Atrioventricular Canal Anatomy and Differences from Normal Hearts, a Systemic Review of Literatures

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Abstract: Objectives: This study was designed for Sound understanding of the pathological anatomy of artioventricular septal defects (AVSD), emphasizing its surgical importance, connecting it to the surgical techniques have developed for proper surgical repair. **Methods:** In a systematic review of the literatures had described the surgical anatomy, techniques of the surgical repair, early or late postoperative complications, and long term outcomes of AVC surgeries, there are 110 8articles published in the time period between 1960 and 2013 directly discussing these varieties. The total of 10 separated important structural differences between AVC anatomy and normal hearts were reported and demonstrated their direct or indirect effects on the surgical techniques and outcomes. **Results:** Along with variations in the degrees of incomplete development of the three basic elements characterizing AVSD, there are other seven important anomalies involving the level of the attachments of the AV valves to the crest of the inter-ventricular septum, abnormal dimension of the inlet and outlet septum to ventricular apex, different contribution of attachments of valve leaflets to common A-V valve annular circumference, variable position of the common A-V valve in relation to the plane of the ventricular septum, different position and pathway of the conductive tissue, and unwedged position of the aortic valve away from AV valves with absent or hypoplastic aortomitral Curtin. **Conclusion:** Pathological anatomy of AVSDs are not only the septal and AV valve malformations to be considered during surgery, they include other many associated structural anomalies that affect the techniques and results of surgical repair.

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1. Introduction

Embryologically, Atrio-ventricular septal defects (AVSD) caused by incomplete embryonic development of superior and inferior endo-cardial cushion tissue. This group of defects subdivided anatomically according to the three basic defects into AV valves and cardiac septae to partial, intermediate and complete forms. In partial AVC, there are ostium primum ASD, common AV fibrous annulus with two separate AV valve orifices, and no VSD. In intermediate form (transient AVC), three are ostium primum ASD, common AV fibrous annulus with two separate AV valve orifices, and restrictive VSD. Whereas in complete AVC, there are ostium primum ASD, common AV fibrous annulus with one common AV valve orifices, and unrestrictive VSD **(McElhinney 1998).**

Objectives

This study was designed for Sound understanding of the pathological anatomy of artioventricular septal defects (AVSD), emphasizing its surgical importance, and connecting it to the surgical techniques have developed for proper surgical repair.

2. Methods

In a systematic review of the literatures published in the last five decades that described the AVC malformations and outcomes of surgical technique, there are about 1108 literatures posted in the time period between 1960 and 2010. Most of these papers described the AVSDs as a spectrum of cardiac malformations characterized apparently by varying degrees of incomplete development of inferior portion of the atrial septum, inflow portion of ventricular septum, and atrioventricular (AV) valves. However, by meticulous collection and analyzing these literatures, we reported the main anatomical features of AVC and the impact of them on timing, techniques, and outcome of surgery. The best surgical options and techniques that suggested by surgeons to deal with these anatomical features to ensure better outcome is highlighted. There are ten differences between the AVC anatomy and normal hearts, all of them directly affect the surgical strategy for repair **(table 1).** Moreover, some of them are more important in identifying these anomalies than the septal and valvular defects. These features considered cardinal features of AVSDs.

3. Results

Almost all pediatric cardiac surgeons describe the **a**trioventicular canal (AVC) as a complex cardiac malformation characterized by varying degrees of incomplete development of inferior portion of the atrial septum, inflow portion of ventricular septum, and atrioventricular (AV) valves. Although this is undoubtedly true, however, these are not the cardinal anatomical features that differentiate the AVC from normal hearts. All types of AVC share intracardiac abnormalities characteristic to it and make it quite different from normal hearts. These anomalies are involving the level of the attachments of the AV valves to the crest of the inter-ventricular septum, abnormal dimension of the inlet and outlet septum to ventricular apex, different contribution of attachments of valve leaflets to common A-V valve annular circumference, variable position of the common A-V valve in relation to the plane of the ventricular septum, different position and pathway of the conductive tissue, and unwaged position of the aortic valve away from AV valves with absent or hypoplastic aortomitral Curtin **(table 1**).

