Carolina Escudero ORCID iD: 0000-0001-9401-3221

Riata Lead Failure in Pediatric and Congenital Heart Disease Patients

Short title: Riata Lead Failure in Pediatric and CHD Patients

Carolina A Escudero[a], MD, MSc, Douglas Y Mah[b], MD, Christina Y Miyake[c], MD, MS, Elizabeth A Stephenson[d], MD, MSc, FHRS, Martin J LaPage[e], MD, MSc, FHRS, Peter Kubuš[f], MD, Mitchell Cohen[g,1], MD, FHRS, Joseph Atallah[a,h], MD, MSc

[a]University of Alberta and Stollery Children's Hospital, 8440 112 St NW, Edmonton, AB, T6G 2B7, Canada, escudero@ualberta.ca; [b]Boston Children's Hospital, 300 Longwood Avenue, Boston, MA, 02115, USA, douglas.mah@cardio.chboston.org; [c]Baylor College of Medicine, One Baylor Plaza, Houston, TX, 77030, USA, cymiyake@bcm.edu; [d]Hospital for Sick Children, 555 University Ave, Toronto, ON, M5G 1X8, Canada, elizabeth.stephenson@sickkids.ca; [e]University of Michigan, 1540 East Hospital Drive, Ann Arbor, MI, 48109, USA, mlapage@med.umich.edu; [f]Motol University Hospital, V Úvalu 84

150 06, Prague, Czech Republic, peter.kubus@fnmotol.cz; [g]Phoenix Children's Hospital, 1919 E Thomas Rd, Phoenix, AZ, 85016, USA, mitchcohenmd@gmail.com; [h]joseph.atallah@ahs.ca

[1]Present address: Inova Children's Hospital, 3300 Gallows Rd, Falls Church, VA, 22042, USA

Corresponding author:

Dr. Carolina Escudero

Address: 4C1.19 WMC, 8440-112 Street, Edmonton, AB T6G 2B7

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Phone number: 780-407-3963; Fax: 780-407-3954; Email address:

carolina.a.escudero@gmail.com

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Abstract

Background: Implantable cardioverter defibrillator (ICD) lead failures occur at higher rates in pediatric and CHD patients.

Objective: To determine the rate and timing of Riata lead failure in pediatric and congenital heart disease (CHD) patients.

Methods: This was a retrospective, multicenter cohort study of pediatric patients and adults with CHD with implantation of a Riata or Riata ST lead between 2002-2009. The prevalence and timing of electrical failure and conductor coil externalization (CCE) were determined.

Results: Fifty-eight patients and 63 leads from 7 centers were included. Median (IQR) age at implant was 14.4 (11.5-18.7) years and median follow-up was 8.7 (7.3-11.1) years. The underlying diagnosis was a primary arrhythmia disorder in 45%, cardiomyopathy in 31%, and CHD in 28% of patients. Electrical failure occurred in 43% and CCE in 16% of leads at median lead ages of 4.7 (3.4-7.5) and 4.3 (3.9-7.0) years, respectively. Median lead survival free from electrical failure or CCE was 7.9 (95% CI 5.8-10.0) years. Forty-one percent of leads were functional at the end of the follow-up period, and 33% were extracted with a complication rate of 5%.

Conclusions: The rate of Riata lead electrical failure was high in children and patients with CHD, while the rate of CCE was comparable to published data. Counselling on lead management should factor in the high rate of electrical failure with considerations for elective replacement.

Keywords: Riata lead, lead failure, pediatric, children, congenital heart disease, PACES

Abbreviations List:

BMI: body mass index

BSA: body surface area

CCE: conductor coil externalization

CHD: congenital heart disease

CI: confidence interval

CPVT: catecholaminergic polymorphic ventricular tachycardia

D-TGA: dextro-transposition of the great arteries

HV: high voltage

ICD: implantable cardioverter-defibrillator

PACES: pediatric and congenital electrophysiology society

REDCap: research electronic data capture

Introduction

Implantable cardioverter-defibrillators (ICDs) are potentially life-saving therapies; however, due to the nature of their design, they carry a risk of system failure, resulting in serious complications and patient morbidity and mortality. ICD lead failure is the most common form of ICD system failure, with lead failure in pediatric and congenital heart disease (CHD) patients being significantly worse than in adult patients[1,2].

