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Original Article

Arrhythmic Burden of Adult Survivors With Repaired Total Anomalous Pulmonary Venous Connection

Mariama Touray, MD,[a] Magalie Ladouceur, MD, PhD,[b] Judith Bouchardy, MD, PD MER,[a,c] Markus Schwerzmann, Prof. MD,[d] Matthias Greutmann, Prof. PD MD,[e] Daniel Tobler, Prof. MD,[f] Reto Engel, MD,[g] Harald Gabriel, Prof. MD,[h] Etienne Pruvot, Prof. MD,[a] Coralie Blanche, MD,[c] Nicole Sekarski, Prof. MD,[i] and Tobias Rutz, MD, PD MER[a]

[a] *Service of Cardiology, Heart and Vessel Department, Lausanne University Hospital and University of Lausanne, Lausanne, Switzerland*

[b] *Adult Congenital Heart Disease Unit, Department of Cardiology, Hôpitaux de Paris, Hôpital Européen Georges Pompidou, Centre de référence des Malformations Cardiaques Congénitales Complexes, M3C, Paris, France*

[c] *Cardiology Unit, University Hospitals of Geneva, Geneva, Switzerland*

[d] *Department of Cardiology, Center for Congenital Heart Disease, Inselspital, University of Bern, Bern, Switzerland*

[e] *Department of Cardiology, University Heart Center, University of Zurich, Zurich, Switzerland*

[f] *Department of Cardiology, University Hospital of Basel, University of Basel, Basel, Switzerland*

[g] *Cardiology, Kantonsspital St. Gallen, St. Gallen, Switzerland*

[h] *Department of Cardiology, Vienna General Hospital, Medical University of Vienna, Vienna, Austria*

[i] *Paediatric Cardiology Unit, Women-Mother-Child Department, Lausanne University Hospital and University of Lausanne, Lausanne, Switzerland*

ABSTRACT

Background: The long-term outcome of adults with repaired total anomalous pulmonary venous connection (TAPVC) is poorly documented. Therefore, the present study aims to provide current clinical data on adult survivors with repaired TAPVC focusing on arrhythmia.

Methods: Clinical and imaging data (prevalence and type of arrhythmias, symptoms, surgical and medical treatment, echocardiographic and cardiac magnetic resonance haemodynamic parameters) were retrospectively collected from 8 European centres and compared between patients with and without arrhythmias.

Results: Fifty-seven patients were included (age 20 [16-67] years [female 28, 49%]). At the last follow-up, that is, 21 (8-51) years after surgery, 79% and 93% of patients were free of symptoms and cardiac medication, respectively. The prevalence of late arrhythmias was 21%;

RÉSUMÉ

Contexte : Les résultats de santé à long terme chez les adultes ayant subi la correction d'un retour veineux pulmonaire anormal total (RVPAT) sont mal connus. Notre étude vise donc à recueillir des données cliniques au sujet des patients ayant subi cette intervention et ayant survécu jusqu'à l'âge adulte, en particulier pour ce qui est des arythmies.

Méthodologie : Les données d'observation clinique et d'imagerie (la prévalence et le type d'arythmies, les symptômes, les traitements chirurgicaux et médicaux, et les paramètres hémodynamiques obtenus par échographie et par résonance magnétique cardiaque) ont été recueillies de façon rétrospective dans huit centres européens et comparées selon que les patients présentaient ou non une arythmie.

Résultats : Cinquante-sept patients ont été retenus (âge médian : 20 [16-67] ans; 28 [49 %] femmes). Au dernier suivi, soit 21 (8-51) ans

Total anomalous pulmonary venous connection (TAPVC) is a rare and complex cyanotic congenital heart defect[1] representing approximately 1%-3% of congenital heart disease (CHD) lesion.[2,3] This pathology is defined by all pulmonary veins aberrantly connecting to the right atrium or a systemic vein. The Darling classification separates TAPVC into 4 anatomic groups, depending on the site of the anomalous connection of the pulmonary veins: supracardiac, cardiac, infracardiac, and mixed.[4] TAPVC is often an isolated anomaly. Approximately 30% of patients with TAPVC have associated complex CHD such as a univentricular heart or isomerism.[5]

