



Sinus node dysfunction in children: different aetiologies, similar clinical course in two-centre experience

Original Article

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Abstract

Aim: This study aims to evaluate the clinical characteristics and outcomes of children diagnosed with sinus node dysfunction. *Methods:* This was a retrospective review of patients diagnosed with sinus node dysfunction in two tertiary paediatric cardiology centres in Turkey from January 2011 to June 2022. *Results:* In all, 77 patients (50, 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. While age-incompatible bradycardia and pauses were the most common rhythm disturbances, syncope, presyncope, and dizziness (n:33, 43%) were the most frequent initial symptoms. Structural heart disease was present in 58 (75.3%) of the 77 patients, 47 (61%) of whom were congenital. The most commonly associated CHDs were transposition of the great arteries (n:8), atrial septal defect (n:7), and atrioventricular septal defect (n:5). Seven of them also had left atrial isomerism. The remaining 19 patients were isolated. Four patients had SCN5A mutation (two of them were siblings) and two of them had Emery–Dreifuss muscular dystrophy. *Conclusion:* Although sinus node dysfunction 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 to atrial tissue. Since sinus node dysfunction 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.

Introduction

Although sinus node dysfunction is most often related to age-dependent progressive fibrosis of the sinus nodal tissue and surrounding atrial myocardium,¹ the disorder is being diagnosed with increasing frequency in paediatric and young adult patients.

Symptoms attributable to sinus node dysfunction can range from mild fatigue to frank syncope. The key to diagnosing sinus node dysfunction is to establish a correlation between the patient's symptoms and the electrocardiogram or 24-hour Holter electrocardiogram findings at the time of symptoms.

Sinus node dysfunction exhibits various non-specific clinical manifestations and may even be asymptomatic. Common symptoms include dizziness, syncope, reduced exercise capacity, and increased fatigue. In the evaluation of unexplained dizziness or syncope in children, consideration of sinus node dysfunction in the differential diagnosis is crucial.

Although there is a large body of literature on sinus node dysfunction in adults, there is a limited amount in children. This study aims to assess the clinical characteristics and follow-up data of paediatric sinus node dysfunction patients in two tertiary paediatric cardiology and heart surgery centres.

Patients and methods

We retrospectively evaluated patients under the age of 18 diagnosed with sinus node dysfunction over an 11-year period, January 2011–June 2022. This retrospective study was approved by the institutional ethics committee and was conducted in accordance with the principles of the Declaration of Helsinki.

Sinus node dysfunction is the inability of the sinoatrial node to produce an adequate heart rate that meets the physiologic needs of the individual. Sinus node dysfunction is defined with

accompanying symptoms with 12-lead electrocardiogram or 24-hour electrocardiogram-Holter monitor findings such as sinus bradycardia (less than appropriate for age), sinus pause or arrest for more than two seconds, sinoatrial exit block, tachycardia-bradycardia syndrome (sinus bradycardia, ectopic atrial bradycardia), or sinus pause alternating with periods of abnormal atrial tachycardia, atrial flutter, or fibrillation.¹ When symptoms are associated with exertion, the patient has an exercise electrocardiogram test to assess chronotropic incompetence. A diagnosis of chronotropic incompetence is made if the patient is unable to meet 80% of the maximum heart rate (220 bpm – age) during exertion.

For each patient enrolled, data regarding demographics, cardiac diagnosis, age at first surgical repair, age at pacemaker implantation, and genetic abnormalities were collected from the system database. The demographic data, symptoms at the time of admission, familial history, 12-lead electrocardiogram, transthoracic echocardiography, 24-hour Holter monitoring (three channels and 12-lead, if required), exercise testing, genetic testing, and electrophysiology study data of the patients were evaluated retrospectively.

The diagnosis of sinus node dysfunction was documented on hospital records, electrocardiogram, 24-hour Holter monitoring, exercise testing, and pacemaker or loop recorder interrogation, whenever available.

