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Clinical features and outcomes of Childhood Hypertrophic Cardiomyopathy: a retrospective study in the United Kingdom

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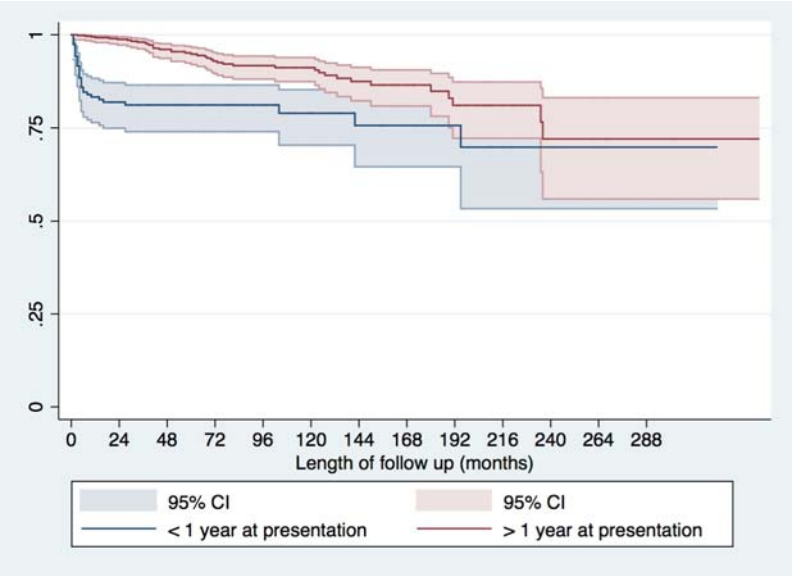
Objectives:
Describe the clinical features and outcomes of Childhood Hypertrophic Cardiomyopathy (HCM) in a national cohort.

Methods:
A retrospective, multi-centre cohort of children diagnosed with HCM in the United Kingdom (UK) since 1980 was formed from 13 paediatric cardiac centres. Anonymised, non-invasive clinical data were collected.



Results:
687 patients with childhood HCM were identified with a median age at presentation of 5.2 years (range 0-16). 23% (n=159) of patients presented during infancy. The underlying aetiology was varied including idiopathic (n=433, 63%), Noonan's syndrome (n-121, 17%), Friedreich's ataxia (n=58, 8%) and inborn errors of metabolism (n=64, 9%). Those presenting under 1 year of age were more likely to have a diagnosis of Noonan's syndrome (39% vs 11%, p = 0.000*) or inborn error of metabolism (18% vs 6.4% p =0.000*).
The majority of patients were asymptomatic, in NYHA/Ross class I at presentation (n=516, 75.4%), however heart failure symptoms were more common in infants (16% vs 2.5%, p =0.000*). A history of unexplained syncope (n=38, 5.6%) or previous aborted cardiac arrest (n=24, 5.6%) was uncommon. Overall the prognosis was good, survival without death or transplant was 91.7% (89.1 – 93.7%) at 5 years. Children diagnosed during infancy or with an inborn error of metabolism had a worse prognosis with a survival of 81% and 82% at 1 year respectively. Cause of death was most commonly sudden cardiac death (n=20) followed by non-cardiovascular causes (n=17) and congestive cardiac failure (n=12). However, sudden cardiac death or an equivalent event (aborted cardiac arrest or appropriate Implantable Cardioverter Defibrillator therapy) was rare, occurring at a rate of 0.012 events/person year follow up.

Conclusions:
This national study of childhood HCM describes a heterogeneous cohort whose outcomes are dependent on underlying aetiology and age of presentation. Further studies are needed to systematically investigate risk factors for prognosis in this patient group.

Legend:
Kaplan Meier curve for survival stratified according to age of presentation. Log rank test $p < 0.0001$