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## Surgical correction of atrioventricular septal defect

Hooohenkerk, G.J.F.

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**Author:** Hoohenkerk, Gerard Joannes Franciscus

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## **Introduction**

## General introduction

Faulty development of the endocardial cushions, which represent the primordia of the atrioventricular septum and atrioventricular valves, plays a central role in the development of atrioventricular septal defects (AVSD) (1). The superior and inferior endocardial cushions appear at 4-5 weeks' gestation. During this time, the common atrioventricular canal is positioned over the primitive left ventricle.

Mesenchymal cells invade these masses of tissue, and, during the fifth week of gestation, the cushions approach each other and fuse. This divides the common atrioventricular canal into right and left canals. The right and left lateral endocardial cushions develop shortly after the appearance of the superior and inferior cushions, followed by the dextrodorsal conus cushion. These structures are involved in the development of the mitral and tricuspid valves and their support apparatus.

The endocardial cushions do not directly form the valve components but play an essential role in the process by which undermining and delamination of the myocardium forms the valve leaflets and chordal attachments (1). Complete failure of fusion of the endocardial cushions results in deficiency of the inlet portion of the interventricular septum, a common atrioventricular valve annulus and common AV valve, as well as deficiency of the inferior (primum) portion of the atrial septum. This constellation of features results in a large defect in communication with all 4 chambers of the heart, and is known as complete AVSD.

AVSD has also been called endocardial cushion defects and involve a spectrum of anomalies which are unified by varying degrees of incomplete development of the septal tissue surrounding the atrioventricular valves along with varying degrees of abnormalities of the atrioventricular valves themselves. The term suggest that the atrioventricular septum is missing or abnormal. However, the atrioventricular septum is not an independent septal component and is formed by the atrial and ventricular muscular septum and fibrous annulus. The AVSD therefore arises as a result of impaired embryological development of both the muscular and the fibrous part of the atrioventricular septum. The spectrum of AVSD can be subcategorized according to whether they have a common or separate right and left atrioventricular orifice. They can be further subdivided according to the integrity of the atrial and ventricular septum and, in case of a common atrioventricular orifice, whether or not the bridging leaflets are attached to the crest of the ventricular septum. Striking feature of AVSD

are the "scooped out" appearance of the septal crest, the deficient inlet septum and the disproportionate size of the inlet and outlet dimensions of the septal surface.

Another important anatomic consideration is the location of the conduction system in AVSD. The location of the AV node is altered in AVSD because the ostium primum ASD often pushes the coronary sinus posteriorly and inferiorly toward the left atrium. This distorts the triangle of Koch and creates a second triangle called the nodal triangle, which is bounded by the coronary sinus, the posterior attachment of the inferior bridging leaflet, and the edge of the atrial septum at the septal defect. The AV conduction axis then runs from the AV node toward the ventricle through the crest of the ventricular septum (2).

There are three different types of AVSD including partial AVSD (p-AVSD), complete AVSD (c-AVSD), and finally, intermediate AVSD (i-AVSD).

## Partial AVSD

In p- AVSD, also known as ostium primum ASD, there is an interatrial communication and separate right and left atrioventricular junctions without an interventricular communication (Fig 1). However, the separate right and left junction in partial AVSD are directly comparable to the common junction in complete AVSD. The only difference is that in the partial defect, a connecting tissue bridge joins the facing portions of the bridging leaflets along the crest of the ventricular septum. The atrioventricular valve apparatus in partial AVSD has been described as having six leaflets. The right superior and right inferior leaflets fuse with the ventricular septum to complete the structure of the right atrioventricular valve, whereas the left superior and left inferior fuse to form the left atrioventricular valve. The commissure between the left superior and inferior leaflets represents the “cleft” of the left atrioventricular valve. In p- AVSD, there is varying degrees of malformation of the left atrioventricular valve, leading to varying degrees of left atrioventricular valve regurgitation (LAVVR).



