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Doubly Committed Ventricular Septal Defect: Single-Centre Experience and Midterm Follow-Up

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Key Words
Aortic valve regurgitation · Aortic valve prolapse · Ventricular septal defect · Doubly committed ventricular septal defect

Abstract
Background: Doubly committed ventricular septal defect (dcVSD) is the least common type of VSD. Because published studies are rather scarce, this study aimed at evaluating the midterm outcome of dcVSDs. ***Methods:*** The records of all patients registered in the database of Paediatric and Congenital Cardiology, University Hospitals Leuven, with a dcVSD at 16 years of age were reviewed. Clinical, electrocardiographic and transthoracic echocardiographic changes from baseline, defined as of the age of 16 years, until the latest follow-up were compared. ***Results:*** Thirty-three patients (20 males, median age 26 years, interquartile range 12) were followed for a median time of 7.9 years (interquartile range 9.8, time range 2–25.9). No deaths occurred. In 15 patients (45%), the defect remained patent at baseline. During follow-up, two spontaneous closures (13%) occurred. Eighteen patients (55%) required closure before the age of 16 years. Five (28%) needed reoperation. In the dcVSD closure group, left ventricular ejection fraction decreased from 69 ± 12 to 61 ± 6% (p = 0.028). No significant changes in pulmonary arterial hypertension were noticed. ***Conclusions:*** Patients with persistently patent dcVSD remained nearly event free during follow-up. Event-free survival after dcVSD closure was markedly lower. These patients developed reduced left ventricular function and had a high risk of reintervention.

Introduction

Doubly committed ventricular septal defect (dcVSD) is the least common type of VSD in the western hemisphere, accounting for approximately 6% of all VSDs. In contrast, up to 33% of VSDs in the Asian population are dcVSDs [1]. A dcVSD is situated directly beneath the ventricular outlet with the adjacent leaflets of the pulmonary and aortic valves in fibrous continuity [2]. There are many synonymous names given to this type of VSD, such as supracristal, subarterial infundibular or subpulmonary, or subpulmonic VSD. Clinically, a thrill is present

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with a holosystolic murmur, diamond shaped and with maximum intensity in the second left intercostal space [3]. Its location adjacent to the arterial valves explains the unique natural history of this defect. Spontaneous closure is a rarity, whereas prolapse of an aortic leaflet is a common feature, with likely progression to aortic valve regurgitation (AR). The size and location of the dcVSD determine the need for surgical repair. Small dcVSDs without haemodynamic repercussion are usually treated conservatively. Larger dcVSDs are often associated with early signs of decompensation from a large left-to-right shunt or severe anatomic alterations that require early repair. In the literature, there are almost no clinical trials about the evolution of dcVSD in a western population. However, such information may be very valuable to patients with regard to their medical prognosis and capabilities and the agreements made with insurance companies. In addition, most published studies focused on the surgical repair and postoperative outcome of dcVSD. The objective of the present study was to determine the evolution and midterm outcome of both repaired and unrepaired dcVSDs in young adult patients followed in our medical centre.

Methods

Patient Population

All patients, ≥16 years and registered with a dcVSD, were selected from the database of Paediatric and Congenital Cardiology of the University Hospitals Leuven. The database contains >30,000 patients with congenital heart defects, of which 7,000 are >16 years old. The diagnosis of dcVSD was made by angiography in the early phase of the database. Later on, when transthoracic echocardiography became available, the diagnosis was confirmed by transthoracic echocardiography, which from then on formed the technique of choice for diagnosis. A dcVSD was echocardiographically defined on a parasternal short-axis and long-axis view as being situated beneath both arterial valves, with continuity of the leaflets of the aortic and pulmonary valves. In all operated patients, the diagnosis of dcVSD was confirmed peroperatively. Patients who had associated complex congenital anomalies were included in the present study. The institutional ethics committee of the hospital approved the study protocol.