A unique form of lead failure has been observed in the St Jude Medical Riata (8 French) and Riata ST (7 French) defibrillation leads, where degradation of the outer insulation results in conductor coil externalization (CCE)[3]. It has been suggested that this mechanism of lead failure is due to movement of the cables within the lead lumen, causing outward forces and disrupting

the outer silicone insulation of the Riata lead, resulting in cable externalization[3]. CCE can predispose to lead malfunction and may lead to cardiac injury, thrombo-embolic events and lead-lead interaction. However, leads with CCE may maintain stable electrical function.

The studies to date investigating structural and electrical failure in Riata leads have been performed in adult patients. Research specifically investigating Riata lead function has not been performed in pediatric patients or patients with congenital heart disease (CHD). Considering the increased failure rate of ICD leads in pediatric patients in general, we hypothesized that Riata lead failure rate is also higher in pediatric patients. In addition, the timing of CCE post-implant is not well defined in pediatric or congenital heart disease patients, nor is the temporal association with electrical failure. The aim of this retrospective study was to determine the rate and timing of Riata lead failure in children and patients with CHD.

Methods

This international, multicenter, retrospective cohort study involved 7 centers from 3 different countries (Canada, United States, Czech Republic). Centers were recruited through the Pediatric and Congenital Electrophysiology Society (PACES). De-identified data were managed using REDCap (Research Electronic Data Capture) hosted at the University of Alberta. REDCap is a secure, web-based application designed to support data capture for research[4]. This study was approved by each institution's research ethics board.

Patients implanted with one of the following Riata or Riata ST lead models were identified by searching each institution's local ICD database: 1560, 1561, 1562, 1570, 1571, 1572, 1580, 1581, 1582, 1590, 1591, 1592, 7000, 7001, 7002, 7010, 7011, 7040, 7041, and 7042. Only pediatric patients (21 years or younger at the time of ICD implantation) or adults with CHD were included in the study.

Lead failure was defined as either structural or electrical failure. Structural lead failure, or CCE, was defined by the presence of outer insulation breach and conductors outside of the lead body on at least one x-ray or fluoroscopic image. Specifically, CCE was confirmed when the suspected lead portion had a width that was larger than the high voltage (HV) coil and when the conductor radius of curvature was larger than that of the lead body. Electrical failure was defined as at least one of the following findings or changes in lead properties from stable post-implantation values: a) non-physiological electrical noise/artifact detected as non-sustained ventricular arrhythmia or causing an inappropriate shock, b) low voltage impedance or high voltage impedance decrease by > 50% or increase by > 100%, c) capture threshold increase by > 100%, d) R wave sensing decrease by > 50%, e) or failed appropriate shock secondary to a change in electrical lead properties.

Patient demographic data was collected including gender, age at ICD implantation, and height and weight at ICD implantation. Clinical data collected included electrophysiologic diagnosis, cardiac structural diagnosis, ICD indication (primary or secondary prevention), and device/lead characteristics. Follow-up data collected included the type and timing of Riata lead structural and electrical failure, lead status at last follow-up, lead extraction data, and patient status at last follow-up.

Statistical analysis

The data were analyzed and are presented as patient-specific and lead-specific variables. Continuous variables were summarized using median and interquartile range. Frequency tables were generated for categorical variables. Differences between groups were assessed using Mann-Whitney U testing for continuous variables and chi square or Fisher exact testing for categorical variables. Time to event analysis was performed using Kaplan-Meier analysis. Average yearly failure rates were calculated using the 7-year actuarial rate. Significance

was set at p = 0.05 for all analyses. Statistical analysis was performed with SPSS v20.0 (IBM Corp, NY).

Results

There were 58 patients included in the study with a Riata or Riata ST lead implanted between 2002 and 2009. Patient demographics are provided in Table 1. Median age at Riata lead implantation was 14.4 (11.5-18.7) years. Children ≤12 and ≤18 years at Riata lead implantation comprised 38% and 78% of the cohort, respectively. The majority (71%) of ICDs were implanted for a primary prevention indication. A single chamber ICD was implanted in 29 (50%), dual chamber in 28 (48%), and CRT-D in 1 (2%). The generator was positioned in the left pectoral region in 52 (90%), right pectoral in 5 (8%) and abdomen in 1 (2%). Table 1 provides the underlying cardiac diagnosis for the patients included in this study. A primary arrhythmia disorder was present in 26 (45%), cardiomyopathy in 18 (31%), and repaired CHD in 16 (28%). The specific disease types for each of these diagnostic categories are provided in Table 1. There were 2 patients with overlap in their primary diagnoses: one with long-QT syndrome and dilated cardiomyopathy, and the other with CHD and dilated cardiomyopathy (see Table 1).

