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ORIGINAL ARTICLE
Pediatric Cardiology and Adult Congenital Heart Disease

Acute and Long-Term Outcomes of Transvenous Cardiac Pacing Device Implantation in Patients With Congenital Heart Disease

Daiji Takeuchi, MD; Keiko Toyohara, MD; Daigo Yagishita, MD; Kyoichiro Yazaki, MD; Satoshi Higuchi, MD; Koichiro Ejima, MD; Morio Shoda, MD; Nobuhisa Hagiwara, MD

Background: Little is known about the acute/long-term outcomes of implantation of cardiac implantable electronic devices (CIED) using a transvenous approach for patients with congenital heart disease (CHD).

Methods and Results: We retrospectively investigated the acute/long-term results and complications associated with transvenous CIED implantation in 140 patients with CHD. We implanted 77 pacemakers, 51 implantable cardioverter defibrillators (ICD), and 12 cardiac resynchronization therapy (CRT) devices. Although we successfully implanted pacemakers and ICD in all patients, we could not place a coronary sinus (CS) lead in 25% of the patients requiring CRT devices due to coronary vein anomalies associated with corrected transposition of the great arteries (cTGA). Overall complication rate, lead failure rate, and incidence of device infection were 16%, 9%, and 0.7%, respectively. There was no significant difference in overall complication rates between the simple (n=22) and complex CHD (n=118) groups (14% vs. 16%). The 10-year lead survival for the ICD leads (77%) was significantly lower than for the pacemaker leads (91%, P=0.0065).

Conclusions: The outcomes of transvenous CIED in patients with CHD seemed acceptable, although there was a relatively high incidence of complications. CS lead placement for cTGA may be hindered by coronary vein anomalies. Lead survival tended to be lower for ICD than for pacemakers in these patients.

Key Words: Cardiac pacing device; Congenital heart disease; Endocardial pacing lead; Transvenous approach

Patients with congenital heart disease (CHD) may have arrhythmic episodes including cardiac conduction disturbances and/or atrial/ventricular tachycardia. These patients may also experience heart failure due to cardiac dysfunction.[1] Furthermore, as the survival of patients with CHD improves, the number of cardiac implantable electronic devices (CIED) implanted in these patients is increasing. Transvenous CIED implantation for CHD, especially in complex CHD, is challenging because of the unique anatomy and physiology involved.[2] In patients with large intracardiac shunts or difficult vascular access, epicardial lead implantation is usually used, but this requires thoracotomy, and is more likely to be associated with lead failure and therefore may require reintervention.[3–5] Transvenous CIED implantation may have the advantage of lead longevity compared with epicardial lead implantation, if cardiac anatomy permits. The use of transvenous CIED implantation in patients with CHD may be increasing in frequency due to several factors: (1) the use of transcatheter intervention for intracardiac shunts and venous stenosis prior to CIED implantation if needed; (2) the use of 3-D mapping systems[6,7] during CIED implantation to better understand cardiac anatomy and low-voltage areas; and (3) developments in lead extraction systems.[8] The complex intracardiac anatomy associated with previous cardiac surgery and low-voltage atrial/ventricular electrical activity, however, may contribute to the difficulty in achieving successful transvenous CIED implantation and to the high rate of CIED-related complications in these patients. Most previous reports regarding the outcomes of pacemaker lead implantation in patients with CHD include both epicardial and endocardial leads.[2,3,9,10] The outcomes of CIED implantation using only endocardial leads by the transvenous approach for patients with CHD, however, have not been studied in detail. The aim of this study was to investigate the acute and long-term results

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Department of Pediatric and Adult Congenital Cardiology (D.T., K.T.), Department of Cardiology (D.Y., K.Y., S.H., K.E., M.S., N.H.), Tokyo Women's Medical University, Tokyo, Japan
N.H. is a member of *Circulation Reports*' Editorial Team.
Mailing address: Daiji Takeuchi, MD, PhD, Department of Pediatric and Adult Congenital Cardiology, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan. E-mail: takeuchi.daiji@twmu.ac.jp or Morio Shoda, MD, PhD, Department of Cardiology, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan. E-mail: shoda.morio@twmu.ac.jp
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and complications associated with transvenous CIED implantation in patients with CHD, particularly complex CHD.

Methods

Study Design and Groups

This was a single-center, retrospective study investigating the acute and long-term results of CIED implantation in patients with CHD between February 1990 and February 2016. The implanted devices were pacemakers, implantable cardioverter defibrillators (ICD), and cardiac resynchronization therapy (CRT) devices. The study investigated only devices implanted using a transvenous approach. A total of 144 consecutive patients with CHD received a cardiac pacing device implanted using endocardial leads. Exclusion criteria included receiving a hybrid device in which endocardial and epicardial leads were used (n=3), and loss to follow-up ≤1 year after initial device implantation (n=1). Subsequently, a total of 140 patients were enrolled in this study. We avoided transvenous pacing device implantation in patients with large intracardiac shunts, but we did perform CIED implantation in patients with small shunts or in those who had shunt closure prior to implantation of the pacing device.

We obtained approval for this study from the institutional review board of Tokyo Women's Medical University Hospital. Informed consent was obtained from the patients in accordance with university hospital policies. According to the complexity of CHD, based on the ICD-9 codes,[1,11] we divided the patients into 2 groups: simple CHD (n=22, 20%) and complex CHD (including moderate/severe CHD; n=118, 80%).

First, we studied the acute results for each of the different types of pacing devices, comparing the complex CHD group with the simple CHD group. In addition, we reviewed the outcomes associated with the use of catheter interventions and 3-D mapping systems. Second, we compared the complication rates between the simple and complex CHD groups. Third, we studied the longevity of the pacemaker, ICD and CRT leads in each group and the risk factors associated with lead failure. Finally, we studied patient survival rates during the follow-up period.

