PEDIATRIC HEART JOURNAL

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TÜRK PEDİATRİK KARDİYOLOJİ VE KALP CERRAHİSİ DERNEĞİ ADINA SAHİBİ
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YAYIN PERİYODU VE TÜRÜ
Pediatric Heart Journal 4 ayda bir olmak üzere yılda 3 sayı yayınlanır (Mart-Ağustos-Aralık).
Yerel süreli yayın.

ADRES DEĞİŞİKLİKLERİ
Derginin yayınlandığı tarihlerden en az 15 gün önce dernek yazışma adresine bildirilmelidir. Zamanında yapılmayan bildirimler nedeniyle derginin aboneye ulaşmamasından yayıncı sorumlu tutulamaz.

BASILDIĞI YER-BASIMCI-YAYIMCI
Ortadoğu Reklam Tanıtım Yayıncılık Turizm Eğitim İnşaat Sanayi ve Ticaret A.Ş. (Türkiye Klinikleri)
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Basıma veriliş tarihi: 08.01.2018

ISSN: 2148-4910
Online ISSN: 2458-7591

THE OWNER ON BEHALF OF TURKISH PEDIATRIC CARDIOLOGY AND CARDIAC SURGERY SOCIETY
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PUBLICATION TYPE AND PERIODS
Pediatric Heart Journal is published 3 times a year (March-August-December).
Local periodic publication.

CHANGE OF ADDRESS
Notify the subscription office for the change of address at least 15 days before the date of issue. The publisher will not be responsible from any lost resulting from an address change if not informed.

PUBLISHING HOUSE-PUBLISHER
Ortadoğu Advertisement Presentation Publication Tourism Education Construction Industry and Trade Co. (Türkiye Klinikleri)
Türkocağı Cad. No:30 06520 Balgat/Ankara/Turkey
Phone : +90 312 286 56 56
Fax : +90 312 220 04 70
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PEDIATRIC HEART JOURNAL

Cilt Vol 3 Sayı No 3 Yıl Year 2016

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2014 *yılında pediatrik kardiyoloji alanında Türkiye'de üretilen çalışmaları yayınlamak üzere yola çıktığımız "Pediatric Heart Journal" dergisinin yayınına üzüntü ile son verdiğimizi sizlere duyurmak isterim.*

Tüm dünyada artan dergi sayısı ile birlikte üretilen bilimsel yayınları yayınlayacak çok seçenek olmaya başlamıştır. Mütevazi bir hedefle yayın yaşamına başlayan dergimiz, 2 yıl pek çok merkezde çalışan yazarlarımızın katkısı ile ayakta kalabilmeyi başardı. Ancak 2017 yılında dergimizi çıkaracak kaliteli yayın bulmakta yaşanılan zorluklar, yayın yaşamamızı sonlandırma konusunu gündeme getirdi. Yönetim Kurulumuzun da kararı ile dergimiz, elimizdeki son yazıları yayınlandıktan sonra işlevini sonlandıracaktır.

Bu süreçte desteklerini esirgemeyen tüm pediatrik kardiyoloji ve kalp damar cerrahisi derneği yönetim kurulu üyelerine, yazılarını esirgemeyen yazarlarımıza, derginin yayınlanması konusunda desteklerini veren Türkiye Klinikleri yönetim ve çalışanlarına, yayın kurulumuzdaki tüm arkadaşlarıma sonsuz teşekkür ederim.

Saygılarımla.

Prof. Dr. N. Kürşad TOKEL

Editör

Basal Electrocardiography and Holter Monitoring Findings in Children Presenting with Vasovagal Syncope

Vazovagal Senkoplu Çocuklarda Bazal Elektrokardiyografi ve Holter Monitorizasyon Bulguları

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Geliş Tarihi/*Received:* 29.09.2016
Kabul Tarihi/*Accepted:* 03.01.2017

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ABSTRACT Objective: This study aims to determine the basal electrocardiography and Holter monitoring findings of the children with vasovagal syncope. **Metarial and Methods:** This is a prospective review of 92 children with vasovagal syncope and 50 healthy children who were matched with respect to age, sex and body mass index. All children had basal electrocardiography so that p-wave dispersion (difference between maximum and minimum p-waves), QT interval (difference between the onset of QRS complex and the end of T-wave), QT dispersion (difference between maximum and minimum QT intervals), T-peak-to-T-end interval (interval between the peak and end of T-wave) and T-peak-to-T-end dispersion (difference between maximum and minimum T-peak-to-T-end intervals) were measured. Then, all children underwent Holter monitoring and heart rate variability was assessed in the time domain analysis. Ninety-two patients with vasovagal syncope underwent tilt-table test which had positive results in 42 children including 4 children with cardioinhibitory response, 12 children with vasodepressor response, 12 children with mixed response and 14 children with postural orthostatic tachycardia syndrome. All two-tailed p values < 0.05 were accepted as statistically significant. **Results:** When compared with healthy controls, tilt test positive children had significantly slower heart rate, longer p minimum, longer p maximum, greater p-wave dispersion, longer QT minimum, longer QT maximum, greater QT dispersion and longer T-peak-to-T-end intervals in V2, V3, V4, V5 and V6 leads. The tilt test positive children had significantly higher heart rate variability and longer T-peak-to-T-end intervals in V2, V5 and V6 leads than the tilt test negative children ($p<0.05$ for all). **Conclusion:** Electrocardiography findings can be used to decrease the need for the tilt-table test in children with vasovagal syncope.

KeyWords: Ambulatory electrocardiography; child; electrocardiography; holter monitoring; tilt-table test; vasovagal syncope

ÖZET Amaç: Vazovagal senkoplu çocuklarda bazal elektrokardiyografi ve Holter monitorizasyonu bulgularının nasıl değiştiğinin belirlenmesi amaçlanmıştır. **Gereç ve Yöntemler:** Yaş, cinsiyet ve vücut kitle indeksi bakımından benzer 92 vazovagal senkoplu çocuk ve 50 sağlıklı çocuk karşılaştırıldı. Olguların tümünde bazal elektrokardiyografi kaydedilerek p-dalgası dispersiyonu (maksimum ve minimum p-dalgalarının farkı), QT aralığı (QRS kompleksinin başlangıcı ve T-dalgasının sonu arasındaki fark), QT dispersiyonu (maksimum ve minimum QT aralıklarının farkı), T-tepe-T-son aralığı (T-dalgasının tepesi ve sonu arasındaki fark) ve T-tepe-T-son dispersiyonu (maksimum ve minimum T-tepe-T-son aralıklarının farkı) ölçüldü. Kalp hızı değişkenliğini ölçmek için olguların tümünde 24 saat boyunca elektrokardiyografi monitorizasyonu uygulandı. Vazovagal senkoplu 92 çocuğa tilt masa testi yapıldı; bu test 50 çocukta negatif sonuçlanırken 42 çocukta pozitif sonuç (dördünde kardioinhibitör yanıt, 12'sinde vazodepresör yanıt, 12'sinde karışık yanıt ve 14'ünde postural ortostatik taşıkardi sendromu) belirlendi. Çift taraflı p değerleri <0.05 ise istatistiksel olarak anlamlı kabul edildi. **Bulgular:** Sağlıklı kontrol olgularıyla karşılaştırıldığında, tilt pozitif çocuklarda kalp atım hızı daha düşüktü, p minimum ve maksimum değerleri daha uzundu, p-dalgası dispersiyonu daha genişti, QT minimum ve maksimum değerleri daha uzundu, QT dispersiyonu daha genişti ve V2, V3, V4, V5 ve V6 elektrotlarındaki T-tepe-T-son aralıkları daha uzundu. Kalp hızı değişkenliği ve V2, V5 ve V6 elektrotlarındaki T-tepe-T-son aralıkları, tilt testi negatif çocuklara göre tilt testi pozitif çocuklarda daha uzundu (hepsi için $p<0.05$). **Sonuç:** Vazovagal senkoplu çocuklarda tilt masa testine duyulan gereksinimi azaltmak için elektrokardiyografi bulgularından yararlanılabilir.

Anahtar Kelimeler: Ambulatuar elektrokardiyografi; çocuk; elektrokardiyografi; holter monitorizasyonu; tilt masa testi; vazovagal senkop

Pediatr Heart J 2016;3(3):89-96

Vasovagal syncope is most commonly observed in children and adolescents, but the precise pathophysiology of this clinical entity still remains obscure. It has been hypothesized that the nucleus tractus solitarii of the brainstem is somehow activated by a triggering stimulus which results in simultaneous enhancement of parasympathetic nervous system and withdrawal of sympathetic nervous system. On one end of the spectrum, the enhancement of parasympathetic tonus leads to the cardioinhibitory response which is characterized by a decrease in heart rate and myocardial contractility. On the other end of the spectrum, the withdrawal of sympathetic nervous system causes vasodilatation which leads to a drop in blood pressure without much change in heart rate. The majority of people with vasovagal syncope have a mixed response somewhere between these two ends of the spectrum.[1-4]

The head up tilt test has been considered as the reference standard for the diagnosis of vasovagal syncope in adults. However, recent guidelines of the European Society of Cardiology report that normal physical examination and electrocardiography are sufficient to diagnose neurocardiogenic syncope in children with a typical history. Thus, tilt test should be used carefully for primary identification of patients with syncope.[5-7]

Electrocardiography findings related with p-wave (the shortest duration, longest duration and p-wave dispersion) reflect atrial depolarization while those associated with QT interval (the shortest interval, longest interval and QT dispersion) indicate ventricular depolarization and repolarization. In addition, electrocardiography findings related with T-wave (T peak-to-T-end interval and dispersion) reflect ventricular repolarization. A 24-hour electrocardiography monitorization also allows the evaluation of heart rate variability which has been recently defined to examine the well-being and autonomous innervation of the heart.[8,9]

The present study aims to determine the basal electrocardiography and Holter monitoring findings of the children with vasovagal syncope.

MATERIAL AND METHODS

This prospective study was approved by the Ethical Committee of Afyon Kocatepe University Hospital where it was conducted at the Department of Pediatric Cardiology from May 2014 to April 2015. All procedures contributing to this work comply with the ethical standards of the Helsinki Declaration of 1975, as revised in 2008. Written informed consent was obtained from all participants and their parents.

STUDY GROUP

The study group included 92 children presenting with vasovagal syncope and 50 healthy children who were matched with respect to age and sex. The children presenting with vasovagal syncope had at least two unexplained syncope attacks. The diagnosis of vasovagal syncope was based on history and symptoms. In order to exclude any structural cardiac diseases, American Society of Echocardiography guidelines were used to carry out an echocardiography examination.[10] The children with structural cardiac diseases, the children with arrhythmias, the children with neurologic diseases, the children with psychiatric diseases and the children who were using any drugs that could alter heart rate and blood pressure were excluded from the study.

ELECTROCARDIOGRAPHY MEASUREMENTS

Standard 12-lead electrocardiography was initially obtained on all patients while they were lying in a comfortable supine position within the outpatient facility. The electrocardiograms were recorded at a paper speed of 50 mm/s and a standardization of 0.5 mm/mV standardization. Such an adjustment was made to prevent the superimposition of R and S waves into the QRS complexes that were located above or below them and, thus, to allow more accurate measurement and calculation. During the recordings, all patients breathed freely and did not speak. All of the electrocardiography measurements were made by the same pediatric cardiologist (A.P.) who was blinded to the patients.

The electrocardiography measurements were made with a ruler at three consecutive cardiac cycles for each lead and the averages of these values were used for final analysis. The intra-observer coefficient of variation was calculated to be 1.0% ± 0.2% for p-wave duration, 1.2% ± 0.4% for QT interval and 1.1% ± 0.1% for T peak-to-T-end intervals.

The onset of the P wave was defined as the junction between the isoelectric line and the beginning of the deflection, and the offset of the P-wave was defined as the junction between the end of the P-wave deflection and the isoelectric line. The P maximum was defined as the longest duration and the P minimum as the shortest duration among the 12 leads. The duration of the P wave was measured in seconds. The difference between maximum and minimum p-wave durations was defined as the p-wave dispersion (Figure 1).

The QT interval was measured in seconds from the onset of QRS complex to the end of T-wave. In order to distinguish between T- and U-waves, the QT-interval was measured in lead II and if there were too many artifacts (i.e. T-wave amplitude very low) either lead III or V5 were used. The QT dispersion was defined as the difference between the maximum and minimum QT intervals. Both maximum QT interval and QT dispersion measurements were corrected for heart rate according to Bazzet's formula (QTc=QT/√RR) (Figure 1).