There were 63 Riata leads in total, with 5 patients (9%) implanted with 2 consecutive Riata leads each. Forty-two (67%) leads were the 8 French Riata lead, with the remainder being the 7 French Riata ST lead. Seven (11%) leads were implanted on the right side. The venous access site for implantation was left cephalic in 12 (19%), left axillary/subclavian in 39 (62%), right axillary/subclavian in 5 (8%), right internal jugular in 1 (2%), and unknown in the remainder.

Patients were followed for a median of 8.7 (7.3-11.1) years. At the end of the follow-up period, 57 (98%) patients were alive. One patient died secondary to endocarditis. Median lead follow-up was 7.1 (4.3-9.1) years. Presentation with Riata lead failure is summarized in Table 2.

Over the follow-up period, 27 (43%) leads experienced electrical failure at a median lead age of 4.7 (3.4-7.5) years, with the most frequent presentation being electrical noise or artifact detected as non-sustained ventricular tachycardia or fibrillation in 78% of leads with electrical failure. An inappropriate shock occurred in 4/20 (20%) patients with lead noise or artifact, with patients receiving 1, 2, 5, and 78 inappropriate shocks each due to lead noise. There were no failed appropriate shocks.

Structural lead failure, or CCE, occurred in 10 (16%) leads at a median lead age of 4.3 (3.9-7.0) years. Seven cases of CCE were identified on x-ray or fluoroscopy and 3 were identified at the time of surgery for lead replacement or extraction, just prior to lead manipulation being performed. Electrical failure and CCE were concomitantly present in 7 patients. Electrical failure was not significantly associated with CCE (p=0.084). In leads with CCE and electrical failure, 3 cases of CCE were identified at 6 months, 10 months, and 2.4 years prior to the onset of electrical failure. Of the remaining 4 cases with CCE and electrical failure, 2 were diagnosed radiographically at the time of electrical failure and 2 were diagnosed by direct inspection at the time of surgical intervention for lead replacement or extraction. Thirty (48%) leads experienced either electrical or structural failure during the follow-up period. Using Kaplan-Meier analysis, the median lead survival from electrical failure or CCE was 7.9 (95% CI 5.8-10.0) years. Survival curves are provided in Figure 1. Based on the 7 year failure rate, the actuarial average yearly failure (electrical failure or CCE) rate was 7.9%/year.

There were no differences in electrical failure (48% vs 33%, p=0.280), CCE (17% vs 14%, p=1.0), or either CCE or electrical failure (52% vs 38%, p=0.285) between the 8 and 7 French Riata leads. Table 3 provides a comparison between patients who did not have Riata lead failure and those that experienced either CCE or electrical lead failure for the initial Riata lead. There were no differences in age at Riata implantation, BMI, BSA, or the presence of CHD between

patients with and without lead failure. Lead failure was less likely to occur in patients with cardiomyopathy (p=0.004), and more likely to occur in patients with a primary arrhythmia disorder (p=0.039). Table 3 provides a comparison of lead characteristics between the Riata leads that had electrical or structural failure and the leads that did not fail during the follow-up period for all Riata leads.

At the end of the follow-up period, 26 (41%) leads were functional, 12 (19%) were abandoned, 4 (6%) were conditionally functional, and 21 (33%) leads were extracted. Of the leads that were conditionally functional, 2 Riata leads were used only for sensing and pacing, and 2 were used only to deliver a shock (additional pace/sense lead inserted). The distribution of lead outcome is displayed in Figure 2. Electrical failure was the indication for lead extraction in 12 (57%) leads, with 5 of these leads having concomitant CCE. In leads with only CCE, the indications for extraction were structural failure in 1 (5%) and elective extraction for advisory in 1 (5%) lead. In leads without structural or electrical failure, 7 were extracted for indications of device infection in 2 (9%), lead dislodgement in 2 (9%), cardiac transplantation in 2 (9%) and during concomitant cardiac surgery for tricuspid and pulmonary valve replacement in 1 (5%) patient. The leads were extracted with simple traction alone in 2 (10%), using locking stylets in 5 (24%), using a laser sheath in 10 (48%), and surgically extracted in 4 (19%). One (5%) patient had a complication of lead extraction, with development of a hemothorax that was noted post-operatively. The hemothorax was treated with chest tube placement and did not require open surgical intervention as the bleeding resolved with observation. The indication for lead extraction in this patient was electrical failure.

