An Unmet Need: Arrhythmia Detection by Implantable Loop Recorder in the Systemic Right Ventricle

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Structured Abstract

Background and Aims: Patients with systemic right ventricles are at high risk of sudden cardiac death. Arrhythmia is a significant risk factor. Routine Holter monitoring is opportunistic with poor adherence. Continuous rhythm monitoring with an implantable loop recorder (ILR) could allow early detection of clinically important arrhythmias.

Methods: ILR implantation was offered to patients with atrial switch repair for transposition of the great arteries. Recordings were made with symptoms or automatically for pauses, significant bradycardia or tachycardia and reviewed by the muti-disciplinary team.

Results: 24/36 eligible patients underwent ILR implantation with no complication. 42% had preserved ventricular function, 75% were NYHA functional class I, 88% had low sudden cardiac death risk. 33% had previous IART and none had known conduction disease. 18/24 (75%) patients made 52 recordings (52% automated) over 39.5 months (1.6 –72.5). 32/52 (62%) recordings in 15/24 (63%) of the cohort were clinically significant and included sinus node disease (2 patients), atrio-ventricular block (2 patients), intra-atrial re-entry tachycardia [IART] (7 patients) and IART with sinus node disease or atrio-ventricular block (4 patients). ILR recordings prompted medication change in 11 patients [beta blockers (n=9), anti-coagulation (n=5), stopping anticoagulation (n=1)] and device therapy recommendation in 7 patients [5 pacemakers (3: atrioventricular block), 2 defibrillators]. 2 patients declined intervention; one suffered an arrhythmic death. IART and clinically relevant conduction disease were detected in patients irrespective of sudden cardiac death risk.

Conclusion: Continuous monitoring with an ILR in patients with systemic right ventricle following atrial switch detects clinically relevant arrhythmias that impact decision-making. In this cohort, clinically relevant arrhythmias did not correlate with sudden cardiac death risk.

Keywords: Transposition of the great arteries, systemic right ventricle, arrhythmia surveillance, sudden cardiac death, implantable loop recorder

What's New?

Adults with atrial switch repair for transposition of the great arteries are 50 times more likely to experience sudden cardiac death than the general population. Arrhythmia contributes significantly to sudden death and incipient heart failure although mechanisms are unclear.

- Implantable loop recorders [ILR] offer a convenient, minimally-invasive strategy for continuous rhythm monitoring to enable earlier detection and intervention than conventional ECG and Holter screening.
- In this low-risk cohort of adults with systemic right ventricle after atrial switch, 66% had clinically significant arrhythmia detected by ILR [atrial tachyarrhythmia and/or conduction disease] which changed management in 63% [medication, pacemakers, defibrillators]. Conventional surveillance and risk scoring would not have identified these issues in the same timeframe.
- The prognostic impact of early intervention is unclear, however, continuous monitoring with ILR in this population is feasible and safe and impacts clinical decision making. Further studies should determine prognostic benefit and cost-effectiveness.

INTRODUCTION

Sudden cardiac death is a serious concern for adults following atrial switch for patients with transposition of the great arteries (TGA), with an incidence between 1.7 – 9.5 per 1000 patient-years [1]. This represents a 50 times increased risk compared with the age matched general population [2] and 10 times increased risk compared with young people with hypertrophic cardiomyopathy [3]. After heart failure, sudden death is the second most common cause of mortality associated with this physiology [4] and an unresolved issue despite advances in surgical and heart failure management. A better strategy to identify patients at risk of sudden cardiac death is of paramount importance particularly with the predicted growth in heart failure and arrhythmia within this ageing population. The precise mechanism of sudden cardiac death is often unclear [1], however arrhythmias, particularly intra-atrial re-entrant tachycardia (IART), are amongst the strongest identified risk factor [5-7]. Early recognition and treatment of IART may preserve cardiac output, avoid myocardial ischaemia, reduce the risk of ventricular arrhythmias and prompt anticoagulation to reduce the considerable risk of thromboembolism [1, 6, 8, 9]. It remains unclear whether conduction disease contributes to sudden cardiac death within this physiology although it is an established risk factor within the structurally normal heart [10, 11]. A recently developed risk score is promising in identifying those at greatest risk who may benefit from a primary prevention intracardiac defibrillator (ICD) but did not find conduction disease to be an independent predictor [4]. However, arrhythmias including IART and conduction disease, may carry a significant burden of morbidity and potential mortality irrespective of risk scores [5-7].

