## Words and how we use them – which is to be the master?

Robert Anderson<sup>1</sup>, Diane E. Spicer<sup>2</sup>, James Quintessenza<sup>2</sup>, Hani Najm<sup>3</sup>, and Justin Tretter<sup>3</sup>

<sup>1</sup>IInstitute of Genetic Medicine, Newcastle University, Newcastle-upon-Tyne, United Kingdom Birmingham Children's Hospital, Birmingham, United Kingdom <sup>2</sup>Johns Hopkins All Children's Hospital <sup>3</sup>Cleveland Clinic

May 4, 2022

### Abstract

This is an invited commentary and does not need a commentary.

Words and how we use them – which is to be the master?

# Robert H. Anderson,<sup>1</sup> Diane E. Spicer,<sup>2</sup> James A. Quintessenza,<sup>2</sup>Hani K. Najm,<sup>3</sup> and Justin T. Tretter<sup>4</sup>

<sup>1</sup> – Cardiovascular Research Centre, Biosciences Institute, Newcastle University, Newcastle upon Tyne, UK

<sup>2</sup> – Heart Institute, Johns Hopkins All Children's Hospital, St. Petersburg, FL, USA

<sup>3</sup> – Department of Pediatric Cardiac Surgery, Cleveland Clinic Children's, and The Heart, Vascular, and Thoracic Institute, Cleveland Clinic, Cleveland, OH, USA

<sup>4</sup> – Department of Pediatric Cardiology, Cleveland Clinic Children's, and The Heart, Vascular, and Thoracic Institute, Cleveland Clinic, Cleveland, OH, USA

Key words: Aortic root; Arterial valves; Sinuses; Leaflets; Nomenclature

Corresponding author: Justin T. Tretter, M.D., Pediatric and Adult Congenital Heart Center, Cleveland Clinic, 9500 Euclid Avenue, M-41, Cleveland, OH, 44195. Fax: 216-445-3692. Phone: 216-219-6374. Email: trettej3@ccf.org

In a brief report published in the current issue of the Journal, the group working at Brussels in Belgium describe their experience with "bicuspidisation" of a congenitally unicuspid aortic valve.<sup>1</sup> We applaud the Belgian group in terms of its meticulous reparative approach, which is based on the understanding of the three-dimensional anatomy of the aortic root. We support the general concepts as described in their current account as the potential way forward in treating those with functionally unileaflet aortic valves amenable to surgical repair. In this regard, those attempting to repair the congenitally malformed aortic root should remember that, when repositioning the commissures, account should also be taken of the asymmetry of the leaflets and their sinuses.<sup>2</sup>

The general approach described by the Belgian group, which we have adopted ourselves,<sup>3</sup> is founded on unambiguous communication between the imager and the surgeon. In this light, during the review process culminating in the publication of their significant contribution, several exchanges took place between the referee, who was one of ourselves (RHA), and the authors. The content of the exchanges is also pertinent to

recent exchanges in the pages of JCTVS Open, in which one of the Belgian authors was a contributor.<sup>4,5</sup> The exchanges related to a "consensus" document regarding the description of the normal and abnormal aortic root.<sup>6</sup> They reveal that the schisms identified in the questionnaire recognising the "Tower of Babel", which itself was said to exist with description of the root,<sup>7</sup> have yet to be resolved. The reasons for the ongoing disagreements are multiple.

In the first instance, the view obtained of the aortic root in the operating room, or in the echocardiographic laboratory, is quite different from that seen in the dissecting or autopsy room. This difference can now be overcome by the sophistication provided by clinical techniques such as computed tomography. These techniques are able to reveal the three-dimensional features of the arterial roots as seen in the living heart.<sup>3</sup> A second problem relates to the frequency with which the different abnormal aortic phenotypes are encountered by those working in adult as opposed to pediatric cardiac surgical practises. Also of significance is the use of words by those who seek to establish the superiority of their own preferred nomenclature. The authors who responded to our own letter on "the aortic valve with two leaflets" cited the oft-used aphorism originating from the works of Lewis Carroll.<sup>5</sup> As they emphasised, the question in terms of the words used is which feature is to be the master. Should it be the general meaning of the word, or the way it is defined by those who choose to use the given word for a specific purpose?