The T-peak-to-T-end interval was measured in milliseconds for each precordial lead and obtained from the difference between the peak of the T-wave and the end of T-wave. T-peak-to-T-end dispersion was defined as the difference between the maximum and minimum T-peak-to-T-end intervals in precordial leads V1 to V6 during a single beat (Figure 1).

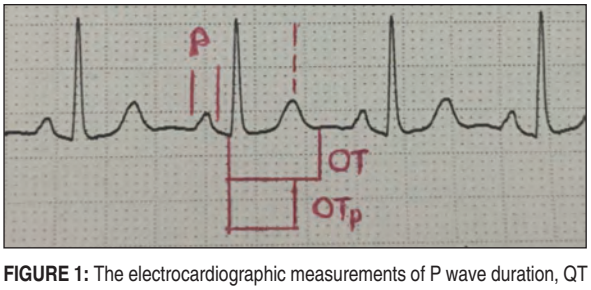

FIGURE 1: The electrocardiographic measurements of P wave duration, QT interval and T-peak-to-T-end interval.

HOLTER MONITORING

In order to assess heart rate variability, all patients underwent 24-hour electrocardiography monitoring (Reynolds Medical, Pathfinder Software Version V8.255). All tapes were subsequently analyzed by the same pediatric cardiologist (A.P.) who was blinded to the patients. Heart rate variability was measured in the time domain, using HRT View program (version 0.60-0.1 Software Program, Munich, Germany). The normal and aberrant complexes were discriminated, and all adjacent intervals between normal beats (NN) were collected over a period of 24 hours. All normal intervals were analyzed employing the time domain method. The time domain analysis included the mean of all normal R-R intervals (N-N), the standard deviation of all normal RR intervals (SDNN), the standard deviation of all the 5-minute normal RR interval means (SDANN), the square root of the mean of the squared differences between successive normal RR intervals over 24 hours (RMSSD), and the percentage of differences between successive normal RR intervals over 24 hours that are greater than 50 ms (PNN 50%).

HEAD UP TILT TEST

All patients presenting with vasovagal syncope underwent head up tilt test. The tilt test was performed in a quiet room with low lighting after 12 h of patient fasting from 8 PM to 8 AM.[11,12] No intravenous fluid infusions or pharmacological provocation was used during the test. Patients were monitored for continuous assessment of heart rate and rhythm and conventional automatic arm cuff blood pressure measurement in every 5 min. The tilt test protocol consisted of the patient lying in a supine position for 10 minutes while baseline electrocardiogram and blood pressure recordings were performed; then the patient was subsequently

tilted to a head-up position at 70° for 20 minutes. Whenever symptoms or alterations in blood pressure or heart rate were observed, blood pressure was recorded more frequently manually (in 30 second intervals). Previous investigations documented that relatively short protocols are convenient and satisfactory to address the sensitivity and specificity concerns.[13,14]

Forty-two patients reproducing syncope or presyncope during tilting were defined as tilt test-positive while fifty patients with no response at the end of 20 min were defined as tilt test-negative according to the Guidelines of Syncope of the European Society of Cardiology (version 2009).[7] Positive response was also defined as at least one of the following signs with or without syncope: (1) bradycardia, which was characterized by heart rate <75 bpm in children of 4–6 years old, heart rate<65 bpm in children of 7–8 years old, heart rate <60 bpm in children over 8 years old, sinus arrest, second degree atrioventricular block or higher, and asystole for 3 seconds; (2) hypotension defined as <80 mmHg in systolic blood pressure or decrease of >15 mmHg and/or diastolic blood pressure <50 mmHg; and (3) junctional rhythm together with escaped rhythm and accelerated idioventricular rhythm.[13,14]

STATISTICAL ANALYSIS

Collected data were analyzed by Graphpad Prism 6.1. software program on computerized media. Continuous variables were expressed as mean ± standard deviation (range: minimum-maximum) and categorical variables were denoted as numbers or percentages where appropriate. Statistical comparisons were performed by using unpaired Student's t test or one-way ANOVA followed by the Bonferroni post hoc test. Two-tailed p values less than 0.05 were considered to be statistically significant.

RESULTS

This study reviews 50 healthy children, 50 children presenting with vasovagal syncope and negative tilt test and 42 children presenting with vasovagal syncope and positive tilt test. The healthy controls, tilt test negative children and tilt test positive children were statistically similar in aspect of age, sex, height, weight and body mass index (p>0.05 for each) (Table 1).

Table 2 summarizes the electrocardiography findings of the study cohort. When compared with the healthy controls, the tilt test positive children had significantly slower heart rate, longer p minimum, longer p maximum, greater p-wave dispersion, longer QT minimum, longer QT maximum, longer OTc , greater QT dispersion and longer T-peak-to-T-end intervals in V2, V3, V4, V5 and V6 leads (p<0.05 for each). The tilt test positive children had significantly longer T-peak-to-T-end interval peaks in V2, V5 and V6 leads than the T-peak-to-T-end intervals in V2, V5 and V6 leads of the tilt test negative children (p<0.05 for each).

Table 3 shows the Holter monitoring findings of the children presenting with vasovagal syncope. When compared with the tilt test negative children, the tilt test positive children had significantly higher RMSSD and PNN 50% (p<0.05 for both).

DISCUSSION

Vasovagal syncope is characterized with relative inhibition of the sympathetic tonus and activation of the parasympathetic tonus within the heart.[15] The head up tilt test has been traditionally regarded as the gold-standard test for vasovagal syncope. An analysis of five studies that evaluate the head up tilt test in adults indicates the sensitivity rates as 13%, 25%, 31%, 35% and 75% (median of 31%) whereas the specificity rates were 100%, 100%, 95%, 92% and 89% (mean of 95%) respectively.[16] In a few pediatric studies, the sensitivity for head up tilt test varies from 43% to 49% and the specificity changes between 93% and 100%.[17] Therefore, the latest approach is that normal physical examination and electrocardiography are sufficient to stop further assessment in children with a typical history of reflex syncope.[7] The reason is that tilt test seems to have high false-negative and -positive rates and negative tilt test results do not exclude the diagno-

TABLE 1: Demographic and clinical characteristics of the study group.

	Healthy controls (n=50)	Head up tilt test negative (n=50)	Head up tilt test positive (n=42)
Age (years)	13.7±0.5	13.6±0.4	13.5±0.3
Male / Female	18/32 (36.0%/64.0%)	11/39 (22.0%/78.0%)	14/28 (33.3%/66.7%)
Height (cm)	158.1±1.2	156.3±1.8	159.1±1.7
Weight (kg)	49.2±2.1	47.8±1.7	48.7±1.9
Body mass index (kg/m2)	19.7±0.5	19.3±0.5	18.9±0.5

sis of neurally mediated syncope. That's why, the tilt test should be used to support a diagnosis of reflex syncope if this diagnosis has not been proved initially.[18,19]

It is well known that increased p-wave and QT dispersion can be a sign of autonomic dysfunction and autonomic tone may alter the duration of the P wave by affecting atrial conduction velocity.[20] P-wave dispersion is a simple and useful electrocardiographic marker reflecting inhomogeneous and discontinuing propagation of sinus impulses.[21] It has been reported to be a useful marker in autonomic neuropathy associated with diabetes mellitus and sickle cell disease.[22,23] Kose et al. were the first to evaluate the p-wave dispersion in patients with cardiogenic syncope. They stated that p maximum and p-wave dispersion were significantly greater in children with vasovagal syncope than in controls. Moreover, p-wave dispersion was significantly greater in the tilt test negative group than in controls.[12] Likewise, the tilt test positive children had significantly longer p minimum, longer p maximum and greater p-wave dispersion than the healthy controls in this study. These alterations in p-waves of the children with vasovagal syncope can be attributed to the imbalance between sympathetic and parasympathetic innervation which ultimately impairs the electrical conduction system within the atria.

The QT dispersion has been identified as a non-invasive tool for the assessment of the heterogeneity of repolarization within the ventricular myocardium. The larger the QT dispersion, the higher is the risk of reentry arrhythmias, and it is not age or gender dependent.[24,25] In apparently

TABLE 2: Electrocardiography findings of the study group.

	Healthy controls (n=50)	Head up tilt test negative (n=50)	Head up tilt test positive (n=42)
Heart rate (beats/min)	86.8±2.5	81.0±2.1	77.5±1.9*
P minimum (sec)	0.03±0.001	0.04±0.001	0.04±0.001*
P maximum (sec)	0.07±0.002	0.08±0.002	0.09±0.003*
P dispersion	0.04±0.001	0.05±0.001	0.05±0.001*
QT minimum (sec)	0.33±0.01	0.36±0.02	0.36±0.02*
QT maximum (sec)	0.31±0.02	0.31±0.03	0.32±0.03*
QT dispersion	0.04±0.01	0.05±0.001	0.05±0.001*
QTc	0.36±0.02	0.39±0.02	0.39±0.01*
T-peak-to-T-end (V1)	0.05±0.001	0.05±0.001	0.07±0.006
T-peak-to-T-end (V2)	0.05±0.004	0.06±0.001	0.06±0.002*†
T-peak-to-T-end (V3)	0.06±0.003	0.07±0.002	0.07±0.002*
T-peak-to-T-end (V4)	0.06±0.002	0.07±0.004	0.06±0.003*
T peak-to-T-end (V5)	0.06±0.003	0.06±0.004	0.07±0.005*†
T peak-to-T-end (V6)	0.06±0.005	0.06±0.002	0.07±0.002*†
T-peak dispersion	0.04±0.002	0.04±0.002	0.03±0.003

*There is statistically significant difference between healthy controls and tilt positive children ($p<0.05$).

† There is statistically significant difference between tilt negative and tilt positive children ($p<0.05$).

TABLE 3: Holter monitoring findings of the children with vasovagal syncope.

	Head up tilt test negative (n=50)	Head up tilt test positive (n=42)
Mean heart rate (beats/min)	82.3±1.3	81.3±1.4
Mean RR (msec)	734.7±12.2	740.9±13.3
SDNN (msec)	159.0±5.4	159.1±6.7
SDANN / 5 min (msec)	138.3±5.3	128.1±7.1
RMSSD (%)	53.3±3.1	65.0±4.3*
PNN (50%)	25.0±16.4	30.3±17.2*

*p<0.05 was accepted to be statistically significant.
SDNN-standard deviation of all RR intervals, SDANN/5 min-standard deviations of 5 minute mean values of RR, RMSSD-root mean square of successive difference of RR intervals, PNN (50%)-percentage of successive difference of RR>50 msec for each 5 minute interval

healthy populations, QT dispersion greater than 50 ms is considered abnormal, indicating an increased risk of arrhythmia and sudden death in various clinical entities including cardiomyopathies, mitral valve prolapse, ischemic heart disease, long QT syndromes and renal failure.[26] Pathologic QT dispersion also seems to predict premature atrial and ventricular contractions with electroconvulsive therapy.[27] In accordance, this study also shows significantly longer QT minimum, longer QT maximum and greater QT dispersion for the tilt test positive children when compared with the controls. These changes related with QT interval can be due to the autonomic dysfunction which simultaneously causes vasovagal syncope and contravenes the propagation of electrical stimuli throughout the ventricular myocardium.

T-peak-to-T-end interval is a marker of transmural dispersion of surface electrocardiography, which indicates ventricular repolarization. This interval also points out the risk of arrhythmia for both congenital and acquired ion channel diseases that lead to ventricular arrhythmias.[28] Prolonged T-peak-to-T-end interval was associated with the ventricular tachycardia in high risk patients who have organic heart disease.[29,30]

In an Iran study, the T-peak-to-T-end interval in lead V1 and T-peak-to-T-end dispersion were detected to be significantly larger in patients with a positive tilt test. Thus, it was suggested that ventricular repolarization might have been impaired in children presenting with vasovagal syncope.[31] In this study, the tilt test positive children had significantly longer T-peak-to-T-end intervals in V2, V3, V4, V5 and V6 leads than those of the same leads in healthy children. When compared with tilt test negative children, the tilt test positive children were also found to have significantly longer T-peak-to-T-end intervals in V2, V5 and V6 leads. The significant prolongation in T-peak-to-T-end intervals of the tilt test positive children may designate a delay in the ventricular repolarization process which may have been induced by the disturbance in the autonomic innervation of the heart.

