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Dysrhythmias in patients with a complete atrioventricular septal defect: From surgery to early adulthood

Charlotte A. Houck MD^{1,2} | Reinder Evertz MD³ | Christophe P. Teuwen MD¹ |
 Jolien W. Roos-Hesselink MD, PhD¹  | Janneke A. E. Kammeraad MD, PhD⁴ |
 Anthonie L. Duijnhouwer MD³  | Natasja M. S. de Groot MD, PhD³ |
 Ad J. J. C. Bogers MD, PhD² 

¹Department of Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands

²Department of Cardiothoracic Surgery, Erasmus University Medical Center, Rotterdam, The Netherlands

³Department of Cardiology, Radboud University Medical Center, Nijmegen, The Netherlands

⁴Department of Pediatric Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands

Correspondence

Ad J. J. C. Bogers, MD, PhD, Department of Cardiothoracic Surgery, Erasmus University Medical Center, RG-619, Dr. Molewaterplein 40, Rotterdam 3015 GD, The Netherlands.
 Email: a.j.j.c.bogers@erasmusmc.nl

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Abstract

Objective: Outcomes after surgical repair of complete atrioventricular septal defect (cAVSD) have improved. With advancing age, the risk of development of dysrhythmias may increase. The aims of this study were to (1) examine development of sinus node dysfunction (SND), atrial and ventricular tachyarrhythmias, and (2) study progression of atrioventricular conduction abnormalities in young adult patients with repaired cAVSD. **Study design:** In this retrospective multicenter study, 74 patients (68% female) with a cAVSD repaired in childhood were included. Patients' medical files were evaluated for occurrence of SND, atrioventricular conduction block (AVB), atrial and ventricular tachyarrhythmias.

Results: Median age at repair was 6 months (interquartile range 3-10) and median age at last follow-up was 24 years (interquartile range 21-28). SND occurred after a median of 17 years (interquartile range 11-19) after repair in 23% of patients, requiring pacemaker implantation in two patients (12%). Regular supraventricular tachycardia was observed in three patients (4%). Atrial fibrillation and ventricular tachyarrhythmias were not observed. Twenty-seven patients (36%) had first-degree AVB, which was self-limiting in 16 (59%) and persistent in 10 (37%) patients. One patient developed third-degree AVB 7 days after left atrioventricular valve replacement. Spontaneous type II second-degree AVB occurred in a 28-year-old patient. Both patients underwent pacemaker implantation.

Conclusions: Clinically significant dysrhythmias were uncommon in young adult patients after cAVSD repair. However, three patients required pacemaker implantation for either progression of SND or spontaneous type II second-degree AVB. Longer follow-up should point out whether dysrhythmias will progress or become more prevalent with increasing age.

KEYWORDS

adult congenital heart disease, atrial tachyarrhythmias, atrioventricular conduction block, complete atrioventricular septal defect, sinus node dysfunction

1 | INTRODUCTION

The first report of successful repair of complete atrioventricular septal defect (cAVSD) dates from 1955 by C.W. Lillehei.¹ In the following decades, alterations in surgical techniques, increased knowledge of the anatomy of the conduction system and improved perioperative care led to improved outcomes after cAVSD repair.²

New problems may arise in this aging population, including brady- and tachyarrhythmias. Previous studies have shown that the incidence of dysrhythmias in patients with congenital heart disease (CHD) steadily increases with increasing age.^{3,4} In patients with an atrial septal defect (ASD), age at repair—and thus duration of volume overload and subsequent arrhythmogenic atrial stretch⁵—is associated with development of atrial tachyarrhythmias (AT).⁵⁻⁷ The reported incidence of AT was lower when ASD repair was performed at a younger age.^{6,7}

Since the majority of patients with cAVSD undergo surgical repair at young age, the duration of volume overload and its consequences is relatively short. Nonetheless, these patients not only develop AT—which may also result from surgical scars and suture lines—but also bradyarrhythmias, including sinus node dysfunction (SND) and late atrioventricular conduction block during follow-up.⁸⁻¹¹

However, the number of studies explicitly focusing on dysrhythmias in patients with cAVSD repaired in childhood in a recent surgical era is scarce.⁹ Additionally, the time line of dysrhythmias, including age and relation to (redo) surgery, has less well been studied. When patients with a surgically repaired cAVSD grow older and reach (early) adulthood, it is likely that the risk of development of various dysrhythmias may increase.

Therefore, the aims of this study were to (1) examine the occurrence of SND, atrial and ventricular tachyarrhythmias and (2) study time course and progression of atrioventricular (AV) conduction abnormalities in a cohort of young adult patients after surgical repair of cAVSD in childhood.

2 | METHODS

2.1 | Study population

This retrospective multicenter study was part of the Dysrhythmias in Patients with Congenital Heart Disease (DANARA) project, which was approved by the local ethics committee in the Erasmus University Medical Center Rotterdam (MEC-2012-482). Informed consent was not required.