Mortality is high without immediate surgical repair with approximately 80% of patients dying during the first year after birth.[6] Advances in surgical techniques have drastically reduced mortality, enabling the majority of patients to reach

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Corresponding author: Dr Tobias Rutz, Service of Cardiology, Lausanne University Hospital, Rue du Bugnon 46, 1011 Lausanne, Switzerland. Tel.: +41-21 314 48 00; fax: +41-21 314 00 13

E-mail: tobias.rutz@chuv.ch

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9 (16%) patients showed intra-atrial re-entrant tachycardia (IART) and 2 (4%) ventricular arrhythmias. Patients with IART were older ($P = 0.018$) and 4 (7%) required antiarrhythmic medication. Three patients (5%) underwent an electrophysiological study, and another 3 (5%) underwent pacemaker implantation within 36 months after surgical correction, which were removed in 2 patients after 7 years. Early postoperative arrhythmias ($P = 0.005$), right ventricular dilatation ($P = 0.003$), and valvulopathy ($P = 0.009$) were more often present in patients with late IART.

Conclusions: Adult survivors after isolated-TAPVC repair presented a high prevalence of arrhythmias. Age, right ventricular dilatation, early arrhythmias, and valvular lesions are risk factors for IART. Long-term follow-up is important as some of these currently asymptomatic patients will probably develop arrhythmias in the future.

après l'intervention chirurgicale, 79 % des patients ne présentaient pas de symptômes et 93 % des patients ne prenaient pas de médicaments pour des troubles cardiaques. La prévalence d'arythmies tardives s'élevait à 21 %; neuf patients (16 %) présentaient une tachycardie par réentrée intra-atriale (TRIA) et deux patients (4 %) présentaient des arythmies ventriculaires. Les patients qui présentaient une TRIA étaient plus âgés ($P = 0,018$) et quatre d'entre eux (7 %) devaient prendre des médicaments antiarythmiques. Trois patients (5 %) avaient subi des études électrophysiologiques et trois autres patients (5 %) avaient subi l'implantation d'un stimulateur cardiaque au cours des 36 mois suivant la correction chirurgicale; le stimulateur cardiaque a été retiré sept ans plus tard dans deux de ces cas. Les arythmies postopératoires précoces ($P = 0,005$), la dilation du ventricule droit ($P = 0,003$) et la valvulopathie ($P = 0,009$) étaient plus fréquentes chez les patients qui présentaient une TRIA tardive.

Conclusions : La prévalence d'arythmies chez les patients survivant jusqu'à l'âge adulte après la correction isolée d'un RVPAT était élevée. L'âge, la dilation du ventricule droit, les arythmies précoces et les lésions valvulaires sont des facteurs de risque de TRIA. Il est important d'effectuer un suivi à long terme des patients ayant subi une RVPAT puisque certains d'entre eux, asymptomatiques pour le moment, présenteront sans doute des arythmies dans les années à venir.

adulthood.[7] Risk factors for a worse outcome are complex forms of TAPVC such as atrial isomerism, single ventricle, infracardiac, mixed anatomic variants and preoperative pulmonary venous obstruction, and younger age at surgery.[8] The morphologic heterogeneity of TAPVC illustrates the complexity and existence of various surgical techniques.[9]

Contemporary studies have illustrated an acceptable outcome after repair of isolated TAPVC.[8,9] Survival rates observed 3 years after surgery to adulthood range from 73.9% to 97%.[8-11]

Most studies on outcome focus on paediatric patients; literature addressing adult patients with TAPVC is, however, scarce.[11]

Frequent complications in patients late after CHD repair are arrhythmias that have an important impact on morbidity and mortality.[12-14] The prevalence of arrhythmias is higher in patients with CHD than the general population, particularly in patients with complex cyanotic CHD.[12] The aim of the present study was therefore to retrospectively evaluate the prevalence of late complications focusing on arrhythmias in adult survivors after isolated-TAPVC repair.

