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# The characteristic of fusion bicuspid aortic valve in pediatric patients at Ramathibodi Hospital

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**Abstract**

Bicuspid aortic valve disease is considered a congenital condition. The severity of the disease depends on the effects on the valve, whether it is stenosis or leakage, or whether it causes aortic dilatation. The cause is related to genes and genetics causing the cusp to fuse or malfunction of commissure. The most common types are left and right coronary cusp fused (L+R). The echocardiography is considered the Golden standard equipment of imaging to rule out and classification fused especially valvular, subvalvular and supravalvular structure and complication. The search for pathology required observation of the parasternal short axis at the aortic valve level, cross section of aortic valve opening and closed was show raphe scar and coronary was identified cusp. Echocardiographic data will inform the rationale of treatment selection, patient behavior, and postoperative care. Especially the left and right coronary cusp of aortic fused was a common type. This data was similar and nearly another study in western**.** Therefore, cardiologist, cardio thoracic technologists or sonographer should have the knowledge and skills to analyze and diagnose bicuspid aortic valve abnormalities**.**

**Keyword** : Aortic valve, Bicuspid aortic valve, Echocardiography, Pediatrics

## Introduction

Bicuspid aortic valve (BAV) fusion is a common cardiac abnormality found on echocardiography, in which the semilunar valve commissure is fused. There are only two or one gap openings, instead of the original three aortic valve leaflets and three commissures. (Figure 1 A-B) This condition causes blood to flow inconveniently, leading to a narrowing of the blood passageway around the heart valve, called aortic stenosis (AS). And when the heart relaxes, when the valves are connected, the deformed sutures cannot close together tightly, causing blood to flow back into the left ventricle, called aortic regurgitation (AR). In normal people, the aortic valve consists of three leaflets that fit together and act like a door that opens and closes to allow blood to flow through. However, in some patients, there may be abnormalities in the structure and function of the aortic valve due to some of the leaflets being connected. This makes the images found on the echocardiogram appear to have only two leaflets (Bicuspid aortic valve; BAV). This echocardiogram can diagnose which leaflets are connected. Bicuspid aortic valve is a common condition compared to other heart abnormalities, most of which are congenital and genetic, causing abnormal aortic valve function, such as stenosis and regurgitation. Prevalence figures vary widely depending on study method, with most reporting a higher incidence in males than females, and if female, the prevalence is more likely to be associated with Turner syndrome. Alan C Braverman et al. reported that bicuspid aortic valve is found in approximately 0.5-1.4% of the general population, which is similar to the study of Hammond GL Et al And the study of Nuran Yener et al. which found in approximately 0.9-2.0% of the general population, with a prevalence of 54% in males. [1,2] Currently, echocardiography is considered the standard method for characterizing pathology, although the results of this instrument are highly dependent on the experience of the examiner (Depend on operator). This article aims to give knowledge to cardio-thoracic technologists and health care personnel involved in cardiac imaging diagnosis to know the types, numbers and be able to classify the characteristics of aortic valve leaflet fusion, the author has created this article.

**Objective**

This article aim to present type and classification of leaflet fusion in bicuspid aortic valve.

**Source of information**

Review articles and research related to aortic valve abnormalities, especially bicuspid aortic valve, both domestically and internationally. Also presents incidence figures from the echocardiography database, Department of Pediatrics, Faculty of Medicine, Ramathibodi Hospital, Mahidol University.