Heart rate variability has become a popular indicator for autonomic imbalance.[32] Longin et al. compared short-term heart rate variability indices of healthy children and children with syncope or pre-syncope symptoms, and came up with sympathetic predominance.[33] However, Akcaboy et al. failed to show that the children with vasovagal syncope had significantly different heart rate variability with respect to healthy controls.[34] On the other hand, two studies reported that heart rate variability indices were augmented by parasympathetic predominance in syncope patients.[32,35] Similarly, tilt test positive children in this study had significantly higher RMSSD which referred to an increased parasympathetic tone of the heart. Such discrepancy might be attributed to the differences in the methodology adopted for the assessment of heart rate variability.

To the best of our knowledge, this is the first study to evaluate the resting electrocardiography findings of the vasovagal syncope patients within a larger context. This context includes the data related with p-wave, QT-interval, T-wave and heart rate variability. The present study indicates that the children presenting with vasovagal syncope have significantly greater p-wave dispersion, greater QT dispersion, prolonged T-peak-to-T-end intervals and increased heart rate variability than the

healthy children. These findings imply that resting electrocardiography can be used to indicate autonomic dysfunction in the heart and, thus, to decrease the need for the head up tilt test in children presenting with vasovagal syncope.

The power of the present study is limited by the small sample size, the lack of frequency domain measures for heart rate variability, the absence of longitudinal data and the controversial interpretation of heart rate variability patterns in patients with vasovagal syncope. Further research is warranted to analyze the significance of resting electrocardiography findings in children with vasovagal syncope and to include heart rate variability time and frequency domain analysis of heart rate variability in this analysis.

Acknowledgement

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

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Role of the Cardiac Magnetic Resonance Imaging in Young Patients with Cardiomyopathies and Atypical Ventricular Arrhythmias

Atipik Ventriküler Aritmili ve Kardiyomyopatili Çocuklarda ve Genç Erişkinlerde Kardiyak Manyetik Rezonans Görüntülemenin Rolü

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Geliş Tarihi/*Received:* 24.04.2017
Kabul Tarihi/*Accepted:* 03.08.2017

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ABSTRACT Objective: Cardiac magnetic resonance imaging (CMRI) is a useful tool in assessing patients with cardiomyopathy. The current study evaluated the CMRI results of children and young adults with suspected cardiomyopathies and atypical ventricular arrhythmias. **Material and Methods:** A total of 55 patients who underwent CMRI for suspected cardiomyopathy between July 2012 and February 2014 were analyzed the results of CMRI. **Results:** The mean patient age was 11.59±5.6 years and the mean patient weight was 42.3±23.1 kg. Echocardiographic findings were as follows: normal in 26 (47%), hypertrophic cardiomyopathy (HCM) in 13 (24%), right ventricular dysfunction and dilatation in 7 (13%), left ventricular noncompaction in 5 (9%); restrictive cardiomyopathy (RCMP) in 1 (2%), and borderline left ventricular dilatation and dysfunction in 3 (5%) patients. In patients with normal echocardiographic findings (n: 26), CMRI was obtained due to atypical ventricular arrhythmias or clinical complaints suggestive of significant arrhythmias. CMRI was unremarkable in 9/26 patients (35%), arrhythmogenic right ventricular dysplasia (ARVD) was found in 6 (23%), left ventricular non-compaction in (LVNC) 1 (4%), borderline septal hypertrophy in 2 (8%), and non-specific abnormal findings in 8 (30%). CMRI results in the rest of the patients were as follows: ARVD or suspected-ARVD in 7, HCM in 13, left ventricular noncompaction in 5, RCMP in 1, and non-specific minor findings in 3. **Conclusion:** Cardiac magnetic resonance imaging is a safe and useful tool and has a critical role in the evaluation of children and young adults with suspected cardiomyopathies and atypical ventricular arrhythmias and is often superior to echocardiography.

Keywords: Cardiomyopathy; children; magnetic resonance imaging; arrhythmias

ÖZET Amaç: Kardiyak Manyetik Rezonans Görüntüleme (CMRI), kardiyomiyopatili hastaların değerlendirilmesinde yararlı bir araçtır. Bu çalışma, kardiyomiyopati ve atipik ventriküler aritmi şüphesi olan çocukların ve genç erişkinlerin CMRI sonuçlarını araştırmıştır. **Gereç ve Yöntemler:** Temmuz 2012 ile Şubat 2014 arasında kardiyomiyopati şüphesiyle CMRI yapılmış 55 hastanın sonuçları analiz edildi. **Bulgular:** Ortalama hasta yaşı 11,59±5,6 yıl ve ortalama hasta ağırlığı 42,3±23,1 kg idi. Ekokardiyografi bulguları şunlardı: 26'sında (% 47) normal, 13'ünde (% 24) hipertrofik kardiyomiyopati (HKM), 7'sinde (%13) sağ ventrikül disfonksiyonu ve dilatasyon, 5'inde (%9) sol ventrikül nonkompaction; 1'inde (%2) restriktif kardiyomiyopati (RCMP) ve 3 (%5) hastada borderline sol ventrikül dilatasyonu ve disfonksiyonu vardı. Normal ekokardiyografi bulguları olan hastalarda (n: 26), atipik ventriküler aritmiler veya belirgin aritmileri düşündüren klinik yakınmalara bağlı olarak CMRI çekildi. Kardiyak manyetik rezonans görüntüleme 9/26 hastada (%35) özelliksiz iken, aritmojenik sağ ventrikül displazisi (ARVD) 6 hastada (%23), sol ventrikül non-kompaksiyon 1 hastada (%4), borderline septal hipertrofi 2 hastada (%8) ve non-spesifik anormal bulgular 8 hastada (%30) saptandı. Hastaların geri kalanında Kardiyak manyetik rezonans görüntüleme sonuçları şunlardı: ARVD veya şüpheli-ARVD; 7, HCM; 13, sol ventrikül non-kompaksiyonu; 5, RCMP; 1 ve non-spesifik minör bulgular; 3 kişide saptandı. **Sonuç:** Kardiyak manyetik rezonans görüntüleme, güvenli ve kullanışlı bir araçtır. Kardiyomiyopati ve atipik ventriküler aritmilerden şüphelenilen çocukların ve genç erişkinlerin değerlendirilmesinde kritik bir role sahip olup genellikle ekokardiyografiden daha üstündür.

Anahtar Kelimeler: Kardiyomyopati; çocuk; magnetik rezonans görüntüleme; aritmiler

Pediatr Heart J 2016;3(3):97-104

Cardiovascular magnetic resonance imaging (CMRI) is an effective noninvasive technique in evaluating the impact of congenital and acquired pediatric heart diseases on the functional and anatomic structures of the heart.[1,2] It has the abilities of presenting 3D data, measuring the ventricular volumes ,ejection fraction and great vessel flow, providing myocardial tissue characterization, and assessing myocardial perfusion.[2] CMRI emerges as an important research development area in the field of cardiomyopathies (CMPs).[3]

Since its emergence, CMR has become a popular imaging technique for the diagnosis and follow-up of CMPs. While echocardiography, being usually the first step in evaluation of the CMPs, involves hidden risks with its limited acoustic window, cardiac MRI, on the other hand, provides replicable, accurate evaluation of myocardial morphology, function, perfusion, and tissue damage in a non-invasive and all-included approach.[4,5] Therefore, CMRI has evolved to be a widespread valuable diagnostic tool for CMP, and to be a new standard in the evaluation of cardiac function.

CMRI is used for various purposes such as: the evaluation of pre/post therapy of hypertrophic and dilated CMPs, in differential diagnosis of RCM and constrictive pericarditis, the evaluation of myocardial damage in acute and chronic CMPs, and the assessment of myocardial involvement in systemic diseases such as amyloidosis and sarcoidosis.[6] In dilated CMP, CMRI can aid in determining the aetiology of the disease including coronary artery anomalies, idiopathic disease, secondary causes such as myocarditis or neuromuscular disorders.[7]

Following the intravenous (IV) administration of gadolinium, late-enhancement MRI is crucial in the diagnosis of myocardial tissue abnormalities[6] such as infarction, fibrosis, and deposition of extracellular materials like amyloid proteins.[8-10] Therefore, CMR is important in the diagnosis, risk assessment, and the management of such conditions.[11]

In this manuscript, performance of CMRI is evaluated in young patients with suspected cardiomyopathy and atypical ventricular arrhythmias. The results are presented in details.

MATERIAL AND METHODS

STUDY POPULATION

A retrospective study was conducted in 55 patients with mean age of 11.59±5.6 years (range 1-30 years) who underwent CMRI for suspected cardiomyopathy at the Pediatric and Genetic Arrhythmia Center at …….. University Hospital between July 2012 and February 2014. Clinical features and the physical examinations were documented from medical records and the echocardiographic images were reviewed from the computer database. Data included age at diagnosis, gender, weight, height, clinical presentation, family history, physical examination findings (cyanosis, heart murmur, arrhythmia, heart failure), electrocardiography (ECG), 24-hour ambulatory ECG recording results, initial and last echocardiogram findings, and CMRI results. The patients with congenital heart diseases were excluded from the study. Findings in echocardiography and CMRI were classified as normal, LV noncompaction, suspected-ARVC, DCM, RCM, HCM, and other non-specific findings. As non-specific findings, LV diastolic dysfunction, LV dilatation, RV diastolic dysfunction, biventricular systolic dysfunction and mild ventricular dilatation were found.

DIAGNOSTIC EQUIPMENT

Rhythm Assessment

A 12-lead ECG (Mortara, VERITAS, Milwaukee, WI, USA) and a 24-hour ambulatory ECG monitoring (Lifecard CF, Spacelife healthcare, Germany) were obtained in all patients. Event recorder (Beam, BHC Health Care, Stolberg, Germany) was used in patients with suspected paroxysmal arrhythmias.

Echocardiographic Imaging

Echocardiographic examination was performed using Vivid-6 ultrasound device with 6S or 3S (Vivid S6, GE Medical Systems, Milwaukee, WI, USA) and 3 MHz transducers equipped with harmonic imaging. Two-dimensional, Doppler (pulse wave, continuous wave and color), and M-mode

echocardiography were performed at resting condition by one of the investigators. Images were taken in the long axis parasternal view, the three short axis views (basal, mid, apical) and the 2, 3 and 4 chamber apical views. All images were obtained according to AHA segmental analysis standards.[12] An ECG was recorded simultaneously with the echocardiogram.

Offline analysis was then performed on the digitally stored images (EchoPac, GE vingmed, Horthen, Norway)

Cardiovascular Magnetic Resonance Imaging

Our CMRI protocol was performed using a 1.5-T scanner (Achieva Nova, Philips Healthcare) with maximum gradient strength of 33 mT/m and a 5-element phased-array coil (SENSE [sensitivity encoding] Cardiac, Philips Healthcare). The study usually includes morphologic fast spin-echo black blood sequences with and without fat suppression, cine single-shot free-precession (steady-state free-precession) sequences, phase-contrast sequences, and late-enhancement 3D T1-weighted fast-field echo inversion recovery sequences obtained 10-15 minutes after the IV administration of 0.2 mmol/kg of a gadolinium-based contrast agent. All images were obtained with breath-holding along the two-chamber plane, the four-chamber long-axis plane, and the short-axis plane.

Functional evaluation is implemented on the cine short-axis images, encompassing the LV and RV from base to apex (8-12 contiguous slices) to obtain a volumetric evaluation using a dedicated workstation (ViewForum, Philips Healthcare).

The pediatric patients, who were not able to hold the breath on MRI device, received general anesthesia under the supervision of an experienced cardiac anesthesist with a good understanding of congenital heart disease.

STATISTICAL ANALYSIS

The statistical evaluation of the data was performed using the SPSS for Windows, Version 17.0 software package (SPSS Inc, Chicago, IL). Categorical variables were presented as absolute and percent frequencies, whereas quantitative variables were summarized as means-standard deviations (SD). The chi-square test was used in comparing the parameters.

RESULTS

The demographics of patients were as follows: 60% male vs 40% female gender, mean age (years) of 11.59±5.6 (range 1-30), mean weight (kg) of 42.3±23.1 (range: 6-88), mean height (cm) of 143.03±32.5 (range:63-183).

The echocardiographic findings of patients were as follows: 47% normal (n:26), 24% HCM (n:13), 13% right ventricular dysfunction and dilatation (suspected ARVC, n:7), 9% LV non-compaction (n:5), 2% RCM (n:1), 5% borderline left ventricular dilatation and dysfunction (n: 3) (Table 1).

CMR findings of patients: 22% normal (n:12), 18% ARVC or suspected-ARVC (n:10), 27% HCM (n:15), 11% LV non-compaction (n:6), 2% RCM (n:1), 20% non-specific minor findings (n:11), (Table 2, Figure 1-4).

TABLE 1: Echocardiographic findings of patients who underwent *CMR imaging (n:55).

