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A Review of Bicuspid Aortic Valve Anomaly

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Abstract

Bicuspid aortic valve (BAV) is a common congenital heart valve disease, often accompanied by aortic valve stenosis or regurgitation, infective endocarditis, and aortic diseases such as aortic dilation and dissection. This article aims to review the definition, pathogenesis and genetic basis, classification methods, clinical features and current status of research and treatment and prognosis of BAV, and provide reference for the diagnosis and treatment of BAV. The content and structure of this article are as follows: The first part introduces the definition of BAV; The second part introduces the etiology and classification methods of BAV; The third part briefly describes the clinical manifestations and diagnosis of BAV; The fourth part discusses the treatment strategies and prognosis prediction of BAV; Finally, the article summarizes and looks forward to the future research directions.

Keywords

Bicuspid Aortic Valve, Congenital Heart Disease, Aortic Valve Disease, Transcatheter Aortic Valve Replacement

1. Introduction

Bicuspid Aortic Valve (BAV) is a congenital heart valve disease where the aortic valve has two leaflets instead of the normal three. BAV is one of the most common types of congenital heart disease in adults, with a prevalence of approximately 0.5% - 2% in the population [1]. BAV is often associated with aortic valve stenosis or insufficiency, as well as aortic dilation and aortic dissection [1] [2]. The occurrence of BAV is related to various factors, including genetics, environment, and development, and there are significant individual differences in its classification,

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progression, and prognosis [3] [4]. In recent years, with the advancement of imaging technology, the diagnosis and assessment of BAV have become more precise. Additionally, surgical procedures and transcatheter aortic valve replacement (TAVR) have been continuously improved, providing more options for BAV patients [5].

2. Causes and Classification of Bicuspid Aortic Valve (BAV)

2.1. Causes of Bicuspid Aortic Valve (BAV)

The occurrence of BAV is a multifactorial process involving genetic, molecular biological, embryological, and hemodynamic factors [2]. The genetic basis of BAV remains unclear, but it is known to have familial aggregation and is associated with chromosomal abnormalities (such as Turner syndrome) or other congenital heart diseases (such as Shone syndrome) [2] [6]. The molecular biological mechanisms of BAV include regulation at various levels, such as transcription factors, signaling pathways, extracellular matrix, and epigenetics. The embryological origin of BAV is related to the developmental process of the aortic valve, primarily due to abnormal proliferation, migration, or apoptosis of neural crest endothelial cells, leading to changes in the number, morphology, and function of the leaflets. The hemodynamic impact of BAV mainly manifests during the opening and closing of the aortic valve. Due to the asymmetry or fusion of the leaflets, the stress distribution on the aortic valve is uneven, resulting in deformation, calcification, and fibrosis of the leaflets, as well as dilation, rupture, and dissection of the aortic wall [6].

2.2. Classification Methods of BAV

There are various classification methods for BAV, each with different bases, advantages, disadvantages, and clinical applications. The common classification methods include:

2.2.1. Sievers Classification

This classification is based on the number, location, and function of the raphes between leaflets, dividing BAV into three types: Type 0 (no raphe), Type 1 (one raphe), and Type 2 (two raphes). Each type is further subclassified according to the spatial arrangement of the leaflets/raphes and the function of the leaflets [7]. This classification method is straightforward, practical, easy to remember, and widely used in surgical practice, correlating with the choice of surgical procedures and outcomes. It is also adopted by the STS surgical database. However, this method does not encompass all BAV malformation types, and some Type 2 BAV cases may actually be variants of unicuspid or tricuspid valves. Schematics can be found in **Figure A1** and **Figure A2**.

2.2.2. Schaefer Classification

This classification is based on the fusion position of the leaflets relative to the coronary arteries and the structural characteristics of the aorta, dividing BAV into three types: Type 1 (right-left fusion), Type 2 (no right fusion), and Type 3 (no left fusion). Each type is further divided into three subtypes according to aortic structure: N (normal), A (ascending aorta dilation), and E (no sinus). This classification method, based on echocardiographic diagnosis, is the first to integrate BAV and aortic structure into a unified classification, providing insights into the progression and risk assessment of BAV complications. However, this method does not describe the detailed geometry of the leaflets or the degree of fusion at the commissures or raphes, which may be critical factors affecting clinical outcomes of TAVR. Schematics can be found in **Figure A3**.