**Structure of the aortic valve**

The structure of the aortic valve consists of three semilunar valves that act like a door to prevent blood from flowing back during relaxation. The aortic valve rim also has a bulging shape that acts as a reservoir for storing blood before it is sent to the heart muscle via the coronary artery, including The left coronary artery arises from the left cusp of the aortic valve to give off branches to the anterior (left anterior descending; LAD) and lateral (left circumflex artery; LCX) sides. Right coronary artery; RCA exits from the right cusp of aortic valve and is divided into Sinoatrial nodal artery; SNA, Conus artery; CA, Right acute marginal artery, Posterior descending artery; PDA and Posterior interventricular artery; PIA supplying the posterior artery may originate from either RCA or LCA. If it comes from LCA, it is called Left dominance, which is found in about 29-56.8% of people in general, with 90% having the length of left main segment coronary artery; LMCA is the beginning of LCA before giving LAD branches, approximately 5 mm in size. [3] (Figure 1C)

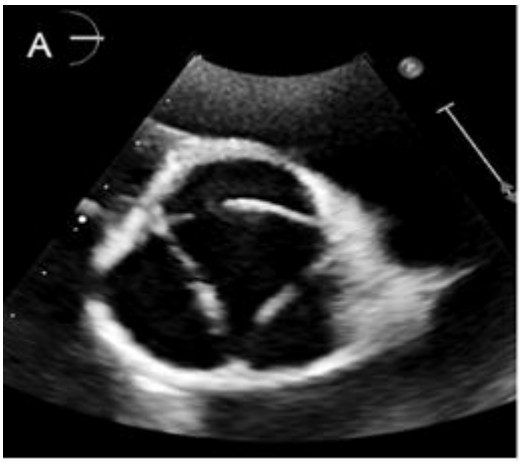



**Figure 1A** Echocardiogram in parasternal short axis echocardiogram at aortic valve level shows the number and characteristics of the aortic valve leaflets during opening in a normal person

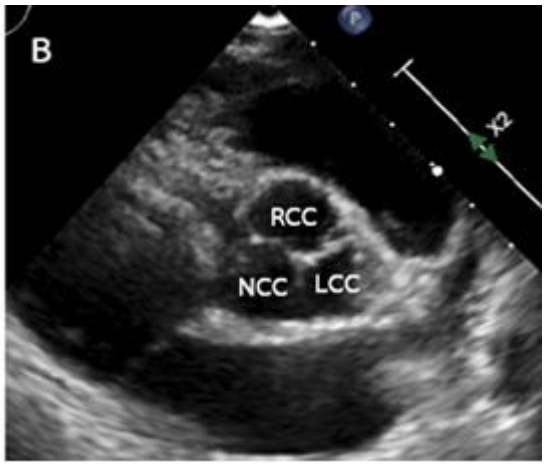



**Figure 1B** shows the number and characteristics of the aortic valve leaflets during closing. (Right coronary cusp; RCC, Left; LCC, None; NCC)

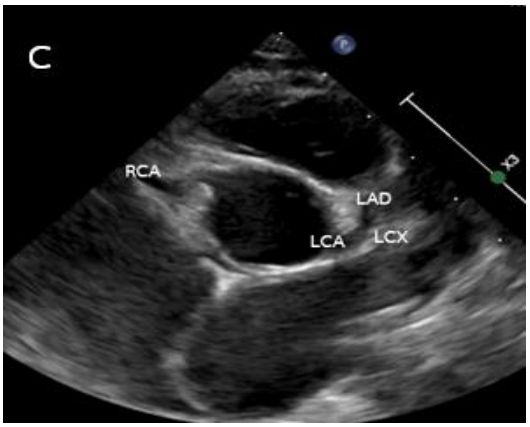



**Figure 1C** shows the number of coronary artery characteristics from the aortic valve in both Right cusp and Left cusp

In this condition where the heart valves are connected to each other with only two leaflets remaining, it usually occurs at the commissure. Originally, there were three leaflets and three commissures that functioned to open and close tightly, but if any of the leaflets are connected, only two or one leaflet remains that can open and close. The remaining raphe/pseudo commissure may be seen as a non-separate raphe. In most cases, the shape and size of the heart valve may be abnormal. In addition to fusion of the septum, it is also caused by loss of function or atrophy of some valve leaflets. When fused, the atrophied valve is pulled in the same direction as the larger valve. The echocardiography image shows a single large flap valve. Diagnosis requires motion pictures to observe the line of the suture. Many studies have

stated that the fusion of the two remaining aortic valve leaflets is also associated with aortic stenosis (Coarctation of aorta) in 20-85%.[1] It is also associated with interrupted aortic arch up to 27%, with several studies indicating a relationship with Shone complex, which is a narrowing of the left side of the heart at various levels.

