ASSESSMENT OF RELIABILITY AND VALIDITY OF THE MALAY VERSION PedsQL™ 3.0 CARDIAC MODULE

BY

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LIST OF ABBREVIATION

ALCAPA	Anomalous origin of the Left Coronary Artery from the Pulmonary Artery
ASD	Atrial Septal Defect
AVSD	AtrioVentricular Septal Defect
AP Window	AortoPulmonary Window
AV	AtrioVentricular
CDC	Center of Disease Control
CHD	Congenital Heart Disease
COA	Coarctation of aorta
CCTGA	Congenitally Corrected Transposition of the Great Arteries
DILV	Double Inlet Left Ventricle
DORV	Double Outlet Right Ventricle
EFA	Exploratory Factor Analysis
HRQOL	Health Related Quality Of Life
HLHS	Hypoplastic Left Heart Syndrome
ICC	Intraclass Correlation Coefficient
MAPCAS	Major AortoPulmonary Collateral Arteries

PAIVS	Pulmonary Atresia with Intact Ventricular Septum
PAVSD	Pulmonary Atresia with Ventricular Septal Defect
PDA	Patent Ductal Arteriosus
PedsQL	Pediatric Quality of Life inventory
POS	Pulse Oxymetry Screening
PAF	Principal Axis Factoring
PS	Pulmonary Stenosis
QOL	Quality Of Life
TOF	Tetralogy of fallot
TGA	Transposition of great arteries
TAPVD	Total Anomalous Pulmonary Venous Drainage
VA	VentriculoArterial
VSD	Ventricular Septal Defect
WHO	World Health Organization

ABSTRAK

TAJUK

Penilaian tentang kebolehpercayaan dan kesahihan PedsQL™ 3.0 Modul Kardiak versi Bahasa Melayu.

OBJEKTIF

Untuk menghasilkan PedsQL™ 3.0 Modul Kardiak versi bahasa Melayu yang boleh dipercayai dan sahih.

TATACARA

Ini merupakan kajian keratan rentas melibatkan kanak-kanak yang menghidapi penyakit jantung kongenital di antara umur 5-18 tahun dan ibubapa yang mana anaknya berumur 2-18 tahun. Sejumlah 528 peserta terlibat dalam kajian ini adalah yang hadir di klinik pakar kanak-kanak kardiologi dan klinik ekokardiografi di Hospital Raja Perempuan Zainab II dan Hospital Universiti Sains Malaysia. Tatacara yang digunakan termasuklah penterjemahan ke hadapan dan ke belakang, kesahihan isimuka dan kandungan soalan PedsQL™ 3.0 Modul Kardiak versi bahasa Melayu.

KEPUTUSAN

Kajian ini menyokong kebolehpercayaan dan kesahihan PedsQL™ 3.0 Modul Kardiak dalam menilai tahap kesihatan kualiti hidup kanak-kanak yang menghidapi penyakit jantung di masa hadapan.

KESIMPULAN

PedsQL™ 3.0 Modul Kardiak versi Bahasa Melayu ini terbukti akan kebolehpercayaan dan kesahihannya. Soalan PedsQL™ 3.0 Modul Kardiak ini sangat berguna untuk menilai tahap kualiti hidup pesakit yang menghidapi masalah jantung.

ABSTRACT

TITLE

Assessment of reliability and validity of the Malay version PedsQL^{TM} 3.0 Cardiac Module

OBJECTIVE

To produce a reliable and validated Malay version of the PedsQL^{TM} 3.0 Cardiac Module.

METHODS

This is a cross sectional study involving children with congenital heart disease between 5-17 years old and parent of children with congenital heart disease aged 2-18 years old. A total of 528 participants were enrolled into this study when they presented to the pediatric cardiology clinic and echocardiography clinic at Hospital Raja Perempuan Zainab II and Hospital Universiti Sains Malaysia. The methods used including forward and backward translation, face and content validation of Malay version of the PedsQL^{TM} 3.0 Cardiac Module questionnaires.