2.2.3. TAVR-Oriented Classification

Based on the number of leaflet commissures, presence of raphes, and leaflet opening direction observed in CT images, this classification divides BAV into three types: Tricommissural (three commissures), Bicommissural raphe-type (two commissures with raphes), and Bicommissural non raphe-type (two commissures without raphes). Each type is further subclassified into coronary leaflet fusion type and mixed leaflet fusion type based on leaflet opening direction or fusion involvement. This classification method, based on CT structural analysis, considers the geometric features of the leaflets and the structure of the aortic root, providing guidance for the technical success and clinical outcomes of TAVR. However, the Tricommissural type in this classification may represent a variant of the tricuspid valve rather than a true BAV. Schematics can be found in **Figure A4**.

2.2.4. 2021 BAVcon Classification

This classification, based on the number of Valsalva sinuses, the degree of raphe fusion between leaflets, and the spatial geometry of the leaflets, divides BAV into three types: Fused BAV (three Valsalva sinuses with raphes), 2-sinus BAV (two Valsalva sinuses without raphes), and Unicuspid aortic valve (one Valsalva sinus without raphes). Each type is further subclassified based on leaflet fusion position or geometric characteristics [8]. This classification method integrates previous research findings and considers a comprehensive set of factors, including the morphology of the leaflets, the structure of the aortic root, and features of the ascending aorta, providing a more accurate and detailed description. It is the latest international expert consensus classification. Schematics can be found in **Figure A5**.

2.3. The Correlation between BAV Classification, Complications, and Prognosis

The correlation between BAV classification and its complications and prognosis is a hot research area, with different classification methods yielding varying results. Generally, right-left fusion BAV is more prone to aortic valve stenosis and calcification, no-right fusion BAV is more prone to aortic valve regurgitation and aortic dilation, and no-left fusion BAV is more prone to aortic dissection. The geometric morphology, opening direction, and degree of fusion of the leaflets can also affect BAV function and the occurrence of complications. Additionally, the structure and size of the aortic root and ascending aorta are crucial factors affecting BAV prognosis. Therefore, BAV classification should comprehensively consider multiple aspects of the leaflets and aorta to conduct risk assessment and make treatment decisions [9].

3. Clinical Manifestations and Diagnosis of BAV

3.1. Clinical Manifestations of BAV

The clinical manifestations of BAV depend on the valve function, aortic lesions, and the presence and severity of other complications. The common clinical manifestations and complications of BAV include:

3.1.1. Aortic Valve Stenosis

BAV is the most common cause of aortic valve stenosis in adults, accounting for about 50% of cases. Due to the asymmetry or fusion of the leaflets in BAV, deformation, calcification, and fibrosis are likely to occur, leading to reduced valve orifice area, increased transvalvular pressure gradient, left ventricular hypertrophy, and decreased compliance. The severity of aortic valve stenosis can be graded based on the orifice area, transvalvular pressure gradient, and peak aortic ejection velocity. Typical symptoms of aortic valve stenosis include angina, syncope, and heart failure. The diagnosis of aortic valve stenosis mainly relies on echocardiography, supplemented by CT or MRI for evaluation [1] [10]. Treatment options for aortic valve stenosis include medication, transcatheter aortic valve replacement (TAVR), and surgical aortic valve replacement (SAVR).

3.1.2. Aortic Valve Regurgitation

BAV is one of the common causes of aortic valve regurgitation, accounting for about 10% of cases. Due to the asymmetry or fusion of the leaflets in BAV, deformation, tearing, or thickening may occur, leading to incomplete valve closure, increased regurgitant volume, left ventricular dilation, and decreased contractile function. The severity of aortic valve regurgitation can be graded based on regurgitant volume, regurgitant fraction, regurgitant area, and effective regurgitant orifice area. Typical symptoms of aortic valve regurgitation include palpitations, dyspnea, and heart failure. The diagnosis of aortic valve regurgitation mainly relies on echocardiography, supplemented by CT or MRI for evaluation. Treatment options for aortic valve regurgitation include medication, transcatheter aortic valve repair (TAVR), and surgical aortic valve repair (SAVR) [1] [11].

3.1.3. Infective Endocarditis

BAV is a risk factor for infective endocarditis, accounting for about 10% of cases. The asymmetry or fusion of the leaflets in BAV makes them more susceptible to damage or inflammation, leading to the attachment and growth of bacteria or fungi, forming vegetations, causing valve dysfunction, embolic complications, or pericarditis. Typical symptoms of infective endocarditis include fever, chills, anemia, clubbing, and petechiae. The diagnosis of infective endocarditis mainly relies on blood cultures and echocardiography, supplemented by CT or MRI for evaluation. Treatment includes antibiotics and transcatheter or surgical valve intervention [1] [12].