**Incidence and classification**

In the past, Brandenberg classified aortic valve fusion into 3 types based on the location of the rape, similar to Sievers and Schmidtke who studied the classification of bicuspid aortic valve based on the difference in the number of rapes, until the "Sievers and Schmidtke classification" was created.[3-4] The fusion of the aortic valve leaflets to only two leaves is mostly a congenital anomaly, associated with family incidence and Turner syndrome. Marfan syndrome is also occasionally found, but its association has not been clearly reported. This condition is more common in males than females, especially in those over 15 years of age, accounting for 0.5-1.4 % of general patients.[1-3] And from a random survey of Ramathibodi Hospital in 2021-2024, it was found that in a 3year period, out of 4,113 examinations. Approximately 1.43% (59 cases) of examinations related to bicuspid aortic valve were found. (Table 1 and Figure 2)

**Table 1** Shows the number and type of bicuspid aortic valve fusions. Department of Pediatrics, Faculty of Medicine, Ramathibodi Hospital, 2021-2024

| Fusion | Amount | Percent | Gender | | Age |
|---|---|---|---|---|---|
| | | | Male | Female | |
| L+R | 26 | 44% | 14 | 12 | 14 days – 27 years |
| R+N | 21 | 36% | 8 | 13 | 2 months – 20 years |
| L+N | 12 | 20% | 3 | 9 | 2 months – 17 years |
| SUM | 59 | 100% | 25 | 34 | 14 days – 27 years |

**Note :** L+N; Left coronary cusp and Non coronary cusp fusion , R+N; Right coronary cusp and Non coronary cusp fusion, L+R; Left coronary cusp and Right coronary cusp fusion

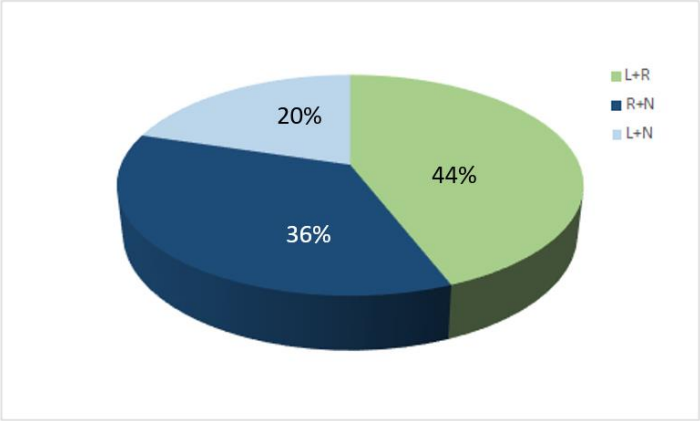



Figure 2. Chart showing the number of pediatric patients and types of septal defects at Ramathibodi Hospital.

**Effects or complications**

From the survey of pediatric patients at Ramathibodi Hospital, as shown in Figure 6, the graph showing complications of bicuspid aortic valve, aortic stenosis was found to be the most common at 42%, with aortic regurgitation at only 8.5%. However, combination of aortic stenosis and regurgitation was found in 27% and aortic root dilate was found in only 5.2%. Similarly, the study by Nuran Yener et al. Found that aortic valve stenosis was most commonly associated with bicuspid aortic valve, followed by aortic valve regurgitation, aortic dissection, and infective endocarditis, respectively.[3] Bicuspid aortic valve bulging can also occur in patients with aortic valve disease. Some patients are unaware that they have a disease, but it is usually detected when the valve becomes infected (infective endocarditis), has calcification, loses function (dysfunction), or has side effects on the aortic valve. Although there are currently standard treatment methods, there are still limitations. For example, if surgery or artificial heart valves are performed at a young age, there is a high chance that repeat surgery will be required because artificial heart valves have a limited lifespan, It does not expand in size, and may require long-term use of anticoagulants or immune suppressants. Recognizing the abnormality and treating it early, and having regular follow-up visits with cardiologist will help manage in

symptoms and maintain native heart valve for a long period of time. [7,9]