Materials and Methods

All adult patients diagnosed with TAPVC and included from centres participating in the Swiss Adult Congenital Heart Registry (6 Swiss centres, 1 Austrian centre) and 1 further centre in France were enrolled in the study.[15] A researcher (MT) visited all 8 centres to collect reports from the pre-, perioperative period and at the last follow-up. Patients with associated complex CHD lesions (eg, single ventricle, atrial isomerism, or transposition of the great arteries) were excluded because of the additional impact of those lesions on the general outcome. Outpatient and inpatient reports, electrocardiogram (ECG) and Holter-ECG, surgical notes, reports from cardiac magnetic resonance (CMR) and computer tomography examinations, cardiac catheterization, echocardiography, exercise testing, and cardiopulmonary exercise testing, were obtained for each patient when available. The study protocol was approved by the leading (Swiss Adult Congenital Heart Registry centres) and local ethics committees (Paris, Vienna), and the need for informed consent was waived.

Haemodynamic parameters

Haemodynamic parameters were collected retrospectively through medical reports for all patients. Cardiac shunt was defined by the pulmonary-systemic blood flow ratio determined from echocardiography, cardiac catheterization, or CMR reports. Pulmonary hypertension was defined by an estimated systolic pulmonary artery pressure of $\geq$36 mm Hg.[16-18] The left (LV) and right ventricular (RV) dimensions and volumes were obtained from echocardiography and CMR reports, respectively. The RV and LV were considered dilated when the volume or end-diastolic diameter exceeded defined norms according to published CMR and echocardiography reference values.[19,20] The RV systolic function on echocardiography was considered abnormal if the tricuspid annular plane systolic excursion was $<$16 mm, the pulsed Doppler peak S′ wave $<$10 m/s, or if described as impaired by visual assessement.[17] A CMR indexed RV ejection fraction $<$51%/52% was considered reduced for men and women, respectively.[21] An echocardiographic LV ejection fraction was considered normal above 50%.[19] A CMR LV ejection fraction $<$52%/50% was considered reduced for men and women, respectively.[20]

Information on obstruction of the pulmonary venous return was obtained from echocardiography, CMR, or angiography. An obstruction was considered significant when exceeding a velocity of $>$1.6 m/s by Doppler echocardiography or when the diameter of the pulmonary vein was reduced by 50%.[22,23]

Arrhythmias were assessed through resting ECG, Holter-ECG report, cardiopulmonary exercise testing, and electrophysiological study (EPS) documented during follow-up.

Supraventricular arrhythmia consisted of sinus node dysfunction, atrial fibrillation, or intra-atrial re-entrant tachycardia (IART), which includes atrial flutter and atrial tachycardia. Ventricular arrhythmias (VA) were characterized by sustained and nonsustained tachycardia (VT). Arrhythmias were differentiated according to the time of presentation: preoperative, early postoperative (within 30 days), and late postoperative (after 30 days) arrhythmias. Information on treatment of arrhythmias was obtained: medication, ablation during EPS, or pacemaker implantation.

Statistical analysis

Results are expressed in frequencies, means and standard deviation, median, and ranges where appropriate.

Normal distribution for continuous parameters was evaluated by the Shapiro-Wilk test. The Student t test or the Mann-Whitney U test was used, where appropriate, for comparison of continuous parameters between groups of patients with and without arrhythmia. The χ^2 test was used for dichotomous variables to compare patients with IART with those without. It was not possible to take into consideration the type of surgical procedure for the assessment of predictive factors of arrhythmia due to the heterogeneity and lack of data. The P value for statistical significance was set to $P < 0.05$. The software SPSS was used (version 25; IBM, Chicago, IL).

Results

Patients' characteristics and clinical data on latest follow-up

A total of 57 adult patients born between 1950 and 2000 were identified. Patients' characteristics are summarized in Table 1. Patients underwent surgery during the neonatal period at a median age of 1 month (range: 1 day to 14 years). One patient, currently 70 years old, presented with atrial septal defect (ASD) and pulmonary stenosis and was operated at the age of 14 years as she was initially considered not suitable for surgical correction in the 1960s.