Seventy-four patients who underwent surgical repair of cAVSD <18 years of age between 1986 and 1999 were collected from databases of the participating academic hospitals (Erasmus University Medical Center: $N = 52$, Radboud University Medical Center: $N = 22$). This study period was based on stable operator and institutional experience in cAVSD repair and perioperative management, as well as consistent availability of patient data. Patients <18 years of age at last follow-up and patients with cAVSD unsuitable for biventricular repair or other types of major CHD were excluded.

Data were collected from patients' medical records, starting from the moment of first diagnosis of cAVSD. Intervals between clinical evaluations depended on the presence of relevant sequelae or residual lesions according to the guidelines.¹² Clinical evaluation of patients included history, physical examination, electrocardiography (ECG) and, when available, 24-hour Holter recording, echocardiography, and exercise testing. Echocardiographic characteristics at last follow-up were available in 63 patients; in the other 11 patients, latest echocardiographic characteristics dated from follow-up encounters at a median of 16 months (range 4 months to 4.5 years) before last follow-up. The following interpretations of echocardiographic parameters were included: left or right ventricular function, moderate/severe left or right atrioventricular valve insufficiency; severe left or right atrial dilatation, and residual ASD and/or ventricular septal defect (VSD). Echocardiographic definitions were based on guidelines.^{13,14}

2.2 | Dysrhythmias

All available rhythm registrations and correspondence between first diagnosis of cAVSD and last follow-up were included and evaluated for occurrence of the following dysrhythmias: (1) SND, (2) first-, second-, and third-degree AVB, (3) atrial fibrillation (AF), (4) regular supraventricular tachycardia (SVT; including atrial flutter, intraatrial reentrant tachycardia, ectopic atrial tachycardia, atrioventricular nodal reentrant tachycardia, and atrioventricular reciprocating tachycardia), and (5) ventricular tachyarrhythmia (VTA; including sustained ventricular tachycardia and ventricular fibrillation).

Dysrhythmias were defined according to the guidelines.^{15,16} SND included sinus bradycardia or chronotropic incompetence without identifiable causes, paroxysmal or persistent sinus arrest with replacement by subsidiary escape rhythms in the atrium, AV junction, or ventricular myocardium, or tachy-brady syndrome.¹⁵ We subsequently defined sinus bradycardia in patients >16 years of age as sinus rhythm <50 beats/min without use of β -blockers.¹⁷ In patients <16 years of age, heart rate <2nd percentile and PR-duration >98th percentile according to age as recommended by Rijnbeek et al were applied for retrospective diagnosis of, respectively, sinus bradycardia and first-degree AVB.¹⁸ Sinus bradycardia was diagnosed on an ECG made by day or a 24-hour Holter recording. In case of the latter, SND did not include (1) strictly nocturnal sinus rates under the lower limit appropriate for age and (2) an overall average heart rate under the lower limit over 24 hours. First-degree AVB was diagnosed in the absence of use of drugs potentially causing PR-prolongation. We did not differentiate between typical (counter) clockwise atrial flutter, intra-atrial reentrant tachycardia, or ectopic atrial tachycardia.

2.3 | Statistical analysis

Normally distributed continuous variables were expressed as mean \pm SD and skewed continuous variables as median (interquartile range (IQR)). Categorical variables were expressed as numbers and percentages. Cumulative freedom from SND was estimated using a right-censored Kaplan-Meier curve, constructed with age as time scale.³

A *P* value < 0.05 was considered statistically significant. Statistical analysis was performed with SPSS version 24 (IBM Corporation, Armonk, New York).

3 | RESULTS

3.1 | Study population

Clinical characteristics of 74 patients (female: *N* = 50, 68%) with a repaired cAVSD are listed in Table 1. Fifty patients (68%) were additionally diagnosed with Down syndrome and three (4%) had impaired mental functioning with (1) or without (2) a known underlying chromosomal abnormality. Given similar difficulties in reporting of symptoms, these patients will be combined in 1 group ("syndromic") for the sake of this study. These patients did not differ from nonsyndromic patients regarding characteristics listed in Table 1 (all *P* > 0.05).

Repair of cAVSD was performed at a median age of 6 months (3-10). Typically, a 2-patch technique was applied throughout the series.^{19,20} In one patient, the atrial component was closed with a patch and the ventricular component was directly closed. Another patient

underwent additional infundibulectomy for infundibular pulmonary stenosis. A preparatory pulmonary banding had been performed in 1 patient and in another patient an aortic coarctation had been repaired before cAVSD repair. Median age at last follow-up was 24 years (IQR: 21-28, range: 18-37).

During follow-up, 17 patients (23%) underwent 1 or more additional surgical procedure(s) including AV valve surgery (*N* = 13), AV valve surgery and resection of subaortic stenosis (*N* = 3), resection of subaortic stenosis (*N* = 3), and AV valve surgery and repair of residual ASD/VSD (*N* = 2).

