Long-Term Outcome of Atrioventricular Septal Defect Surgical Repair: Impact of Down Syndrome

Résultats À Long Terme De La Réparation Chirurgicale Du Canal Atrioventriculaire : Impact Du Syndrome De Down

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Summary

Introduction : Given the strong association between the atrioventricular septal defect (AVSD) and the Down syndrome (DS), it is impossible to discuss surgical repair of an AVSD without also taking into account the issues raised by the presence of DS. The primary endpoint of this study was to compare the long-term results of surgical repair of the AVSD in patients with and without DS.

Methods and results: 200 patients who underwent surgical biventricular repair of AVSD between 1995 and 2020 were included in our study. 49.5% of patients had DS (87.9% with compete AVSD). Concerning the complete AVSD form, left atrioventricular valvar (LAVV) dysplasia but also common atrium were significantly less frequent in patients with DS (p=0.022 and p=0.012 respectively). Actuarial survival was 83.9% among patients with DS versus 89.3% of the group of patients without DS (p=0.365). LAVV reintervention were more frequent in the group of patients without DS (p=0.034). Regarding the partial AVSD form, there was no significant correlations between DS and any anatomic characteristics. DS was not associated with higher mortality or reinterventions.

Conclusions: The presence of DS does not represent a risk factor for late mortality after biventricular repair of AVSD. Furthermore, DS appears to be a protective factor for long-term reintervention on the LAVV in patients with complete AVSD. An accurate pre-operative assessment of pulmonary pressures is essential to ensure the best outcomes.

Keywords

Atrioventricular septal defect; Trisomy 21; surgery; long-term; survival

Résumé

Introduction : Compte tenu de la forte association entre le canal atrioventriculaire (CAV) et le syndrome de Down (SD), il est impossible de considérer la réparation chirurgicale du CAV sans prendre en compte les enjeux soulevés par la présence d'un SD.

L'objectif principal de cette étude était de comparer les résultats à long terme de la réparation chirurgicale du CAV chez les patients avec et sans SD.

Méthodes et résultats : 200 patients ayant bénéficié d'une réparation chirurgicale bi-ventriculaire du CAV entre 1995 et 2020 ont été inclus dans notre étude. 49,5% des patients avaient un SD (87,9% avec un CAV complet). Concernant la forme complète du CAV, la dysplasie de la valve auriculo-ventriculaire gauche (VAVG) mais aussi l'oreillette commune étaient significativement moins fréquentes chez les patients avec SD (p=0,022 et p=0,012 respectivement). La survie actuarielle était de 83,9% chez les patients avec SD contre 89,3% dans le groupe des patients sans SD (p=0,365). Les réinterventions de la VAVG étaient plus fréquentes dans le groupe de patients sans SD (p=0.034). En ce qui concerne la forme partielle du CAV, il n'y avait pas de corrélation significative entre le SD et les caractéristiques anatomiques du CAV. Le SD n'était pas associé à une mortalité ou à un taux de réintervention plus élevé.

Conclusions : La présence d'un SD ne représente pas un facteur de risque de mortalité tardive après une réparation bi-ventriculaire d'un CAV. En outre, le SD semble être un facteur protecteur de la réintervention à long terme de la VAVG chez les patients présentant un CAV complet. Une évaluation préopératoire minutieuse des pressions pulmonaires est essentielle pour garantir les meilleurs résultats.

Mots-clés

Canal atrioventriculaire; trisomie 21; chirurgie; survie à long terme.

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INTRODUCTION

The prevalence of AVSD is 0.19 per 1000 live births [1]. It accounted for 2.9% of patients with congenital heart defects [1]. The association between AVSD and Down syndrome (DS) is frequent: The AVSD is the most common congenital heart disease in patients with DS [1,2].