Discussion

To the best of our knowledge, this is the largest published experience of Riata lead failure in children and patients with CHD, and provides long-term follow-up data on the

performance of the Riata lead in this unique population. We found that the Riata lead failure rate was high in our patient cohort, at an average of 7.9%/year, mainly driven by a high rate of electrical lead failure occurring at a median lead age of 4.3 years. At the end of the follow-up period, only 41% of the Riata leads remained fully functional.

The rate of Riata electrical failure, reported at 43% in this study, is much higher than what has been reported to date in predominantly adult patients. A recent study of 3763 Riata leads found a cumulative incidence of electrical failure of 5.2% at 8 years, while a recent meta-analysis found an overall electrical failure rate of 6.3%[5,6]. In the large study by Parkash *et al.* (2016), predictors of electrical failure included higher ejection fraction and lower age[5]. Compared to our series, the older age of their cohort and the higher rate of patients with cardiomyopathies and reduced ejection fraction may explain some of the difference in electrical failure rates[5]. The finding of a high rate of electrical failure in our study is concordant with previous studies demonstrating that rates of ICD lead failure are higher in pediatric and CHD patients than in adult populations[1,2]. The rate of electrical lead failure was even higher in our cohort of patients with Riata leads than has been identified in other cohorts of pediatric and CHD patients with variable proportions of non-Fidelis ICD leads[1,2,7-9]. This finding may be related to the Riata lead being more susceptible to the factors influencing lead failure in children and patients with CHD, including lead stress related to somatic growth and higher activity levels[1,2,7-9].

In contrast to the higher rate of electrical failure, the 16% rate of CCE seen in our cohort is in keeping with previously reported rates of CCE in adult patients. Parkash *et al.* (2016) found a cumulative incidence of CCE of 9.2% at 8 years, while a meta-analysis by Zeitler *et al.* (2015) found an overall rate of CCE of 23.1%[5,6]. The number of leads with both electrical and structural failure in our study was small, not allowing inferences on the temporal correlation. Nevertheless, this population specific data with variable rates of electrical and structural failure

is important for proper counselling of patients and families, allowing for more informed decision making on lead management.

On univariate analysis, there was no difference in the age at ICD implantation in patients with and without Riata lead failure in our study, although previous studies have shown that earlier age at lead implantation is associated with an increased rate of ICD lead failure in children and patients with CHD[1]. There was no significant association between CCE and electrical failure in our patient cohort, with a similar number of cases of CCE being recognized at the time of identification of electrical failure as those recognized before the onset of electrical lead failure. It is possible that the lack of association between CCE and electrical failure in our study is due to being underpowered to detect this relationship, as recent studies have demonstrated an association between Riata lead electrical failure and CCE[5,6,10,11]. Electrical failure occurs more frequently in Riata leads with CCE than without CCE, occurring in 14% of cases of CCE in the series by Parkash *et al.* (2016), and in 17% in a recent meta-analysis[5,6]. We found that patients with Riata lead failure had a lower frequency of cardiomyopathy and a higher frequency of a primary arrhythmia disorder compared to patients without Riata lead failure. In a large multicenter study on a similar pediatric and adult congenital patient population, the frequency of inappropriate shocks was significantly lower in the cardiomyopathy subgroup compared to the subgroups of primary electrical disease and CHD[2]. Although the frequency of lead failure was not compared among those subgroups, lead failure was the most common attributable cause for inappropriate shocks, which may be in keeping with our finding of decreased lead failure in cardiomyopathy patients[2].

Analysis of the survival curve demonstrates a sharp increase in lead failure rate at an approximate lead age of 4 years, after which the lead failure rate remains stable over time. The steady rate of failure after the inflection point in the survival curve is in keeping with recent data

demonstrating a steady Riata lead failure rate in adult patients[5]. At the end of the follow-up period, nearly half of the patients in this study had experienced either electrical or structural failure. The high and steady rate of lead failure identified in our study supports ongoing close monitoring for Riata lead failure in children and patients with CHD. Although no failed appropriate shocks were reported in this study, data published on adult patients reported failed appropriate shocks secondary to lead electrical failure, with associated fatality[5,12]. Remote monitoring should be implemented for all patients with a Riata or Riata ST lead, potentially allowing for prompt detection of signs of electrical failure. In addition, the high rate of Riata lead electrical failure would argue for a more aggressive approach to lead replacement, usually at the time of generator or other lead replacement/revision, with or without lead extraction.