Fig 1. p-AVSD

## Complete AVSD

c- AVSD results from complete deficiency, of both its fibrous and muscular structures of the atrioventricular septum (Fig2). In addition to the deficiency of the atrioventricular septal structures, c- AVSD has a common atrioventricular junction rather than the normal separate right and left junction. The leaflets guarding the common junction differ markedly from the normal leaflets and are best interpreted as a five leaflet valve. The anterosuperior and inferior leaflets contain entirely within the right ventricle and are comparable with these leaflets in the normal tricuspid valve. In contrast to the normal mitral valve, where the mural leaflet occupies two-thirds of the circumference of the atrioventricular junction, the mural leaflet of the left atrioventricular valve in AVSD is small and tethered between paired left ventricular papillary muscles. The other two leaflets of the five leaflet valve are tethered to papillary muscles in both the right and left ventricle and are known as the superior and inferior bridging leaflets. They are not fused to the “scooped out” crest of the ventricular septum. The degree of bridging and chordal attachment by the superior bridging leaflet forms the basis for the Rastelli classification. The effect of the five leaflet arrangement on the left atrioventricular junction is a trileaflet valve with the aorta that cannot wedge between the normal separate right and left junction and the left ventricular outflow tract anterior to the superior bridging leaflet.



Fig 2. c-AVSD

## Intermediate AVSD

p- AVSD and c-AVSD represent a spectrum of cardiac pathologic conditions. An intermediate form in the middle of this spectrum has been described and termed i-AVSD (Fig 3). This is a rare form of the AVSD characterized by an atrial septal defect (ASD) just above and a ventricular septal defect (VSD) just below the atrioventricular valves. The VSD is often restrictive. A single valvular anulus is usually present, where the superior and inferior bridging leaflets are fused above the crest of ventricular septum. The tissue bridge between superior and inferior bridging leaflets gives the appearance of a separate right and left atrioventricular orifice and partially fuse with the typical "V-shaped" and "scooped out" crest of the ventricular septum.



Fig 3. i-AVSD

The clinical manifestations and natural history of the three types of AVSD are different. p-AVSD is characterized by an ostium primum ASD. The pressure gradient between the two atria and the amount of left-to-right shunt flow is often small and depends upon the size of the defect, and the relative distensibility of the right and left sides of the heart. There is no VSD. p-AVSD is relatively often diagnosed in adults because the defect does usually not cause symptoms in infancy and childhood.

In i-AVSD there is an ASD and often a moderate to small VSD. Clinical manifestations present early in infancy. In i-AVSD the left AV-valve is many times dysplastic and regurgitant. This and the relatively small size of the left ventricle is responsible for earlier symptomatology than is p-AVSD.

In contrast to i-AVSD, c-AVSD is characterized by a large ASD and VSD. Most patients have clinical signs in infancy, according to a large left-to-right shunt and development of pulmonary hypertension.

A number of other cardiac anomalies and abnormalities of the left atrioventricular valve are associated with AVSD including tetralogy of Fallot and double-orifice of the left atrioventricular valve. A thorough understanding of the anatomy, physiology and pathophysiology of these defects is essential for successful diagnosis and correction.

In children with Down's syndrome 40%-50% have congenital heart disease. c-AVSD occurs in 25%, p-AVSD in 8% of the patients.

The aim of surgical treatment for both short and long-term results is closure of the atrial and ventricular defects while preserving or rather improving atrioventricular valve function. In 1955, Lillehei and colleagues (3) reported the first successful repair of an AVSD. Early mortality rates for AVSD repair were then up to 50%. Complications, including complete heart block and recurrent atrioventricular valve regurgitation were common. In 1958, Lev delineated the bundle of His, which helped to decrease the incidence of heart block following surgery. An improved understanding of the structure and function of the common AV valve and a realization of the importance of closing the gap between the anterior and posterior portions of the septal leaflet (the "cleft") led to refinements in surgical technique. This in turn has resulted in a decrease in the short and long-term incidence of left atrioventricular valve regurgitation (LAVVR). Further improvement in the surgical techniques over the last few decades has enabled repair of AVSD with a significant decrease in morbidity and mortality (4, 5). Several other factors are believed to have led to improved survival. The introduction of echocardiography has immensely improved the accuracy of diagnosis of AVSD, and associated congenital cardiac anomalies as well as providing a better understanding of