Results	n
Normal	26
Hypertrophic cardiomyopathy	13
Right ventricular dysfunction and dilatation (suspected ARVC**)	7
Left ventricular noncompaction	5
Restrictive cardiomyopathy	1
Borderline left ventricular dilatation and dysfunction	3

*CMR: Cardiac magnetic resonance imaging; **ARVC: Arrhythmogenic right ventricular cardiomyopathy.

TABLE 2: CMR* findngs of patients.

Results	n
Normal	12
Arryhythmogenic right ventricular cardiomyopathy or suspected ARVC**	10
Hypertrophic cardiomyopathy (one case with partial noncompaction)	15
Right ventricular dysfunction and dilatation (suspected ARVC**)	7
Left ventricular noncompaction (two cases with partial noncompaction)	6
Restrictive cardiomyopathy	1
Non-specific findings	11

*CMR: Cardiac magnetic resonance imaging; **ARVC: Arrhythmogenic right ventricular cardiomyopathy.

FIGURE 1: The cardiac magnetic resonance ımage showing the left ventricular non- compaction in a 16 years old male patient.

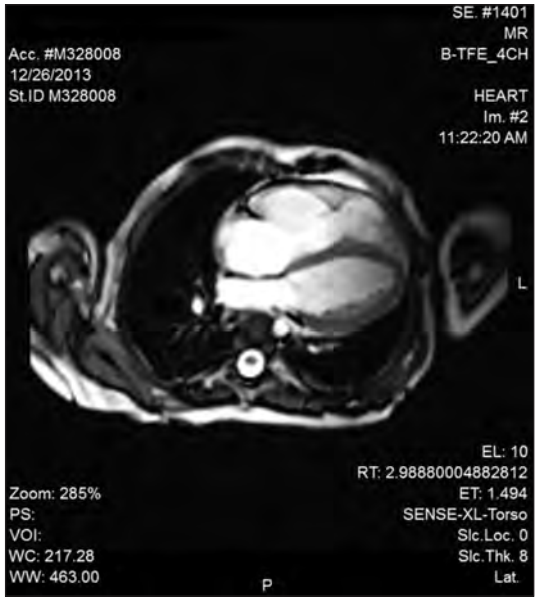

FIGURE 2: The cardiac magnetic resonance ımage of the arrhythmogenic right ventricular dysplasia ın a 4 years old female patient.

The comparison of echocardiography vs. CMRI in terms of the number of diagnoses was summarized in Figure 5. The break down of diagnoses is as following where the first figures are of echocardiography and the second figures are of MRI; Normal findings (26 vs 12), HCM (13 vs 15), ARVC (7 vs 10), LVNC (5 vs 6), RCM (1 vs 1), and non-specific findings (3 vs 11). It is remarkable that number of diagnoses in CMRI is considerably higher than echocardiography (p=0.0001) (Figure 5).

Patients with normal echocardiography findings and those with arrhythmias noted to have following findings: 65% atypical PVCs (n: 17), %19 polymorphic-atypical VT (n: 4), 16% palpitations and syncope (n: 5). The results of CMR in these patients showed: normal findings in 35% (n: 9), ARVC in 23% (n: 6), LV non-compaction in 4% (n: 1), borderline septal hypertrophy in 8% (n: 2), non-specific findings in 30% (n:8).

Among total 15 HCM patients, four indiduals (27%) had myocardial scars and three had arrhythmias (PVCs). While two of 11 patients (18%) without scars showed arrhythmias, ventricular arrhythmias were noted in three of four patients (75%) with myocardial scars (Figure 6). An ICD was implanted in 3 of 4 patients with scars since they also had history of syncope and sudden cardiac deaths in the family.

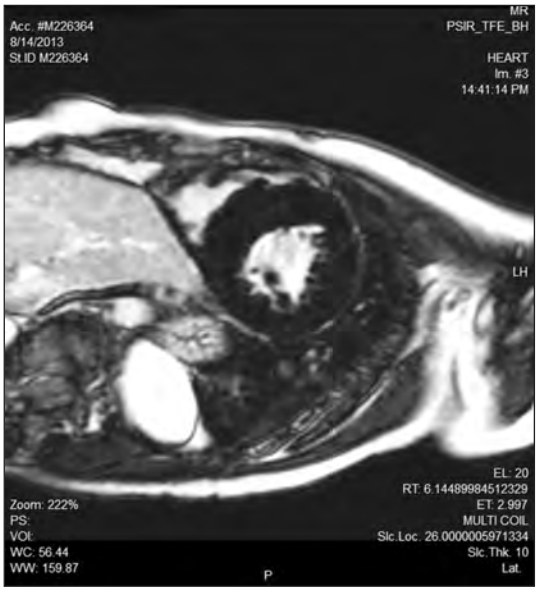

FIGURE 3: The cardiac magnetic resonance ımage of the hypertrophic cardiomyopathy without scar ın a 12 years old male patient.

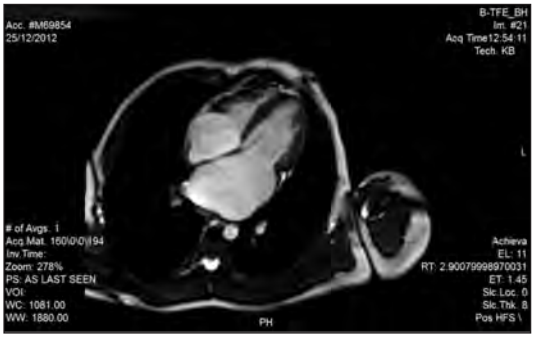

FIGURE 4: The cardiac magnetic resonance ımage showing the restrictive cardiomyopathy ın a 10 years old female patient.

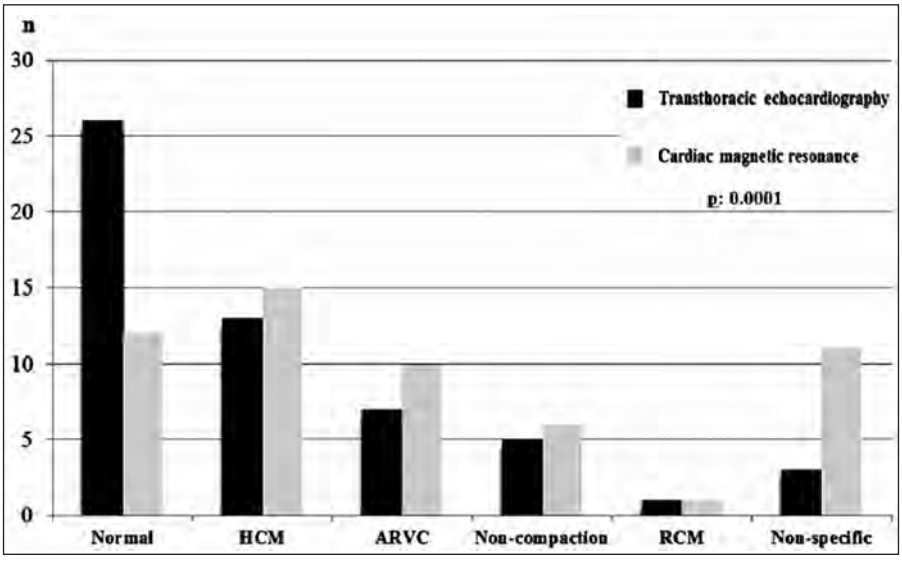

FIGURE 5: Comparison of echocardiography and CMR imaging in terms of diagnostics. Y- axis shows number of patients with diagnosis using echo or CMR.

DISCUSSION

Cardiovascular magnetic resonance imaging is a useful technique allowing the identification of many types of cardiac pathologies from ischemic myocardial disease to myocardial inflammatory disease and different types of cardiomyopathies.[11,13] This study demonstrated significant benefits of CMRI in young patients with cardiomyopathies and atypical ventricular arrhythmias.

An important advantage of CMRI over other imaging techniques is the detection of myocardial scarring and fibrosis with gadolinium contrast enhancement technique. Although transthoracic echocardiography (TTE) is a practical, cheaper and non-invasive method which is accepted generally as "gold standard" for the diagnosis of CMPs, it has some technical constraints; reliable and quantitative determination of LV wall thickness depends on adequate acoustic windows. Therefore, a normal TTE might possibly underdiagnose the disease in the case of suspected cardiomyopathy. Although the efficacy and superiority of CMRI for the diagnosis of CMPs in adult patients have been shown in many studies, pediatric studies are limited in this area.[14-16] Our study was performed at a pediatric center with majority of patients in the childhood and young adults.

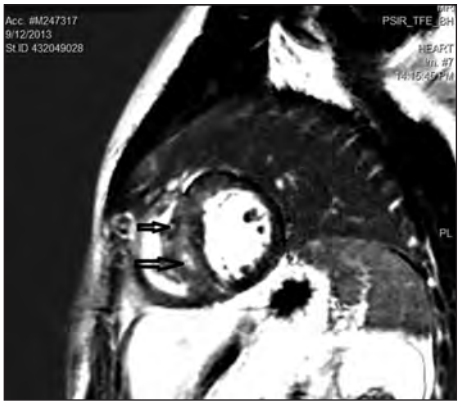

FIGURE 6: A 14 years old male patient with hypertrophic cardiomyopathy. The arrow shows the scar tissue in interventricular septum detected by cardiac magnetic resonance image.

Hypertrophic cardiomyopathy is one of the most common hereditary cardiac diseases and is the most common type of cardiomyopathy. CMRI is an auxiliary and reliable technique in the diagnosis or confirming the diagnosis of HCM. It has been shown to be superior to TTE in atypical segment involvement such as mild hypertrophy or hypertrophy of the apical and lateral walls where diagnosis can be overlooked with TTE. More importantly, massive hypertrophy, which is important for the individual risk grading and the decision of Implantable Cardioverter Defibrillatior (ICD) implantation, can be determined easily and more accurately with CMR rather than TTE [17]

Comparing TEE and CMR in the diagnosis of HCM; Rickers et al. have reported that despite the LV wall thickness in all segments was found to be normal (≤12 mm) with TTE in 3 (6%) of 48 patients including in the study, hypertrophy was detected in the anterolateral LV free wall with CMR imaging.[16] In the same study, it has been shown that despite maximum wall thickness was detected as <30 mm in 42 patients with TTE, more significant wall thickness (≥ 30 mm) has been shown in 6 of these patients with CMRI. They concluded that TTE is inadequate compared to CMRI in determination of the magnitude of the hypertrophy especially more so in the basal anterolateral LV free wall.

The presence of myocardial fibrosis detected by late gadolinium enhancement technique in the CMRI has been shown to play an important role in determining morbidity and mortality in adult patients with HCM. Presence of scar tissue by CMR imaging has been associated with arrhythmias, sudden death, ventricular dilation and heart failure. Therefore besides diagnostic role, CMR imaging also provides important information about the management of the treatment.[17] On the other hand, Campton et al reported that echocardiography identified myocardial scarring with a high sensitivity and negative predictive value when compared to the presence of LGE by CMRI and TTE screening cannot substitute for CMRI.[18] In addition, Slesnick et al. studied 34 pediatric HCM patients with CMR using 1.5 Tesla magnet and observed that six of those patients had evidence of late gadolinium enhancement (LGE).[19] The incidence of ventricular tachycardia (VT) in these patients was higher compared to those without VT (including non-sustained VT) and an ICD was implanted in 5 of them. Furthermore, El Saiedi et al recommended that CMR should be performed for patients with HCM to detect fibrosis and patients at high risk for developing heart failure as there is significant correlation between percentage LGE in children with HCM and parameters of NYHA classification, LVOT pressure gradient and LV myocardial mass.[20] In our study, similarly, the frequency of ventricular arrhythmias was 75% in patients with scar and 18% in those without scar. An ICD was implanted in 3 of 4 patients with scars since they also had history of syncope and sudden cardiac deaths in the family. Finally, an LGE prevelance of 27% in our patient populations is in line with the literature.[2]

Arrhythmogenic right ventricular cardiomyopathy is a genetic cardiomyopathy where right ventricular muscle is replaced by the fibro-fatty tissue. ARVC is characterized by a high incidence of ventricular arrhythmias with an increased risk of sudden cardiac death. Cardiac magnetic resonance imaging is an important method for the diagnosis of ARVC because it allows for three-dimensional imaging of ventricles. Several studies demonstrated that CMR imaging has high sensitivity and specificity in the diagnosis of ARVC.[21] Liu et al. have reported that the diagnosis could be performed with CMR imaging in cases referred to their center with suspected ARVC even in those who did not meet Task Force Criteria. In addition, in some cases who were not diagnosed yet, it has been determined with CMRI that there were structural or functional abnormalities, which would change the management.[22] In our study, while 7 patients were diagnosed with ARVC using TEE, 10 patients were diagnosed with CMRI. In our study, while nonspecific minor findings such as left ventricular dilatation and borderline dysfunction were detected in only 3 patients with TTE, similar findings were found in 11 patients with CMRI.

