

## Editorial



# Clinical Implication of Multifocal Atrial Tachycardia in Children for Pediatric Cardiologist

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► See the article “The Complexity of Pediatric Multifocal Atrial Tachycardia and Its Prognostic Factors” in volume 48 on page 148.

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Multifocal atrial tachycardia (MAT), also known as chaotic atrial arrhythmia, is rare and arises from multiple foci of increased automaticity located within the atria. MAT is defined by the presence of 3 or more P wave morphologies, irregular P-P, R-R, and P-R intervals, an isoelectric baseline between the P waves, and ventricular rate over 100 beats/min.<sup>1)</sup>

Although the prevalence of pulmonary disease in MAT has been well established in adult MAT patients, particularly those with pulmonary diseases including chronic obstructive pulmonary disease, it is relatively rare in the pediatric ages. Therefore, the clinical feature of MAT in children is not well known with several studies of small cases.<sup>2)</sup>

Possible mechanism for MAT has been suggested such as right atrial enlargement, hypercapnia, hypoxia, or adrenergic stimulation in pulmonary disease.<sup>3)</sup> The majority of MAT reports in pediatric patients have also reported not so low coexisting pulmonary disease (>20%) compared to the higher rate in adults (up to 60%). Unlike pulmonary physiology in adults, growth and development of bronchopulmonary system and pulmonary vessel continues for at least 2 years. It could be some role for the predominant infantile onset of MAT. Also, many of MAT have been detected in utero indicating that the immaturity and vulnerability of the atrium may also contribute to the infant-predominant age distribution of MAT.<sup>4)</sup> Therefore, immaturity of both the lungs and the heart might play a key role of infant-predominant age distribution and its favorable outcome in idiopathic infant cases.

Bradley et al.<sup>5)</sup> reported the clinical course of MAT in infants and children in 2001. This retrospective study included 21 patients (14 boys, 7 girls [median age 1.8 months]) and comorbidities of 6 respiratory illness and 7 structural heart disease (SHD). Four of 15 patients (27%) had diminished ventricular function. Fifty percent of patient reverted to sinus rhythm within 5 months. There were no late arrhythmias. The extended follow-up of this study confirms the low mortality and likely good outcome and long-term health appears related primarily to underlying conditions. Response to antiarrhythmic agents is limited, although amiodarone shows promise when control of the rhythm is necessary. Direct current cardioversion is ineffective and should be avoided. They conclude that long-term cardiovascular and developmental outcome depends principally on the overall clinical state; for otherwise healthy children, it is excellent.

Baek et al.<sup>6)</sup> reported the clinical outcome of MAT and potential prognostic factors. Compared to the previous reports, this study has relatively large number of patients and composed of various etiologies despite of limitation of retrospective study from single tertiary center. Among 33 patients with identified MAT, 27 (82%) were infantile onset and 10 patients (30%) had fetal diagnosis. Incidental detection without significant clinical manifestation is rather high (27%). Comorbidities had a variety of SHD (42%) and lung disease (24%). Interestingly, syndromic diagnosis, including 3 with Costello syndrome and 2 with Noonan syndrome, and one suggestive of RASopathy were noted in infantile onset group. Among 27 patients with infant onset of MAT, 11 patients (41%) were included in the idiopathic group. Accompanying arrhythmias was revealed in 4 patients (2 atrioventricular reentrant tachycardia prior to MAT diagnosis; 2 catecholaminergic polymorphic ventricular tachycardia [CPVT] after MAT diagnosis). The arrhythmia control rate was higher in the infant group (85%) than in the non-infant group (67%), although this trend was not statistically significant. There was a significantly lower rate of unfavorable outcomes in the idiopathic infant group (n=11) than in the other groups (p=0.008). Considering the findings of previous studies, the mortality rate was significantly higher in patients with SHD than in patients without (21% vs. 5%, p=0.01). The idiopathic infant group had a significantly lower rate of unfavorable outcomes than did the others (0% vs. 47%, p=0.008).

For the pediatric practitioners, 4 issues have arisen from above studies regarding MAT in children as follows: 1) how to detect early, 2) how to control, 3) how deep to investigate etiologies of MAT, and 4) how to predict another arrhythmia and outcome.

Firstly, early detection is very important to prevent worse outcome in infantile onset MAT. Tachycardia is usually first detected during the newborn period and incidental detection not based on clinical suspicion is rather high. Clinical suspicion of infantile onset of MAT is important for early detection. If tachycardia last long over several days without proper management, myocardial dysfunction can develop resulting in congestive heart failure. due to tachycardia-induced cardiomyopathy. So early detection and immediate proper management for tachyarrhythmias is necessary.

Secondly, complete control of MAT is not easily achievable with combination of multiple antiarrhythmic medications, even in high-dose combinations. A more realistic treatment goal is initially reducing the percentage of MAT and achieving ventricular rate control. Various drugs have been used for the purpose, including beta blocker, digoxin, and amiodarone, but there is no data to support the superiority of any one approach.

Thirdly, because of variety of etiology of MAT in children, delineation of etiology should be done to treat underlying problems and get better clinical outcome. Idiopathic infantile onset group shows a favorable outcome compared to the other groups including SHD and syndromic disease. RASopathy has been reported to be associated with high incidence of atrial arrhythmias.<sup>6,7)</sup> MAT in children should be checked the association of RASopathy and vice versa.

Fourthly, further lethal arrhythmias could not be predicted not only by MAT but also by additional studies. Atrial premature beats, atrial fibrillation (AF), or atrial flutter are known to accompany MAT in both adults and pediatric patients.<sup>5,6,8)</sup> MAT may be an early manifestation of CPVT and also additional findings of atrioventricular nodal reentrant tachycardia. Phenotypical progression of MAT into CPVT and an association between

the RyR2 mutation and AF and ectopic atrial tachycardia have reported.<sup>6,9)</sup> MAT in young children may be the initial manifestation of a potentially life-threatening arrhythmia of CPVT. Therefore, non-infantile form of MAT with structurally normal hearts might need aggressive evaluations and close follow-up.

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