The matter of "unbalance" in right dominant atrioventricular septal defect

Ignacio Lugones[1], María Fernanda Biancolini[2], Germán Lugones[3], Julio César Biancolini[2], Ana MS de Dios[2]
[1]Cardiac Surgery Department, "Dr. Pedro de Elizalde" General Children´s Hospital, Buenos Aires, Argentina, [2]Pediatric Cardiology Department, "Dr. Pedro de Elizalde" General Children´s Hospital, Buenos Aires, Argentina, [3]Physics Department, Center of Natural and Human Science, Federal University of ABC, San Pablo, Brazil

ABSTRACT

Unbalance in atrioventricular septal defect can be found in more than one anatomic level and in different degrees at each level. The definition of "unbalance" has historically been focused in comparing the dimensions of main cardiac structures, such as the atrioventricular valve and the ventricles. However, the hemodynamic aspects of unbalance need to be considered as having, at least, similar relevance. New concepts and already described parameters must be combined and understood as a whole to help the surgical decision-making process.

Keywords: Common atrioventricular canal defect, common atrioventricular valve, echocardiography, functionally univentricular heart, left ventricle, ventricular septal defect

The term "unbalance" has been used for decades in atrioventricular septal defect (AVSD) to describe a lack of symmetry between left and right sides of the heart. Even though we all think to understand its meaning, the concept it alludes to is quite vague. What does then "unbalance" truly mean? We still strive to find a precise definition.

Perhaps, the main issue is that we are trying to sort out a heart with AVSD as if it only had two pieces (left and right) that could be similar in size or not. Actually, unbalance can strike different levels independently. Therefore, the affected anatomic level should always be specified (i.e., unbalance at the atrioventricular valve [AVV] level). Being "balanced" at one level does not necessarily mean that this condition will be fulfilled in the rest. Besides, finding a certain degree of asymmetry at one level does not imply that the same degree will be found in the others.

Unbalance is related to a lack of symmetry in: (a) size or (b) distribution of blood flow between the pulmonary and systemic circulations. The first might also be described as "anatomic unbalance" and reflects the actual absence of symmetry in size (diameter, length, and volume) between the right and left structures. On the other hand, asymmetry in blood flow distribution can be conceived as a "physiologic unbalance." Certain anatomic features such as the sizes of the atrial septal defect and ventricular septal defect (VSD) or malalignment of the atrial septum and/or ventricular septum have a higher impact on distribution of blood flow between the systemic and pulmonary circulations than on the presence or absence of symmetry in size between right and left structures (although they might contribute to the development of anatomic asymmetry).[1]

Perhaps, after all, the precise definition of "unbalance" could be no more than a semantic issue. The core challenge in this disease is to predict the ability of the left ventricle (LV) to maintain systemic cardiac output and according to that be able to identify the best surgical approach in terms of morbidity and mortality.

Several echocardiographic indexes are currently being used to evaluate these patients. One of them is

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Address for correspondence: Dr. Ignacio Lugones, "Dr. Pedro de Elizalde" General Children´s Hospital, Buenos Aires, Argentina,
40 Manuel Montes de Oca Ave, C1270AAN Buenos Aires, Argentina. E-mail: cardiocongenitas@gmail.com

the modified AVV index (mAVVI), derived from the previously described AVV index (ratio between the smaller and the larger areas of the AVV).[2] This index has proved to accurately identify unbalance at the AVV level. In 2013, a multi-institutional study revealed another variable: right ventricle [RV]/LV inflow angle.[3] This angle was more obtuse in "more balanced" cases, while patients with more acute angles had greater degree of unbalance. Recently, our group showed that this angle is essentially an indirect way of assessing the VSD size. Thus, the indexed VSD (inVSD) emerged as a new echocardiographic variable.[4] It is calculated as the ratio between VSD size and total AVV diameter in apical four-chamber view. This parameter emphasizes the importance of assessing the physiologic unbalance determined by the presence of a shunt at the ventricular level.[5] A small inVSD means that the LV is already maintaining the systemic cardiac output by itself in the preoperative state. Therefore, one-stage biventricular repair (BVR) might be tolerated. Conversely, a large VSD might preclude this approach as it has been related to increased mortality, probably because the LV is unloaded into the RV and the pulmonary circulation, and its ability to manage the systemic circulation is at least uncertain.

Even with the introduction of the inVSD, we are facing an incomplete preoperative evaluation. Malformations of the AVV and its subvalvar apparatus are frequent associations that can modify the "true LV inflow." In other words, even achieving a good diameter of the mitral component of the AVV during BVR, the presence of subvalvar derangements can cause obstruction at the true entrance to the LV and affect its performance. Taking this into consideration, Szwast *et al.* described in 2011 the left ventricular inflow index (LVII) and found that patients with values under 0.55 had increased risk of mortality of BVR.[6] This index is defined as the ratio between the secondary annulus (SA) and the left primary annulus (LPA), both measured in apical four-chamber view. The SA is represented by the smallest diameter of color Doppler at LV inflow and is obtained by tracing a line between the crest of the interventricular septum and the LV wall. The LPA is measured as a line extended from the left hingepoint of the AVV to the crest of the interventricular septum. In our geometric model, this line represents the hypotenuse of the left right-angled triangle [Figure 1]. Its size depends on the size of the other two sides of the triangle, given by the VSD and the left component of the AVV. A large VSD determines a large LPA. Given the fact that the LVII is calculated as the SA/LPA ratio, an increase in LPA will determine a low value of LVII. This is why this index is not a "pure" measure of the true LV inflow since it is markedly influenced by the VSD size.

