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Review Articles

Atrioventricular Septal Defect

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The defining characteristic of an atrioventricular (AV) septal defect is deficiency or the absence of the atrioventricular septum. In patients with complete AV septal defect (CAVSD) the deficiency in the inlet portion of the ventricular septum is greater than that in those with partial defects (PAVSD). In both anomalies an ostium primum defect is immediately above the AV valves. Usually five or more atrioventricular valve leaflets of variable size are present, and often a variability of the commissures or insertion of chordae or number of papillary muscles occurs [6]. Even though there are some revised ideas about bridging leaflets [18, 27], the description of the AV valve leaflets by Rastelli et al. [19] is still accepted and useful for actual surgery. Generally these defects can be associated with a variety of minor or major cardiovascular anomalies. Down's syndrome is common, particularly in patients with an interventricular communication.

The indication for operation in patients with PAVSD is elective in general mainly in the first decade of life. Symptoms depend primarily on the degree of left AV valve incompetence. Ten percent to 15% of these patients have important left AV valve incompetence and may require treatment in infancy [3]. Clinical presentation, chest radiograph, and two- or three-dimensional echocardiography with Doppler studies are sufficient for diagnosis. Patients with CAVSD usually become symptomatic within the first year of life. Pulmonary vascular disease develops within a few months; thus, early operation is recommended [11, 15]. A small group of patients (about 2%) have so-called intermediate AV canal morphology. Their clinical symptoms depend on the size of the interventricular communication, being small in general, and the amount of AV valve incompetence. In this group of patients the indication for operation is elective.

For diagnosis, echocardiography in the hands of an experienced team will provide full information for all forms of atrioventricular defects. Cardiac catheterization is indicated in cases of associated cardiac anomalies, e.g., tetralogy of Fallot, transposition of the great arteries, double-outlet right ventricle, unroofed CS, additional ventricular septal defect (VSD) or questionable hypoplastic left ventricle, especially in patients with suspicion of pulmonary vascular disease [11].

Operative Technique

The standard approach is a median sternotomy. In children with PAVSD and more than 20 kg body weight an anterolateral thoracotomy has been used occasionally. In our early experience cardiopulmonary bypass, profound hypothermia, and total circulatory arrest without cardioplegia were used. Currently, we prefer two venous cannulas, moderate hypothermia (22°C) and low flow (1,2 L/min^{-1} m^{-2}) and one shot of 2°C cold antegrade cristalloid cardioplegia (Bretschneider, Dr. F. Köhler Chemie, Neue Bergstraße 3-7, 64665 Alsbach-Hähnlein, Germany) (40 ml/kg) [5]. After opening the right atrium the morphology of the AV valve is analyzed by injection of cold saline, thus defining the points of valve coaptation or possible regurgitant leaks; at these points, marking sutures are placed. In the past, a single patch was used; however, since 1978 the double-patch technique has been applied. For VSD closure a Dacron patch is sutured with continuous 5–0 polypropylene more on the right side of the crest of the ventricular septum, in particular below the inferior leaflet of the tricuspid valve. The stitch ends at the edge of this leaflet above the coronary sinus. If the McGoon method for avoiding heart block is used, the sutures are placed on the left superior side of the VSD [12]. Then the edges of the anterior and inferior leaflets are sutured to the appropriate upper points of the VSD patch using the marking stitches previously placed. For this interrupted mattress, sutures (5–0 Dacron) are used in very small children, with soft tissue autologous to pericardial pledgets serving as reinforcement. Then the valves are tested by injection of cold saline and the anterior commissure or cleft of the left AV valve is closed by single propylene stitches. Saline solution is injected again to ensure the closure pattern and

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competence of the valves. In case of a central leak, a stitch on the commissures (e.g., of the left anterior and lateral leaflets) may be helpful. Usually, we assess the diameter of the left AV valve with a Hegar dilator, applying the nomograms of Rowlatt et al. [21]. There are rare cases in which, due to the preoperative morphology, it is not possible to achieve a competent valve. The repair is completed by suturing a second patch for closing the ostium primum defect. Some colleagues prefer a pericardial patch in order to avoid hemolysis in cases of left AV valve incompetence; others, like us, have good experience with a Dacron patch. In cases of sparse tissue in front of the coronary sinus, the suture line passes above the AV node, placing the coronary sinus on the left side; otherwise, superficial stitches along the valve ring serve for anchoring the atrial patch.

