CASE REPORT

Successful cryoablation of incessant fast-slow atypical atrioventricular nodal reentrant tachycardia in a child with tachycardia-induced cardiomyopathy

Taşikardi ilişkili kardiyomiyopatili bir çocukta "incessant fast-slow" atipik atriyoventriküler nodal reentran taşikardinin başarılı kriyoablasyonu

Yakup Ergül, M.D.,[1] Ahmet İrdem, M.D.,[1] Osman Esen, M.D.,[2] Alper Güzeltaş, M.D.[1]

[1]Department of Pediatric Cardiology, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

[2]Department of Anesthesia, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

Summary– A 15-year-old female patient presented at the clinic with heart failure (HF). A 12-lead electrocardiogram showed a heart rate of 170 bpm; negative P waves in leads DII, DIII, aVF; and long RP tachycardia (LRPT). Echocardiography demonstrated a shortening fraction (SF) of 20%. An electrophysiology study during tachycardia revealed an atrial-His time of 52 milliseconds and a His-atrial interval of 295 milliseconds. During ventricular entrainment, the postpacing interval-tachycardia cycle length was measured at 225 milliseconds. There was a pseudo V-A-A-V response. These findings confirmed the diagnosis of atypical atrioventricular nodal re-entrant tachycardia (aAVNRT). Successful slow pathway cryoablation was performed with an 8-mm-tip cryocatheter. After 2 weeks, the SF was measured as 34%. During a 2-year follow-up period, no recurrence was observed. In conclusion, fast-slow aAVNRT should be a part of the differential diagnosis of incessant LRPT leading to HF. Cryoablation can be used successfully in cases of aAVNRT.

Özet– On beş yaşındaki kız hasta merkezimize kalp yetersizliği ile başvurdu. On iki kanal elektrokardiyografisinde kalp hızı 170/dk olan hastada DII-III ve avF'de negatif P dalgası olan uzun RP aralıklı taşikardi mevcuttu. Ekokardiyografide sol ventrikül fonksiyonları azalmış olan hastada fraksiyone kısalma %20 idi. Elektrofizyolojik çalışmada atriyal-His zamanı 52 ms, HA 295 ms olan hastada ventriküler "entrainment" ile PPI-TCL (postpacing interval-tachycardia cycle length) 225 ms idi ve yalancı V-A-A-V yanıtı vardı. Bu veriler ile atipik atriyoventriküler reentran taşikardi (aAVNRT) tanısı konulan hastaya 8 mm uçlu kriyokateter ile başarılı kriyoablasyon uygulandı. İşlemden iki hafta sonra sol ventrikül fonksiyonları düzeldi ve kısalma fraksiyonu (KF) %34'e yükseldi. İki yıllık izlem boyunca taşikardi nüksü izlenmedi. Sonuç olarak kalp yetersizliği ile başvuran uzun RP'li taşikardilerde ayırıcı tanıda aAVNRT'de düşünülmelidir. Bu olgularda kriyoablasyon başarılı şekilde kullanılabilir.

The atrioventricular node is known to have a dual conduction system that can cause reentrant tachycardia. Atrioventricular nodal reentrant tachycardia (AVNRT) is a common, regular type of tachycardia. While the slow pathway is antegrade and the fast pathway is retrograde in typical AVNRT, the reverse is true for atypical AVNRT (aAVNRT). As the fast pathway is in the interatrial septum and the slow pathway is adjacent to the coronary sinus, the earliest retrograde atrial activation is recorded in the interatrial septum in typical AVNRT and in the coronary sinus in aAVNRT.[1] While rare, incessant AVNRT can lead to tachycardia-induced cardiomyopathy.[2–5]

In this report, the case of a 15-year-old female patient who presented with palpitations, shortness of breath, and early fatigue is described; she was found to have tachycardia-related left ventricular dysfunction, diagnosed with incessant aAVNRT, and was successfully treated with cryoablation.

Received: August 20, 2017 *Accepted:* October 20, 2017

Correspondence: Dr. Yakup Ergül. Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Pediatrik Kardiyoloji Kliniği, İstanbul, Turkey.
Tel: +90 212 - 692 20 00 e-mail: yakupergul77@hotmail.com

