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Original Article

Modified Ross score and echocardiographic score in children with heart failure: a subgroup analysis

I Made Karma Setiyawan, Ni Putu Veny Kartika Yantie, Anak Agung Ngurah Ketut Putra Widnyana, I Gusti Lanang Sidiartha, I Putu Gede Karyana, I Made Arimbawa

Abstract

Background Over the past two decades, heart failure in children has increased in terms of symptom recognition and prevalence. The initial clinical manifestations of heart failure in children are non-specific. Therefore, diagnosis requires the support of echocardiography. The severity of heart failure symptom in children can be classified through a simple scoring system such as Ross score. The cardiac remodeling status, duration of heart disease and treatment may have clinical and anatomical effects on the disease.

Objective To analyze correlation between modified Ross score and echocardiographic score by subgroup analysis consisting of cardiac remodeling status, duration of heart disease and treatment.

Methods This cross-sectional study included children aged 1 month to 18-years-old with heart failure who sought treatment at Prof.Dr. I.G.N.G Ngoerah Hospital, Denpasar from June 2019 to February 2020. Cardiac remodeling was defined as >20% increase in left ventricle internal end diastolic dimension (LVIDd) compared to normal values, based on body surface area. Spearman's correlation test was used for statistical analysis.

Results A total of 30 subjects were analyzed in this study. The median modified Ross score and echocardiography score were 3 (range 2-11) points and 4 (range 2-6) points, respectively. The median durations of heart disease and preventive heart failure treatment were 2 years (range 7 days-15 years) and 1 year (range 7 days-15 years), respectively. The mean LVIDd was 4.3 (SD 1.4) cm. Twenty-one out of 30 subjects experienced a ≥ 20% increase of LVIDd from baseline. The modified Ross score and echocardiographic score had no significant correlation (r=0.18; P=0.33). However, the modified Ross score had significant correlations with duration of heart disease (r=-0.632; P<0.001) as well as duration of treatment (r=-0.584; P=0.001). In addition, no correlation was found between echocardiographic score with heart disease and treatment duration (P>0.05). Echocardiography score and remodeling process was significantly correlated (r=0.64; P<0.001).

Conclusion There is no correlation between modified Ross score and echocardiographic score. Duration of heart disease and treatment are significantly negatively correlated with modified Ross scores. The remodeling process is positively correlated with echocardiographic score. Further research on acute heart failure and validated echocardiographic scores are needed.**[Paediatr Indones. 2024;64:202-8; DOI: 10.14238/pi64.3.2024.202-8**].

Keywords: *children; heart failure; modified Ross score; echocardiographic score; remodelling*

Over the past two decades, heart failure in children has increased in terms of symptom recognition and prevalence. Heart failure may result from anatomical and functional abnormalities that can be detected using echocardiography. The severity of heart failure symptom in children can be classified through a simple scoring system.[1]

The etiologies of heart failure in children are generally divided into congenital heart disease

From the Department of Child Health, Medical Faculty of Universitas Udayana/Prof.Dr.I.G.N.G Ngoerah Hospital, Denpasar, Bali, Indonesia.

Corresponding author: Ni Putu Veny Kartika Yantie. Department of Child Health, Medical Faculty of Universitas Udayana/Prof.Dr.I.G.N.G Ngoerah Hospital. Jalan Diponegoro, Denpasar Barat 80113, Denpasar, Bali, Indonesia. Telp. +62-811398802; Email: kartika.veny@gmail.com.

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(CHD) and acquired heart disease. Congenital heart disease is further categorized into acyanotic and cyanotic. Acquired heart disease includes cardiomyopathy, carditis, Kawasaki disease, heart rhythm abnormalities, and rheumatic fever.[2] Heart failure due to CHD in pediatric patients was estimated to be 1-2 per 1,000 live births and 65% of CHD was hospitalized in American children.[3,4] Congenital heart disease, which contributes to 55-60% of heart failure, is more often diagnosed when the child is less than one year old.[5] Other non-cardiac conditions that cause heart failure in children include fluid overload, anemia, sepsis, systemic hypertension, and vasculitis.[6]

The clinical manifestations of heart failure in children are initially non-specific but the symptoms become more distinctive in a severe stage. The original Ross score, the *New York Heart Association* (NYHA) score, and the most frequently used modified Ross score in diagnosing heart failure are based on clinical symptom-based assessment.[7] Assessment of heart failure by echocardiography in children uses variables that describe the function of the right and left ventricles. These variables are simplified in the echocardiographic score according to the underlying cause of heart disease.[8]

Acute compensation for heart failure symptoms involves activation of the sympathetic nervous system and renin-angiotensin-aldosterone system to maintain cardiovascular homeostasis. However, chronic system stimulation worsens myocardial function and affects remodeling.[9] In children with heart failure, the main purposes of medication are to decrease pulmonary wedge pressure, increase cardiac output, and improve end-organ perfusion, therefore delaying disease progression.[10] Sub-optimal treatment or failure to follow the suggested regimen might generally aggravate the disease and cause decompensation, both clinically and anatomically.[11] Based on the rationale above, we would like to analyze for correlation between modified Ross score and echocardiographic score by subgroup analysis consisting of cardiac remodeling, duration of heart disease and treatment.