In patients with a bridging left anterior leaflet (Rastelli type C) the VSD patch slides underneath the leaflets between the chordae; sometimes, an incision of the free edge of the leaflet in the direction of the anulus is required. In case of an intermediate type the small subvalvar interventricular communications have to be identified and closed with interrupted sutures that attach that portion of the leaflets to the ventricular septum, avoiding the crest in the posterior region. In case of additional anomalies a different operative management may be indicated.

Results

Our study comprises 586 patients with atrioventricular septal defect operated on between October 1974 and December 1995. The complete form of AVSD was present in 385 patients and 190 had partial defects. Eleven patients showed an intermediate type of AV septal defect. Of the 385 patients with complete atrioventricular septal defect 65 had major associated cardiovascular anomalies. Of the 320 patients with isolated CAVSD 274 underwent repair: 167 (60.9%) were female and 107 (39.0%) male. Down's syndrome was present in 67.1% (184 patients). According to Rastelli's classification, 67.9% (186 patients) presented type A, 3.6% (10 patients) type B, and 28.5% (78 patients) type C. Age at repair ranged between 1 month and 15.2 years (median, 0.9 years). Weight varied between 2.8 and 46.2 kg (median; 6.3 kg); 56.6% (155 patients) of repairs were performed on children <1 year old and 23% (63 patients) on children <6 months old. Since primary repair is our main goal, 198 (72.3%) patients had primary repair, 144 (72.7%) of these were <1 year old. Seventy-six (27.7%) patients underwent palliative surgery 1 month to 11.9 years (median, 2.9 years) before repair. Operative death of 274 patients with isolated CAVSD who underwent repair was 6.6% (late death, 12.4%). Of 198 primary corrected patients, 15 (7.6%) died early and 28 (14.1%) late, mainly due to

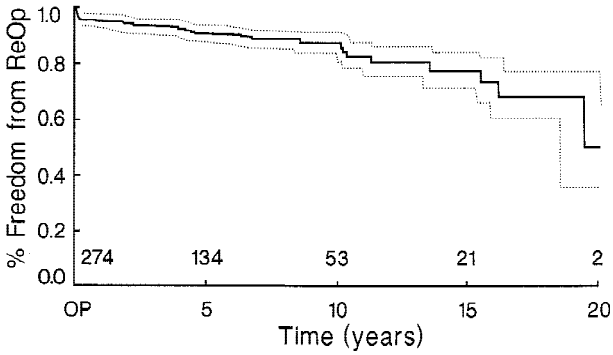

Fig. 1. Kaplan–Meier estimate of freedom from reoperation for postrepair complete atrioventricular septal defect (CAVSD), with 70% confidence intervals. Freedom from reoperation at 10 and 20 years is 82.5 ± 3.8% and 50 ± 16%, respectively.

congestive heart failure and/or pulmonary problems. During the years, operative death decreased from 17.6% (1974–1979) to 5.0% (1990–1995) despite an increase in the number of primary corrected patients younger than 6 months. Patients with pulmonary artery banding before repair showed an operative death of 3.9% (late death, 7.9%).