Latest echocardiography interpretations described moderately impaired left ventricular function in 4 of 73 available reports (5%) and moderately impaired right ventricular function in 4 of 69 reports (6%). Moderate to severe left atrioventricular valve insufficiency was present in 32% of patients.

3.2 | Sinus node dysfunction

As demonstrated in Figure 1, SND was identified in 17 patients at a median age of 18 years (12-24), corresponding to 23% of syndromic

TABLE 1 Patient characteristics

Number of patients	74
Female gender	50(68)
Down syndrome	50(68)
Impaired mental functioning—other	3(4)
Age at cAVSD repair (months)	6(3-10)
Duration postoperative follow-up (years)	23(20-27)
No. of surgical procedures	
• 1	57(77)
• 2	12(16)
• 3	4(5)
• 4	1(1)
LAVV replacement	3(3)
Echocardiographic characteristics	
Left ventricular function	
• Normal/mildly impaired	69/73(95)
• Moderately impaired	4/73(5)
Right ventricular function	
• Normal/mildly impaired	65/69(94)
• Moderately impaired	4/69(6)
Severe left atrial dilatation	4/56(7)
Severe right atrial dilatation	1/34(3)
Moderate/severe LAVV insufficiency	24/74(32)
Moderate/severe RAVV insufficiency	6/73(8)
Residual shunt	0/41

Values are presented as *N*(%) or median (interquartile range). For each echocardiographic parameter the number of patients with available data is shown.

Abbreviations: cAVSD, complete atrioventricular septal defect; LAVV, left atrioventricular valve; RAVV, right atrioventricular valve.

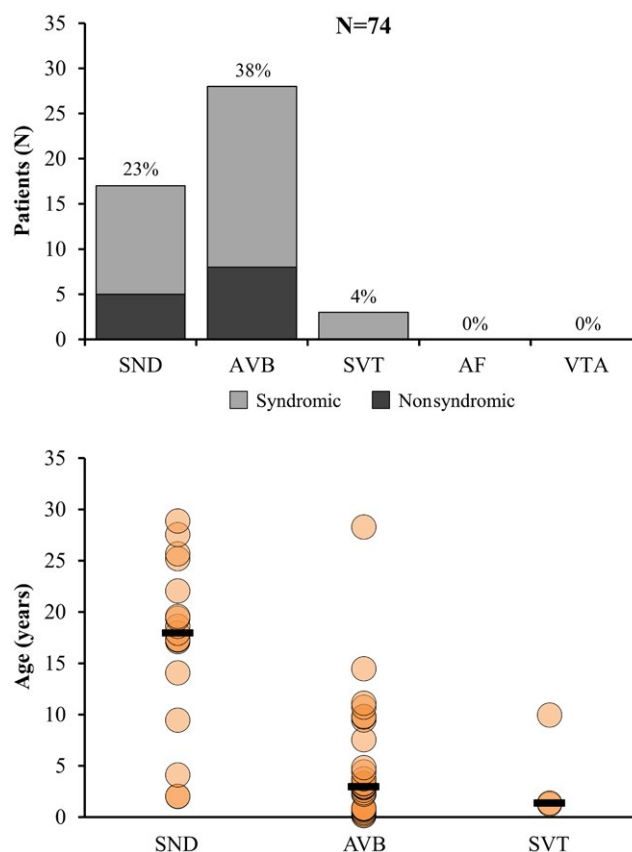


FIGURE 1 (Upper panel) Occurrence of dysrhythmias in syndromic and nonsyndromic patients. (Lower panel) Age at development of dysrhythmias. Bars indicate median age. Abbreviations: AF, atrial fibrillation; AVB, atrioventricular conduction block; cAVSD, complete atrioventricular septal defect; SND, sinus node dysfunction; SVT, supraventricular tachycardia; VTA, ventricular tachyarrhythmia

TABLE 2 Characteristics of patients requiring pacemaker implantation

Patient	Gender	Down syndrome	Age	Indication	Symptoms	Diagnosis	(Redo) surgery
1	F	Yes	17.5y	SND	Syncope	Symptomatic SND (ILR)	17y after repair
2	F	No	18.5y	AVB-III	Asymptomatic	ECG, bedside telemetry	7d after LAVV-replacement
3	F	Yes	24.4y	SND	Presyncope	Progressive symptoms, preexistent SND	24y after repair
4	M	Yes	28.7y	AVB-II	Presyncope	Symptomatic AVB (Holter)	28y after repair

Abbreviations: AVB(-II, -III), (second-, third-degree) atrioventricular conduction block; ILR, implantable loop recorder; LAVV, left atrioventricular valve; SND, sinus node dysfunction.

patients and 24% of nonsyndromic patients ($P = 1$). Types of SND at presentation included sinus bradycardia ($N = 15$) and sinus arrest ($N = 2$); all were initially documented on a routine ECG. The 2 patients with sinus arrest underwent 24-hour Holter monitoring to confirm the diagnosis.