Latest follow-up was at a median of 21 (8-51) years after surgery. The majority (79%) of patients were asymptomatic and free (93%) of cardiac medication (Table 1). All patients on medication (n = 4, 7%) were treated for IART (Table 1). In patients diagnosed with IART, 1 patient was treated with oral anticoagulation (Table 1).

The latest results of echocardiography and CMR are shown in Tables 2 and 3, respectively. Thirty percent of patients had an exercise test during follow-up, which is described in Table 4.

Nine (16%) patients required reintervention 8 ± 5 years after initial repair surgery. Noticeably, the need for reintervention did not occur in adulthood. The main indication for reintervention was the presence of stenosis of a pulmonary vein, which occurred in 5 (9%) patients. Two (4%) individuals had reintervention for a residual ASD. The other 2 (4%) patients had a reintervention because of a persistent right to left shunt: 1 patient had subhepatic veins connected to the left atrium and the other inferior vena cava to the left atrium.

Table 1. Patients' characteristics

Characteristics	Patients with total anomalous pulmonary venous connection (n = 57)
Female	28 (49)
Type of TAPVC	
Supracardiac	26 (46)
Cardiac	6 (11)
Infracardiac	11 (19)
Unknown	14 (25)
Associated lesions	
None/unknown	17 (29)
Ductus arteriosus	5 (9)
VSD	2 (4)
PLSVC	1 (2)
ASD	32 (56)
Pulmonary venous obstruction preoperatively	14 (25)
Preoperative complications	
Pulmonary hypertension	10 (18)
Valvulopathy*	6 (11)
Arrhythmia	0
Median age at correction (mo)	1 (1 d-14 y)
Median time since surgery (y)	21 (8-51)
Median age at latest follow-up (y)	20 (16-67)
Symptomatic at latest follow-up	12 (21)
NYHA class II	4 (7)
Palpitations	6 (11)
Chest pain	5 (9)
Treatment at latest follow-up	4 (7)
Beta-blocker	3 (5)
Calcium channel blocker	1 (2)
Anticoagulation	1 (2)

Data are expressed in n (%) and median (interquartile range).

ASD, atrial septal defect; PLSVC, persistent left superior vena cava; TAPVC, total anomalous pulmonary venous connection; VSD, ventricular septal defect.

*Tricuspid regurgitation in 3 (1 severe and 2 unknown), severe mitral regurgitation in 1, pulmonary stenosis in 1, and unknown in 1.

Early arrhythmias

During the early postoperative period, 5 (9%) patients developed the total of 7 arrhythmias: IART (n = 3, 5%), sinus node dysfunction (n = 2, 4%), third-degree atrioventricular block (AV block, n = 1, 2%), and sustained VT (n = 1, 2%). Supplemental Table S1 describes the arrhythmias and their evolution during the follow-up of these 5 patients. Interestingly, all patients requiring pacemaker implantation in our population presented either sinus node dysfunction (2 patients) or third-degree AV block (1 patient) within the first month after surgical correction.

Late arrhythmias

Late arrhythmias were observed in 12 (21%) patients during follow-up (Supplemental Table S2). Seventeen percent of patients with arrhythmia were symptomatic at latest follow-up (n = 2, 17%). IARTs occurred in 9 (16%) patients, of whom 3 also showed bradyarrhythmias. Two further patients presented with isolated bradyarrhythmias. Among the total of 5 patients with bradyarrhythmias, 3 (5%) required a pacemaker implantation. The first patient had an epicardial pacemaker implantation 2 months after surgery for sinus node dysfunction. This device was explanted 6 years later (reasons

Table 2. Latest available echocardiography

Echocardiographic parameters	Patients with total anomalous pulmonary venous connection (n = 57)
Echocardiography	54
Median time after initial surgery (y)	21 (2 mo-52 y)
LVEF %	62 ± 6
RV dilatation	5 (9)
RV visual dysfunction	3 (6)
Left to right shunt*	5 (9)
Valvulopathy†	3 (6)
S wave (cm/s)	9 ± 2
TAPSE (mm)	15 ± 2
Median systolic pulmonary artery pressure (mm Hg)	25 (19-51)

Data are mean ± standard deviation or n (%).