Within an observation period of more than 20 years, 31 (11.3%) patients required 35 reoperations 7 days to 8.0 years (mean, 1.2 ± 2.0 years) after repair (Fig. 1). Half of these patients underwent reoperation within 2 months after repair. In 88.5% (31/35) reoperations was significant left AV valve (LAVV) incompetence. Refixation and repair of LAVV was possible in 87% (27/31 cases), except in 5 patients requiring valve replacement. Mechanical prostheses (size, 19–22 mm) were used in 3 cases, 1 patient received a biological prosthesis, and another received a stentless mitral prosthesis size 14. None of our patients underwent LAVV replacement during the original operation. Analysis of potential risk factors that might be associated with a higher reoperation rate showed no influence of weight <5.0 kg ($p = 0.09$) or age <6 months ($p = 0.13$). Rastelli type C was more prevalent in reoperated patients (42% vs 25.5%). However, this difference was not significant ($p = 0.29$). Operative technique (one-patch vs two-patch technique) ($p = 0.50$) and incision of anterior and/or posterior bridging leaflets ($p = 0.77$) had no significant influence on reoperation rate.

Regarding patients with PAVSD, 97 were females and 93 males. The indication for operation was elective at an average age of 9.27 ± 11.5 years (18 days to 58 years). Seventy-nine patients (42%) had operative correction before entering school, e.g., <5 years. An indication for operation in childhood is given by severe congestive heart failure and/or the presence of significant mitral valve incompetence, failure to thrive, or the presence of additional cardiovascular anomalies such as co-

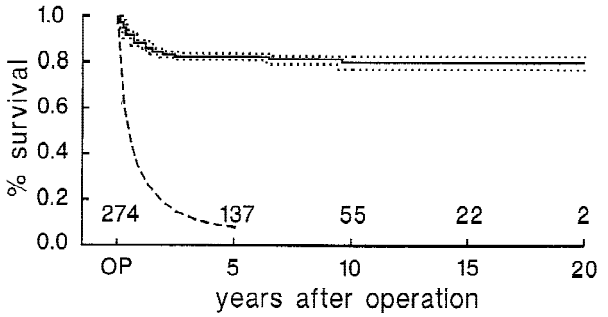

Fig. 2. Kaplan–Meier estimate of survival and 70% confidence intervals for 274 patients undergoing complete atrioventricular septal defect (CAVSD) repair. The actuarial survival at 20 years is 79 ± 2.8% compared to the natural course according to Berger et al. (*Ann Thorac Surg 27*:104–111, 1979).

arctation. Operative death is low (<2%). However, there is a certain rate of reoperation mainly for valve incompetence. Some of these patients show deficiency of the left inferior leaflet of their AV valve, making effective repair very difficult. In our group, this occurred in 14 patients (8%), 4 of which had two reoperations and 1 had four reoperations. Thus, in most children repair of the incompetent mitral valve was possible but in 7 patients (3.7%) implantation of a mechanical valve was necessary, and in 2 patients tricuspid valve replacement was indicated. Overall, 7 patients (3.7%) required a pacemaker, and some developed complete AV block years after correction. Even though right bundle branch block is rare in this malformation, in the long-term follow-up there is a higher rate of atrial arrhythmias compared to the group of CAVSD. Ninety percent of the operated patients show a normal development.

Follow-Up

Follow-up data of 204 (92%) of 222 late survivors are available. The observation time ranged between 1 year and 21. 5 years, with a mean of 7.2 years or 1593 patient years.

Actuarial survival rate after 20 years is 79 ± 2.8% (Fig. 2). Eighty-seven percent (193 patients) are in NYHA class I, 81.5% (181 patients) do not need any medication, and 90.5% present sinus rhythm. Repeated echocardiographic evaluation of LAVV function in 155 patients (70% of late survivors) revealed insignificant valve incompetence in 58% (90/155), moderate incompetence in 35.5% (55/155), and severe incompetence in 6.5% (10/155).

Discussion

With increasing experience, surgical repair of AV septal defects has become a standard procedure with excellent results [2, 24–26, 30].

