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Research on Children With Hypertrophic Cardiomyopathy

The Hypertrophic Cardiomyopathy Association is a 501c3 nonprofit organization founded in 1996 to serve the hypertrophic cardiomyopathy (HCM) community in matters of advocacy, support, and education. The Hypertrophic Cardiomyopathy Association monitors publications on HCM daily, and after review of the paper by Sreeram et al. (1) from Germany and the United Kingdom regarding the use of a novel strategy of radiofrequency ablation in HCM children with obstruction, I was left wondering why these children were experimented on at all.

HCM patients, particularly children, have a history of being harmed by research and unbridled enthusiasm to use novel techniques, unfortunately at great risk to these already compromised patients. I remain perplexed with why the authors aggressively have promoted an invasive intervention to reduce outflow obstruction in children with an experimental technique when suitable therapies have been used, tested, and proven in adults and then used safely in children.

The standard of care for patients with HCM with obstruction (American College of Cardiology [2,3] as well as American Heart Association [3] and European Society of Cardiology [2] through their consensus recommendations and guidelines [2003 and 2011]), was abandoned in this study. In the past decade, these societies have agreed that invasive interventions to remove outflow obstruction in HCM should be reserved for those with drug-refractory symptoms and not as a first-line approach. Further, these recommendations have served patients well: complication rates have decreased and quality of life has improved. The use of tiredness as an inclusion criterion is highly disturbing because it is a difficult symptom to interpret in HCM patients of any age and, when reported by a third party (parent or caregiver), highly imprecise and is not a recognized symptom of obstruction.

The results of this study are more disturbing than the concept alone, reporting 2 deaths and 7 serious complications, including ventricular fibrillation, need for permanent pacing, valve replacement, implantable cardioverter-defibrillator implantation, failure and need for second attempt, burns, and myectomy—totalling a complication rate of more than 20%.

Although the HCM community of patients appreciates the investigation of new approaches to therapy, research on children with HCM must be held to the highest level of ethical and scientific standards. In our opinion, this research fell short. We disagree with the authors. This research does not merit further investigation in children and should be abandoned.

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Reply

We thank Dr. Maron and the Hypertrophic Cardiomyopathy Association for their interest in our paper (1). Both take issue with the fact that one of the symptoms in this patient cohort was tiredness. Symptoms have to be understood in the context of being of such severity as to interfere with the child's daily activity and quality of life, and parents are often the best judge of that.

The standard of care for patients with hypertrophic cardiomyopathy (HCM), as set out in the most recent version of the consensus guidelines, is curiously reticent when it comes to the care of children with symptomatic HOCM (2). It makes no mention of radiofrequency ablation, although this has been reported (3). The only reference to invasive therapy in childhood is the surgical series by Theodoro et al. (4) from the Mayo Clinic. In this small series (n = 25), 20% of patients had no reported symptoms. Does this imply that surgery in these children was experimental or unnecessary? The overall complication rate was similar to that reported in our study (intra-aortic balloon pump, n = 2; early AICD or pacemaker implantation, n = 3; and pericardiectomy, n = 1). Should we accept this uncritically as the gold standard?

There are few studies reporting on the efficacy of pharmacologic therapy for symptomatic children with HOCM. Reasonable questions that may be asked are: which drug(s) and at what level of evidence? It would be quite wrong to state as established fact that suitable therapies have been "used, tested, and proven in adults and then used safely in children," when this is clearly not the case.

Symptomatic children with severe obstruction may represent the worst end of the spectrum of HCM. One may argue that adult patients have selected themselves out as having a better prognosis simply by having lived longer. The absence of an alternative to open heart surgery undoubtedly has resulted in a conservative approach to HOCM in childhood. Maintaining the status quo is not of benefit for symptomatic children with an incurable and progressive disease. Most parent groups support efforts by the medical profession to improve the quality of life of their children. The theoretical considerations and rationale for the use of radiofrequency ablation are described clearly in our paper. Avoiding complications is paramount, but any new approach has an element of a learning curve; this was minimized by the authors' experience in catheter ablation techniques, but lessons learned will improve outcomes further.

Measuring ventricular septal thickness is meaningless, because this will remain unchanged regardless of whether a patient undergoes surgery or pharmacological therapy. What is important is

preservation of left ventricular function in the long term, allowing children to reach adult life. HCM is a leading cause of sudden death in young people worldwide (5). Restricting physical activity in young persons is impracticable, placing the onus further on the pediatric cardiologist to make timely management decisions. Newer techniques that are effective, repeatable, reproducible, easier to apply, and result in a shorter hospital stay and reduced costs therefore deserve a fair hearing and should be welcome.

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