EDITORIAL

Coronary Artery Anomalies: A New Mystery of the Bicuspid Aortic Valve

Las anomalías coronarias: Una nueva incógnita de la válvula aórtica bicúspide

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The bicuspid aortic valve (BAV) is the most common congenital heart defect and although it is considered a relatively benign condition, it has been associated with valvular dysfunction, aortic dilation or aortic dissection throughout its progress. (1, 2) The association with coronary artery anomalies had been described in isolated cases, but, in the past 5 years different studies have highlighted the association between both entities, probably related to the higher demand for computed tomography (CT) scans in this population. In a series with more than 400 patients, Michalowska et al. explored this association and did not find significant differences in the presence of coronary anomalies among patients with BAV and those with tricuspid aortic valve (TAV). (3) Conversely, the large series by Naito et al., which included 1099 patients undergoing aortic valve replacement or aortic repair, demonstrated that the prevalence of coronary anomalies was greater in patients with BAV compared with those with TAV during the preoperative evaluation with coronary angiography (in most cases) or CT scan. (4)

The present article by Carrero et al. (5) further emphasizes the importance of this association after finding in their prospective series of patients evaluated with CT scan a higher prevalence of coronary anomalies in patients with BAV compared with controls with TAV (26.1% vs. 2.2%, p = 0.001). In contrast to previous series, Carrero et al. included patients at earlier stages of the disease and not before surgery; nevertheless, this phenomenon was still present.

The estimated prevalence of coronary anomalies diagnosed by angiography, CT scan and autopsy is variable and ranges from 0.21% to 5.8%. (6-8) The association between BAV and coronary anomalies has been observed in recent years, and the embryogenesis of the aortic root and coronary arteries has been postulated as a probable cause. In fact, BAV is the result of aortic cusp development during valvulogenesis with fusion or absence of division of adjacent cusps. The prevalence of BAV in first-degree relatives is 6%, (9) indicating the genetic origin of this entity; moreover, its association with other congenital heart defects as coarctation of the aorta, aneurysms of the thoracic aorta, ventricular septal defect or mitral valve abnormalities has also been demonstrated. Although the genetic substrate of BAV remains unclear, it would probably help to understand BAV embryogenesis and its association with abnormal development of the coronary arteries.

The present study classifies coronary anomalies using the classification of Naito et al. as separate ostium, anomalous origin and anomalous course. (4) However, the classification of coronary artery anomalies varies depending on the study. In 2016, the European Society of Cardiology Working Group published a consensus statement reviewing the mechanisms of embryonic development and established a more extensive anatomical classification based on embryological development. (8) Probably, in view of the recent scientific evidence on this association, a universal and standard classification should be established to unify criteria among future studies.

High take-off of the coronary arteries has proved to be one of the most common anomalies. As the use of CT scan in this population increases, more cases of these anomalies will probably be detected than when identified for clinical reasons or before surgery or coronary intervention. Its clinical significance is uncertain; while the potential risk of sudden cardiac death has been reported in isolated cases, experts consider this anatomic feature has little clinical relevance. (10-12) Although probably most coronary anomalies do not have clinical significance, some studies have shown that clinical implications, while rare, may be relevant in this population. Case series of patients with BAV and TAV have shown that perioperative myocardial injury is greater and the need for postoperative coronary angiography for suspected coronary artery obstruction is higher in patients with coronary anomalies. (4) Even though previous reports have not demonstrated the need for any specific coronary intervention in the surgical procedure of patients with coronary anomalies, repositioning of the ostium by reimplantation of the coronary arteries may be required during David or Bentall-De Bono procedures, increasing the risk of surgery.

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The present study calls for the characterization of the coronary anatomy before heart surgery in patients with BAV to detect any potentially significant coronary anomaly. Despite clinical practice guidelines might not recommend preoperative coronary angiography in young patients without cardiovascular risk factors, an imaging test should always be considered in the population with BAV, though in most cases they are already performed to rule out aortic dilation that cannot be detected by conventional echocardiography. Thus, these findings support the idea of routine CT scanning in this population during follow-up or before any procedure.

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