Table 1, ten anatomical differances between AVSDs and normal hearts

AV canal defects	Normal heart
• Ostium primum ASD • Variable degree of underdevelopment of the inlet portion of the inter-ventricular septum • Common AV ring with variable degree of separation into two AV orifices	Intact interatrial septum Well-development inlet portion of the inter-ventricular septum Competent separate two AV valves with separate rings
• Apical displacement of the attachments of the AV valves to the crest of the inter-ventricular septum. The mitral and tricuspid valves are displaced into the ventricles	Tricuspid valve in more apical position than the mitral valve
• The left lateral leaflet occupies only 25% of the left AV valve annular circumference (figure 1)	The corresponding posterior leaflet of the normal mitral valve occupies 65%of the left AV valve annular circumference
• The left superior and left inferior leaflets together occupy 75% of the left AV valve annular circumference (figure 1)	The corresponding anterior leaflet of the normal mitral valve occupies 35% of the left AV valve annular circumference
• Variable positions of the common A-V valve in relation to the plane of the ventricular septum.	Central position of each separate A-V valve
• Shortened dimension of the inlet septum-to-ventricular apex ("scooped-out" appearance) and lengthened dimension of the outlet septum-to-ventricular apex ("goose-neck" appearance). (figure 3,4)	Inlet septum-to-ventricular apex length and the outlet septum-to-ventricular apex length are equal
• Inferior displacement of conductive tissue. The AV node located at the base of the triangle of Koch in the atrial wall just above the crux cordis (It is in the area between the coronary sinus and the junction of the left and right inferior leaflets). The bundle of His is displaced inferiorly, coursing at the inferior rim of the scooped-out basal portion of the inter-ventricular septum. It extends on the crest of the septum and divides into bundle branches before reaching the septal commissure.	AV node located at the tip of the triangle of Koch Bundle of His is located superiorly piercing the membranous septum
• Superior (rightward) and anterior deviation of the aortic valve away from AV valves ("un-wedged position") (figure 5).	Wedged position of the aortic valve between the AV valves

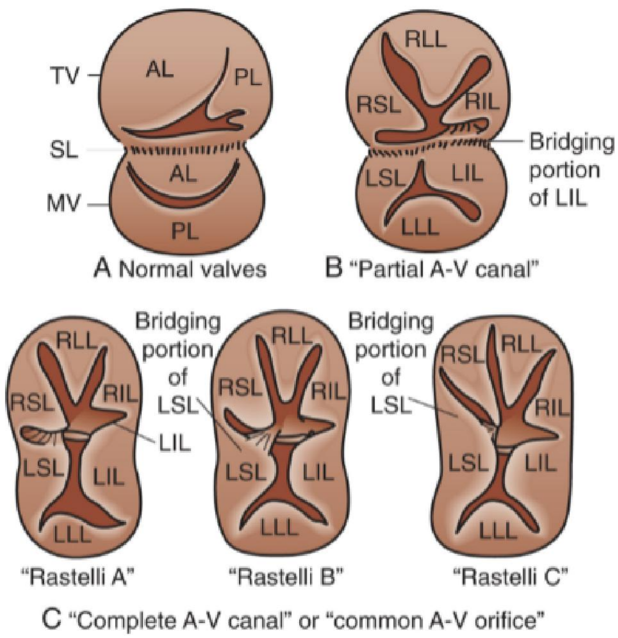

Figure 1: The difference in valve morphology in a normal, partial canal, and complete canal defect, including Rastelli classification type A, B, or C. AL, anterior leaflet; A-V, atrioventricular; MV, mitral valve; PL, posterior leaflet; RIL, right inferior leaflet; RLL, right lateral leaflet; RSL, right superior leaflet; TV, tricuspid valve. (Kirklin et al 1985)

