

Treatment of junctional ectopic tachycardia before and during total cavopulmonary connection procedure for the patients with asplenic heart.

Kosei Tanaga, M.D., Ph.D.¹⁾, Shigeru Tateno, M.D., Ph.D.¹⁾,
Yasunori Hiranuma, M.D., Ph.D.¹⁾, Takayasu Ishikawa, M.D., Ph.D.¹⁾,
Kouzo Matsuo, M.D., Ph.D.¹⁾, Koichirou Niwa, M.D., Ph.D.²⁾

¹⁾Department of Cardiology, Chiba Cerebral and Cardiovascular Center,
575 Tsurumai, Ichihara, Chiba 290-0512, Japan

²⁾Department of Cardiology, Cardiovascular Center, St Luke's International University Hospital,
9-1 Akashi-cho, Chuo-ku, Tokyo 104-8560 Japan

Abstract

Background: Non-surgically related junctional ectopic tachycardia (JET) in asplenic heart has been rare, however if it occurs, such tachycardia make serious influence on morbidity and mortality in patients with total cavopulmonary connection (TCPC). Our aim of this study was to evaluate efficacy and safety of atrio-ventricular node (AVN) modification and DDD pacemaker implantation during TCPC procedure in patients with asplenic heart and JET.

Methods: A retrospective study was performed on the medical records and electronically recorded data of electro-physiological studies.

Results: A total of 3 patients with asplenic heart were diagnosed as paroxysmal JET prior to TCPC. First onset of JET were 9 days of age in case 1, 7 year of age in case 2 and 1 year of age in case 3. On electro-physiological studies, narrow QRS tachycardia with irregular RR interval was induced and terminated by ventricular pacing, and no retrograde conduction via AVN and no twin functional AVN were observed in any patient. DDD pacemaker implantation and cryo-ablation for AVN were performed during TCPC procedure in all 3 patients. Anti-arrhythmic treatment prior to TCPC procedure were radiofrequency catheter ablation in 2 and oral amiodarone administration in the other one. Transient complete atrio-ventricular block occurred in 2 patients. JET had never recurred after the operation in all patients during about 10 years follow up.

Conclusion: DDD pacemaker implantation and cryo-ablation for AVN modification during TCPC procedure could be method of choice for elimination of the JET in patients with asplenic heart.

Key words : junctional ectopic tachycardia, total cavopulmonary connection, asplenia

Background

Recent surgical advances made it possible that many children with complex heart disease survive after corrective and palliative repair. Tachycardia after cardiac repair is recognized to be an important source of early and late morbidity and mortality. Since Fontan's report the first successful right heart bypass directing the entire systemic venous blood flow to the

pulmonary arteries in a patient with tricuspid atresia¹⁾, several studies have reported that supraventricular tachycardia (SVT) due to atrial flutter, atrial fibrillation, incisional atrial reentrant tachycardia, or atrioventricular reciprocating tachycardia occurs frequently as a major arrhythmia after Fontan repair, especially in asplenic heart^{2,3)}. Total cavopulmonary connection (TCPC) resulted in less frequency of

Received July 27, 2017 Accepted September 25, 2017

Corresponding author : Kosei Tanaga, M.D., Ph.D., Department of Cardiology, Chiba Cerebral and Cardiovascular Center,
575 Tsurumai, Ichihara, Chiba 290-0512, Japan, E-mail: t.kosei.5150@m8.dion.ne.jp

tachyarrhythmia compared with the original atriopulmonary connection^{4,5,6}). Although the incidence of SVT was reduced, asplenic heart is often associated with SVT, which may compromise hemodynamic status. Junctional ectopic tachycardia (JET) is an uncommon SVT thought to arise from abnormal automaticity at the atrioventricular node (AVN) or His bundle⁷. JET is an increasingly recognized as a malignant arrhythmia arising in the postoperative setting after surgery for congenital heart disease⁸. JET in children is classified into three categories: congenital JET without structural heart disease, JET immediate after cardiac surgical repair of complex congenital heart disease, and non-surgically related JET with congenital heart disease⁹. Non-surgically related JET with congenital heart disease is very rare and the exact electrophysiological mechanisms have not been elucidated. Furthermore, treatment for non-surgically related JET during TCPC has not been established. We experienced 3 consecutive patients with asplenic heart and non-surgically related JET prior to TCPC. In this study, we performed electrophysiological study to reveal the mechanism of non-surgically related JET and evaluated the efficacy and safety of DDD pacemaker implantation and AV node modification during TCPC for patients with asplenic heart.