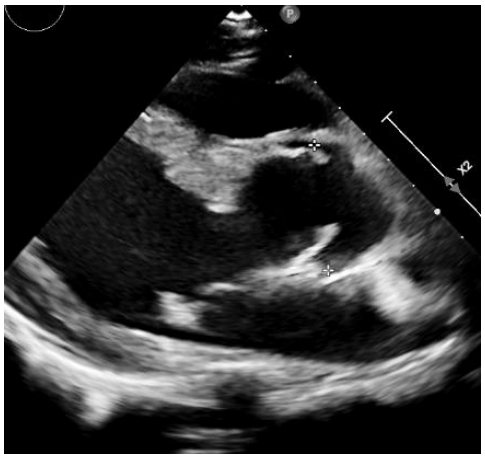


**Figure 3.** Echocardiogram in parasternal long axis view in 2D shows aortic root dilated with limitation of aortic valve opening or doming form. Aortic stenosis also results in left ventricle hypertrophy, and regurgitation, leading to excessive backflow into the left ventricle, resulting in left ventricular dilation.

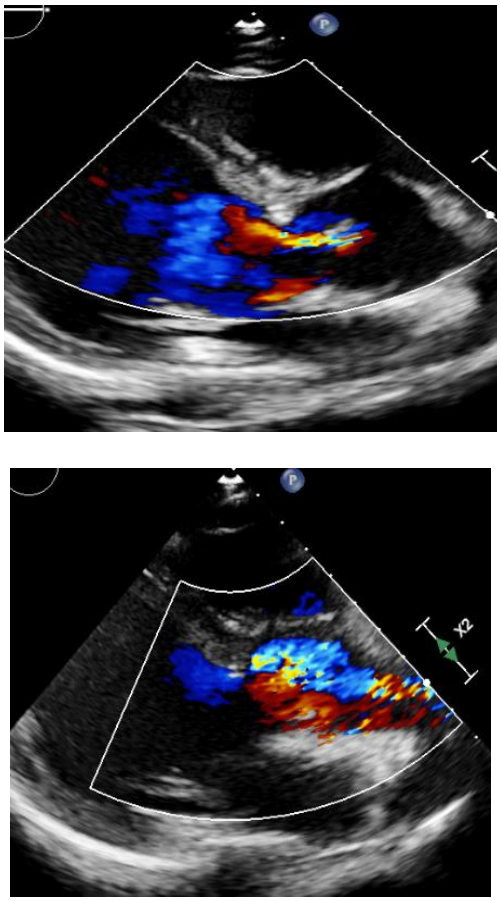


**Figure 4-5.** Echocardiogram in parasternal long axis view in with color mode shows aortic regurgitation and stenosis was combine complication of bicuspid aortic valve.

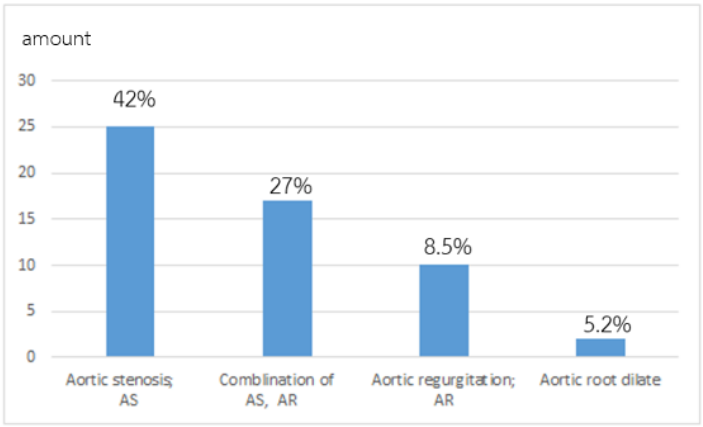



**Figure 6.** Diagram showing type and amount of side effects associated with bicuspid aortic valve. Department of Pediatrics, Faculty of Medicine, Ramathibodi Hospital, 2021-2024

