European Journal of Cardio-Thoracic Surgery 0 (2019) 1–8
doi:10.1093/ejcts/ezz085

ORIGINAL ARTICLE

CONGENITAL

Cite this article as: Pontailler M, Haidar M, Méot M, Moreau de Bellaing A, Gaudin R, Houyel L *et al.* Double orifice and atrioventricular septal defect: dealing with the zone of apposition. Eur J Cardiothorac Surg 2019; doi:10.1093/ejcts/ezz085.

Double orifice and atrioventricular septal defect: dealing with the zone of apposition†

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Received 17 September 2018; received in revised form 13 January 2019; accepted 30 January 2019

Key question

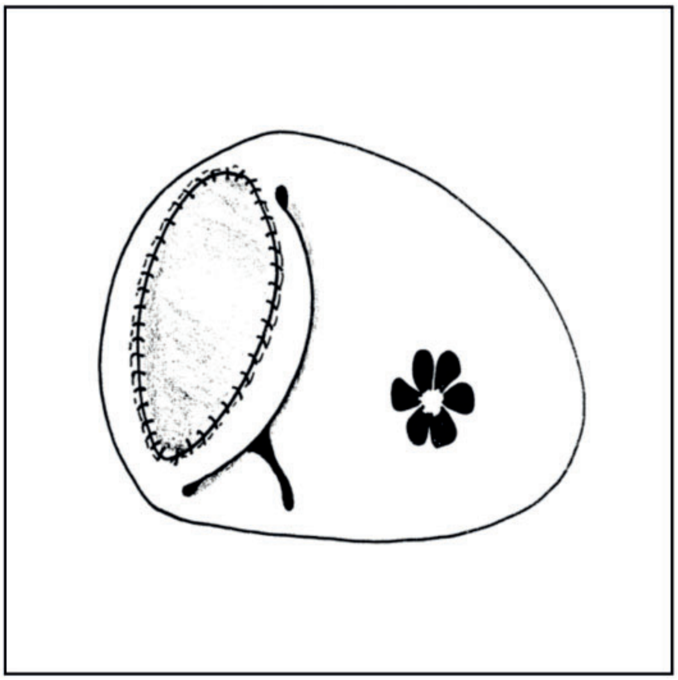
What is the outcome of an atrioventricular defect associated with a double orifice of the left atrioventricular valve?

Key finding(s)

Reoperation rate of 15%, especially if the ventricles are unbalanced, subvalvular apparatus is abnormal, and regurgitation > grade 2

Take-home message

The mortality rate has decreased but the reoperation rate remains a key problem. Anterior hemivalve pericardial enlargement can be an alternate strategy.



Abstract

OBJECTIVES: A double orifice of the left atrioventricular valve (LAVV) associated with atrioventricular septal defects (AVSD) can significantly complicate surgical repair. This study reports our experience of AVSD repair over 3 decades, with special attention to the zone of apposition (ZoA) of the main orifice, and presents a technique of hemivalve pericardial extension in specific situations.

METHODS: We performed a retrospective study from 1987 to 2016 on 1067 patients with AVSD of whom 43 (4%) had a double orifice, plus 2 additional patients who required LAVV pericardial enlargement. Median age at repair was 1.3 years. Mean follow-up was 8.2 years (1 month–32 years).

†Presented at the 32nd Annual Meeting of the European Association for Cardio-Thoracic Surgery, Milan, Italy, 18–20 October 2018.

