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CLINICAL REVIEW

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Permanent junctional reciprocating tachycardia in infants and Children

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Abstract

Permanent junctional reciprocating tachycardia (PJRT) is a rare form of supraventricular tachycardia (SVT). It generally presents in infants but can be difficult to diagnose. The characteristic EKG findings, response to Adenosine and persistence or frequent recurrences are helpful in making the diagnosis. It is usually difficult to manage with the initial and single medications used in SVT. Many patients are misdiagnosed and not treated effectively and end up having end stage cardiomyopathy and are diagnosed in patients referred for transplant. Hence all patients referred for a cardiac transplant with dilated cardiomyopathy need to be evaluated for this arrhythmia. If appropriate treatment is started early in the course, the failure can be ameliorated, and the cardiomyopathy can be resolved.

KEYWORDS

ablation, permanent junctional reciprocating tachycardia, supraventricular tachycardia, tachycardia induced cardiomyopathy

1 | INTRODUCTION

Supraventricular tachycardia (SVT) is an abnormally fast heart rhythm in which the initiation or persistence of the arrhythmia requires electrical activity originating above the ventricles.[1] SVT is the most common form of cardiac tachyarrhythmia in infants and children.[1–3] The reported incidence varies widely from 1 in 250 to 1 in 25 000.[1–3] If the origin of the aberrant focus is in the sinoatrial region, it is called sinoatrial nodal re-entrant tachycardia.[1] If the arrhythmia originates in the atria, it could manifest as ectopic unifocal atrial tachycardia, multifocal atrial tachycardia or atrial flutter. If the substrate involves the atrioventricular (AV) region, it manifests as AV nodal reentrant tachycardia (AVNRT), AV reciprocating tachycardia (AVRT) or as junctional ectopic tachycardia (JET). In children, AVRT, is the most common form of SVT (Figure 1). While all of these tachyarrhythmias originate above the ventricle, the term SVT, by convention, usually refers to AVRT or AVNRT.[4]

Permanent junctional reciprocating tachycardia (PJRT) is a subtype of AVRT. It is also known as persistent junctional reciprocating tachycardia. PJRT is a rare form of refractory and persistent SVT occurring predominantly in infants and children, accounting for 1% of SVT in this age group. PJRT is caused by AV re-entry using the AV node as the antegrade limb and a slowly conducting accessory pathway (AP) as the retrograde limb.[2,5–8] The diagnosis is rarely made in the neonatal period and the electrocardiogram (EKG) during sinus rhythm is normal, without any manifestation of pre-excitation. As the name implies, PJRT is incessant and if untreated can lead to cardiac decompensation and development of tachycardia induced cardiomyopathy (TIC).[2,8–10] TIC may resolve if early diagnosis and appropriate management is instituted.[10] Hence, prompt diagnosis is imperative.

Most forms of SVT including AVRT, AVNRT, and PJRT are reentrant rhythms.[11] Reentrant tachycardia requires, in addition to the AV node, one additional pathway for conduction, with unidirectional