The current strategy of rhythm surveillance with ECGs and intermittent ambulatory monitoring has significant limitations related to its short temporal coverage. It frequently fails to detect paroxysmal IART [12], offers a narrow window for symptom-rhythm correlation and has poor adherence within this population (supplementary table 2). Furthermore, there is a high burden of care to the patient and health care provider in delivering this sub-optimal monitoring strategy which is expected to grow with ageing of the ACHD population. The current generation of the injectable implantable loop recorder (ILR) presents a convenient alternative for continuous rhythm monitoring. Limited data on symptom driven application in unselected congenital heart disease has shown good tolerance and a high detection of atrial and ventricular arrhythmias with clinical implications [13, 14]. ILRs have also been shown to have benefit in empirical arrhythmia surveillance in other high-risk populations such as Fabry's disease and hypertrophic cardiomyopathy [15, 16].

The aim of this study was to prospectively assess the feasibility and utility of arrhythmia detection with ILR in a single centre population of adults with systemic right ventricle following atrial switch.

METHODS

Study Population

Since 2016, all adults (> 18 years) with a biventricular circulation and systemic right ventricle following atrial switch repair (Mustard or Senning procedure) for TGA attending the ACHD clinic at Newcastle upon Tyne NHS Foundation Trust were considered for an empirical ILR

implant. Those who had subsequent heart transplant, ventricular assist device (VAD) implantation or surgical restoration of a systemic left ventricle (i.e. Rastelli procedure and arterial switch) were excluded. Final screening also excluded those with a pre-existing intra-cardiac monitoring device or were deemed excessively frail by their responsible clinician (Figure 1). Thirty-six patients were identified, and their responsible clinician discussed the rationale for ILR implantation at their routine clinical review. Twenty-four patients agreed to ILR implantation (Figure 1). All patients subsequently agreed to automatic, elective replacement of their ILR at the end of battery life.

Data Collection

Demographic, anatomical, arrhythmia history, arrhythmia (symptomatic or automatic) during follow-up, clinical action following recordings and clinical outcomes following intervention were recorded. Survival outcome at the end of the study was recorded as alive, transplant, transplant assessment, ventricular assist device or death.

ILR Implantation, Arrhythmia Detection and Management

Implantation was conducted under local anaesthetic in the usual fashion [17]. One patient underwent Medtronic Reveal XT® implantation with all subsequent patients having an injectable Medtronic Reveal Linq® in the left parasternal position. Patients were instructed to activate a recording if they experienced palpitations, dizziness or syncope. Automated recordings were made for brady-arrhythmias, tachy-arrhythmias or atrial fibrillation. Brady-arrhythmia detection was set for sinus pauses of greater than 3 seconds or bradycardia less than 30bpm for 12 consecutive beats. Tachy-arrhythmias were set to detect a suspected supraventricular tachycardia of greater than 194bpm for 16 beats.

All recordings were transmitted remotely and reviewed by a cardiac physiologist within one day. The patient's responsible clinician was informed in the event of suspected arrhythmia and management agreed in the adult congenital heart disease and electrophysiology multi-disciplinary team meeting (ACHD-EP MDT). Recommendation for intervention included review of consensus guidelines [18], expert opinion and, following publication, calculation of the risk of sudden cardiac death using the published scoring system utilising age, heart failure, syncope, right ventricular function, left ventricular outflow tract obstruction and QRS duration [4].

Ethical Considerations and Data Protection

The decision to offer ILR implantation was a clinical consensus reached in the ACHD-EP MDT after an audit of the existing practise of Holter monitoring confirmed poor adherence and thus limited arrhythmia detection. Written informed consent was taken from patients prior to ILR implantation. This study represents a re-audit following a change in clinical practise and as such did not require separate ethical approval. The audit was registered with the clinical effectiveness database of The Newcastle upon Tyne NHS Foundation Trust. Data was managed in accordance with GDPR.

Patient and Public Involvement

This study reports the results of a change in clinical practise that was instigated after audit of the recommended arrhythmia monitoring strategy for this patient group identified poor adherence. Whilst patients and the public were not directly involved in the design of this study, the results have led to subsequent engagement with the Sommerville Foundation and patient focus groups funded by the British Heart Foundation and led by the Senior Author to

understand what issues matter to patients with respect to monitoring and how this can help inform future study design.