Data Collection

The work-up of the study is analogical to the study of Soufflet et al. [4] concerning perimembranous VSDs. The patient records were primarily reviewed for demographic data, clinical characteristics and events after the age of 16 years. The events were defined as death, spontaneous VSD closure, surgical VSD closure, valve surgery, and implantation of a pacemaker or cardioverter defibrillator.

Secondarily, the electrocardiographic and transthoracic echocardiographic data were collected. Atrioventricular (AV) conduction time, QRS duration, type of QRS morphology, and type of rhythm (sinus rhythm or not) were reviewed. The left ventricular ejection fraction (obtained by M-mode and the Teichholz formula), left ventricular diameters (obtained by M-mode through a parasternal long-axis view) and volumes (automatically calculated by the software of the echocardiographic machine from the M-mode data), stroke volume, valve function, and pulmonary arterial hypertension (PAH, defined as a pulmonary acceleration time of <100 ms on pulsed wave Doppler or a tricuspid valve regurgitation velocity of >2.8 m/s) were assessed. Grading of valve regurgitation was done by Doppler echocardiography, on a scale (grade 0–4) according to the length of the regurgitant jet.

Statistical Analysis

Continuous variables are reported as the mean ± SD. If a non-normal distribution was present, the data were plotted as the median and interquartile range (IQR). Proportions are reported as numbers and percentages. Continuous variables between baseline (i.e. at the age of 16 years) and the latest follow-up were compared using the paired t test. The proportions between baseline and the latest follow-up were compared using the McNemar test in case of a 2 × 2 table or the Sign test where applicable. The patient outcome is displayed by using a Kaplan-Meier survival curve. Differences at $p < 0.05$ were considered statistically significant. All statistical analyses were performed using Statistical Package for Social Sciences for Windows, version 16 (SPSS, Chicago, Ill., USA).

Results

Patient Characteristics

There were 38 patients in the database who met the inclusion criteria, of whom 5 were excluded because of lack of data. A total of 33 patients (20 males, male/female ratio 1.54:1, median age 26 years, IQR 12) were included in the present study. Associated congenital anomalies were present in 5 patients: coarctation of the aorta in 3, patent ductus arteriosus in 3, atrial septal defects in 1, other VSDs in 2 and ruptured sinus of Valsalva in 1 patient. In 1 patient with a persistently patent dcVSD, the VSD extended to the perimembranous part of the interventricular septum. Another patient also had a small muscular VSD. All other patients in our study had isolated dcVSDs. Associated complex congenital malformation was seen in 1 patient who also had a transposition of the great arteries. Events were registered from the age of 16 years, for a median follow-up time of 7.9 years (IQR 9.8, time range 2–25.9). The patients were classified into two groups. One group included patients with a persistently patent interventricular communication at 16 years of age (n = 15). The other group included those patients with a closed dcVSD before the age of 16 years (n = 18). The demographic and clinical characteristics at baseline and at latest follow-up are summarized in table 1.

Table 1. Patient characteristics at baseline and latest follow-up

Variable	Patent dcVSD (n = 15)		Closed dcVSD (n = 18)	
	at baseline	at latest follow-up	at baseline	at latest follow-up
Gender				
Male	7	7	13	13
Female	8	8	5	5
Age, years				
Median	17	29	16	22
IQR	3	10	2	13
Height, cm	173 ± 6	179 ± 5	174 ± 8	177 ± 7
Weight, kg	73 ± 26	77 ± 13	58 ± 9	68 ± 8
Body surface area, m^2	1.84 ± 0.25	1.90 ± 0.14	1.70 ± 0.15	1.81 ± 0.15
Systolic blood pressure, mm Hg	115 ± 10	118 ± 13	121 ± 17	123 ± 16
Diastolic blood pressure, mm Hg	77 ± 10	74 ± 10	66 ± 13	73 ± 8

Data are presented as the mean ± SD, unless otherwise noted.