As illustrated in the upper panel of Figure 2, SND occurred (relatively) late after repair of cAVSD in all 17 patients (median: 17 years (IQR 11-19, range: 1.5-27)). Three of these patients underwent redo surgical procedures during follow-up: SND occurred long

(respectively, 18 and 20 years) after redo AV valve surgery in two patients and 6 years before redo AV valve surgery in one patient. Two patients (12%) received a pacemaker for symptomatic SND at the age of, respectively, 18 and 24 years. Characteristics of these patients are listed in Table 2. Both patients initially had asymptomatic sinus bradycardia for several years before they developed progressive symptoms that were correlated to SND on, respectively, implantable loop recorder and 24-hour Holter monitoring. The lower panel of Figure 2 shows that at the age of 35 years, an estimated 65% of cAVSD patients remained free from SND.

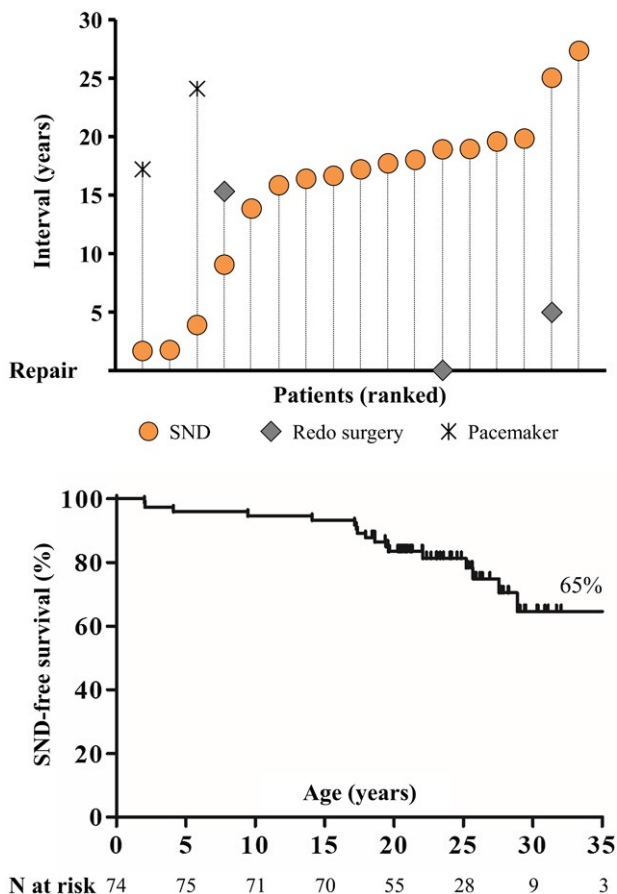


FIGURE 2 (Upper panel) Time course of identification of SND, redo surgical procedures and pacemaker implantation in relation to cAVSD repair. (Lower panel) SND-free survival curve. Abbreviation: SND, sinus node dysfunction

3.3 | Atrioventricular conduction block

AVB was observed in 28 patients (38%) at a median age of 3 years (9 months to 9 years; Figure 1). The incidence did not differ between syndromic and nonsyndromic patients (both 38%, $P = 1$).

The majority of patients with AVB presented with first-degree AVB on a routine ECG ($N = 27$; upper left panel Figure 3). In four patients first-degree AVB presented before cAVSD repair. As illustrated in the upper right panel of Figure 3, first-degree AVB was self-limiting in 16 (59%) and persistent in 10 (37%) of 27 patients. Self-limiting first-degree AVB was present for a median duration of 5 years (11 months to 7 years) before normal AV conduction returned (lower panel Figure 3).

Pacemaker implantation was required in two patients with AVB (Table 2). One patient underwent left atrioventricular valve replacement and developed postoperative third-degree AVB, 18 years after initial presentation of first-degree AVB. This patient had normal sinus rhythm with first-degree AVB for the first 7 postoperative days, before development of third-degree AVB. Another patient presented with symptomatic type II second-degree AVB without apparent cause at the age of 28 years, which was diagnosed on 24-hour Holter monitoring. ECG at this time showed preexistent bifascicular block and new onset first-degree AVB. Both patients received a pacemaker at, respectively, 28 (second degree) and 18 years (third degree) of age.