Repair of isolated PAVSD carries a low hospital death rate (<1%) [24] whereas the death rate of isolated CAVSD varies between 2.5% and 13% [2, 25, 26, 30]. Patients with complex defects are known to carry a higher risk and require different surgical strategies [9, 16, 28].

Most early deaths are due to acute cardiac failure or failure of LAVV repair [13]. Poor preoperative clinical status is known to be a risk factor for hospital death in both groups [24]. Hospital death in patients with advanced preoperative cardiorespiratory instability differs significantly. Left ventricular hypoplasia, additional AV valve anomalies (e.g., double-orifice mitral valve), or a singulary papillary muscle are also known risk factors for increased death [2, 7, 8, 24] (Table 1). In contrast to these reports, none of our patients with double-orifice LAVV or solitary left papillary muscle died, but in our study the presence of a severe dysplastic left LAVV was associated with a higher risk for operative death ($p = 0.001$). We have changed our operative approach from the one-patch technique to the two-patch technique. We discontinued the one-patch technique due to a higher rate of postoperative valve incompetence. Since 1978 the two-patch repair has been our method of choice. However, the one-patch technique proved to be satisfactory in quite a number of publications [1, 4, 14, 22, 29]. Williams et al. [29], for instance, applied the one-patch method for CAVSD repair in 32 children without early death. By analyzing the influence of operative technique, it was determined that there was a trend toward better results in the two-patch group but this failed to reach significance ($p = 0.06$). Today, primary repair of CAVSD is the treatment of choice [2, 25, 30]. Most authors advocate primary repair in children <1 year or 6 months of age [4, 14, 22, 25, 30]. Yasui et al. [30] analyzed 40 patients (23 within the first year of life) and reported a 2.5% hospital death rate. As in other studies, in our series the number of patients who underwent repair at an age of <1 year constantly increased over the years with a simultaneous reduction in death [26]. Timing of operation is an important factor; it must be carried out before development of irreversible pulmonary vascular changes [4, 15]. Pulmonary vascular disease in CAVSD develops during the first year of life, e.g., intimal fibrosis of the pulmonary vasculature can be found within 6 months of life [15]. Increasing degree of pulmonary vascular resistance is a known risk factor for hospital death in patients with complete AV septal defects but also in patients with partial defects [13]. Another argument in favor of repair in early childhood is the possible increase in degenerative changes of the atrioventricular valve as age increases [30]. Studer et al. [24] stated that young age was an incremental risk factor in the early period of their study (1967–1976) but disappeared as a risk factor after 1976. McGrath and Gonzalez-Lavin [13] found that a smaller body surface area

Table 1. Summary of potential risk factors associated with operative death

Factor	Reference
	PAVSD
Date of operation	McGrath and Gonzalez-Lavin [13]
Young age at repair	McGrath and Gonzalez-Lavin [13]
High pulmonary vascular resistance	McGrath and Gonzalez-Lavin [13]
Severe AV valve incompetence	Studer et al. [24]
Poor preoperative clinical status	Studer et al. [24]
	Isolated CAVSD
Date of operation	Studer et al. [24], Tweddell et al. [26], DHM (1997)
High pulmonary vascular resistance	Rizzoli et al. [20], McGrath and Gonzalez-Lavin [13]
Smaller size	Rizzoli et al. [20], McGrath and Gonzalez-Lavin [13]
Longer cross-clamp time	McGrath and Gonzalez-Lavin [13]
Severe preop AV valve incompetence	Studer et al. [24]
Poor preoperative clinical status	Studer et al. [24[, Alexi-Meskishvili et al. [2], DHM (1997)
Additional AV valve anomalies	Studer et al. [24], Clapp et al. [8], Alexi-Meskishvili et al. [2], DHM (1997)
Left ventricular hypoplasia	Studer et al. [24], Clapp et al. [8]
Postoperative pulmonary hypertension	Alexi-Meskishvili et al. [2]