Statistical methods

Continuous data were summarised as median and range (following confirmation of non-normality with the Shapiro-Wilk test) and categorical data as count and percentage of the whole cohort. Freedom from arrhythmia was described with the Kaplan-Meier method. Comparison of different arrhythmia events were compared with a 2 tailed log rank test. Group analysis was conducted for five variables (right ventricular function, left ventricular function, tricuspid regurgitation severity, type of surgical repair and previous arrhythmia) for dependence on survival outcome (death, transplant or VAD), detected arrythmia and future intervention using Fischer's exact test. Ventricular function and tricuspid regurgitation were grouped as normal or mild and moderate or severe as assessed by echocardiography. A p value < 0.05 was considered significant.

Analysis was conducted in GraphPad Prism® 9.5 (GraphPad Software LLC, 2022).

RESULTS

Study Population

24 patients (29–49 years) underwent ILR implantation. Half had residual cardiac lesions and 46% had additional non-cardiovascular co-morbidities. 75% were NYHA functional class 1. 42% had moderate or severe systemic right ventricular impairment and 30% had moderate or severe tricuspid regurgitation. All but one patient had baseline sinus rhythm with a third having previous documented IART and a third taking regular beta blocker therapy. None

were taking a class I or III anti-arrhythmic agent. Demographics of the study population are shown in Table 1. Most patients, 88%, had a low predicted risk of sudden cardiac death (less than 5%) (Figure 3).

ILR Implantation, Arrhythmia Detection and Management

All ILR implantations were conducted without procedural complication, and all had interpretable electrograms. Two patients had a second ILR implant following battery depletion of their first device. There were no complications related to the devices during the surveillance period. The median duration of surveillance was 39.5 months (1.6 –72.5) during which a total of 52 recordings (52% automated, 48% activated) were made in 18 patients. This equated to a median of 2.8 recordings per patient (range 0-8) or 0.5 recordings per patient-year (range 0 – 14.5).

25% (6/24) of the cohort had no arrhythmia detected during the monitoring period. Anticoagulation was stopped in one individual based on multiple sinus rhythm recordings that correlated with symptoms and patient discussion of the risk (Table 2).

61.5% of the total recordings made in the remaining patients were clinically significant (significant sinus pause, atrioventricular block or IART). ILR detected rhythms directly changed clinical management in 15/24 patients (64%) (Table 2).

IART was detected in 46% (11/24) of the cohort with half (5 of 11 patients) having no previously documented atrial tachyarrhythmia. Anticoagulation was newly commenced in five patients.

Significant conduction disease was detected in 33% (8/24), 50% co-existing with IART, with one patient having a previously documented short sinus pause. Six patients had an indication for pacing; five agreed to proceed with this intervention. One patient, with sinus pauses and non-sustained IART, preferred an initial conservative approach with ongoing ILR surveillance rather than pacing. At 1072 days post ILR implant, they experienced further pauses and atrioventricular block and then agreed to pacemaker implantation. Three patients with tachy-brady arrhythmia were taking a beta blocker which was continued following permanent pacing. None of the patients with atrioventricular block were taking a beta blocker.

Conduction disease and IART were detected over the entire surveillance period and appeared unrelated to each other in onset.

An additional patient, with pulmonary hypertension and recurrent IART, had a high predicted risk of sudden cardiac death and was offered a primary prevention ICD. The patient declined the ICD, preferring a conservative approach, and subsequently had a sustained episode of symptomatic IART resulting in a pulseless electrical activity cardiac arrest shortly after presenting to hospital. Unfortunately, the patient was not a candidate for advanced heart failure therapies and resuscitation was unsuccessful.

There was no association between the type of surgical repair, right ventricular function or tricuspid regurgitation severity with detected arrhythmia, adverse outcome or intervention. In this cohort, those with previously documented IART were not more likely to have further IART compared with those without a documented arrhythmia history ($p=0.08$). Moderate or severely impaired LV systolic function was associated with an adverse survival outcome but

not with detected arrhythmia. IART and conduction disease was detected in patients despite low sudden death risk scores (Figure 3).

Three patients had incidental ambulatory ECG monitors during ILR surveillance. None detected any arrhythmia whereas their ILR detected IART in all patients and significant conduction disease requiring eventual permanent pacing in one patient.

DISCUSSION

Empirical ILR implantation in patients with a systemic right ventricle following atrial switch repair for TGA detected a high burden of arrhythmia which resulted in a change in clinical management in the majority of patients. This is despite our cohort representing a healthy subpopulation with predominately preserved systemic RV function, near normal NYHA functional class and low predicted risk of sudden cardiac death (Table 1).

