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Case Report

Quadricuspid Aortic Valve: An Overview

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Abstract

Quadricuspid aortic valve (QAV) is a rare congenital heart disease. The functional status of QAV is predominantly a pure aortic regurgitation. Clinical manifestations of patients with a QAV depend on the functional status of the QAV and the associated disorders. Significant valvular regurgQuadricuspid Aortic Valve: An Overviewitation and (or) stenosis is often present with subsequent operation performed at the fifth to sixth decade of life. The functional status of QAV is predominantly regurgitant; whereas pure stenotic QAV can be as few as in only 0.7% of the patients. QAV is usually an isolated anomaly, but other congenital heart defects can be present in 18- 32% of the patients. About one-fifth of them require a surgical operation. Tricuspidalization is a preferred technique for QAV repair. As not all the patients with a QAV necessarily warrant a surgical operation, decision-making in patient selection and surgical procedure of choice are crucial. Antibiotic prophylaxis against infective endocarditis is necessary in the QAV patients with unequal-sized cusps.

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Introduction:

With the advent of echocardiography and other imaging diagnostic techniques, QAVs are increasingly reported. Perhaps the first retained human awareness of the valvulopathy preceded even medical journals, as Leonardo da Vinci's detailed anatomical illustrations and notes on the typical tricuspid aortic valve dated to 1512 - 13 also includes quadricuspid and bicuspid examples.[1] Dr. Benjamin Guy Babington reported the first known case of quadricuspid aortic valve (QAV) in an 1847 London Medical Gazette article after observing it incidentally while conducting an autopsy on a 34-year-old woman with stunted development.[2] The estimated incidences of QAVs ranges from as low as 0.003% to as high as 1.46%,[3] with its most commonly accepted incidence rate to be between 0.013% and 0.043%.[4-7] The average age of diagnosis is 43.5 ± 21.8 years, with a range of 2 days to 84 years of age.[8] A small male predominance is generally acknowledged, although it has been proposed to be as high as 62%. However, one publication has documented a slightly-higher female prevalence of 52%, and claims the aforementioned male predominance was perhaps due to their higher likelihood of males undergoing aortic valve surgery, for which they accounted for 75% of cases.[8,9] Bicuspid (approximately 2% of the population) aortic valves are the most prevalent aortic anomaly, followed by unicuspid (0.02%) then QAVs, although the borderline scarcity between unicuspid and QAVs generate conflicting data to said rates of occurrences.[10-13] There have only been eight reported cases of quinticuspid, also referred to as pentacuspid aortic valves as of 2016.[14] Quadricuspid pulmonary valves are nine times more prevalent than QAVs, with a comparable minor male predominance.[15]

About 18% to 32% of QAV patients present with an additional congenital heart defect, such as coronary artery and ostium abnormalities, atrial and ventricular septal defect (ASD and VSD, respectively), patent ductus arteriosus, tetralogy of Fallot, sinus of Valsalva fistula, subaortic fibromuscular stenosis, regurgitation and prolapse of the mitral valve, hypertrophic nonobstructive cardiomyopathy, and transposed great

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arteries.[8,16-26] The most common cardiovascular irregularities observed with QAV are deformities in coronary arteries and ostia,[25] with atypical coronary arteries, usually singular, associated with 10% of cases; this is of particular significance due to a reported sudden cardiac death of a previously unremarkable 16-year-old while walking owed to a left coronary ostium dome-like occlusion, with ventricular fibrillation noted as the immediate cause of death.[27] Furthermore, a single case has been reported in a patient with Ehlers-Danlos syndrome,[16] as well as another case amongst identical twins.[28] QAVs' correlation with aortic irregularities is ambivalent; one publication reported it was not associated with aortic dilation (unlike bicuspid aortic valves),[7] while another found common concurrence with ascending aortic dilation and aneurysms.[29]

Classifications

There are two utilized QAV anatomical classification systems. The first one is Hurwitz and Roberts's classification system, seven subtypes were included, lettered A to G, based on the four cusps' relative sizes,[4] with an additional type H supplemented by Vali et al.[30] (Table I, Fig. 1). Based on their findings, approximately 85% of QAVs are of type A, B, or C. Based on a review of the literature by Timperley et al, type B has been reported as the most prevalent,[8] although a later meta-analysis supported type A as being the most common.[31]