Even though the LVII has been considered a strong discriminator of risk of mortality of BVR, limitations have been reported in its use. Some high values of this index, which are identified as implying low risk of mortality, may in fact be problematic. For example, a patient with a very small LPA and an SA with a similarly small size will certainly have LV inflow derangements, despite having a high value of LVII. This is the reason why Szwast *et al.* found that the LVII was not a useful parameter for cases displaying more severe degrees of unbalance. As they stated, although uniform blood flow could be found across the left component of the AVV and the "true" LV inflow, both structures can be so markedly hypoplastic that would preclude BVR.

For these reasons, our group is now working on a new metric, the indexed SA, which might have the ability to assess the severity of the subvalvular obstruction in a pure fashion. It is calculated as the ratio between the SA and total AVV diameter. Our geometric model provides the critical values below which this index would suggest that malformations of the mitral subvalvar apparatus need to be addressed if BVR is to be considered.

So far, we have not been collectively able to identify a set of measures with precise cutoffs that allow a trustworthy surgical decision-making. Correlation between mAVVI, RV/LV inflow angle, and LVII has recently been assessed in a large cohort of patients.[7] These parameters correlate poorly, or not at all, with another. There is no significant correlation between the mAVVI and the angle, and the reason for that is that they evaluate different things (unbalance at the AVV level and VSD

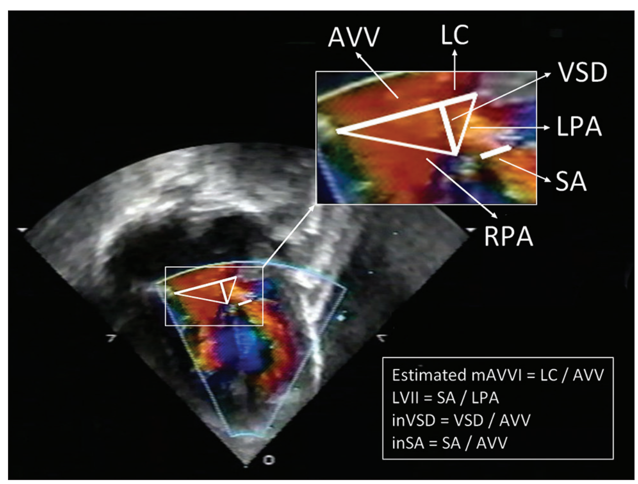

Figure 1: In echocardiographic four-chamber view, the main structures of the crux of the heart are depicted. The AVV, the LPA, and the RPA form the main triangle, which in turn is formed by two right-angled triangles. The left right-angled triangle is composed by three elements: The LC of the AVV valve, the VSD, and LPA. Simple geometric rules apply to these triangles, and therefore, relations between the different elements can be established. AVV: Atrioventricular valve, LC: Left component, LPA: Left primary annulus, RPA: Right primary annulus, SA: Secondary annulus, VSD: Ventricular septal defect, mAVVI: Modified atrioventricular valve index, LVII: Left ventricular inflow index, inVSD: Indexed ventricular septal defect, inSA: Indexed secondary annulus

size, respectively). The angle was found to correlate only moderately with the LVII, and that is because both parameters are influenced by the VSD size. Similarly, the moderate degree of correlation found between the mAVVI and the LVII is explained by the fact that both variables are influenced by the size of the mitral component of the AVV.

Many other variables have been described and must be assessed for proper surgical decision-making. This long list includes (but is not limited to) the ventricular cavity ratio,[2] LV-to-RV long-axis ratio,[8] ventricular volumes, apex forming ventricle, AVV overriding and/or straddling,[9] AVV z-score,[10] ductus arteriosus dependency, atrial to ventricular septum malalignment,[1,11] AVV insufficiency, and pulmonary hypertension.[11] Associated malformations, such as aortic coarctation or double-outlet RV,[12] can also modify AVSD physiology and should be carefully evaluated.

Other imaging technologies can help defining the patient's status. Three-dimensional echocardiography opened a new window in the assessment of the AVV and its apparatus.[9,13,14] Magnetic resonance imaging is the gold standard to determine ventricular volumes and can also assess ventricular function.[9,14]

One thing we know for sure: unbalanced AVSD is a really complex puzzle.[5] Several variables have been described, but the physiologic consequence of the interplay between them has not been elucidated. New concepts and already described parameters must be combined and understood as a whole to fit together the loose pieces of this puzzle.

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Conflicts of interest

There are no conflicts of interest.

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