LVEF, left ventricular ejection fraction; RV, right ventricle; TAPSE, tricuspid annular plane systolic excursion.

*Persistent pulmonary vein anomaly connected in 3 patients.

†Tricuspid regurgitation in 2 (mild in one and unknown in another) and moderate pulmonary regurgitation in 1.

unkown due to lack of data). After pacemaker explantation, this patient maintained a junctional rhythm through follow-up. The second patient received an epicardial pacemaker 1 month after TAPVC repair due to a third-degree AV block. Two years after implantation, a permanent sinus rhythm was observed. The pacemaker was removed 7 years after implantation due to atrial and ventricular lead fracture. After explantation, episodes of IART were detected 21 years after surgery. This patient underwent EPS with successful ablation (for details please see the Electrophysiological studies section below). The third patient requiring pacemaker implantation was diagnosed with sinus node dysfunction within the first month after surgical correction. A pacemaker was implanted 36 months after surgery. Battery replacement was required 9 years after implantation.

No atrial fibrillation was observed. Two (4%) patients presented with VA that consisted in nonsustained VT (Supplemental Table S2).

Table 3. Latest available cardiac magnetic resonance

Parameters	Patients with total anomalous pulmonary venous connection (n = 57)
CMR	14
Median time after initial surgery (y)	20 (15-51)
LV size (mL/m^2)	94 ± 18
RV size (mL/m^2)	114 ± 25
Dilated RV	4 (29)
Shunt	2 (14)
LVEF (%)	61 ± 6
RVEF (%)	52.5 ± 9
Stenosis of the pulmonary venous return	1 (7)

Data are mean ± standard deviation or n (%).

CMR, cardiac magnetic resonance; LEVF, left ventricular ejection fraction; LV, left ventricle; RV, right ventricle; RVEF right ventricular ejection fraction.

Table 4. Latest available exercise test/cardiopulmonary exercise testing

Parameters	Patients with total anomalous pulmonary venous connection (n = 57)
Exercise testing (%)	17 (30)
Median time after initial surgery (y)	21 (10-68)
VO2 max (mL/kg/min) (n = 9)	35 ± 9
VO2 max, % (n = 9)	92 ± 16
VE/VCO2 slope (n = 7)	26 ± 3
BPM, % predicted (n = 13)	94 ± 11
MET, % predicted (n = 12)	104 ± 21

Data are mean ± standard deviation or n (%).

BPM, beat per minute; MET, metabolic equivalent of task.

Clinical variables related to IART

Table 5 shows comparison of parameters between patients with and without IART. IARTs were more frequently observed in patients who had presented early postoperative arrhythmias as well as RV dilatation or valvulopathies on latest echocardiography. Patients with IART were significantly older than those without (Table 5). Residual shunt, tricuspid annular plane systolic excursion, S wave, and pulmonary artery pressure were not related to IART. No IARTs were observed in patients who required reintervention during childhood.

Noticeably, 1 patient with late IART was operated at the age of 14 years and was 68 years old at latest follow-up (patient 5 in Supplemental Table S2). When excluding this particular patient from the statistical analysis, older age, early postoperative arrhythmias, and RV dilatation but not valvulopathies are more significantly observed in patients with IART.

Electrophysiological studies

Three patients (5%) underwent an EPS 20 years after surgery for IART in 2 patients and for nonsustained VT in 1 patient. The first patient had developed asymptomatic arrhythmia in the first month after surgery that consisted of nonsustained atrial tachycardia and ventricular extrasystoles that persisted through follow-up. The EPS failed to reproduce IART with isoproterenol and ajmaline provocation test. Moreover, the EPS showed polymorphic ventricular extrasystoles that did not originate from the outflow tracts. No ablation was performed. The second patient had previously undergone pacemaker implantation with subsequent removal and underwent an EPS and ablation of IART 21 years after surgical repair of TAPVC. Two circuits, 1 in the basal and 1 in the posterolateral part of the right atrium, were identified and successfully ablated. Electroanatomic mapping revealed significant scaring of both atria. No recurrence of IART occurred during follow-up. The third patient underwent EPS for nonsustained VT. Ventricular stimulation and isoproterenol provocation testing failed to provoke VT. An idiopathic nonsustained VT was concluded.