Most arrhythmias were IART with half occurring in patients without a recognised history of arrhythmia. The incidence is consistent with previous reports in cohorts with a heterogenous risk of sudden cardiac death (38% compared with 14-44%) utilising intermittent surveillance and symptom-based monitoring [4, 5], as well as ILR studies in unselected populations with congenital heart disease (30-53%) [13, 14]. As atrial arrhythmias are typically progressive in this population, it is possible that these arrhythmias would have eventually been detected by conventional monitoring. There is, however, significant potential clinical benefit to the early detection and intervention afforded by an ILR surveillance strategy. This is of particular importance given the strong relationship between IART with heart failure and sudden cardiac death in this population [1, 5-7]. The

pathophysiological mechanisms are incompletely understood however it is increasingly recognised that the association is robust and early intervention can mitigate adverse outcomes and improve quality of life [19].

Strategies to maintain sinus rhythm are limited with significant adverse effects experienced from the few anti-arrhythmic agents suitable for this population [20]. Our unit, in line with others [21], prefers early ablation as the strategy to manage sustained IART. However, recurrence is frequently observed and the prognostic value of repeated interventions, the acceptable burden of paroxysmal arrythmias and the threshold to pursue advanced heart failure therapies influence clinical decision making [19, 20, 22].

Conduction abnormalities, predominantly sinus node disease, are common following atrial switch and may promote IART, reduce cardiac output and potentially contribute to sudden cardiac death [18]. Consequently, there is an accepted lower threshold for pacing [18] however patient selection and approach remains challenging given the high incidence of device related complications [23]. Although a significant proportion of our screened cohort had a pre-existing intracardiac pacing device (32%), an additional 11% of patients were identified to have a pacing indication over the course of the study. Of note, half of these patients (three out of six) had daytime atrioventricular block, without reversible trigger, which presents a clear risk factor for sudden cardiac death. In TGA with concordant atrioventricular connection, in contrast to congenitally corrected transposition, disruption of the atrioventricular node conduction is unusual and the mechanism for block is unclear. It is possible that accelerated fibrosis following initial surgical repair and subsequent structural or electrophysiological procedural interventions are contributory. In addition, diseased conduction tissue may be particularly vulnerable to sudden changes in depolarisation that

occurs with IART and/or sinus pauses and result in prolonged tissue refractoriness resulting in atrioventricular block (type 3 and type 4 block) [24].

Conduction abnormalities were not independently associated with sudden cardiac death in a recent large retrospective analysis [4], although the cohort was limited to intermittent rhythm surveillance with ECGs and Holter monitoring. Increasing ventricular conduction delay (QRS duration) and repolarisation instability (QTc dispersion) over time can predict SCD across a spectrum of ACHD disease [25]. Whether this association is due to elevated risk of atrioventricular block, re-entrant tachycardia or purely worsening heart failure is unclear. Correlation of sequential ECG and echo studies with continuous rhythm monitoring could offer important mechanistic insight into arrhythmogenesis and sudden cardiac death, further guiding timely intervention, and is an important area for further study. The only death, thus far, in our monitored cohort was associated with IART followed by low cardiac output circulatory failure.

The alternative strategy of clinic ECGs and intermittent ambulatory ECG recordings capture short, unselected periods [12] with poor adherence within this population (Supplementary Table 1) and do not reliably detect clinically relevant arrhythmias. By comparison, ILRs were well tolerated and had a high rate of clinically significant findings. Given these findings, alongside the added value of prospective symptom-rhythm correlation and response to interventions, our unit currently recommends lifelong ILR surveillance in this population who do not have a pre-existing cardiac monitoring device. It is hoped that this will improve detection and understanding of clinically relevant arrhythmias with the potential to reduce the burden of sudden cardiac death. How those with ACHD experience monitoring and surveillance is not fully understood and is an important component that requires further

evaluation if care is to be delivered holistically as well as effectively from a clinical perspective. At this stage, costs are not fully evaluated. The incremental costs and benefits of ILR surveillance for this small cohort, should also be considered in terms of their expected lifetime healthcare costs and relative to the whole cohort of patients referred for ILR implantation. To accurately assess cost-effectiveness, economic evaluation should be incorporated into a prospective clinical study that compares this approach to usual care. Finally, we should consider the rapidly increasing diagnostic utility of wearable smart watches that is already informing clinical decision making in many clinical fields [26]. Future studies are required to investigate their use in ACHD to enable a more patient centred approach to surveillance and potentially reduce the care burden for patients and healthcare providers [27].

Limitations

This was a single centre study not sufficiently powered for robust subgroup analysis. Nevertheless, the clinical event rate was sufficiently high to be confident about the likely benefit of this approach over routine and ad-hoc monitoring based on symptoms and risk-scoring. In fact, it clearly identifies a concern over the conventional monitoring approach in ACHD guidelines in missing clinically relevant arrhythmias in low-risk individuals.