Fewer than half of all leads were functional at the end of the follow-up period. Lead extraction was performed for 33% of leads and was associated with a complication in 1 of the 21 extracted leads. Published reports show that Riata lead extraction is associated with increased procedural complexity and associated higher complication rates of 2-19%[5,13,14]. Decision making regarding lead extraction is particularly important in children and patients with CHD given their young age, need for lifetime device therapy and their predisposition to a higher risk of a complicated or failed extraction. Overall, it is important to consider an individualized decision-making approach, with careful analysis of the risk to benefit ratio for each patient, and alternative therapeutic options including subcutaneous ICDs.

Limitations of the present study include its retrospective design and the relatively small number of patients. The small patient cohort in our study may have led to being underpowered to detect differences between patients with and without lead failure. The number of patients with CCE was also low in our study population and we were underpowered to detect risk factors for CCE. The timing of assessment of structural lead

integrity using chest radiography or fluoroscopy was non-standardized, making the temporal relationship between CCE and electrical failure, and the reported rate of CCE, potentially inaccurate. This study may have also been underpowered to detect a relationship between CCE and electrical failure. The timing of routine interrogation and the use of remote monitoring was not standardized given the retrospective nature of this study, which may have influenced the time to detection of electrical failure in our cohort. There is the potential for selection bias to have affected our results if the centers included in this study were influenced to participate by higher local Riata failure rate than may be present in the broader pediatric and CHD population.

Conclusions

This study of Riata lead failure in pediatric and CHD patients demonstrated a very high and steady rate of electrical failure, while the rate of CCE is similar to what has previously been reported in adult populations. We did not identify a temporal correlation between electrical and structural failure. Less than half of all leads were functional at the end of the study and one-third were extracted. This population specific data will allow for more informed and focused counseling of affected patients, including the use of remote monitoring and consideration for elective lead replacement.

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Figure Legends:

Figure 1: Kaplan-Meier estimate of lead survival from electrical or structural failure for all Riata leads. The 95% confidence intervals for the survival curve are represented by the dotted lines. The vertical lines represent censored patients. The number of leads at risk at each 2 yearly time point is provided at the bottom of the figure. Median lead survival was 7.9 years. CI = confidence interval, CCE = conductor coil externalization

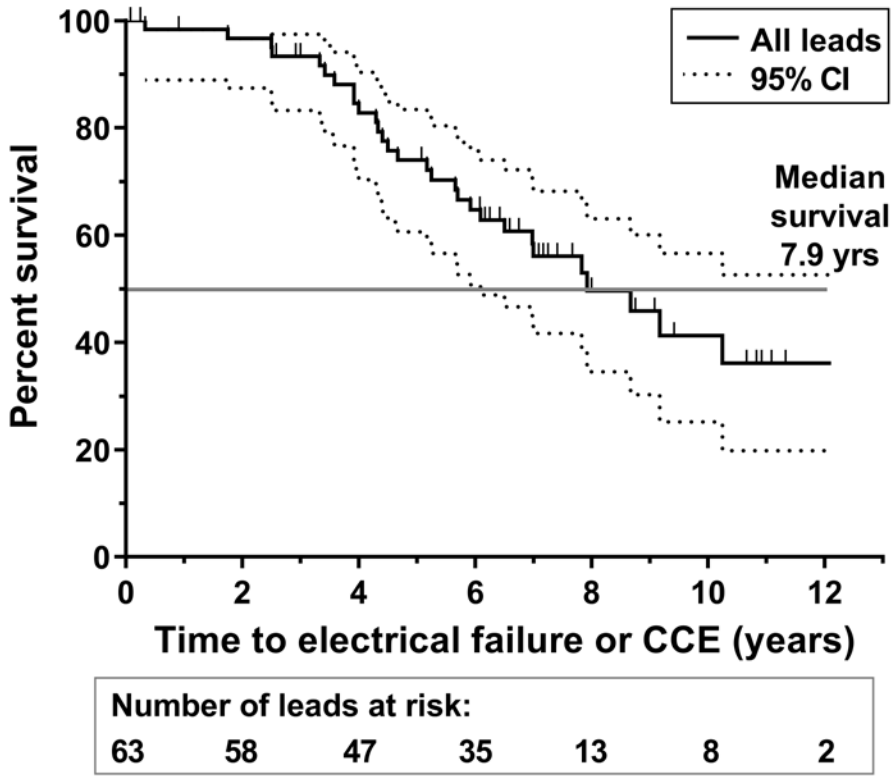

Figure 2: Flowchart of lead outcomes for all Riata leads. CCE = conductor coil externalization.