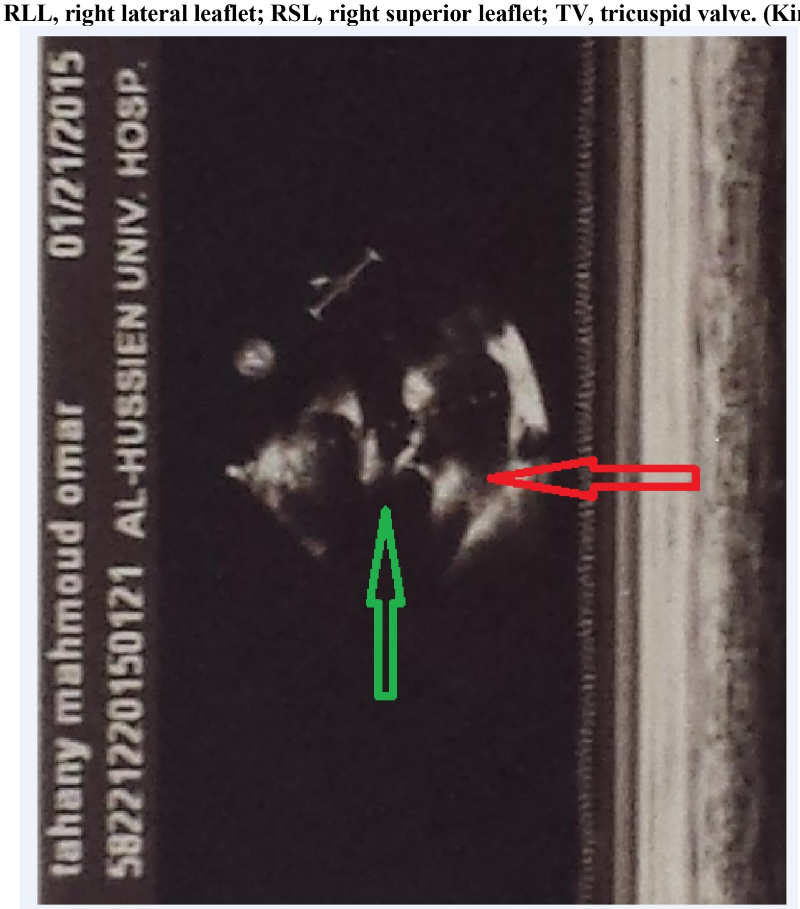

Figure 2: long axis parastaernal view shows prosthetic mitral valve (red transverse arrow) causing significant subaortic obstruction with mean gradient 66mmHg (green vertical arrow) in female patient, 27 years, with AVC anatomy.