RESULTS: Associated abnormalities of the LAVV subvalvular apparatus were found in 7 patients (5 parachute LAVV and 2 absence of LAVV subvalvular apparatus). ZoA was noted in 4 patients (9%): partially closed in 15 (35%) and completely closed in 24 (56%). Four patients required, either at first repair or secondarily, a hemivalve enlargement using a pericardial patch without closure of the ZoA. The early mortality rate was 7% ($n = 3$), all before 2000. Two patients had unbalanced ventricles and the third had a single papillary muscle. There were no late deaths. Six patients (14%) required 7 reoperations (3 early and 4 late reoperations) for LAVV regurgitation and/or dysfunction, of whom 4 (9%) required mechanical LAVV replacement (all before 2000). Freedom from late LAVV reoperation was 97% at 1 year, 94% at 5 years and 87% at 10, 20 and 30 years. Unbalanced ventricles ($P = 0.045$), subvalvular abnormalities ($P = 0.0037$) and grade >2 LAVV postoperative regurgitation ($P = 0.017$) were identified as risk factors for LAVV reoperations. Freedom from LAVV mechanical valve replacement was 95% at 1 year, 90% at 5 years and 85% at 10, 20 and 30 years. An anomalous LAVV subvalvular apparatus was identified as a risk factor for mechanical valve replacement ($P = 0.010$). None of the patients who underwent LAVV pericardial extension had significant LAVV regurgitation at the last follow-up examination.

CONCLUSIONS: Repair of AVSD and double orifice can be tricky. Preoperative LAVV regurgitation was not identified as an independent predictor of surgical outcome. LAVV hemivalve extension appears to be a useful and effective alternate surgical strategy when the ZoA cannot be closed.

Keywords: Atrioventricular septal defects • Atrioventricular canal defects • Double orifice • Mitral valve repair • Mitral valve replacement • Surgical technique • Outcomes • Congenital heart disease

INTRODUCTION

A double orifice (DO) of the left atrioventricular valve (LAVV) is a rare valvular cardiac abnormality that can be encountered in patients with congenital heart disease. A DO is rarely isolated but is usually associated with atrioventricular septal defects (AVSD), occasionally in association with obstructive left-sided lesions, cyanotic congenital heart disease and non-compaction of the left ventricle [1].

The incidence of DO LAVV has been reported in the literature to be around 3.6–7.5% [2–6].

DO LAVV in AVSD can significantly complicate surgical repair. Closure of the zone of apposition (ZoA) usually limits regurgitation (if the left mural valve of the orifice is correctly developed) but can create LAVV stenosis. Presence of a DO LAVV in AVSD has a bad reputation, leaving the patient at risk for LAVV dysfunction, reoperation and death [6–11].

This study reports our experience of AVSD repair over 3 decades, with or without closure of the ZoA, and presents a technique of hemivalve pericardial extension.

MATERIALS AND METHODS

Patient characteristics

From 1987 to 2016, 1067 patients with AVSD underwent surgical biventricular repair in our institution. Among them, 43 patients (4%) had a DO LAVV. We excluded patients who presented with AVSD and tetralogy of Fallot and patients who had severe unbalanced ventricles leading to univentricular repair. Patients with unbalanced ventricles but who underwent biventricular repair were included.

Two patients with AVSD and DO LAVV who were operated on by our surgical team in another institution and who required anterior hemivalve pericardial extension (need for extension was suspected from preoperative echocardiogram) were also included in this study (but not in the cohort data or statistical analysis) to report our experience with this surgical alternative.

DO anatomy and position were clearly documented in all patients but 1 (Table 1): the DO was most frequently described as posterior in 31 patients (72%). Abnormalities of the left subvalvular apparatus were described in 7 patients, most frequently a parachute LAVV (5 patients).

Patients were divided in 2 groups, depending on the surgical ZoA operative strategy: either completely closed ($n = 24/43$) or respected ($n = 19/43$, no or partial closure).

Surgical management

Surgical technique for repair of atrioventricular septal defects. Surgical repair of AVSD was performed via a median sternotomy, with standard aortic and bicaval venous cannulation and normothermic cardiopulmonary bypass (35–37°C). Cardioplegia was obtained by infusion of anterograde normothermic hyperkalaemic blood in the aortic root and subsequently repeated every 10 min or less when myocardial activity resumed.