3.4 | Atrial and ventricular tachyarrhythmias

Only 3 patients (4%; all with Down syndrome) had regular SVT, which was diagnosed based on postoperative rhythm monitoring early after cAVSD repair (1) or left atrioventricular valvuloplasty (1) and did not recur

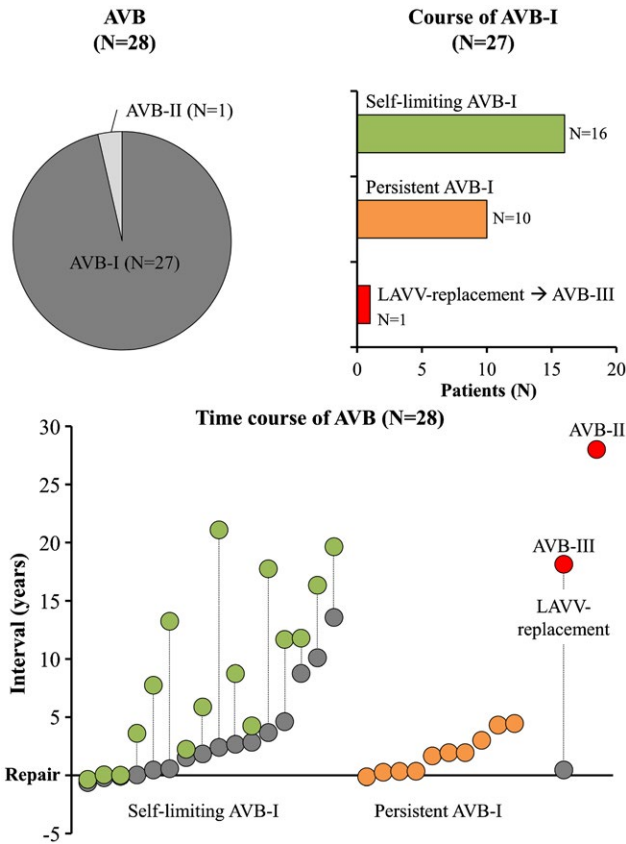


FIGURE 3 Upper left panel: type of AVB at presentation. Upper right panel: self-limiting or persistent first-degree AVB, or progression of first-degree AVB. Lower panel: time course of AVB in relation to cAVSD repair. AVB(-I, -II, -III): (first-, second-, third-degree) atrioventricular conduction block, LAVV, left atrioventricular valve

after the early postoperative period in both patients. Another patient developed SVT 9 months after initial repair in the setting of severe residual left atrioventricular valve insufficiency and residual ASD and VSD. This patient underwent redo surgery, after which SVT did not recur. Median age at occurrence of SVT was 16 months (range 15 months-10 years). AF and VTA were not observed in this study population.

4 | DISCUSSION

This study demonstrated development of late postoperative SND in 23% of patients with cAVSD repaired at a median age of 6 months and a median postoperative follow-up duration of 23 years. Most patients remained asymptomatic, but pacemaker implantation was required in 12% of these patients. One patient developed spontaneous symptomatic type II second-degree AVB 28 years after cAVSD repair, requiring pacemaker implantation. Fortunately, regular SVT were less common (4%) and AF and VTA were not observed, as may be expected in this relatively young study population.

Prior studies including arrhythmic outcomes during long-term follow-up after surgical repair of cAVSD in childhood are scarce. Either surgery was performed decades ago—thus with considerable differences in surgical techniques and perioperative care^{2,8,21}—patients were older at the time of repair,¹⁰ long-term follow-up until adulthood was missing,¹¹ or outcomes for complete and partial AVSD were only partially separated.^{9,10} One of these studies included 238 patients with repaired cAVSD and 177 with repaired partial AVSD and a mean postoperative follow-up duration of 9 years.⁹ SND was observed in 6.7% and late complete AVB in 0.7%, although these outcomes were not further specified according to AVSD type. First-degree AVB was identified in 19% of patients, as opposed to 36% in our study. Use of age-specific criteria¹⁸ for retrospective diagnosis of first-degree AVB in the present study may explain the difference in these findings. Soufflet et al evaluated long-term outcomes in 71 patients after surgical repair of cAVSD at a median age of 3.8 years (0.2-61.3).¹⁰ AT occurred more often compared to our study (respectively, 11% vs 4%), which might be explained by the difference in age at repair. Suzuki et al included 116 patients undergoing surgical cAVSD repair at a median age of 4.8 months (9 days to 5.4 years), who were followed for a median duration of 27 months.¹¹ Permanent pacemaker implantation was performed in 2/116 patients (2%), although the indication was not provided. The somewhat higher percentage of patients requiring pacemaker implantation in our study (5%) may be explained by the considerable difference in follow-up duration.