In seeking to justify their need to achieve compromise rather than consensus, the authors of the response claim to have illustrated the deficiencies of our own chosen preference.<sup>4</sup> There are, however, fundamental flaws in their described justifications.<sup>5</sup> This now becomes of significance with regard to the description by the Belgian authors of their approach to "bicuspidisation".<sup>1</sup> The evidence provided by the three-dimensional reconstructions,<sup>3</sup> to which we will return, is obviously of equal, if not greater, significance. But first consider the linguistic problems. It was the disagreements in the use of words that came to the fore in the "Tower of Babel" identified by the questionnaire circulated by the German cardiac surgeons.<sup>7</sup> Such disagreements are unlikely properly to be demolished until, as we suggested in our letter,<sup>4</sup>distinction is made between the moving components of the arterial roots and the arterial valvar sinuses that support them. Our preference for description of the moving components of the atrioventricular valves. In describing their technique for "bicuspidisation",<sup>1</sup> the Belgian authors have also, on this occasion, described the moving components as "leaflets". The response to our letter in JCTVS Open,<sup>4</sup> of which de Kerchove is a co-author,<sup>5</sup> along with the account of bicuspidisation,<sup>1</sup> suggests that he, and his colleagues, would much prefer to have used the word "cusp" when accounting for the moving components in the arterial roots.

We had pointed out the deficiency of such usage in our letter.<sup>4</sup> We emphasised that the authors of the consensus document that prompted our letter<sup>6</sup> had themselves created confusion when suggesting that the coronary arteries arose from the "cusps" of the aortic root. In their response,<sup>5</sup> the authors indicated that this was no more than a mistake, or an "oversight". Cardiac surgeons are well aware that mistakes, or oversights, made in the operating room can have catastrophic consequences. Attention to the current cardiological literature, furthermore, particularly when used by electrophysiologists, will show that "cusp" is used in equal fashion not only to describe the arterial valvar leaflets, but also their supporting sinuses. The latest issue of the European Heart Journal – Case Reports, for example, contains a report of premature ventricular complexes ablated from the non-coronary cusp.<sup>8</sup> "Cusp" is similarly used to account for the valvar sinuses in a recent publication in "Heart Rhythm".<sup>9</sup> Or do the authors of the response<sup>5</sup> imagine that the electrophysiologists are ablating the valvar leaflets? Perhaps this again would be no more than an "oversight"? If so, ablation of the leaflet is unlikely to be advantageous to post-intervention valvar function.

There are also, as we have stated, additional major flaws in the reasons provided by the responding authors, including de Kerchove, to substantiate their preference for "cusp" over "leaflet".<sup>5</sup> In the first place, they suggest that "the term "cusp" is specific to the semilunar valves". There is an additional problem here, in that the valves to which they refer are arterial, whereas it is the hinges of their moving parts that are semilunar. But that is a mere bagatelle when set against their assertion that "cusp" is specific to the arterial valves. Could it be that they have forgotten that the counterpart of the mitral valve is usually described

as the tricuspid valve? Or perhaps they are suggesting that the valve guarding the right atrioventricular junction of the normal heart should now be re-named as the "trileaflet" valve? We find it unlikely that such a suggestion would find general favour. It is incorrect, therefore, to suggest that "cusp" is limited to the moving components of the arterial valves.