Methods

Patients

From February 2006 to September 2007, we performed DDD pacemaker implantation and

AV node modification during TCPC for 3 consecutive patients with asplenic heart and non-surgically related JET. JET was defined in terms of the following electrocardiographic (ECG) and electrophysiological characteristics: (1) narrow QRS tachycardia with the same morphology as in sinus or atrial rhythm; (2) features of either ventriculoatrial (VA) dissociation or variable VA conduction; (3) in the case of a 1:1 AV relation, an atrium not actively involved in tachycardia maintenance; and (4) no signs of typical AV reentrant or AV nodal reentrant tachycardia⁹.

Methods

A retrospective study was performed on the medical records and electronically recorded data of electro-physiological studies (EPS).

Results

Table 1 shows patient characteristics, features of JET and treatment. Table 2 shows electrophysiological characteristics and drug efficacy of JET.

Case 1

Four-year-old boy had been diagnosed at birth with single right ventricle (SRV), common atrio-ventricular canal (CAVC), pulmonary valve stenosis (PS) and bilateral superior vena cava (SVC). He had undergone bilateral bidirectional Glenn (BDG) shunt at 1 year of age. He had suffered from frequent episodes of paroxysmal SVT since 9 days of age (Figure 1). He was noted to have a narrow QRS tachycardia with the same

Table 1

Pt no.	Sex	JET onset	Mean heart rate at presentation (bpm)	R-R irregularity	Age at operation (y)	Medication before TCPC	Radiofrequency catheter ablation before TCPC	Cryo-ablation during TCPC
1	M	1 year	250	+	4	—	+	+
2	M	7 years	190	+	13	Beta-blocker and digoxin	+	+
3	M	1 year	240	+	13	Amiodarone	—	+

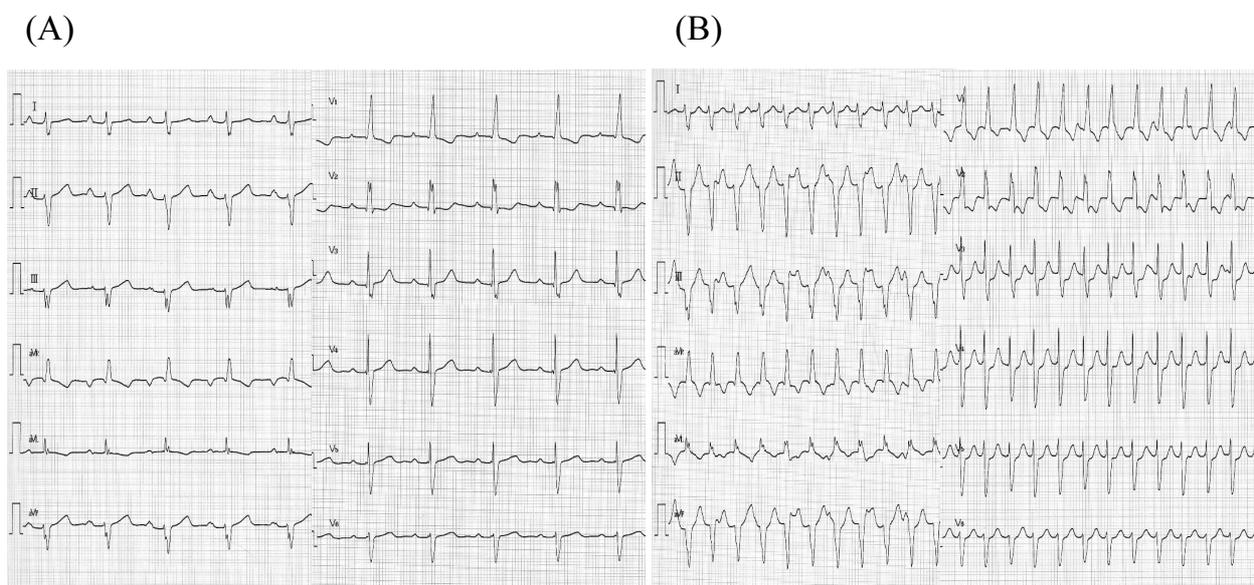
Patient characteristics, features of junctional ectopic tachycardia (JET) and treatment.