Diagnosis of DO LAVV was made with preoperative transthoracic echocardiography and/or identified at surgery. Closure of the ZoA was performed when the residual orifices were operatively evaluated as non-restrictive; otherwise, ZoA was partially

Table 1: Anatomy and fate of direct orifice left atrioventricular valve

LAVV anatomy and fate	*N* (%)
DO anatomy (data missing from 1 patient)	
Posterior	31 (72)
Superior	7 (16)
Inferior	3 (7)
Septal	1 (2)
LAVV subvalvular apparatus abnormalities	
Absence	2 (5)
Parachute LAVV	5 (12)
ZoA closure	
Complete	24 (56)
Partial	15 (35)
None	4 (9)
LAVV MVR	4 (9)

DO: double orifice; LAVV: left atrioventricular valve; MVR: mechanical valve replacement; ZoA: zone of apposition.

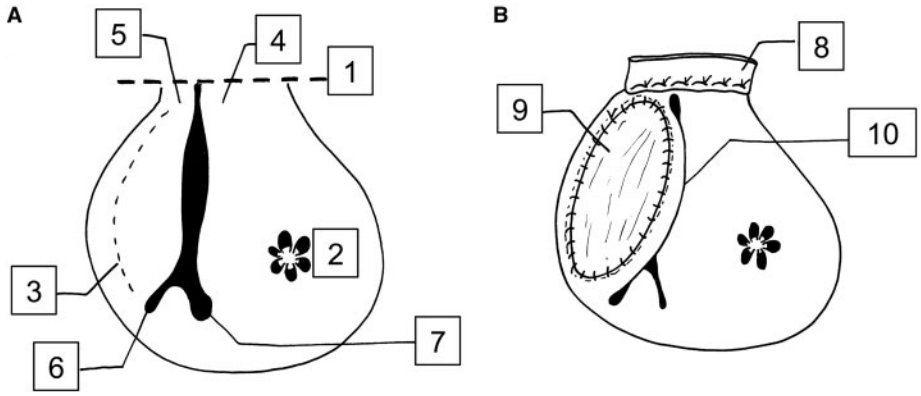


Figure 1: Enlargement of an anterior pericardial patch hemivalve. (**A**) Double-orifice left atrioventricular valve before repair: (1) ventricular crest; (2) accessory orifice; (3) line of incision for pericardial extension; (4) posteroinferior bridging leaflet; (5) anterosuperior bridging leaflet; (6) anterosuperior commissure; and (7) posteroinferior commissure). (**B**) The anterior bridging leaflet is detached and the treated autologous pericardial patch is sutured to increase leaflet coaptation; the zone of apposition and the double orifice are left open: (8) interatrial patch; (9) autologous pericardial patch extension; and (10) zone of coaptation.

closed or left open. None of the accessory DO were surgically closed; all of them were conserved because none was found to be significantly regurgitant at preoperative evaluation. If both VSD and ASD required closure, the double patch technique was used, preferentially with a heterologous pericardial patch. An LAVV commissuroplasty was performed in 11 patients.

The median age and weight at operation were 1.3 years (range 1 month–30 years) and 9 kg (range 3.2–50 kg), respectively.

Hemivalve pericardial patch enlargement. Four patients, 2 of our cohort and 2 patients operated on by our surgical team at another institution, required an anterior hemivalve pericardial patch enlargement (Fig. 1). These patients presented with a specific anatomical configuration whereby complete closure of the ZoA was mandatory to obtain acceptable competence but would have been responsible for a significant degree of stenosis. In 2 cases, despite complete closure of the ZoA, residual regurgitation was significant (hypoplasia of the mural leaflet was related to the main orifice).

An autologous pericardial patch was then harvested and treated in glutaraldehyde for 8–10 min. The hemivalve enlargement technique consisted of detaching the anterior bridging leaflet, 1 mm from the annulus and from the ZoA to the commissure. This technique can be applied to the posterior hemivalve. We do not divide the papillary muscle, even localized in the axis of the inflow, in order to avoid any prolapse. With this technique, the stenosis is not an issue because of the absence of complete closure of the ZoA. An oval autologous treated patch is sewn using 6/0 or 7/0 polypropylene running sutures, depending on the age of the patient at repair and the thickness of the leaflet. The ZoA is left totally or partially open to avoid tension and tearing. The increased hemivalve surface allows better LAVV coaptation. If deemed necessary, a commissuroplasty was performed to increase leaflet coaptation, only if a stenosis was not to be feared.