Data Collection

We reviewed the medical records of patients with CHD who had received a CIED at Tokyo Women's Medical University Hospital between February 1990 and February 2016. We examined many variables including diagnosis of cardiac anatomy and surgical history, lead information, indication for pacing device, the use of 3-D mapping and transcatheter intervention combined with pacing device implantation, complications (including lead failure), device revision, clinical course and mortality. Device-related complications were defined as pneumothorax/hemothorax, hematoma, cardiac tamponade, lead dislodgement, lead failure, perforation, superior vena cava (SVC) occlusion, systemic embolization, and device infection. Lead failure

Table 1. Patient Characteristics

	Overall patients (n=140)	Simple/complex CHD group	
		Simple CHD group (n=22)	Moderate/severe CHD group (n=118)
Male	80 (57)	13 (59)	67 (57)
Age at implantation (years)	34 (25–43)	50 (39–64)	32 (23–42)
Initial implanted pacing device			
PM	77 (55)	16 (73)	61 (52)
ICD	51 (36)	6 (27)	45 (38)
CRT	12 (9)	0 (0)	12 (10)
Situs			
Inversus	10 (7.1)	0 (0)	10 (8.4)
Intra cardiac shunt	5 (3.6)	1 (4.5)	4 (3.4)
Cardiac disease			
Simple CHD			
VSD	7 (5.0)	7 (32)	0 (0)
ASD	13 (9.3)	13 (59)	0 (0)
PDA	2 (1.4)	2 (9)	0 (0)
Complex CHD			
CTGA	37 (26)	0 (0)	37 (31)
TOF	38 (27)	0 (0)	38 (32)
dTGA	11 (7.9)	0 (0)	11 (9.3)
AVSD	7 (5.0)	0 (0)	7 (5.9)
DORV	4 (2.9)	0 (0)	4 (3.4)
Single ventricle	4 (2.9)	0 (0)	4 (3.4)
TA	2 (1.4)	0 (0)	2 (1.7)
Truncus arteriosus	1 (0.7)	0 (0)	1 (0.8)
IAA complex	1 (0.7)	0 (0)	1 (0.8)
Others	12 (9.2)	0 (0)	12 (10)

(**Table 1** continued the next page.)

	Overall patients (n=140)	Simple/complex CHD group	
		Simple CHD group (n=22)	Moderate/severe CHD group (n=118)
Cardiac surgery			
ICR	89 (64)	18 (86)	71 (43)
Atrial switch operation	10 (7.1)	0 (0)	10 (8.4)
Mustard	5	0 (0)	5
Senning	5	0 (0)	5
Double switch operation	11 (7.8)	0 (0)	11 (9.3)
Senning+Rastelli	5	0 (0)	5
Mustard+Rastelli	5	0 (0)	5
Senning+arterial switch	1	0 (0)	1
Arterial switch operation	5 (3.5)	0 (0)	5 (4.2)
Rastelli operation	7 (5.0)	0 (0)	7 (5.9)
Fontan-type surgery	5 (3.6)	0 (0)	5 (3.4)
Ventricular septation	2 (1.4)	0 (0)	2 (1.7)
PDA ligation	2 (1.4)	2 (9.0)	0 (0)
ASD occluder	2 (0.7)	2 (4.5)	0 (0)
No cardiac operation	7 (5.0)	0 (0)	7 (5.9)
Indication			
Pacemaker	n=77	n=16	n=61
SSS	33 (43)	8 (50)	25 (41)
AVB	44 (57)	8 (50)	36 (59)
ICD	n=51	n=6	n=45
Primary prevention of SCD	5 (9.8)	0 (0)	5 (11)
Secondary prevention of SCD	46 (90)	6 (100)	40 (89)
CRT	n=12	n=0	n=12
Systemic ventricular dysfunction	12 (100)	0 (0)	12 (100)
Pacing mode			
AAI	17 (12)	1 (4.5)	16 (14)
VVI	11 (7.9)	2 (9.1)	9 (7.6)
VDD	5 (3.6)	1 (4.5)	4 (3.4)
DDD	107 (76)	18 (82)	89 (75)
Pacing lead conversion from epicardial to endocardial leads	18 (13)	1 (5)	17 (15)
Remote monitoring system	62 (44)	7 (32)	55 (47)
Late death	12 (8.3)	1 (5)	11 (9.3)

Data are given as median (IQR) or n (%). ASD, atrial septal defect; AVB, atrioventricular block; AVSD, atrioventricular defect; CHD, congenital heart disease; CRT, cardiac resynchronization therapy; CTGA, congenitally corrected transposition of the great arteries; DORV, double outlet right ventricle; dTGA, d-loop transposition of the great arteries; IAA, interrupted aortic arch; ICD, implantable cardioverter defibrillator; PDA, patent ductus arteriosus; PM, pacemaker; SCD, sudden cardiac death; SSS, sick sinus syndrome; TA, tricuspid atresia; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

included high pacing thresholds, pacing failure, lead fracture, leakage, sensing failure, and insulation failure.

3-D Navigation System

In line with previous studies,[6,12] we used a 3-D navigation system (Ensite NavX™, St. Jude Medical, MN, USA) for lead implantation in the patients with complex CHD. In brief, a steerable catheter was advanced through the SVC or femoral vein and connected to the NavX system to visualize and guide movement. Then, a 3-D geometry (shell) of the atria or ventricle was created. Atrial or ventricular voltage mapping was also obtained if needed. After constructing the anatomical shell, the pacing or ICD lead of the distal and ring electrodes was connected to the NavX system and advanced into the heart to visualize the movement and position of the pacing lead tip in real time. Sensing and pacing threshold parameters and twitching site by phrenic nerve capture were checked using the Ensite system. If acceptable, the site for lead implantation was confirmed and we fixed the lead at the adequate pacing site.