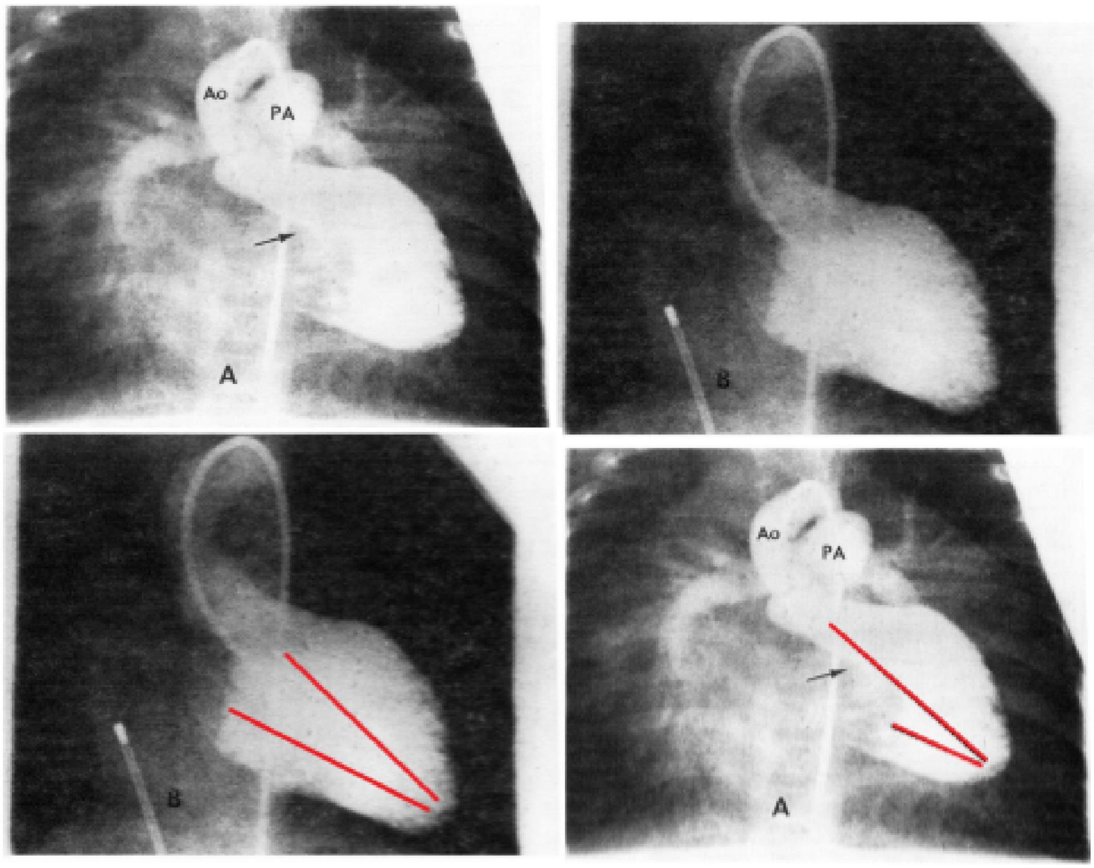

Figure 3: Left ventricular angiograms obtained from a case of AVC before correction, the A-P view in end-diastole shows the typical gooseneck deformity arrow (A). Left ventricular angiograms obtained from a case of normal heart reveals normal left ventricular outflow tract (B). Ao aorla; PA = pulmnoniary artery (Culpepper et al 1978). The lower pictures are the same after adding a red line by us to demonstrate the difference.

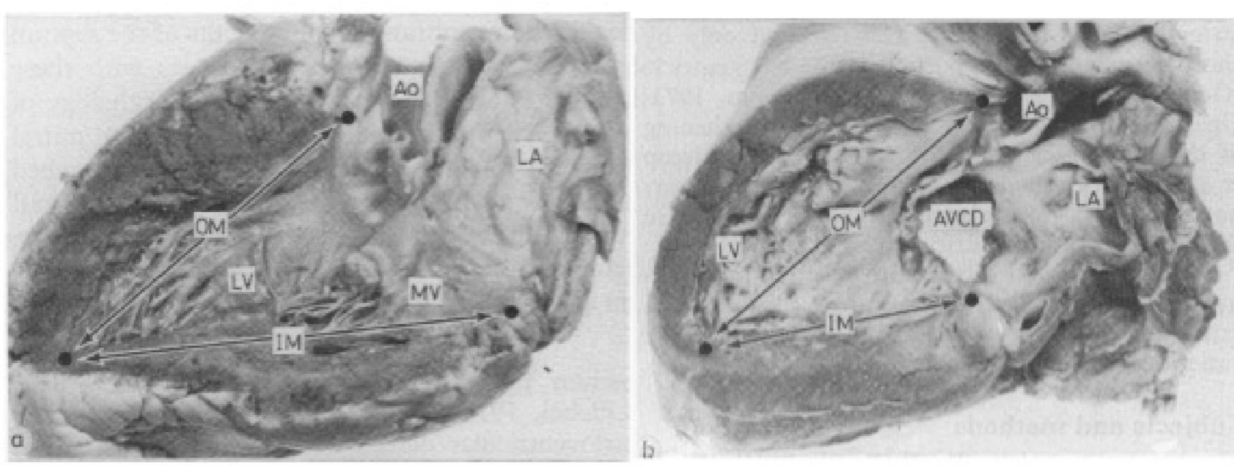

Figure 4: Photographs of longitudinal sections of the left ventricle in a normal heart (Fig. la) and the left ventricle in an atrioventricular defect (Fig. lb). The photographs show the inlet (IM) and outlet (OM) measurements taken. Note that in the atrioventricular defect there is disproportion between these measurements. LA, left atrium; LV, left ventricle; MV, mitral valve; Ao, aorta; AVCD, atrioventricular canal defect (Piccoli et al 1979).