Outcome of Patients with Persistently Patent dcVSD

In 15 patients (45%), the defect remained patent at the age of 16 years. Closure was not needed because of a low pulmonary-to-systemic blood flow ratio (Qp/Qs < 1.5/1) on heart catheterization and the absence of associated valvar dysfunction. During follow-up, two spontaneous closures (13%) occurred late in life. No operative closures were needed after the age of 16 years. None of the patients died during the follow-up period.

The electrocardiographic and echocardiographic findings are summarized in table 2. All patients were in sinus rhythm at baseline. During follow-up, 1 patient developed atrial fibrillation. None of the patients developed conduction disorders. At baseline, 63% of the patients had normal QRS morphology. The remaining 37% were characterized by left or right bundle branch block or left or right hemiblock or had signs of left or right ventricular hypertrophy.

There were 6 patients with grade 1 AR at baseline, of whom 2 had associated aortic valve prolapse (AVP). We found that in the midterm follow-up, this trivial AR did not increase.

Outcome of Patients with Closed dcVSD

In 18 patients (55%), the defect was closed before the age of 16 years. The median age at closure was 5.4 years (IQR 5.9). No spontaneous closures occurred. All defects were closed by surgery. In 6 patients (33%), the VSD was primarily closed. In the remaining 12 patients (67%), the VSD was closed by patch. Main reasons for early closure were VSD together with AVP or AR, double-chambered right ventricle, net left-to-right shunting with Qp/Qs > 1.5/1. None of the patients died during the follow-up period. Two patients had a small residual VSD with a limited left-to-right shunt. Two patients with mildly persistent PAH at 16 years regained normal pulmonary artery pressure later in life.

The electrocardiographic and echocardiographic findings are summarized in table 3. All patients were in sinus rhythm at baseline and at latest follow-up. During follow-up, 2 patients required pacemaker implantation because of complete AV block. At baseline, 65% of the patients had normal QRS morphology.

Before dcVSD closure, AVP without AR was observed in 2 patients (11%). Both developed grade 2 AR during follow-up. Six patients (33%) had both AVP and AR preoperatively and all underwent aortic valvoplasty in association with VSD closure. Five of 6 (83%) patients showed progressive AR in time and 1 patient had a stable grade 1 AR. Two patients required aortic valve replacement during follow-up (15 and 32 years after VSD closure). Two patients (11%) had AR without AVP before dcVSD closure. One of these patients was diagnosed preoperatively with a ruptured sinus of Valsalva. The other patient had a bicuspid aortic valve with severe AR before dcVSD closure. Despite aortic valve repair in combination with VSD closure, both patients required aortic valve replacement later in life (19 and 9 years after closure, respectively). One patient developed complete AV block directly postoperatively requiring pacemaker implantation. Overall, 4 of 8 (50%) patients with AR before dcVSD closure required aortic valve replacement later in life. There was no AVP or AR present in 8 patients (44%) before sur-

Table 2. Changes in electrocardiographic and echocardiographic variables in patients without VSD closure (n = 15)

Variable	At baseline	At latest follow-up[1]	p value
Electrocardiographic			
Atrioventricular conduction time, ms	158 ± 32	168 ± 33	0.102[2]
QRS duration, ms	94 ± 16	95 ± 17	0.604[2]
Prevalence sinus rhythm	100	92	1.000[3]
Prevalence normal QRS morphology	63	50	1.000[3]
Echocardiographic			
Left ventricular end-diastolic diameter, mm	46 ± 5	48 ± 9	0.459[2]
Left ventricular end-systolic diameter, mm	30 ± 4	30 ± 4	0.988[2]
Left ventricular end-diastolic volume, mm	100 ± 24	112 ± 44	0.331[2]
Left ventricular end-systolic volume, mm	38 ± 11	38 ± 12	0.966[2]
Left ventricular ejection fraction, %	69 ± 7	69 ± 16	0.872[2]
Stroke volume index, ml/m^2	38 ± 7	41 ± 16	0.636[2]
Prevalence degree mitral valve regurgitation (x/4)			0.250[3]
0	83	58	
1	17	42	
2	0	0	
3	0	0	
4	0	0	
Prevalence degree AR (x/4)			1.000[3]
0	50	58	
1	50	42	
2	0	0	
3	0	0	
4	0	0	
Prevalence degree tricuspid valve regurgitation (x/4)			1.000[4]
0	17	8	
1	75	92	
2	8	0	
3	0	0	
4	0	0	
Prevalence degree pulmonic valve regurgitation (x/4)			0.250[3]
0	17	42	
1	83	58	
2	0	0	
3	0	0	
4	0	0	
Prevalence pulmonary valve stenosis	8	0	1.000[3]
Prevalence pulmonary hypertension	17	17	1.000[3]

