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ARTIKEL PENELITIAN

REVISITING THE ROLE OF ELECTROCARDIOGRAPHY FOR SCREENING OF CONGENITAL HEART DISEASE IN YOUNG ADULTS

PERAN ELEKTROKARDIOGRAFI DALAM PENAPISAN PENYAKIT JANTUNG BAWAAN PADA DEWASA MUDA

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ABSTRAK
Pendahuluan: *Penyakit jantung bawaan (PJB) merupakan cacat lahir yang paling sering ditemukan. Banyak kasus PJB tidak terdeteksi hingga terjadi komplikasi, umumnya pada usia dewasa muda. Pemeriksaan ekokardiografi transtorakal dan Doppler merupakan modalitas pilihan untuk mendeteksi PJB, tetapi modalitas ini terbatas di Indonesia. Elektrokardiografi (EKG) merupakan modalitas yang banyak digunakan dan berbiaya rendah. Penelitian ini bertujuan untuk mengetahui peranan EKG sebagai modalitas penapisan PJB.*
Metode: *Studi potong lintang dilakukan di Fakultas Kedokteran dan Ilmu Kesehatan Unika Atma Jaya pada Agustus-November 2019. Subjek adalah mahasiswa berusia 18 tahun ke atas. Subjek dengan riwayat PJB sebelumnya dieksklusi dari penelitian. Data dikumpulkan berdasarkan anamnesis, pemeriksaan antropometri, pengukuran tekanan darah, dan EKG 12 sadapan. Hasil EKG diinterpretasikan oleh dua kardiolog secara independen untuk mengurangi bias.*
Hasil: *Total subjek berjumlah 193 orang, terdiri dari 78 pria dan 115 wanita. Rerata usia subjek 19,22 ± 1,85 tahun. Didapatkan hasil berupa 57 orang (29,5%) dengan abnormalitas EKG berupa tanda chrocetage 15 orang, deviasi aksis ke kanan 13 orang, deviasi aksis ke kiri tiga orang, hipertrofi ventrikel kanan dan kiri masing-masing sembilan orang, blok cabang berkas kiri enam orang, dan defektif gelombang T sebanyak dua orang. Evaluasi lanjutan dengan ekokardiografi dilakukan pada 20 partisipan, ditemukan satu subjek memiliki prolaps katup mitral.*
Simpulan: *EKG dapat mendeteksi karakteristik yang sugestif terhadap PJB, terapi pemeriksaan EKG tunggal tidak cukup untuk menegakkan adanya abnormalitas struktural jantung.*
Kata Kunci: *penyakit jantung bawaan, ekokardiografi, elektrokardiografi, penapisan*

ABSTRACT
Introduction: Congenital heart disease (CHD) is the most prevalent congenital disability found in newborns. Many cases remain unknown until complications occur, usually in young adults. Transthoracic and Doppler echocardiography are modalities of choice for CHD detection, but these are limited in Indonesia. Electrocardiography (ECG) is a widely available and low-cost test. This study investigated the role of ECG as a screening modality for CHD.
Methods: A cross-sectional study was held at Atma Jaya School of Medicine and Health Science from August to November 2019. Participants were students aged 18 years old or more. Exclusion criteria was previously detected for CHD. Data were collected through history taking, anthropometrics, blood pressure measurement, and 12 leads ECG. ECG results were interpreted independently by two cardiologists to minimize observer bias.
Results: A total of 193 students, 78 male and 115 female, participated. The mean age was 19.22±1.85 years. ECG abnormalities were discovered in 57 (29.5%) participants:15 with crochetage sign, 13 right axis deviation, three left axis deviation, nine right ventricle hypertrophy, and nine left ventricle hypertrophy, six left bundle branch block, and two defective T wave. Further evaluation was done with echocardiography in 20 participants, which resulted in one participant having mitral valve prolapsed (MVP).

Conclusion: ECG could detect the characteristic patterns suggestive of CHD, but ECG alone is insufficient to confirm cardiac structural abnormalities.
Key Words: Congenital heart disease, echocardiography, electrocardiography, screening

INTRODUCTION

Congenital heart disease (CHD) is the most common congenital disease found in newborns. Congenital heart disease affects approximately 6-8 per 1000 live births worldwide and becomes the leading cause of mortality from birth defects.[1,2] The incidence of CHD is increasing over time, especially in mild types.[3] In Asian and Black ethnic groups, the incidence of CHD is significantly higher.[2] About 40.000 children per year are born with CHD in the United States.[3] We found there is no national report on CHD in Indonesia. Hospital-based study in Sardjito hospital, a tertiary referral hospital in Yogyakarta and Central Java, the incidence of CHD in 2014 was 134/10.000 person-years newly diagnosed by echocardiography. These consisted of 22% adults with mean age of 32.7 ± 13.5 years old.[4]

Although all newborns underwent congenital defect screening, delayed diagnosis still occurs in over 10% of critical congenital heart disease cases. The leading causes are associated with delivery outside a tertiary hospital and isolated CHD.[5] In mild cases, it is not uncommon to remain undiagnosed until a complication occurs.