Left ventricular non-compaction is an uncommon and poorly understood myocardial dysfunction characterized by prominent trabeculations with deep intertrabecular recesses in the left ventricular myocardium. The diagnosis of LVNC is typically made via TTE, however today there are no universally accepted evaluation criteria for TTE results. Cardiac magnetic resonance imaging is increasingly being used in the evaluation of suspected LVNC adult patients.[23] Thuny et al. have performed both TTE and CMR imaging within the same week on 16 adult patients with a diagnosis of LVNC.[24] In their study, it was reported that CMR imaging was superior to TTE in determining the extension of non-compaction. In addition, CMRI was found to be effective in providing complementary morphological information to TTE results.

Cardiovascular magnetic resonance imaging may also help clarify (and even alter) diagnosis by demonstrating the presence of prominent trabeculations consistent with a diagnosis of LV noncompaction in patients initially diagnosed as apical HCM. A previous pediatric study showed that CMRI had a high sensitivity and specificity in differentiating LVNC from the normal heart or other CMPs.[13] In our study, while five patients were diagnosed with LVNC using TTE, CMR was able to diagnose six patients with LVNC.

Ventricular tachyarrhythmia is the most common cause of sudden cardiac death in the developed countries.[25] Under the age of 30, HCM, myocarditis and congenital heart diseases are among the common causes of ventricular tachycardia. The relationship between CMP and ventricular arrhythmias is well known. Electrocardiography was found to be abnormal in most of the patients with CMP. Therefore, normal ECG and 24-hour Holter monitoring may be helpful to exclude the diagnosis of CMP.[24] Our findings demonstrate the importance of CMR in the cases with clinical suspicion of CMP due to atypical ventricular arrhythmias despite a normal TTE assessment.

In conclusion, the current study demonstrates that CMR is superior to TTE in the clinical diagnosis of CMP in the pediatric age group as well as in young adults. In general, although TTE is recommended to be done before CMR for the diagnosis of CMP, CMR is highly useful in the diagnosis of cases with normal TTE findings if clinical suspicion of CMP is high. Further studies are needed to better delineate the indications of CMR in such patients.

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Salmeterol Kullanımına Bağlı Gelişen Nadir Bir Aritmi: Akselere Ventriküler Ritim

A Rare Arrhythmia Due to Salmeterol Treatment: Accelerated Ventricular Rhythm: Case Report

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Geliş Tarihi/*Received:* 25.11.2016
Kabul Tarihi/*Accepted:* 16.01.2017

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ÖZET Uzun etkili beta2-agonistleri kronik obstrüktif akciğer hastalığının tedavisinde sık kullanılan ajanlardandır. Bu ajanlarla olan beta adrenerjik stimulasyon kalp üzerinde aritmilere neden olabilir, kalp hızını arttırabilir. Literatürde bu ajanların kullanımı sonrası aritmi oluşan az sayıda vaka tanımlanmıştır. Bu yazıda salmeterol kullanımı sonrası akselere ventriküler ritim gelişen ondört yaşında bir kız hasta sunulmuştur.

Anahtar Kelimeler: Akselere ventriküler ritim; elektrokardiyografi; salmeterol; tedavi

ABSTRACT Long acting beta agonists are among commonly used agents in the treatment of chronic obstructive pulmonary disease. Beta adrenergic stimulation caused by these agents may lead to arrhythmias and increase in heart beat. In the existing literature, there are few cases defined with arrhythmia after the use of these agents. Here we presented a fourteen-years-old female patient with accelerated ventricular rhythm after the use of salmeterol.

Keywords: Accelerated ventricular rhythm; electrocardiography; salmeterol; therapy

Pediatr Heart J 2016;3(3):105-7

Uzun etkili beta$_2$-agonistleri kronik obstrüktif akciğer hastalığının (KOAH) tedavisinde sık kullanılan ajanlardandır. Bu ajanlarla olan beta adrenerjik stimulasyon kalp üzerinde aritmilere neden olabilir, kalp hızını arttırabilir.[1,2] Literatürde bu ajanların kullanımı sonrası aritmi oluşan az sayıda vaka tanımlanmıştır. Bu yazıda salmeterol kullanımı sonrası akselere ventriküler ritim gelişen bir kız hasta sunulmuştur.

OLGU SUNUMU

Ondört yaşında astım/KOAH nedeniyle takipli kız hastaya salmeterol tedavisi başlandıktan sonra çarpıntı hissetmesi nedeniyle polikliniğimize başvurdu. Hastanın elektrokardiyografik incelemesinde kalp hızı: 73 atım/dk, normal sinüs ritmi, QTc: 413 msn olarak saptandı. ST-T değişikliği yoktu. Kalp hızı değişkenliği incelemesinde zaman ölçütlerinden SDNN: 232 msn, SDANNİ: 242 msn, rMMSD: 75 msn, pNN50: %38, frekans ölçütlerinden Toplam güç: 8920 msn^2, LF: 1498.1 msn^2, HF: 980.5 msn^2, LF/HF: 1.5 olarak

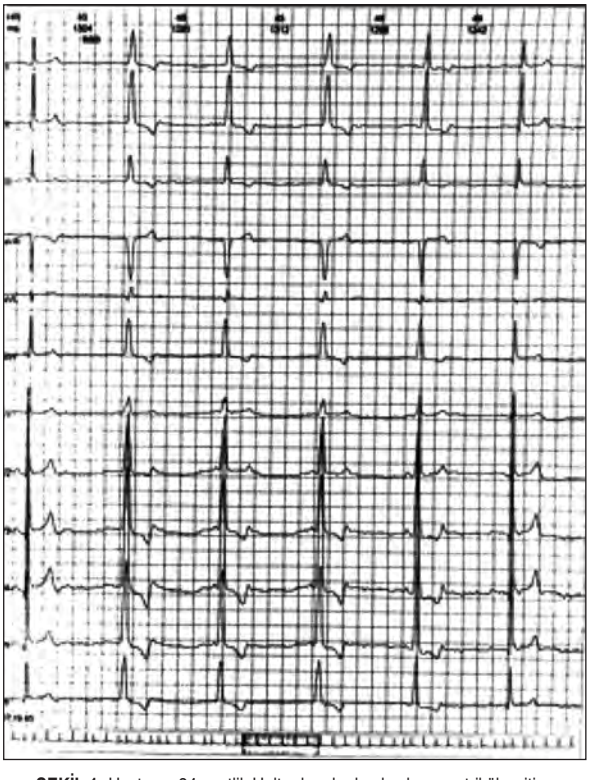
saptandı. Ekokardiyografisinde normal bulgular mevcuttu. Hastanın laboratuar incelemesinde hemogram, biyokimya ve tiroid fonksiyon testleri normal olarak saptandı. 24 saatlik ritim holter kaydının incelemesinde tüm derivasyonlarda, normal sinüs ritminin devamında tüm gün sık sık tekrarlayan, 5-6 atım süren, geniş QRS ve bifazik T dalgası olan, daha sonra tekrar normal sinüs ritmine dönen akselere ventriküler ritim görülmesi üzerine salmeterol tedavisi kesildi (Şekil 1). Tedavi kesildikten sonra şikayeti gerileyen hastanın çekilen kontrol 24 saatlik ritim holteri normal olarak saptandı.

TARTIŞMA

Salmeterol KOAH tedavisinde sık kullanılan uzun etkili beta$_2$-agonist ajanlardır. Literatürde bu ajanlarla aritmi tanımlanan vaka sayısı azdır. Literatürde yapılan birçok çalışmada bu ajanların güvenilir olduğu ve aritmi riskini arttırmadığı belirtilmiştir. Hanrahan JP. ve ark. 1429 hasta ve 5226 holter kaydını incelemişler ve uzun etkili beta agonist verilen grup ile plasebo verilen grup arasında aritmi açısından anlamlı bir fark bulamamışlardır.[2] Ferguson GT. ve ark. Salmeterol verilen grup ile plasebo verilen grup arasında kardiyak yan etkiler yönünden anlamlı fark saptamamışlardır.[3] Kusunoki Y. ve ark. ise bronkodilatör kullanımı ile supraventriküler prematür atım arasında bir ilişki bulmuşlardır. Aynı zamanda KOAH hastalarında ventriküler ekstra atımın da sık olduğunu fakat bunun bronkodilatör kullanımı ile ilişkisi olmadığını belirtmişlerdir.[4] Lee CH. ve ark. 3312 yeni gelişen aritmi vakasını incelemiş ve uzun etkili beta$_2$-agonist kullanımının taşiaritmi gelişimi ile ilişkili olduğunu bulmuşlardır.[5]

Olgumuzda EKG'de, geniş QRS kompleksinin olduğu vurularda (özellikle V2-V4 de daha belirgin olmak üzere) 2., 3. ve 4. vuruda geniş QRS'den önce P dalgası görülmekte olup, intermittan preeksitasyon (WPW) görüntüsü vermektedir ancak geniş QRS in olduğu, ilk vuruda çok temiz ve net bir şekilde geniş QRS ten önce P dalgası olmadığı için EKG intermittan WPW değil, accelere ventriküler ritim olarak yorumlanmıştır, 2., 3. ve 4. vurularda görülen sinüs P dalgalarının tamamen tesadüfi bir zamanlama olduğu düşünülmüştür.

Literatürde salmeterol kullanımına bağlı akselere ventriküler ritim gelişen olgu bulunmamaktadır. Akselere ventriküler ritim genellikle birkaç atım devam eden geniş QRS kompleksli bir ritimdir. Doğumsal kalp hastalıkları, miyokardit, dijital toksisitesi, hipertansiyon, kardiyomiyopati, miyokart infarktüsü ve metabolic anomaliler sonucunda oluşabilir. Salmeterol tedavisi alan olgularda da bu aritminin gelişebileceğine dikkat çekmek için bu olguyu sunduk.



ŞEKİL 1: Hastanın 24 saatlik Holter kaydında akselere ventriküler ritim.

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Successful Treatment of Protein-Losing Enteropathy with Heparin in a Case with Dilated Cardiomyopathy: Case Report

Dilate Kardiyomiyopatili bir Olguda Protein Kaybettiren Enteropatinin Heparin ile Başarılı Tedavisi

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Geliş Tarihi/*Received:* 11.01.2017
Kabul Tarihi/*Accepted:* 08.02.2017

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Bu çalışma 15. Ulusal Pediatrik Kardiyoloji ve Kalp Cerrahisi Kongresi (13-16 Nisan 2016, Antalya)n'de olgu sununmu olarak sunulmuştur.

ABSTRACT Protein-losing enteropathy (PLE) can occur as a complication of the constrictive pericarditis, restrictive cardiomyopathy, congestive heart failure and after Fontan procedure. The diagnosis of PLE should be considered in patients with hypoproteinemia after other causes, such as malnutrition, proteinuria, and impaired protein synthesis, have been excluded. The diagnosis of PLE is most commonly based on the determination of fecal-1 antitrypsin. Treatment strategies, tailored to the severity of the disease, include symptomatic relief with diuretics and supplemental protein, attempts at halting intestinal protein leak using steroids or heparin, and alteration of cardiovascular physiology via fenestration creation, atrial pacing, or heart transplantation. Here, we present successful treatment of PLE with heparin in a case with dilated cardiomyopathy.

Keywords: Heparin; protein-losing enteropathy; dilated cardiomyopathy

ÖZET Protein kaybettirici enteropati (PKE), konstriktif perikardit, restriktif kardiyomiyopati, konjestif kalp yetmezliği ve Fontan ameliyatı sonrası komplikasyon olarak ortaya çıkabilir. Hipoproteinemisi olan hastalarda PKE' nin tanısı, malnütrisyon, proteinüri ve bozulmuş protein sentezi gibi diğer nedenler dışlandıktan sonra düşünülmelidir. PKE tanısı çoğunlukla fekal -1 antitripsin tayinine dayanır. Hastalığın ciddiyetine uygun tedavi stratejileri, diüretik ve ek protein ile semptomatik iyileşmeyi, steroidler veya heparin kullanarak bağırsak protein sızıntısını durdurma girişimlerini ve fenestrasyon oluşturma, atriyal pacing veya kalp transplantasyonu yoluyla kardiyovasküler fizyolojideki değişiklikleri içerir. Burada, dilate kardiyomiyopatili bir olguda PKE'nin heparin ile başarılı bir şekilde tedavisini sunuyoruz.