The second one is Nakamura's classification system, four subtypes, numbered I to IV, based on the supernumerary cusps' relative position to the left, right, and noncoronary cusps (Table II, Fig. 2) were included. According to this classification's initial publication, the most frequent type encountered is type II, with types I and II respectively corresponding to Hurwitz and Roberts types A and B. However, it should be noted that the most commonly used Hurwitz and Roberts's classification system, as well as Nakamura's by its namesake's own admission, often lacks correspondence with patient treatment and management, as well as with surgical and echocardiographic findings. Thus, additional subtyping categories have been proposed, such as a condensed revision of Hurwitz and Robert's seven subtypes into four concerning their clinical implications on management by Jagannath et al.[9]

Causes

QAVs have been reported in both human and non-human mammals, such as dogs, shrews, and hamsters.[34,35] Both the aortic and pulmonic semilunar valves emerge from mesenchymal ridges in the post-division truncus arteriosus, with the usual three nodules materializing within the vascular lumen after arterial trunk septation during the fifth and ninth weeks of gestation at the junction between the conus and truncus in the aortic and pulmonary ridges.[36] Irregular aortic valves can occur from disordered semilunar cusps primordia development within the aortic trunk wall.[31] The specific etiology of QAVs is yet undetermined.[6] Multiple pathophysiological mechanisms at various points of development have been proposed that might alter the valve cusp number, such as irregular septation of the conotruncus causing unequal distribution of the

Table-I

Hurwitz and Roberts's seven subtypes with an additional Type H[6]

Types	Prevalence	Description
Type A	32%	Four equal sized cusps
Type B	41%	three equal-sized larger cusps and one smaller cusp
Type C	15%	two equal-sized larger cusps and two equal-sized smaller cusps
Type D	3%	one larger cusp, two intermediate cusps, and one smaller cusp
Type E	2%	one larger cusp and three equal-sized smaller cusps
Type F	2%	two equal-sized larger cusps and two unequal-sized smaller cusps
Type G	5%	four unequal-sized cusps Type H: one larger cusp, one intermediate cusp, and two equal-sized smaller cusps
Type H		one larger cusp, one intermediate cusp, and two equal-sized smaller cusps