Sinus node dysfunction was defined as “early” if it occurred within 30 days after the first surgery, persisting for at least 7–10 days. It was considered “late” sinus node dysfunction if diagnosed more than 30 days after the first surgery, requiring pacemaker (PM) implantation with Class I indication according to the ACC/AHA/NASPE and ACCF/AHA/HRS guidelines and recent literature data.¹

In eligible patients (≥ 6 years), exercise stress testing was carried out using the modified Bruce treadmill protocol. During the exercise stress test, 12-lead electrocardiogram and blood pressure were monitored. The exercise test was suggested to have stopped when there was a failure of heart rate to increase with exercise, severe hypotension or progressive fall in systolic blood pressure, the presence of ≥ 2 mm ST segment changing, increase in ventricular ectopy with exercise or the presence of ventricular tachycardia, and, finally, if complaints or signs of fatigue, dyspnoea or dizziness occurred in patients during exercise testing.²

Patients diagnosed with sinus node dysfunction were categorised into two groups: those presenting with structural heart disease, including CHD or cardiomyopathy, and those without structural heart disease, termed idiopathic. The groups were compared in terms of gender, age at diagnosis, weight, need for pacemaker implantation, and prognosis.

Categorical variables are expressed as absolute numbers and percentages. Continuous variables are expressed as mean \pm standard deviation. Differences between means were calculated using the *t* test, where appropriate. Categorical variables were compared using the χ^2 test. Statistical analyses were performed using SPSS version 21.0 (SPSS, Inc., Chicago, Illinois). A 2-tailed value of *p* ≤ 0.05 was considered statistically significant.

Results

All demographic and clinical characteristics of patients are listed in Table 1. A total of 77 patients (50/77, 64.9% males) were identified. The mean age at diagnosis was 8.2 ± 6.3 years, and the mean weight was 28.2 ± 18.8 kg.

Mode of presentation

The initial symptoms of patients at diagnosis were syncope in 18 (23.4%), fatigue in 17 (22.1%), dizziness and presyncope in 15 (19.5%), and palpitations in 11 (14.3%). Sixteen (20.8%) of the patients were asymptomatic at the time of diagnosis. Fifty-eight (75.3%) of the patients had structural heart disease, while 47 (61%) had CHD, most commonly univentricular heart physiology (n:9), transposition of the great arteries (n:8), atrial septal defect (n:7), and atrioventricular septal defect (n:5). Seven of them also had left atrial isomerism. Ten (13%) patients had cardiomyopathy. Of these, five had left ventricular noncompaction cardiomyopathy, two had hypertrophic cardiomyopathy and restrictive cardiomyopathy, and one had dilated cardiomyopathy.

Of the total patient population, 44 (57.1%) had previously undergone a cardiac operation, and all but one patient had developed sinus node dysfunction after a surgical procedure. The exception was a patient with left atrial isomerism in whom sinus node dysfunction was diagnosed during 24-hour Holter monitoring before surgery. The most frequently performed surgical procedures were atrial septal defect (7), Fontan surgery (6), atrioventricular septal defect (5), Senning surgery (5), and abnormal pulmonary venous connection anomaly repair (3). Thirty-nine (50.6%) patients underwent pacemaker implantation (n = 25, 64.1% of whom were epicardial). Twenty-two of them were implanted after cardiac surgery. The mean interval between pacemaker implantation and the previous operation was 5.1 years (range, 0–18.2 years). Indications for permanent pacemakers were symptoms correlated with severe bradycardia or pauses in 29 patients and significant bradycardia that did not improve in the early postoperative period in eight patients. One patient had left atrial isomerism, and sinus node dysfunction was already diagnosed during 24 hours of Holter monitoring before surgery. Based on her symptoms, pacemaker implantation was performed intraoperatively (Table 2).

The following pacing system was applied to 36 patients: dual chamber (DDDR), atrial demand (AAIR), and ventricular demand (VVI/VVIR) pacing systems, applied to 16/13/7 patients, respectively. Of the remaining three patients, two had cardiac resynchronisation therapy, and one had implantable cardioverter-defibrillator implanted (Table 2). Implantable cardioverter-defibrillator implantation was performed on one patient who was followed up in another centre with a diagnosis of left ventricular noncompaction cardiomyopathy and sinus node dysfunction and was admitted to our hospital after cardiac arrest due to ventricular fibrillation. Cardiac resynchronisation therapy implantation was performed in a two-year-old patient with transposition of the great arteries who underwent a Senning operation and in a patient with an Ebstein anomaly diagnosis who underwent a cone operation due to systolic dysfunction in the follow-up.