One in 150 adults is expected to have some form of CHD. The severity of lesions varies from mild lesions without dysfunction to severe lesions such as hypoplastic left heart syndrome. Atrial septal defect (ASD) and Tetralogy of Fallot (TOF) are the most common CHD in adults.[6] From 2015 to 2016, the death rate from congenital cardiovascular defects increased from 0.30 to 0.60 per 100.000 people in adults 25 years and above. For ages 15 to 24 years old, there were 0.40 deaths per 100.000 people.[6]

Early recognition of CHD in adults is essential to prevent further decrement of cardiac function. Transthoracic and Doppler echocardiography is the most widely used imaging tool in diagnosing CHD. Other options include cardiac magnetic resonance (CMR), cardiac computed tomography, and nuclear scintigraphy.[7] Echocardiography is still a limited resource in Indonesia. It is unavailable in primary health care and only performed in higher health care facilities when there is a suspicion of cardiac abnormalities. On the other side, electrocardiography (ECG) is a well-known modality with low cost and is widely used in health care for cardiac assessment. Some reports state that patients with ECG abnormalities have CHD using further evaluation.[8,9]

Abnormalities in ECG may refer to lesions found in CHD.[10] First-degree heart block, third-degree heart block, right bundle branch block, and right or/and left ventricular hypertrophy may be found in ventricular septal defect (VSD).[11] Previous studies reported incomplete right bundle branch block and crochetage R wave presence in inferior derivations in patients with ASD.[12] Other than that, defective T wave (DTW) has recently

been described as a sensitive marker for ASD.[13] In the patient with patent ductus arteriosus (PDA), the ECG may demonstrate sinus tachycardia, atrial fibrillation, left ventricular hypertrophy, right ventricular hypertrophy, right bundle branch block, and left atrial enlargement in moderate to large ductus shunts.[14,15] Electrocardiogram in a most patients with ToF shows right bundle branch block (RBBB) with or without QRS fragmentation.[16]

It is now recognized that CHD is associated with lifelong comorbidities such as arrhythmias, pulmonary hypertension, and a repeated need for surgery.[17] After reaching 25 years old, the heart will undergo permanent remodeling. Early recognition and repair prevent morbidity and mortality caused by CHD.[18] As reported in several studies, ECG findings may provide important clues in patients with CHD.[11–16] We, therefore, examine ECG as an alternative early screening modality to assess cardiac abnormalities in CHD.

METHODS

A descriptive cross-sectional study was done at the Faculty of Medicine and Health Sciences, Atma Jaya Catholic University of Indonesia. Participants were all students above 18 years old and willing to be assessed. Participants with a history of CHD will be excluded from this study.

Data were collected between August and November 2019. After giving informed consent, participants underwent history taking, anthropometry, blood pressure measurement using a digital sphygmomanometer, and 12 lead Electrocardiogram (ECG) measurement. Two cardiologists interpreted the results independently to minimize observer bias.

Table 1. Criteria for ECG Findings

Findings	Criteria
Right axis deviation	Axis > 90°
Left axis deviation	Axis < -30°
Right atrial enlargement	P waves amplitudes > 2.5 mm
Left atrial enlargement	P waves duration > 0.12s
Right ventricular hypertrophy	R:S ratio in V1 > 1
Left ventricular hypertrophy	S in V1 + R in V5 or V6 >35 mm
Right bundle branch block	RSR' pattern in V1-V3 QRS duration >0.10s
Left bundle branch block	QS pattern in V1 R notch in V5, V6 QRS duration >0.10s
Crochetage R wave	A notch near the apex of the R wave in the ECG inferior limb leads
Defective T wave	Inverted displacement of the T wave in V1-V6

RESULTS

One hundred ninety-three participants were assessed during the study period, consisting of 78 males and 115 females. The average age of the participant was 19 years old, ranging from 18 to 24 years old. Among participants, most of them never smoke, and almost half of them do not do routine physical activity.