Table 5. Clinical and echocardiographic parameters associated with IART

	With IART (n = 9)	Without IART (n = 48)	*P* value
Median age at latest follow-up (y)	25.5 (16-67)	20 (16-41)	0.011
Early postoperative arrhythmias	3 (33)	2 (4)	0.005
Symptomatic at latest follow-up	2 (22)	10 (21)	0.925
RV dilatation*	3 (33)	2 (4)	0.003
Valvulopathy*	2 (22)	1 (2)	0.009
LVEF (%)*	64 ± 9	61 ± 5	0.6490
S wave (cm/s)*	8 ± 3	9 ± 2	0.4431
TAPSE (mm)*	14 ± 3	16 ± 2	0.2404
Median systolic pulmonary artery pressure (mm Hg)*	25 (20-51)	25 (19-30)	0.388
Presence of residual shunt*	2 (22)	3 (6)	0.120
Reintervention	0	9 (19)	0.157

Data are mean ± standard deviation or n (%).
IART, intra-atrial re-entrant tachycardia; LVEF, left ventricular ejection fraction; RV, right ventricle; TAPSE, tricuspid annular plane systolic excursion.
*Parameters obtained on latest echocardiography.

Discussion

The present study documents in adult survivors after TAPVC repair a high burden of arrhythmias, occurring in 21% of patients. The majority of arrhythmias were supraventricular arrhythmias and particularly IART, which are typically observed in patients with CHD.[12,24,25] We identify age, RV dilatation, early postoperative arrhythmias, and valvular lesions as risk factors for the occurrence of late IART.

A strength of our study is the information on outcome in adult patients. Studies are extremely scarce in this patient population due to the rarity of the defect and have almost exclusively been conducted in paediatric populations.[26-30] In 1991, Saxena et al.[28] described the presence of significant arrhythmias in 6 of 16 (37%) children, on 24-hour ECG monitoring at a mean time of 8 years after isolated-TAPVC repair. The arrhythmias noted were supraventricular tachycardia (n = 3), bradyarrhythmia (n = 2), sick sinus syndrome (n = 2), and multiform supraventricular and ventricular ectopic beats (n = 2).[28]

Tanel et al.[30] described sinus node dysfunction in most of the children with repaired isolated TAPVC, and rarely VA with no predictors found requiring no particular intervention.

Korbmacher et al.[29] described arrhythmias in a 11-year follow-up among 24 patients including children and adults. All 24 TAPVC survivors had a 24-hour ECG monitoring that revealed significant arrhythmia in 11 (46%), sinus node dysfunction but also VA. Predictors were type of incision and the site of cardiopulmonary bypass cannulation.[29]

Our present study has a lower prevalence of arrhythmias than the studies mentioned above, which probably could be explained by the absence of routine use of 24-hour ECG monitoring and the exclusion of patients with associated complex lesions.

The genesis of late arrhythmias in these patients is not yet fully elucidated and is probably multifactorial. As described for patients with ASD, prolonged volume overload, pulmonary hypertension, ventricular dysfunction, and atrial stretch are factors that contribute to the development of arrhythmias.[31] However, in contrast to patients with ASD, patients with TAPVC undergo surgery early in infancy and with rapid restoration of the normal haemodynamic situation. In the present study, persistent haemodynamic lesions after repair (valvulopathies and RV dilatation) were related to a higher prevalence of IART. Our observations confirm for this very particular population similar risk factors for IART as has previously been described for the general CHD population.[14] Ávila et al.[14] identified among others previous intracardiac repair, pulmonary hypertension, pulmonary regurgitation, subpulmonary AV valve regurgitation, and subaortic and subpulmonary ventricular dysfunction as predictors of IART in CHD.