Familial sinus node dysfunction was identified in four patients. Among them, two were siblings with heterogeneous mutations found in the SCN5A and TTN genes, and they also presented with hypertrophic cardiomyopathy. Another patient exhibited a homozygous mutation in the RYR2 gene, while his older brother, with the same mutation, received an implantable cardioverter-defibrillator due to sustained ventricular tachycardia.

Thirty (39%) additional arrhythmia substrates were detected at the time of diagnosis or during follow-up of the patients, and some patients had more than one (Table 1). The most common rhythm disorders were intra-atrial re-entrant tachycardia (n:6), atrial

Table 1. Clinical characteristics of patients

Patients	77
Male	50(64.9)
Age at diagnosis (years) Mean (\pm SD)	8.2 \pm 6.3
Weight at diagnosis(kg) Mean (\pm SD)	28.2 \pm 18.8
Initial symptom or signs (n-%)	
• Syncope	18 (23.4)
• Fatigue	17 (22.1)
• Presyncope and Dizziness	15 (19.5)
• Palpitation	11 (14.3)
• Asymptomatic	16 (20.8)
Structural heart disease	58 (75.3)
<i>Congenital heart disease*</i>	
• Univentricular heart	9
• Transposition of great arteries	8
• Atrial septal defect	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 arrhythmia substrates **	30(39)
Supraventricular tachycardia	
• Atrial fibrillation	4
• Intraatrial reentrant tachycardia	6
• Short RP SVT	4
• Focal atrial tachycardia	3
• Multi focal atrial tachycardia	1
• Junctional ectopic tachycardia	1
Ventricular arrhythmias	
• Ventricular fibrillation	1
• Idiopathic ventricular rhythm	2
• Premature Ventricular contraction	3
Others	
• Complete AV block	2
• Long QT	2
• Short QT	1
Follow up time(years) Mean (\pm SD)	4.3 \pm 3.8

Values mean \pm SD or n (%)

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

** : Some patients had more than one *arrhythmia* substrate.

fibrillation (n:4), short RP supraventricular tachycardia (n:4), and focal atrial tachycardia (n:3). Two patients had complete atrioventricular block, in addition to sinus node dysfunction. Electrophysiological study \pm transcatheter ablation was performed in 13 patients. Because some patients had more than one arrhythmia, multiple different transcatheter ablations were performed at the same time. Details are shown in Table 3.

A patient diagnosed with restrictive cardiomyopathy and implanted with a pacemaker in single-chamber AAI mode presented with syncope again. The patient developed a second- and third-degree atrioventricular block. Cardiac functions deteriorated, and it was decided to put them on the heart transplant list. The single-chamber pacemaker was upgraded to dual-chamber implantable cardioverter-defibrillator.

During a mean follow-up period of 4.3 ± 3.8 years, two patients (one from noncardiac causes) died. A nine-month-old female patient died due to metabolic disease and pneumonia. The second one underwent a cone operation due to an Ebstein anomaly in another centre and cardiac resynchronization therapy pacemaker implantation due to complete atrioventricular block, sinus node dysfunction, and poor cardiac functions; he was referred to us due to the development of intra-atrial re-entrant tachycardia in the follow-up. After the first successful ablation, intra-atrial re-entrant tachycardia relapsed. Extracorporeal membrane oxygenation was applied. A haemodynamically unstable patient underwent a second intra-atrial re-entrant tachycardia ablation but died under extracorporeal membrane oxygenation.

No statistically significant difference was observed in terms of age at diagnosis, gender, weight, pacemaker implantation rate, or prognosis between patients diagnosed with sinus node dysfunction due to structural heart disease and sinus node dysfunction of idiopathic origin ($p > 0.05$). However, the follow-up period in patients diagnosed with sinus node dysfunction due to structural heart disease was longer than in the patient group with idiopathic origin ($p:0.052$) (Table 4).

