

Clinical aspects of pediatric Brugada syndrome

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Introduction: Brugada syndrome (BrS) is an inherited arrhythmia characterized by a coved-type ST elevation and sudden death, especially in middle-aged males and more common in Asia. Mutations in SCN5A are detected in 15–20% and reported to be associated with poor prognosis. Among children, BrS is rare and the risk factors in pediatric BrS are unknown, especially in Asian population.

Purpose: The purpose of this study is to elucidate the risk factors for fatal arrhythmic events in Japanese pediatric patients with BrS.

Methods: We enrolled 52 Japanese children with BrS younger than 20 years, and performed genetic analysis and collected the clinical information.

Results: The mean age of initial symptoms was 10.7±5.5 years, and the mean follow-up period was 3.9±5.5 years. Ninety percent of patients were

probands. No subjective symptom was confirmed in 28 of the patients, but aborted cardiac arrest (ACA) in 4, ventricular tachycardia in 4, ventricular fibrillation in 1, and syncope in 11. We identified mutations in SCN5A in 63%. There was no significant gender difference in ≤10 years, but a significant male predominance appeared in >10 years. And no gender difference was confirmed in the incidence of severe cardiac events in ≤10 years.

Conclusion: No gender difference was confirmed in ≤10 years in this study about Asian children. And being girls did not reduce the risk in ≤10 years. The frequency of SCN5A mutations was higher than adults, but decreased from childhood (68%) to adolescence (59%). In BrS, genetical and environmental factors may be more effective in childhood and adulthood, respectively.

Difference by the age and SCN5A status

	Age			SCN5A mutations		
	≥10y (n=25)	>10y, ≤20y (n=27)	p value	(+) (n=33)	(-) (n=33)	p value
Sex: Male	15 (60)	24 (89)	0.025	23 (70)	16 (84)	NS
Age (y)				10.2±5.8	11.6±4.8	NS
SCN5A mutation	17 (68)	16 (59)	NS			
Spontaneous type1 BrS ECG	23 (92)	18 (67)	0.040	28 (85)	13 (68)	NS
ACA or VT or VF	5 (20)	4 (15)	NS	6 (18)	3 (16)	NS
FH of BrS	8 (32)	11 (41)	NS	16 (48)	3 (16)	0.035
FH of sudden death	4 (16)	9 (33)	NS	6 (18)	7 (37)	NS
SSS	1 (4)	6 (22)	NS	7 (21)	0 (0)	0.039
Syncope	2 (8)	9 (33)	0.040	6 (18)	5 (26)	NS

Values are presented as n (%) or mean ± SD. y = years old; BrS = Brugada syndrome; ACA = aborted cardiac arrest; VT = ventricular tachycardia; VF = ventricular fibrillation; FH = family history; SSS = sick sinus syndrome.