Data are presented as the mean ± SD or percentages.
[1] Median follow-up time = 7.9 years. [2] Paired t test. [3] McNemar's test. [4] Sign test.

Table 3. Changes in electrocardiographic and echocardiographic variables in patients with VSD closure (n = 18)

Variable	At baseline	At latest follow-up[1]	p value
Electrocardiographic			
Atrioventricular conduction time, ms	146 ± 17	149 ± 22	0.532[2]
QRS duration, ms	116 ± 30	118 ± 31	0.650[2]
Prevalence sinus rhythm	100	100	1.000[3]
Prevalence normal QRS morphology	65	65	1.000[3]
Echocardiographic			
Left ventricular end-diastolic diameter, mm	49 ± 9	52 ± 6	0.104[2]
Left ventricular end-systolic diameter, mm	33 ± 8	37 ± 6	0.040[2]
Left ventricular end-diastolic volume, ml	119 ± 55	131 ± 35	0.212[2]
Left ventricular end-systolic volume, ml	48 ± 25	64 ± 22	0.024[2]
Left ventricular ejection fraction, %	69 ± 12	61 ± 6	0.028[2]
Stroke volume index, ml/m^2	39 ± 27	36 ± 8	0.691[2]
Prevalence degree mitral valve regurgitation (x/4)			0.125[4]
0	65	41	
1	35	47	
2	0	6	
3	0	6	
4	0	0	
Prevalence degree AR (x/4)			0.059[4]
0	17	12	
1	59	41	
2	12	35	
3	12	12	
4	0	0	
Prevalence degree tricuspid valve regurgitation (x/4)			0.250[4]
0	6	0	
1	82	77	
2	12	23	
3	0	0	
4	0	0	
Prevalence degree pulmonic valve regurgitation (x/4)			0.508[4]
0	23	53	
1	65	35	
2	6	6	
3	6	0	
4	0	6	
Prevalence pulmonary valve stenosis	12	29	0.250[3]
Prevalence pulmonary hypertension	12	0	0.500[3]

Data are presented as the mean ± SD or percentages.
[1] Median follow-up time = 7.9 years. [2] Paired t test. [3] McNemar's test. [4] Sign test.

gery, of which 2 patients developed no AR and 6 patients developed only grade 1 AR during follow-up.

In a subgroup analysis, we found that once operated, the risk of reintervention was high (5 of 18, 28%). The corresponding Kaplan-Meier curve is plotted in figure 1. An event was defined as redo surgery for dcVSD complications. Four out of 5 events were related to aortic valve dysfunction. Event-free survival at 10 years after dcVSD closure was 88%, and at 20 years, 64%. The mean interval between dcVSD closure and reintervention was 16 years.