If left unrepaired, a complete AVSD can result in pulmonary arterial hypertension and irreversible alterations in pulmonary vascular resistance. The results of surgical repair of AVSD are generally good with a 15 year-survival rate exceeding 85% [3] . Long-term complications may occur, mainly due to the risk of progressive left atrioventricular valve (LAVV) regurgitation or left ventricular outflow tract obstruction (LVOTO) requiring reoperation on one hand, and pulmonary arterial hypertension, on the other hand. It is impossible to discuss surgical repair of an AVSD without also taking into account the issues raised by the presence of DS. There are two main concerns. First, it's frequently reported that pulmonary hypertension is more common in children with DS. Second, in comparison to their chromosomally normal peers, children with Down syndrome have a different anatomic spectrum.

The primary endpoint of this study was to compare the long-term results of surgical repair of the AVSD in patients with and without Down syndrome, in terms of survival, re-interventions on the mitral valve and pulmonary hypertension. The secondary endpoints were to compare the clinical and anatomical characteristics of the AVSD in the two groups.

MATERIALS & METHODS

We conducted a single-center, retrospective and descriptive study including patients with or without DS who underwent surgical repair of AVSD, confirmed per-operatively between 1995 and 2020. They were not included in this study: patients with associated tetralogy of Fallot or other complex congenital heart disease, and patients with unbalanced AVSD who required uni-ventricular palliative surgery. Patients lost to follow-up were excluded from this study.

The patients were divided into two groups according to the AVSD type: Patients with complete AVSD defined by the presence of a common atrioventricular valve per-operatively, and patients with partial AVSD defined by the presence of separate right and left atrioventricular orifices within the common junction [1]. For each AVSD type, patients were grouped into patients with DS and patients without DS. The diagnosis of DS was made in the presence of an obvious facial dysmorphia (hyperthelorism with slanting eyes and macroglossia) and/or a Trisomy 21 karyotype.

Demographic, clinical, echocardiographic, hemodynamic, and operative data were collected for all patients from hospital records. The pulmonary artery banding (PAB) prior surgical repair was performed in the presence of poor clinical tolerance with a low weight of less than 4.5 kg or coexisting coarctation. Cardiac catheterization was performed before surgical correction to evaluate systemic and pulmonary pressure, resistance, and blood flow in case the age of patients with complete AVSD and without prior palliative surgery exceeded 8 months or in case of suspicion of Eisenmenger syndrome.

Long-term mortality was defined by mortality beyond the first postoperative month and/or during the same postoperative hospitalization. LAVV reinterventions were reported with details of the timing after the surgical repair and the surgical procedure performed. Latest clinical and echocardiographic findings were documented. The assessment of the severity of LAVV regurgitation was semi-quantitative graded from 1 to 4. Signs of pulmonary artery hypertension were investigated according to the 2022 ESC guidelines [4], with high echocardiographic probability based on a peak tricuspid regurgitation velocity > 3.4m/s, or a peak tricuspid regurgitation velocity between 2.9 and 3.4 m/s in the presence of at least two of the additional echocardiographic signs suggestive of PH. Right heart catheterization with the threshold of mean pulmonary artery pressure > 20 mmHg was proposed to confirm the diagnosis when there was a need for pulmonary hypertension treatment.

Statistical analysis

Statistics were analyzed using the Statistical Package for the Social Sciences software (version 25 for Windows; SPSS). Qualitative variables were expressed by their frequencies and percentages. Quantitative variables were expressed by their means or medians, with the standard deviations or interquartile range as appropriate.

In order to compare the two groups of patients (patients with DS and patients without DS), the following parametric and non-parametric tests were used: t-test or Mann-Whitney test to compare quantitative variables and Chi Square or Fisher's exact to compare qualitative variables.

A significance cut-off of p strictly less than 0.05 was used.