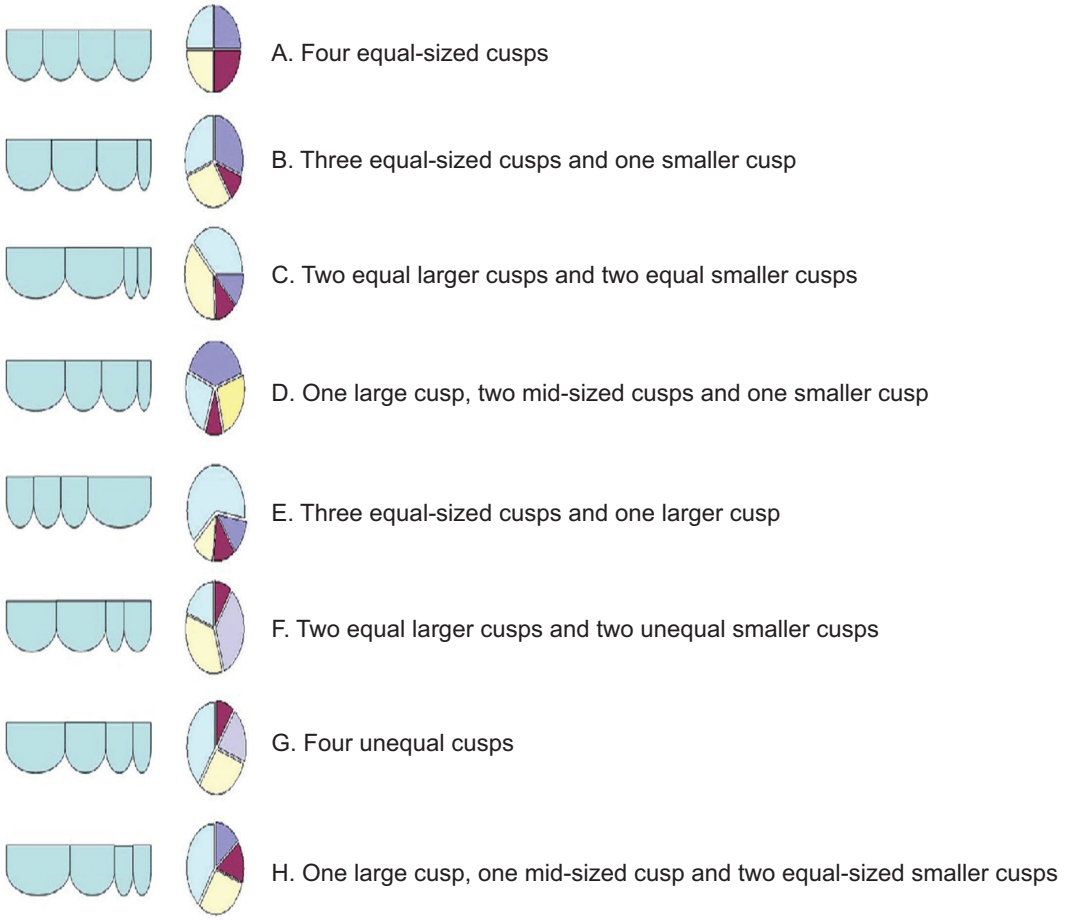

Figure 1: *Hurwitz and Roberts' classification subtypes of quadricuspid aortic valves, including Vali et al's type H supplement. The figure was reprinted from Yuan.*[32]

Table-II
Nakamura's Four Subtypes[33]

Types	Prevalence	Description
Type I	23.8%	Supernumerary cusp between the left and right coronary cusps
Type II	30.9%	Supernumerary cusp between the right and noncoronary cusp
Type III	7.1%	Supernumerary cusp between the left and noncoronary cusp
Type IV	9.5%	Supernumerary cusp indistinguishable due to two equal-sized smaller noncoronary cusps

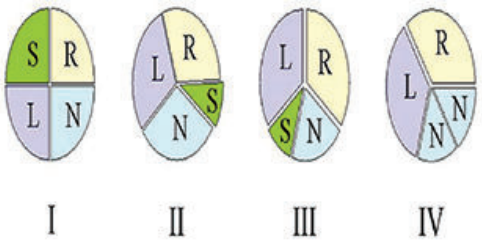

Figure 2: *Nakamura's simplified classification subtypes of quadricuspid aortic valves.*[32]
S: supernumerary cusp; R: right coronary cusp; L: left coronary cusp; N: noncoronary cusp.

distention in each of the great arteries, anomalous proliferation of mesenchymal ridges, or valve cusp division during its formation.[36] The current leading hypothesis involves the partition of one of the three valve cushions due to an invagination of the endothelial layer on the luminal side during an early stage of valve development.[34]

Mode of Presentation

QAV patients may be asymptomatic until their sixth decade, with subsequent symptoms

experienced correlating with the valve's functional status and any associated abnormalities.[32] Congestive heart failure is the most common prevailing presenting diagnosis. As previously mentioned, QAVs can be nonpathological, with symptomatic degeneration cases often associated with prolapsing, conjoint larger cusps, and poor coaptation due to smaller cusps.[37] The most common clinical manifestation observed is aortic regurgitation (AR) without aortic stenosis. [32] Tutarel et al. reported the following in QAV cases: pure AR in 74.7%, AR with stenosis in 8.4%, pure stenosis in 0.7%, and normal function in 16.2%.[38] It has been proposed that Hurwitz and Roberts' type B have a higher likelihood of developing AR due to the single undersized cusp progressively leading to uneven stress distribution, trauma, and malcoaptation,[39] although it has also been suggested that AR is just as likely with type A.[6] The most commonly accepted explanation proposed for said AR is unequal shear stress leading to progressive leaflet fibrosis and partial coaptation eventually failing. Notably, the accessory cusps location has not been found to correspond with the likelihood of developing AR.[40]