Statistical Analysis

Data regarding patient characteristics and various biomarker levels are given as percentage, mean±SD, or median (IQR). We used the chi-squared test to compare the proportion of categorical variables (e.g., complications) between the 2 groups. We used the Mann-Whitney U-test to compare the various biomarker levels between the groups. The Kaplan-Meier method was used to estimate pacing lead survival. We compared freedom from lead complications using log-rank test. Event-free survival in lead failure was calculated from the date of lead implantation to the date of lead failure. We used multivariate logistic regression analysis to analyze the risk factors associated with lead failure, including age <16 years at the time of lead placement, atrial lead, moderate to severe CHD, and ICD lead. Age <12 years and atrial leads were reported as risk factors for lead failure.[4,13,14] Given that only a small

Table 2. Pacemaker, ICD or CRT Patient Characteristics

	Pacemaker (n=77)	ICD (n=51)	CRT (n=12)	
			CRT-D (n=7)	CRT-P (n=5)
Male	42	32	1	4
Age at implantation (years)	30 (23–48)	37 (28–46)	50 (29–67)	28 (19–35)
Situs				
Inversus	5 (6.5)	5 (9.8)	0 (0)	0 (0)
Cardiac disease				
Simple CHD				
VSD	5 (6.5)	2 (3.9)	0 (0)	0 (0)
ASD	9 (12)	4 (7.8)	0 (0)	0 (0)
PDA	1 (1.3)	1 (2.0)	0 (0)	0 (0)
Complex CHD				
CTGA	23 (30)	6 (12)	5 (71)	4 (80)
TOF	11 (14)	25 (49)	2 (29)	0 (0)
dTGA	8 (10)	3 (5.9)	0 (0)	0 (0)
AVSD	5 (6.5)	2 (3.9)	0 (0)	0 (0)
DORV	0 (0)	4 (7.8)	0 (0)	0 (0)
Single ventricle	3 (3.9)	0 (0)	0 (0)	1 (20)
TA	2 (2.6)	1 (1.9)	0 (0)	0 (0)
Truncus arteriosus	1 (1.3)	0 (0)	0 (0)	0 (0)
IAA complex	1 (1.3)	0 (0)	0 (0)	0 (0)
Others	6 (7.8)	3 (5.9)	0 (0)	0 (0)
Cardiac surgery				
ICR	43 (56)	37 (73)	5 (71)	4 (80)
Atrial switch operation	9 (12)	1 (2.0)	0 (0)	0 (0)
Mustard	5	0	0	0
Senning	4	1	0	0
Double switch operation	9 (12)	2 (3.9)	0 (0)	0 (0)
Senning+Rastelli	3	2	0	0
Mustard+Rastelli	5	0	0	0
Senning+arterial switch	1	0	0	0
Arterial switch operation	4 (5.2)	1 (2.0)	0 (0)	0 (0)
Rastelli operation	1 (1.3)	6 (12)	0 (0)	0 (0)
Fontan-type surgery	4 (5.2)	1 (2.0)	0 (0)	0 (0)
Ventricular septation	1 (1.3)	0	0 (0)	1 (20)
PDA ligation	1 (1.3)	1 (2.0)	0 (0)	0 (0)
ASD occluder	0 (0)	2 (3.9)	0 (0)	0 (0)
No cardiac operation	5 (6.5)	0 (0)	2 (29)	0 (0)
Indication				
Pacemaker				
SSS	33			
AVB	44			
ICD				
Primary prevention of SCD		5	3	
Secondary prevention of SCD		46	4	
CRT				
Systemic ventricular dysfunction			7	5
Pacing mode				
AAI	17	0	0	0
VVI	8	2	0	0
VDD	2	2	0	0
DDD	50	41	7	4
Pacing lead conversion from epicardial to endocardial leads	15	2	0	1
Remote monitoring system	20 (26)	32 (63)	5 (71)	5 (100)
Late death	8 (10)	4 (7.8)	0 (0)	0 (0)

Data given as median (IQR) or n (%). ICR, intracardiac repair. Other abbreviations as in Table 1.

Table 3. Pacing Lead Characteristics

Leads	Manufacturer	Model	n	Fixation	Polarity	Diameter	Steroid
Pacemaker leads (n=183)							
Fineline/Thinline II Sterox EZ	Boston/Intermedics	438-35S	71	Screw-in	Bipolar	5F	Yes
Fineline/Thinline II Sterox EZ	Boston/Guidant	4469	13	Screw-in	Bipolar	5F	Yes
Fineline/Thinline II Sterox EZ	Boston/Guidant	4470	46	Screw-in	Bipolar	5F	Yes
Fineline/Thinline II Sterox EZ	Boston/Guidant	4471	22	Screw-in	Bipolar	5F	Yes
ThinlineEZ	Intermedics	438-10	4	Screw-in	Bipolar	5F	No
Capsure-Fix	Medtronic	4068	1	Screw-in	Bipolar	2.3 mm	Yes
Capsure	Medtronic	4504	1	Tined	Bipolar	2 mm	Yes
CapSureFix Novus	Medtronic	5076	5	Screw-in	Bipolar	2 mm	Yes
CapSureFix silicon	Medtronic	5068	2	Screw-in	Bipolar	2.4 mm	Yes
CapSureZ Novus silicon	Medtronic	5054	3	Tined	Bipolar	2 mm	Yes
IS-1 sutureless	Medtronic	5071	1	Screw-in	Unipolar	2.2 mm	No
Screw-in lead	Medtronic	6957	1	Screw-in	Unipolar	2.2 mm	No
Capsure-VDD-2	Medtronic	5038	1	Tined	Bipolar	2.65 mm	Yes
Selectsecure	Medtronic	3830	1	Screw-in	Bipolar	1.4 mm	Yes
Capsure-IS-1	Medtronic	4004M	1	Tined	Bipolar	2.4 mm	Yes
Solia S	Biotronics	Solia S	3	Screw-in	Bipolar	5.6F	Yes
Safio S	Biotronics	Safio S	2	Screw-in	Bipolar	5.6F	Yes
Tendril Optim	St. Jude Medical	1888TC	3	Screw-in	Bipolar	1.87 mm	Yes
Optisense	St. Jude Medical	1999	1	Screw-in	Bipolar	1.82 mm	Yes
Brilliant plus	Vitatron B.V	IMR16Q	1	Tined	Bipolar	2.65 mm	Yes
ICD leads (n=58)							
Durata ICD	St. Jude Medical	7120	2	Screw-in	Bipolar	2.34 mm	Yes
Durata ICD	St. Jude Medical	7120Q	10	Screw-in	Bipolar	2.34 mm	Yes
Durata ICD single coil	St. Jude Medical	7122Q	4	Screw-in	Bipolar	2.34 mm	Yes
Durata ICD single coil	St. Jude Medical	7122	4	Screw-in	Bipolar	2.34 mm	Yes
Durata ICD single coil	St. Jude Medical	7131	2	Screw-in	Bipolar	2.34 mm	Yes
Optisure	St. Jude Medical	LDA 210Q	2	Screw-in	Bipolar	2.4 mm	Yes
Sprint tined	Medtronic	6932	3	Tined	Bipolar	2.6 mm	Yes
Sprint Fidelis	Medtronic	6949	8	Screw-in	Bipolar	2.2 mm	Yes
Sprint Quattro	Medtronic	6935	7	Screw-in	Bipolar	2.8 mm	Yes
Sprint Quattro MRI	Medtronic	6935M	7	Screw-in	Bipolar	2.8 mm	Yes
Sprint screw-in	Medtronic	6943	1	Screw-in	Bipolar	2.6 mm	Yes
Reliance 4-front	Boston	0693	1	Screw-in	Bipolar	2.4 mm	Yes
Linox Smart Pro SD	Biotronik	Linox proMRI 65/18	3	Screw-in	Bipolar	2.7 mm	Yes
Linox Smart Pro SI DX	Biotronik	Linox proMRI S DX65/15	3	Screw-in	Bipolar	2.7 mm	Yes
Isoline	Sorin CRM	2CR6	1	Screw-in	Bipolar	2.8 mm	Yes
CRT (CS) lead (n=14)							
QuickFlex Micro	St. Jude Medical	1258T	1		Bipolar	1.42 mm	Yes
QuickFlex	St. Jude Medical	1156T	2		Bipolar	1.42 mm	Yes
Quartet quadripolar lead	St. Jude Medical	1458Q	6		Quadripolar	4.7Fr	Yes
Attain OTW	Medtronic	4193	1		Unipolar	1.3 mm	Yes
Attain Ability Plus	Medtronic	4296	1		Bipolar	1.57 mm	Yes
Attain Performa MRI	Medtronic	4298	1		Quadripolar	1.75 mm	Yes
Easytrack 2	Guidant	4542	1		Bipolar	1.8 mm	Yes
Corox OTW BP	Biotronik	Corox 85-BP	1		Bipolar	1.8 mm	Yes