Data collection and follow-up

For this retrospective monocentric study, we reviewed our Paediatric Cardiac Surgery database to identify the patients who required biventricular repair for AVSD. The Paris V University Ethics Committee granted approval for review of health records. The need for individual consent was waived due to the retrospective nature of the study.

The mean follow-up was 8.2 years (1 month–32 years). The surviving patients had an annual examination by their referring cardiologist.

Statistical analysis

Continuous data are expressed as median and interquartile range or mean and standard deviation. Categorical data are expressed as number and percentage. Univariable analysis was performed using either the χ^2 or the Fisher's exact test for categorical data when cell frequencies were ≤ 5 or the Student's t-test for continuous data. A P-value <0.05 was considered significant. Overall survival and freedom from events were calculated using the Kaplan–Meier method with curves compared using the log-rank test. Data were analysed using GraphPad Prism version 6 software (GraphPad Software, La Jolla, CA, USA).

Although we report our experience in 45 patients, statistical analyses were performed only on the 43 patients who underwent initial repair in our institution.

RESULTS

Operative and postoperative data

Patient pre-, peri- and postoperative data are presented in Table 2. The sex ratio was 1/2 (men/women).

AVSD was associated with Down syndrome in 6 (14%) patients. Major associated cardiac lesions included coarctation in 4 (9%) patients and a left partial anomalous pulmonary venous return in 1 patient (2%). Seven patients had unbalanced ventricles: mild to moderate hypoplasia of the left ventricle in all, 2 with associated coarctation of the aorta. Two patients (5%) required previous palliation with pulmonary artery banding.

Thirteen patients (30%) were operated on before 2000.

Preoperative LAVV regurgitation was described as none to trivial in 10 (23%) patients, grade 1 in 13 (30%) patients, grade 2 in 12 patients (28%), grade 3 in 4 patients (9%) and grade 4 in 4 patients (9%). Twenty-four patients (56%) underwent complete

Table 2: Patient demographics

	ZoA complete closure ($n = 24$)	ZoA conserved (partial closure or left open) ($n = 19$)	P value
Preoperative data			
Male gender	8 (33)	6 (32)	1
Age (years)	1.7 (3.8)	0.8 (4.2)	0.53
Weight (kg)	9.1 (8)	8.3 (13)	0.92
Year <2000	5 (21)	8 (42)	0.19
AVSD type			0.76
cAVSD	10 (42)	10 (53)	
iAVSD	2 (8)	1 (5)	
pAVSD	12 (50)	8 (42)	
Unbalanced ventricles	1 (4)	6 (32)	0.032
Previous PAB	0	2 (11)	0.19
Peri- and postoperative data			
Preoperative LAVV regurgitation grade >2	4 (17)	4 (21)	1
CPB	97 (40)	108 (96)	0.22
Aortic clamping	62 (27)	79 (72)	0.085
Postoperative LAVV regurgitation grade >2	2 (8)	0	0.49
Inotropic support (days)	2 (4)	1 (3)	0.85
Ventilation time (h)	10 (44)	8 (39)	0.14
ICU (days)	3 (5)	2 (5)	0.40
Hospital stay (days)	8 (4)	7 (6)	0.52
Outcomes			
Deaths	1 (4)	2 (11)	0.58
Reoperation LAVV	3 (12)	3 (16)	1
LAVV MVR	2 (8)	2 (11)	1
Pericardial enlargement	1 (4)	1 (5)	1
Pacemaker for AV block	1 (4)	0	1

Categorical values are expressed as n (%) and quantitative values are expressed as median (interquartile range).

AV: atrioventricular; AVSD: atrioventricular septal defect; cAVSD: complete AVSD; CPB: cardiopulmonary bypass; iAVSD: intermediate AVSD; ICU: intensive care unit; LAVV: left atrioventricular valve; MVR: mechanical valve replacement; PAB: pulmonary artery banding; pAVSD: partial AVSD; ZoA: zone of apposition.

closure of the ZoA, whereas 15 (35%) had a partial closure. ZoA was left open in 4 patients (9%). All DO were surgically conserved because none were found to be significantly regurgitant at evaluation.