CASE REPORT

A 15-year-old female patient presented with heart failure (HF) findings that had persisted for 3 months. An electrocardiogram (ECG) revealed a heart rate of 176 bpm; negative P waves in leads DII, DIII, and aVF; and long RP tachycardia (Fig. 1). Echocardiography showed left ventricular (LV) dilation and dysfunction, with a shortening fraction (SF) of 20%. Holter monitoring demonstrated incessant long RP tachycardia with an intermittent atrioventricular (AV) block. When tachycardia persisted despite milrinone and propafenone treatment, a decision was made to perform an electrophysiology study and ablation. Under general anesthesia, right atrium mapping was performed using the EnSite mapping system (St. Jude Medical, Inc., St. Paul, MN, USA). The differential diagnosis included focal atrial tachycardia (FAT), permanent junctional reciprocating tachycardia (PJRT), nodofascicular reentrant tachycardia (NFRT), and aAVNRT. During tachycardia, the atrial-His (AH) time was 52 milliseconds and His-atrial (HA) time was 295 milliseconds. Entrainment of sustained tachycardia was performed during ventricular overdrive pacing with a supraventricular tachycardia (SVT) cycle of 20 to 30 milliseconds, a postpacing interval-tachycardia cycle length (PPI-TCL) interval of 586-340=246 milliseconds, an interval between the last pacing stimulus and the last entrained sinoatrial (SA) electrogram minus the tachycardia ventriculoatrial (VA) interval (SA-VA interval) of 430-205=225 milliseconds, and a corrected PPI-TCL of 208 milliseconds (Fig. 2). When pacing was stopped, there was a pseudo V-A-A-V response. In addition, when His-refractory ventricular pace (HRVP) was performed, tachycardia was not affected and A was not advanced. These findings confirmed the diagnosis of aAVNRT. During EnSite system-guided mapping, the earliest VA interval was 153 milliseconds, observed near the coronary sinus ostium. As slow pathway ablation was to be performed in this area, the procedure was continued with a cryocatheter. Slow pathway cryoablation-mapping was performed with an 8-mm-tip cryocatheter at -70°C, and tachycardia ended with VA block after 3 seconds (Figs. 3a and 3b). A total of 7 lesions were treated. After the ablation, the VA conduction was concentric, decremental, and the VA block was 400 milliseconds. There was no retrograde jump. When SVT could not be induced spontaneously or with orciprenaline after a 30-minute waiting period, the procedure was completed without complications. The total procedure time was 140 minutes; the fluoroscopy time was 0 minutes. The patient was discharged with a normal sinus rhythm 1 week later. Congestive HF findings improved in follow-up;

Abbreviations:

aAVNRT	*Atypical AVNRT*
AH	*Atrial-His*
AV	*Atrioventricular*
AVNRT	*Atrioventricular nodal re-entrant tachycardia*
FAT	*Focal atrial tachycardia*
HA	*His-atrial*
HF	*Heart failure*
HRVP	*His-refractory ventricular pace*
LRPT	*Long RP tachycardia*
LV	*Left ventricular*
NFRT	*Nodofascicular reentrant tachycardia*
PJRT	*Permanent junctional reciprocating tachycardia*
PPI	*Postpacing interval*
PPI-TCL	*Postpacing interval–tachycardia cycle length*
SA	*Sinoatrial*
SF	*Shortening fraction*
SVT	*Supraventricular tachycardia*
VA	*Ventriculoatrial*
VOP	*Ventricular overdrive pacing*

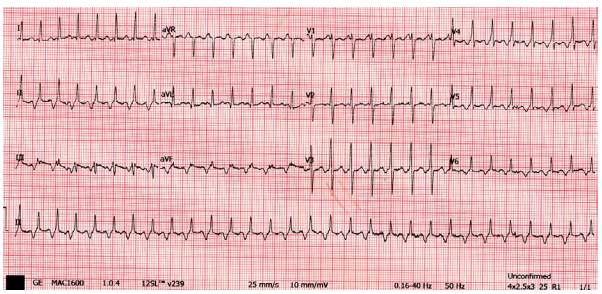

Figure 1. Initial electrocardiogram demonstrating atypical atrioventricular nodal re-entrant tachycardia.

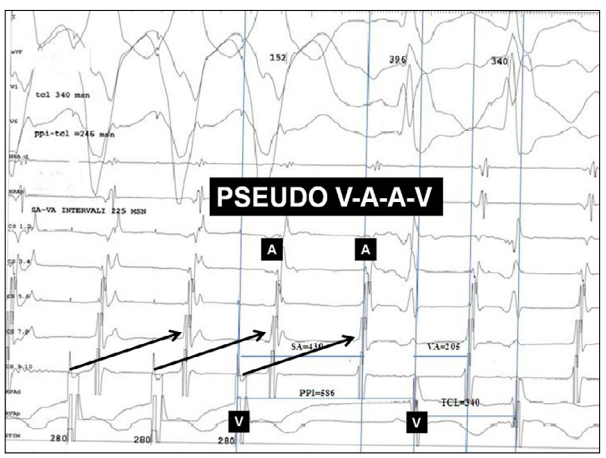

Figure 2. Postpacing interval–tachycardia cycle length and the interval between the last pacing stimulus and the last entrained sinoatrial electrogram minus the tachycardia ventriculoatrial interval as seen during the electrophysiology study.

