



Revista Argentina de Cardiología

ISSN: 0034-7000

ISSN: 1850-3748

revista@sac.org.ar

Sociedad Argentina de Cardiología
Argentina

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Revista Argentina de Cardiología, vol. 86, no. 4, 2018, July-, pp. 249-250
Sociedad Argentina de Cardiología
Argentina

DOI: <https://doi.org/10.7775/rac.es.v86.i4.11821>

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Long-term Spontaneous Remission of Congenital Junctional Ectopic Tachycardia

Remisión espontánea a largo plazo de la forma congénita de la taquicardia ectópica de la unión

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ABSTRACT

Background: The congenital form of junctional ectopic tachycardia (JET) is a rare supraventricular tachycardia with high morbidity and mortality. Its treatment frequently requires the prolonged use of multiple drugs and, in some cases, an invasive procedure, such as cryoablation, is necessary. Some studies have demonstrated the existence of spontaneous remission of arrhythmia in the long term (with reversal to sinus rhythm or to an accelerated nodal rhythm), without need of drugs or any other form of intervention.

Objectives: The aim of this study was to evaluate the long-term incidence of spontaneous remission of congenital JET in a pediatric population.

Methods: Twenty-three consecutive patients with congenital JET were evaluated at Hospital J.P. Garrahan and Hospital Italiano de Buenos Aires, between 1999 and 2017, with a mean follow-up of 8.8 years [interquartile range (IQR): 5.5-14.1]. The incidence of spontaneous arrhythmia remission was evaluated in the long term.

Results: Median presentation age was 2 months (IQR: 0.625-3); 15 patients (62.5%) presented signs of heart failure and 11 patients (45.8%) tachycardiomyopathy. Two patients underwent successful cryoablation. The spontaneous remission rate was 52%. In patients with follow-up above 10 years (15 subjects), remission was 62.5%. Mean remission age was 9.6 years.

Conclusions: Congenital JET is a potentially severe tachycardia in the first months of life, but with a high rate of long-term spontaneous remission.

Key Words: Junctional ectopic tachycardia, congenital JET, JET remission

RESUMEN

Introducción: La forma congénita de la taquicardia ectópica de la unión (junctional ectopic tachycardia, JET) es una taquicardia supraventricular poco común, con alta morbimortalidad. Su tratamiento requiere, frecuentemente, el uso de múltiples fármacos durante períodos prolongados. En algunos casos se necesita, además, realizar un procedimiento invasivo, como la crioblación. Algunos estudios han demostrado la existencia de casos de remisión espontánea de la arritmia a largo plazo (con la reversión a ritmo sinusal o a un ritmo nodal acelerado), sin requerimiento de fármacos y sin mediar intervención alguna.

Objetivos: Evaluar la incidencia de remisión espontánea de la JET congénita en una población pediátrica en el seguimiento a largo plazo.

Material y métodos: Se incluyeron 23 pacientes consecutivos con JET congénita evaluados en el Hospital J.P. Garrahan y en el Hospital Italiano de Buenos Aires, entre los años 1999 y 2017, con un seguimiento medio de 8,8 años (rango intercuartil, RIC: 5,5-14,1). Se evaluó a largo plazo la incidencia de remisión espontánea de la arritmia.

Resultados: La mediana para la edad de presentación fue 2 meses (RIC: 0,625-3); 15 pacientes (62,5%) presentaron signos de insuficiencia cardíaca y 11 pacientes (45,8%), taquicardiomiopatía. A 2 pacientes se les realizó una crioblación en forma exitosa. La tasa de remisión espontánea fue del 52%. En aquellos pacientes con un seguimiento mayor a 10 años (15 sujetos), la remisión fue del 62,5%. La media para edad de remisión fue 9,6 años.

Conclusiones: La JET congénita es una taquicardia potencialmente grave en los primeros meses de vida, pero con alta tasa de remisión espontánea a largo plazo.