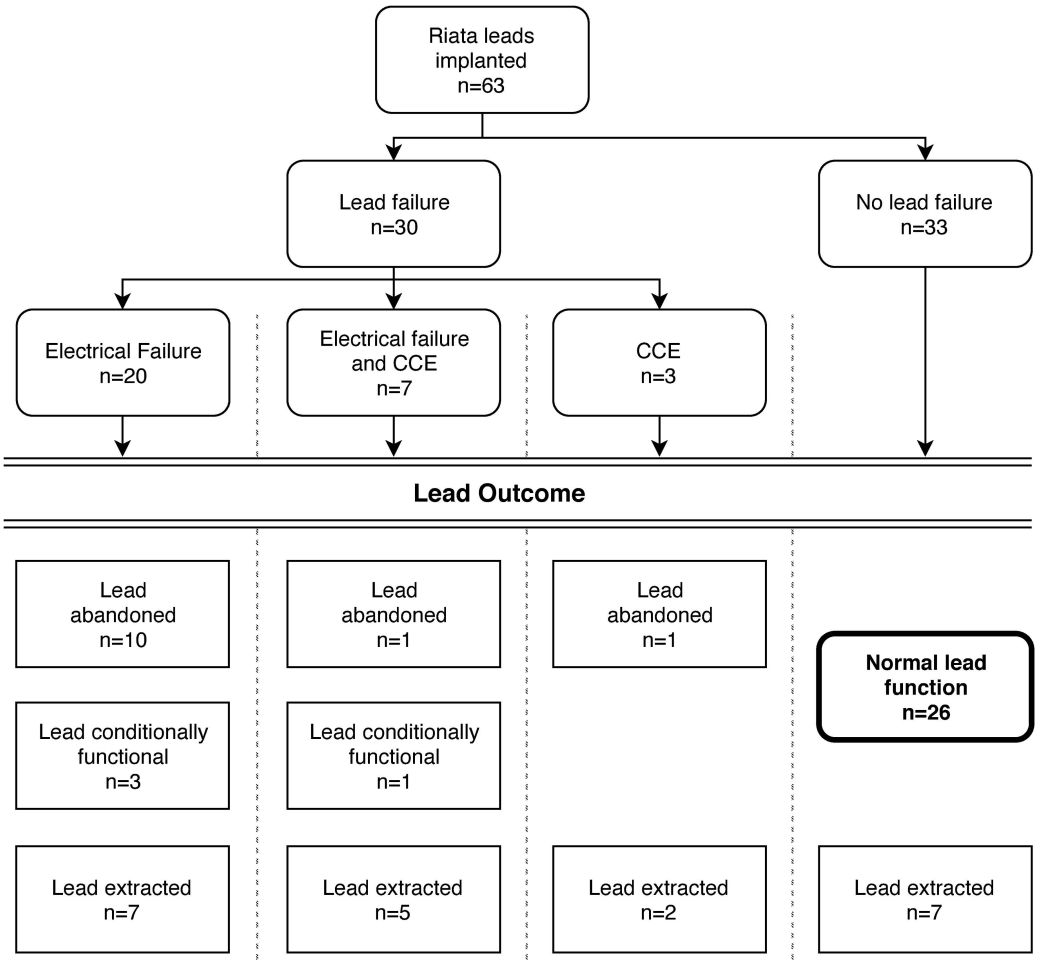

Table 1. Baseline Characteristics and Patient Diagnoses

Variable	N=58
Gender (male)	33 (57%)
Age at implantation (years)	14.4 (11.5-18.7)
Height at implantation (cm)	160 (147-171)
Weight at implantation (kg)	58.9 (37.9-71.2)
BMI at implantation (kg/m^2)	22.4 (17.2-25.7)
BSA at implantation (m^2)	1.61 (1.22-1.83)
Patients with ≥2 Riata leads	5 (9%)
Primary prevention indication	41 (71%)
Diagnosis:	
Primary arrhythmia disorder	26 (45%)
Long-QT syndrome	19 (33%)
CPVT	4 (7%)
Brugada syndrome	1 (2%)
Idiopathic ventricular fibrillation	1 (2%)
Other[†]	1 (2%)
Cardiomyopathy	18 (31%)
Hypertrophic cardiomyopathy	8 (14%)
Arrhythmogenic cardiomyopathy	6 (10%)
Restrictive cardiomyopathy	1 (2%)
Dilated cardiomyopathy	2 (3%)
Severe post-transplant coronary vasculopathy	1 (2%)