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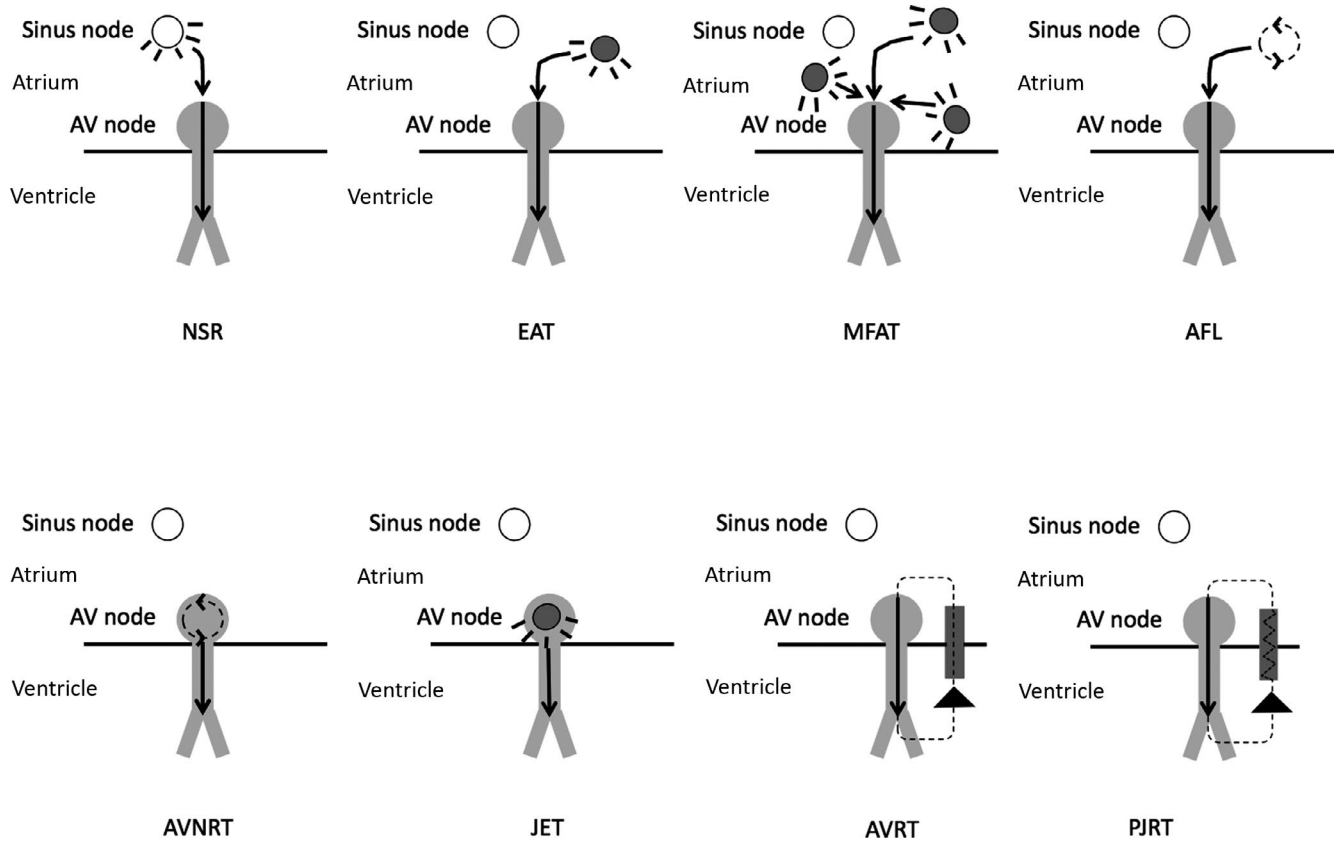

FIGURE 1 Various mechanisms of SVT. Diagrammatic representation of various mechanisms of supraventricular tachycardia. Each figure depicts a schematic of electrical impulse conduction. AFl, atrial flutter; AVNRT, AV nodal reentrant tachycardia; AVRT, AV reciprocating tachycardia; EAT, ectopic atrial tachycardia; MFAT, multifocal atrial tachycardia; NSR, normal sinus rhythm; PJRT, permanent junctional reciprocating tachycardia.

block in one of the two pathways. In AVRT, this additional route is usually because of an AP outside the AV node connecting the atrium and ventricle, allowing direct electrical communication between the two chambers. APs are abnormal tracts made of myocardial fibers that span the AV groove and enable conduction to bypass the AV node. APs can conduct forward from atrium to ventricle (antegrade), or backward from ventricle to atrium (retrograde), or both ways.

When SVT develops, a reentrant circuit for SVT is formed by allowing the electrical impulse to cycle repetitively in one direction, with rapid and regular ventricular activation. Most APs are inserted along the free wall of the mitral valve. About 25% of APs are septal pathways and they insert along the septal aspect of the tricuspid or mitral valve.[12] About 10%-15% are right free-wall pathways. In infants and young children, right sided APs are described more often because of congenital anomalies such Ebstein's anomaly of the tricuspid valve. Other extremely rare APs include atriofascicular (right atrium to the distal right bundle), nodoventricular (AV node to the right ventricular myocardium), nodofascicular (AV node to the specialized conduction system), and atrionodal (right atrial myocardium to the AV node) pathways.[12] In the normal setting, during sinus rhythm, AV conduction occurs exclusively through the AV node. The AV nodal conduction is marked by a delay of several milliseconds before the impulse is transmitted to the ventricle. Additionally, the AV node possesses a characteristic known as decremental conduction, in which the conduction time actually prolongs as the impulse rate increases. In the presence of an AP that is able to conduct in the antegrade direction, the impulse arrives at the ventricle, without the delay imposed by the AV node. This causes ventricular pre-excitation with a characteristic EKG during sinus rhythm consisting of a short PR interval, a delta wave and a widened QRS complex, which is called the Wolff-Parkinson-White (WPW) pattern or ventricular preexcitation. APs are also known to exist where impulses are conducted only in the retrograde direction. These are known as concealed pathways, and the EKG during sinus rhythm appears normal.