Postoperative LAVV regurgitation was none to trivial in 35 patients (81%), grade 2 in 6 (14%) and grade 4 in 2 (5%). Both patients with grade 4 postoperative LAVV regurgitation underwent subsequent early reoperation.

The ZoA tended to be more frequently only partially closed or left open in patients with unbalanced ventricles ($P = 0.032$) and who had required previous palliation by pulmonary artery banding (not significant) (Table 2).

Mortality rate

The early mortality rate was 7% ($n = 3$), all before 2000. There were no late deaths (Fig. 2A).

The first patient had a partial AVSD with balanced ventricles and a single papillary muscle associated with the DO LAVV. The ZoA had been completely closed during repair but postoperative echocardiography showed grade 4 LAVV regurgitation. He underwent LAVV mechanical valve replacement (MVR) after a second LAVV repair failed, 12 days after the initial repair, but he did not survive.

The other 2 patients had unbalanced ventricles (left ventricular end-diastolic volume: z score -4 DS) and the ZoA had only been partially closed. The first patient had been diagnosed with complete AVSD and grade 4 preoperative LAVV regurgitation. He died the night after surgery of pulmonary oedema and biventricular dysfunction. Necropsy found a stenotic LAVV. The second patient had a post-natal diagnosis of partial AVSD associated with aortic coarctation. He underwent ostium primum closure, partial ZoA closure and coarctation repair when he was 1 month old. He died 75 days after repair of septic shock complicating a chylothorax that had required thoracic duct ligation. At the last echocardiogram, the mean LAVV gradient was 2 mmHg, associated with grade 2 LAVV regurgitation.

Table 3 summarizes the univariable analysis of risk factors for mortality. Surgery before 2000 was identified as a risk factor for mortality ($P = 0.045$). Early MVR and presence of unbalanced ventricles showed a trend but no statistical significance was found (P-value: 0.070 and 0.064, respectively).

There was no statistically significant difference regarding the preoperative and postoperative LAVV regurgitation (P-value: 0.47 and 0.14, respectively) or the ZoA surgical strategy ($P = 0.58$).

Reoperations

Nine patients (21%) required subsequent reoperation after surgical repair: 6 for LAVV dysfunction, 1 pacemaker implantation for complete AV block, 1 for pericardial effusion and 1 for subaortic resection 12 years after the initial surgery.

Six patients (14%) required 7 reinterventions on the LAVV. Three patients required early reintervention at 3, 12 and 14 days after surgery. Two patients underwent subsequent LAVV repair (1 with an anterior hemivalve pericardial enlargement), whereas the third patient required LAVV MVR because the re-repair failed, leading to death. Three patients from the initial cohort required late reoperation consisting of LAVV MVR in all cases, 5 months,

