TITLE: CLINICAL CHARACTERISTICS, AND COURSE OF SINUS NODE DYSFUNCTION IN CHILDREN: TWO CENTER EXPERIENCE FROM TURKEY

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Abstract

Aim: To evaluate the clinical characteristics and outcomes of children diagnosed with sinus node dysfunction (SND).

Methods: This was a retrospective review of patients diagnosed with sinus node dysfunction in two tertiary paediatric cardiology center in Turkey from January 2011 to June 2022.

Results: In all, 77 patients (50/77, 64.9% males) were included, with a mean age of 8.2±6.3 years and a mean weight of 28.2±18. 8 kg.The age inappropriate bradycardia and pauses were the most common rhythm disturbance, and syncope and dizziness (n:29, 37.6%) were the most frequent initial symptoms. Fifty eight of the 77 patients (75.3%) had structural heart disease, 47 (61%) of them had congenital heart disease, most commonly transposition of great arteries (n:8), atrial septal defect (n:7) and AV septal defect (n:5). Seven of them had also left atrial isomerism. The remaining 19 patients were isolated. Four patients had SCN5A mutation and two of them were siblings, One patient had DSP and ANK2 mutation, one had AKAP9, one of them had Emery–Dreifuss muscular dystrophy. Of the total patient population, 44 patients (57.1%) had previously undergone a cardiac operation and except one patient sinus node dysfunction developed after a surgical procedure. This patient had left atrial isomerism and SND was diagnosed during 24 hours Holter monitorization before surgery. The most common surgical procedures were closure of atrial septal defect in seven, fontan operation in six, correction of AV septal defect in five, Senning operation in five, and abnormal pulmonary venous connection anomaly repairment in three. Thirty-nine (50.6%) patients (22 of them were after cardiac surgery) were underwent pacemaker implantation. The mean interval between pacemaker implantation and the previous operation was 5,1 years (range, 0-18.2 years). Eight (8/22, 36%) of them had pacemaker implantation within the first 30 days after cardiac surgery. All symptomatic patients noted resolution of symptoms after pacemaker implantation. During the mean follow-up time of 4.3±3.8 years, no mortality was seen.

Conclusion: Although SND is rare in children, it has been diagnosed with increasing frequency with structural heart disease especially in patients who have undergone corrective cardiac surgery related with atrial tissue. Since SND can occur at any time postoperatively, these patients should be kept under constant control. If symptomatic sinus node dysfunction is confirmed, permanent pacing is an effective therapeutic modality

Table 1: Clinical characteristics of patients

Patients	77
Male	50/64.9%
Age at diagnosis (months) Mean (±SD)	8.2±6.3
Weight at diagnosis(kg) Mean (±SD)	28.2±18.8
Initial symptom or signs	
✓ *Syncope& Dizziness*	29
✓ *Palpitation*	11
✓ *Bradicardia*	15
✓ *Fatique*	6
Asymptomatic	16
Structural heart disease	**58 (75.3)**
✓ ***Congenital heart disease****	
○ *Univentricular heart*	9
○ *Transposition of great arteries*	8
○ *Atrial septal defekt*	7
○ *Atrioventricular septal defect (Complete or partial)*	5
○ *Partial/total anomalous pulmonary venous connection*	3
○ *Ventricular septal defect*	3
○ *Others*	11
○ *Left atrial isomerism*	7
✓ **Cardiomyopathy**	
○ Left ventricular noncompaction cardiomyopathy	5
○ Hypertrophic cardiomyopathy	2
○ Restrictive cardiomyopathy	2
○ Dilated cardiomyopathy	1
✓ **Myocarditis**	1
Additional arrythmia substrates **	**30(39)**
✓ *Supraventricular tachycardia*	
○ Atrial fibrilation	4
○ Intraatrial reentran tachycardia	6
○ Short RP SVT	4
○ Focal atrial tachycardia	3
○ Multi focal atrial tachycardia	1
○ Junctional ectopic tachycardia	1
✓ Ventricular *arrythmias*	
○ Ventricular fibrilation	1

○ Idioventricular rhythm	2
○ Premature Ventricular contraction	3
✓ Others	
○ Complete AV block	2
○ Long QT	2
○ Short QT	1
Follow up time(years) Mean (±SD)	4.3±3.8

Values mean±SD or n (%)

*: Some patients had multipl or complex congenital cardiac disease.

**: Some patients had more than one *arrythmia* substrat.

Table 2. Pacemaker data of patients

Patients	39(50.6)
Gender(male)	27(69.2)
Age(years)	8.7±6.1(0.5-18)
Postoperative	22(56.4)
Isolated	17(43.6)
Pacemaker Type	
✓ *Epicardial*	25 (64.1)
✓ *Transvenous*	14 (35.9)
Pacemaker indication	
✓ *Symptoms correlate with* severe bradycardia or pauses	29
✓ Significant bradycardia that does not improve in the early postoperative period	8
✓ *Preoperative symptomatic SND (in left atrial isomerism petient)*	1
✓ *Cardiac arrest*	1
Pacemaker modes	
✓ *DDD*	16
✓ *AAI*	13
✓ *VVI*	7
✓ *ICD*	1
✓ *CRT*	2
Pacemaker complications	
✓ *Pocket infection*	1
✓ *Lead fracture*	1
✓ *Lead detechment*	1
✓ *Lead Insulation defect*	1

Values mean±SD or n (%)