Figure 3. **(A)** EnSite mapping system (St. Jude Medical, Inc. St. Paul, MN, USA) 3-dimensional mapping-guided cryoablation. **(B)** Successful ablation during the electrophysiology study.

the SF measured 2 weeks later was 34%. No recurrence of tachycardia was observed during 2 years' of follow-up.

DISCUSSION

To the best of our knowledge, this is the first reported case of incessant fast-slow AVNRT treated with electroanatomic mapping-guided cryoablation in a child with tachycardia-induced cardiomyopathy.

AVNRT usually presents as a paroxysmal form of tachycardia with each episode lasting for a short period of time. Approximately 10% of AVNRT cases are atypical (also known as fast-slow and sometimes slow-slow AVNRT). Retrograde slow pathway conduction plays a key role in the origin and permanence of tachycardia.[1] In AVNRT, the AH interval is short (average: 80 milliseconds) and the HA interval is long (average: 260 milliseconds). As retrograde conduction is via the slow pathway in this type of AVNRT, retrograde P waves are recorded in the second diastolic phase (long RP tachycardia). Earliest retrograde activation is recorded in the anteroseptal area. A second-degree AV block may occur during tachycardia. A negative P wave is seen in the inferior leads; a positive or isoelectric P wave is seen in D1. In a study conducted by Kaneko et al.,[7] an 81-year-old patient with aAVNRT originating from the noncoronary sinus of Valsalva was successfully treated with radiofrequency ablation. There is only 1 report of fast-slow aAVNRT presenting as incessant tachycardia-induced cardiomyopathy in an adult.[2] Jastrzębski et al.[6] reported pseudo-PJRT in an 8-year-old boy with acute decompensated HF and recurrent episodes of ventricular fibrillation. The cardiomyopathy was caused by incessant long RP tachycardia resistant to pharmacotherapy. With the help of an electrophysiology study performed due to initially suspected PJRT, aAVNRT was diagnosed and treated with radiofrequency ablation.

Differentiating between aAVRNT and long RP supraventricular tachycardia can be a challenge. The differential diagnosis must include sinus tachycardia, FAT, NFRT, and PJRT. FAT is diagnosed based on surface ECG and the clinical features of the arrhythmia.[1,7–9] In FAT, the RP interval is variable or independent. Increased pace and decreased AV node conduction may lead to tachycardia with a markedly short RP interval. Additionally, a heart rate increase or decrease in the space of 30 seconds as a result of warm-up and cool-down heart rate variability also points to FAT.[7] In PJRT, retrograde earliest activation is recorded in the posteroseptal area. No AV block is observed during this type of tachycardia, which ends with VA block. The lack of warm-up and cool-down heart rate variability, retrograde earliest activation in the anteroseptal area, and the lack of VA block in our patient led us to eliminate FAT and PJRT. Also, the ventricular overdrive pacing (VOP) maneuver provides the diagnosis in the majority of supraventricular tachycardia (SVT) cases. A post-VOP response that is ventricular-atrial-atrial-ventricular (V-A-A-V) rules in FAT while a post-VOP response that is ventricular-atrial-ventricular (V-A-V) rules out FAT (effectively ruling in AVRT or AVNRT). But, when the retrograde limb of the circuit conducts slowly, as it might in aAVNRT (such as in so-called "fast-slow" AVNRT), or slowly conducting accessory pathway (PJRT), the VA interval after the last paced beat may be longer than the pacing cycle length such that the second atrial electrogram after the last paced ventricular beat may in fact be the last atrial electrogram that was accelerated to the pacing cycle length. This is called a

pseudo V-A-A-V response, which may be incorrectly interpreted as indicating a diagnosis of FAT.[1,3,9]

Standard diagnostic criteria, starting with common pacing maneuvers, are applied to more common short RP supraventricular tachycardias. Thus, in long RP tachycardias, the diagnostic values of these pacing maneuvers are lower and there are different data regarding cut-off values.[9,10] However, as a long post-pacing interval (PPI) is very common in aAVNRT, a PPI–TCL >115 milliseconds, or paradoxically, when the AH (SVT)<AH (normal sinus rhythm), AVNRT can be differentiated from orthodromic reciprocating tachycardia and PJRT. The presence of SA-VA >85 milliseconds is also a typical finding in aAVNRT. Presence of other entrainment criteria, like PPI <110 milliseconds, VA <85 milliseconds, and HA <0 milliseconds, is only relatively sensitive, but 100% specific for orthodromic reciprocating tachycardia. For PJRT, the criteria are PPI–TCL <115 milliseconds (or corrected PPI-TCL <110 milliseconds), VA <85 milliseconds, HA <0 milliseconds, AH <20 milliseconds.[4,8]

This case also demonstrates the feasibility of a limited fluoroscopy approach with the use of an electroanatomical mapping system for cryoablation of the slow pathway in AV nodal reentrant tachycardia in children. Catheter ablation is the established curative therapy for pediatric tachyarrhythmias and electroanatomical mapping systems have been used for more than a decade to localize tachycardia substrates.[11,12] Also, as we know, cryoablation has been suggested as a safe and effective alternative to RF ablation in cases where the target sites are located close to perinodal structures, such as the slow pathway in AVNRT and septal accessory pathways. Cryoablation with an 8-mm-tip ablation catheter has been shown to be a low-risk and effective option to treat AVNRT in pediatric patients.[12] In our case with aAVNRT, we performed successful ablation with an 8-mm cryocatheter and we did not observe recurrence in 2 years of follow-up.