CHD	16 (28%)
Atrial switch for D-TGA	5 (9%)
Tetralogy of Fallot	4 (7%)
Atrioventricular septal defect	2 (3%)
Other[‡]	5 (9%)

BMI = Body mass index, BSA = body surface area, CPVT = Catecholaminergic polymorphic ventricular tachycardia, CHD=congenital heart disease, D-TGA = D-transposition of the great arteries

[†]One patient was incorrectly diagnosed with long QT syndrome

[‡]The other types of CHD included double outlet right ventricle; ventricular septal defect with post-surgical aortic insufficiency requiring mechanical aortic valve replacement; atrial septal defect, ventricular septal defect, and patent ductus arteriosus with later development of pregnancy-induced dilated cardiomyopathy; and Marfan's syndrome associated with mitral valve replacement.

Table 2. Presentation of Riata lead failure

Variable	N=63
Electrical failure	27 (43%)
Lead age at electrical failure (years)	4.7 (3.4-7.5)
Type of electrical failure:	
Non-physiologic noise/artifact	21 (78%)
Capture threshold increase by >100%	5 (19%)
Impedance increase by > 100%	4 (15%)
R wave decrease by >50%	4 (15%)
Impedance decrease by >50%	3 (11%)

Structural failure (CCE)	10 (16%)
Lead age at structural failure (years)	4.3 (3.9-7.0)
Location of CCE:	
Proximal to distal coil	6 (60%)
ICD pocket	3 (30%)
Unspecified	1 (10%)

CCE = conductor coil externalization

Table 3. Patient and lead characteristics between those with and without lead failure

Patient Variable[†]	No lead failure (n=31)	Lead failure (n=27)	p value
Gender (male)	19 (61%)	14 (52%)	0.469[‡]
Age at implantation (years)	13.7 (11.4-17.2)	15.2 (12.4-22.9)	0.307[§]
Age ≤ 12 years	14 (45%)	9 (33%)	0.358[‡]
Height at implantation (cm)	160 (147-171)	162 (151-168)	0.992[§]
Weight at implantation (kg)	59.4 (36.3-75.1)	55.9 (44.0-70.0)	0.804[§]
BMI at implantation (kg/m^2)	21.9 (16.5-26.8)	22.4 (17.3-25.4)	0.852[§]
BSA at implantation (m^2)	1.64 (1.20-1.87)	1.57 (1.36-1.78)	0.772[§]
Primary arrhythmia disorder	10 (32%)	16 (59%)	0.039[‡]
Cardiomyopathy	14 (48%)	3 (11%)	0.004[¶]
CHD	8 (26%)	8 (30%)	0.745[‡]
Primary prevention indication	22 (71%)	19 (70%)	0.960[‡]
Patient follow-up (years)	7.8 (6.8-10.4)	10.7 (8.3-11.6)	0.006[§]
Lead Variable[†]	**No lead failure (n=33)**	**Lead failure (n=30)**	
Lead implantation site			0.083[‡]
Axillary/cephalic	3 (9%)	9 (30%)	

Subclavian	25 (76%)	19 (63%)	
Other/unspecified	5 (15%)	2 (7%)	
Left sided lead implantation	29 (88%)	27 (90%)	1.0[¶]
Active fixation lead	20 (61%)	18 (60%)	0.961[‡]
Dual coil lead	11 (33%)	12 (40%)	0.583[‡]
8 French model	20 (61%)	22 (73%)	0.285[‡]

[†]Patient characteristics were compared for patients with and without failure of the initial Riata lead, while lead characteristics were compared for all Riata leads with and without failure

BMI = Body mass index, BSA = body surface area, CHD = congenital heart disease

Analysis performed with chi square ([‡]), Mann-Whitney U test ([§]), or Fisher exact ([¶])