Anahtar Kelimeler: Heparin; protein kaybettiren enteropati; dilate kardiomiyopati

Pediatr Heart J 2016;3(3):108-10

Protein-losing enteropathy (PLE) is a rare condition characterized by a loss of serum protein into the gastrointestinal tract resulting in hypoproteinemia. It has also been described in the setting of a number of rare cardiac conditions including constrictive pericarditis, restrictive cardiomyopathy and after Fontan surgery.[1] Congestive heart failure may lead to PLE such as dilated cardiomyopathy, to our knowledge there is only one documented case in the literature.[2] We report the second case of PLE of a 17 year-old patient who suffered from dilated cardiomyopathy and he responded to heparin therapy after unsuccessful steroid and dietary therapy.

CASE REPORT

A 17-year-old male patient with a history of non-ischemic dilated cardiomyopathy and end-stage congestive heart failure presented to our institution for a cardiac transplantation evaluation. During follow up implantable cardiac defibrillator was placed him because of unsuccessful medical treatment of sustained ventricular tachycardia.

Six months later the patient was admitted to our hospital with increased peripheral edema and ascites. In this process, levels of albumin and total protein (2,2 and 4.3 g per 100 ml) (respectively) were markedly decreased. Further evaluation of the severe hypoalbuminemia included a 24-h urine protein measurement, serologic testing for celiac disease and intrinsic liver diseases, and abdominal imaging were performed and remained normal. The patient's central venous pressure was increased (mean 14 mmHg). Finally, fecal -1 antitrypsin (FA1AT) was assessed and found to be markedly elevated at a level of 6,2 mg/g (normal, < 2 mg/g) consistent with PLE. Despite modifying his diet (medium-chain fatty acids and high protein), his albumin and protein levels did not change. We administered oral steroid and did not note improvement on protein levels. He required albumin infusion twice weekly while he was on steroid therapy. Treatment with enoxaparin 60 mgr (6000 IU)/day sub-cutaneously was started and he showed dramatic improvement in symptoms, marked elevation in serum albumin levels within a few weeks of beginning therapy. After 2 months of treatment, our patient showed normal albumin levels and FA1AT (< 2 mg/g).

DISCUSSION

The pathophysiology of congestive heart failure associated PLE appears to be secondary to lymphatic obstruction and rupture of lacteals in the gut due to an increase in central venous pressure. Chronic microemboli in the mesenteric circulation have been described as a potential causative factor in the development of PLE. As there is no specific treatment of PLE, its treatment should be directed at the underlying condition. Treatment options may include dietary, pharmacological or surgical intervention, or a combination of these. Dietary manipulation (medium-chain fatty acids and high protein), albumin replacement constitute the principal elements of medical management.[1,2] Steroids can also be helpful, but the beneficial effect tends to be transient.[2,3] Heparin produces benefits independent of its anticoagulant effect. It is theorized that heparin may stabilize the cell–matrix interactions at the capillary endothelium or at the intestinal mucosa to decrease the leakage of protein into the extravascular space or into the intestinal lumen, respectively.[4] To date, few cases that responded to high molecular weight heparin therapy have been reported.[3-5] However only one case reported with protein-losing enteropathy after Fontan palliation which was successfully treated with low-molecular weight heparin (LMWH).[6] The improvement in our patient using low-molecular weight heparin therapy supports the efficacy of this agent in the PLE. Although cardiac transplantation has been used infrequently, it should be considered for patients who have PLE associated with deteriorating hemodynamic variables and poor response to medical treatment. Indeed, longstanding PLE can produce irreversible changes in the enteric lymphatic system.[7] In our case, heparin treatment was started at early stage of illness.

In conclusion, LMWH may be an important treatment in dilated cardiomyopathy who developed a protein-losing enteropathy. We need more experience to understand the pathophysiology of the PLE and more studies to prove the efficacy of this treatment in this illness.

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Right Cervical Aortic Arch Presenting with a Pulsatile Neck Mass in a Child

Boyunda Pulsatil Kitle ile Prezente Olan Sağ Servikal Arkus Aorta

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Geliş Tarihi/*Received:* 09.04.2017
Kabul Tarihi/*Accepted:* 18.08.2017

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KeyWords: *Aort, torasik; boyun*

Anahtar Kelimeler: *Aorta, thoracic; neck*

Pediatr Heart J 2016;3(3):111-2

A 9-year-old Syrian girl was referred to our pediatric cardiology clinic for evaluation of a pulsatile neck mass. She has been examined for Turner Syndrome due to short stature but chromosomal analysis demonstrated a 46 XX normal karyotype. Physical examination revealed a prominent pulsation above the right sternoclavicular area. A systolic murmur, grade 1/6, in the aortic position was heard. Transthoracic echocardiography (TTE) revealed normal ventricular morphology and functions; tricuspid aortic valve with no regurgitations or stenosis. However images of the aortic arch could not be obtained with the usual suprasternal angulation. We were able to demonstrate the aortic arch by angulation of the probe slightly to the right. However branching of the aortic arch could not be demonstrated clearly. As TTE was insufficient for determination of detailed configuration of the aortic arch, cardiac catheterization and angiography were performed. Aortography in left lateral position demonstrated a high-lying aortic arch extending cranially to the toracic aperture (Figure 1A). Normally the aortic arch lies behind the lower part of the manubrium sterni and does not exceed to the sterno-clavicular joint. In this case, a portion of the ascending aorta and the aortic arch exceeded the sterno-clavicular angle and extended to the neck. An angulation which did not create any obstruction in the aortic lumen was seen in the descending aorta. Although the branching pattern of the aorta was usual, all 3 branches were thinner than normal (Figure 1B). Pressure was recorded in 3 major branches of the aorta and no stenosis was detected. Computed tomographic angiography revealed a right cervical aortic arch extending to the neck and turned downward on itself to become the descending aorta which was the cause of angulation in descending aorta detected by angiography (Figure 1C and 1D). In the cardiology and cardiac surgery joint meeting we decided to follow this patient clinically since she was asymptomatic.

In conclusion if the aortic arch could not find in usual echocardiographic position, first of all sided aortic arch must be considered. Also in addition cervical arch must be remembered. A cervical aortic arch has been

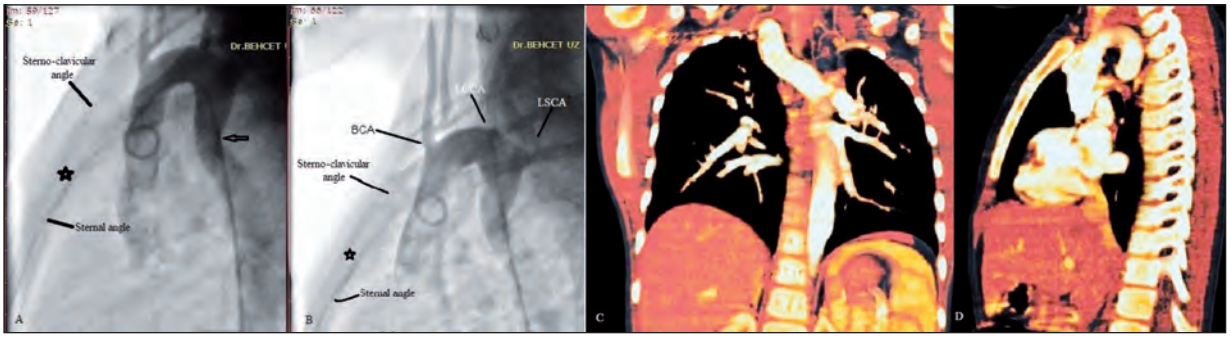

FIGURE 1: A) Aortography in left lateral position demonstrated a high-lying aortic arch extending cranially to the toracic aperture. A portion of the ascending aorta and the aortic arch exceeded the sterno-clavicular angle and extended to the neck. An angulation which was not create any obstruction in the aortic lumen showing by the arrow. **B)** Aortographic view (90º Left lateral 10°cranial) showing a branching of the aortic ach. **C)** A coronal view of the CT angiogram demonstrating a right cervical aortic arch extending to the neck. **D)** A sagittal view of the CT angiogram revealed a high-lying aortic arch extending to the neck.
BCA: Brachiocephalic artery; LCCA: Left common carotid artery; LSCA: Left subclavian artery; star: manubrium sterni.

considered to result from the regression of the fourth aortic arch with persistence of the third aortic arch. Although patients are usually asymptomatic, symptoms related to compression of the trachea and oesophagus such as dysphagia, wheezing,coughing and stridor may be present. Surgical treatment is usually indicated for symptomatic patients.

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YAZARLARA

MAKALE GÖNDERMEK İÇİN

Pediatric Heart Journal'e makale göndermek için; *www.turkpedkar.org.tr* adresindeki "Online Makale" linkini tıklayınız (Yalnızca bu yolla gönderilen makaleler işleme alınmaktadır). Makalelerinizle ilgili tüm işlemleri de bu adresten takip edebilirsiniz.

GENEL BİLGİLER

Pediatric Heart Journal, retrospektif, prospektif veya deneysel araştırmalar, derlemeler, olgu sunumları, editöryal yorum/tartışmalar, editöre mektuplar, tıbbi eğitimler, bilimsel mektuplar, cerrahi teknikler, ayırıcı tanılar, orijinal görüntüler, tanınız nedir? ler, tıbbi kitap değerlendirmeleri, soru-cevaplar ve tıp gündemini belirleyen güncel konuları yayımlayan, ulusal ve uluslararası tüm tıbbi kurum ve personele ulaşmayı hedefleyen bilimsel bir dergidir.

Dergiler; yayımladıkları makalelerde, konu ile ilgili en yüksek etik ve bilimsel standartlarda olması ve ticari kaygılarda olmaması şartını gözetmektedir.

Editörler ve yayınevi, reklam amacı ile verilen ticari ürünlerin özellikleri ve açıklamaları konusunda hiçbir garanti vermemekte ve sorumluluk kabul etmemektedir.

Yayımlanmak için gönderilen makalelerin daha önce başka bir yerde yayımlanmamış veya yayımlanmak üzere gönderilmemiş olması gerekir. Eğer makalede daha önce yayımlanmış; alıntı yazı, tablo, resim vs. mevcut ise makale yazarı, yayın hakkı sahibi ve yazarlarından yazılı izin almak ve bunu makalede belirtmek zorundadır. Bilimsel toplantılarda sunulan özetler, makalede belirtilmesi koşulu ile kabul edilir.

Dergiye gönderilen makale biçimsel esaslara uygun ise, editör ve en az yurt içi-yurt dışı iki danışmanın incelemesinden geçip, gerek görüldüğü takdirde, istenen değişiklikler yazarlarca yapıldıktan sonra yayımlanır.

Makale işleme alındıktan sonra, yayın hakları devir formunda belirtilmiş olan yazar isimleri ve sıralaması esas alınır. Bu aşamadan sonra;

- Makaleye hiçbir aşamada, yayın hakları devir formunda imzası bulunan yazarlar dışında yazar ismi eklenemez ve yazar sırası değiştirilemez.

YAYIN KURALLARI

BİLİMSEL SORUMLULUK

Tüm yazarların gönderilen makalede akademik-bilimsel olarak doğrudan katkısı olmalıdır. Yazar olarak belirlenen isim aşağıdaki özelliklerin tamamına sahip olmalıdır:

- Makaledeki çalışmayı planlamalı veya yapmalı,
- Makaleyi yazmalı veya revize etmeli,
- Son halini kabul etmelidir.

- Makalelerin bilimsel kurallara uygunluğu yazarların sorumluluğundadır.