RESULT

Most of 27 items in PedsQL^{TM} 3.0 Cardiac Module questionnaires showed satisfactory internal consistency with Cronbach Alpha exceeding 0.7 and good correlation with intraclass correlation coefficient 0.76-0.98. There were no missing data.

CONCLUSION

The result generally supports reliability and validity of Malay version PedsQL^{TM} 3.0 Cardiac Module in assessing HRQOL among children with heart disease.

ABSTRACT

ASSESSMENT OF RELIABILITY AND VALIDITY OF THE MALAY VERSION PedsQL^{TM} 3.0 CARDIAC MODULE

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Introduction: The improvement in medical and surgical treatment over the last decades has led to a dramatic increase in survival rates of patients with congenital heart disease (CHD). Hence, it is a very important to focus on HRQOL among children with CHD. Up to date, there is no assessment tool in Bahasa Melayu to measure HRQOL among children with heart disease. The validated tool is available is in English and other languages. The PedsQL^{TM} 3.0 Cardiac Module was chosen because it has been validated, developmentally appropriate for our subjects, disease specific, includes both self and proxy-report and covering a wider age group for

children 5-18 years old and parent-proxy age 2-18 years old, it can be conducted within 15 minutes, the questionnaires also multidimensional and had minimal missing data.

Objective: To produce a reliable and validated Malay version of the PedsQL^{TM} 3.0 Cardiac Module.

Patients and Methods: This is a cross sectional study involving children with CHD between 5-17 years old and parent of children with CHD aged 2-18 years old. A total of 528 participants were enrolled into this study when they presented to the pediatric cardiology clinic and echocardiography clinic at Hospital Raja Perempuan Zainab II and Hospital Universiti Sains Malaysia. The methods used including linguistic validations of Malay version of the PedsQL^{TM} 3.0 Cardiac Module questionnaires.

Results: Most of 27 items in Malay version PedsQL^{TM} 3.0 Cardiac Module questionnaires showed satisfactory internal consistency with Cronbach Alpha exceeding 0.7 and good correlation with intraclass correlation coefficient 0.76-0.98. The validity also showed all the domains are significantly correlated among each other. There were no missing data.

Conclusion: The result generally supports reliability and validity of Malay version PedsQL^{TM} 3.0 Cardiac Module in assessing HRQOL among children with congenital heart disease.

Associate Prof Dr Ariffin Nasir (Supervisor)

Associate Prof Dr Norsarwany Mohamad (Co-Supervisor)

Prof Col (B) Dr. Wan Pauzi (Co-Supervisor)

1. INTRODUCTION

According to National Institutes of Health (National heart, lung and blood institute, 2011), Congenital heart disease (CHD) is defined as problems with the heart's structure that are present at birth. The defects may involved the interior wall, valves and vessels of the heart. There are several types of congenital heart disease that range from a simple lesion to the complex defects. Congenital heart disease is the most common type of birth defect (National heart, lung and blood institute, 2011).

In Malaysia, there are many pediatric cardiology centres and Hospital Raja Perempuan Zainab II (HRPZ II) and Hospital Universiti Sains Malaysia (HUSM) are the tertiary centers in Kelantan. It is the regional centre of the east coast of peninsular Malaysia and received patients from the state of Kelantan and Terengganu.

The global incidence of congenital heart disease is about 8/1000 live birth (Hoffman, 1995). According to Malaysian Ministry of Health in 2007, they estimated about 4000 to 5000 children were born with congenital heart disease yearly (Annual report Ministry of Health, 2007). There is no local data on the incidence of congenital heart disease in Kelantan. A total of 2073 congenital heart disease was detected in children born in the state of Johor from 2006-2012 with the prevalence of 5.5 per 1000 live birth (Mat Bah, 2014) as presented in table 1.1. The most common type of congenital heart disease in Johor is ventricular septal defect. The severity of congenital heart disease among 2073 Johorean patient, 891 (43%) was severe and 964 (46%) were mild lesions. Regarding the prognosis of Johorean patient, 28% required no intervention and 16% of the cardiac lesions close spontaneously and the overall mortality rate was 12.2% (Mat Bah, 2014).