CRT, cardiac resynchronization therapy; CS, coronary sinus; ICD, implantable cardioverter defibrillator.

number of patients were age <12 years (n=3), we used age <16 years as a variable on multivariate analysis. JMP Pro version 11 (SAS Institute, Cary, NC, USA) was used for all analyses. P<0.05 was considered significant.

Results

Baseline Patient Characteristics

Baseline patient characteristics are summarized in **Table 1**. Twenty percent of the patients (n=22) were categorized as having simple CHD, while the remaining 80% (n=118)

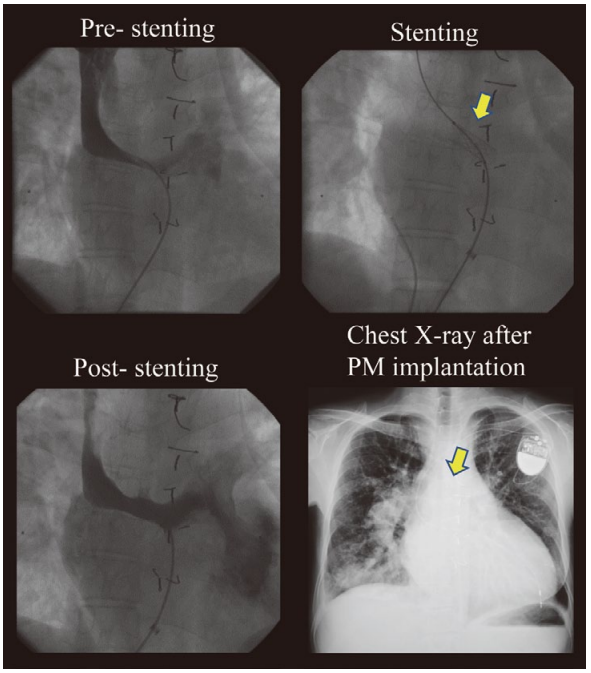

Figure 1. Stent implantation for superior vena cava (SVC) stenosis prior to pacemaker (PM) lead implantation in a patient with corrected transposition of the great arteries who had previously undergone Mustard operation, and subsequently developed atrial fibrillation with bradycardia and severe heart failure. (**Left upper**) Venography showing SVC stenosis; (**Right upper**) stenting for stenotic SVC (yellow arrow); (**Left lower**) dilated SVC stenotic lesion after stenting; (**Right lower**) chest X-ray after stenting showing the implanted stent (yellow arrow).

were categorized as having complex CHD.

In the simple CHD group, there were no CRT devices initially implanted. Overall, 95% of patients (n=133) had a history of previous cardiac surgery. This cardiac surgery included complex procedures such as 10 atrial switch operations, 11 double switch procedures, and 5 Fontan operations. Thirteen percent (n=18) of patients who previously had pacemaker implantation using epicardial leads underwent pacing lead conversion from epicardial to endocardial leads due to epicardial lead failure (n=16) or infection (n=2). One patient in the simple CHD group and 4 patients in the complex CHD group had intracardiac shunt. Patient characteristics according to the different types of pacing devices are listed in **Table 2**. Of the patients, 45% (n=77) received a pacemaker, 36% (n=51) received an ICD, and 9% (n=12) received a CRT device. Of the patients undergoing pacemaker implantation, the most common cardiac disease (30% of patients) was corrected transposition of the great arteries (cTGA). For the patients having ICD, the most common cardiac disease (49% of patients) was tetralogy of Fallot (TOF). In addition, with regard to CRT devices, cTGA was the most common disease (75% of patients).