3.1.4. Aortic Dilation

BAV is a risk factor for aortic dilation, accounting for about 50% of cases. The asymmetry or fusion of the leaflets in BAV leads to abnormal hemodynamic effects, resulting in uneven stress distribution on the aortic wall, causing dilation, rupture, or dissection of the aortic root and/or ascending aorta. The severity of aortic dilation can be graded based on aortic diameter, dilation rate, and extent. Typical symptoms of aortic dilation include chest pain, cough, dyspnea, and hoarseness. The diagnosis of aortic dilation mainly relies on echocardiography, supplemented by CT or MRI for evaluation. Treatment includes medication, transcatheter, or surgical aortic intervention [1] [13].

3.2. Diagnosis of BAV

There are various diagnostic methods for BAV, each with different accuracy, sensitivity, specificity, and feasibility. Common diagnostic methods include:

3.2.1. Echocardiography

Echocardiography is the preferred method for diagnosing BAV [14], offering high accuracy, sensitivity, and specificity, and is also a crucial tool for evaluating valve function, aortic lesions, and other complications. The advantages of echocardiography include being non-invasive, convenient, cost-effective, and repeatable. However, it depends on the operator's skill and experience and sometimes finds it difficult to distinguish between bicuspid and tricuspid valve variants, and it cannot provide comprehensive three-dimensional structural information. The diagnostic criteria for echocardiography include observing two leaflets or a raphe in the short-axis view or asymmetrical or eccentric leaflet opening in the long-axis view. Echocardiography is indicated for patients with heart murmurs, symptoms, or a family history, as well as for patients with other congenital heart diseases or aortic lesions [1] [6] [15]. It should be performed at the initial detection of abnormalities or when symptoms appear, and regularly during follow-up.

3.2.2. CT

CT is an auxiliary method for diagnosing BAV [14], with high accuracy, sensitivity, and specificity. It is also an important tool for evaluating the structure and morphology of the aorta. The advantages of CT include providing clear three-dimensional structural information, well demonstrating the geometric characteristics of the leaflets, the degree of fusion, and calcification, and guiding the suitability and prognosis of TAVR. However, CT has disadvantages such as radiation exposure, the need for contrast agents, high cost, and inability to dynamically observe valve function. The diagnostic criteria for CT include observing two leaflets or a raphe in the short-axis view or asymmetrical or eccentric leaflet opening in the long-axis view. Indications for CT include patients with unclear or inconsistent echocardiography results or those with indications for TAVR [1] [8] [16]. The timing for CT diagnosis is when echocardiography cannot confirm the diagnosis or further evaluation is needed, or before TAVR.

3.2.3. MRI

MRI is an auxiliary method for diagnosing BAV [14], with high accuracy, sensitivity, and specificity. It is also a key tool for evaluating aortic function and hemodynamics. The advantages of MRI include providing comprehensive three-dimensional structural and functional information, well demonstrating the morphology, movement, and regurgitant volume of the leaflets, and evaluating the stress, elasticity, and shear force of the aortic wall, which is significant for assessing the progression and risk of aortic lesions. However, MRI is non-invasive, has high costs, cannot use metal devices, and is affected by breathing and heart rate. The diagnostic criteria for MRI include observing two leaflets or a raphe in the short-axis view or asymmetrical or eccentric leaflet opening in the long-axis view. Indications for MRI include patients with unclear or inconsistent echocardiography or CT results or those with abnormal aortic function or hemodynamics [1] [8] [16]. The timing for MRI diagnosis is when echocardiography or CT cannot confirm the diagnosis or further evaluation is needed, or during regular follow-up.

3.3. Diagnostic Criteria, Indications, and Timing for BAV

The diagnostic criteria for BAV include observing two leaflets or a raphe on echocardiography, CT, or MRI, or observing asymmetrical or eccentric leaflet opening in the long-axis view [15] [16]. BAV is classified based on the fusion position and direction of the leaflets into right-left fusion type, no right fusion type, and no left fusion type [4] [7]-[9]. The indications for BAV diagnosis include patients with heart murmurs, symptoms, or a family history, as well as patients with other congenital heart diseases or aortic lesions [1] [5] [6]. The timing for BAV diagnosis is at the initial detection of abnormalities or when symptoms appear, and during regular follow-up [1] [5].

The diagnostic process and timing for BAV should be individualized based on the patient's clinical situation and available resources. Generally, the following is a possible diagnostic workflow (**Figure 1**).