Methods

We recruited subjects using a consecutive sampling method for this cross-sectional study, conducted from June 2019 to February 2020. Inclusion criteria were all patients aged one month to 18-year-old with congestive heart failure, fulfilled the original Ross score (≥ 2), and who were seen or admitted to the Pediatric Cardiology Outpatient Clinic and/or Pediatric Ward of Prof.Dr. I.G.N.G Ngoerah Hospital, Denpasar, Bali. Children with renal failure, musculoskeletal injury, sepsis, lung infection, human immunodeficiency virus infection, pulmonary stenosis, and those who lacked parental consent were excluded from the study. Written informed consent was obtained from parents before recruitment.

The diagnosis of heart failure was established according to the original Ross score classification, physical examinations, and echocardiography. A modified Ross score was used for clinical evaluation of heart failure severity related to age. The symptoms of heart failure were graded on a scale of 0, 1, or 2 points, according to severity. This score was used to classify patients as having no heart failure (0-2 points), mild heart failure (3-6 points), moderate heart failure (7-9 points), or severe heart failure (10-12 points).[1] Echocardiographic score was designed according to cardiac disease type. For congenital heart defects, the score consisted of the size of the defect, Doppler gradient across the defect, and left ventricle diastolic dimension (LVd) *z*-score. For valvular heart disease, the score consisted of the echo severity of the valvular lesion, pulmonary hypertension, and LV(d) *z*-score. Score of ≥ 4 are considered high score.[8] The left ventricular end-diastolic dimension [LV (d)] parameter represents the left ventricle during the diastolic phase obtained according to the body surface area (BSA). LV (d) is obtained with the equation: $3.5(\text{BSA}-0.1215) + 4.9$, SD = 0.0788, and is expressed in centimeters.[8,12] Pediatric cardiologists repeated the echocardiography measurements three times to obtain the mean echocardiographic parameters. Duration of heart disease was defined as the duration of heart disease experienced before being diagnosed based on history of symptoms. Cardiac remodeling was defined as a >20% increase in LVIDd compared to normal values based on BSA.[13,14]

Descriptive data were presented as median or mean. Normality test (Shapiro-Wilk) was performed on the data. Spearman's test was used to analyze correlations of variables. A P value of <0.05 was used to determine statistical significance. We analyzed all

data with *SPSS 22.0* software. The study protocol was approved by the Health Research Review Committee of Prof.Dr. I.G.N.G Ngoerah Hospital.

Results

From June 2019 to February 2020, 30 subjects were included. Subjects' median age was 6.5 years (range 3 months - 17 years). A combination of furosemide, captopril, spironolactone, and digoxin was used as oral anti-heart failure agents. The median durations of heart disease and preventive heart failure treatment were 2 years (range 7 days - 15 years) and 1 year (range 7 days - 15 years), respectively.

Twenty out of 30 subjects had received treatment for more than 1 year. The mean LVIDd was 4.3 (SD 1.4) cm, with 2/30 subjects experienced an increase of ≥ 20% from baseline. The main characteristics of subjects are shown in **Table 1**.

Frequent sweating during infant feeding or dispneu d'effort in older children were heart failure symptoms commonly found using the original Ross classification. Subjects' median modified Ross score was 3 (range 2-11) points. The most common physical examination finding for heart failure based on modified Ross scores was increased heart rate (28/30 subjects). Subjects' median echocardiographic score was 4 (range 2-6), with 24/30 subjects had scores that were considered high (≥ 4). A total of 8 subjects with a modified Ross score of ≥ 7 points had high echocardiographic score. **Table 2** shows the Ross score and echocardiographic score.

There was no significant correlation between modified Ross score and the echocardiographic score. The modified Ross score significantly correlated with duration of the disease and treatment, while the echocardiography score substantially correlated with the remodeling process (**Table 3**).