Palabras clave: taquicardia ectópica de la unión, JET congénita, remisión JET

REV ARGENT CARDIOL 2018;86:239-243. <http://dx.doi.org/10.7775/rac.v86.i4.11821>

Received: 03/12/2018 – Accepted: 06/27/2018

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INTRODUCTION

The congenital or primary form of junctional ectopic tachycardia (JET) is a generally incessant and, at times, paroxysmal arrhythmia, with high morbidity and mortality, described by Coumel et al. in 1976. (1, 2) The mechanism of tachycardia is due to an abnormal automatism originating in the region of the atrioventricular (AV) junction and in rare cases to triggered activity. (3, 4)

Clinically, JET usually manifests as heart failure with compromised myocardial function secondary to incessant behavior, elevated heart rate, and the AV dissociation that characterizes this peculiar arrhythmia.

Treatment strategies for patients with congenital JET include pharmacological treatment, often combining different antiarrhythmic agents and cryoablation. Some studies document the existence of patients whose arrhythmia was self-limiting in the long-term follow-up, without need of pharmacological or invasive treatment. (5-7)

In this study we evaluated the population with congenital JET diagnosis treated in two high complexity referral centers, and reported the behavior, clinical characteristics, treatment scheme and remission rate in the long-term follow-up of this infrequent and severe pediatric arrhythmia.

METHODS

A retrospective and observational cohort study was performed including all consecutive patients diagnosed with congenital JET controlled in the arrhythmia clinic of Hospital de Pediatría Prof. Dr. Juan P Garrahan and in Hospital Italiano de Buenos Aires between January 1999 and May 2017.

Mean follow-up was 8.8 years (IQR: 5.5-14.1). Junctional ectopic tachycardia was diagnosed if the following criteria were met in an electrocardiographic recording: narrow QRS complex tachycardia, usually with AV dissociation, occasion-

al sinus capture beats and less frequently with 1:1 or variable AV conduction, and heart rate above the 95th percentile for the age group. (Figure 1)

All patients were evaluated with a complete clinical examination, electrocardiogram (ECG), color Doppler echocardiography, and 24-hour Holter monitoring. The reversal to sinus rhythm or to JET heart rate control, with or without intermittent periods of sinus rhythm, and with recovery of myocardial function, was considered as response to medical treatment.

Once treatment response was ensured, the slow and progressive decrease of the antiarrhythmic medications was initiated with control Holter monitoring for each change of treatment dose, and spontaneous remission was considered in the absence of JET in two consecutive 6-month interval 24-hour Holter evaluations, or in the presence of accelerated nodal rhythm with heart rates below the 95th percentile of the corresponding age group in the absence of symptoms, without need of pharmacological or invasive treatment.

Statistical analysis

Data was stored in Microsoft Office Excel 2010 and Stata 12.0 statistical software package was used to perform the analysis.

The distribution of frequencies or percentages in relation to the total number of cases was established for all variables, and the values were accordingly expressed as proportions, mean and standard deviation or median and interquartile range (IQR).

Continuous variables were compared with Student's t test or the Mann-Whitney test, as appropriate, and Fisher's exact test or the chi-square test were used to analyze proportions, according to the sample size. A significant difference was considered for $p < 0.05$.

Ethical considerations:

The retrospective nature of the study complied with the ethical committee regulations of the participating institutions, with care of patients' personal data integrity.