4. Treatment and Prognosis of BAV

4.1. Treatment of BAV

The treatment principles for BAV are to formulate individualized treatment plans based on the patient's symptoms, valve function, aortic lesions, and other complications to improve quality of life, slow disease progression, prevent complications, and reduce mortality risk [1] [10]. The treatment goals for BAV are to maintain valve function, protect the aortic wall, prevent infective endocarditis, and provide anticoagulant therapy. The treatment strategies for BAV include medication, surgical intervention, and transcatheter aortic valve replacement (TAVR) [1] [5] [11]. Different treatment methods have varying effects, safety, and indications.

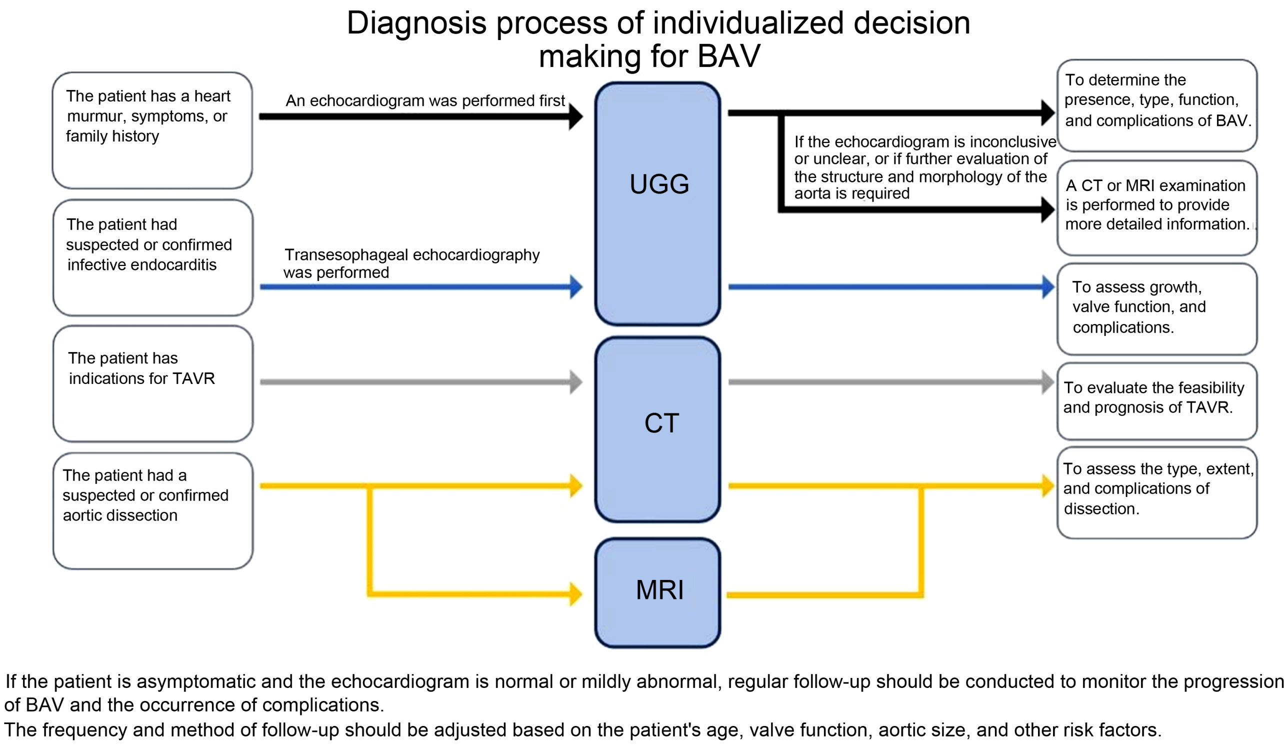

Figure 1. Possible diagnostic process.

4.1.1. Medication

Medication is the foundational treatment for BAV [1] [5], mainly used for asymptomatic or mildly symptomatic patients and as a bridge therapy for surgery or TAVR. The goals of medication are to control blood pressure, reduce cardiac load, improve heart function, prevent infective endocarditis, and provide anticoagulant therapy. Types of medications include angiotensin-converting enzyme inhibitors (ACEIs), angiotensin receptor blockers (ARBs), beta-blockers, calcium channel blockers, diuretics, antibiotics, and anticoagulants. The advantages of medication are non-invasiveness, convenience, and cost-effectiveness, while the disadvantages include the inability to change the structure and function of the valve, prevent the progression of aortic lesions, or reduce the risk of surgery or TAVR [8] [12].