Pacing Lead Characteristics

Pacing lead characteristics are listed in **Table 3**. Overall, a total of 255 leads were analyzed in this study. This included 183 pacemaker leads (including 123 atrial leads), 58 ICD high-voltage shock leads, and 14 coronary sinus (CS) leads implanted in 140 patients. Of the pacemaker leads, 96% (n=176) were thin leads with diameter ≤6 Fr. Moreover, in 85% of the pacemaker leads, we used either Fineline leads (Boston Scientific, MN, USA) or Thinline II leads (Intermedics, TX, USA). These were fixed screw leads with a narrow diameter (≤5 Fr). With regard to the ICD leads, 9 were officially recalled (8 Sprint Fidelis and 1 Isoline). In particular, 1 of these recalled leads (Sprint Fidelis 6949) showed oversensing of noise during follow-up. None of the Riata leads used in this study was recalled. With regard to the CS leads, 50% (n=7) were quadripolar leads.

Transcatheter Interventions

In this study, 2 of the patients underwent transcatheter atrial septal occlusion using an Amplatzer Septal Occluder device (St. Jude Medical) before ICD implantation. One of the patients had an unclosed atrial septal defect (ASD) and the other had a residual ASD with TOF following intracardiac repair. In addition, in another patient, urgent stent implantation for SVC stenosis was performed at the same time as VVI pacemaker implantation (**Figure 1**). This patient had TGA and, after Mustard operation, had symptomatic bradycardia associated with chronic atrial fibrillation and severe systemic right ventricle failure. Furthermore, epicardial lead implantation requiring thoracotomy for this patient was deemed too invasive.

3-D Navigation System

We used a 3-D navigation system (Ensite NavX, St. Jude Medical) in 11 patients for 8 pacemakers, 2 ICD, and 1 CRT defibrillator (CRT-D) implantation. All patients had complex CHD. Specifically, there were 3 patients with TGA after Mustard operation, 1 patient with isolated atrial inversion after Mustard operation, 4 patients with cTGA after double switch operations (including 2 Mustard/Rastelli operations and 2 Senning/Rastelli operations), 2 patients with double outlet ventricle after intracardiac repair, and 1 patient with TOF with severe left ventricular dysfunction. Forty-five percent (n=5) of these 11 patients had situs inversus.

Using the NavX system for various pacing lead implantations, we were able to achieve successful device implantation without any complications in all 11 cases. The system enabled us to map atrial and ventricular geometry and voltage in real time and therefore determine the optimal placement of the pacing lead tip. We were also able to view and mark points of twitching by phrenic nerve capture, pacing-p wave duration and pacing-QRS width during the procedure (**Figure 2**).

Pacing Devices: Acute Results

In the simple CHD group, 16 patients were initially planned for pacemaker implantation and 6 patients were scheduled for ICD implantation (**Figure 3**). All of these devices were successfully implanted and therefore the acute success rate was 100% for both pacemaker and ICD implantation. There were no CRT device implantations performed as the initial device implantation in the simple CHD group.

In the complex CHD group, 60, 43 and 15 patients were initially scheduled for pacemaker, ICD and CRT implantations, respectively. In this group, all pacemaker and ICD implantations were successful, but in 3 cases in which the initial plan was to implant a CRT device, this was unsuccessful due to the inability to place the CS leads (acute