The authors do proceed accurately to account for the derivation of "cusp".<sup>5</sup> As they describe, the word "indicates a pointed end formed by the intersection of 2 arcs or curved lines that meet (as in the tip of a spear)".<sup>5</sup> We agree with this definition. But they are in error when suggesting that such arcs and points are integral to the arterial valves, but not the atrioventricular valves. The edges of the leaflets of the atrioventricular valves, at the level of the atrioventricular junction, come together to produce such meetings of curved lines (Figure 1A). These meetings are directly comparable to those of the arterial valves at the sinutubular junction (Figure 1B). The area that is identified by the joining of the curved lines at the sinutubular junction, and hence correctly identified as the "cusp", demonstrates the peripheral end of the zone of apposition between the adjacent leaflets. This feature is usually described as the "commissure". It does not account for the entirety of the moving components of the arterial valves (Figure 1B).

Description of the "cusps" of the arterial valves in linguistically appropriate fashion would create still further problems. This is because, as emphasised above, the feature of the meeting of the two arcs is usually described as the valvar commissure. These "commissures" are the peripheral attachments, at the sinutubular junction, of the zones of apposition between the moving components of the roots. Suggestions that "cusp" be restricted to the moving components,<sup>5</sup>therefore, does no more than add to the potential confusion. Such commissures are also readily identified between the moving components of the valve itself usually described as being "tricuspid" (Figure 1A). The reason that the leaflets of the arterial valves were initially considered to be "cusps" is more likely to be because of their resemblance, when viewed in closed position from their ventricular aspect, to the surfaces of the molar and premolar teeth. The arrangement of the leaflets of the tricuspid valve, furthermore, would present a similar appearance when viewed in closed position from the atrial aspect. The argument that the derivation of "cusp" justifies use of the word to describe only the moving components of the arterial valves, therefore, is both illogical and unjustified.

Of far greater importance, as was acknowledged by the authors of the response to our letter,<sup>5</sup> is the ability to distinguish between the moving components of the arterial valves and the sinuses that support them. The very fact that "cusp" is currently used interchangeably to describe both features, in our opinion, disqualifies it from use exclusively to describe the moving components. Far better to use "leaflet" to describe these components in both sets of valves, not least because it is the competence or incompetence of these components in both settings that determines valvar function.

There is then another problem in the use of words that has emerged from the recommendations of the consensus document,<sup>6</sup> the response to our letter,<sup>5</sup> and the approach now taken by those recommending "bicuspidation" for some variants of the unicuspid and unicommissural aortic valve.<sup>1</sup> This is in the use of "annulus". As was shown by the questionnaire prompting the recognition of the "Tower of Babel",<sup>7</sup> not all surgeons equated this ring with the diameter measured by echocardiographers, better described as the virtual basal ring. This virtual plane is created by joining together the nadirs of the moving components. In the response to our letter,<sup>5</sup> it was indeed the virtual basal ring that was illustrated as representing the "annulus". In the described approach to "bicuspidisation", however, the Belgian authors now argue in favour of a "functional annulus", which they argue extends from the virtual basal ring to the sinutubular junction. It would seem that this corresponds with the hinges of the moving components. And the response to the German questionnaire showed that some surgeons considered these attachments to represent a "semilunar ring".<sup>7</sup> Could it be, therefore, that the group are now recognising both potential candidates as the valvar "annulus"? Such an approach must provide the potential for still further confusion, and additional "oversights", the more so since some surgeons in the response to the questionnaire equated the virtual basal ring with the ventriculo-arterial junction.<sup>7</sup>

As we indicated in our opening paragraph, the ability now to use computed tomographic interrogation so as to demonstrate the three-dimensional arrangement in the living heart emphasises the need to use words