Because ambulatory monitoring had poor adherence in our cohort and in view of the high rate of sudden cardiac death, randomising patients to this option was felt not to be clinically justifiable and therefore a control group remaining in usual care was not included.

We also acknowledge an inherent selection bias given a third of potentially eligible patients refused or were not offered ILR implantation. However, based on these results, we have

subsequently re-approach this omitted group and the majority have agreed to future ILR surveillance.

Conclusion

Continuous rhythm monitoring with an ILR in patients with systemic right ventricle following atrial switch is well tolerated and detects a high burden of clinically relevant tachy- and brady-arrhythmias that impact decision-making. The existing sudden cardiac death risk scoring system did not predict arrhythmias detection in this cohort and the long-term significance of these findings are unclear.

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CONFLICT OF INTERESTS

Dr Stephen Murray has received honoraria from Acutus Medical and Boston scientific. Dr Louise Coats receives funding from the British Heart Foundation and collaborates with Cardiacsense. All remaining authors have declared no conflicts of interest.

FUNDING STATEMENT/FINANCIAL DISCLOSURE

There was no funding for this study.

DATA AVAILABILITY STATEMENT

Data will be supplied by direct request to the authors.

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Table 1 Study population.

Demographic	Median (Range)	Number (%)
Age (Years)	35 (29 – 49)	
Male		18 (75)
Mustard Repair		13 (54)
Senning Repair		11 (46)
Left ventricular outflow tract obstruction		4 (17)
Ventricular Septal Defect		5 (21)
Baffle stenosis/occlusion		5 (21)
Single coronary		2 (8)
Pulmonary hypertension (known)		2 (8)
Straddling tricuspid valve		1 (4)
Epilepsy/Prior Stroke		5 (20)
Thyroid Disease		2 (8)
Type 2 Diabetes Mellitus		1 (4)
Depression		1 (4)
Moderately/severely impaired right ventricular function		10 (42)
Moderately/severely impaired left ventricular function		2 (8)
Moderate/severe tricuspid regurgitation		7 (29)
NYHA 1		18 (75)
NYHA 2/3		6 (25)
Sinus rhythm		23 (96)
Junctional rhythm		1 (4)
QRS duration >120ms		4 (17)
Previously documented IART [ablated]		8 (33) [2(8)]
Previously documented NSVT		1 (4)
Previously documented sinus pause (<6 seconds)		1 (4)
No medication		16 (66)
Beta blocker		8 (33)

IART: Intra-atrial re-entrant tachycardia, NSVT: Non-sustained ventricular tachycardia.

Table 2 Characteristics and clinical outcomes of ILR recordings.

Recording	Number (%)	Clinical Management (Number)	Current clinical status
No recording	6 (25)	-	All alive and well
Insignificant recording	**3 (13)**		
Sinus rhythm	2 (8)	Reassurance (1) Anticoagulation stopped (1)	All alive and well
Short sinus pause (<6 seconds) ¥	1 (4)	Observation (1)	Alive and well
Conduction disease	**4 (17)**		
Long sinus pause (>6 seconds)	2 (8)	Exercise test & observation (1) PPM (1)	Alive and well Alive and well
Atrioventricular block	2 (8)	PPM (1) ICD (1)	Alive. transplant assessment Alive and well
Atrial arrhythmia	**11 (46)**		
Non-sustained IART*	2 (8)	Observation (1) Beta blocker (1)	Alive and well Further runs of non-sustained IART
Sustained IART†	5 (21)	Beta blocker (1)	Further runs of non-sustained IART
		Beta blocker, anticoagulation (1)	Further runs of non-sustained IART
		Beta blocker, declined primary prevention ICD (1)	Heart failure, IART, cardiac arrest, death
		Beta blocker, anticoagulation, ablation (1)	Heart failure, ventricular assist device, tricuspid valve replacement
		Beta blocker, anticoagulation, awaiting ablation (1)	Alive, transplant assessment
Tachy-brady arrhythmia ‡	4 (17)		
• Sinus node disease	2 (8)	Beta blocker, declined PPM (1) DCCV, beta blocker, anticoagulation, awaiting ablation (1)	Alive and well Alive. Further admission with IART expediting ablation
• Atrioventricular block	2 (8)	PPM, anticoagulation, ablation (1) Beta blocker, PPM (1)	Alive and well Alive and well