There has been one published case in which AI resulting from a QAV progressed to aortic disease, ultimately concluding in patient mortality due to cardiac tamponade after an ascending aortic aneurysm.[41] Structural valve degeneration can increase susceptibility to certain cardiac pathologies, such as rheumatic valves and bacterial endocarditis, which may mask or complicate QAV diagnosis.[42] The associated risk of infective endocarditis was recorded in 1.4% of QAV cases, with a small supernumerary cusp as an anticipating risk factor.[7,42] This risk is believed to be lessened in patients with Hurwitz and Roberts' type A/Nakamura type I, as the equally-sized cusps lack asymmetry or flow disturbance. Diagnosis of a QAV with AR has been recognized as an important risk factor for endocarditis.[43]

Diagnosis

The majority of QAV-presenting cases are only found to be such in their later years. As compared to other aortic valve anatomies, QAV patients may present with AR and be symptomatic at relatively earlier age or sometimes incidentally discovered in pediatric patients.[44] QAV is a differential diagnosis of aortic valve neoplastic involvement, valvular degeneration with or without calcifications, and adherent thrombus or vegetations, such as aortic valve tumors (e.g., papillary fibroelastoma, myxoma).[45,46] True congenital QAVs can be distinguishable by the presence of corpus nodules of Arantius at the center of the free border on each of the four valve's cusps.[46] Modern technological advancements in common clinical practice diagnostics have contributed to a recent increase in QAV detection, as more than 80% of reports have been published after 1980, with echocardiography responsible for 60% of such. Presently, most QAV diagnoses (51.1%) are made via two-dimensional transthoracic or real-time three-dimensional transesophageal echocardiography (TTE and TEE, respectively).[31,47] TTE and TEE can both visualize aortic valve morphology (i.e., cusp number and thickening degree, plus possible vegetations), as well as the size of the aortic root and left ventricle, with TEE being the more sensitive method as it can additionally visualize the coronary ostia. Fewer recent cases of QAV have been initially detected by the relatively historical methods of surgical examination (22.6% of documented incidences) and pathological inspection (15.6%), and rarely is it diagnosed by aortography (6.5%) or the few cases by cardiac magnetic resonance imaging (CMR).[46] It has been proposed that current cardiac imaging, such as CMR and multi-detector cardiac computerized tomography (CCT) can detect QAV via cross-sectional imaging.[48] In addition to diagnoses, CMR can detect QAV morphology, AR presence and volume, valve stenosis due to calcification, cardiac function and chamber sizes, as well as possible associated disorders (e.g., stenotic pulmonary valve, hypertrophic cardiomyopathy, a patent ductus arteriosus, sinus of Valsalva aneurysm, and coronary artery conditions.[46-49]

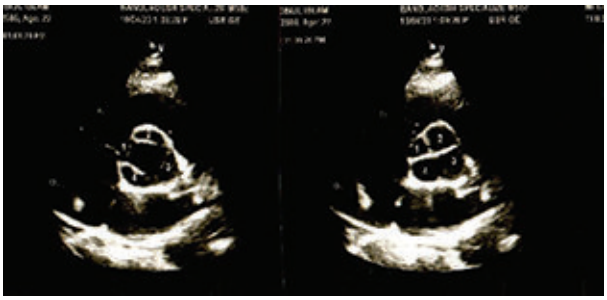

Figure 3: *A Hurwitz and Roberts' type B/ Nakamura type I quadricuspid aortic in both diastolic and systolic phases*

Management

Surgical indications include severe aortic stenosis,[50] but principally severe AR and dysfunctional QAV linked with other lesions (e.g., left coronary ostium occlusion).[8] The various methods by which these can be surgically corrected include biological or mechanical valve implantation, Ross procedure, Bentall procedure, and bi-/tricuspidization (with or without root replacement).[51] Approximately one-fifth of QAV cases require surgical intervention. Valve repair, via the aforementioned methods collectively referred to as aortic valve reconstruction surgery (AVRS), favored versus replacement.[44]