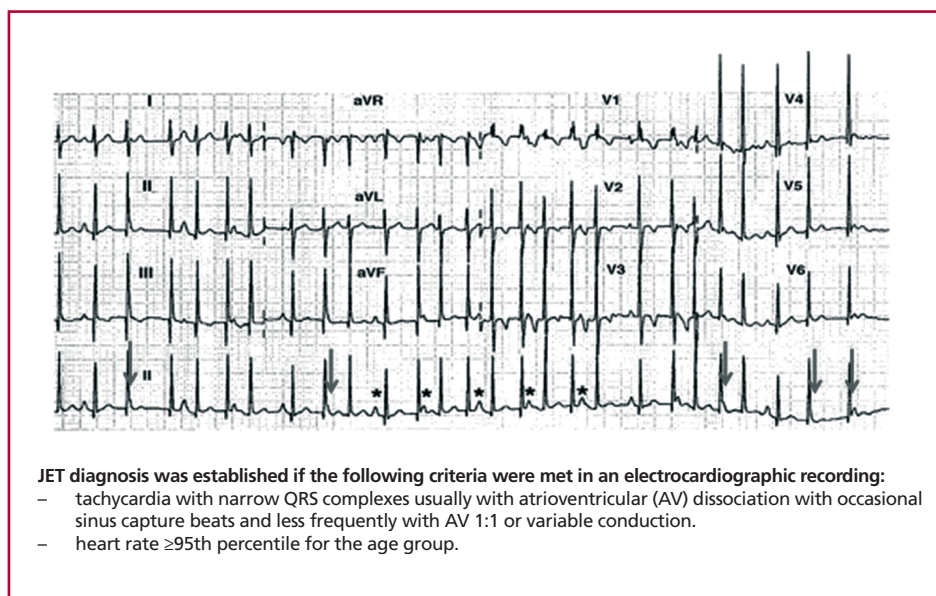


Fig. 1. Diagnostic criteria of junctional ectopic tachycardia (JET)

RESULTS

Clinical characteristics

Twenty-three patients were evaluated, 15 (62.5%) male, with median age of 2 months (IQR: 0.625-3). Junctional ectopic tachycardia was incessant in 17 patients (70.8%), with a maximum heart rate detected in Holter monitoring of 192 beats per minute (IQR: 170-233). Eighteen patients (75%) required hospitalization; 15 (62.5%) were admitted with heart failure, and 11 (45.8%) had compromised myocardial function, with shortening fraction <30% (IQR: 23-30). Antiarrhythmic treatment could control JET in an average time of 20 days (IQR: 11-60), with Holter monitoring reaching an average heart rate of 119 beats per minute (IQR: 107-135). Two drugs (IQR: 1-2) were used for the chronic pharmacological management of the arrhythmia; in all cases, amiodarone was used at an intravenous loading dose of 10 mg/kg/day for 48-72 h, together with oral amiodarone, at maintenance dose between 5 and 20 mg/kg/day, and then only orally. Other drugs, such as digoxin, were associated at 5-10 g/kg/day orally, to control signs of heart failure or in the presence of associated ventricular dysfunction; atenolol at a dose of 1-2 mg/kg/day orally; and flecainide, 3-5 mg/kg/day orally. Three patients presented adverse effects due to amiodarone: one had hypothyroidism, another corneal dystrophy and a third presented both effects. Two patients underwent successful cryoablation of the focus after 10 and 12 years of pharmacological treatment, due to the adverse effects caused by the medication and lack of spontaneous remission, with persistence of symptomatic JET. None of the cases presented with complications during the procedure.

Twelve patients (52%) had spontaneous remission during follow-up. The average age of spontaneous re-

mission was 9.31 ± 5.1 years. At the 4-year follow-up, the remission rate was 15% and at 10 years, 62.5% (Figure 2). The rest of the patients continue with pharmacological treatment.

DISCUSSION

Congenital JET is an infrequent supraventricular arrhythmia that appears at an early age with high morbidity and mortality. There are very few studies in which the spontaneous remission of JET has been evaluated over time, and none performed in Latin America. In our study we have demonstrated a high resolution rate to either sinus rhythm or to accelerated nodal arrhythmia (52%), at an average age of 9.31 ± 5.1 years.

In a 2009 publication, Collins et al. reported a spontaneous resolution of the arrhythmia in 33.1% of the patients at a mean age of 3 years (range: 0.3-20 years). In the study by Villain et al., (5) 23% of the patients evolved with resolution or control of the arrhythmia at a mean age of 3.6 years (range: 0.5-8 years), whereas in the registry of Sarubbi et al., (6) including 9 patients with a follow-up of 2.6 to 21 years, the pharmacological treatment could not be discontinued in any case due to JET recurrence.

The differences observed with the studies of Collins et al. and Villain et al. can be explained, on the one hand, by the difference of follow-up period, since, as demonstrated in our work, the remission rate was greater with the passage of time; on the other hand, since the form of discontinuation was not homogeneously standardized, probably many of the patients could have received pharmacological treatment for an unnecessary period of time, since the discontinuation of the drug was not attempted earlier to evaluate the resolution of the arrhythmia.