4.1 | Sinus node dysfunction

In the present study, SND occurred late after cAVSD repair and was not related to redo surgical procedures. These findings are comparable to those of a previous study including 34 patients with a surgically repaired ASD and SND.¹⁷ Median interval between ASD repair and SND was 16 years and late SND was not related to redo surgical procedures. Similar findings were reported by Roos-Hesselink et al, who studied outcomes of surgical repair of VSD at young age after 22-34 years of follow-up.²² A pacemaker was implanted in 4/95 patients because of sick sinus syndrome more than 15 years after surgery. The progressive nature of the disease in these patients was illustrated by the presence of minor indicators of SND after 10-22 years of follow-up.²³ Development of (late) SND in CHD patients was also previously studied by Goldman et al, who reviewed indications for permanent pacing in 132 patients after surgery for CHD.²⁴ Specific results for patients with cAVSD ($N = 4$) were not provided, but overall, late SND was the indication for permanent pacing in 14% of patients. In the present study, two patients requiring pacemaker implantation for symptomatic SND had preexistent asymptomatic sinus bradycardia for over a decade before they became progressively symptomatic. It was previously demonstrated that the incidence of SND in the general population increased with age: from 0% to 1.2% at 45-54 years of age to 6%-23% in subjects 75-84 years of age.²⁵ Thus, it should be taken into account that SND in cAVSD patients may also be a progressive disease and may become more severe as these patients are aging. The cause of SND in cAVSD

patients is unknown, but is probably multifactorial. A congenital origin, direct surgical damage, late fibrosis after surgery, sinus node abnormalities due to atrial stretch, persistence of electrophysiological abnormalities beyond surgical repair, and aging may all contribute to development of SND in patients with septal defects.²⁴⁻²⁷

4.2 | Atrioventricular conduction block

In line with previous findings, first-degree AVB was relatively often observed.²⁸ The anatomy of the conduction system in patients with cAVSD is abnormal, with a more posterior position of the atrioventricular node (AV) and His bundle compared to patients with a structurally normal heart.² First-degree AVB in these patients is most commonly caused by intraatrial conduction delay,²⁸ which is probably due to the altered course of the atrial conduction system and thus atrial activation in the presence of the atrial component of cAVSD. In the present study, a large atrial component of the defect was found in two of four patients with preoperative first-degree AVB. Regression of first-degree AVB occurred in 59% of patients. We assume that in some patients, regression may be caused by inversed atrial remodeling after cAVSD repair. Furthermore, the relative difference in PR-duration initially caused by intraatrial conduction delay as described above may become less outspoken with increasing age compared to healthy peers.

Increased knowledge of the course of the conduction system in cAVSD patients and improved surgical techniques resulted in a dramatic decrease of surgically induced third-degree AVB.² In the present study, not one patient developed immediate postoperative third-degree AVB. One patient did develop third-degree AVB 7 days after left atrioventricular valve replacement, necessitating pacemaker implantation. A prior study demonstrated a relatively high incidence of early postoperative third-degree AVB after left atrioventricular valve replacement compared to left atrioventricular valvuloplasty in cAVSD patients, which—the authors suggested—may be explained by mechanical compression of the prosthesis on the conduction system.²⁹ However, our patient had normal sinus rhythm with a previously known first-degree AVB for 7 days before development of third-degree AVB. This course implicates that AVB in this patient was not caused by direct surgical injury or compression of the prosthesis to the AV conduction system. We assume that third-degree AVB in this patient was caused by development of scar tissue after left atrioventricular valve replacement.

One 28-year-old patient developed spontaneous symptomatic type II second-degree AVB without apparent cause. Concomitant documentation of preexistent bifascicular block and de novo first-degree AVB might imply a progressive deterioration of the AV conduction system. A previous study on dysrhythmias in patients with a surgically repaired ASD showed that development of AVB was not related to a redo surgical procedure in 6 of 10 patients with late third-degree AVB.¹⁷ Development of AV conduction abnormalities in patients with septal defects may be a progressive disease²² influenced by (a combination of) several factors including abnormal anatomy, a genetic origin, direct surgical damage, late fibrosis

after cardiac surgery, and aging.^{24,30} Given the overall young age at cAVSD repair, fibrosis near the AV node caused by hemodynamic effects of the shunt seems less likely but cannot be ruled out as contributing cause.

4.3 | Aging and atrial tachyarrhythmias

It was previously demonstrated that aging is associated with an increased incidence of AT.³¹ However, the incidence of AT in aging patients with CHD is strikingly higher compared to the general population.⁴ The underlying mechanism of AT in CHD patients is variable: “incisional” AT are directly related to prior surgical lesions, whereas AT may also occur as a result of longstanding volume and/or pressure overload leading to atrial stretch.^{5,32} Our data did not allow us to differentiate between these types of AT.

As expected given the young age of our study population, the number of patients with AT was relatively low. These findings are in line with previously reported incidences of AT in adult cAVSD patients (~2%-10%).^{8,10} In addition, it can be postulated that AT are also less common in patients with cAVSD due to generally early closure of the defect. Prior studies including patients with an ASD demonstrated that young age at repair resulted in significantly lower incidences of AT.^{5,6} This observation is most likely based on earlier termination of the left-right shunt, thereby preventing further right atrial dilatation and consequent vulnerability to development of AT.⁵ Longer follow-up of surgically repaired cAVSD patients should point out whether AT become more frequent as these patients become older.