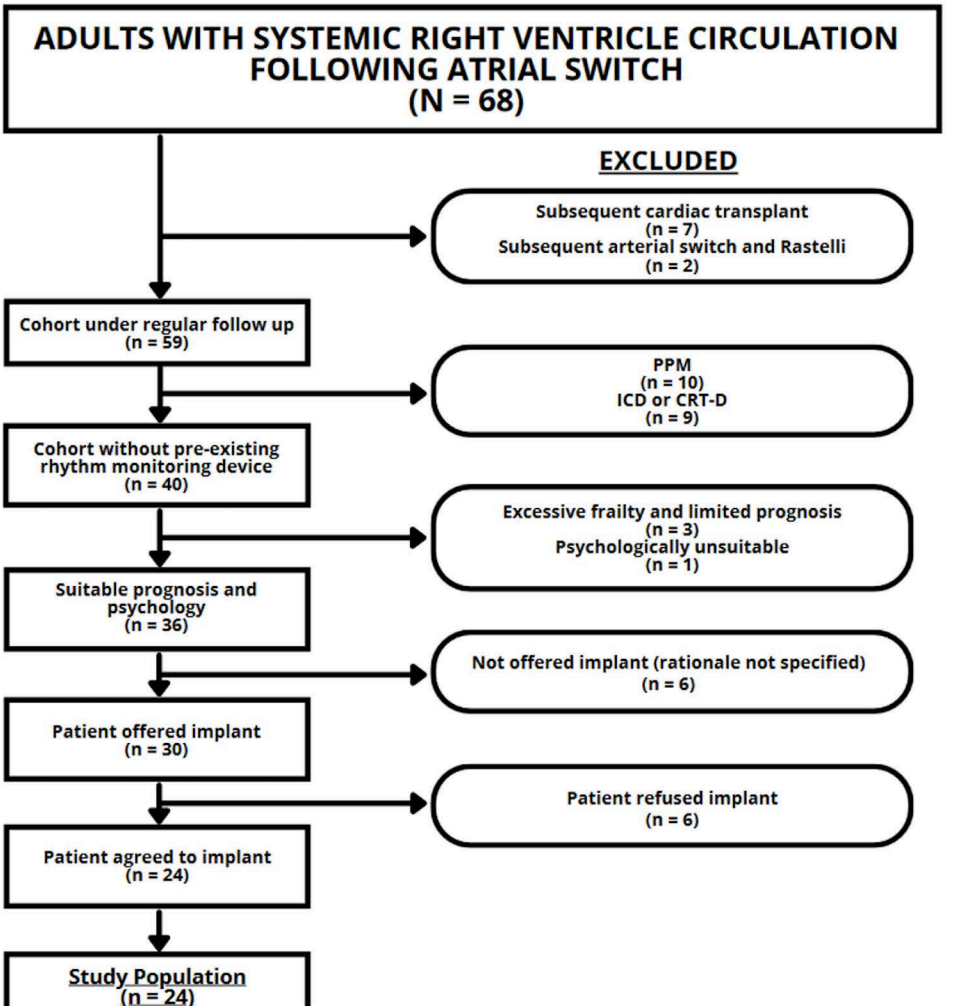
IART: Intra-atrial re-entrant tachycardia, PPM: permanent pacemaker, ICD: Implantable cardiac defibrillator, DCCV: direct current cardioversion, * regular narrow complex tachycardia lasting less than 30 seconds, † regular narrow complex tachycardia lasting 30 seconds or greater, ‡ IART and significant sinus or AV node disease recorded within the