4.1.2. Surgical Intervention

Surgical intervention is the traditional treatment for BAV [1] [5], primarily used for symptomatic patients or those with severe valve dysfunction, aortic lesions, or other complications. The goals of surgical intervention are to repair or replace the valve and to repair or replace the aorta. Types of surgical procedures include aortic valve repair (AVR), surgical aortic valve replacement (SAVR), aortic root (sinus) repair or replacement, and ascending aorta replacement [11]. The advantages of surgical intervention are the effective improvement of valve function and aortic lesions, increased survival rate, and improved quality of life. However, it is invasive, has high risks and costs, and may lead to complications such as bleeding, infection, arrhythmias, stroke, and long-term issues like prosthetic valve deterioration or thrombosis [8] [12].

4.1.3. TAVR

TAVR is an emerging treatment for BAV [8], mainly used for symptomatic patients or those with severe valve dysfunction who cannot undergo surgical intervention or are at high surgical risk. The goal of TAVR is to improve valve function by implanting a prosthetic valve within the native valve via a catheter. Types of TAVR valves include self-expanding valves and balloon-expandable valves, with various catheter access routes such as femoral artery, subclavian artery, carotid artery, and direct aortic access. The advantages of TAVR are minimally invasive, convenient, and rapid, effectively improving valve function and survival rates. However, it is costly, has multiple complications, and may result in aortic valve regurgitation, cardiac conduction system damage, vascular complications, pericardial tamponade, stroke, and long-term issues like prosthetic valve deterioration or thrombosis [8] [12].

4.2. Prognostic Factors of BAV

There are various prognostic factors for BAV, each with different levels of influence and mechanisms. The common prognostic factors include:

4.2.1. Symptoms

Symptoms are a critical prognostic factor for BAV [1] [14]. Patients with symptoms generally have a worse prognosis than those without, primarily because symptoms reflect the severity of valve dysfunction and cardiac impairment. The type and severity of symptoms are also related to prognosis. Typically, angina, syncope, and heart failure are symptoms of aortic valve stenosis; palpitations, dyspnea, and heart failure are symptoms of aortic valve regurgitation; chest pain, cough, difficulty breathing, and hoarseness are symptoms of aortic dilation; fever, chills, anemia, clubbing, and petechiae are symptoms of infective endocarditis. The emergence and worsening of symptoms are important indicators for surgery or TAVR, and timely treatment can improve prognosis.

4.2.2. Valve Function

Valve function is a significant prognostic factor for BAV [1] [14]. The degree and type of valve dysfunction are related to prognosis because valve dysfunction affects cardiac load and output, leading to changes in cardiac function and ventricular remodeling. The evaluation of valve function includes indices such as orifice area, transvalvular pressure gradient, peak aortic ejection velocity, regurgitant volume, regurgitant fraction, regurgitant area, and effective regurgitant orifice area, which can be measured and graded using echocardiography, CT, or MRI. Improving valve function is a primary treatment goal, and surgery or TAVR can effectively enhance valve function and reduce mortality risk.

4.2.3. Aortic Lesions

Aortic lesions are significant prognostic factors for BAV [1] [6] [14]. The degree and type of aortic lesions are related to prognosis because they affect the stress, elasticity, and shear force of the aortic wall, leading to dilation, rupture, or dissection of the aortic root and/or ascending aorta. The evaluation of aortic lesions includes indices such as aortic diameter, dilation rate, and extent, which can be measured and graded using echocardiography, CT, or MRI. Controlling aortic lesions is a primary treatment goal, and medication, surgery, or TAVR can effectively manage aortic lesions and reduce complications.

4.2.4. Other Complications

Other complications are significant prognostic factors for BAV [1] [14]. The presence of other complications is related to prognosis because they increase the burden and damage to the heart, leading to worsening cardiac function and increased mortality risk. The evaluation of other complications includes indicators such as infective endocarditis, arrhythmias, pericarditis, and stroke, which can be diagnosed and graded using blood cultures, ECG, pericardiocentesis, head CT, or MRI. The prevention and treatment of other complications are primary treatment goals, and antibiotics, anticoagulants, anti-arrhythmic drugs, pericardiocentesis, and vascular interventions can effectively prevent and treat other complications, reducing mortality risk.