Table 2

Pt no.	Adenosine sensitivity	Verapamil sensitivity	Valsalva maneuver	Induction by RAP/VAP	Termination by RAP/VAP	VA conduction (V pacing/JET)
1	+	+	n.d.	+/+	-/+	Diss/Diss
2	n.d.	n.d.	+	+/+	-/+	Diss/Diss
3	n.d.	+	n.d.	+/+	n.d.	Diss/Diss

n.d.=not done

Electrophysiologic characteristics and drug efficacy of junctional ectopic tachycardia (JET). We performed rapid atrial pacing (RAP) and ventricular apical pacing (VAP) in electro-physiological studies.

**Figure 1**

Twelve lead electrocardiogram of the patient. (A) is normal sinus rhythm and (B) is junctional ectopic tachycardia.

QRS as in sinus rhythm. Heart rate during tachycardia was 250/min. Tachycardia was terminated by intravenous verapamil. We performed EPS at 4 year of age, prior to TCPC (Figure 2). Narrow QRS tachycardia without VA conduction was induced by atrial burst pacing and terminated by adenosine injection or rapid ventricular burst pacing. Tachycardia could not be terminated by atrial burst pacing. QRS morphology was same as sinus rhythm. RR

interval was irregular. Antegrade AV conduction only via posterior AVN was confirmed. EPS showed no VA conduction during ventricular pacing or no overdrive suppression. We created electroanatomical map of the atrium and performed radiofrequency catheter ablation (RFCA) around the posterior AVN avoiding the area where His bundle electrograms (HBEs) were recorded (Figure 3). During sinus rhythm or atrial pacing, we carefully performed RFCA

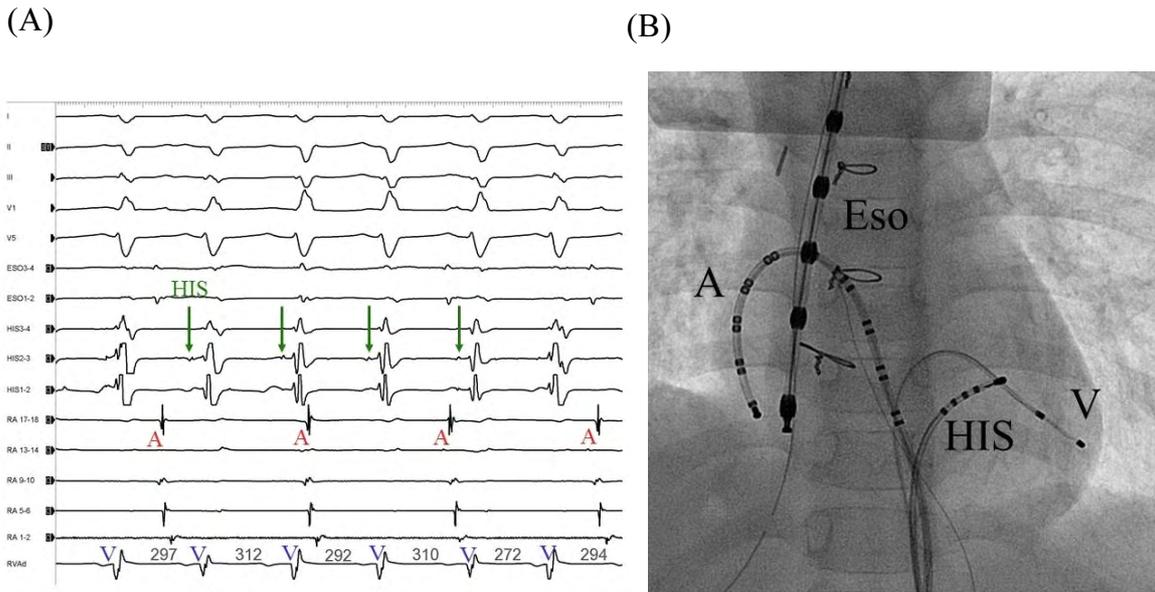


Figure 2

- (A) Intracardiac electrocardiography recorded during junctional ectopic tachycardia (JET). Electrograms of right atrium (A), right ventricle (V) and His bundle (HIS) were showed by arrows.
- (B) Fluoroscopic anterior-posterior view showing catheter positions. Catheters are positioned in the right atrium (A), the single right ventricle (V), common atrio-ventricular where His bundle electrogram was identified (HIS), and the esophagus (Eso).