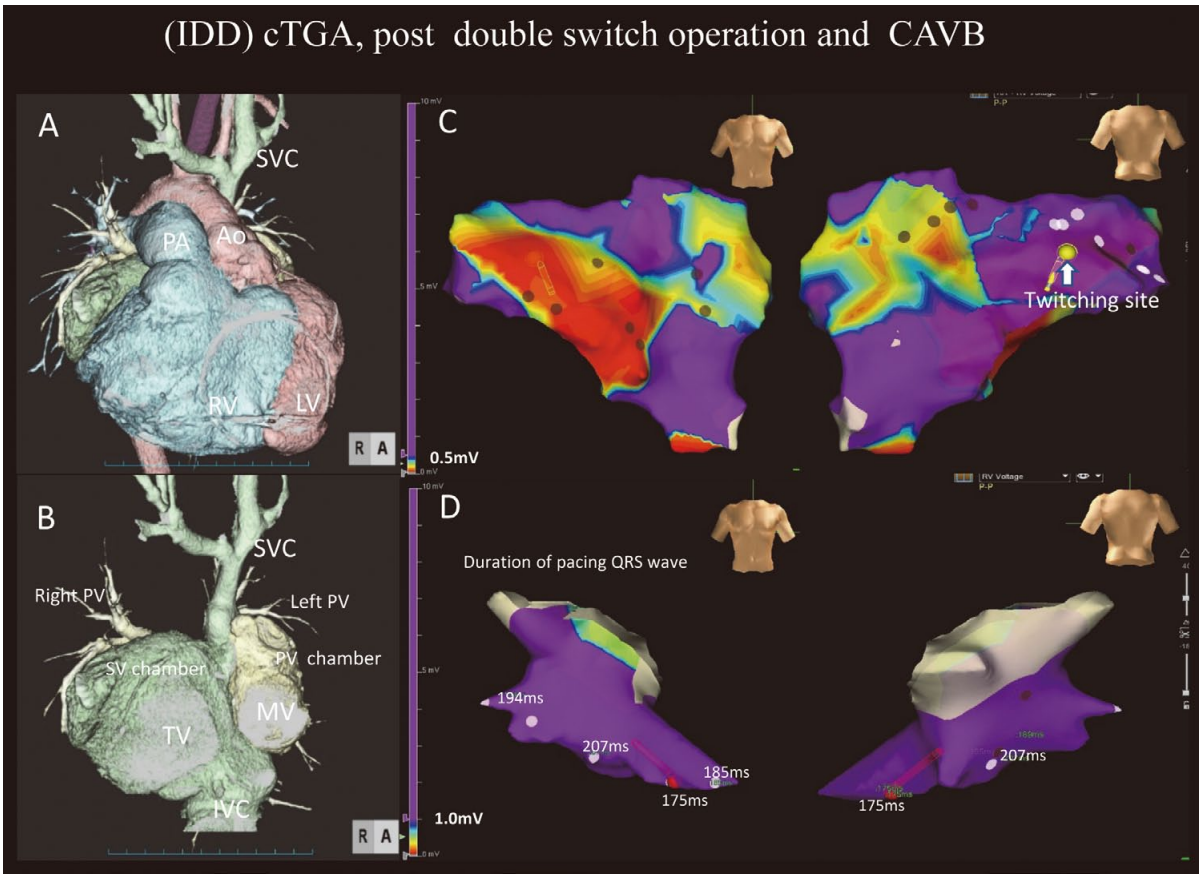

Figure 2. 3-D mapping system (Ensite NavX)-guided pacemaker implantation for a patient with situs inversus and corrected transposition of the great arteries (who had previously undergone a double switch operation) and subsequently developed complete atrioventricular block. (**A**,**B**) 3-D computed tomography. (**C**) Voltage map of the atrium. White arrow, twitching site due to phrenic nerve capture. (**D**) Voltage map of the right ventricle. White points, pacing sites and duration of paced QRS waves. Ao, ascending aorta; CAVB, complete atrioventricular block; IDD, situs inversus and D-loop atrioventricular discordance; IVC, inferior vena cava; LV, left ventricle; MV, mitral valve; PA, pulmonary artery; PV, pulmonary venous or vein; RV, right ventricle; SV, systemic venous; SVC, superior vena cava; TV, tricuspid valve.

success rate, 80%). All 3 of these failed cases involved patients with cTGA and coronary vein (CV) anomalies. Specifically, 2 of the patients had abnormal CV distribution without CV return to the CS, and 1 patient had a severely stenotic CV. Consequently, the initially planned pacing devices for these 3 patients were changed, with 1 patient receiving a pacemaker and the other 2 patients receiving ICD. We were able to implant 1 DDD pacemaker and 1 ICD using a CS lead as the ventricular pacing lead or ICD lead via the CS for 2 patients with tricuspid atresia who had previously undergone classical Fontan operation. The ventricular lead placement by the transvenous approach was not previously possible in these 2 patients after Fontan operation. We also successfully implanted a CRT pacemaker (CRT-P) device in a patient with a double-outlet left ventricle after ventricular septation and mechanical right-side atrioventricular valve replacement, using 2 CS leads via the CS and anterior cardiac vein origin from the right atrium.[15]

Procedural Complications and Management

Complication rates associated with each of the pacing devices are listed in **Table 4**. The overall complication rate was 16% (n=22). There was no significant difference in complication rate between the simple and complex CHD groups (14% vs. 16%, P=0.95). The complication rates in pacemaker, ICD, and CRT implantations were as follows: 12% (n=9), 20% (n=10), and 25% (n=3), respectively. All 4 cases of complications associated with the CRT devices involved CRT-P implantation. The complications of pacemaker implantations included 1 hematoma, 7 lead failures (3 pacing failures, 3 lead fractures and 1 sensing failure), 1 SVC occlusion, and 1 case of device infection. Pacemaker lead failures occurred in 3 atrial and in 4 ventricular leads. Complications from ICD implantation included 2 hematomas, 1 atrial pacing lead dislodgement by Ratchet syndrome,[16] 1 ICD lead dislodgement, and 6 cases of lead failure. In particular, the cases of lead failure included 1 lead fracture of the atrial pacing lead, 1 high pacing threshold in the ICD lead, 3 sensing failures in the ICD lead, and 1 insulation failure in the ICD lead. One of the 3 sensing failures of the ICD lead was associated with the officially recalled Sprint Fidelis lead. Four of five ICD lead failures were related to sensing/pacing leads and an

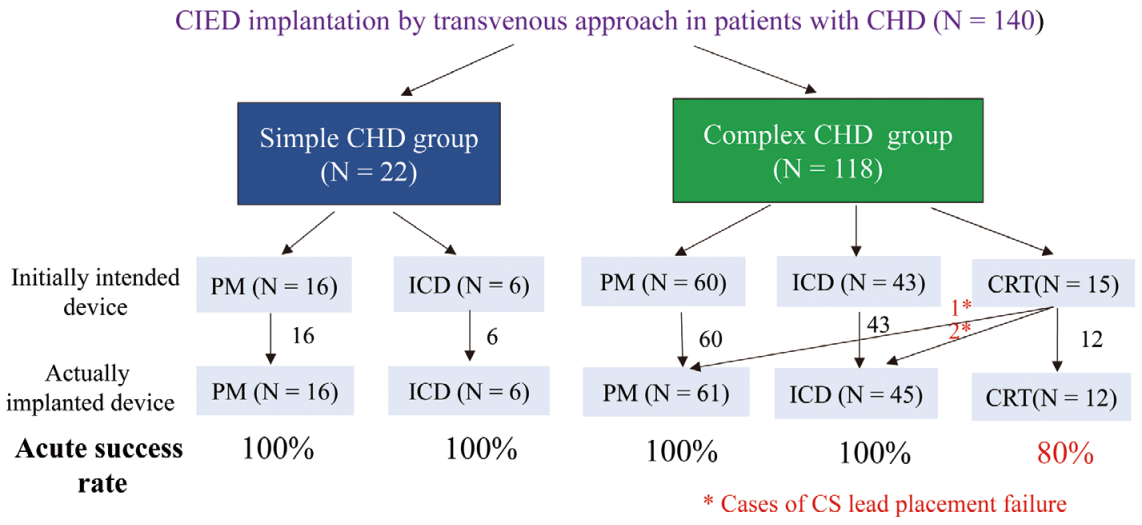

Figure 3. Acute success rate of transvenous cardiac implantable electronic device (CIED) implantation in congenital heart disease (CHD). *All 3 cases of failed coronary sinus (CS) lead placement involved corrected transposition of the great arteries and coronary vein anomalies. CRT, cardiac resynchronization therapy; ICD, implantable cardioverter defibrillator; PM, pacemaker.