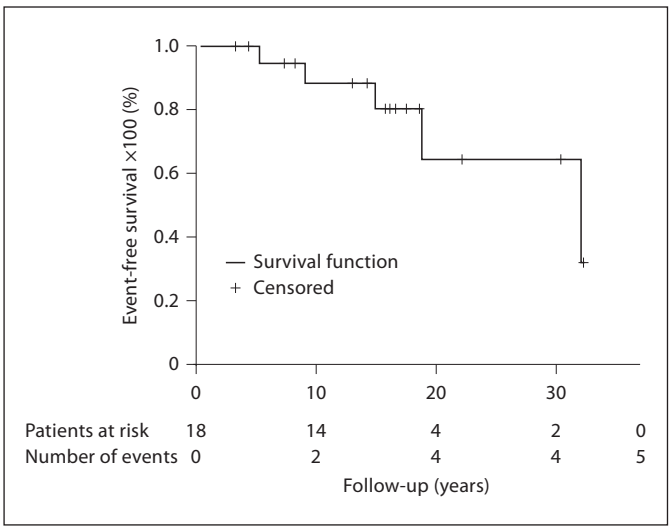

Fig. 1. Kaplan-Meier curve of event-free survival, with event defined as redo surgery after dcVSD closure.

Discussion

In the present study, 18 of 33 patients (55%) with a dcVSD required VSD closure. Reintervention was needed in 28%. Patients with unrepaired dcVSD almost completely remained event-free at follow-up. None of the patients died in our study.

Prevalence of dcVSD and Associated Lesions

VSDs can occur anywhere along the ventricular septum, and their size and location determine the clinical manifestations. dcVSDs are rare and account for approximately 6% of VSDs in the western population [1]. In our centre, there are more than 3,000 patients registered with a VSD, of whom only 38 patients (1%) have a dcVSD. These defects can occur isolated or in association with other congenital malformations [5]. In the present study, dcVSDs were most often associated with coarctation of the aorta (9%) and patent ductus arteriosus (9%). There was 1 patient with a transposition of the great arteries and 1 patient had a small ostium secundum atrial septal defect and a small apical muscular VSD. As seen in 1 patient in the study, a dcVSD can sometimes extend to the perimembranous part of the interventricular septum. Most dcVSDs have a muscular postero-inferior rim separating the leaflets of the aortic and tricuspid valves. This muscular rim separates the atrioventricular conduction axis from the postero-inferior margin of the dcVSD and thus protects the conduction axis from injury during operative VSD closure. On occasion, dcVSDs can extend so that there is fibrous continuity, not only between both arterial valve leaflets, but also between the aortic and tricuspid valve leaflets. Such defects are both doubly committed and perimembranous and present a higher risk of atrioventricular bundle injury by operative VSD closure [6].

In our study, we found a predominance of male patients (72%) in the operated group. This finding is also reported in other studies. Chang et al. [7] documented a male/female ratio of 74:46 for operated dcVSD. A possible explanation could be the observation of a higher incidence of aortic prolapse and AR in male patients with dcVSD [8].

dcVSD Closure

Small defects having small left-to-right shunts, no left ventricle volume overload and no PAH usually present as systolic murmurs without significant symptomatology and are mostly treated conservatively [1]. Most of the patients with large defects undergo an early closure procedure [9, 10]. Other indications for dcVSD closure include patients with congestive heart failure, involvement of the aortic valve (i.e. prolapse of the aortic valve leaflet into the defect, with or without AR), severe right ventricular outflow tract obstruction or multiple respiratory infections [2, 10]. In our study, 8 patients were operated because of aortic valve involvement, 1 patient because of recurrent respiratory infections and 1 because of severe right ventricular outflow tract obstruction. VSD closure is also indicated when there is a large left-to-right shunt across the dcVSD with a Qp/Qs of >2.0, and is reasonable when the Qp/Qs exceeds 1.5 [1, 10]. A Qp/Qs of >1.5 formed the indication for surgical VSD closure in 4 of 18 patients in the present study (22%). Three patients underwent VSD closure in combination with repair of associated congenital anomalies. Echocardiography showed the presence of a double-chambered right ventricle (DCRV) in 4 patients (12%), which was also corrected together with the procedure for VSD closure. Because dcVSDs are not surrounded by muscular tissue, spontaneous closure is less common [9]. In our study, two spontaneous closures (13%) occurred after the age of 16 years.