4.4 | Study limitations

Our selection of the study period (1986-1999) obviously resulted in a young study population at follow-up. This may explain the relatively low incidence of dysrhythmias, which may be expected to increase with increasing age, as seen with other CHD types.^{3,4} However, in our opinion, this time period represents stable surgical experience—about 10 years after introduction of the 2-patch technique²—and encompasses patients currently entering (early) adulthood, who may be at risk of development of dysrhythmias.

Due to the retrospective nature of this study, patients did not receive the exact same diagnostic tests at the same time points during follow-up. In accordance with the guidelines, frequency of follow-up depended on the presence and severity of residual lesions.¹² Although the majority of patients was seen at follow-up intervals up to 5 years, some patients had a gap >6 years in follow-up: 12% of patients with SND and/or AVB had a gap as opposed to 30% of patients without. Furthermore, 24-hour Holter recordings were performed at the discretion of the treating physician. Our results showed that dysrhythmias were not documented on 24-hour Holter recordings that were performed for screening purposes only, without a specific indication. These differences in follow-up intervals and rhythm monitoring methods may result in an underestimation of the number of patients with (asymptomatic) dysrhythmias. Furthermore, asymptomatic events may have occurred before the first actual

documentation of dysrhythmia. The number of dysrhythmias in patients with impaired mental functioning or Down syndrome may be underestimated due to potential underreporting of symptoms.

5 | CONCLUSION

Incidence of clinically significant dysrhythmias in young adult patients after surgical repair of cAVSD in childhood was low, as indicated by the absence of atrial fibrillation and ventricular tachyarrhythmias. In addition, there was a low incidence of regular SVT, most of which only occurred in the immediate postoperative period. Although SND was not uncommon, most patients remained asymptomatic. However, pacemaker implantation was required in 12% of these patients. One patient developed spontaneous type II second-degree AVB requiring pacemaker implantation. SND and AV conduction abnormalities in cAVSD patients are probably caused by multiple factors and might have a progressive course in some patients. Longer follow-up of these young adult patients should point out whether dysrhythmias will progress or become more prevalent with increasing age.

CONFLICT OF INTEREST

The authors declare that they no conflict of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

Conceptualization; data curation; formal analysis; writing—original draft: Charlotte A. Houck

Data curation; formal analysis; writing—review & editing: Reinder Evertz
Data curation; writing—review & editing: Christophe P. Teuwen.

Writing—review & editing; expertise on adult congenital heart disease: Jolien W. Roos-Hesselink

Writing—review & editing; expertise on pediatric cardiology: Janneke A. E. Kammeraad

Writing—review & editing; expertise on adult congenital heart disease: Anthonie L. Duijnhouwer