Fig. 2. Spontaneous remission (Kaplan Meier curve)

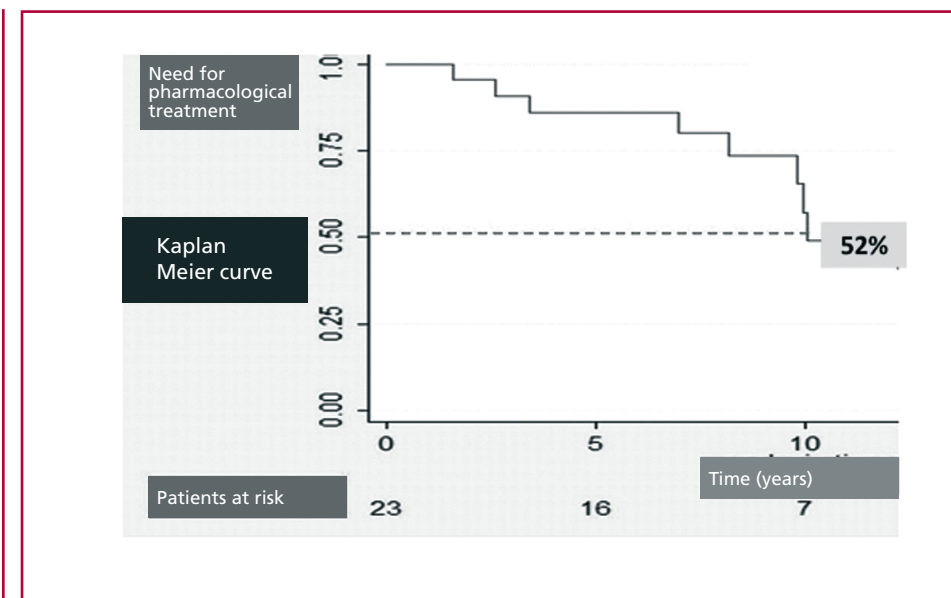


Table 1 shows the most relevant published studies on congenital JET. There are two therapeutic strategies for this condition: pharmacological and cryoablation. During the first years of life, the first one is chosen, since ablation in patients of less than 15 kg is associated with a higher risk of complications. (8) Among the drugs, the most effective is amiodarone, but its use for prolonged periods is associated with the development of adverse effects, such as thyroid or corneal dystrophies, and, less frequently, liver and lung dystrophies. (9) Other alternative drugs may be propafenone or flecainide associated with a beta-blocker, and recently, a study has reported the use of ivabradine as adjuvant, mainly of amiodarone, with very good results. (10, 11) The cryoablation strategy is generally reserved for those patients with greater weight, in whom the pharmacological treatment could not be discontinued, or due to the development of adverse effects. The arrhythmogenic substrate is located very close to the compact AV node and the bundle of His, and therefore the risk of AV block is high. But with the advent of cryoablation, it has been possible to attempt its elimination with very low risk of complications. (12, 13)

With the results obtained in our study, where the rate of spontaneous resolution was high, there is need to reassess a new approach for the follow-up of patients with congenital JET. According to the findings of our and published studies, the strategy in our institutions is as follows: in the initial stage when the diagnosis is established and incessant and very rapid tachycardias are usually present with risk of tachycardiomyopathy, cardiogenic shock and death, the most effective pharmacological treatment, which is amiodarone, should be initiated, in many cases requiring the combination with other drugs. The aim of the treatment is the reversal to sinus rhythm, which is achieved in few cases, or the control of heart rate, with intermittent periods of sinus rhythm. Once the goal is achieved, the scheme is maintained between 2 and 3 years, with weight-adjusted dosing. The selected time is based on the results