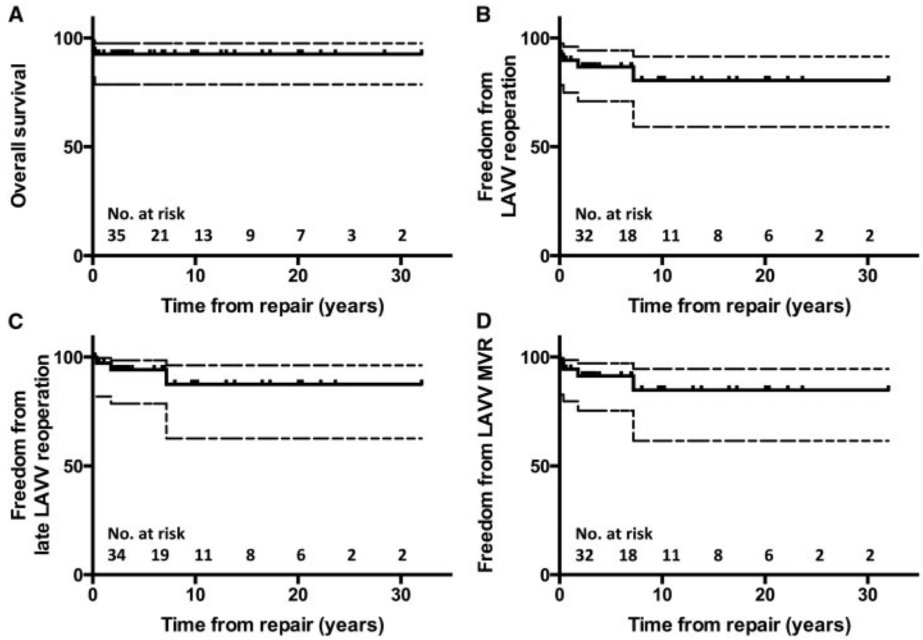


Figure 2: Survival and freedom from reoperation after surgical repair of atrioventricular septal defect with direct orifice left atrioventricular valve. (**A**) Overall survival was 95% [95% confidence interval (CI): 92–100] at 30 days and 93% (95% CI: 88–100) at 1, 5, 10, 20 and 30 years. (**B**) Freedom from left atrioventricular valve reoperation was 93% (95% CI: 87–100) at 30 days, 90% (95% CI: 83–100) at 1 year, 87% (95% CI: 79–100) at 5 years and 80% (95% CI: 70–100) at 10, 20 and 30 years. (**C**) Freedom from late left atrioventricular valve reoperation was 97% at 1 year (95% CI: 95–100), 94% at 5 years (95% CI: 90–100) and 87% (95% CI: 79–100) at 10, 20 and 30 years. (**D**) Freedom from LAVV MVR was 95% at 1 year (95% CI: 90–100), 90% at 5 years (95% CI: 86–100) and 85% (95% CI: 75–100) at 10, 20 and 30 years. LAVV: left atrioventricular valve; MVR: mechanical valve replacement.

Table 3: Univariable analysis of risk factors for mortality in patients with atrioventricular septal defect and direct orifice left atrioventricular valve

Risks factors for death	*P*-value
Preoperative factors	
AVSD type	1.0
Down syndrome	1.0
Preoperative LAVV regurgitation grade >2	0.47
Unbalanced ventricles	0.064
Subvalvular apparatus abnormalities	0.064
Perioperative factors	
Surgery before 2000	0.045
ZoA closure	0.58
Postoperative factors	
Postoperative LAVV regurgitation grade >2	0.14
Early MVR	0.07

Only surgery before 2000 was statistically significant. Unbalanced ventricles, subvalvular abnormalities and early MVR showed a trend but no statistical significance was reported.

AVSD: atrioventricular septal defect; LAVV: left atrioventricular valve; MVR: mechanical valve replacement; ZoA: zone of apposition.

2 years and 7 years, respectively, after the initial repair. All 3 patients are doing well at a median interval of 13 years after the initial surgery (5–15 years).

Unbalanced ventricles ($P = 0.045$), subvalvular abnormalities ($P = 0.0037$) and grade >2 LAVV postoperative regurgitation ($P = 0.017$) were identified as risk factors for LAVV reoperations in patients with DO LAVV (Table 4).

Long-term overall freedom from LAVV reoperation, freedom from late LAVV reoperation and freedom from LAVV MVR were 80% [95% confidence interval (CI) 70–100], 87% (95% CI 79–100) and 85% (95% CI 75–100), respectively (Fig. 2B–D).

Subvalvular abnormalities were identified as a risk factor for MVR ($P = 0.010$).

Anterior hemivalve pericardial enlargement

Four patients, 2 of our cohort and 2 patients operated on by our surgical team in another institution, required an anterior hemivalve pericardial patch enlargement. This technique was performed during the initial repair in 2 patients and when the LAVV reoperation was required in the other 2 patients (1 early reoperation 3 days after initial repair and 1 late repair 3 years after surgery). Patient characteristics are described in Table 5. There were 3 partial AVSD and 1 intermediate AVSD. Two patients had a single papillary muscle.

All operations were uneventful. Apart from the patient who underwent a reoperation 3 days after the initial repair and had to remain in the intensive care unit (ICU) for 5 days, the other 3 patients were weaned from mechanical ventilation and inotropic support in the first hours after surgery (h 2–h 4) and were discharged from the ICU the day after surgery with a mean hospital stay of 10 days.