RESULTS

200 patients meeting the inclusion criteria underwent surgical repair of AVSD during the study period, of which 99 patients (49,5%) had DS, 87 patients (87,9%) with compete AVSD and 12 patients (12.1%) with partial AVSD. In patients without DS, 55.4% of patients had the complete type of AVSD. Our study revealed significant association between the complete AVSD type and DS (p-value <0.001) (Table 1). Antenatal diagnosis was made in seven patients: four patients with DS and complete AVSD, a patient with DS and partial AVSD; the other two patients didn't have DS, with the complete type in one patient and the partial type in the other one. For patients with complete AVSD as well for patients with partial AVSD, DS patients were non-significantly younger than patients with normal karyotype (Table 1). There was a female predominance in our study with a sex-ratio of 0.8 (Table 1). Parental consanguinity was found in 60 patients (30.8%). There was no statically significant relationship between the parental consanguinity and the type of AVSD (p= 0.841) nor with the presence of DS (p= 0.633).

Table 1. Different types of AVSD and Patient demographic data in our study

	Patients with Down syndrome (n=99)	Patients without Down syndrome (n=101)	p Value
AVSD type, n (%) - Complete AVSD (%) - Partial AVSD	87 (87.9) 12 (12.1)	56 (55.4) 45 (44.6)	<0.001
Diagnosis age in months, Mean ±SD - Complete AVSD - Partial AVSD	4.21 ±4.614 24.09 ±37.676	6.65 ±14.291 57.12 ±67.074	0.497 0.124
Number of males, n (%)	48 (48,5)	41 (40.5)	0.253
Parental consanguinity, n (%)	28 (29.2%)	32 (32.3%)	0.633

AVSD: Atrioventricular septal defect; SD: standard deviation.

The complete avsd type

Preoperative findings and surgical data: (table 2)

For patients with complete AVSD, shortness of breath was the most common symptom in patients with and without DS. Feeding difficulties was significantly higher in patients with DS than in patients without DS. Signs of heart failure were present in 9.2% and 12.5% of patients with and without Down syndrome respectively with no significant difference between the two groups (p=0.529).

The PAB was performed in 9 patients (6.3%) prior to the surgical repair. The indications were poor tolerability with low weight in 8 patients and coexisting coarctation in a patient.

Transthoracic echocardiography (TTE) revealed a good systolic left ventricle function in both groups of patients with and without DS. Our study didn't reveal a significant difference of the overall associated cardiac anomalies between the groups of patients with and without DS (p=0.265). However, when analyzing the subgroups of associated cardiac defects, common atrium was significantly higher in the group of patients without DS. Sub-aortic stenosis was present in a patient with DS, with no significant difference between the two groups (p= 0.421). Dysplastic LAVV was significantly more important in patients without DS (p=0.022), with severe LAVV regurgitation significantly higher in this group of patients (p= 0.011). Pulmonary hypertension was higher, but with no significant difference, in the group of patients with DS (p=0.062). Fourteen patients (9.8%) underwent cardiac catheterization before surgical repair (DS: 8 patients (9.2%) versus No DS: 6 patients (10.7%)). There was no significant difference in pulmonary vascular resistance between the two groups (p=0.603).

All patients underwent surgical repair of the complete AVSD. No significant difference was found in regard the mean age of repair (p=0.816) and weight at surgical repair (0.454) in the group of patients with and without DS. The surgical procedure consisting of the double-patch technique were more used in the group of patients with DS (p <0.001). There was no significant difference between the two groups of patients concerning the associated LAVV repair (p=0.290). LAVV replacement was performed in one patient without DS, aged 3-year-old who underwent PAB prior to the surgical repair, with severe dysplastic AV valve. Additional surgical procedures at repair included debanding in 9 patients, closure of PDA in 12 patients, resection of sub-aortic stenosis in one patient, and closure of additional muscular ventricle septal defect in two patients.