ETİK SORUMLULUK

- Dergi, "İnsan" öğesinin içinde bulunduğu tüm çalışmalarda Helsinki Deklerasyonu Prensipleri'ne uygunluk *(http://www.wma.net/en/30publications/10policies/b3/ index.html)* ilkesini kabul eder. Bu tip çalışmaların varlığında yazarlar, makalenin GEREÇ VE YÖNTEMLER bölümünde bu prensiplere uygun olarak çalışmayı yaptıklarını, kurumlarının etik kurullarından ve çalışmaya katılmış insanlardan "Bilgilendirilmiş olur" ***(informed consent)*** aldıklarını belirtmek zorundadır.
- Çalışmada "Hayvan" öğesi kullanılmış ise yazarlar, makalenin GEREÇ VE YÖNTEMLER bölümünde *Guide for the Care and Use of Laboratory Animals (www.nap.edu/catalog/5140.html)* prensipleri doğrultusunda çalışmalarında hayvan haklarını koruduklarını ve kurumlarının etik kurullarından onay aldıklarını belirtmek zorundadır.
- Olgu sunumlarında hastanın kimliğinin ortaya çıkmasına bakılmaksızın hastalardan "Bilgilendirilmiş olur" *(informed consent)* alınmalıdır.
- Eğer makalede direkt-indirekt ticari bağlantı veya çalışma için maddi destek veren kurum mevcut ise yazarlar; kullanılan ticari ürün, ilaç, firma... ile ticari hiçbir ilişkisinin olmadığını ve varsa nasıl bir ilişkisinin olduğunu (konsültan, diğer anlaşmalar), editöre sunum sayfasında bildirmek zorundadır.
- Makalede "Etik Kurul Onayı" alınması gerekli ise; alınan belge online olarak, *www.turkiyeklinikleri.com* adresinde yer alan "Makale Gönderim" linkindeki bölümden, makale ile birlikte gönderilmelidir.
- Makalelerin etik kurallara uygunluğu yazarların sorumluluğundadır.
- Makalenin değerlendirilmesi aşamasında, yayın kurulunun gerek görmesi halinde, makale ile ilgili araştırma verilerinin ve/veya etik kurul onayı belgesinin sunulması yazarlardan talep edilebilir.

BİYOİSTATİSTİKSEL DEĞERLENDİRME

Tüm retrospektif, prospektif ve deneysel araştırma makaleleri biyoistatistiksel olarak değerlendirilmeli ve uygun plan, analiz ve raporlama ile belirtilmelidir.

Makalelerde p değerleri açık olarak verilmelidir (p= 0.025; p= 0.524 gibi).

Araştırma makaleleri dergiye gönderilmeden önce, biyoistatistik uzmanı tarafından değerlendirilmeli ve uzmanın ismi yazarlar arasında yer almalıdır.

Biyomedikal dergilere gönderilen yazıların biyoistatistiki uygunluğunun kontrolü için ek bilgi *www.icmje.org* adresinden temin edilebilir.

- Makalelerin biyoistatistiksel kurallara uygunluğu yazarların sorumluluğundadır.

YAZIM DİLİ YÖNÜNDEN DEĞERLENDİRME

Derginin yayın dili Türkçe ve İngilizce'dir.

Türkçe makalelerde Türk Dil Kurumu'nun Türkçe sözlüğü veya *www.tdk.gov.tr* adresi esas alınmalıdır.

İngilizce makaleler ve İngilizce özetler, dergiye gönderilmeden önce dil uzmanı tarafından değerlendirilmeli ve uzman onayı editöre sunum sayfasında özellikle belirtilmelidir. Makaleyi, İngilizce yönünden değerlendiren, yazarlardan biri değil ise bu kişinin ismi makalenin sonunda bulunan TEŞEKKÜR (Acknowledgement) bölümünde belirtilmelidir.

Ayrıca gönderilmiş olan makalelerdeki yazım ve dilbilgisi hataları, makalenin içeriğine dokunmadan, redaksiyon komitemiz tarafından düzeltilmektedir.

- Makalelerin yazım ve dil bilgisi kurallarına uygunluğu yazarların sorumluluğundadır.

YAYIN HAKKI

1976 Copyright Act'e göre, yayımlanmak üzere kabul edilen yazıların her türlü yayın hakkı dergiyi yayımlayan kuruma aittir. Yazılardaki düşünce ve öneriler tümüyle yazarların sorumluluğundadır. Makale yazarlarına, yazıları karşılığında herhangi bir ücret ödenmez.

Yazarlar, *http://turkishclinics.com/Log/phj* internet adresinden ulaşacakları "Yayın Hakları Devir Formu"nu doldurup, online olarak, ***www.turkpedkar.org.tr*** adresinde yer alan "Online Makale" linkindeki bölümden, makale ile birlikte göndermelidirler.

YAZI ÇEŞİTLERİ

Dergiye yayımlanmak üzere gönderilecek yazı çeşitleri şu şekildedir;

Orijinal Araştırma: Kliniklerde yapılan prospektif-retrospektif ve her türlü deneysel çalışmalar yayımlanabilmektedir.

Yapısı:

- Özet (Ortalama 200-250 kelime; amaç, gereç ve yöntemler, bulgular ve sonuç bölümlerinden oluşan, Türkçe ve İngilizce)
- Giriş
- Gereç ve Yöntemler
- Bulgular
- Tartışma
- Sonuç
- Teşekkür
- Kaynaklar

Derleme: Doğrudan veya davet edilen yazarlar tarafından hazırlanır. Tıbbi özellik gösteren her türlü konu için son tıp literatürünü de içine alacak şekilde hazırlanabilir. Yazarın o konu ile ilgili basılmış yayınlarının olması özellikle tercih nedenidir.

Yapısı:

- Özet (Ortalama 200-250 kelime, bölümsüz, Türkçe ve İngilizce)
- Konu ile ilgili başlıklar
- Kaynaklar

Olgu Sunumu: Nadir görülen, tanı ve tedavide farklılık gösteren makalelerdir. Yeterli sayıda fotoğraflarla ve şemalarla desteklenmiş olmalıdır.

Yapısı:

- Özet (ortalama 100-150 kelime; bölümsüz; Türkçe ve İngilizce)
- Giriş
- Olgu Sunumu
- Tartışma
- Kaynaklar

Editöryel Yorum/Tartışma: Yayımlanan orijinal araştırma makalelerinın, araştırmanın yazarları dışındaki, o konunun uzmanı tarafından değerlendirilmesidir. İlgili makalenin sonunda yayımlanır.

Editöre Mektup: Son bir yıl içinde dergide yayımlanan makaleler ile ilgili okuyucuların değişik görüş, tecrübe ve sorularını içeren en fazla 500 kelimelik yazılardır.

Yapısı:

- Başlık ve özet bölümleri yoktur.
- Kaynak sayısı 5 ile sınırlıdır.
- Hangi makaleye (sayı, tarih verilerek) ithaf olunduğu belirtilmeli ve sonunda yazarın ismi, kurumu, adresi bulunmalıdır. Mektuba cevap, editör veya makalenin yazar(lar)ı tarafından, yine dergide yayımlanarak verilir.

Bilimsel Mektup: Genel tıbbi konularda okuyucuyu bilgilendiren, basılmış bilimsel makalelere de atıfta bulunarak o konuyu tartışan makalelerdir.

Yapısı:

- Özet (ortalama 100-150 kelime; bölümsüz, Türkçe ve İngilizce)
- Konu ile ilgili başlıklar
- Kaynaklar

Cerrahi Teknik: Ameliyat tekniklerinin ayrıntılı işlendiği makalelerdir.

Yapısı:

- Özet (ortalama 100-150 kelime; bölümsüz, Türkçe ve İngilizce)
- Cerrahi teknik
- Kaynaklar

Ayırıcı Tanı: Güncel değeri olan olgu sunumlarıdır. Benzer hastalıklarla ilgili yorumu içermektedir.

Yapısı:

- Özet (ortalama 100-150 kelime; bölümsüz, Türkçe ve İngilizce)
- Konu ile ilgili başlıklar
- Kaynaklar (3-5 arası)

Orijinal Görüntüler: Literatürde nadir gözlenen açıklamalı tıbbi resim ve fotoğraflardır.

Yapısı:

- 300 kelimelik metin, orjinal resimler, kaynaklar

Tanınız Nedir?: Nadir görülen, tanı ve tedavide farklılık gösteren hastalıklar hakkında, soru-cevap şeklinde hazırlanmış makalelerdir.

Yapısı:

- Konu ile ilgili başlıklar
- Kaynaklar (3-5 arası)

Tıbbi Kitap Değerlendirmeleri: Güncel değeri olan ulusal veya uluslararası kabul görmüş kitapların değerlendirmeleridir.

Soru Cevaplar: Tıbbi konularda bilimsel eğitici-öğreticiliği olan soru ve cevaplar.

YAZIM KURALLARI

Dergiye yayımlanması için gönderilen makalelerde aşağıdaki biçimsel esaslara uyulmalıdır:

- Makale, PC uyumlu bilgisayarlarda **Microsoft Word programı** ile yazılmalıdır.

KISALTMALAR: Kelimenin ilk geçtiği yerde parantez içinde verilir ve tüm metin boyunca o kısaltma kullanılır. Uluslararası kullanılan kısaltmalar için "Bilimsel Yazım Kuralları"[1] kaynağına başvurulabilir.

ŞEKİL, RESİM, TABLO VE GRAFİKLER:

-Şekil, resim, tablo ve grafiklerin metin içinde geçtiği yerler ilgili cümlenin sonunda belirtilmelidir. Şekil, resim, tablo ve grafiklerin açıklamaları makale sonuna eklenmelidir.

-Şekil, resim/fotoğraflar ayrı birer .jpg veya .gif dosyası olarak (pixel boyutu yaklaşık 500x400, 8 cm eninde ve 300 çözünürlükte taranarak), sisteme eklenmelidir.

- Kullanılan kısaltmalar şekil, resim, tablo ve grafiklerin altındaki açıklamada belirtilmelidir.
- Daha önce basılmış şekil, resim, tablo ve grafik kullanılmış ise yazılı izin alınmalıdır ve bu izin açıklama olarak şekil, resim, tablo ve grafik açıklamasında belirtilmelidir.
- Resimler/fotoğraflar renkli, ayrıntıları görülecek derecede kontrast ve net olmalıdır.

EDİTÖRE SUNUM SAYFASI: Gönderilen makalenin kategorisi, daha önce başka bir dergiye gönderilmemiş olduğu, varsa çalışmayı maddi olarak destekleyen kişi ve kuruluşlar ve varsa bu kuruluşların yazarlarla olan ilişkileri belirtilmelidir.

KAPAK SAYFASI: Makalenin başlığı (Türkçe ve İngilizce), tüm yazarların ad-soyadları, akademik ünvanları, kurumları, iş telefonu-GSM, e-posta ve yazışma adresleri belirtilmelidir. Makale daha önce tebliğ olarak sunulmuş ise tebliğ yeri ve tarihi belirtilmelidir.

ÖZETLER: YAZI ÇEŞİTLERİ bölümünde belirtilen şekilde hazırlanarak, makale metni içerisine yerleştirilmelidir.

ANAHTAR KELİMELER:

- En az 2 adet, Türkçe ve İngilizce yazılmalıdır.
- Kelimeler birbirlerinden noktalı virgül (;) ile ayrılmalıdır.
- İngilizce anahtar kelimeler "Medical Subject Headings (MESH)"e uygun olarak verilmelidir (Bkz: *www.nlm.nih.gov/mesh/MBrowser.html*).
- Türkçe anahtar kelimeler Türkiye Bilim Terimleri (TBT)'ne uygun olarak verilmelidir (Bkz: *www.bilimterimleri.com*).

TEŞEKKÜR: Eğer çıkar çatışması, finansal destek, bağış ve diğer bütün editöryal (istatistiksel analiz, İngilizce/Türkçe değerlendirme) ve/veya teknik yardım varsa, metnin sonunda sunulmalıdır.