PAVSD, partial atrioventricular septal defect; CAVSD, complete atrioventricular septal defect.

was an incremental risk factor for hospital death in CAVSD patients. Patients younger than 6 months and weighing less than 5 kg needed longer ventilatory assistance (mean, 8.4 ± 12.3 days) compared to those more than 5 kg (mean, 3.3 ± 4.3 days; $p = 0.001$) and a longer intensive care unit stay (mean, 12.2 ± 12.9 days vs 7.3 ± 8.8 days; $p = 0.004$). However, by analyzing different time periods—age <6 months ($p = 0.03$) and weight <5.0 kg ($p = 0.44$)—no significant influence after 1984 was shown, indicating that increasing research is of germane importance. Of particular interest is the long-term function of the repaired AV valve. It appears that patients without interventricular communication are at greater risk of valve repair failure than are those with CAVSD [24]. Late valve replacement is more common in the group with partial AV septal defect than in the latter. Thus, 7 (3.7%) of 190 patients with PAVSD in our series needed valve replacement compared to 5 (1.8%) of 274 who underwent CAVSD repair. In 155 of our patients (70% of late survivors after CAVSD repair) AV valve function has been analyzed by Doppler echocardiography. Fifty-eight percent (90/155) revealed insignificant valve incompetence, 35.5% (55/155) moderate incompetence, and 6.5% (10/155) severe incompetence. The incidence for LAVV reoperation after CAVSD repair was 11.3%. According to the reports in the literature the rate of reoperation varies between 6% and 9% [2, 10, 25, 26]. A rare complication presents left ventricular outflow tract obstruction that in some cases can be repaired through the aortic valve; very rarely are a repositioning of the VSD patch and re-repair of the LAVV necessary. The rate of complete AV block after CAVSD repair varies in the literature between 2% and 4% [2, 25, 26]; in our hospital 14 CAVSD patients (5.1%) needed a pacemaker within an observation period of more than 20 years.

Another moot point is the role of pulmonary artery banding in patients with CAVSD. In the early studies (1974–1978) CAVSD repair was associated with a higher death rate than it is today. Some authors used pulmonary artery banding (PAB) to increase the age at correction while offering protection from the rapidly progressive pulmonary vascular disease [8, 13, 15]. Others were discouraged with PAB because of inadequate palliation in children with moderate to severe LAVV incompetence, high operative death (5–40%), and difficulty with removal of the band [22]. Despite a policy toward primary repair in young children, we do have a subgroup of 76 patients who underwent a two-stage procedure with PAB on average 3.5 years before repair. Today, a hypoplastic left ventricle (left ventricular end diastolic volume <50% of predicted normal), elevated pulmonary vascular resistance (>6 U/m^2 not reactive to oxygen test or nitric oxide), a severe dysplastic LAVV, coexisting coarctation, and low weight (<2800 g) are indications for palliative surgery. Comparing operative death of patients who underwent a two-stage procedure to primary corrected patients, we found no significant difference [$p = 0.28$). This coincides with the data published by Clapp et al. [8] and Alexi-Meskishvili et al. [2]. Tweddell et al. [26] report that none of their patients who underwent PAB died. In a subgroup of 46 CAVSD patients with advanced pulmonary vascular disease, PAB was the only therapy possible. Operative death in this group was 13%, with 11 late deaths (26%) within 22 years.