Table 2. Preoperative findings and surgical data of patients with complete AVSD type in our study

	Patients without DS (n=56)	Patients without DS (n=56)	p Value
Symptoms, n (%)			
- Shortness of breath	63 (72.4)	32 (57.1)	0.059
- Feeding difficulties	57 (65.5)	25 (44.6)	0.014
- Recurrent respiratory infections	54 (62.1)	33 (58.9)	0.707
- Growth failure	50 (57.5)	25 (44.6)	0.134
Signs of heart failure, n (%)	8 (9.2)	7 (12.5)	0.529
TTE findings:			
- LV fractional shortening, Mean ±SD	45.02 ±7.409	44.29 ±7.962	0.633
- Dysplastic LAVV, n (%)	15 (17.2)	19 (33.9)	0.022
- LAVV regurgitation Grade III-IV, n (%)	6 (6.9)	15 (26.8)	0.011
- PASP, Mean ±SD	63.93 ±18.262	57.57 ±20.662	0.062
- Associated abnormalities, n (%)			0.265
• Other trabecular VSD	3 (3.4)	0 (0)	0.168
• Sub-aortic stenosis	1 (1.1)	0(0)	0.421
• ASD type OS	16 (18.4)	4 (7.1)	0.064
• Common atrium	0 (0)	4 (7.1)	0.012
• Aortic coarctation	0 (0)	1 (1.8)	0.211
• PDA	12 (13.8)	5 (8.9)	0.380
PAB prior to surgical repair, n (%)	8 (9.2)	1 (1.8)	0.075
Age at surgical repair in months, Mean ±SD	18 ±26.850	19.66 ±20.575	0.816
Weight at surgical repair in Kg, Mean ±SD	6.989 ±3.817	8.035 ±3.609	0.454
Surgical procedure:			<0.001
- Single-patch technique	36 (41.4)	44 (78.6)	
- Double-patch technique	51 (58.6)	12 (21.4)	
LAVV repair	5 (5.7)	7 (12.1)	0.290
LAVV replacement	0 (0)	1 (1.8)	0.211
RAVV repair	0 (0)	1 (1.8)	0.211

ASD: atrial septal defect, DS: Down syndrome, LAVV: left atrioventricular valve, LV: left ventricle, OS: ostium secundum, PAB: pulmonary artery banding, PASP: pulmonary artery systolic pressure, PDA: patent ductus arteriosus, RAVV: right atrioventricular valve, SD: standard deviation, VSD: ventricle septal defect, TTE: transthoracic echocardiography.

Post-operative results and long-term follow-up (long-term follow-up data are summarized in table 3).

Fifteen patients (10.5%) died in the early postoperative period, with no statistically significant difference between the two groups of patients with and without DS (p=0.295). The main causes of death were pulmonary hypertensive crises, infective endocarditis, septic shock, and severe heart failure. Reintervention for severe LAVV regurgitation was performed in 6 patients (4.7%) during this period with no difference between the two groups (p=0.388). LAVV repair was the procedure performed in all these patients. Pulmonary hypertension was documented in 29 patients (22.7%), with no difference between the two groups of patients (p=0.248). Pulmonary hypertensive crises were noted in fourteen and in four patients with and without Down syndrome respectively (p=0.115). Complete atrioventricular block was objected in 12 patients. It was more frequent in patients without Down syndrome (p=0.041), but indication for permanent pacemaker implantation was only made in five patients with no significant difference between the two groups.

For patients who survived the early postoperative period, the mean follow-up period in our study was 10.63 ±59 years. Late mortality was documented in five patients (3.9%) with an 8.3±54-year delay following the surgical repair. There was no difference between the 2 groups of patients with and without DS (p=0.977). For patients with DS, the causes of death were: Pulmonary hypertension, severe mitral regurgitation complicated with LV dysfunction, and an extracardiac cause. For the patient who died because of the pulmonary hypertension, he underwent a PAB at the age of four months for severe congestive heart failure and growth failure. Three years later, he underwent the surgical repair without cardiac catheterization before the surgery. Pulmonary hypertension was confirmed with cardiac catheterization and the patient died two years following the surgical repair. Considering patients without DS, severe LAVV regurgitation complicated with left ventricle dysfunction was the cause of death in both patients. Actuarial survival was 83.9% among patients with DS versus 89.3% of the group of patients without DS (p=0.365).