that are specific for the different components that come together to form competent arterial roots. These images show that, as also demonstrated in the drawing provided by the authors responding to our letter,<sup>5</sup> the arterial roots extend between the virtual basal ring and the sinutubular junction (Figure 2, Video 1). It is the extent of the semilunar hinges of the moving components within the roots that delimits these boundaries. If we understand their definition correctly, it is these semilunar hinges that the Belgian group now define as the "functional annulus".<sup>1</sup>Rather than producing confusion with those who continue to recognise the virtual basal ring as the echocardiographic annulus,<sup>7</sup> it would surely be preferable simply to describe the attachments of the moving components as the semilunar hinges. And, as shown in Figure 2, it is now an easy matter to reconstruct these hinges. When viewed in three-dimensions, they produce a crown-like configuration. Equally important, the reconstruction shows that, interposing between the adjacent hinges, and separating the valvar sinuses, are to be found the fibrous interleaflet triangles.<sup>10</sup> It is the extent of formation of these interleaflet triangles that serves to identify the different phenotypes encountered when the arterial roots are congenitally malformed.<sup>11</sup> As the Belgian authors recognise in their account, the phenotype that they treat on the basis of "bicuspidisation" is built on the basis of a trisinuate scaffold.<sup>3</sup> It is because of hypoplasia and incomplete formation of two of the three interleaflet triangles that the skirt of leaflet tissue takes on a unicuspid and unicommissural appearance. And it is because of the presence of a solitary zone of apposition within the skirt of leaflet tissue that the phenotype lends itself to creation of a new "bicuspid" valve, as described in their brief report.<sup>1</sup> It also follows that the commonest variants of "bicuspid" valves are similarly built on a trisinuate scaffold, but with incomplete formation of only one of the interleaflet triangles.<sup>3</sup> The variation in such bileaflet valves housed within trisinuate roots then depends on which interleaflet triangle is incompletely formed. In the rarest variant of the "bicuspid" valve, there are but two sinuses formed.<sup>3</sup>This phenotype then varies according to whether the missing sinus, and its supporting leaflet, is the non-coronary primordium or one of the primordiums that gives rise to a coronary artery. It is a surprising paradox that, when the interleaflet triangles are incompletely formed, as in the so-called unicuspid variant, then the hinges of the skirt of leaflet tissue are much more annular, rather than showing the crown-like configuration of the normal root.

There is then an even bigger paradox emerging from the brief report of the Belgian group,<sup>1</sup> and the other recent publications we have highlighted in our commentary.<sup>4-6</sup> We all have the same goal – namely to demolish the Tower of Babel. And we all share the desire to use words in their most appropriate fashion. We submit, however, that the reasons offered thus far for using "cusp" exclusively to account for the moving components of the arterial roots are without any logical foundation. It is spurious to suggest that the use of "cusp" to account for the valvar sinuses rather than the moving components is no more than an oversight. The tower will only properly be demolished when separate words are used to account for the moving components of the roots and the walls of the root that support them. This, we submit, is best achieved by describing the moving parts as the leaflets, and recognising that such moving components are also to be found within the atrioventricular valves. In both sets of valves, it is snug apposition of these moving parts that ensures valvar competence. And the achievement of valvar competence is the major goal of cardiac surgeons when operating on malformed valves, be they arterial or atrioventricular.

### $References\ cited$

- 1. Jahanyar J, Aphram,G, Munoz, DE, Mastrobuoni, S, Navarra, E, de Kerchove, L, El Khoury, G. Congenital Unicuspid Aortic Valve Repair without Leaflet Patch Augmentation. J Card Surg 2022; in the press
- 2. Aicher D, Kunihara T, Abou Issa O, Brittner B, Gräber S, Schäfers H-J et al. Valve configuration determines long-term results after repair of the bicuspid aortic valve. Circulation 2011; 123: 178–85
- Tretter JT, Izawa Y, Spicer DE, Okada K, Anderson RH, et al. Understanding the Aortic Root Using Computed Tomographic Assessment: A Potential Pathway to Improved Customized Surgical Repair. Circulation: Cardiovascular Imaging. 2021; 14: e013134.
- 4. Tretter JT, Spicer DE, Jacobs JP, Anderson RH. The aortic valve with two leaflets. JTCVS Open. 2022; 9: 89-90
- 5. Della Corte, A, Maleszewski, JJ, Fernandez, B, De Paulis, R, de Kerchove L, et al. Reply: Sometimes