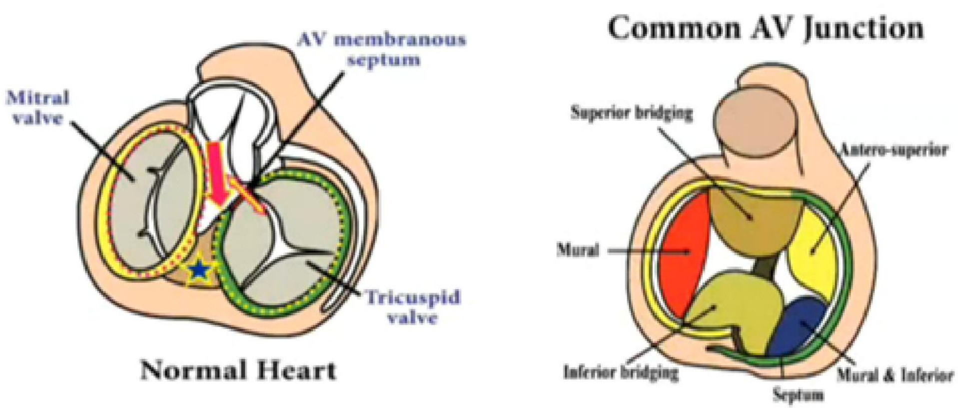

Figure 5: position of the aortic valve; Wedged position of the aortic valve between the AV valves in normal hearts. Superior (rightward) and anterior deviation of the aortic valve away from AV valves ("un-wedged position") in AVCs

4. Discussion

AVSDs are uncommonly accompanied by an intact atrial septum, either in partial or complete form. Such hearts have all of the typical features of a complete A-V septal defect with a common AV valve annulus, but the septum primum is intact, with the AV valve leaflets either attached to the underside of the atrial septum or abutting it during ventricular systole. In either case, significant interatrial shunting is prevented. This anatomy may be confusing to the surgeons when see it through a right atrial incision, and is clarified by opening the atrial septum to inspect and repair the left-ward components of the AV valve. **Piccoli GP et al** in their series of AVC malformations, observed a ventricular septal defect in association with a cleft mitral valve but with an intact atrial septum in four cases out of 104, these cases were having all the features of an atrioventricular defect. They describe them as 'atrioventricular canal ventricular septal defect'. They recommended, therefore, that term, if used at all, should be reserved for description of such cases which have the cardinal features of AVSDs. **Goor DA et al c**ategorised these cases as complete atrioventricular defects without an ostium primum component. While **Neufeld HN et al** said, these cases must be distinguished from some perimembranous defects excavating the inlet portion of the inlet septum and do not possess the stigmata of the atrioventricular defect itself.

Absence of inlet VSD is a hollomark for partial form, however, associated other types of VSDs, for example, perimembrous, outlet, subpulmonic, or muscular, don't make it complete form and must be treated as partial AVCs. **Calabro R** described a fibrous tissue connection between the left superior and inferior leaflets over the ventricular septum in intermediate AVC, This connection somewhat obliterates the bare area of ventricular septum, which is always found in the complete form but is not sufficient for the mitral and tricuspid portions of the valve to be considered separate entities. Tiny defects in the attachment of the mitral valve to the crest of the ventricular septum may result in small interventricular communications. **Pacifico AD** reported that, intermediate form is sometimes amenable to the surgical repair used for the partial AVSDs, but more often must be converted at operation to the complete form and repaired as such.