Discussion

To the best of our knowledge, this is one of the largest paediatric studies to evaluate the clinical and genetic characteristics of sinus node dysfunction and its outcomes in children.

The most noteworthy findings are as follows:

- Although sinus node dysfunction usually occurs in the elderly, it is also common in children with structural heart disease, especially in patients who have undergone cardiac surgery.
- Sinus node dysfunction presents with a wide variety of symptoms and prognoses, ranging from no symptoms to the presence of syncope. Sinus node dysfunction should be considered in the differential diagnosis of any child with unexplained dizziness or syncope.
- The diagnosis of sinus node dysfunction in paediatric patients is usually based on a combination of clinical settings and rhythm documentation. Routine electrophysiological studies are not recommended.
- Since sinus node dysfunction can occur at any time after surgery, these patients should be kept under constant monitoring or surveillance.
- Permanent pacemaker implantation is a safe and effective treatment method for patients with symptomatic sinus node dysfunction.

Table 2. Data of patients with pacemakers

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)
Type of Pacemaker	
• Epicardial	25 (64.1)
• Transvenous	14 (35.9)
Pacemaker indication	
• Symptoms associated 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 patient)	1
• Cardiac arrest	1
Pacemaker modes	
• DDDR	16
• AAIR	13
• WDIR	7
• ICD	1
• CRT	2
Pacemaker complications	
• Pocket infection	1
• Lead fracture	1
• Lead detachment	1
• Lead Insulation defect	1

CRT = cardiac resynchronization therapy; ICD = implantable cardioverter-defibrillator; SND = Sinus node dysfunction.

Table 3. Electrophysiological study± ablation data

TEEPS (positive SND findings)	1
EPS (positive for sinus node dysfunction)	4
IART RF ablation	5
Focal atrial tachycardia irrigated RF ablation	2
Multifocal atrial tachycardia RF ablation	1
Purkinje fiber-induced PVC irrigated RF ablation	1
Typical AVNRT cryoablation	1
Junctional ectopic tachycardia cryoablation	1

AVNRT = Atrioventricular nodal reciprocating tachycardia; EPS = Electrophysiology study; IART = Intraatrial reentrant tachycardia; RF = Radiofrequency; PVC = Premature ventricular contraction; SND = Sinus node dysfunction; TEEPS = Transesophageal electrophysiological study.

The clinical manifestations of sinus node dysfunction vary depending on the age, underlying haemodynamic status, and functionality of the remaining conduction system. It is important to evaluate the patient's rhythm when a child presents with

unexplained seizures or syncope. Syncope, presyncope, and dizziness were the most common signs in our patients and were seen in 43% of the patients. Conversely, 20.8% of the patients were asymptomatic at the time of diagnosis.

Although sinus node dysfunction usually occurs in the elderly, it is increasingly diagnosed in paediatric and young adult patients. Sinus node dysfunction in young patients is most common in children with CHDs, especially in patients who have undergone cardiac surgery.³ Despite the development of surgical techniques in recent years, sinus node dysfunction is still seen in the postoperative period. The incidence is highest in patients who have undergone atrial repair (Mustard or Senning) for transposition of the great arteries.⁴ Five of our patients underwent atrial repair (Senning) for transposition of the great arteries. Sinus node dysfunction may develop immediately after the first surgical repair or may occur years later. The early onset of postoperative sinus node dysfunction may be the result of surgical procedures near the sinus node, while the late onset of sinus node dysfunction occurs because of progressive fibrosis at the surgical site near the sinus node. The optimal time for pacemaker implantation in postoperative patients is unknown. If sinus node dysfunction is due to oedema of the node, improvement may be noted over time, and the need for permanent cardiac pacing may be circumvented. In other patients, irreversible damage to the node may cause immediate dysfunction or problems that arise many years later.⁵ In the present study, the mean time between surgery and pacemaker implantation was 5.1 years, while eight patients underwent PM implantation within the first 30 days postoperatively.

