

2 THE RELATIONSHIP BETWEEN LEFT VENTRICULAR OUTFLOW TRACT GRADIENT AND SUDDEN CARDIAC DEATH IN CHILDHOOD HYPERTROPHIC CARDIOMYOPATHY

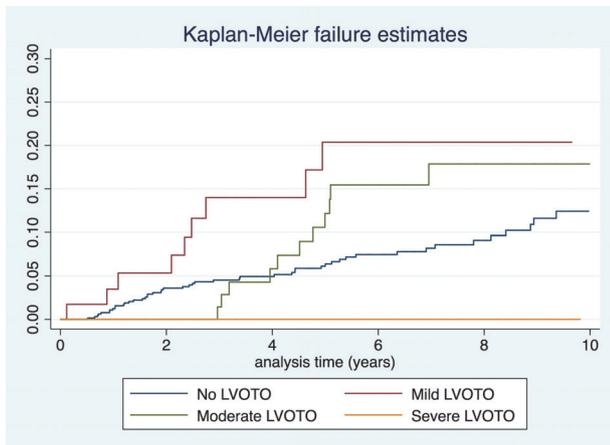
Gabrielle Norrish. *Institute of Cardiovascular Science*

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Introduction The most common mode of death in childhood hypertrophic cardiomyopathy (HCM) is sudden cardiac death (SCD). Left ventricular outflow tract (LVOT) obstruction is an established risk factor for SCD in adults with the disease. In contrast, the prognostic implications of left ventricular outflow tract obstruction (LVOTO) in childhood disease is unclear, with recent studies suggesting that it may have an inverse relationship with the risk of SCD. The aim of this study was to explore the role of LVOTO and the risk of SCD in childhood HCM.

Methods A multi-centre, retrospective, longitudinal cohort of 871 children (diagnosed with HCM <16 years of age) was used to explore the relationship between SCD and LVOTO (LVOT gradient ≥ 30 mmHg).

Results 189 patients (23%) had LVOTO, which was mild (30-50mmHg), moderate (50-100mmHg) or severe (>100mmHg) in 58 (6.7%), 98 (11.3%) and 33 (3.8%), respectively. The risk of SCD showed an inverse relation to LVOT gradient severity compared to those with no obstruction: mild HR 1.75 (95% CI 0.89-3.44), moderate HR 1.04 (95% 0.55-1.98), and severe HR 0.7 (0.36-1.35) [figure 1]. On univariable analysis [table 1] LVOTO was associated with heart failure symptoms (NYHA>1) [p <0.001], maximal wall thickness (MWT) [p <0.001], left atrial (LA) diameter [p <0.001], and future myectomy occurring during follow up [p <0.001]. The



Abstract 2 Figure 1

Abstract 2 Table 1 Demographics and clinical characteristics by LVOT gradient

| | <30mmHg (n=682) | 30- 50mmHg (n=58) | 50- 100mmHg (n=98) | >=100mmHg (n=33) | P value |
|-----------------------------------|----------------------|-------------------------|--------------------------|----------------------|------------|
| B Blocker therapy | 288 (42.3%) | 30 (51.7%) | 50 (51.6%) | 19 (57.6%) | 0.085 |
| NYHA>1 | 137 (20.5%) | 17 (29.3%) | 37 (37.8%) | 14 (43.8%) | <0.001 |
| NSVT | 39 (6.8%) | 5 (10.4%) | 7 (8.5%) | 2 (8.7%) | 0.771 |
| Unexplained syncope | 70 (10.3%) | 9 (15.5%) | 6 (6.1%) | 4 (12.1%) | 0.296 |
| Z score MWT | 10.3 (+/- 6.6) | 15.1 (+/-7.6) | 15 (+/-8.1) | 16.6 (+/-8.3) | <0.001 |
| Z score LA | 1.7 (+/-2.3) | 2.8 (+/-2.4) | 3 (+/-2.4) | 3.4 (+/-2.5) | <0.001 |
| Myectomy during follow up | 20 (2.9%) | 9 (15.5%) | 29 (29.9%) | 12 (36.4%) | <0.001 |
| SCD event | 54 (7.9%) | 11 (18.9%) | 13 (13.2%) | 1 (3.0%) | 0.009 |
| Incidence of SCD/ 100 pt years | 1.40 (1.06- 1.80) | 3.5 (1.90- 6.30) | 2.08 (1.21- 3.58) | 0.42 (0.06- 2.98) | 0.219 |

inverse relationship observed was not altered by the presence or absence of other traditional risk factors.

Conclusions LVOT gradient has a complex relationship with the risk of SCD in childhood with multiple contributing factors. The pathophysiological mechanisms behind this observation need further exploration, which may be limited by low patient numbers.

Conflict of Interest Nil

3 RESIDENTIAL EXPOSURE TO FINE PARTICULATE MATTER AIR POLLUTION IS ASSOCIATED WITH IMPAIRED CARDIAC PHENOTYPES IN DILATED CARDIOMYOPATHY

¹Upasana Tayal, ¹Daniela Fecht, ¹Marc Chadeau, ²John Gulliver, ¹James Ware, ³Stuart Cook, ¹Sanjay Prasad. ¹Imperial College London; ²University of Leicester; ³National University of Singapore

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Background Air pollution might contribute to adverse ventricular remodelling in healthy populations. A recent study on a cohort of 500,000 participants (UK Biobank) showed that residential exposure to particulate matter with aerodynamic diameter <2.5 μ m (PM2.5) and nitrogen dioxide (NO2) was associated with cardiac chamber dilatation and increased left ventricular mass.

Dilated cardiomyopathy (DCM) has marked structural and functional phenotypic heterogeneity. The biological basis for this is undefined, but environmental factors are plausible phenotypic modifiers.

Abstract 3 Table 1 Hazard ratio for primary composite outcome and exposure to small particle pollutants. Volumes and masses are indexed to body surface area. Hazard ratios presented with 95% confidence intervals

| Exposure | Absolute left ventricular ejection fraction (%) | Indexed left ventricular mass (g/m ²) | Indexed left ventricular end diastolic volume (ml/m ²) | Absolute right ventricular ejection fraction (%) | Indexed right ventricular end diastolic volume (ml/m ²) |
|---|---|---|--|--|---|
| PM2.5 per 1 unit (μ g/m ³) | -0.6 (-1.1 to 0.0), p=0.05 | 1.4 (0.18 to 2.60), p=0.02 | 0.39 (-1.3 to 2.1), p=0.64 | -0.03 (-0.7 to 0.6), p=0.93 | -1.2 (-2.3 to -0.03), p=0.04 |
| NO2 per 1 unit (μ g/m ³) | -0.1 (-0.2 to -0.04), p= 0.004 | 0.27 (0.10 to 0.43), p=0.001 | 0.20 (-0.03 to 0.4), p=0.09 | -0.03 (-0.11 to -0.06), p=0.52 | -0.06 (-0.22 to 0.09), p= 0.42 |