To achieve a competent AV valves, it is important to consider all of its elements and not assume that simple suture closure of the septal commissure will result in a competent valve. While most surgeons agree that the cleft is a "normal" part of the valve architecture in AVC defects and that reconstruction does not ever re-create anterior leaflet of a normally formed mitral valve, closure of the cleft is performed routinely by most surgeons. In Partial AV-septal defect, the mitral valve was described as a trileaflet structure by **Rastelli G et al**, whose anatomy and mode of function are different from those of the normal mitral valve **(figure 1).** In Complete AVC, the key element is the presence of a common (single) A-V valve orifice that is common to the right and left A-V chambers. In both anomalies, the lateral leaflet occupies only 25% of the annular circumference, and thus it is distinctly different from the corresponding posterior leaflet of the normal mitral valve, which occupies 65%. Similarly, the left superior and left inferior leaflets together occupy 75% of the annular circumference, and thus they are distinctly different from the anterior leaflet of the normal mitral valve, which occupies 35% **(figure 1).** This difference in the AV valve annular circumference requires special consideration for proper valve repair. Consequently, **Suzuki K et al observed that** closure of the clefts may result in significant stenosis of the valve orifice or even worsen the regurgitation. The technique used for repair of CAVC also might contribute to occurrence of valve incompetence, as Rastelli single-patch technique with dividing the common leaflets consume a lot of the valve tissues, and the modified "Australian" technique of one patch, transforming the complete AVSD into a partial AVSD, may also lead to mitral insufficiency due to changing the chordae geometry and potentiate posterior leaflet prolapse. Moreover, Australian" technique may lead to subaortic stenosis as a postoperative gradient being reported postoperatively by **Metras D et al.** Whereas, **Puga FJ** reported an inherent problem of keeping the competence of the valve even after complete repair, **McElhinney DB et al and Najm HK et al** reported annular dilatation and degeneration of the leaflets play a major role. **Yasui H et al** noticed that degeneration of the atrioventricular valve increases with age. **Michielon G et al** also described annular dilatation as an important factor for regurgitation. **Peter BM** reported Combinations of the Annuloplasty, commissuroplasty, or Augmentation of dysplastic superior and inferior bridging leaflets using glutaraldehyde-fixed autologous pericardium. The dysplastic superior and inferior bridging leaflets are detached radially along the anterior annulus. They are then augmented allowing for better leaflet coaptation. In rare instances double-orifice valve may be needed, however, a few cases may require valve replacement which could be difficult due to the annulus projecting towards the left ventricle outflow tract and the possibility of causing LVOTO by the mechanical valve **(figure2)**. This is because the inlet septum is shortened and the outlet septum is lengthened. A crescent-shaped patch may be used to lengthen the mitral-to-aortic continuity along the circumference of the annulus in the section of A1 and P1 due to hypoplastic aorto-mitral curtin. In this way, a low-profile mechanical valve can be positioned away from the left ventricle outflow, with a projection into the left atrium. Leaflet remnants are used to anchor the valve along the other segments of the annulus.

