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What is the best wall thickness measure for indicating risk of sudden death in childhood hypertrophic cardiomyopathy?

Östman-Smith I. (1), Sjöberg G. (2), Larsson P. (3), Rydberg A. (4), Fernlund E. (5)
Queen Silvia Chuildren´s Hospital, Gotenburg, Sweden (1); Astrid Lindgren Children´s Hospital, Stockholm, Sweden (2); Academic Hospital, Uppsala, Sweden (3); University Hospital, Umeå, Sweden (4), University Hospitals, Linköping and Lund, Sweden (5)

BACKGROUND: In hypertrophic cardiomyopathy risk for sudden death is influenced by degree of cardiac hypertrophy but there is no agreement about which measure of hypertrophy is most useful in the growing child. A Z-score of >6 has been a proposed cut-off but not validated to be useful, other measures proposed have been a max wall thickness >2 cm, or septal thickness >190% of the 95th percentile value for age (SEPPER).
PATIENTS AND METHODS: Patients with a diagnosis of HCM before age 19 years attending all five regional centres of paediatric cardiology in Sweden have been studied. There were 28 patients with sudden death or re-suscitated cardiac arrest, and 99 patients with at least two years follow-up without heart-failure death (mean follow-up 11.1yrs). The wall thickness measure at last follow-up were quantified as maximall wall thickness, Z-score for maximal wall thickness (Detroit Z-score), SEPPER, or septum-to-cavity ratio, and frequency distribution histograms were used to find optimal cut-off denoting high risk, and subsequently relative risk, sensitivity and specificity was determined.
RESULTS Maximal wall thickness >2 cm had a relative risk of 5.1 [95%CI 2.1-12.6], a sensitivity of 83 [64-94)% and a specificity of 62 [52-72]; Z-score >6 a relative risk of 5.7 [3.0-10.8], sensitivity 56 [35-76]%, specificity 92 [84-96]%, whereas a lower cut-off Z-score >4.5 gave a relative risk of 9.9 [3.1-31.4], sensitivity of 88 [69-97]% and specificity of 69 [59-79]%. SEPPER >190 gave a relative risk of 8.0 [3.3-19.5], sensitivity of 82 [63-94]% and specificity 77 [67-85], and septum-to-cavity ratio >0.45 a relative risk of 6.3 [2.5-15.7], sensitivity of 79 [58-93]% and specificity of 73 [63-82]%. Most subjects with cardiac arrest had shown progressive disease with median increase in Z-score of +0.48 over time, but the majority of survivors had reduced their Z-scores in response to therapy, median -0.29, (p=0.021).
CONCLUSION: The measures relating wall thickness to body size (Detroit Z-score) or age (SEPPER) discriminated best between a high and low risk, but a Z-score >6 is too high a cut-off to give a reasonable sensitivity; a cut-off >4.5 gives the highest sensitivity but at the cost of a lower specificity.