obtained in the works of Villain et al. and Collins et al., in which the mean age of spontaneous remission was in that range. After obtaining two 6-month interval Holter monitorings in sinus rhythm or heart rate control, the dose of amiodarone is not modified and the associated drugs are decreased. This is followed by the progressive decrease and discontinuation of amiodarone if no JET relapse is observed. If the arrhythmia reappears when the drugs' dosage descends, the therapeutic dose is optimized up to 8-9 years, with strict control of adverse effects. In case of presenting any of them and with weight under 15 kg, a new pharmacological scheme is tried until the appropriate conditions to perform the cryoablation with the lowest possible risk are achieved. If JET is not controlled with the new scheme or the patient presents an adequate weight, cryoablation is performed. The choice of that age arises from our work, in which patients with follow-up above 9 years had up to 62% spontaneous JET remission. In the period between 3 and 9 years, the reduction of drugs should always be evaluated sporadically to determine if the arrhythmia has self-limited. (Figure 3)

Limitations

This is an observational and retrospective study, but it includes a significant number of cases and a prolonged follow-up time for an infrequent pediatric arrhythmia.

CONCLUSIONS

Junctional ectopic tachycardia is a potentially severe tachycardia in the first months of life, but with a high rate of spontaneous remission in the long term; therefore, the indication for ablation should not be premature, except with refractoriness or adverse effects to the treatment.

Conflicts of interest

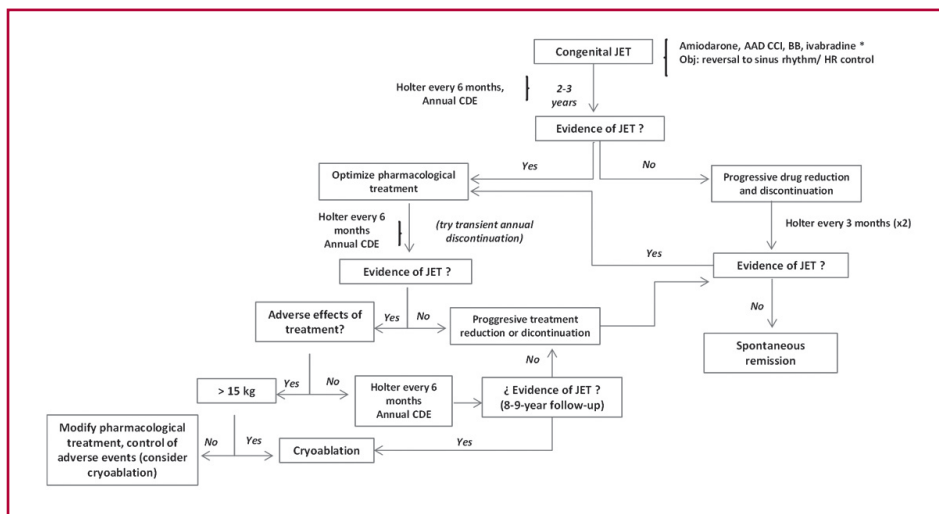
None declared. (See authors' conflicts of interest forms on the website/Supplementary material).

Table 1. Published registries of patients with congenital junctional ectopic tachycardia.

Author (year)	N° pts.	Age at presentation (months)	RF Abl. (%)	Cryoablation (%)	Pacemaker (%)	Follow-up (years)	Death rate (%)	Age of SR (years)	SR rate (%)
Villain et al. (1990)	26	1	-	-	15	6.1	34	3.6 (range: 0.5-8)	23
Sarubbi et al. (2002)	9	3.8	-	-	0	12.4	0		0
Collins et al. (2009)	92	9.6	18	27.6	14	4.5	4	3 (range: 0.5-8)	33.1
Garrahan Hospital (2016)	31	2	3.2	19.3	3.2	8.8	0	9.31±5.1	51.6

N.° Pts: Number of patients. RF Abl: Radiofrequency ablation. SR: Spontaneous remission.

Fig. 3. Therapeutic algorithm for congenital JET



JET: Junctional ectopic tachycardia. AAD: Antiarrhythmic drugs. CCI: Calcium channel inhibitors. BB: Beta-blockers. HR: Heart rate. CDE: Color Doppler echocardiography. * Mainly as amiodarone coadjutant

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