Sinus node dysfunction can also be observed due to acquired or familial myocardial diseases, such as cardiomyopathies and inflammatory or ischaemic diseases.³ Ten (13%) of our patients had cardiomyopathy. Five had left ventricular noncompaction cardiomyopathy, two each had hypertrophic cardiomyopathy and restrictive cardiomyopathy, and one had dilated cardiomyopathy. Sinus node dysfunction is called idiopathic when no other cause is found. Nineteen of our patients were idiopathic. Familial transmission or inheritance have been reported but are probably uncommon.⁶ Two of our patients were siblings. The SCN5A gene is the most commonly identifiable gene responsible for inherited conduction disease and accounts for only 5% of progressive conduction disease cases.¹ Four patients had SCN5A mutation, and two of them were siblings. Mutations in the SCN5A gene have been reported in association with several cardiac electrophysiological disorders, including long QT syndrome type 3, Brugada syndrome, progressive cardiac conduction defects, and sinus node dysfunction.⁷ In addition to providing appropriate medical intervention for affected cases, identifying genetic defects would help provide proper genetic counselling to patients and their families. Moreover, testing at-risk family members for carrier status and targeted premarital screening would also be possible to prevent the recurrence of a potentially fatal disease.⁸

The diagnosis of sinus node dysfunction in paediatric and young adult patients is usually based on a combination of clinical settings and rhythm documentation. Patients with sinus node dysfunction due to a surgical injury have symptomatic arrhythmias that may be detected by monitoring in the immediate and late postoperative periods. In contrast, sinus node dysfunction often goes undiagnosed in young patients with normal cardiac anatomy. In these patients, sinus bradycardia or intermittent tachyarrhythmias can easily be overlooked, especially when pronounced sinus arrhythmia is considered physiological. Also, the disorder is unlikely to be detected in these patients unless they have syncope or

Table 4. Characteristics of patients with and without structural heart disease

	Structural heart disease (n:58) (44 / 76% operated)	Idiopathic (n:19)	p value
Gender (Male)	51 (88%)	15(79%)	0.207
Age at diagnosis (years) Mean (min-max)	8 (33 days–18.2 years)	8.7(20 days–17 years)	0.906
Weight at diagnosis(kg) Mean (min-max)	26.8 (4–64)	31.8(4–70)	0.478
Follow up time(months) Mean (\pm SD)	57.8 \pm 45.9	34.3 \pm 45.9	0.052
Additional arrhythmia substrates	26 (45%)	6(32%)	0.207
Pacemaker implantation	27(47%)	12(63%)	0.298
Exitus	1	1*	0.432

*: noncardiac cause.

dizziness. The diagnosis of sinus node dysfunction has been on the rise due to increased awareness of the disorder and the availability of cardiac monitoring in recent years. Electrophysiology study is not routinely performed for the diagnosis of sinus node dysfunction. Electrophysiology study may be considered in symptomatic patients suspected of sinus node dysfunction who are also undergoing an electrophysiology study for another indication or when the diagnosis remains uncertain after an initial non-invasive evaluation (Class IIb).¹

In left atrial isomerism, there is a lack of a normal sinus node and atrioventricular node, which predisposes these patients to bradyarrhythmia.⁹ A study including left atrial isomerism patients aged 15–30 years demonstrated that half of these patients had sinus node dysfunction.¹⁰ Ozawa et al. also reported that 10/16(62%) of their left atrial isomerism patients had SND.¹¹ In the present study, seven (9%) patients had left atrial isomerism. Sinus node dysfunction was detected postoperatively in six patients with left atrial isomerism, while sinus node dysfunction was detected preoperatively in one patient, and pacemaker implantation was performed intraoperatively because the patient was symptomatic.