At the last follow-up (1–7 years), all patients are doing well, with none-to-trivial LAVV regurgitation and no LAVV stenosis.

DISCUSSION

Atrioventricular septal defects and double-orifice left atrioventricular valve

Survival. The presence of a DO LAVV has often been incriminated as putting the patient at risk for LAVV dysfunction and reoperation after AVSD repair and even for death after reoperation [6–12]. Only a few publications describe the outcome of patients with AVSD with LAVV DO. Hoohenkerk *et al.* [4] reported their experience with 21 patients: no early deaths and 3 late deaths with an overall survival of 84% at 15 years. Although the mortality rate was high (14%), survival did not significantly differ between patients with AVSD with and without a DO-LAVV. Sharma *et al.* [5] had similar findings, with a 15-year survival rate of 86%. We report a higher survival rate, over 90% long-term. The overall mortality rate has dramatically dropped since 2000 and now appears to be close to that of standard AVSD without associated lesions. We are strongly convinced that the decrease in the mortality rate is directly correlated with the achievement of an optimal repair.

Table 4: Univariable analysis of risk factors for left atrioventricular valve reoperation in patients with atrioventricular septal defect and direct orifice left atrioventricular valve

Risks factors for LAVV reoperation	*P*-value
Preoperative factors	
AVSD type	1.0
Down syndrome	1.0
Preoperative LAVV regurgitation grade >2	1.0
Unbalanced ventricles	0.045
Subvalvular apparatus abnormalities	0.004
Perioperative factors	
Surgery before 2000	1.0
ZoA closure	1.0
Postoperative factors	
Postoperative LAVV regurgitation grade >2	0.017

Unbalanced ventricles, subvalvular abnormalities and postoperative regurgitation were identified as risk factors for LAVV reoperations.
AVSD: atrioventricular septal defect; LAVV: left atrioventricular valve; ZoA: zone of apposition.

Management of the double orifice. The Mayo Clinic team suggested the following management for DO-LAVV: do not divide the tissue bridge; cleft closure must be achieved except when the valve area is small, the mural leaflet is dysplastic and/or a single papillary muscle is present; a competent DO must be conserved but closed with a patch if regurgitant; if the DO must be closed, the ZoA should not be completely closed [5]. We agree with these recommendations even though some details need to be clarified. Respect for the bridging tissue valve and ZoA closure are the 2 key determinants for an easy and effective repair. Nevertheless, ZoA closure can be problematic in specific situations. If the residual orifice is too small, organic stenosis can be encountered, especially when there is a single papillary muscle or an underdeveloped mural leaflet. It is always difficult to appreciate accurately the functional residual orifice, although we can estimate the inflow by adding the surface area of each orifice. Calibration of the residual orifice can lead to tears in the repair, and the functional orifice is also correlated with the anatomy of the subvalvular apparatus, which can create a restriction. We cannot say that, in our experience, none of the ZoA have been left open in the presence of a single papillary muscle or dysplastic mural leaflet, but clearly, it is a complex repair, and partial closure, no closure or primitive patch augmentation has to be considered.

Surgical management of the ZoA remains the challenge of the AVSD repair, because incomplete ZoA closure has been reported in previous publications to be associated with poor outcome, death and reoperations [6, 13, 14]. Association with abnormalities of the subvalvular apparatus, especially a single papillary muscle, can complicate surgical repair and make one reconsider the initially planned surgical management of the LAVV. In our series, abnormalities of the subvalvular apparatus have been identified as a risk factor for both LAVV reoperations and MVR. Definitively, transoesophageal echocardiography remains mandatory in this situation (accepted mean gradient up to 5 mmHg).