Conclusion

Our results indicate that despite being rare, fast-slow aAVNRT must be a part of the differential diagnosis of incessant long RP tachycardias leading to HF. Electroanatomic 3D-mapping helps reduce the fluoroscopy exposure when localizing the tachycardia.

Peer-review: Externally peer-reviewed.

Conflict-of-interest: None.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Authorship contributions: Concept: Y.E., A.G.; Design: Y.E.; Supervision: Y.E., A.G.; Materials: Y.E., O.E., A.İ.; Data collection: Y.E., O.E., A.İ.; Literature search: Y.E.; Writing: Y.E.

REFERENCES

1. Katritsis DG, Josephson ME. Classification, Electrophysiological Features and Therapy of Atrioventricular Nodal Reentrant Tachycardia. Arrhythm Electrophysiol Rev 2016;5:130–5. [CrossRef]
2. Hwang C, Martin DJ, Goodman JS, Gang ES, Mandel WJ, Swerdlow CD, Peter CT, et al. Atypical atrioventricular node reciprocating tachycardia masquerading as tachycardia using a left-sided accessory pathway. J Am Coll Cardiol 1997;30:218–25. [CrossRef]
3. Selvaraj R, Ananthakrishnapillai A, Sadasivam R, Balachander J. "Pseudo PJRT"–fast-slow AV nodal reentrant tachycardia presenting with tachycardia-induced cardiomyopathy. Pacing Clin Electrophysiol 2013;36:e4–6. [CrossRef]
4. Furushima H, Chinushi M, Sugiura H, Aizawa Y. Radiofrequency catheter ablation for incessant atrioventricular nodal reentrant tachycardia normalized H-V block associated with tachycardia-induced cardiomyopathy. J Electrocardiol 2004;37:315–19. [CrossRef]
5. Selvaraj R, Arunprasath P, Karthikeyan B, George G, Balachander J. Uncommon presentation of a common tachycardia. Indian Pacing Electrophysiol J 2010;10:426–8.
6. Jastrzębski M, Pitak M, Rudziński A, Czarnecka D. Pseudo-PJRT in an 8 year-old boy with severe decompensated heart failure and recurrent episodes of ventricular fibrillation. [Article in Polish] Kardiol Pol 2013;71:310–4. [CrossRef]
7. Kaneko Y, Nakajima T, Irie T, Suzuki F, Ota M, Iijima T, Tamura M, et al. Successful ablation of atypical atrioventricular nodal reentrant tachycardia from a noncoronary sinus of Valsalva. Int Heart J 2014;55:84–6. [CrossRef]
8. Roberts-Thomson KC, Kistler PM, Kalman JM. Focal atrial tachycardia II: management. Pacing Clin Electrophysiol 2006;29:769–8. [CrossRef]
9. Ho RT, Frisch DR, Pavri BB, Levi SA, Greenspon AJ. Electrophysiological features differentiating the atypical atrioventricular node-dependent long RP supraventricular tachycardias. Circ Arrhythm Electrophysiol 2013;6:597–605. [CrossRef]
10. Michaud GF, Tada H, Chough S, Baker R, Wasmer K, Sticherling C, et al. Differentiation of atypical atrioventricular node re-entrant tachycardia from orthodromic reciprocating tachycardia using a septal accessory pathway by the response to ventricular pacing. J Am Coll Cardiol 2001;38:1163–7. [CrossRef]

11. Tuzcu V. A nonfluoroscopic approach for electrophysiology and catheter ablation procedures using a three-dimensional navigation system. Pacing Clin Electrophysiol 2007;30:519–25. [CrossRef]
12. Silver ES, Silva JN, Ceresnak SR, Chiesa NA, Rhee EK, Dubin AM, et al. Cryoablation with an 8-mm tip catheter for pediatric atrioventricular nodal reentrant tachycardia is safe and efficacious with a low incidence of recurrence. Pacing Clin Electrophysiol 2010; 33:681–6. [CrossRef]

Keywords: Atypical atrioventricular nodal reentrant tachycardia; child; cryoablation; heart failure.

Anahtar sözcükler: Atipik atrioventriküler nodal reentran taşikardi; çocuk; kriyoablasyon; kalp yetersizliği.