PJRT is a subset of this type of concealed pathway causing AVRT. The AV node acts as the antegrade limb and a unique concealed AP acts as the retrograde limb. The retrograde conduction in the AP is characteristically slow and decremental, compared to the conduction velocity of normal myocardium, and is often similar to the normally slow antegrade conduction through the AV node. This similarity in conduction characteristics to the AV node thereby creates a stable reentrant circuit and results in narrow complex tachycardia. It is also the reason for the persistence and refractoriness of SVT, which can lead to left ventricular dilatation and TIC.[2,6,9,12–16] The heart rates in PJRT can range from 200 to 300 in infancy and later on decreases from 250 in early childhood to 120 in adults.[7–9,15]

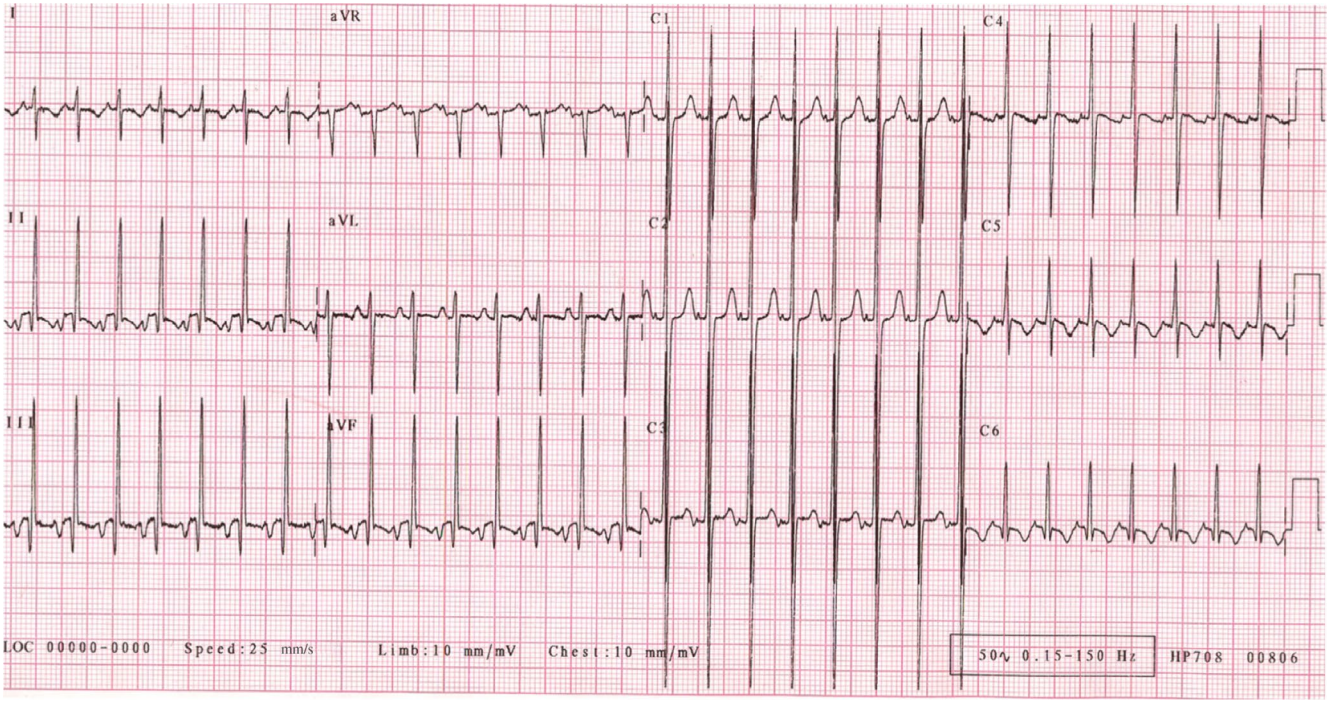

FIGURE 2 12-lead EKG recording of PJRT. Long RP interval and negative P waves in II, III, & aVF. (With permission from Europace, Oxford University Press from Shebani SO, Ng GA, Stafford P, Duke C. Radiofrequency ablation on veno-arterial extracorporeal life support in treatment of very sick infants with incessant tachymyopathy. Europace. 2015(4):622-7)

1.1 | Diagnosis

It is important in the management, to distinguish between the different types of narrow complex reentrant tachycardia, especially AVRT from PJRT. The appearance of the EKG, with special reference to QRS duration, P wave morphology and response to vagal maneuvers or adenosine may help in their distinction. In the differential diagnosis of tachyarrhythmias in infants, the initial steps are to identify if it is a regular or irregular rhythm and then if the QRS complex is broad or narrow complex. The next steps are to identify if P waves are visible and if the P wave and QRS rates are similar. If there are temporal differences in the P and QRS relationship, there is AV dissociation. The next step would be to determine the ratio of RP to PR interval. The EKG criteria for the diagnosis of PJRT include (Figure 2),

1. RP interval is longer than PR interval (because of the location and decremental conduction properties of the AP)
2. 1:1 AV ratio (no dissociation)
3. Inverted P wave are often visible in the inferior leads, II, III, and AVF
4. Does not require a critically timed extrasystole for initiation
5. Not associated with frequent PACs