Emery-Dreifuss muscular dystrophy is an inherited muscular dystrophy often accompanied by cardiac abnormalities in the form of supraventricular arrhythmias, conduction defects and sinus node dysfunction. Cardiac findings typically occur years after skeletal muscle involvement but are important because they can be severe and life-threatening. The clinical picture characterised by joint contractures, progressive muscle weakness, and atrophy and cardiac symptoms is observed in the third decade of life. Although cardiac abnormalities are not common in paediatric patients with Emery-Dreifuss muscular dystrophy, the clinical course and sequence of muscle and cardiac manifestations may vary due to genotypes.¹² The diagnosis was made in two of our patients with Emery-Dreifuss muscular dystrophy while investigating one for drop foot and the other for joint contractures. Although they did not describe any obvious cardiac symptoms, one patient was in atrial fibrillation at presentation and also had frequency premature ventricular contraction, and the other had pauses of four seconds. While dual chamber pacemaker implantation was performed in DDDR mode in the patient with pauses, implantable cardioverter-defibrillator implantation was performed in the patient who had atrial fibrillation, frequent premature ventricular contraction, and non-sustained ventricular tachycardia after synchronised cardioversion.

Permanent pacemaker placement is recommended only in patients with symptomatic sinus node dysfunction and documented bradycardia that is not caused by correctable extrinsic factors.¹³ There is no clear consensus regarding the minimum heart rate or maximum pause time for pacemaker implantation, as with a complete atrioventricular block.¹⁴ In accordance with the guidelines, in our clinic, we performed pacemaker implantation in symptomatic patients with documented bradycardia or secondary symptoms of long pauses, as well as in patients with chronotropic incompetence or impaired systemic ventricular functions as a result of bradycardia. We performed a routine 24-hour Holter electrocardiogram evaluation in the preoperative period for left atrial isomerism patients. Accordingly, we applied simultaneous pacemaker implantation during the operation in patients with symptomatic sinus node dysfunction.

The choice of permanent pacemaker is either an AAIR pacemaker with just an atrial lead or a DDDR pacemaker with both atrial and ventricular leads. Most guidelines recommend DDDR pacing as the first choice of pacing. The major disadvantage of AAIR pacing is the possibility of developing an atrioventricular block in the future, which will require the addition of a ventricular lead. Atrial pacing is associated with a reduced incidence of atrial fibrillation compared with ventricular pacing,^{15–17} but atrioventricular block may occur even in carefully selected patients. We also preferred the atrial-based pacing mode in symptomatic patients with sinus node dysfunction. For patients with dual chamber pacemakers and intact atrioventricular conduction, we also programme the dual chamber pacemaker to minimise ventricular pacing (using managed ventricular pacing (managed ventricular pacing (MVP) mode)). In many studies, right ventricular pacing is not recommended, as it may cause ventricular dyssynchrony in the future.^{18,19} However, we preferred single-chamber ventricular pacing techniques, especially in some patients who were underweight, had comorbidities, were suspected of having progressive conduction disease, or might have developed atrioventricular block in the future.

Although pacing in the paediatric population has been characterised by poor performance of leads and generators in the past, according to recent advances in technology, permanent cardiac pacing has become more reliable and more applicable to paediatric patients. During the follow-up period, lead fracture, lead detachment, and lead insulation defects occurred in one patient each, and lead replacement was made.

Conclusion

While sinus node dysfunction is rare in children, it is more common in patients who have undergone corrective cardiac surgery involving atrial tissue. As sinus node dysfunction can manifest at any time postoperatively, it is essential to maintain continuous monitoring of these patients. Permanent pacemaker implantation has proven to be a safe and effective treatment for patients with symptomatic sinus node dysfunction. Additionally, sinus node dysfunction can be caused by various aetiologies. However, regardless of the specific aetiology, as observed in the present study, there was no significant difference in terms of demographic characteristics, treatment strategies, or prognosis.

Authors contributions. GT, CK, SO, and HK had primary responsibility for the data collection and analysis of the results. GT, YE, and FS contributed to the paper in all stages by systematically organising the data, while GS and YE critically appraised and wrote of the manuscript. SH and AG critically appraised and edited the paper.

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Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975 and the Istanbul Mehmet Akif Ersoy Research and Training Hospital Institutional Review Board (2023-09/03.01.2023) approved the study.

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