Table 5: Characteristics of patients who required hemivalve pericardial patch enlargement

Patient number	Diagnosis	Age at initial repair	Indication for patch enlargement	Preoperative LAVV regurgitation	Last follow-up results
1	pAVSD; posterior DO; single papillary muscle	1.2 years	LAVV re-repair 3 days after initial repair Initial repair: partial ZoA closure Reoperation: left hemivalve patch enlargement; ZoA left open; commissuroplasty	Grade 2 (initial repair)/grade 4 (reoperation)	At 7 years: grade 1 regurgitation; no LAVV stenosis; sports in elementary school
2	iAVSD; posterior DO	4.5 years	Initial repair: left hemivalve patch enlargement; partial ZoA closure	Grade 3	At 3 years: no regurgitation; mean gradient 2 mmHg
3	pAVSD; anterior DO	9 months	Initial repair: left hemivalve patch enlargement; ZoA left open	Grade 1	At 1 year: no regurgitation; mean gradient 2 mmHg
4	pAVSD; single papillary muscle	1.1 years	LAVV re-repair 3 years after initial repair Initial repair: ZoA partial closure, single papillary muscle splitting, section of accessory chordae Reoperation: Left hemivalve patch enlargement, partial ZoA closure, commissuroplasty	No regurgitation (initial repair)/grade 4, no stenosis (reoperation)	At 1 year after reoperation: trivial LAVV regurgitation; mean gradient 7 mmHg

AVSD: atrioventricular septal defect; DO: double orifice; iAVSD: intermediate AVSD; LAVV: left atrioventricular valve; pAVSD: partial AVSD; ZoA: zone of apposition.

Reoperations for left atrioventricular valve dysfunction. Postoperative LAVV dysfunction is the main indication for AVSD reoperations [9, 15, 16]. Risk factors such as preoperative LAVV regurgitation, unbalanced ventricles, incomplete cleft closure, DO-LAVV and the absence of Down syndrome have been reported in the literature [4, 6, 7, 14, 17–20]. When reoperation is required, one should always try to re-repair the LAVV because MVR still remains associated with a worse outcome than valvuloplasty in patients with AVSD [6, 12, 21].

Freedom from LAVV reoperation in our cohort goes along with previously reported outcomes after AVSD and DO-LAVV repair [4, 5]. We also confirmed the bad reputations of postoperative LAVV regurgitation, unbalanced ventricles and subvalvular apparatus abnormalities. Although early MVR in 1 patient led to death, the other 3 patients who required late MVR are still alive after a median follow-up of 13 years. Evolution in surgical strategies, postoperative management of the patients with AVSD in the ICU and improvements in the supervision of anticoagulation therapy decreased the morbidity and mortality rates of paediatric patients with mechanical valves, but valve repair has to be the first choice and, most of the time, can be achieved in expert hands.

Pericardial patch enlargement as an alternate surgical strategy

Different surgical strategies can be used for LAVV re-repair in AVSD: complete cleft closure; cleft patch augmentation with bovine or pericardial patch or via the use of a mitral valve homograft; leaflet augmentation technique; and Gore-Tex artificial chordae placement [22–24].

As described in this manuscript and by others, we initially used pericardial patch bridging leaflet extension in AVSD with DO LAVV in re-repair for residual regurgitation with an excellent early outcome. Secondly, this technique has been considered at the first repair when ZoA closure was ineffective (residual regurgitation or estimated low coaptation surface height) or obviously stenotic. These situations were particularly encountered in cases of subvalvular apparatus abnormalities, underdeveloped mural leaflet and, rarely, asymmetric bridging leaflets. We are strongly convinced that this technique can be helpful in patients in whom competence is difficult to achieve without stenosis. Because leaflet extension does not meet the surface of coaptation, we expect the repair to last and to avoid the adverse outcomes of partial or no closure.

CONCLUSION

Repair of AVSD and DO LAVV is every paediatric cardiac surgeon's challenge. Preoperative LAVV regurgitation is not reliable for predicting results of surgery whereas significant postoperative LAVV regurgitation was identified as a risk factor for reoperation but not for survival, as were unbalanced ventricles and abnormalities of the subvalvular apparatus.

LAVV hemivalve pericardial extension appears to be a useful and effective alternate surgical strategy when leaflets and subvalvular apparatus appear to be deficient and the ZoA cannot be closed.

Conflict of interest: none declared.

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