

Unicuspid aortic valves are no bicuspid aortic valves – It’s time to retire the Sievers-classification

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Abstract

A general misconception that unicuspid aortic valves (UAV) are just a subtype of bicuspid aortic valves (BAV) has now been perpetuated in the medical and surgical community for nearly two decades[1](#ref-0001), and there is no end in sight.

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It’s time to retire the Sievers-classification

Short title: Bicuspid aortic valve classification

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A general misconception that unicuspid aortic valves (UAV) are just a subtype of bicuspid aortic valves (BAV) has now been perpetuated in the medical and surgical community for nearly two decades¹, and there is no end in sight.

The Sievers classification distinguishes between 3 types of bicuspid aortic valves. Type 0 is an aortic valve with two symmetric leaflets, but no raphe (a raphe is a non-functional commissure, as two leaflets fuse at the commissure to form a conjoined leaflet). Type I is a bicuspid aortic valve with a raphe, and the raphe can occur at any commissure (71% left/right-, 15% right/non-, 3% left/non-commissural fusion). Type II is described as a bicuspid aortic valve with 2 rapheae. But here also lies the fundamental mistake. We know that unicuspid aortic valves can present as unicommissural- or acommisural valves². Thus, an aortic valve with only one commissure is simply a unicuspid aortic valve and not a bicuspid aortic valve. With this, the Sievers classification is now reduced to 2 types only. The question therefore arises: how helpful is this classification really?

In a recent study from Denmark, the prevalence of bicuspid aortic valves in newborns was investigated³. The authors hereby found an overall prevalence for BAV of 0.77%. Within this population, 8.7% of valves had a Type 0 phenotype, 90.8% were Type I and 0.5% Type 2 BAV. In this study, as well as a recent editorial in the Journal of Cardiac Surgery, the authors have been mistakenly under the impression that unicuspid aortic valves are just a subtype of BAV^{3, 4}. Based on the Sievers classification unfortunately, this is a common notion shared throughout the medical and scientific community.

Categorization of valve subtypes, however, mainly carries importance in the surgical arena where different phenotypes require different surgical approaches for valve repair, with their respective complexities. But in a surgical setting, to summarize >90% of BAV phenotypes with a raphe to just one subtype is utterly useless. Type I BAVs can range from perfectly symmetric valves to almost tricuspid-like phenotypes (forme-fruste BAV)⁵. The Sievers classification is hence too simplistic and does not provide any roadmap for potential surgical repair.

Our understanding of BAV phenotypes however, has fundamentally changed and evolved over the last few years with the landmark study by De Kerchove et al⁶. In this study, the investigators elucidated that commissural orientation, length of cusp fusion of the conjoined leaflets, and raphe height are intrinsically linked to each other. A true renaissance in our understanding of BAV morphologies, as now a wide spectrum of potential BAV phenotypes. This spectrum was further subcategorized into symmetric-, asymmetric-, and very asymmetric phenotypes. Symmetric valves thus comprise Sievers Type 0 and I, and the latter 2 only comprise Type I valves. This improved understanding, however, has led to a repair-oriented surgical classification for BAV^{5, 7}, which is based on commissural orientation. The commissural orientation is easily assessed on echocardiography and has implication for length of cusp fusion and raphe height. This provides the surgeon with a clear picture of the valve, in anticipation of surgical repair. This classification also doesn't mistakenly entail unicuspid aortic valves as a subtype of bicuspid aortic valves anymore.

The Sievers classification is a relic of the past when our understanding of BAV morphology was still very limited. With this limited understanding of valve morphology, a fundamental mistake was introduced into a widely used BAV classification. Sadly, to this day it is still confusing the scientific community. The main aim of this editorial, is to set the record straight. *Unicuspid aortic valves are unicuspid aortic valves*, and not just a subcategory of bicuspid aortic valves. Moreover, with a fundamentally improved understanding of bicuspid aortic valves, more sophisticated classifications are now available^{5, 7}. These classifications can guide surgeons in surgical repair of bicuspid aortic valves by providing a clear picture of valve morphologies, with accordingly, an improved ability to plan the correct surgical repair strategy ahead of the operating room.

Author contributions:

JJ – writing, conceptualization, final approval

LDK – conceptualization, revision, final approval

PIT – conceptualization, writing, revision

SMS – conceptualization, drafting, revision

GEK – conceptualization, revision, final approval

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