If feasible, particularly among younger candidates, repair is preferred as to avoid valve replacement-associated risks such as bleeding, endocarditis, thromboembolism, and prosthetic valve deterioration.[44] The objective of AVRS is to reestablish proper coaptation while minimizing transvalvular gradient and turbulent flow to achieve lasting durability.[52] This intention is predominantly undertaken by varied tricuspidization techniques, although QAV bicuspidization has been documented twice. Bicuspidization was first reported in 2010 by Luciani et al as the four unequaled cusps of a Hurwitz and Roberts' type G QAV posed geometrical challenges hindering tricuspidization.[53] Shimamoto et al published the second instance of QAV bicuspidization in 2014 after an unsuccessful tricuspidization produced an incompetent valve, although they still recommend routinely attempting tricuspidization first as it is surgically simpler and physiologically corresponds better.[54]

Surgical decision making can be influenced by a multitude of factors, including patient symptoms, AR severity, left ventricular dilation or dysfunction, and corresponding aortopathy with aneurysm.[55] Additionally, a few Ross procedures (subcoronary inclusion technique), also known as pulmonary autograft procedures, have successfully treated QAV while reducing aortic root dilation risk.[56,57] Compared with traditional mechanical or bioprosthetic replacements, autografting a patient's own pulmonary valve (identical in size, shape, and strength to their aortic valve) minimizes future valve re-replacement probability and does not require lifetime blood thinner medication use. A less invasive and significantly less popular alternative to AVRS specifically for stenosed QAVs is transcatheter aortic valve replacement (TAVR), also referred to as transcatheter aortic valve implantation (TAVI), with its first documented case and TAVR/TAVI as its treatment published in 2011 by Blanke et al.[58]

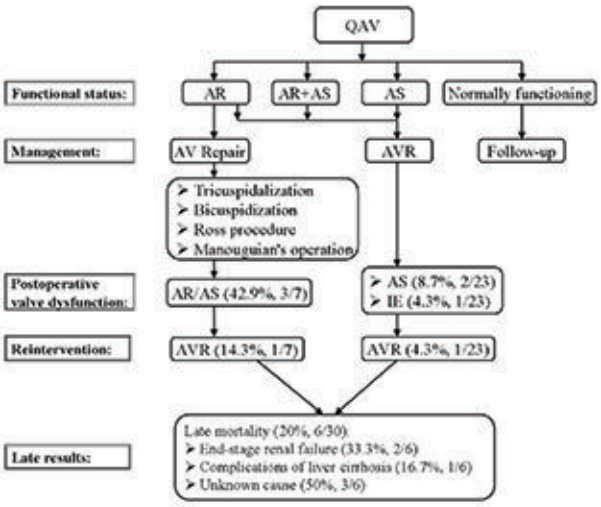

Figure 4: *Management and prognosis of patients with a quadricuspid aortic valve.*[59]

AR=aortic regurgitation; AS=aortic stenosis; AV=aortic valve; AVR=aortic valve replacement; IE=infective endocarditis; QAV=quadricuspid aortic valve

Follow-Up

Due to the aforementioned small number of QAVs documented in medical literature, its natural history of disease is not of defined conclusions. Nonpathological QAV cases still justify attentive recurring monitoring and clinical follow-ups, as frequently the valve can develop progressive hemodynamic concessions.[60] This is especially true for younger QAV presenting patients, considering their recurrent need for AVRS later in life. Echocardiograms are customarily performed both pre- and postoperative alongside surgical correction, as well as at hospital discharge and annually if no complications present.[52] Long-term follow-ups to gauge the lastingness of an operation are warranted as this is still a relatively rare occurrence with only a few recent review publications.[52]

Conclusions:

Most of the patients with a QAV develop aortic valve incompetency at the fifth to sixth decade of life. About one-fifth of them require a surgical operation. Although tricuspidalization is a preferred repair technique for QAV with significant AR, the associated aortopathy could be a predictive risk factor of late failure of aortic repair. As not all the patients with a QAV necessarily warrant a surgical operation, decision-making in patient selection and surgical procedure of choice are crucial. The aortic valve repair of panegyric was started later and the procedural choice was determined by the feasibility concerning the QAV condition and surgeon's preference. Antibiotic prophylaxis against infective endocarditis is necessary in the QAV patients with unequal-sized cusps.

Conflict of Interest - None.

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