

Outcomes following general anaesthesia in Paediatric Hypertrophic Cardiomyopathy

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Introduction (or Basis or Objectives):

Hypertrophic cardiomyopathy (HCM) is the second most common cause of cardiomyopathy in childhood. These patients have historically been considered to be high risk candidates for general anaesthesia, however there is currently a paucity of evidence regarding the safety of anaesthesia and peri-operative outcomes in this population.

Methods:

A retrospective single centre cohort study of all paediatric patients (< 18 years) with hypertrophic cardiomyopathy undergoing general anaesthesia (GA) between 2000-2016 was performed to assess peri-anaesthetic outcomes. Patients undergoing cardiac surgery were excluded. Primary outcome was peri-operative mortality or need for re-hospitalisation within 30 days of anaesthesia.

Results:

86 patients aged 0-18 years (median 12.4 years) with HCM underwent 164 separate general anaesthetic procedures over the study period. General anaesthetic was most commonly required for a non-surgical cardiac procedure (n = 87, 53%), followed by a general surgical, radiology or orthopaedic procedure (n=17, n=16, n=12 respectively). The underlying aetiology was heterogenous and included sarcomeric/idiopathic disease (56%), a malformation syndrome (26%), inborn error of metabolism (12%) or neuromuscular disease (4%). Maximal wall thickness (MWT) ranged from 4-49mm. 23 patients (1%) had severe left ventricular hypertrophy (MWT >30mm or Z score >+6) and 47 patients (30.5%) had left ventricular outflow tract obstruction at rest. The majority of patients (n=143, 77%) had no complications. Length of stay ranged from 0-21 days although two thirds of patients (n=104, 65%) were performed as day-cases. 20 patients (12%) had minor peri-anaesthetic complications which included intra-operative bradycardia or hypotension requiring treatment. 1 patient (0.6% of GA procedures) experienced a cardiac arrest during anaesthetic induction which required extra-corporeal membrane oxygenation (ECMO) supported cardiopulmonary resuscitation. The presence of left ventricular systolic impairment was the only clinical feature associated with a peri-anaesthetic complication.

Conclusions:

We report the largest series of paediatric HCM patients undergoing general anaesthesia to date with a low incidence of minor complications (12%) and a mortality rate of 0.6%. In an experienced centre, paediatric patients with HCM can be anaesthetised with a low risk of adverse events. Future studies are required to identify clinical features that may predict anaesthetic risk.