Table 4. CIED-Associated Complications

	Overall patients (n=140)	CHD group	
		Simple CHD (n=22)	Moderate/severe CHD (n=118)
Follow-up (years)	8.1±5.3	7.0±4.4	8.3±5.5
Overall complication	22 (16)	3 (14)	19 (16)
Perioperative period			
Pneumothorax/Hemothorax	0 (0)	0 (0)	0 (0)
Hematoma	5 (3.6)	0 (0)	5 (4.2)
Cardiac tamponade	0 (0)	0 (0)	0 (0)
Pacemaker lead dislodgement	1 (0.7)	0 (0)	1 (0.8)
ICD lead dislodgement	1 (0.7)	0 (0)	1 (0.8)
Perforation	0 (0)	0 (0)	0 (0)
Long-term period			
Lead failure	13 (9.3)	3 (14)	10 (9.3)
Pacemaker lead			
High pacing threshold/pacing failure	3 (2.1)	0 (0)	3 (2.5)
Fracture	3 (2.1)	0 (0)	3 (2.5)
Sensing failure	1 (0.7)	1 (4.5)	0 (0)
ICD lead			
High pacing threshold	1 (0)	1 (4.5)	0 (0)
Sensing failure of ICD lead	3 (2.1)	1 (4.5)	2 (1.7)†
Insulation failure of ICD leads	1 (0.7)	0 (0)	1 (0.8)
CRT(CS) lead			
High pacing threshold of CRT lead	1 (0.7)	0 (0)	1 (0.8)
Perforation	0 (0)	0 (0)	0 (0)
SVC occlusion	1 (0.7)	0 (0)	1 (0.8)
Systemic embolization	0 (0)	0 (0)	0 (0)
Device infection	1 (0.7)	0 (0)	1 (0.8)
Revision	12 (8.5)	1 (4.5)	11 (9.3)
Device-related mortality	0 (0)	0 (0)	0 (0)

Data given as mean±SD or n (%). †One lead failure was associated with the recalled ICD lead: Sprint Fidelis 6949 (Medtronics). CIED, cardiac implantable electronic devices; SVC, superior vena cava. Other abbreviations as in Tables 1,2.

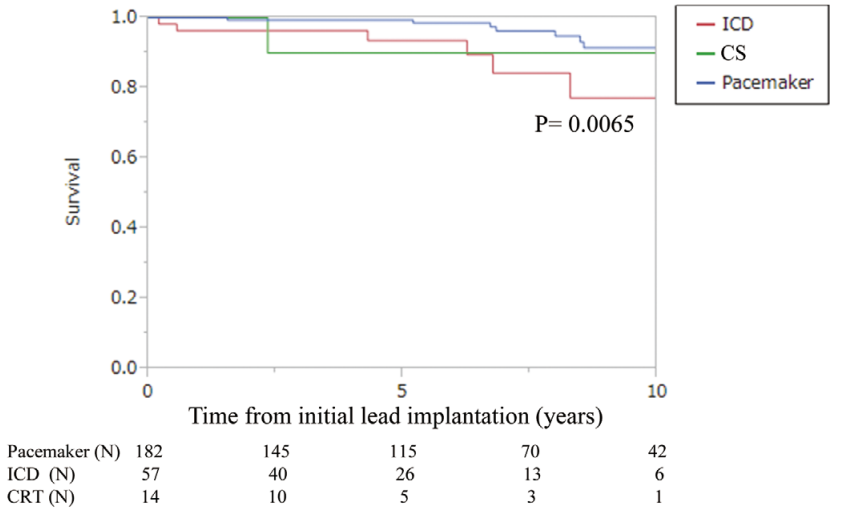

Figure 4. Kaplan-Meyer survival rates for freedom from lead failure in pacemaker, implantable cardioverter defibrillator (ICD) and coronary sinus (CS) leads. Two dislodged leads (1 pacemaker and 1 ICD lead) were excluded.

insulation failure, which are presumed externalizations of defibrillator lead conductors. Complications in CRT-P implantation included 2 hematomas and 1 high pacing threshold in the CRT lead. Device infection occurred in 0.7% of all patients (n=1). We performed 12 revisions for complication management including: 1 lead extraction, 5 lead extraction and new lead implantations, 2 lead repositions, and 4 additional lead placements. There were no procedural complications. There were 2 patients in whom CEID implantation was performed under 14 years of age in this study, including 1 pacemaker implantation at 14 years old and 1 ICD implantation at 12 years old. Fortunately, they had no lead failure associated with lead extension due to body growth.

Lead Survival Rate and Risk Factors for Lead Failure

Kaplan-Meier estimates for freedom from lead complications for pacing, ICD and CS leads are shown in **Figure 4**. The 10-year survival of the pacemaker, ICD, and CS leads was 91%, 77%, and 91%, respectively. Over 10 years, ICD lead survival was significantly lower than pacemaker lead survival (ICD vs. pacemaker leads, P=0.0065, ICD vs. CRT leads, P=0.96). Taking into account the 9 recalled ICD leads, ICD lead survival was still significantly lower than pacemaker lead survival (P=0.0076).

There was no significant difference in pacing and ICD lead survival between the simple and complex CHD groups (simple vs. complex CHD: pacemaker leads, 96% vs. 91%, P=0.68; ICD leads, 50% vs. 81%, P=0.30). On multivariate analysis, there were no risk factors associated with lead failure (atrial lead: OR, 0.90; 95% CI: 0.25–3.65; P=0.88; age <16 years: OR, 1.4; 95% CI: 0.07–8.16; P=0.76; ICD lead: OR, 2.6; 95% CI: 0.74–10.2; P=0.13; complex CHD: OR, 0.80; 95% CI: 0.23–3.69; P=0.76).

Outcomes of Pacing Device Implantation

During the 8±5 years of follow-up, 3 DDD pacemaker upgrades for VVI pacing, 2 CRT upgrades (1 upgrade to CRT-P and 1 upgrade to CRT-D) due to systemic ventricular dysfunction, and 2 device changes from pacemaker to ICD due to episodes of ventricular tachycardia were performed. Appropriate therapy occurred in 25% (n=14) of the 51 patients with ICD, while inappropriate therapy occurred in 16% (n=8) of these patients. Twelve patients (1 patient in the simple CHD group and 11 patients in the complex CHD group), died during the follow-up period (**Table 1**). Specifically, 8 patients died from heart failure and 1 died from liver cancer. Moreover, 3 patients died suddenly after pacemaker implantation (2 repaired TOF, 1 repaired complete atrioventricular septal defect), suggesting sudden cardiac death. There were no pacing device implantation-related deaths.

