Bicuspid Aortopathy and Sports Clearance

Bicuspid aortic valve is the most common congenital heart defect, with a population prevalence estimated at 1% to 2%.[1] This condition has a variable clinical course − while many have mild disease, longitudinal studies have shown that up to 50% of patients will have some degree of aortic dilatation by age 18.[2,3] Within this setting, sports clearance is a common scenario faced by pediatric cardiologists. Traditionally, a conservative, risk-averse approach has predominated, given the magnitude of incorrectly clearing those with truly pre-morbid disease. However, the benefit of exercise for all patients,[4] including those with congenital heart disease, has become increasingly clear, with improvement in physical development, mental health, and perceived quality of life all reported.[5] How then, do we best counsel our patients regarding both the hypothetical risk and plausible benefit?

The 36 Bethesda conference in 2005 provided guidelines for clearance, with straightforward, yes-or-no advisements based on mostly expert opinion of each diagnosis covered.[6] With the 2015 American Heart Association (AHA) and/or American College of Cardiology statement, recommendations for eligibility and disqualification are given in a more nuanced manner.[7] Specifically, the authors acknowledge the uncertainty, when present, for each clinical scenario and are transparent in showing levels of evidence and strengths of each recommendation. Class II recommendations should be a focus, as they indicate conflicting evidence and opinion and thus, from our viewpoint, give flexibility to physicians when applying them to patients. This is germane to our topic, as Class II recommendations predominate in those with bicuspid aortic valve and mild-moderate aortic dilation. For athletes with mild to moderate dilation (Z-score 2-2.5 − which are utilized in the pediatric population to adjust the aortic dimensions to the patient's body surface area, or diameter 40 to 42 mm in men, or 36 to 39 mm in women), the recommendation (class IIb) is to consider less strenuous competitive sports. For those with aortas measuring 43 to 45 mm, even less strenuous competitive sports may be considered (class IIb). The wording here is critical; it should not be interpreted as stronger than it is, and thus replacing the individualized data interpretation, risk assessment, and counseling we all perform with these patients.

Evidence regarding bicuspid aortopathy, risk of dissection, and sports participation continues to emerge and should be incorporated into up-to-date counseling. While the risk of dissection in those with bicuspid aortopathy has been described to be up to eight-times that of the general population,[8] the rates of aortic dissection in children are vanishingly low. In a population based study that reviewed all cases of aortic dissection in pediatric hospitals in the state of Texas over 7 years (2004 to 2011), representing almost 3.9 million hospitalizations, there were 110 patients with an aortic dissection.[9] The vast majority of these cases occurred following a recent cardiac procedure or surgery. In fact, there were only 10 cases of aortic dissection that occurred apart from this circumstance, and then only 2 of these patients had bicuspid aortopathy. While the anatomic and clinical details surrounding these individuals are unknown, both patients did survive to hospital discharge. Thus, the incidence of presentation to a pediatric hospital with an aortic dissection, without a prior inciting procedure, was found to be 0.25 per 100,000 hospitalizations. By comparison the risk of having a non-fatal drowning event in the United States from 2006 to 2011 was 8.8 per 100,000.[10] In spite of the 35-fold higher risk associated with swimming, many families accept this risk for their children. The familiarity with swimming and thus the inherently known risks, likely explains much of this paradox, but also underscores the role that a pediatric cardiologist can play in bridging the educational gap in informed decision making.

While risk of aortic dissection is the primary concern, cardiologists also fret over the theoretical risk of progression in aortic dilation or aortic valve dysfunction with more intensive exercise. Recent results, however, have been consolatory. Boraita et al[11] evaluated the behavior of bicuspid aortopathy in elite athletes, and found no difference in aortic diameters in the athlete cohort compared with sedentary controls, and furthermore no acceleration in aortic growth over the medium term. These findings are similar to those reported by Stefani et al[12] previously. With reference to these and other recent data, the 2020 European Society of Cardiology guidelines on sports cardiology and exercise[13] have been published and are less stringent when providing guidance on bicuspid aortopathy. Specifically, there is no recommendation for any exercise restriction in those with aortic diameters < 4 cm. Furthermore, for those with mild dilation, continued participation in noncontact and/or non"-power sports" is permissible. In the end, the pediatric cardiology office should not function merely as a conduit by which any guidelines, foreign or domestic, are stamped on to the next patient passing through. The ramifications of such an approach are known - a recent review of the strict application of the AHA guidelines to a representative patient cohort showed that 34% of children with a bicuspid aortic valve would be at least partially restricted due to some degree of aortic dilation.[14] How then should we proceed?