atrioventricular valve morphology. Further development of perfusion techniques and myocardial protection has decreased the deleterious effects of cardiopulmonary bypass in pediatric surgery, while ongoing improvements in miniaturization of all components of the bypass system are promising (6, 7). Refined anaesthesiological techniques and intensive care management, for instance inhaled Nitric Oxide are responsible for improvements in intraoperative and postoperative support (8, 9). The aforementioned factors have facilitated the introduction of more complex surgical procedures as well as reoperations after previous correction of AVSD. Coincidentally with the transition to more complex surgical procedures we have seen an increasing interest in repair of AVSD in infancy. This transition to repair in early infancy avoids progressive ventricular dilatation from volume overload with atrioventricular valve annular dilatation and valve insufficiency. In addition, the use of palliative procedures such as pulmonary artery banding is no longer considered necessary in surgical treatment of all balanced types of AVSD. Nowadays, c-AVSD is repaired in the first three months of life, i-AVSD is typically repaired before the age of one year and p-AVSD should have been surgically corrected in the first few years of life.

## **Purpose and outline of this thesis**

The aim of this thesis was to analyse and review the results of more than 30 years' experience in the correction of various types of AVSD at Leiden University Medical Centre, Leiden, The Netherlands. Most investigations that evaluate outcome of surgical correction of AVSD focus on one type of AVSD, comprise small number of patients or have short time follow-up. The advantage of a large study that contains surgery in the whole spectrum of AVSD over a period of time, enables us to study different operative techniques, operative strategy, risk factors and outcome. Moreover, a large study can compare surgical results in different time periods. Concerning our operative strategy, we have seen a tendency towards correction of AVSD earlier in life. (8-11) We strongly believe that, with the exception of unbalanced forms, repair of all types of AVSD can and should be surgically corrected early in life. The timing of surgical intervention is an important factor, in ensuring that surgical repair is performed prior to the development of irreversible pulmonary vascular changes (12,13). Intimal fibrosis of the pulmonary vasculature can be found already at the age of 6 months. Many reports have mentioned non-cleft closure to be a risk factor for reoperation. As do others we recommend to close the cleft during repair of AVSD to such an extent that

regurgitation is managed optimally without creating a valvular stenosis. Several other risk factors, including severe preoperative atrioventricular valve insufficiency, additional atrioventricular valve anomalies and coexisting cardiac anomalies are reported in the literature. Controversy still exists regarding the impact of these factors on reoperation and mortality.

We studied a group of 312 patients who were operated for AVSD over a period of more than 30 years. All underwent primary repair and we evaluated the prospect of AVSD type as a risk factor for mortality and reoperation. Other risk factors that were studied included, associated cardiovascular anomalies, left AV- valve dysplasia and absence of cleft closure, as well as era of surgery. This study is presented in chapter 1.

In chapter 2 we present our study regarding patients that were re-operated as a result of left atrioventricular valve regurgitation (LAVVR) following previous correction of AVSD. Reoperation for LAVVR occurs in 4% to 15% after primary repair of AVSD and is an important cause of postoperative morbidity and mortality (14-17). If reoperation is unavoidable, the choice has to be made between left atrioventricular repair and valve replacement. Valve replacement, especially in young children, is accompanied with a high risk of mortality, and pacemaker implantation is not unthinkable. In this study we reviewed our experience with correction of all types of AVSD and included the entire patient population to identify risk factors for reoperation for LAVVR and to determine long-term outcome of the patients that were reoperated for LAVVR. The overall survival of patients reoperated for LAVVR was evaluated by comparing their results to those of patients not reoperated for LAVVR.