**Genetic causes and factors**

Several studies have reported that the incidence of bicuspid aortic valve is related to genetic abnormalities up to 14.6%, as in the study by Emanuel et al. who found a male to female ratio of approximately 2:1 in the general population.[1] Associated conditions include aortic root dilation, aortic aneurysm, and aortic dissection.[5] It comes from the first hypothesis about the enlargement of the aorta, such as in patients with Marfan syndrome, there will be a change in the structure and function of the Protein Fibrillin-1 affecting the location of TGF-beta, causing apoptosis of smooth muscle cells in the blood vessel wall until they lose their condition and bulge into an aneurysm.[2] Hypothesis 2: Animal experiments have found that the bicuspid aortic valve is associated with abnormalities of endothelial nitric oxide synthase at the NKX 2.5 and NOTCH loci, especially the NOTCH-1 locus, which has been previously studied. Hypothesis 3: Thoracic aortic aneurysm and thoracic aortic dissection are associated with the presence of ACTA-2 gene mutations.[1]

**Diagnosis**

Based on physical examination, medical history, and heart auscultation. But the tool that can definitely confirm the results, is radiation-free, easy to move, and inexpensive is the trans-thoracic echocardiography (TTE). However, in adult patients and those with unclear or poor acoustic window, For more definitive confirmation, a modality of trans-esophageal echocardiography (TEE) may be necessary, which

provides a clearer image only in cases with indications.[3] Echocardiography uses two-dimensional and three-dimensional diagnostic methods, especially the cross-section of the heart valve leaflets, where three-dimensional examination can clearly show the characteristics of the disease. The angle or view that helps to diagnose bicuspid aortic valve and is popular according to international recommendations is parasternal short axis window at aortic valve level as the main method for distinguishing the type of fusion. (Figure 4-6) The next is the parasternal long axis to assess the severity of stenosis and leakage, including the lateral closure and opening conditions. However, it still requires other perspectives according to the general examination standards to assess the overall function of the heart. There are 3 types of fusion consist of 1.1) Left coronary cusp type; LCC fused to Right coronary cusp; RCC (L+R) 1.2) Right coronary cups type; RCC fused to Non coronary cusp; NCC (R+N) 1.3) Left coronary cups type; LCC fused to Non coronary cusp; NCC (L+N)

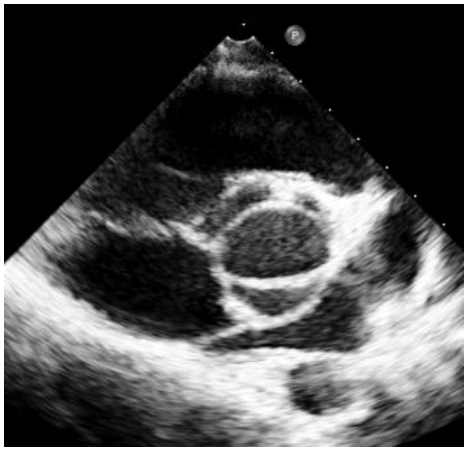


Figure 7. Echocardiogram shows raphe scar with Left coronary cusp and right coronary cusp fused. (L+R)