The model of shared decision-making is increasingly being recognized as an ideal way to counsel our patients through more difficult decisions. Indeed, within the AHA guidelines themselves, use of shared decision-making for patients with long QT syndrome and hypertrophic cardiomyopathy is evident. In 2019, the American College of Cardiology published practical steps for implementing shared decision-making when caring for athletes,[15] with emphasis on determining each individual's risk and benefits, providing education, and then having a detailed discussion with the athlete and other interested parties (family, coaches, etc.) to make sure all concur with the formulated plan. This is a time to also delineate what is unknown, which often then allows an athlete and family to use their personal beliefs to bridge the gap toward an acceptable decision. When done appropriately, this can result in 2 similar athletes with similar medical issues coming to divergent decisions about sports participation. Shared decision-making does not absolve the cardiologist from participating in the decision,

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but instead allows us to properly advise our patients on a decision that should be both medically reasonable and patient copacetic. Documentation of these discussions and outcome is essential, both for future reference and to address legal ramifications. Finally, decisions are dynamic, and reassessment remains an important component of the evaluation. Most commonly, this would be done on an annual basis and as data continues to be gathered, ultimate decisions can change (and should be specified as such at the onset).

Dictating to children and their families that an underlying heart condition means one is not healthy enough to participate in sports can have significant consequences beyond the decision itself. On the heels of such a unilateral decision, a previously asymptomatic patient not uncommonly turns into a chronically unwell and perpetually unhappy patient going forward (if they keep seeing you at all). Shared decision-making, on the other hand, empowers patients and families to better understand, in this case, the heart condition and how it impacts their health. A stronger doctor-patient bond is also formed by the process. Bicuspid aortopathy and sports clearance are common issues encountered by pediatric cardiologists, and we need not be at odds with our more athletic patients and families when these entities comingle. Instead, the process of shared decision-making provides the framework to reach a decision acceptable to all.

Declaration of Interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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1. Verma S, Siu SC. Aortic dilatation in patients with bicuspid aortic valve. *N Engl J Med* 2014;370:1920–1929.
2. Mahle WT, Sutherland JL, Frias PA. Outcome of isolated bicuspid aortic valve in childhood. *J Pediatr* 2010;157:445–449.
3. Holmes KW, Lehmann CU, Dalal D, Nasir K, Dietz HC, Ravekes WJ, Thompson WR, Spevak PJ. Progressive dilation of the ascending aorta in children with isolated bicuspid aortic valve. *Am J Cardiol* 2007; 99:978–983.
4. Norman RS, Blyholder L, Sanfilippo J. Sport and physical activity level impacts health-related quality of life among collegiate students. *Am J Heal Promot* 2019;33:675–682.
5. Dean PN, Gillespie CW, Greene EA, Pearson GD, Robb AS, Berul CI, Kaltman JR. Sports participation and quality of life in adolescents and young adults with congenital heart disease. *Congenit Heart Dis* 2015;10:169–179.
6. Maron BJ, Co-chair C, Zipes DP, Co-chair C. 36th bethesda conference 36th bethesda conference : eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol* 2005:45.
7. Maron BJ, Zipes DP, Kovacs RJ. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: preamble, principles, and general considerations. *Circulation* 2015:132.
8. Michelena HI, Khanna AD, Mahoney D, Margaryan E, Topilsky Y, Suri RM, Eidem B, Edwards WD, Sundt TM, Enriquez-Sarano M. Incidence of aortic complications in patients with bicuspid aortic valves. *JAMA - J Am Med Assoc* 2011;306:1104–1112.
9. Shamszad P, Barnes JN, Morris SA. Aortic dissection in children and young adults; a multiinstitutional study. *Congenit Heart Dis* 2014; 9:54–62.
10. Felton H, Myers J, Liu G, Davis DW. Unintentional, non-fatal drowning of children: US trends and racial/ethnic disparities. *BMJ Open* 2015;5:1–8.
11. Boraita A, Morales-Acuna F, Marina-Breysse M, Heras ME, Canda A, Fuentes ME, Chacón A, Diaz-Gonzalez L, Rabadan M, Parra Laca B, Pérez De Isla L, Tuñón J. Bicuspid aortic valve behaviour in elite athletes. *Eur Heart J Cardiovasc Imaging* 2019;20:772–780.
12. Stefani L, Galanti G, Innocenti G, Mercuri R, Maffulli N. Exercise training in athletes with bicuspid aortic valve does not result in increased dimensions and impaired performance of the left ventricle. *Cardiol Res Pract* 2014;2014:23869.
13. Task A, Members F, Pelliccia A, France JC, Drezner JA, States U, Germany MH, Francesco M, Denmark EP, Stuart AG, Kingdom U, Taylor RS, Kingdom U, Thompson PD, States U, Gale CP, Kingdom U, Hermann K, France BI, Germany HAK, Israel AK, France CL. 2020 ESC guidelines on sports cardiology and exercise in patients with cardiovascular disease. *Eur Heart J* 2020:1–80.
14. Baleilevuka-Hart M, Teng BJ, Carson KA, Ravekes WJ, Holmes KW. Sports participation and exercise restriction in children with isolated bicuspid aortic valve. *Am J Cardiol* 2020;125:1673–1677.
15. Kolwgi GM. Shared Decision Making: The New Frontier. Available at: http://www.acc.org/latest-in-cardiology/articles/2019/07/30/15/30/shared-decision-making-the-new-frontier.