Long-term reintervention on the LAVV (2.4%) was statistically more frequent in the group of patients without DS (p= 0.034). The different procedures were: a mechanical mitral valve replacement 11-year following the surgical procedure in a patient who underwent mitral valve repair in the early postoperative period; and LAVV repair in two patients 2 and 8 years following the surgical repair. Subaortic obstruction relief was performed in two patients without DS five and eight years following the surgical repair respectively. Despite pulmonary hypertension being the cause of death in a patient with complete AVSD and DS, there was no significant difference in long-term pulmonary hypertension frequency between the two groups of patients with and without DS (p=0.589). At last check-up, 110 patients (89.4%) were asymptomatic (p=0.127). The most frequent symptom was dyspnea stade II NHYA in 10 patients (8.1%). Last TTE check-up revealed that LAVV regurgitation was present in 84 patients (68.3%), and it was more frequent in the

group of patients without Down syndrome (p <0.001). However, severe LAVV regurgitation was documented in seven patients (8.3%) with no significant difference between the two groups of patients (p=0.263).

Table 3. Long-term follow-up in patients with repaired complete AVSD

	Patients with DS (n=76)	Patients without DS (n=52)	p Value
Mortality, n (%)	3 (3.9)	2 (3.8)	0.977
Reinterventions on the LAVV, n (%)	0 (0)	3 (5.8)	0.034
Reinterventions on the LVOTO, n (%)	0 (0)	2 (4)	0.085
Pulmonary hypertension, n (%)	8 (10.5)	4 (7.7)	0.589
Mitral regurgitation, n (%)	41 (56.2)	43 (86)	<0.001

DS: Down syndrome, LAVV: left atrioventricular valve, LVOTO: left ventricle outflow tract obstruction.

The partial avsd type

Preoperative findings and surgical data: (table 4)

For the partial AVSD type, recurrent respiratory infections were the most common symptom in patients with and without DS. There was no significant difference in symptoms between patients with and without DS. Signs of heart failure were absent in all patients with Down syndrome and present in two patients without Down syndrome. These two patients had severe LAVV regurgitation. Pre-operative TTE revealed good systolic left ventricle function in both groups of patients with and without DS. Dysplastic LAVV and severe LAVV regurgitation were higher in the group of patients without DS, but without significant statistically difference (p=0.124 and p=0.063 respectively). There was also no significant difference of associated cardiac abnormalities in the two groups of patients with and without DS (p=0.602). Pulmonary hypertension was comparable in these two groups of patients (p=0.460). Cardiac catheterization was performed in only one patient with DS. There was no statically difference of the age and weight at surgical repair between the two groups of patients (p=0.230 and p= 0.305 respectively). The single-patch surgical technique was the approach used in all patients with partial AVSD. Closure of the LAVV cleft was performed in almost all patients with and without DS. There was no significant difference between the two groups of patients concerning the associated LAVV repair during the surgical correction.

Table 1. Different types of AVSD and Patient demographic data in our study

	Patients with DS (n=12)	Patients without DS (n=45)	p Value
Symptoms, n (%)			
- Shortness of breath	3 (25)	14 (31.1)	0.351
- Feeding difficulties	3 (25)	7 (16.3)	0.880
- Recurrent respiratory infections	4 (33.3)	21 (48.8)	0.199
- Growth failure	4 (33.3)	13 (30.2)	0.848
- No symptoms	4 (33.3)	14 (32.6)	0.811
TTE findings:			
- LV fractional shortening, Mean ±SD	43 ±10.231	41.28 ±7.275	0.607
- Dysplastic LAVVV, n (%)	1 (8.3)	13 (30.2)	0.124
- LAVV regurgitation Grade III-IV, n (%)	0 (0)	9 (20)	0.063
- PASP, Mean ±SD	44.58 ±18.522	40.33 ±11.942	0.460
- Associated abnormalities, n (%)			0.602
• Trabecular VSD	0 (0)	1 (2.2)	0.608
• ASD type OS	1 (8.3)	2 (4.44)	0.680
• Common atrium	1 (8.3)	7 (15.6)	0.522
• PDA	0 (0	1 (2.3)	0.598
Age at surgical repair in months, Mean ±SD	49.9 ±41.562	76.58 ±66.127	0.230
Weight at surgical repair in Kg, Mean ±SD	14.6 ±7.756	20.81 ±14.12	0.305
Surgical procedure			
- Single-patch technique	12 (100)	45 (100)	
Closure of the left AV valve cleft (%)	12 (100)	43 (95.6)	0.487
Parental consanguinity, n (%)	28 (29.2%)	32 (32.3%)	0.633