4.3. Prognostic Evaluation Methods for BAV

There are various prognostic evaluation methods for BAV, each with different bases, advantages, disadvantages, and application scenarios. The common prognostic evaluation methods include:

4.3.1. Risk Scores

Risk scores are commonly used for prognostic evaluation of BAV [1] [14] [17], mainly to assess the risk and prognosis of surgery or TAVR. The basis of risk scores is to calculate the probability of mortality or other adverse events based on multiple clinical variables and statistical models. The advantages of risk scores are simplicity, speed, and objectivity, while the disadvantages are the inability to reflect individual differences, adapt to new treatments, or predict long-term prognosis. Types of risk scores include the European System for Cardiac Operative Risk Evaluation (EuroSCORE), the American Heart Association/American College of Cardiology (AHA/ACC) risk score, and the Society of Thoracic Surgeons (STS) risk score. The application scenarios for risk scores are preoperative risk assessment and decision-making for surgery or TAVR, as well as postoperative prognostic evaluation and follow-up.

4.3.2. Survival Curves

Survival curves are commonly used for prognostic evaluation of BAV [1] [12] [14]

[17], primarily to assess the prognostic differences of different treatment methods or classification methods. The basis of survival curves is to plot the relationship between survival time and survival probability based on follow-up data, comparing whether there are significant differences in survival curves between different groups. The advantages of survival curves are visual clarity, objectivity, and dynamic representation, while the disadvantages are being influenced by follow-up time, follow-up rate, and follow-up methods, and not reflecting the quality of life and functional status of patients. Types of survival curves include Kaplan-Meier curves, Cox proportional hazards models, and cumulative risk curves. The application scenarios for survival curves are postoperative prognostic comparison and analysis, as well as prognostic monitoring and evaluation during follow-up.

4.3.3. Other Evaluation Methods

Besides risk scores and survival curves, there are other evaluation methods mainly used to assess the pathophysiological characteristics, valve function, aortic lesions, cardiac function, and myocardial ischemia in BAV. The basis of these evaluation methods is to reflect the cardiovascular status and risk of patients based on different examination techniques and indicators. The advantages of these evaluation methods are comprehensive, thorough, and accurate, while the disadvantages are complexity, time consumption, and high cost. Types of these evaluation methods include echocardiography, electrocardiogram, electrocardiographic stress test, cardiac nuclear imaging, cardiac CT, and cardiac MRI [14]-[17]. The application scenarios for these evaluation methods are preoperative condition assessment and risk stratification for surgery or TAVR, as well as postoperative efficacy evaluation and complication monitoring.

4.4. Improvement Measures for BAV

The main improvement measures for BAV [1] [5] [8] [12] include medication, surgical intervention, and transcatheter aortic valve replacement (TAVR). Different measures are suitable for different patient groups and should be selected based on the patient’s condition, prognosis, risk, and preferences.

5. Conclusions

This article aims to review the latest developments in the epidemiology, etiology, classification, clinical manifestations, diagnosis, treatment, and prognosis of BAV, providing references for clinical practice and scientific research. The main points and conclusions are as follows:

1) BAV is a common congenital heart disease with a prevalence of about 1% to 2%. Its pathogenesis is unclear, but it may be related to genetic, environmental, and developmental factors.

2) The clinical manifestations of BAV vary, depending on valve function, aortic lesions, and the presence and severity of other complications. Common clinical manifestations and complications of BAV include aortic valve stenosis, aortic

valve regurgitation, infective endocarditis, aortic dilation, and dissection.

3) There are various diagnostic methods for BAV, each with different accuracy, sensitivity, specificity, and feasibility. Echocardiography is the preferred method for diagnosing BAV, while CT and MRI are auxiliary methods. The diagnostic criteria for BAV include observing two leaflets or a raphe in the short-axis view or asymmetrical or eccentric leaflet opening in the long-axis view. Indications for BAV diagnosis include patients with heart murmurs, symptoms, or a family history, as well as those with other congenital heart diseases or aortic lesions. The timing for BAV diagnosis is at the initial detection of abnormalities or when symptoms appear, and during regular follow-up.

4) There are various treatment methods for BAV, each with different effects, safety, and indications. Medication is the foundational treatment for BAV, surgical intervention is the traditional treatment, and TAVR is the emerging treatment. The treatment principles for BAV are to formulate individualized treatment plans based on the patient's symptoms, valve function, aortic lesions, and other complications to improve quality of life, slow disease progression, prevent complications, and reduce mortality risk. The treatment goals for BAV are to maintain valve function, protect the aortic wall, prevent infective endocarditis, and provide anticoagulant therapy.