Apical displacement of the attachments of the AV valves to the crest of the inter-ventricular septum together with Shortened dimension of the inlet septum-to-ventricular apex ("scooped-out" appearance) and Lengthened dimension of the outlet septum-to-ventricular apex ("goose-neck" appearance) **(figure 3)** i.e. distance from mitral valve annulus to apex is shorter than the outlet distance apex to aortic valve annulus. In the normal heart, these distances are nearly equal **(figure 4)**. These features lead to the characteristic "gooseneck" deformity seen on anteroposterior angiography. Although this leads to a left ventricular outflow tract (LVOT) diameter that is smaller than normal, it does not usually cause clinically significant obstruction by itself, but, contribute to an LVOT obstruction when associated with a subaortic membrane, accessory atrioventricular valve tissue, and large patch inserted to close the VSD. In addition, the LVOT left ventricular outflow tract including the aortic root are displaced anteriorly away from the normal wedged position between both atrioventricular valves. **Piccoli et al** defined AVC malformation by disproportion between the ventricular inlet and outlet dimensions and a malorientation of the aortic valve relative to the atrioventricular valve or valves (figure 2C). Associated with this there is a characteristic 'scooped-out' appearance of the muscular ventricular septum, gross abnormalities of the membranous components of the septum as compared with the normal heart, and narrowing of the aortic outflow tract. Autopsy and echocardiographic studies done by **Peter BM** have shown that this anomaly leads to LVOT obstruction (LVOTO) in as many as three quarters of AVC patients. The incidence of LVOTO may be more pronounced in partial AVC than in complete forms because of the firm fixation of the valve leaflets to the scooped-out. In addition, abnormal positioning of papillary muscles or accessory attachment of cordae to the septum, in some cases, may contribute to this potential. The risk for reintervention for LVOT stenosis does not appear to decline even many years following initial operation, and even more concerning is that the need for second reoperation for LVOT stenosis has been observed by **Gurbuz AT et al** to be 35% to 45%. **Van Arsdell GS et al** described resection of fibrous tissue that may represent accessory endocardial cushion tissue, anomalous cordal attachments, or reaction as component of the first repair. In the reoperation, septal myomectomy or modified Konno technique to create a VSD with patch closure from the right side similar to what has been successfully reported in other forms of tunnel-like subaortic stenosis. While **Van Son JAM et al** recommend detaching all bridging leaflet components of both AV valves from the ventricular septal crest, essentially re-creating an inlet VSD analogous to a Rastelli type C complete AVC defect, then patching the VSD with reattachment of the valves to the top of the patch. The anterior aspect of this patch directly augments the dimension of the septum in the subaortic region. The rationale for this stems from the observation that LVOT stenosis is rare after repair of Rastelli type C complete AVC compared with type A, transitional, or partial AVC; all of which share the distinction of having attachments of valve to the superior septal crest and may be repaired primarily without significant augmentation of the superior portion of the ventricular septum. Other technique involves mobilization of the left-sided valve components only, either the superior portion, or the entire reconstructed septal leaflet were described by **Ross in 1991**. Pericardial patch augmentation of the base of the leaflet similarly moves the left AV valve farther into the left atrium during ventricular systole, thus augmenting the dimension of the LVOT. Such leaflet augmentation may also be beneficial in addressing residual left AV valve insufficiency. In addition, the un-wedged position of the aortic valve away from AV valves make the known posterior root dilatation procedure to insert prosthetic valves are difficult because of the changed relations and higher possibilities to go outside the heart **(figure 5)**.

Inferior displacement of main elements of the conductive tissue are present in all cases of AVC. The AV node located at the base of the triangle of Koch in the atrial wall just above the crux cordis (It is in the area between the coronary sinus and the junction of the left and right inferior leaflets). The bundle of His is displaced inferiorly, coursing at the inferior rim of the scooped-out basal portion of the inter-ventricular septum. It extends on the crest of the septum and divides into bundle branches before reaching the septal commissure. This anomaly leads to postoperative surgical heart block (HB), however, better recent techniques have made as lower as less than 1% of patients with a range of 0.1–3.5%, as reported by **Julien IE**. The risk of complete block is much higher if the valve has to be replaced. **Pacifico AD** illustrated that, during suturing in the posteroinferior aspect of the ventricular septum crest, the objective is to bring the suture line completely away from the area of the conduction tissue.

The surgeon must ensure that repair will leave adequate orifices to each ventricle because of the variable position of the common AV valve in relation to the plane of the ventricular septum in cases of CAVC. When this adequacy is in doubt, it is helpful to compare the diameters of these orifices, measured with Hegar dilators, with the normal mitral and tricuspid dimensions. In the right dominant form of the complete A-V septal defect, the right ventricle is large, the left ventricle is smaller than normal and the position of the common A-V valve in relation to the septum is right-ward. Whereas in the left dominant form, the left ventricle is larger than normal, the right ventricle is smaller than normal, and the position of the valve is left-ward.

Conclusion

Pathological anatomy of AVSDs is not only the septal and AV valve malformations to be considered during surgery, but also they include other many associated structural anomalies that affect the techniques and results of surgical repair. The hallmarks of AVCs are scooped-out ventricular septum, goose-neck appearance of LVOT, unwaged aortic valve and different attachment of valve leaflets to annular circumference.

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