ASD: atrial septal defect, DS: Down syndrome, LAVV: left atrioventricular valve, LV: left ventricle, OS: ostium secundum, PASP: pulmonary artery systolic pressure, PDA: patent ductus arteriosus, SD: standard deviation, VSD: ventricle septal defect, TTE: transthoracic echocardiography.

Post-operative results and long-term follow-up (long-term follow-up data are summarized in table 5).

There was no mortality in the early postoperative period in both groups of patients with and without DS. Reoperation for severe LAVV regurgitation was performed in three patients (5.3%) prior to hospital discharge with no significant difference between the patients with and without DS (p=0.592). Pulmonary hypertension was documented in six patients (10.5%), with no significant difference between the two groups. Pulmonary hypertension crises were documented in one patient with DS. Complete atrioventricular block was objected in five patients without Down syndrome (p=0.227), but there was no indication for permanent pacemaker implantation in any of them.

Long-term mortality in patients with partial AVSD was documented in two patients (3.5%) at a 16.5-year delay

following the surgical repair. There was no significant difference between the two groups of patients with and without DS (p=0.457). Both of the patients didn't have DS. The cause of death was severe LAVV regurgitation in both cases: one patient died post mitral valve replacement reintervention and the other one died because of severe LAVV regurgitation complicated with left ventricle dysfunction. Actuarial survival was 100% among patients DS versus 95.6% of the group of patients without DS (p= 0.457).

Reinterventions on the LAVV were performed in two patients without DS with favorable surgical outcome (p=0.452). The first patient underwent mitral valve replacement for LAVV stenosis five years following the surgical repair. The second patient underwent mitral valve replacement for severe LAVV regurgitation 12 years following the surgical repair. There was no significant difference in long-term pulmonary hypertension frequency between the two groups of patients with and without DS (p= 0.457). At last check-up, 51 patients (92.7%) were asymptomatic (p=0.614). Dyspnea stade II NYHA was the most frequent symptom noted in three patients (5.5%).

Last TTE check-up revealed the presence of LAVV regurgitation in 39 patients (70.9%), with no difference between the two groups of patients with and without DS (p=0.714). Severe LAVV regurgitation was identified in three patients without DS (p=0.224).

Table 5. Long-term follow-up in patients with repaired partial AVSD

	Patients with DS (n=12)	Patients without DS (n=45)	p Value
Mortality, n (%)	0 (0)	2 (4.4)	0.457
Reinterventions on the LAVV, n (%)	0 (0)	2 (4.5)	0.452
Pulmonary hypertension, n (%)	0 (0)	2 (4.4)	0.457
LAVV regurgitation, n (%)	8 (66.7)	31 (72.1)	0.714

DS: Down syndrome, LAVV: left atrioventricular valve.

DISCUSSION

A common atrioventricular junction together with a deficient atrioventricular septation are the anatomic hallmark of the AVSD. There is a common atrioventricular junction whether the valve itself is a common structure, traditionally described as the complete form of the AVSD, or whether there are separate atrioventricular valvar orifices, often referred to as the partial form or ostium primum defect [1]. The relationship between the valvar leaflets defines the type of the AVSD based on the arrangement of the two bridging leaflets. The common atrioventricular valvar orifice (the complete form) is the most common form, characterized by the presence of a space between the two bridging leaflets. When there is a connecting tongue of valvar tissue attaching the bridging leaflets to one another, the atrioventricular junction is divided into right and left orifices within the common atrioventricular junction (the partial form) [1]. The complete AVSD form represented 71.5% and 63.4% of patients in our study and Sarisoy et al work [5] respectively. In the majority of studies, a slight female predominance was noted [5–7], as in the case of our study where the sex-ratio was 0.8. The AVSD is strongly associated with DS. An AVSD was documented in 45% of DS infants having congenital heart defects [8]. Other studies demonstrated that DS was present in about half of individuals with AVSD[5,9,10]. Our study confirmed these findings, showing that 49.5% of our patients had DS. Complete AVSD is the common form in DS patients [5]. 60.8% and 71.6% of patients with complete AVSD had DS in our study and in Lange et al study [11] respectively.