5) There are various prognostic factors for BAV, each with different levels of influence and mechanisms. Symptoms, valve function, aortic lesions, and other complications are important prognostic factors affecting the patient's survival rate and quality of life. There are various prognostic evaluation methods for BAV, each with different bases, advantages, disadvantages, and application scenarios. Common methods include risk scores and survival curves, with other methods including echocardiography, electrocardiogram, electrocardiographic stress test, cardiac nuclear imaging, cardiac CT, and cardiac MRI.

6. Limitations and Shortcomings

The main limitations and shortcomings of this article include:

1) The literature review is mainly based on foreign studies, with fewer domestic studies, which may result in regional differences and a lack of representativeness.

2) The research methods are primarily literature analysis, lacking empirical studies and unable to provide direct evidence and data support.

3) The research results are mainly theoretical, lacking practical applications, and unable to provide specific operational guidance and recommendations.

7. Outlook and Recommendations

The main outlook and recommendations include:

1) Strengthen domestic and international cooperation and exchange to improve the research and clinical levels of BAV, providing better services and protection for BAV patients.

2) Deepen the investigation of BAV pathogenesis and genetic basis, searching for molecular markers and predictive models of BAV to provide evidence and means for early diagnosis and prevention.

3) Optimize BAV evaluation methods and improvement measures to enhance diagnostic accuracy and treatment efficacy, reduce complications and mortality risk, and improve prognosis and quality of life for BAV patients.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Appendix Figures

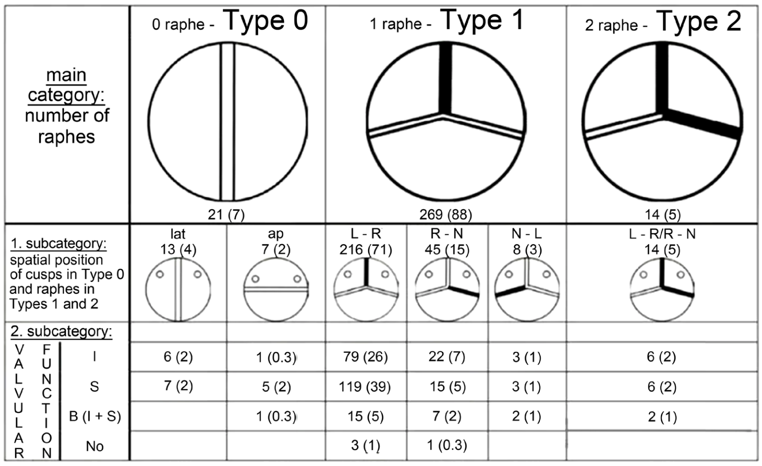


Figure A1. Sievers classification diagram.

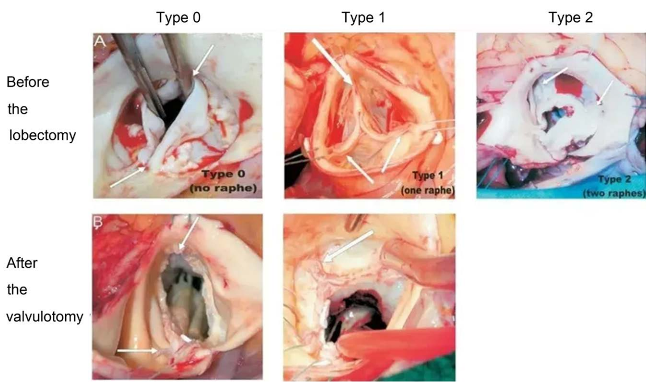


Figure A2. Sievers classification actual case diagram.