Valvular heart disease had median modified

Table 1. Baseline characteristics of subjects

Characteristics	(N=30)
Male, n	19
Nutritional status, n	
Severe malnutrition	2
Malnutrition	18
Well-nourished	9
Overweight	1
Stunted, n	13
History of medication, n	
Two-drug combination	3
Three-drug combination	17
Four-drug combination	10
Heart disease, n	
Congenital heart defect, n	21
Ventricular septal defect (VSD)	12
Patent ductus arteriosus (PDA)	9
Valvular heart disease, n	9
Congenital mitral stenosis	1
Rheumatic heart disease (RHD)	8
RHD mitral regurgitation	4
RHD mitral and aortic regurgitation	4
Median duration of heart disease (range)	
Congenital heart defect	3 years (3 months-15 years)
More than 1 year, n	20
Valvular heart disease	14 days (7 days-10 years)
Less than 1 year, n	5
Median duration of therapy (range)	
Congenital heart defect	1 year (1 month-15 years)
Valvular heart disease	14 days (7 days-10 years)

Table 2. Ross score and echocardiographic score

Score	(N=30)
Original Ross	
Class II	13
Class III	10
Class IV	7
Modified Ross Score	
Score 0-2	9
Score 3-6	12
Score 7-9	6
Score 10-12	3
Echocardiographic Score	
High score (≥ 4)	24
Low score (< 4)	6
Moderate to severe modified Ross score (≥ 7) + high echocardiographic score (≥ 4)	8
Mild modified Ross score (< 7) + high echocardiographic score (≥ 4)	16

Ross score of 9 points (range 3-11), while CHD had modified median Ross score of 3 points (range 2-9). Valvular heart disease had a strong positive correlation with increased modified Ross score (r=0.7; P<0.001). Subjects with modified Ross scores of ≥ 7 points had a median duration of heart disease of 14 days (range 7 days - 2 years) and a median duration of treatment of 14 days (range 7 days - 2 years). The cardiac remodeling process occurred in 21 subjects. Thirteen out of 21 subjects with cardiac remodeling processes had a modified Ross score of < 7 points and the majority of subjects received therapy for > 1 year (10/13 subjects). The proportion of high echocardiography scores was higher in valvular heart disease than in CHD (8 *vs.* 16 out of 30 subjects). The median durations of heart disease and treatment in subjects with high echocardiographic scores were 1.5 years (range 7 days-12 years) and 1 year (range 7 days-12 years), respectively. Echocardiographic score did not correlate with heart disease type (r=0.088; P=0.64). A total of 20/30 subjects with remodeling process had high echocardiographic scores; 15/20 subjects had been suffering from heart disease for >1 year and the majority had received therapy for >1 year (12/20 subjects) (**Table 4**).

Discussion

Most of our subjects had mild heart failure and the majority of abnormalities were ventricular septal defects. A previous study found that most of their subjects had moderate heart failure according to their modified Ross score and ventricular septal defects were the most common abnormality.[4] This difference may have been due to our clinical practice in using the original Ross score to diagnose heart failure before the symptoms were reevaluated using the modified Ross score.

Most of our subjects were male and subjects' median age was 6.5 years (range 3 months-17 years). The median age of CHD diagnosis was 3 years old (8 months-15 years) in our study. The median duration of heart disease in those with valvular heart disease was 14 days (range 7 days-10 years). The heart failure classification system in children varies according to age due to differences in clinical symptoms.[1] A study found that CHD most often presents at a median age of 11 (range 4-22) years old.[15] This difference may have been due to the fact that large-sized CHD symptoms begin to show clinical symptoms after 3

Table 3. Analysis of modified Ross score and echocardiographic score with duration of heart disease, duration of therapy, and remodeling process

Variables	Echocardiographic score	Duration of heart disease (year)	Duration of therapy (year)	Remodeling process
Modified Ross score	r= 0.18*	r= -0.63**	r= -0.58**	r=0.33*
Echocardiographic score	-	r= -0.06*	r= -0.12*	r=0.64**

*P value ≥ 0.05, **P value < 0.05

Table 4. Subgroup characteristics related to modified Ross score and echocardiographic score

Variables	Moderate to severe modified Ross score (≥ 7), n=9	High echocardiographic score (≥ 4), n=24
Type of heart disease		
Valvular heart disease	8	8
Congenital heart defect	1	16
Duration of heart disease > 1 year	3	19
Duration of therapy < 1 year	6	16
Remodeling process	8	20

months of age and require actions to close the defect.[16] Also, our study subjects were patients with CHD who had heart failure.