The Complete AVSD form

Anatomical characteristics of complete AVSD in the presence of DS include essentially the presence of less LAVV dysplasia [11,12]. Our study confirmed these findings. Overall minor associated cardiac anomalies were present with no significant difference in both groups of patients with and without DS in our study and in Lange et al study [11]. However, when analyzing the subgroups of these cardiac anomalies, the common atrium was significantly higher in the group of patients without DS in these two studies. Left ventricle obstructive lesions were not significantly more frequent in the group of patients without DS in our study.

If left uncorrected, a complete AVSD can result in pulmonary arterial hypertension and irreversible alterations in pulmonary vascular resistance. Pulmonary vascular disease is usually noted at around six months of age in infants with complete AVSD [13]. In children with DS, these alterations may be more pronounced and start earlier in life [13,14]. For patients aged less than 6 months, pulmonary vascular resistance was significantly

higher in DS patients compared to patients without DS [11]. In our study, Pulmonary hypertension was not significantly higher in the group of patients with DS on TTE assessment. Only 9.8% of patients with complete AVSD underwent cardiac catheterization in our study, and there was also no statistically significant difference in pulmonary vascular resistance between the two groups of patients with and without DS. Our findings may be partially explained by the fact that some patients have undergone a PAB at the time of the evaluation.

To prevent irreversible modifications of pulmonary vascular disease, early diagnosis and repair are mandatory. The PAB, a palliation surgery to delay surgical correction beyond the first year of life, is nowadays abandoned in the majority of cardiac centers in favor of elective surgical repair early in life. A pulmonary artery banding was performed before surgical repair in 6.3% and in 20% of patients with a complete AVSD in our study and in Lange et al study [11] respectively. The higher rate of this surgical palliation in the latter study is probably because it started earlier in 1974. The older mean age at surgical repair in our study (18±26 in patients with DS versus 19±20 in patients without DS) is probably due to the fact that some patients underwent a PAD which delayed the surgical repair for years.

Like most studies [5,11,15], our study revealed that there were no significant differences in early mortality between DS and non-DS patients.

The impact of DS on late mortality in patients with complete AVSD was controversial. According to the Reller and Morris study [16], late mortality was significantly higher in the presence of DS. In contrast, in most studies [11,17–19] as in ours, the presence of DS was not associated with more significant mortality. Even when associated Major cardiac anomalies (unbalanced AVSD, Tetralogy of Fallot, Heterotaxy, or any other complex congenital heart defect) were included in the Miller et al study, DS was not a risk factor for mortality [17].

The main cause of reintervention after surgical repair of complete AVSD is severe LAVV regurgitation. As in Tumanyan et al work [12], long-term reinterventions on the LAVV was rarer in DS patients in our study. Freedom from reintervention was also lower in the group of patients without DS in the Formigari et al [19] and in Lange et al [11] studies. LAVV repair is generally possible. However, some patients require valve replacement. In our study, reinterventions on LAVV were less frequent in the long-term follow-up than in the early postoperative period. In contrast to Formigani and coworkers [19], our study didn't describe a higher prevalence of reoperation for left ventricular outflow tract obstruction in patients without DS.

Despite the absence of a significant difference in the age of patients with and without DS at the time of surgical repair, our study did not demonstrate a higher frequency of pulmonary hypertension in patients with DS at long-term follow-up. Beyond the age of 6 months, cardiac catheterization remains the preferred method for calculating pulmonary vascular resistance and making the decision of surgical repair. In our study, only one patient died because of pulmonary hypertension at long-term follow-up, and he didn't undergo cardiac catheterization before the surgery because the clinicians though that the PAB was efficient. It is pulmonary vascular resistance, itself related to age, and other associated problems that determine the risk for these patients [11].