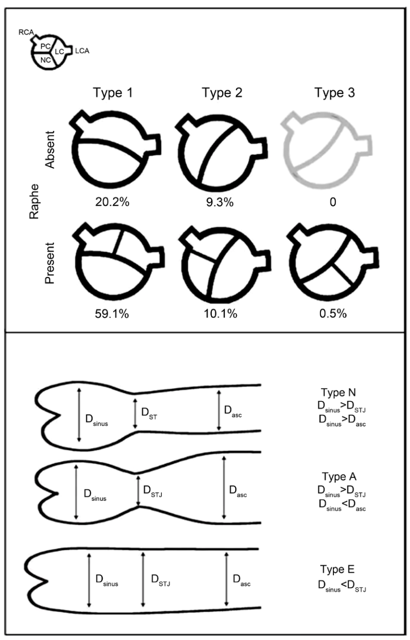
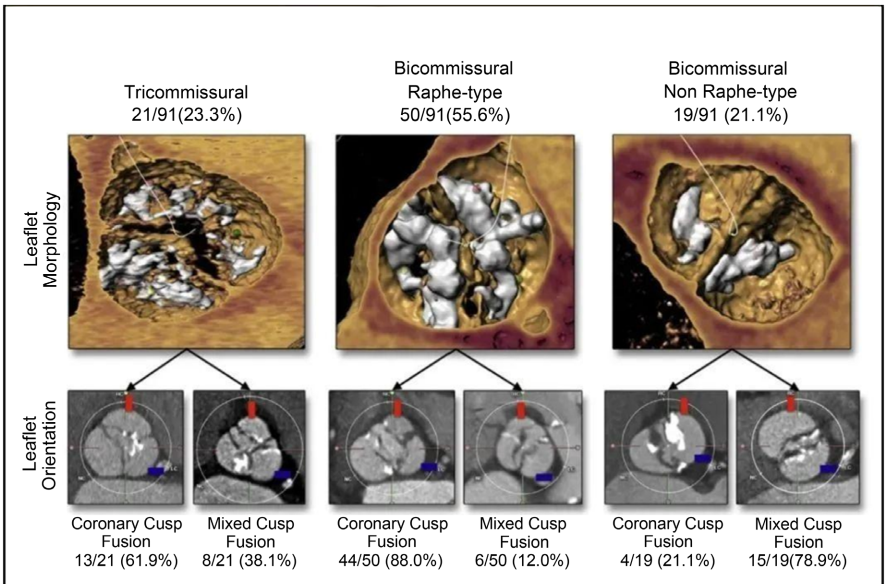


Figure A3. Schaefer classification diagram.



(Top) Leaflet morphology is classified on the basis of number of commissures (2 or 3) and, in the presence of 2 commissures, the presence or absence of a raphe. This classification yields tricommissural, bicommissural raphe type, and bicommissural non-raphe types. **(Bottom)** Leaflet orientation is classified on the basis of cusp fusion, which is either coronary cusp fusion or mixed non-coronary-coronary cusp fusion. Take off of the right coronary artery is indicated by the **red line**: take off of the left coronary artery is indicated by the **blue line**, Values, for example. 21 of 91 (23.39%), are overall frequency of BAV treated with TAVR relative to overall TAVR cases, BAV = bicuspid aortic valve; TAVR = transcatheter aortic valve replacement.

Figure A4. TAVR oriented classification diagram.

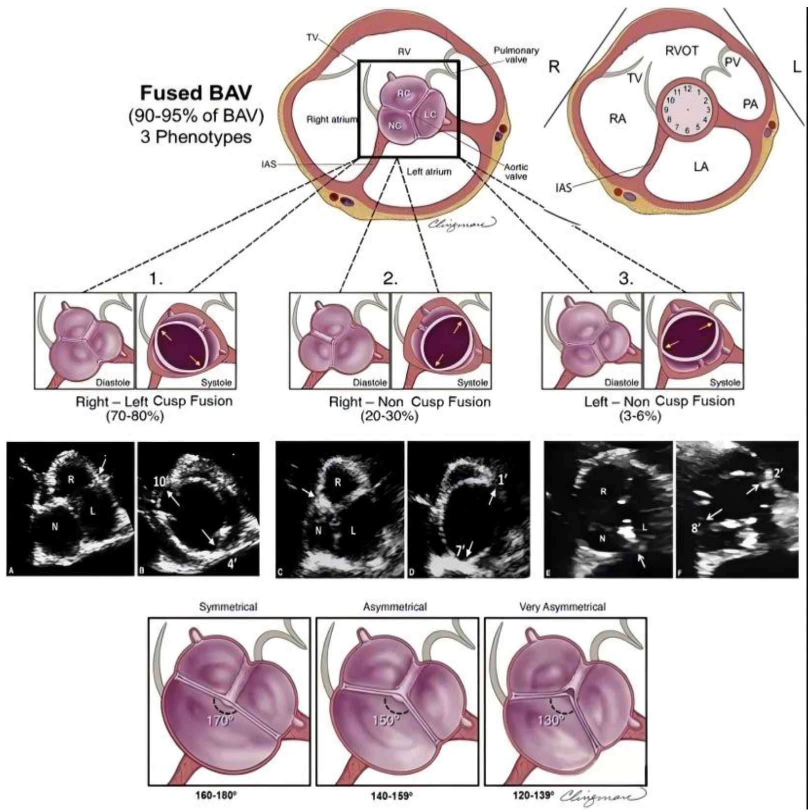


Figure A5. 2021 BAVcon classification diagram.