KAYNAKLAR: İngilizce yazılmalıdır. Orijinal basımı Türkçe olan kaynakların başlığı [ ] içinde ve İngilizce olarak yazılmalıdır. Kaynaklar makalede geliş sırasına göre yazılmalı ve metinde cümle sonunda noktalama işaretlerinden hemen sonra "Üst Simge" olarak belirtilmelidir. Makalede bulunan yazar sayısı 6 veya daha az ise tüm yazarlar belirtilmeli, 7 veya daha fazla ise ilk 6 isim yazılıp "et al." eklenmelidir. Kaynak yazımı için kullanılan format *Index Medicus*'ta belirtilen şekilde olmalıdır (Bkz: *www.icmje.org*). **Kongre bildirileri, kişisel deneyimler, basılmamış yayınlar, tezler ve internet adresleri kaynak olarak gösterilemez.**

Kaynakların yazımı için örnekler ***(Noktalama işaretlerine lütfen dikkat ediniz):***

Makale için; Yazar(lar)ın soyad(lar)ı ve isim(ler)inin başharf(ler)i, makale ismi, dergi ismi, yıl, cilt, sayı, sayfa no'su belirtilmelidir. Örnek:

Arıcı C, Oğuz V**.** [Surgical Treatment Options According to Inferior Oblique Hyperfunction in Superior Oblique Palsy]**.** Turkiye Klinikleri J Med Sci 2011;31(5)**:**1160-6**.**

Kitap için; Yazar(lar)ın soyad(lar)ı ve isim(ler)inin başharf(ler)i, bölüm başlığı, editörün(lerin) ismi, kitap ismi, kaçıncı baskı olduğu, şehir, yayınevi, yıl ve sayfalar belirtilmelidir. Örnek:

Yabancı dilde yayımlanan kitaplar için;

Underwood LE, Van Wyk JJ**.** Normal and aberrant growth. In: Wilson JD, Foster DW, eds. Wiliams' Textbook of Endocrinology**.** 1st ed**.** Philadelphia**:** WB Saunders**;** 1992**.** p.1079-138**.**

Türkçe kitaplar için;

Tür A**.** [Emergency airway management and endotracheal intubation]**.** Şahinoğlu AH, editör**.** Yoğun Bakım Sorunları ve Tedavileri**.** 2. Baskı**.** Ankara: Türkiye Klinikleri**;** 2003**.** p.9-16**.**

Yazar ve editörün aynı olduğu kitaplar için; Yazar(lar)ın/editörün soyad(lar)ı ve isim(ler)inin başharf(ler)i, bölüm başlığı, kitap ismi, kaçıncı baskı olduğu, şehir, yayınevi, yıl ve sayfalar belirtilmelidir. Örnek:

Yabancı dilde yayımlanan kitaplar için;

Solcia E, Capella C, Kloppel G**.** Tumors of the exocrine pancreas**.** Tumors of the Pancreas**.** 2nd ed**.** Washington**:** Armed Forces Institute of Pathology**;** 1997**.** p.145-210**.**

Türkçe kitaplar için;

Eken A**.** [Cosmeceutical ingredients: drugs to cosmetics products]**.** Kozmesötik Etken Maddeler**.** 1. Baskı**.** Ankara**:** Türkiye Klinikleri**;** 2006**.** p.1-7**.**

Sadece on-line yayınlar için;

DOI tek kabul edilebilir on-line referanstır.

İletişim:

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Hoşdere Caddesi No: 180/4 Çankaya, Ankara/Türkiye
Tel : (0312) 212 02 00
Faks : (0312) 212 02 00
e-posta : turkpedkar@gmail.com
web : www.turkpedkar.org.tr

[1] *Scientific Style and Format: The CBE Manuel for Authors, Editors, and Publishers, 6th ed. New York: Cambridge University Press, 1994.*

PEDIATRIC HEART JOURNAL

INFORMATION FOR AUTHORS

SUBMITTING AN ARTICLE

In order to submit an article for the Pediatric Heart Journal, you click "Online Article" link in *www.turkpedkar.org.tr* address (Only internet submitting will be considered). You also may follow up all the procedures related with your articles from this web site.

GENERAL INFORMATION

Pediatric Heart Journal is a scientific journal that aims to reach all national/international medical institutions&personnel and to publish retrospective, prospective or experimental researchs, reviews, case reports, editorial comment/discussions, letters to the editor, medical education, scientific letters, surgical techniques, distinctive diagnosis, original images, "what is your diagnosis?", medical book reviews, questions-answers and recent issues that determine medical agenda.

The *Journals* commits to rigorous peer review, and stipulates freedom from commercial influence, and promotion of the highest ethical and scientific standards in published articles.

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The writers' names and placement that are indicated in the copyright transfer form will be considered after the process of the manuscript is started. After this stage;

- A writer name cannot be added to the manuscript in any time except the writers who signed the copyright transfer form, and the order of the writers' names cannot be changed.

EDITORIAL POLICIES

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All authors should have contributed to the article directly either academically or scientifically. All persons designated as authors should meet all of the following criterias:

- Planned or performed the study,
- Wrote the paper or reviewed the versions,
- Approved the final version.

- It is the authors' responsibility to prepare a manuscript that meets scientific criterias.

ETHICAL RESPONSIBILITY

- *The Journal* adheres to the principles set forth in the Helsinki Declaration *(http://www.wma.net/en/30publications/10policies/b3/ index.html)* and holds that all reported research involving "Human beings" conducted in accordance with such principles. Reports describing data obtained from research conducted in human participants must contain a statement in the MATERIAL AND METHODS section indicating approval by the institutional ethical review board and affirmation that INFORMED CONSENT was obtained from each participant.
- All papers reporting experiments using animals must include a statement in the MATERIAL AND METHODS section giving assurance that all animals have received humane care in compliance with the *Guide for the Care and Use of Laboratory Animals (www.nap.edu/catalog/5140.html)* and indicating approval by the institutional ethical review board.
- Case reports should be accompanied by INFORMED CONSENT whether the identity of the patient is disclosed or not.
- If the proposed publication concerns any commercial product, the author must include in the cover letter a statement indicating that the author(s) has (have) no financial or other interest in the product or explaining the nature of any relation (including consultancies) between the author(s) and the manufacturer or distributor of the product.
- If "Ethics Commitee Approval" must be obtained for an article, document obtained from ethics commitee should be sent to "Online Article Processing" link on *www.turkiyeklinikleri.com*
- It is the authors' responsibility to prepare a manuscript that meets ethical criteria.
- During the evaluation of the manuscript, the research data and/or ethics committee approval form can be requested from the authors if it's required by the editorial board.

STATISTICAL EVALUATION

All retrospective, prospective and experimental research articles must be evaluated in terms of biostatics and it must be stated together with appropriate plan, analysis and report.

p values must be given clearly in the manuscripts (e.g. p= 0.025; p= 0.524).

Research articles must be evaluated by a biostatistician prior to submission and the name of the biostatistician should be placed among the authors' names.

Additional information in order to control the biostatistical convenience of the papers that are submitted to biomedical journals can be obtained from the web page *www.icmje.org.*

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The official languages of the *Journals* are Turkish and English.

Manuscripts and abstracts in English must be checked for language by an expert and this should be mentioned particularly in the cover letter. The expert responsible for English Language editing should be listed in the ACKNOWLEDGEMENTS if he/she is not in the author list.

All spelling and grammar mistakes in the submitted articles, are corrected by our redaction commitee without changing the data presented.

- It is the authors' responsibility to prepare a manuscript that meets spelling and grammar rules.

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All manuscripts submitted must be accompanied by the "Copyright Transfer and Author Declaration Statement form" that is available in *http://turkishclinics.com/Log/phj.*

CATEGORIES OF ARTICLES

The *Journal* publishes the following types of articles:

Original Research Articles: Original prospective or retrospective studies of basic or clinical investigations in areas relevant to medicine.

Content:

- Abstract (200-250 words; the structured abstract contain the following sections: objective, material and methods, results, conclusion; English)
- Introduction
- Material and Methods
- Results
- Discussion
- Conclusion
- Acknowledgements
- References

Review Articles: The authors may be invited to write or may submit a review article. Reviews including the latest medical literature may be prepared on all medical topics. Authors who have published materials on the topic are preferred.

Content:

- Abstract (200-250 words; without structural divisions; English)
- Titles on related topics
- References

Case Reports: Brief descriptions of a previously undocumented disease process, a unique unreported manifestation or treatment of a known disease process, or unique unreported complications of treatment regimens. They should include an adequate number of photos and figures.

Content:

- Abstract (average 100-150 words; without structural divisions; English)
- Introduction
- Case report
- Discussion
- References

Editorial Commentary/Discussion: Evaluation of the original research article is done by the specialists of the field (except the authors of the research article) and it is published at the end of the related article.

Letters to the Editor: These are the letters that include different views, experiments and questions of the readers about the manuscripts that were published in this journal in the recent year and should be no more that 500 words.

Content:

- There's no title and abstract.
- The number of references should not exceed 5.
- Submitted letters should include a note indicating the attribution to an article (with the number and date) and the name, affiliation and address of the author(s) at the end.
- The answer to the letter is given by the editor or the author(s) of the manuscript and is published in the journal.

Scientific Letter: Presentations of the current cardiovascular topics with comments on published articles in related fields.

Content:

- Abstract (100-150 words; without structural division; English)
- Titles on related topics
- References

Surgical Technique: These are articles in which surgical techniques are explained.

Content:

- Abstracts (100-150 words; without structural division; English)
- Surgical techniques
- References

Differential Diagnosis: These are case reports which have topical importance. They include commentaries related with similar diseases.

Content:

- Abstract (100-150 words; without structural divisions; English)
- Titles related with subject
- References

Original Images: Self-explanotory figures or pictures on rare issues in literature.

Content:

- Text with 300 words, original images, references

What is Your Diagnosis?: These articles are related with diseases that are seen rarely and show differences in diagnosis and treatment, and they are prepared as questions-answers.

Content:

- Titles related with subject
- References (between 3 and 5)

Medical Book Reviews: Reviews and comments on current national and international medical books.

Questions and Answers: Scientific educational questions and answers on medical topics.

MANUSCRIPT PREPARATION

Authors are encouraged to follow the following principles before submitting their material.

-The article should be written in IBM compatible computers with Microsoft Word.

ABBREVATIONS: Abbrevations that are used should be defined in parenthesis where the full word is first mentioned. For commonly accepted abbreviations and usage, please refer to *Scientific Style and Format.*[1]

[1] *Scientific Style and Format: The CBE Manuel for Authors, Editors, and Publishers, 6th ed. New York: Cambridge University Press, 1994.*

FIGURES, PICTURES, TABLE S AND GRAPHICS:

-All figures, pictures, tables and graphics should be cited at the end of the relevant sentence. Explanations about figures, pictures, tables and graphics must be placed at the end of the article.

-Figures, pictures/photographs must be added to the system as separate .jpg or .gif files (approximately 500x400 pixels, 8 cm in width and scanned at 300 resolution).

- All abbrevations used, must be listed in explanation which will be placed at the bottom of each figure, picture, table and graphic.
- For figures, pictures, tables and graphics to be reproduced relevant permissions need to be provided. This permission must be mentioned in the explanation.
- Pictures/photographs must be in color, clear and with appropriate contrast to separate details.

COVER LETTER: Cover letter should include statements about manuscript category designation, single-journal submission affirmation, conflict of interest statement, sources of outside funding, equipments (if so), approval for language for articles in English and approval for statistical analysis for original research articles.

TITLE PAGE: A concise, informative title (English), should be provided. All authors should be listed with academic degrees, affiliations, addresses, office and mobile telephone and fax numbers, and e-mail and postal addresses. If the study was presented in a congress, the author(s) should identify the date/place of the congress of the study presented.

ABSTRACT: The abstracts should be prepared in accordance with the instructions in the "Categories of Articles" and placed in the article file.

KEY WORDS:

-They should be minimally two, and should written English.

-The words should be separated by semicolon (;), from each other.

- Key words should be appropriate to "Medical Subject Headings (MESH)" (Look: www.nlm.nih.gov/mesh/MBrowser.html).

ACKNOWLEDGEMENTS: Conflict of interest, financial support, grants, and all other editorial (statistical analysis, language editing) and/or technical asistance if present, must be presented at the end of the text.

REFERENCES: References in the text should be numbered as superscript numbers and listed serially according to the order of mentioning on a separate page, double-spaced, at the end of the paper in numerical order. All authors should be listed if six or fewer, otherwise list the first six and add the *et al.* Journal abbreviations should conform to the style used in the *Cumulated Index Medicus* (please look at: *www.icmje.org*). Declarations, personal experiments, unpublished papers, thesis can not be given as reference.

Examples for writing references ***(please give attention to punctuation):***

Format for journal articles; initials of author's names and surnames, titles of article, journal name, date, volume, number, and inclusive pages, must be indicated. Example:

Stephane A**.** Management of Congenital Cholesteatoma with Otoendoscopic Surgery: Case Report**.** Turkiye Klinikleri J Med Sci 2010**;**30(2)**:**803-7**.**

Format for books; initials of author's names and surnames, chapter title, editor's name, book title, edition, city, publisher, date and pages. Example:

Underwood LE, Van Wyk JJ**.** Normal and aberrant growth**.** In: Wilson JD, Foster DW, eds**.** Wiliams' Textbook of Endocrinology**.** 1st ed**.** Philadelphia**:** WB Saunders**;** 1992**.** p.1079-138**.**

Format for books of which the editor and author are the same person; initials of author(s)' editor(s)' names and surnames chapter title, book title, edition, city, publisher, date and pages. Example:

Solcia E, Capella C, Kloppel G**.** Tumors of the exocrine pancreas**.** Tumors of the Pancreas**.** 2nd ed**.** Washington**:** Armed Forces Institute of Pathology**;** 1997**.** p.145-210**.**

Format for on-line-only publications; DOI is the only acceptable on-line reference.

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