Echocardiography is an essential examination in heart failure for evaluating cardiac structure, chamber volumes and diameters, myocardial contractility, wall thickness, and pulmonary pressure. Neurohormonal activation and clinical deterioration have been shown to be correlated with increase of clinical scores of heart failure.[1,10,17] However, we found no correlation between modified Ross score and echocardiographic score (r=0.18; P=0.33). Since most of the participants in our study had chronic CHD, this correlation was not observed. This is most likely because the development of pulmonary vascular obstructive disease (Eisenmenger's syndrome) reduced the severity of the clinical symptoms. This result is consistent with a previous study on older children with CHD.[8] To date, no other studies have shown the correlation between modified Ross score with echocardiographic score. It is possible to determine whether non-specific symptoms like tachypnea, dyspnea, and failure to thrive are related to heart failure with the use of an echocardiogram, electrocardiography, and chest X-ray.[18] When symptoms of acute heart failure appear, echocardiography is performed to evaluate the hemodynamic condition, with parameters including left ventricular shortening fraction (<25%) or ejection fraction (<45%).[11] Most pediatric ventricular function, size, mass, and volume measurements have been normalized to account for age and body size differences.[19]

Children with CHD should receive therapy to control the chronic heart failure process due to structural abnormalities.[11,19] Treatment options for chronic heart failure include a combination of ACE- inhibitors, β-blockers, aldosterone antagonists, and digoxin.[10,11] There was a moderate negative correlation (r=-0.584; P=0.001) between severity of the symptoms based on the modified Ross score and the duration of treatment. This finding was consistent with previous evidence showing that administering heart failure medications can overcome hemodynamic problems, thereby reducing the clinical decompensation process.[6] The duration of heart disease can also contribute to clinical symptoms. Generally, the symptoms of heart failure depend on the presence or absence of a congestive process in chronic heart failure or hypoperfusion due to acute heart failure. We found a strong negative correlation between clinical symptoms based on modified Ross score and duration of heart disease (r=-0.632; P<0.001). In contrast, we found a strong positive correlation between valvular heart disease and increased modified Ross score (r=0.7; P<0.001). We noted that the manifestations of heart failure symptoms can vary, either appearing acutely or developing earlier and becoming chronic, affecting the severity of the symptoms.[20] We found no correlation between modified Ross score and cardiac remodeling (r=0.33; P=0.08). One hypothesis stated that the scope of the effect of anti-remodeling therapy administration on LVID depends on the baseline severity of remodeling, which the outcome will correspond to the magnitude of reversal in remodeling.[21] These processes will cause symptoms of heart failure that are not visible clinically.

Children are at risk of experiencing symptoms of heart failure which are not always clinically visible. Therefore, the timing of therapy for CHD can be considered based on anatomical and hemodynamic conditions that take into account myocardial cell adaptation and chamber remodeling. Children with left-to-right-shunt heart disease can develop obstructive pulmonary vascular disease (Eisenmenger syndrome) if left untreated for years. Valvular type heart disease can increase volume load in both ventricles

(mitral regurgitation), increase the diastolic pressure gradient between the two atria (mitral stenosis), and increase volume load in the left ventricle (aortic regurgitation).[22] We found that the measurement of echocardiographic score parameters is not influenced by the duration of the disease and treatment (r=-0.124; P=0.51 and r=-0.06; P=0.75, respectively). These results were consistent with a past study that stated the provision of treatment aims to maintain cardiovascular homeostasis, but corrective surgery is the main approach in children with CHD or valvular heart disease.[13] The majority of chronic heart failure in our study (whether stable, progressively worsening or decompensated) requires consideration of therapy to prevent progression, decompensation, or death.[23] There is insufficient evidence to support the efficacy of standard heart failure therapy in both pediatric and adult patients with CHD.[24] We did note a correlation between echocardiographic scores and the cardiac remodeling process (r=0.64; P<0.001). Aminullah et al. reported that pediatric patients who underwent surgery to close the VSD defect had improved LVIDd by 9.89-10.85% within 3 months.[25] The parameters arranged in the echocardiographic score can indicate the occurrence of a remodelling process in the heart, in the form of excessive compensation for heart injury, and manifest as changes in the size, shape and function of the left ventricle.

Limitations of this study were the fact that the majority of subjects suffer from chronic heart disease and have received oral anti-heart failure therapy. The echocardiographic scores in this study have not been used or validated in other studies. Validation is needed before this echocardiographic scoring system can be accepted for general use.

In conclusion, the modified Ross and echocardiographic scores had no significant correlation in children with heart failure. Subgroup analysis revealed that heart disease and treatment duration significantly correlated with the modified Ross score. In addition, the remodeling process correlates with echocardiographic score. Further studies on acute heart failure and validated echocardiographic scores are needed.

Conflict of interest

None declared.

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