6. PR interval is never prolonged
7. The rates are typically slightly slower than typical SVT (can be confused with sinus tachycardia)
8. Response to vagal maneuvers with gradual slowing of the tachycardia because of prolongation of both RP and PR intervals and eventual termination but with recurrence shortly afterwards
9. The AV conduction is usually sensitive to adenosine with tachycardia terminating with AV or VA block but again recurring shortly afterwards
10. QRS is normal and narrow during sinus rhythm and in tachycardia.[3,8,9,12,15]

When evaluating the EKG of narrow complex tachycardia with a short RP interval (QRS complex to P wave activation), the diagnosis is narrowed to the typical form of AVNRT, AVRT using accessory pathways, atrial tachycardia with long first-degree AV block, atrial tachycardia (AT) originating from the os of the coronary sinus or junctional tachycardia. In SVT with long RP interval, the possible diagnoses include atypical AVNRT, PJRT, atrial tachycardia, sinus tachycardia, sinus node reentry tachycardia and atrial flutter with 1:1 conduction. In AT, the RP/PR ratio is higher, the inverted P wave in inferior leads is uncommon, the P wave duration is longer and there is persistence of the tachycardia with P waves marching through during AV block.[17,18] Definitive diagnosis of the arrhythmia substrate requires intracardiac electrophysiologic (EP) study.

1.2 | Cardiac electrophysiology

Intra-cardiac EP studies allow for detailed analysis of the mechanism of the abnormal rhythm especially, precise localization of the site of origin of the AP. After venous vascular access is obtained, electrode catheters are typically placed in the high right atrium (HRA), anterior tricuspid valve (bundle of His), the right ventricle apex (RVA) and coronary sinus (CS). Left-sided electrodes are needed occasionally for left

sided APs or left sided SVTs. Generally the EP study involves the simultaneous recording of surface and intra-cardiac electrograms. After baseline recordings, burst pacing and programmed electrical stimulation (PES) are performed to assess the AV conduction and to induce an arrhythmia. Cardiac mapping during EP testing identifies the spread of activation from its initiation to its completion of electrical potentials generated by the myocardium during normal and abnormal rhythms.