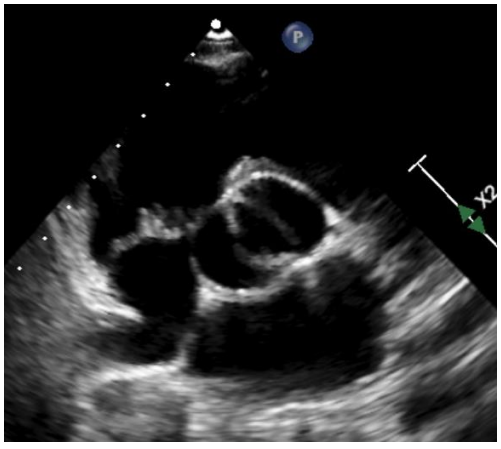
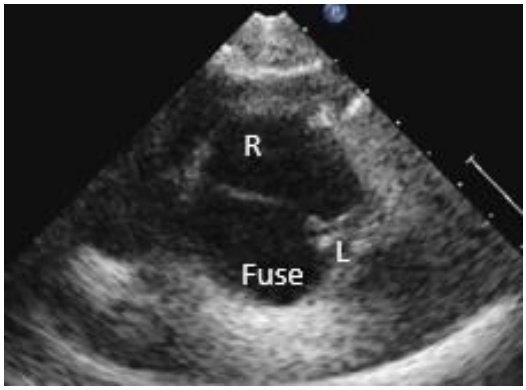


Figure 8. Echocardiogram shows the scars are falling down or rape was the signature of the right coronary cusp and non coronary cusp fused (R+N)



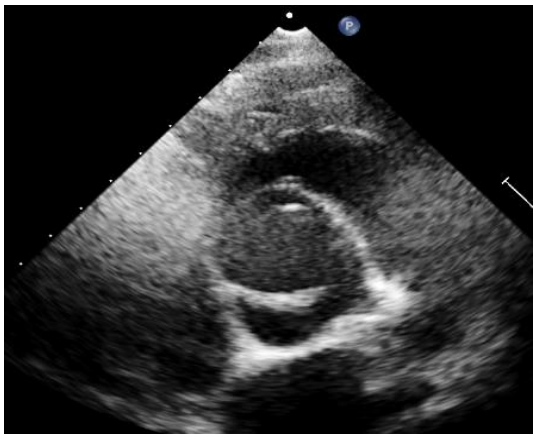



Figure 9-10. Echocardiogram shows the atrophy of left coronary cusp and fused with non coronary cusp.(L+N) The left main coronary artery took off left coronary cusp (L) was the landmark, and right cusp (R) was normal.

Other forms of fusion have also been reported, such as the unicuspid aortic valve,[6] where all the plates are fused together leaving only a single separation in the center. Sometimes, aortic valve atresia occurs to the point where all the valves are connected, leaving no opening for a guide wire to pass through. This condition is called aortic valve atresia. This often occurs in conjunction with a very small left ventricular outflow tract and a very small aorta.

**Treatments**

A cardiologist will consider the severity of the symptoms and the advantages, disadvantages, and risks of each method. Treatment approaches range from medication to reduce symptoms, cardiac catheterization to use balloons to expand or reduce stenosis, and surgery, depending on the