The partial AVSD form

Partial AVSD accounted for only 6% of patients with Down syndrome who underwent surgery for a congenital heart defect, according to a national clinical database [2]. Strong association between partial AVSD and DS was not established. In our study, only 12.1% of patients with DS had DS. Our study did not reveal any significant association between the presence of DS and this form of AVSD. DS was not associated with higher long-term mortality or reinterventions. Actuarial survival among patients with DS was excellent achieving 100%.

CONCLUSION

The presence of Down syndrome does not represent a risk factor for late mortality after biventricular repair of complete AVSD. Furthermore, due to the less frequency of LAVV dysplasia in this group of patients, DS appears to be a protective factor for long-term surgical reintervention on the LAVV in patients with complete AVSD. An accurate pre-operative assessment of pulmonary pressures is

essential to ensure the best outcomes.

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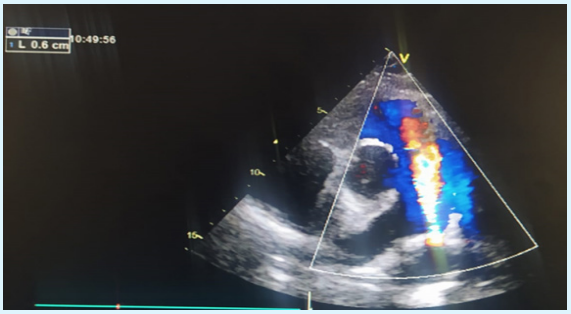

Figure 1. Ductus arteriosus 6 mm wide in a 42-year-old woman

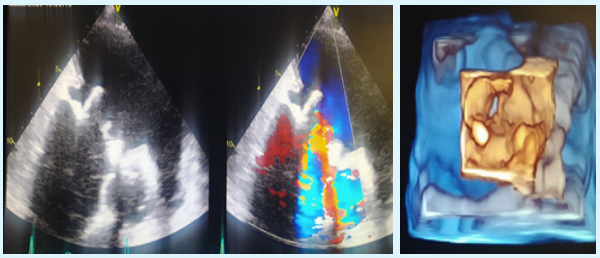

Figure 2. Ebsteine's disease with degeneration of the tricuspid prosthesis which has become stenosing and leaking

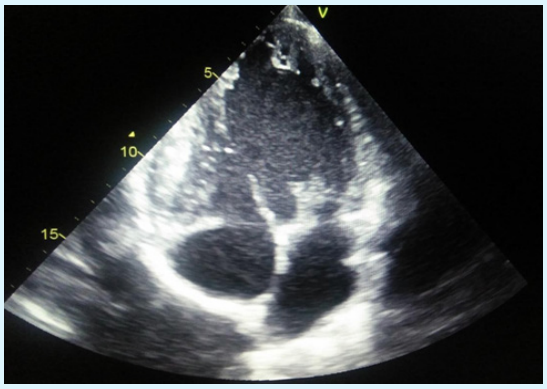

Figure 3. Double-inlet single ventricle

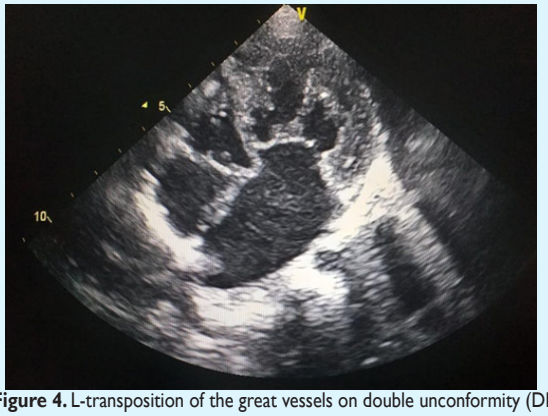

Figure 4. L-transposition of the great vessels on double unconformity (DD)