Supervision; writing—review & editing: Natasja M. S. de Groot

Conceptualization; supervision; writing—review & editing: Ad J. J. C. Bogers

ORCID

Jolien W. Roos-Hesselink  <http://orcid.org/0000-0002-6770-3830>

Anthonie L. Duijnhouwer  <http://orcid.org/0000-0001-5064-0143>

Ad J. J. C. Bogers  <http://orcid.org/0000-0001-6200-1998>

REFERENCES

- Lillehei CW, Cohen M, Warden HE, Varco RL. The direct-vision intracardiac correction of congenital anomalies by controlled cross circulation; results in thirty-two patients with ventricular septal defects, tetralogy of Fallot, and atrioventricularis communis defects. *Surgery*. 1955;38(1):11-29.
- Backer CL, Stewart RD, Mavroudis C. Overview: history, anatomy, timing, and results of complete atrioventricular canal. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2007;10:3-10.
- Avila P, Oliver JM, Gallego P, et al. History and clinical predictors of atrial tachycardia in adults with congenital heart disease. *Circ Arrhythm Electrophysiol*. 2017;10(9):e005396.
- Bouchardy J, Therrien J, Pilote L, et al. Atrial arrhythmias in adults with congenital heart disease. *Circulation*. 2009;120(17):1679-1686.
- Gatzoulis MA, Freeman MA, Siu SC, Webb GD, Harris L. Atrial arrhythmia after surgical closure of atrial septal defects in adults. *N Engl J Med*. 1999;340(11):839-846.
- Cuypers JA, Opic P, Menting ME, et al. The unnatural history of an atrial septal defect: longitudinal 35 year follow up after surgical closure at young age. *Heart*. 2013;99(18):1346-1352.
- Murphy JG, Gersh BJ, McGoan MD, et al. Long-term outcome after surgical repair of isolated atrial septal defect. Follow-up at 27 to 32 years. *N Engl J Med*. 1990;323(24):1645-1650.
- Daliento L, Rizzoli G, Marchiori MC, et al. Electrical instability in patients undergoing surgery for atrioventricular septal defect. *Int J Cardiol*. 1991;30(1):15-21.
- Kharbanda RK, Blom NA, Hazekamp MG, et al. Incidence and risk factors of post-operative arrhythmias and sudden cardiac death after atrioventricular septal defect (AVSD) correction: Up to 47 years of follow-up. *Int J Cardiol*. 2018;1(252):88-93.
- Soufflet V, Daenen W, Van Deyk K, Troost E, Budts W. Repair for partial and complete atrioventricular septal defect: single centre experience and long-term results. *Acta Clin Belg*. 2005;60(5):236-242.
- Suzuki T, Bove EL, Devaney EJ, et al. Results of definitive repair of complete atrioventricular septal defect in neonates and infants. *Ann Thorac Surg*. 2008;86(2):596-602.
- Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J*. 2010;31(23):2915-2957.
- Lancellotti P, Moura L, Pierard LA, et al. Association of echocardiography recommendations for the assessment of valvular regurgitation. Part 2: mitral and tricuspid regurgitation (native valve disease). *Eur J Echocardiogr*. 2010;11(4):307-332.
- Lang RM, Badano LP, Mor-Avi V, et al. Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *Eur Heart J Cardiovasc Imaging*. 2015;16(3):233-270.
- Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2013;127(3):e283-e352.
- Page RL, Joglar JA, Caldwell MA, et al. 2015 ACC/AHA/HRS guideline for the management of adult patients with supraventricular tachycardia: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Heart Rhythm*. 2016;13(4):e136-e221.
- Houck CA, Evertz R, Teuwen CP, et al. Time course and interrelationship of dysrhythmias in patients with surgically repaired atrial septal defect. *Heart Rhythm*. 2018;15(3):341-347.
- Rijnbeek PR, Witsenburg M, Schrama E, Hess J, Kors JA. New normal limits for the paediatric electrocardiogram. *Eur Heart J*. 2001;22(8):702-711.
- Bogers AJ, Akkersdijk GP, de Jong PL, et al. Results of primary two-patch repair of complete atrioventricular septal defect. *Eur J Cardiothorac Surg*. 2000;18(4):473-479.
- Ten Harkel AD, Cromme-Dijkhuis AH, Heinerman BC, Hop WC, Bogers AJ. Development of left atrioventricular valve regurgitation

- after correction of atrioventricular septal defect. *Ann Thorac Surg.* 2005;79(2):607-612.
21. Culpepper W, Kolff J, Lin CY, et al. Complete common atrioventricular canal in infancy—surgical repair and postoperative hemodynamics. *Circulation.* 1978;58(3 Pt 1):550-558.
 22. Roos-Hesselink JW, Meijboom FJ, Spitaels SE, et al. Outcome of patients after surgical closure of ventricular septal defect at young age: longitudinal follow-up of 22–34 years. *Eur Heart J.* 2004;25(12):1057-1062.
 23. Meijboom F, Szatmari A, Utens E, et al. Long-term follow-up after surgical closure of ventricular septal defect in infancy and childhood. *J Am Coll Cardiol.* 1994;24(5):1358-1364.
 24. Goldman BS, Williams WG, Hill T, et al. Permanent cardiac pacing after open heart surgery: congenital heart disease. *Pacing Clin Electrophysiol.* 1985;8(5):732-739.
 25. Jensen PN, Gronroos NN, Chen LY, et al. Incidence of and risk factors for sick sinus syndrome in the general population. *J Am Coll Cardiol.* 2014;64(6):531-538.
 26. Clark EB, Kugler JD. Preoperative secundum atrial septal defect with coexisting sinus node and atrioventricular node dysfunction. *Circulation.* 1982;65(5):976-980.
 27. Morton JB, Sanders P, Vohra JK, et al. Effect of chronic right atrial stretch on atrial electrical remodeling in patients with an atrial septal defect. *Circulation.* 2003;107(13):1775-1782.
 28. Khairy P, Marelli AJ. Clinical use of electrocardiography in adults with congenital heart disease. *Circulation.* 2007;116(23):2734-2746.
 29. Moran AM, Daebritz S, Keane JF, Mayer JE. Surgical management of mitral regurgitation after repair of endocardial cushion defects: early and midterm results. *Circulation.* 2000;102(19 suppl 3):III160-5.
 30. McCulley DJ, Black BL. Transcription factor pathways and congenital heart disease. *Curr Top Dev Biol.* 2012;100:253-277.
 31. Chow GV, Marine JE, Fleg JL. Epidemiology of arrhythmias and conduction disorders in older adults. *Clin Geriatr Med.* 2012;28(4):539-553.
 32. Kanter RJ, Garson A, Jr. Atrial arrhythmias during chronic follow-up of surgery for complex congenital heart disease. *Pacing Clin Electrophysiol.* 1997;20(2 Pt 2):502-511.

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