

## Congenital Junctional Ectopic Tachycardia: There May Be a Light at the End of the Tunnel

*Taquicardia ectópica congénita de la unión: puede haber una luz al final del túnel*

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Congenital junctional ectopic tachycardia (JET) is one of the most difficult arrhythmias faced within the pediatric population. It is difficult to control and frequently results in hemodynamic instability or a tachycardia induced cardiomyopathy. This arrhythmia can result in both morbidity and mortality. Fortunately, this tachycardia is relatively rare. With the arrhythmia being rare, the long term prognosis of the tachycardia has not been well described in the literature.

This paper by Maldonado and colleagues(1) describes spontaneous remission of JET following initially successful therapy. Successful spontaneous resolution within 3-8 years has been previously reported in studies from the 1990's.(2) However, the overall long-term prognosis of congenital JET that is able to be initially controlled with medications has not been well characterized. The resolution of an ectopic rhythm like JET makes sense considering the spontaneous resolution of other automatic focus tachycardias like atrial ectopic tachycardia and ventricular tachycardia when these tachycardias present in the newborn period.

Ablation is one therapeutic strategy that has been used to treat congenital JET. The success rate for ablation is around 80% but the ablation carries about a 15% chance of permanent AV block requiring a pacemaker, and this risk may even be higher in very young patients. (3) The possibility of medical therapy potentially serving as a bridge to ultimate recovery is a very appealing option if the spontaneous resolution rate of the tachycardia is ultimately high.

This paper by Maldonado and colleagues provides important insights into JET, and indicates that it may take longer for JET to eventually resolve in patients, and that extended medical therapy may be warranted for at least 2-3 years (if not longer, even waiting until patients are 8-10 years old) before attempting an ablation that carries a real risk of permanent AV block requiring a pacemaker. This paper

provides insight into the long-term prognosis of this rare arrhythmia and gives both families and pediatric cardiologists hope for a good long-term outcome if the arrhythmia is able to be controlled initially.

Caution must be exercised as JET can cause mortality and conservative treatment without antiarrhythmic therapy should be reserved for slower, hemodynamically stable JET in patients with normal ventricular function. Aggressive medical therapy should be initiated for patients who present with rapid JET as their clinical status may deteriorate quickly. Patients who cannot be controlled medically should be considered for ablation at any age, as there is a real probability of death in patients with uncontrolled tachycardia. Ablation may be considered as a first line of therapy in larger patients (>15 kg) in whom the risks of ablation are likely lower. As this paper nicely describes, medical therapy may take days to weeks to be effective and this delay in arrhythmia control may allow time for hemodynamic deterioration of the arrhythmia before medical control can be achieved. However, in young patients, particularly less than 1 year of age, aggressive medical therapy with rapid escalation of medication doses and addition of multiple antiarrhythmic agents should be strongly considered before ablation because of the real risk of permanent AV block.

Caution must also be applied when determining if a patient has truly had complete resolution of their tachycardia, as we reported the case of a young boy who had recurrence of his JET following urological surgery. (4) This child continued to have recurrences with catecholamine surges and ultimately required ablation due to symptomatic tachycardia although he remained in sinus rhythm the majority of the time when his catecholamine state was low. He underwent a successful ablation and has had no further recurrences of tachycardia. This case highlights the importance of ablation for JET in patients in whom the tachycardia does not resolve spontaneously. It also highlights the

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long-term need for careful surveillance in patients in whom the JET has seemingly “resolved”.

There is much still to be learned about JET and its long term outcomes and if this arrhythmia predicts any adverse consequences when patients reach adulthood. However, Maldonado and colleagues should be congratulated on this paper that aids in the understanding of this rare arrhythmia providing insight into the long-term management of JET and is an important contribution to the pediatric electrophysiology literature.

#### **Conflicts of interest**

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

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