alive. The median age at time of audit was 12 years (range 9-17). All case records were available for review. The median height lay on the 9th centile, and median weight on the 25th centile. Exercise tolerance was subjectively reduced in 42 (72%) of our patients and formal ETT/CPET, where performed, revealed 45-85% predicted V-max across the population. 39 patients (67%) were referred to medical services with extra-cardiac symptoms (72% of these with more than one complaint. 26% of the total population complained of respiratory symptoms including wheeze, chronic cough and recurrent RTI. 23 patients (40%) had concerns over their educational performance, also, 4 (7%) had been diagnosed formally with ADHD and 4 (7%) with an autistic spectrum disorder. 7% of patients suffered some form of imaging-confirmed cerebral infarction, 3 (5%) of our patients were registered partially sighted and 2 patients (3%) were registered as deaf. Only 9 patients (16%) had no documented referral to medical/psychiatric services, however even within this group, 6 had reduced exercise tolerance and 4 of these had made an application for, or were in receipt of, a Disability Living Allowance.

Conclusions: Survivors of HLHS surgery appear to be at risk for adverse clinical outcome with adverse developmental and psychological outcome particular causes for concern. We aim to follow up this pilot study with further controlled studies to elucidate the impact of HLHS on quality of life in patients surviving to adolescence and beyond.