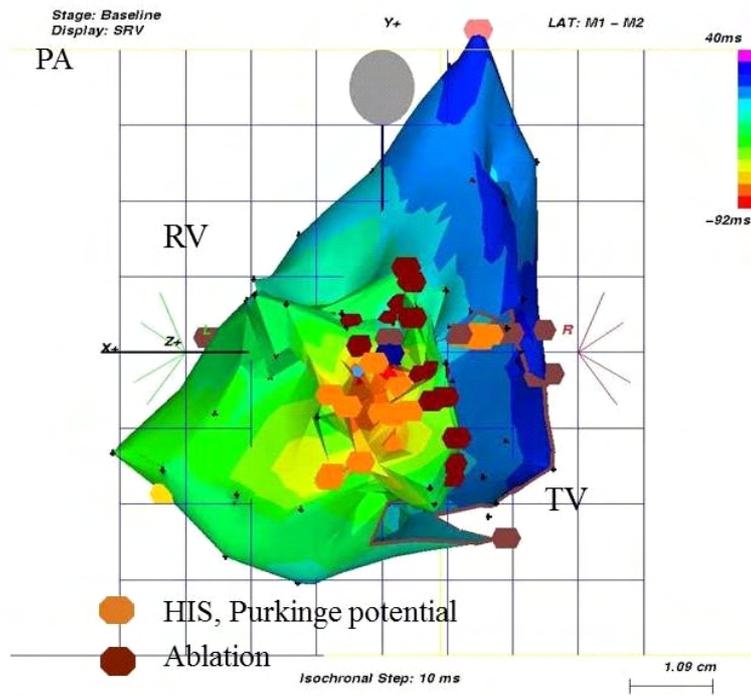


Figure 3

Electroanatomical map of the atrium during junctional ectopic tachycardia (JET). Brown dots denote the radiofrequency catheter ablation (RFCA) sites and orange dots denote the His bundle and Purkinje potentials.

monitoring atrio-His (A-H) interval. If A-H prolongation appeared, we immediately stopped RFCA. We finished RFCA without AV block. After RFCA, JET could not be induced. He underwent surgical intervention 4 days after catheter ablation. However, JET occurred at anesthesia induction. So TCPC, cryo-ablation for posterior AVN and DDD pacemaker implantation were performed. Surgical cryo-ablation was performed based on the previously created electroanatomical map. Accelerated junctional rhythm occurred immediately after the operation. JET has never recurred for 11 years and 11 months without AV block.

Case 2

Thirteen-year-old boy had been diagnosed at birth with SRV, single atrium (SA), patent ductus arteriosus (PDA) and bilateral SVC. Previous intervention were pulmonary artery banding and PDA ligation at 1 month of age and BDG shunt at 3 years of age. He had suffered from paroxysmal SVT several times since 7 year of age. Heart rate during tachycardia was 190/min. Oral Beta-blocker and digoxin were effective for prevention of tachycardia. Tachycardia was

terminated by breath-holding and vomiting. We performed EPS at 13 years of age, prior to TCPC. Narrow QRS tachycardia without VA conduction was induced by atrial and ventricular burst pacing and terminated by ventricular burst pacing. Tachycardia could not be terminated by atrial burst pacing. We recognized two HBEs by electroanatomical mapping, but QRS morphology remained unchanged during atrial pacing around the two HBEs identified sites. We concluded that he had antegrade AV conduction only via functional posterior AVN. RFCA was performed around the posterior AVN. However, we couldn't continue RFCA due to occurrence of AV block. So, RFCA was not effective. AV conduction immediately recovered after RFCA. 1 month later he underwent surgical intervention: TCPC, cryo-ablation of both anterior and posterior AVNs near two HBEs identified sites and tricuspid Isthmus, and DDD pacemaker implantation (Figure 4). Complete AV block (CAVB) and junctional rhythm occurred immediately after the operation and recovered after one week. JET has not recurred for 11 years and 4 months with second degree AV block.