All defects were closed by surgery, and no percutaneous closures were performed. Surgical closure of isolated VSD is a safe, effective therapy. Risk of death, complete heart block and reoperation should be minimal [10]. However, in a subgroup analysis, we found that once operated, the risk of reintervention was high in our population of closed dcVSDs (28%), with an event-free survival at 20 years after dcVSD closure of 64%.

Electrocardiographic Changes during Follow-Up

No significant electrocardiographic changes were noticed in our study. In patients with persistently patent dcVSD, we observed a trend towards increasing AV conduction time. No data were found in literature concerning the relation between persistently patent dcVSD and conduction disturbances. In patients with closed dcVSD, 2 patients required pacemaker implantation because of complete AV block. One developed complete AV block 31 years after dcVSD closure. The other patient developed complete AV block immediately after aortic valve replacement surgery (Bentall operation), 19 years after the VSD closure procedure. Historically, complete heart block has been a serious complication of VSD closure, causing an increased risk of late mortality [10]. The conduction system is particularly at risk during closure of perimembranous VSDs, because of the close anatomic relationship of the AV node and the bundle of His to this type of VSD. However, in dcVSD, the conduction tissue is situated at greater distance of the VSD, with a smaller risk of conduction system injury [6, 11]. Increasing knowledge of the anatomy of the conduction system and advances in surgical technique and perioperative management have greatly decreased the incidence of intraoperative conduction system injury [10]. Still, iatrogenic complete heart block continues to occur after surgical VSD closure, either because of unexpected anatomic variations or because of unawareness of the location of the AV conduction axis [12]. Chronic inflammation or fibrosis is a more important determinant of late-onset AV block after dcVSD closure, making this an important aspect of follow-up [11].

Progression of AR and Need for Aortic Valve Repair/Replacement

The location of dcVSDs results in a markedly increased incidence of associated aortic valve dysfunction as compared to the normal population and patients with other types of VSDs. A study from 2003 with 685 patients with a VSD reported AVP in 8.6% of all VSDs and AR in 6.4%. In contrast, AVP was reported in 69% and AR in 36% of dcVSDs [9]. The anatomic and haemodynamic features in dcVSD have a great impact on the development of AVP and subsequent AR. Some authors have described AVP to be due to a deficiency in the infundibular musculature and subsequent lack of support of the aortic valve [8, 13]. Further, arterial valve offsetting (i.e. both arterial valves are not located at the same level, confined by the presence of the aortic leaflet below the pulmonary valve ring) is one of the major contributing factors to the development of leaflet deformity [2]. Due to offsetting, the aortic valve leaflets, especially the right coronary leaflet, are susceptible to mechanical forces induced by VSD flow, resulting in an aortic valve deformity. Other factors contributing to AVP are aging and the absence of PAH [2, 14]. PAH might prevent the aortic valve leaflet from prolapsing in dcVSD because the increased right ventricular pressure reduces the Venturi effect (i.e. the effect produced by the pressure difference between the right and left ventricle and responsible for an enhanced VSD flow and thus the emergence of AVP). The development of AR may be due to distortion of the herniated aortic sinus or it may result from a long-standing Venturi effect through the dcVSD [13]. As a consequence, the enlarged protruding aortic sinus may partially obstruct the VSD and limit the shunt and thus the development of PAH.

In our study, we found that patients with persistently patent dcVSD only showed minimal aortic valve dysfunction that did not progress during follow-up. In contrast, in patients with a closed dcVSD who had AVP and/or AR preoperatively, we observed a trend towards progression of AR ($p = 0.059$).