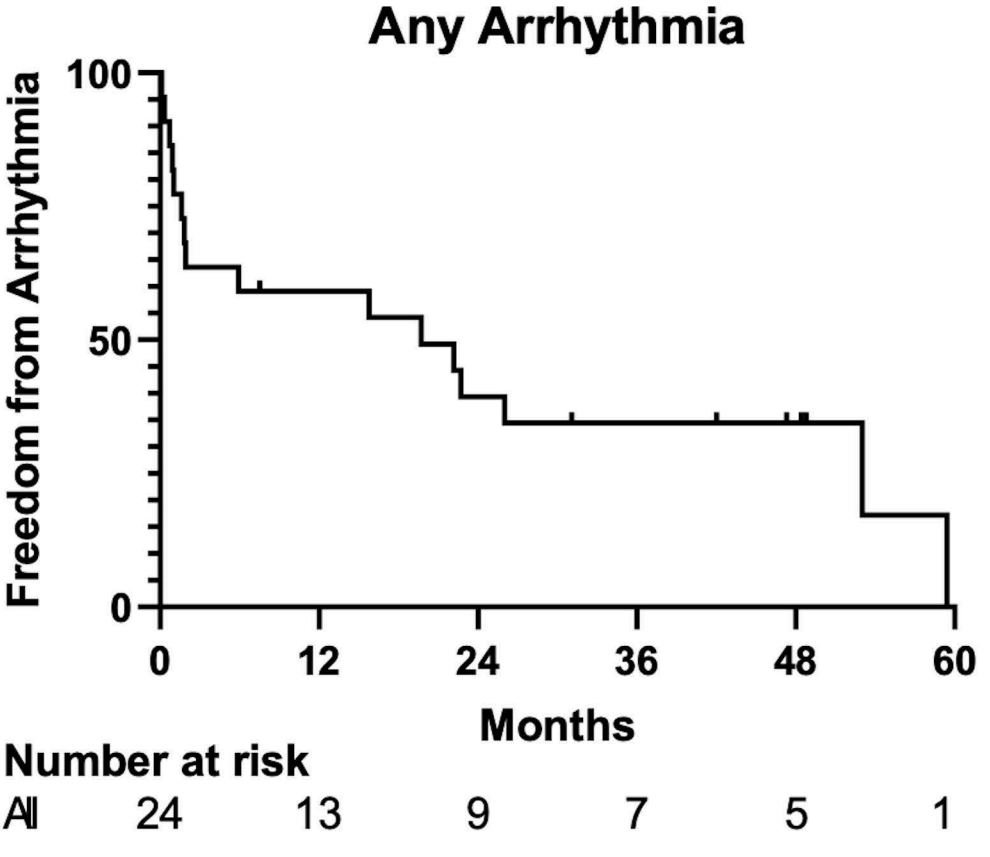
same patient. ¥Limited literature and guidelines suggest potential prognostic implication of shorter pauses in this circulation [21].

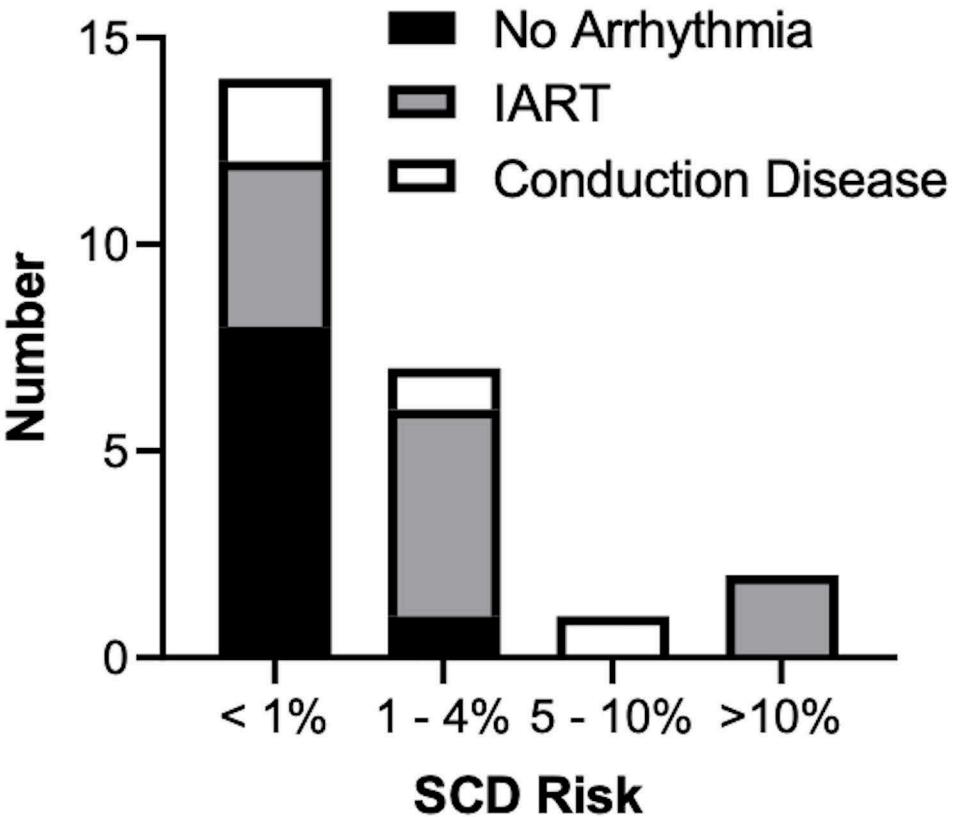
LIST OF FIGURES

Supplementary Table 1 Audit of adherence of biennial ambulatory ECG surveillance within the atrial switch with systemic right ventricle population (over a 10 year period