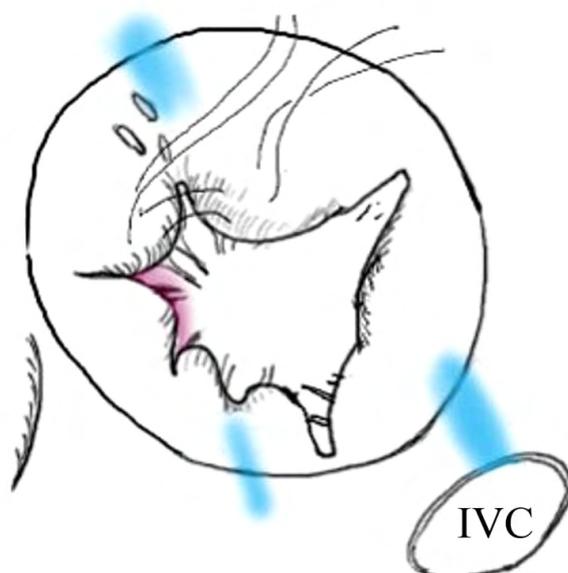


Figure 4

Cryo-ablation was performed for anterior atrioventricular node (AVN), posterior AVN and tricuspid isthmus during total cavo-pulmonary connection procedure. Blue areas show the sites where cryo-ablation was performed. Inferior vena cava (IVC) is presented.

Case 3

Eight-year-old boy had been diagnosed at birth with SRV, SA, PDA and bilateral SVC. He had undergone BT shunt and bilateral BDG shunt. He had suffered from episodes of paroxysmal SVT since 1 year of age. Heart rate during tachycardia was 240/min. Tachycardia was terminated by intravenous administration of verapamil. We performed EPS at 8 years of age, prior to TCPC. Narrow QRS tachycardia without VA conduction was induced by atrial and ventricular burst pacing and terminated spontaneously. Antegrade AV conduction only via posterior AVN was confirmed. Catheter ablation was not performed in this case. He was administered oral amiodarone before surgical intervention. He underwent TCPC, cryo-ablation for posterior AVN and DDD pacemaker implantation, 4 days after EPS. Ventricular tachycardia, CAVB and junctional rhythm occurred immediately after the operation and terminated 5 days after those happened. JET was not recurred for 9 years and 11 months without AV block.

Discussion

The precise etiology of JET is unknown, but JET is believed to originate from abnormal conduction tissue in the region of the AVN, proximal to the bifurcation of the HIS bundle⁷⁾. In our study, tachycardia was induced by both atrial and ventricular burst pacing and terminated by ventricular burst pacing. Our EPS results suggested that triggered activity or reentry could be the mechanism of JET. Furthermore, termination only by ventricular burst pacing in 2 cases suggests that arrhythmogenic area could be below AVN (Figure 5).

In the asplenic heart, isomerism is usually associated with an abnormal conduction system which may involve the coexistence of two distinct twin AVNs with a connecting sling⁹⁾. It is reported that JET is often associated with twin AVNs and AVN modification by RFCA is effective^{10,11)}. In this series, no twin functional AVNs were observed. We performed RFCA around the posterior AVN to 2 patients. One was effective and the other was not effective.

However, JET recurred at anesthesia induction before TCPC in patient whose RFCA was effective. So, we performed cryo-ablation for AVN and DDD pacemaker implantation during TCPC procedure to both patients who underwent RFCA. In Case 2, we recorded two HBEs and it is suggested that he had functional posterior AVN and non-functioning anterior AVN from EPS. SVT originating from the non-functioning AVN is reported¹⁰⁾, so we performed cryo-ablation for both anterior AVN and posterior AVN. Complete AVB before TCPC should be avoided. Due to the risk of AVB, RFCA for AVN for the patients with single AVN is very difficult. In case 3, we performed only EPS to make the guide map for cryo-ablation during TCPC. Instead of RFCA, we performed administration of oral amiodarone before TCPC. Many antiarrhythmic therapies have been employed with variable rates of success in management of JET, and efficacy of the class 3 agent amiodarone is reported⁷⁾. However, long-term therapy with amiodarone is unwarranted due to its side effects. We stopped administration of oral amiodarone 1 month after TCPC.