characteristics of the lesion and symptoms of each patient, including: 1) Medical treatment to reduce symptoms (medical control), such as reducing blood pressure and reducing fluid volume when necessary.[7,11] Antibiotics are given to prevent the spread of infection to the aortic valve leaflet to prevent further valve deterioration.[7] 2) Trans-catheterization treatment includes balloon aortic valvuloplasty and surgical methods, which are divided into valve repair and valve replacement. The current popular treatment is the use of balloons to dilate the sutures, which is intended to widen the blood outlet. This is used to correct the moderate to severe aortic stenosis. Patients who will receive this treatment must have only a mild level of regurgitation because after the stenosis is expanded with a balloon, the expanded valve will leak more. Percutaneous aortic valve implantation involves inserting a new valve into the original valve. In Thailand, this method is still used in small group because it requires a highly experienced team of personnel and the equipment and devices are very expensive.[3,10,13] Cardiologist in many countries often choose this method to avoid various limitations such as surgical scars in the chest area, lifelong immunosuppressive medication, etc. 3) Surgery is considered a standard method that is recommended internationally, and currently the risk of heart valve surgery is only about 3-5%.[8] Consist of 3.1) Aortic valvulotomy is a procedure to repair the heart valve to a near-normal condition. This method is often used in patients whose heart valves are not severely damaged. It may involve suturing or cutting certain parts of the heart valve to allow it to open and close with better quality. The advantage of this surgical method is that after the procedure, there is no need to take immunosuppressive drugs. 3.2) Aortic valve replacement surgery is the replacement of a damaged heart valve with a new one. These heart valves come in many sizes and types. They are classified by material consist of tissue heart valve. For example, heart valves obtained from bovine organs, pig organs, human pericardial parts (Pericardial) and human heart valves (Homograft), etc. Heart valves made from synthetic materials (Mechanical prosthetic heart valve), such as heart valves made from metal, stainless steel, etc.[8,9] If classified by characteristics, they include types with cusps (with cusp) such as bileaflet and types without cusps (Non cusp) such as Ball cage and Tilting disc, etc. 3.3) Aortic root repair surgery (Aortic root remodeling) 3.4) Aortic root replacement surgery (Aortic root replacement) which requires decoration or re-implantation of coronary artery as well.[3]

**Indications for aortic stenosis repair**

In cases with obvious symptoms and echocardiography results showing a valve area of less than 1 square centimeter and a mean pressure gradient > 50 mmHg., according to the recommendations of the American Heart Association Guideline; AHA guideline and the American Society of Echocardiography; ASE, a condition where the Mean Pressure Gradient; Mean PG > 40-50 mmHg. or a velocity exceeding 4 m/s is considered a severe stenosis [11,12] Another group, if the systolic blood pressure is low and there are abnormal symptoms during exertion, along with LV hypertrophy by wall thickness >15 mm. in adolescent and adult, aortic valve area (AVA <0.6 $cm^2$) [8]

**Indications for aortic regurgitation repair**

Must have symptoms along with echocardio-graphic results indicating severe leakage and have symptoms along with New York Heart Association Classification; HYHA in Class 3 with left ventricular end-diastolic size (LVESD) greater than 5.5 cm. And the left ventricular ejection fraction (LVEF) is less than 55%, including in patients with an infected aortic valve that has lost its function.[8,14]

**Monitoring of patients after heart valve surgery**

Mostly to monitor and watch for the following problems: Complications from taking anticoagulants, [6] Heart valve infection (Infective endocarditis; IE) Valve thrombosis and embolization[7,8] Calcification of the newly inserted heart valve, aortic enlargement, and monitoring for dissection of the aorta. In addition, patients with bicuspid aortic valve should have regular oral and dental checkups before and after treatment to prevent infective endocarditis. [9,11-14]

**Summary**

The prevalence of bicuspid aortic valve at Ramathibodi hospital in Thailand was 1.43% of total examination, The left and right coronary cusp of aortic fused was a common type about 44%. This data was similar and nearly another study in western. The echocardiogram remains the gold

standard technique to rule out bicuspid aortic valve and complication.[15-16] Most bicuspid aortic valve disorders are congenital and genetic, and when classified according to the nature of the commissure, cardio-thoracic technologists and echocardiographers have a high chance of being found left coronary cusp and right coronary cusp fusion (44%). Followed by Right coronary cusp and non coronary cusp fusion and Left coronary cusp and non coronary cusp fusion types, respectively. In terms of complications from concomitant abnormalities, aortic stenosis is most commonly found at 42%. Aortic regurgitation is less commonly found. In addition, abnormalities of both stenosis and regurgitation (Combination of aortic stenosis and regurgitation) can also be found in approximately 28.8%. Treatment can be done by both surgery and catheterization depending on the pathological characteristics and limitations, along with consideration of medication to control symptoms and severity.

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