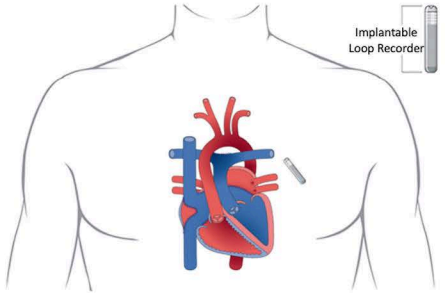
Number of patients	50
Patients who fulfilled audit criteria of biennial ambulatory monitor	0%
Patients with at least one ambulatory monitor	58%
Mean interval between ambulatory monitors (years)	5.3


ADULTS WITH SYSTEMIC RIGHT VENTRICLE CIRCULATION FOLLOWING ATRIAL SWITCH
(N = 68)
EXCLUDED
Subsequent cardiac transplant
(n = 7)
Subsequent arterial switch and Rastelli
(n = 2)
Cohort under regular follow up
(n = 59)
PPM
(n = 10)
ICD or CRT-D
(n = 9)
Cohort without pre-existing rhythm monitoring device
(n = 40)
Excessive frailty and limited prognosis
(n = 3)
Psychologically unsuitable
(n = 1)
Suitable prognosis and psychology
(n = 36)
Not offered implant (rationale not specified)
(n = 6)
Patient offered implant
(n = 30)
Patient refused implant
(n = 6)
Patient agreed to implant
(n = 24)
Study Population
(n = 24)


Any Arrhythmia
Freedom from Arrhythmia
100
50
0
0
12
24
36
48
60
Months
Number at risk
All 24 13 9 7 5 1

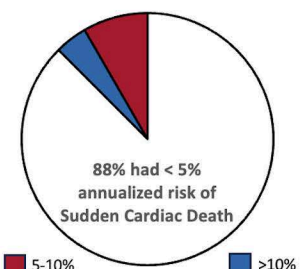

No Arrhythmia
IART
Conduction Disease
15
10
5
0
Number
< 1%
1 - 4%
5 - 10%
>10%
SCD Risk

Arrhythmia Detection in the Systemic Right Ventricle

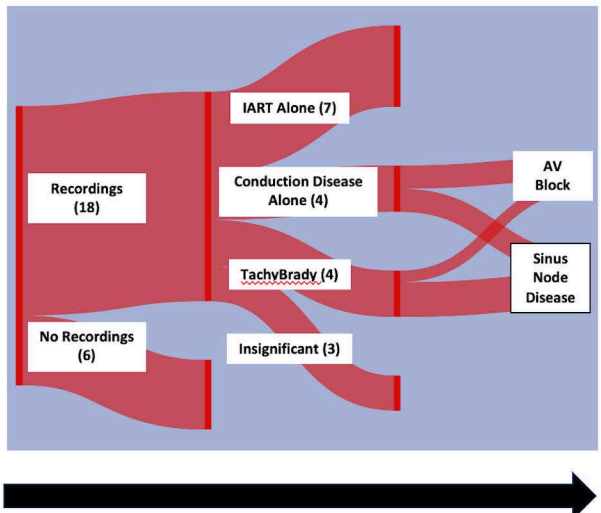


http://www.chd-diagrams.com.

24 Patients
54% Mustard, 46% Senning



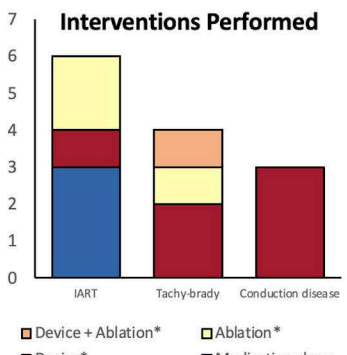
*Ladouceur et al, European Heart Journal (2022)



Follow-Up 39.5 months

Arrhythmia Detection unrelated to:

- Surgical repair
- Ventricular function
- Tricuspid Regurgitation
- Previous arrhythmia
- SCD risk score



*Many patients had additional medication therapy


A
18:58:35
0.1 mV
400 ms
B
14:14:38
0.1 mV
400 ms
Tachy Detected
Terminated
C
20:28:15
0.1 mV
400 ms
520
520
540
580
640
670
700
790
1500
>2000
Pause D
20:28:25
0.1 mV
400 ms
A
D
B
1400
1120
570
520
510
530
570
590
590
570
580