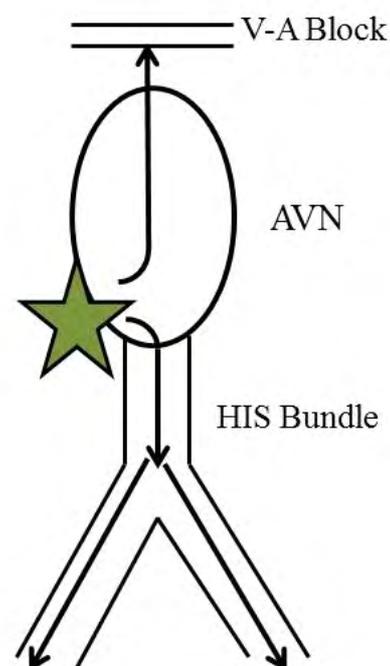


Figure 5

Possible Scheme of the cardiac conduction system in 3 cases. Arrhythmogenic area (☆) could be below AVN and upper HIS bundle.

JET has never recurred after TCPC in all patients during about 10 years follow up, except early postoperative period. Second degree AV block has persisted in only one patient. The others were without AV block and implanted DDD pacemaker did not work during about 10 years follow up. However if JET recurred after TCPC in adulthood and antiarrhythmic medication is not effective, further RFCA for AVN would be considered. Since further RFCA might cause CAVB, DDD pacemaker implantation during TCPC would be necessary. Because dyssynchrony is not favorable for ventricular function, we tried to avoid AV block. If further RFCA for AVN caused CAVB and dyssynchrony due to ventricular pacing caused heart failure, cardiac resynchronization therapy (CRT) could be considered.

Conclusion

DDD pacemaker implantation and cryo-ablation for AVN during TCPC procedure could be method of choice for elimination of the JET in patients with asplenic heart. Preoperative EPS will provide useful information for treatment of JET during TCPC.

Reference

- 1) Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;**26**:240-48.
- 2) Gelatt M, Hamilton RM, McCrindle BW, et al. Risk factors for atrial tachyarrhythmias after Fontan operation. *J AM Coll Cardiol* 1994;**24**:1735-41.
- 3) Bae EJ, Noh CI, Choi JY, et al. Late occurrence of adenosine-sensitive focal junctional ectopic tachycardia in complex congenital heart disease. *J Interv Card Electrophysiol* 2005;**12**:115-22.
- 4) Long-term results of the lateral tunnel Fontan operation. Stamm C, Friehs I, Mayer Jr, et al. *J Thorac Cardiovasc Surg.* 2001;**121**:28-41.
- 5) Stephenson EA, Lu M, Berul CI, et al. Arrhythmias in a contemporary fontan cohort: prevalence and clinical association in a multicenter cross-section study. *J Am Coll Cardiol* 2010;**56**:890-96.
- 6) Balagi S, Daga A, Bradley DJ, et al. An international multicenter study comparing arrhythmia prevalence between the intracardiac lateral tunnel and the extracardiac conduit type of Fontan operations. *J Thorac Cardiovasc Surg* 2014;**148**:576-81.
- 7) Garson A, Gillette PC. Junctional ectopic tachycardia in children: electrocardiography, electrophysiology, and pharmacologic response. *Am J Cardiol* 1979;**44**:298-02.
- 8) Villain E, Vetter VL, Garcia JM, et al. Evolving concepts in the management of congenital junctional ectopic tachycardia. A multicenter study. *Circulation* 1990;**81**:1544-49
- 9) Bae EJ, Noh CI, Choi JY, et al. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. *Pacing Clin Electrophysiol* 2005;**28**:126-34
- 10) Ajiki J, Hayami N, Kasaoka Y, et al. Supraventricular tachycardia originating from the posterior node in the univentricular heart with single atrium. *Int Heart J* 2007;**48**:253-59
- 11) Toyohara K, Yoshimoto J, Ozaki N, et al. Successful catheter ablation of reentrant junctional tachycardia in a patient with asplenia syndrome before total-cavo-pulmonary connection. *Pacing Clin Electrophysiol* 2010;**33**:e43-45