Table 2. Demographic Characteristics

Characteristic	N(%) (N:193)
Gender	
Male	78 (40.4)
Female	115 (59.6)
Age (years old)	19.22 ± 1.85
Height (cm)	161.38 ± 14.28
Weight (kg)	62.62 ± 14.38
Body mass index (km/m^2)	23.65 ± 4.40
Systolic blood pressure (mmHg)	115.84 ± 22.87
Diastolic blood pressure (mmHg)	74.8 ± 15.01
Smoking habit	
Active	3 (1.6)
Stop; less than 12 months ago	3 (1.6)
Never	197 (96.9)
Routine physical activity	
At least once a week	102 (52.8)
Not routinely	91 (47.2)

Generally, all participants were healthy and did not have specific symptoms indicating any cardiac diseases. However, some reported occasional non-specific symptoms, including atypical chest discomfort, shortness of breath, and dizziness or pre-syncope (Table 3). Further exploration indicated that the abovementioned symptoms were mainly related to psychological factors or non-cardiac conditions such as asthma and muscle soreness.

Table 3. Reported Symptoms and Signs

Symptoms	N (%)
Atypical chest discomfort	21 (10.9)
Dizzy or pre-syncope	17 (8.8)
Shortness of breath	24 (12.4)
Heart murmur	1 (0.5)
Increase blood pressure	8 (4.1)
Physical activity limitation	5 (2.6)

Table 4. Electrocardiogram Findings

Characteristic	N (%)
Sinus rhythm	157 (81.3)
Sinus arrhythmia	36 (18.7)
Heart rate (beats/minute)	75.39 ± 12.82
< 60	11 (5.7)
60-100	177 (91.7)
> 100	5 (2.6)
Axis	
Normoaxis	177 (91.7)
Right axis deviation	13 (6.7)
Left axis deviation	3 (1.6)
P wave	
Duration	0.079 ± 0.01
Amplitude	1.256 ± 0.42
PR interval	0.158 ± 0.02
QRS duration	0.085 ± 0.05
QT interval	0.358 ± 0.04
Corrected QT interval	0.398 ± 0.04
S in lead V1/V2	9.78 ± 4.73
R in lead V5/V6	12.61 ± 3.2
Pathologic Q wave	4 (2.1)
ST elevation	6 (3.1)
Inverted T wave	33 (17.1)
Right atrial enlargement	0
Left atrial enlargement	0
Right ventricular hypertrophy	9 (4.7)
Left ventricular hypertrophy	9 (4.7)
Right bundle branch block	6 (3.1)
Left bundle branch block	0
Crochetage sign	15 (7.8)
One lead	7 (4.6)
Two leads	5 (2.5)
Three leads	3 (1.5)
Defective T wave	2 (1.0)
Interpretation	
Normal	136 (70.5)
Abnormal	57 (29.5)

As presented in Table 4, from 193 ECG 57 were interpreted as abnormal. These abnormalities included axis deviation, right and left ventricular hypertrophy, right bundle branch block (RBBB), crochetage sign, and defective T wave. Both crochetage sign and defective T wave are more specific to assess congenital heart disease, especially atrial septal defect. No sign of atrial enlargement and left bundle branch block was found among the participants.

Echocardiography was performed on twenty participants. Others with ECG abnormality were unable to have echocardiography due to conflicting schedules and decided to undergo further evaluation

outside the study. Despite various abnormalities found on ECG, almost all echocardiography results showed normal findings. No congenital heart disease was detected by echocardiography work-up from these participants. There was no echocardiographic sign of right heart strain. One participant with incomplete RBBB was found to have mild mitral valve prolapsed (MVP). Data are shown in Table 5.

Table 5. Echocardiography findings

Number	Gender	Age	BP (mmHg)	HR	Electrocardiogram		Echocardiogram
1	Male	20	120/73	66	Sinus arrhythmia	Crochetage sign	Normal findings
2	Male	18	113/67	78	Sinus rhythm	RAD, incomplete RBBB	Mild MVP
3	Male	20	107/70	54	Sinus arrhythmia	Crochetage sign	Normal findings
4	Male	20	130/72	70	Sinus arrhythmia	LAD	Normal findings
5	Male	19	126/80	80	Sinus arrhythmia	RVH	Normal findings
6	Male	20	136/75	60	Sinus arrhythmia	Incomplete RBBB	Normal findings
7	Male	19	118/78	110	Sinus arrhythmia	RVH	Normal findings
8	Male	19	122/80	68	Sinus rhythm	Crochetage sign	Normal findings
9	Male	19	123/80	80	Sinus rhythm	Crochetage sign	Normal findings
10	Male	18	113/69	68	Sinus rhythm	Crochetage sign	Normal findings
11	Male	18	127/81	60	Sinus rhythm	Crochetage sign	Normal findings
12	Male	23	122/88	60	Sinus arrhythmia	RVH, crochetage sign	Normal findings
13	Female	21	96/72	83	Sinus rhythm	RVH	Normal findings
14	Female	21	113/73	71	Sinus rhythm	RAD, RVH	Normal findings
15	Female	19	134/73	84	Sinus rhythm	Crochetage sign	Normal findings
16	Female	18	128/81	75	Sinus rhythm	RVH	Normal findings
17	Female	18	138/90	88	Sinus rhythm	RVH	Normal findings
18	Female	18	118/70	84	Sinus rhythm	Crochetage sign, incomplete RBBB, LAD	Normal findings
19	Female	18	118/74	60	Sinus rhythm	Crochetage sign	Normal findings
20	Female	18	94/67	88	Sinus rhythm	Incomplete RBBB	Normal findings