In 2002, Cheung et al. [14] conducted a study on the impact of preoperative aortic leaflet prolapse on the long-term outcome after surgical dcVSD closure. Of the 79 patients with no aortic leaflet prolapse and no AR preoperatively, none developed AR during a follow-up time of 6.1 years. None required further interventions. Seventeen patients had moderate to severe AVP and AR and underwent valvoplasty in addition to VSD closure. Regurgitation improved in 10, but remained the same or worsened further in 7 patients at a median follow-up of 4.6 years. Tomita et al. [15] conducted a similar study that followed 55 patients for a period of ≥ 5 years after dcVSD patch closure. They observed the development of trivial or mild AR in 6 out of 23 patients without AVP or AR before dcVSD closure. Two of 15 patients with AVP without AR developed trivial or slight-to-moderate AR after closure. In patients with AVP and AR preoperatively, they found that AR resolved or improved in 12 and remained unchanged in 3 patients.

In summary, surgery for dcVSD, if performed before the onset of AVP, may prevent progressive AR. However, after the onset of AVP, surgical closure without valve repair may not prevent progressive AR. Once AR has occurred, it may still progress despite closure of VSD and valvoplasty. Although valve repair seems to improve and control any progressive AR compared to VSD closure alone, some patients ultimately may still require an aortic valve replacement. The identification of risk groups for developing aortic valve complications which may be pre-

vented by early prophylactic repair of the dcVSD is very important, because AR is often progressive and may be a cause of late morbidity and mortality.

Progression of Pulmonary Valve Dysfunction

Before surgery, we detected valvar pulmonary stenosis (PS) in 2 and infundibular PS associated with DCRV in another 2 patients. DCRV is not uncommonly observed in association with VSDs and pulmonary valve stenosis [16]. PS is important to recognize because of the greater operative risk noted in patients with a dcVSD associated with PS [5]. Therefore, a precise angiographic documentation and localization of PS is needed. Clinical features of dcVSD associated with PS are those of classical tetralogy of Fallot (TOF). Since the association of dcVSD with PS is very uncommon in the western population, it was often mistaken for TOF [17, 18]. In contrary to TOF, the infundibular septum is absent in dcVSD associated with PS [17]. During follow-up, 4 patients with closed dcVSD developed a mild grade of infundibular and/or valvar PS.

Structural Echocardiographic Changes during Follow-Up

The left ventricular ejection fraction, stroke volume, left ventricular diameters and volumes were assessed at baseline and further on. We could not find significant structural echocardiographic changes during follow-up in patients with persistently patent dcVSD. In the group with closed dcVSD, we found a significant reduction in ejection fraction, together with a significant increase in end-systolic diameter and volume during follow-up. This evolution is not reported in literature. A possible explanation could be a diminished left ventricular contractility due to chronic volume overload before VSD closure. In contrast, Soufflet et al. [4] reported an increase in ejection fraction and a decrease in end-systolic diameter with time in patients with persistent perimembranous VSD.

In dcVSD, the position of the defect results in a left ventricle-to-pulmonary artery shunt which almost bypasses the right ventricle. Consequently, dcVSDs tend to produce congestive heart failure and PAH [2]. No significant changes in PAH were noticed in our study.

Limitations

We acknowledge that there are several limitations to our study. First, this study is limited by its retrospective nature and the inherent risks of all retrospective studies, most importantly the risk of missing data. However, we point out that patients with dcVSD are systematically followed up in our institution. Second, we presented a single-centre study, which implies a referral and selection bias. However, due to the healthcare organization in Belgium, almost all children with a VSD are referred to a tertiary care centre. Our hospital is 1 of the 2 main referral centres for congenital heart disease in the Flemish speaking part of Belgium. As a consequence, the risk of data dispersion was low. Finally, the size of our study population was limited wherefore we could only outline some trends but no statistical significant changes. Larger studies will be needed to have a better view on the evolution and complications of repaired and unrepaired dcVSD. However, since the rarity of this type of VSD, no single-centre study in western countries will ever be big enough to reach good statistical power and multi-centre trials will be required.

Conclusion

Patients with unrepaired dcVSD proved to remain quite stable during follow-up, without development of important structural or functional impairment. In contrast, event-free survival after dcVSD closure was markedly lower. This implies important information for both groups of patients, regarding professional choices and insurance policies.

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