In chapter 3 we present our 30 years' experience of surgical repair of AVSD with double-orifice left atrioventricular valve (DO-LAVV), using different strategies to repair the accessory orifice. The outcome of surgical treatment was evaluated by comparing the results of repair of AVSD with DO-LAVV to those of repair of AVSD without DO-LAVV.



Fig 4. DO-LAVV

DO-LAVV was first reported by Greenfield in 1876 (18). In DO-LAVV there are two orifices in the LAVV that are separated by a tissue bridge (Fig 4). These separate orifices, (a cleft containing main orifice and a non-cleft containing accessory orifice), each have their own focus of chordal insertion. The characteristic feature of the DO-LAVV is the “parachute”-like convergent insertion of the tendinous chords in the accessory orifice. It is an uncommon but surgically important condition and may occur as an isolated malformation or in association with other cardiac anomalies but is most often associated with AVSD (19, 20). Autopsy studies have shown a 4,9% to 17,9% incidence of DO-LAVV in patients having an AVSD (21). The presence of DO-LAVV may increase the complexity of the surgical procedure, necessitate technical modifications, and may affect surgical outcome (22). The surgical studies reporting repair of AVSD with DO-LAVV are few and operative mortality has been high, notably in both i-AVSD and c- AVSD. Controversy exists about the surgical management of AVSD with DO-LAVV. Some authors advocate radical repair of a double orifice, combined with suture of the cleft and division of the papillary muscle (22, 23). We, as do others, suggest only suture of the cleft while leaving the accessory orifice and tension apparatus intact and untouched in case no significant regurgitation of the accessory orifice is detected.

In chapter 4 we evaluate the results of concomitant surgical repair of complete AVSD and tetralogy of Fallot (c-AVSD-TOF) repair by a transatrial-transpulmonary approach. The combination of complete AVSD and tetralogy of Fallot is a well recognized congenital heart malformation (24-26). In the early reports the operative mortality of this combined lesion was relatively high, ranging from 29% to 40%, and total correction was thought to be nearly

impossible. Over the last decade, the anatomical features of this malformation have been clarified and diagnostic accuracy, perioperative care and surgical techniques have been improved. The surgical results have now attained acceptable levels with operative mortality ranging from 0% to 11%. However, postoperative complications such as residual ventricular septal defect (VSD), incompetence of the LAVV, pulmonary regurgitation, and residual right ventricular outflow tract obstruction do occur, needing careful management and sometimes require reoperation. The optimal surgical approach to this complex lesion should be designed to minimize the occurrence of these complications. A right ventriculotomy allows a good exposure of the VSD, and the infundibulum can be resected more easily than by a transatrial-transpulmonary approach. However, right ventriculotomy has the potential drawbacks of postoperative right ventricular dysfunction and late dysrhythmias (27). For this reason in Leiden we have always favoured the transatrial-transpulmonary approach.

Two decades ago, expanded polytetrafluorethylene artificial chordae were introduced for mitral valve repair in adults (28, 29). Since then, studies have reported good results, and artificial chordae are now widely accepted for mitral valve repair in the adult (30, 31). Prosthetic valve replacement of native valves in children is preferably avoided because of higher morbidity and mortality due to associated risks of anticoagulation, and a certain risk of reoperation because of patient somatic growth. Therefore, repair should be done whenever possible. However, conventional techniques for mitral or tricuspid valve repair may have limitations because of the wide spectrum of abnormalities of the valvular structures and associated cardiac malformations. Although these artificial chordae lack the potential of growth, they may add to the spectrum of atrioventricular valve repair in children and prevent reoperation. Today, artificial chordae are used for mitral valve repair in children with prolaps of the anterior valve leaflet, and abnormalities of the subvalvular apparatus (32, 33). A few cases have been reported concerning the use of artificial chordae in tricuspid valve repair (34, 35). In some cases of atrioventricular valve repair or re-repair in AVSD, artificial chords may also be considered (36). In chapter 5 we evaluate our 5 years' experience with mitral valve and tricuspid valve repair with artificial chordae in children.

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