BP = Blood pressure, HR = Heart reate, RVH = Right ventricular hypertrophy, RAD = Right axis deviation, LAD =Left Axis Deviation, RBBB = Right bundle branch block, PR = Pulmonary regurgitation, MVP = Mitral valve prolapse

DISCUSSION

Congenital heart disease was once considered a pediatric disease as most patients with severe lesions rarely survived into adulthood. However, advances in early diagnosis and cardiac surgery have shifted the disease burden from pediatrics to adult.[18] In the last decade, the number of grown-up patients with CHD is increasing, constituting 60% of the total CHD.[1] Thus, a screening program in healthy young adults is important to detect any mild or early stage of this abnormality before the symptoms and complications occur.

The electrocardiogram is one of the modalities to detect CHD, especially in rural areas where other modalities, such as echocardiography, are still limited and costly. Together with comprehensive clinical examination, ECG could be valuable in screening cardiac abnormalities, including congenital heart disease. This examination is relatively affordable and practical to conduct. As presented in the table, atrial enlargement, ventricular hypertrophy, conduction block, and other findings can be a sign of CHD.

Table 6. Electrocardiography findings in Congenital Heart Disease

Number	Diseases	Electrocardiography Findings
1	Ventricular Septal Defect[11,19,20]	• Left ventricular hypertrophy • Right ventricular hypertrophy • Biventricular hypertrophy • Right bundle branch block • First-degree AV block • Third-degree AV block
2	Atrial Septal Defect[12,21]	• Right bundle branch block • Crochetage R wave sign • Defective T wave
3	Patent Ductus Arteriosus[14,22]	• Left atrial enlargement • Left ventricular hypertrophy • Right ventricular hypertrophy • Right bundle branch block • Sinus tachycardia • Atrial fibrillation
4	Coarctation of the Aorta[10,23,24]	• Left axis deviation • Left ventricular hypertrophy • Right bundle branch block • Left bundle branch block
5	Congenital Aortic Stenosis[25,26]	• Left ventricular strain • Left ventricular hypertrophy • Left axis deviation

Crochetage pattern is defined as a notch near the apex of the R wave in ECG inferior limb leads. According to Heller et al., its sensitivity and specificity for atrial septal defect reach 73.1% and 92.6% if present in one lead, 58.1%, and 97.2% if present in two or three leads. When the pattern was found in all inferior limb leads, its sensitivity and specificity reached 27.8% and 100%, respectively.[27] Some studies showed crochetage R wave in inferior leads and incomplete right bundle branch block in atrial septal defect (ASD) patients.[21] Other findings, a defective T wave (DTW), has also been described as a sensitive finding for ASD. DTW is defined as an inverted proximal limb of the T wave in the right precordial leads.[12]

In our study, 29.5% of participants had specific abnormalities on ECG, suggesting signs of congenital heart disease. Crochetage sign and DTW were found in 15 and 2 participants, respectively. This result should raise our concern about finding the culprit and preventing early complications. Although ECG findings can indicate the existence of CHD, a normal variation or a false positive result is possible, which are common findings in interpreting ECG in young people.[10] Moreover,

as generally understood, ECG interpretation is more valuable in detecting arrhythmias than cardiac structural abnormalities. Further imaging, echocardiography is necessary and plays an important role in confirming these ECG findings.

In our study, further echocardiography evaluation in 20 out of 57 participants with abnormal ECG findings could not confirm any congenital heart disease. The cardiac dimensions are normal without any sign of right heart strain; hence congenital heart disease tends to be unlikely. These results are probably due to the lack of accompanying incomplete right bundle branch block on ECG as suggested by Deri et al.[21] Although several studies stated that crochetage pattern is an independent marker for ASD,[27,28] its sensitivity and specificity are higher when associated with incomplete right bundle branch block, and other signs of right ventricular hypertrophy.[27] Unfortunately, we cannot explore more confirmative echocardiographic data for participants with DTW patterns on ECG for suggestive of ASD because none consented to further evaluation.

CONCLUSION

Electrocardiography could detect the characteristic patterns suggestive of congenital heart disease. However, wide-range normal variation is commonly encountered, especially in the young population. From our study, ECG alone appears insufficient to confirm cardiac structural abnormalities. More trials are needed to consider ECG as an adjunctive tool in screening for congenital heart disease and its cost-effectiveness in young adults.

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