consensus is a euphemism for compromise. JTCVS Open, 2022; 9, 91 – 92

- Michelena HI, Della Corte A, Evangelista A, Maleszewski JJ, Edwards WD, et al. International consensus statement on nomenclature and classification of the congenital bicuspid aortic valve and its aortopathy, for clinical, surgical, interventional and research purposes. J Thorac Cardiovasc Surg. 2021; 162: 781-797
- 7. Sievers HH, Hemmer W, Beyersdorf F, Moritz A, Moosdorf R, et al. The everyday used nomenclature of the aortic root components: the tower of Babel?. E J Cardio-thor Surg 2012; 41: 478-482.
- 8. Tanaka, A, Nozoe, M, Tsutsumi, T, Kubota, T. Ventricular premature complexes successfully ablated from the non-coronary cusp: a case report, Eur Heart J Case Reports, 2022; 6: ytac129.
- Berte B, Hilfiker G, Mahida S, Wielandts JY, Almorad A, Knecht S, Shah D, Vijgen J, Duytschaever M, Kobza R. High-resolution parahisian mapping and ablation using microelectrode embedded ablation catheters. Heart Rhythm. 2022; 19: 548-559.
- Sutton JPIII, Ho SY, Anderson RH. The forgotten interleaflet triangles: A review of the surgical anatomy of the aortic valve. Ann Thorac Surg 1995; 59: 419-427.
- Tretter JT, Spicer DE, Mori S, Chikkabyrappa S, Redington AN, Anderson RH. The Significance of the Interleaflet Triangles in Determining the Morphology of Congenitally Abnormal Aortic Valves: Implications for Noninvasive Imaging and Surgical Management. J Am Soc Echocardiogr. 2016; 29: 1131-1143.

### Legends to Figures

**Figure 1.** As is correctly stated by the authors of the response to our letter published in JCTVS Open,<sup>4</sup> a "cusp" is the feature "formed by the intersection of 2 arcs or curved lines". As can be seen, such "cusps" are to be found in the atrioventricular valves, as shown for the tricuspid valve in panel A, as well as in the arterial valves (panel B). In both valves, however, the feature is usually described as a "commissure", rather than a "cusp".

Figure 2. The images show selected cuts made from a computed tomographic three-dimensional reconstruction of the normal trileaflet and trisinuate a ortic root. Panels A and B show short (A) and long axis (B) views positioned over a two-dimensional plane constructed at the level of the virtual basal ring. The superior extent of the myocardial crescents incorporated into the coronary sinuses is outlined with an orange line. It is the junction between the sinusal walls and the myocardial crescents that dictates the anatomical ventriculo-arterial junction, which is not present in the non-coronary sinus (N). The semilunar hinge lines of the valvar leaflets, shown by the red lines, extend from their nadir at the plane of the virtual basal ring, shown by the green line, to their zenith at the sinutubular junction, which is marked by the blue line. These hinge lines dictate the hemodynamic ventriculo-arterial junction. The interleaflet triangles, marked by the purple triangles, are positioned between these hinge lines. The interleaflet triangle between the coronary aortic sinuses has muscle at its base. This forms another part of the anatomical ventriculo-arterial junction. The interleaflet triangle between the right (R) and non-coronary sinuses has the membranous septum (colored yellow) at its base, with the septum positioned so as to be transected by the plane of the virtual basal ring. Panels C and D then show further virtual dissections, with panel C showing a cut between the right and non-coronary sinuses, and Panel D showing a cut through the two coronary aortic sinuses. Further abbreviations: L, left coronary sinus; LCA, main stem of the left coronary artery; RCA, right coronary artery.

Video 1. The normal aortic root as shown in Figure 2A and B is shown, initially viewing the root in short axis, and then rotating 360 degrees around its long axis. The non-coronary sinus is seen first, followed by the right, and then the left coronary aortic sinuses.