Discussion

The present study has found a relatively high but acceptable overall CIED-related complication rate in patients with CHD. The ICD leads had significantly lower lead survival due to lead failure compared with the pacemaker leads. No independent risk factors for lead failure were detected. Although the acute success rate of pacemaker and ICD implantation was sufficient (100% success), CRT implantation (CS lead placement) in patients with cTGA had the potential to be unsuccessful due to CV anomalies.

CIED-Related Complications

The overall CIED-related complication rate in non-CHD patients is 5–10% or around 12.5%.[17–20] The incidence may vary according to the length of the follow-up period or the definition of CIED-related complications. The overall complication rate in the present study was 16%, which was slightly higher than had previously been reported in non-CHD patients. The higher incidence of CIED-related complications in patients with CHD (especially in the patients with complex CHD), was predictable due to the difficulties associated with venous route, complex geometry, and widespread low-voltage areas.[9] Although there was a relatively high complication rate overall in the present study, the complication rate was not significantly different between the simple and complex CHD groups. Manipulations such as catheter intervention for stenotic veins or intracardiac shunts prior to device implantation, and the use of 3-D mapping systems at device implantation may have decreased the complication rate in the complex CHD group.

Lead Longevity

We compared lead survival between the pacemaker, ICD, and CS leads, and unexpectedly noted a significantly lower lead survival in the ICD leads compared with the pacemaker leads. Although a high incidence of ICD lead failure has been suggested in patients with CHD,[21] no studies have compared ICD lead failure occurrence to that of pacemaker or CS leads. The reduced ICD longevity may be attributed to the complex anatomy of CHD, the thick and complex composition of ICD leads, and the relatively young patient age (and hence higher level of daily activity placing significant mechanical stress on the leads). The recalled ICD lead also influenced lead longevity results.[22] Although only 1 lead was recalled (out of 9), ICD leads, including 8 Sprint Fidelis leads, had lead failure in this study. Meticulous follow-up of these leads is imperative due to the possibility of future lead failure.[23] In addition, given that the age at which CEID implantation is performed is decreasing, CIED implantation in children should include careful monitoring of lead failure due to lead extension by body growth.

CS Lead Placement for cTGA

All of the pacemaker and ICD leads were successfully placed, even in the complex CHD group; in contrast, however, the success rate of lead placement for the CRT devices in the complex CHD group was comparatively lower (80%). The unsuccessful CS lead placement was associated with CV anomalies associated with cTGA. A high incidence of CV anomalies such as separate CS ostia (CSOS), dual CSOS, and a vein of Marshall without CSOS in patients with cTGA has been reported.[24,25] In this study there were difficulties in CS lead placement for CRT candidates with cTGA. Pre-procedural confirmation of the morphology of the CV and CS opening using various cardiac imaging modalities (e.g., computed tomography, cardiac magnetic resonance imaging, and venous phase of coronary artery angiography) before CS lead implantation is therefore important in cases involving complex CHD, especially cTGA. If CV drainage via CSOS is not observed, lead implantation of the systemic right ventricle using an epicardial lead should be considered.

Other Factors

The successful implantation of CIED in patients with complex CHD may be hindered by venous stenosis, intracardiac shunts, complex geometry, or low-voltage electrical signals. Therefore, using a 3-D navigation system for complicated cases is often useful, especially for atrial lead placement in the double switch operation for congenital cTGA, atrial switch operation for TGA, and in cardiac inversion cases after double switch operation. In these patients the intracardiac structure is complicated, often including the presence of a low-voltage region in the atrium, and the atrial lead can usually be placed only in the left-side atrial appendage (right-side atrial appendage in the case of situs inversus). This placement, however, tends to cause phrenic nerve twitching. A 3-D mapping system can mark the site of twitching. In such cases, if ventricular lead placement is needed, the guidance of a 3-D mapping system is also useful because the ventricular lead should be placed in the contralateral ventricle via the Mustard or Senning route.

The Ensite NavX system is especially beneficial in 3-D mapping because it can display the lead tip in real time and also helps to reduce radiation exposure.[6,7] Intracardiac shunt and venous stenosis increase the risk of a thrombotic event and SVC syndrome.

While cardiac surgery (if possible, minimally invasive surgery using an epicardial lead system) is a viable option for CHD patients with intracardiac shunt or SVC syndrome, an alternative option of closing cardiac shunts and stent implantation for stenotic veins before or during transvenous CIED implantation tends to reduce transvenous approach-related complications.[26] Recent advances in lead extraction techniques have contributed to the increasing number of transvenous lead implantations performed in patients with CHD.[27,28] The relatively young age at CIED implantation of patients with CHD compared with non-CHD patients requires long-term follow-up after implantation. Thus, the number of lead extractions for lead-related issues and device infection will inevitably increase in the future. We encourage the use of a remote-monitoring system[29,30] for early detection of device/lead issues and arrhythmic events in patients with CHD who have undergone CIED implantation.

Study Limitations

This study had several limitations, including small sample size, heterogeneous cardiac disease, lack of a control group without CHD, and its retrospective nature. Furthermore, we could not completely exclude the effects of cardiac disease, age, or device era on lead survival in pacemaker, ICD and CS leads.

Conclusions

The outcomes of transvenous CIED implantation for patients with CHD seem acceptable, although there tends to be a higher incidence of complications in this population than in the general population. Successful CS lead placement for CRT candidates with cTGA may be inhibited by CV anomalies. Moreover, ICD lead longevity tended to be lower than that of pacemaker leads in the CHD population.

Acknowledgment

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Disclosures

N.H. is a member of *Circulation Reports*' Editorial Team. The other authors declare no conflicts of interest.

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