Without doubt, primary repair within the first months of life is the therapy of choice for patients with

Table 2. Analysis of death and morbidity data in the literature

No. of patients	Reference	Operative death (%)	Reoperation rate (%)
	PAVSD		
57	Rizzoli et al. [20]	6	
80	McGrath and Gonzalez-Lavin [13]	5	11
90	Pan-Chih and Chen-Chun [17]	4.4	
	Isolated CAVSD		
34	Abbruzzese et al. [1]	18	8.8
43	Rizzoli et al. [20]	37	
54	McGrath and Gonzalez-Lavin [13]	15	24
70	Clapp et al. [8]	13	
40	Yasui et al. [30]	2.5	
40	Thies et al. [25]	10	7.5
115	Tweddell et al. [26]	13.9	6
120	Alexi-Meskishvili et al. [2]	10	6.8

PAVSD, partial atrioventricular septal defect; CAVSD, complete atrioventricular septal defect.

complete atrioventricular septal defects. In particular cases a two-stage approach permits excellent results. Today this operation can be performed with a low death rate (<5%) [26], an acceptable reoperation rate (<10%) [26], and a very low risk of complete AV block (<2%) [26]. The function of the reconstructed AV valve over more than 20 years is remarkable, making a valve replacement necessary in only <2% of patients [2, 26]. We believe that valve replacement is never indicated at the primary operation. Actuarial survival of patients with CAVSD repair after 20 years is more than 80% [13, 26] (Table 2).

Summary

Anomalies of the atrioventricular septum, such as CAVSD or PAVSD, require surgical therapy, the former early in childhood and the latter electively before entering school.

Primary repair is the treatment of choice. In the case of heart failure and/or moderate or severe LAVV incompetence, operative correction should be performed as early as possible. Otherwise, the operation should be carried out within the first 4–6 months of life. A two-stage approach—that is, PAB followed by later repair—is indicated in patients with heart failure, the absence of valve incompetence, a body weight <2500 g, the presence of coarctation or severe dysplastic LAVV, and/or a hypoplastic left ventricle. In the latter, quite rare group early decision making is required because a Fontan-type repair is hampered by the early development of pulmonary vascular disease. In our study, a two-stage repair permitted very good results even during a long-term observation period. For patients with PAVSD, operation should be completed between 2 and 4 years of age before entering school.

For correction of CAVSD, we and many other surgeons prefer the two-patch technique, which permits more individual and precise repair of the common AV valve. In our more than 20 years' experience the function of the repaired AV valve has been remarkable; the rate for valve repair varies between 6% and 10% in patients with PAVSD. Valve replacement is rarely necessary, occurring in only 1.8% of patients after CAVSD correction and in 3.7% of patients after PAVSD correction. At the primary operation valve replacement is never indicated. The complication of complete AV block has become quite rare and is below 2%. Operative death varies between 1% and 5%. Risk factors include congestive heart failure, additional cardiovascular anomalies (e.g., coarctation), and pulmonary hypertension.

In our more than 20 years' experience, >80% of patients with partial or common AV septal defects show normal development without medication, even after a two-stage approach.

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Congenital Heart Disease Visualized

By G.S. Shirali, F. Lomabno, D. Dayer, R.L. Larsen, D.L. Kearney, and J.T. Bricker; Futura Publishing, Inc., 1997

This CD-ROM presents echocardiographic and pathological findings of the normal heart and a wide array of congenital heart diseases. Normal cardiac anatomy and pathological findings in congenital heart diseases are presented clearly through short echocardiographic looped clips and still pictures of cardiac specimens. The echocardiographic images are exquisitely clear and are accompanied by diagrams illustrating cardiac anatomy and voice elaborating on presented images. Short segments of text are also available. The table of contents is somewhat haphazard; it includes various normal anatomical segments dispersed among congenital and acquired heart diseases. The last four segments deal with echocardiographic images and pathological specimens of some operative repairs.

This CD-ROM is limited to cardiac anatomy and pathology as seen through echocardiography and autopsies, which provides an excellent correlation between the two. Clinical findings, angiography, and management aspect of heart diseases in children are not presented as they are not the focus of this CD-ROM. Correlating echocardiographic and angiographic images to autopsies is a crucial element in learning to construct a mental 3-D image from pictures of a 2-D nature obtained through angiography or echocardiography, and this CD-ROM provides an excellent opportunity to acquire such a skill.

Ra-id Abdulla
Associate Editor