Previous cardiac surgery is known to increase the risk for arrhythmia, due to fibrotic scar formation at the incision or cannulation site.[32] However, in our population, we were unable to demonstrate that, for example, surgical repair techniques were related to arrhythmias, which is certainly due to the population's heterogeneity and incomplete surgical reports.

Nonetheless, one patient in our study population presented with IART originating from scar tissues in the right atrium presumably surgical scars, demonstrated on EPS with atrial mapping.

IARTs are related to an increased morbidity, mortality, and sudden cardiac death in patients with complex CHD.[32] Treatment generally consists of antiarrhythmic and/or ablative treatment as illustrated in the present study.

Interestingly, in our study, only one patient with IART was on anticoagulation, which is surprising as IARTs are considered a risk factor for thromboembolic events, and the latest guidelines propose to consider anticoagulation in patients with moderate or complex CHD.[33]

Our study interestingly reveals 2 patients with VA, which is in line with previous studies.[28-30] These observations are concerning because of the potential increased risk of sudden death in this situation.

The understanding of VA in TAPVC is more complex. Typically, VA in CHD are known for patients, that is, after tetralogy of Fallot repair, in whom risk factors such as age at repair, prior shunt, residual lesions, RV and LV dysfunction, and consequences of ventricular repair are relatively well defined.[13] In patients with TAPVC in contrast, the surgical repair usually does not involve the ventricle and one could speculate fibrotic ventricular tissue remodelling due to cyanosis and volume overload contributing to the development of VA. Both patients with VA in our study had an

excellent exercise capacity with an average of 126% of predicted metabolic equivalent of task and normal haemodynamic parameters on echocardiography and CMR. One patient, with preserved ventricular function and discrete late septal enhancement on CMR, had EPS for asymptomatic polymorphic ventricular and atrial ectopic beats. The treatment of VA remains challenging, and identifying patients who could benefit from catheter ablation without implantable cardioverter-defibrillator therapy remains challenging and needs further research.[34]

Limitations

This retrospective study was limited by the relatively small number of patients due to the rareness of the pathology despite including patients from the total of 8 European centres. Moreover, TAPVC being heterogeneous and complex, most patients had an individualized care management depending on centres' preferences and protocols (eg, surgical procedures, use of Holter-ECG, exercise testing, CMR imaging, etc. during follow-up). Usually, patients underwent at least an echocardiography and ECG during their routine follow-up examinations.

The main limitation of this study is therefore the heterogeneous data. We also acknowledge the lack of data particularly concerning the indication for pacemaker implantation and electrophysiological studies. There is a clear need of prospective studies in this population.

Moreover, the present study reports associations related to late arrhythmia and not necessarily risk factors. Indeed, late arrhythmias occurred for some patients before the last follow-up. Thus, the latest haemodynamic parameters (RV dilatation, valvulopathy, etc.) reported in this study may already be a consequence of late arrhythmia in contrast to, for example, early arrhythmia, which we can consider a risk factor for future events.

The relatively small number of adult patients could also be explained by a loss of patients during the transition phase from paediatric to adulthood. Indeed, as most patients with isolated TAPVC have an excellent long-term outcome, they may therefore not feel the need for continuing their follow-up during adulthood. The prevalence of arrhythmia in our study may consequently not reflect the overall prevalence in this population. However, as age is related to the development of arrhythmias, we want to emphasize the importance of follow-up especially in these arising young adults.

Conclusions

Adult patients with isolated-TAPVC repair have a high prevalence of arrhythmias, which mainly consist of IART. IART appears to be related to RV dilatation, valvular lesions, presence of early postoperative arrhythmias, and age. This study underlies the importance of long-term follow-up as some of the currently asymptomatic patients will probably develop arrhythmias in the future.

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Ethics Statement

The study protocol was approved by the leading (Swiss Adult Congenital Heart Registry centres) and local ethics committees (Paris, Vienna).

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Pediatric and Congenital Heart Disease* at https://www.cjcpc.ca// and at https://doi.org/10.1016/j.cjcpc.2022.08.003