The EP criteria for diagnosis include transient interruption of the PJRT by programmed stimulation with prompt resumption after few sinus beats, delay in atrial electrogram with an identical atrial activation sequence during PJRT via ventricular extrastimulus when the His bundle is refractory; atrial activation sequence during ventricular pacing that is identical to the pattern during tachycardia; and no anterograde conduction through the accessory pathway, but slow and decremental retrograde conduction through the accessory pathway.[17,18] The post pacing interval (PPI) to tachycardia cycle length (TCL) difference of <110 ms and the stimulus to atrial (SA) to venticuloatrial (VA) difference of <85 ms excludes an AVNRT and suggests a septal accessory pathway. The tachycardia response to the His bundle-refractory ventricular extra stimuli can be either advanced or delayed atrial activation. Another point would be that His bundle-refractory ventricular extra stimuli could delay the next atrial activation while maintaining the same atrial activation sequence. This can confirm the existence of an accessory pathway and its involvement in the tachycardia mechanism. Also, an increase in the SA interval with the progressive reduction in coupling intervals of the ventricular extra stimuli occurs, which can demonstrate the decremental behavior of the accessory pathway.

The points in the EP diagnosis includes

1. obligatory 1:1 AV relationship
2. VA/TCL prolongation with the development of ipsilateral bundle branch block (BBB)
3. His-refractory ventricular premature depolarizations (VPDs) reset the atrium or terminate tachycardia with VA block
4. ability to entrain tachycardia from the ventricle with orthodromic capture of the His bundle
5. PPI–TCL<110 ms
6. Ventriculo-atrial (VA) entrainment from ventricle—$VA_{(SVT)}$ = <85 ms
7. His-atrial (HA) entrainment from ventricle—$HA_{(SVT)}$ = <0 ms
8. Atrio-His (AH) atrial pacing/entrainment at/near TCL—$AH_{(SVT)}$ = <20 ms.[12,19]

1.3 | Treatment

1.3.1 | Medical management

PJRT can be difficult to treat and refractory to medical management. The similarity of conduction between the AV node and the AP are such that both antegrade and retrograde limbs are similarly affected by antiarrhythmic agents, making it difficult to achieve sustained block in just one limb of the reentrant circuit. Many infants and children require treatment with more than a single antiarrhythmic agent.[8] Earlier studies reported amiodarone, digoxin, and verapamil in combinations as the most effective regimen with reasonable success.[1] The latter two medications are now rarely used and currently, combination therapy with flecainide and amiodarone has been shown to be slightly more successful in controlling tachycardia and reversing cardiomyopathy, if detected and treated early.[20]

1.3.2 | Interventional management

Ablation is generally indicated when there is difficulty in medical management and some feel that it needs to be attempted early in PJRT after appropriate electrophysiologic mapping. In extremely low birth weight infants there are technical difficulties in obtaining a detailed mapping. Most commonly, the AP is located in the right posterior septum (inferoseptal) near the ostium of the CS or within the proximal CS but locations have been described in almost any position along the AV groove.[21] Multiple pathways have been also reported.[7,22–26] Experience with catheter ablation has accumulated over the years and there are crucial differences in electrophysiological and mapping techniques between PJRT pathways and non-decremental APs. First, advancement of the atrial wavefront over the pathway by His-refractory ventricular pacing can be difficult because of decremental conduction in the pathway by premature stimulation. Second, pathway potentials may be demonstrated in up to 75% of cases but VA intervals are characteristically long and a prominent isoelectric signal between ventricular and atrial signals are not always present. Finally, mapping and ablation of these APs must be performed during tachycardia because it may not be possible to achieve exclusive retrograde pathway conduction while pacing ventricularly, because of retrograde AV node conduction or VA block at cycle lengths longer than the TCL.[21] Radiofrequency ablation has been used more often, but cryoablation is indicated for the rare PJRT pathway located in the mid or anterior region of septum (in proximity of the AV node), as it reduces the risk of AV block.[27,28] Both forms of ablation energy have shortcomings but success rates of more than 80% have been reported.[8,9,25,28–30] Recurrence rates following PJRT ablation are often higher, necessitating more than one procedure for permanent success.

2 | CONCLUSION

PJRT is one of the causes of refractory SVT in infants and children. Even though it can be difficult to manage, it is a potentially treatable arrhythmia once diagnosed. In addition, it is critical to rule out PJRT in any child presenting with dilated cardiomyopathy, intractable heart failure, or for a transplant evaluation, as effective treatment could potentially reverse it.

CONFLICT OF INTEREST

The authors declare no conflict of interests for this article.

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