In the last several decades, advances in the medical and surgical care has increasingly improve the survival of patient living with chronic conditions. Hence, it is also critical to focus on the effort to improve the quality of life for patient with chronic illness such as congenital heart disease.

In Malaysia, no study has been undertaken to evaluate the quality of life in children with cardiac diseases.

Table 1.1 Epidemiology of Congenital Heart Disease in Johor from 2006 to 2012

Diagnosis	Number of patients	Number of CHD in percentage (%)
VSD	711	34.3
PDA	392	18.9
PS	204	9.8
TOF	121	5.8
ASD	112	5.4
TGA	61	2.9
AVSD	52	2.5
COA	43	2.1
Heterotaxia syndrome	36	1.7
PAVSD	35	1.7
PAIVS	32	1.5
DORV	31	1.5
Tricuspid atresia	30	1.4
HLHS	25	1.2
TAPVD	22	1.1
PAVSD, MAPCAs	20	1.0
Ebsteins anomaly	19	0.9
Mitral atresia	18	0.9
Unbalance AV canal	15	0.7
Truncus arteriosus	14	0.7
DILV	14	0.7
Aortic stenosis	13	0.6
Interrupted arch	11	0.5
Vascular rings and slings	8	0.4

Table 1.1 Epidemiology of Congenital Heart Disease in Johor from 2006 to 2012 (continue)

ccTGA	8	0.4
Pulmonary artery abnormality	6	0.3
Coronary artery fistula	5	0.2
Cor triatriatum	4	0.2
ALCAPA	3	0.1
AP window	2	0.1
Mitral stenosis	1	0.0
Single ventricle, other	1	0.0
Hemitruncus	1	0.0
AV discordant, VA concordant	1	0.0
Partial anomalous venous connection	1	0.0
Coronary artery anomalies, others	1	0.0
Total	2073	100

2. LITERATURE REVIEW

2.1 AETIOLOGY OF CONGENITAL HEART DISEASE

The aetiology of congenital heart disease is known to be multi-factorial. These include genetic predisposition, infection, teratology and environmental stimuli. A family history of the heart disease in close relatives increases the chances of congenital heart disease in subsequent pregnancy. A mother with congenital heart disease has 6% risk of having an affected offspring while an affected father has a 2% risk. Study has shown that having a child with congenital heart disease confers a risk of 2 to 3% in subsequent pregnancy but having a child with hypoplastic left heart syndrome (HLHS) or having two or more children with congenital heart disease could increase the risk of congenital heart disease by 10% in a subsequent child (Yogen, Ying-Hui, Renu, 2014).

Recently, the genetic factors are being recognized as playing an important role. They have identified single nucleotide polymorphisms and submicroscopic copy number abnormalities as having a role in the pathogenesis of congenital heart disease (Richards & Garg, 2010). This work has aided the discovery of an increasing number of monogenic causes of human cardiovascular malformations.

2.2 SCREENING OF CONGENITAL HEART DISEASE

The screening of congenital heart disease can be divided into the antenatal and postnatal screening. In the United Kingdom, any suspicion of congenital heart disease on screening or high risk factors in women warrants specialist fetal

echocardiography (Yogen et al., 2014). The indications for fetal echocardiography can be divided into three:

A) Antenatal

1. Fetal factors:
    - Suspected cardiac anomaly on ultrasound
    - Increased nuchal translucency thickness on ultrasound
    - Abnormal fetal karyotype such as trisomies (21, 18, 13), Turner syndrome, 22q11 deletion
    - Other congenital anomalies like an exomphalos and diaphragmatic hernia
    - Fetal hydrops
    - Fetal arrhythmia
    - Twin to twin transfusion syndrome
2. Maternal factors:
    - Increased maternal risk for Down's syndrome and other congenital defects (advanced maternal age or increased risk of Down's syndrome on serum screening)
    - Maternal alcohol abuses
    - Maternal ingestion of teratogenic drugs (anticonvulsants, lithium, retinoic acid, amphetamines)
    - Maternal diseases (insulin-dependent diabetes mellitus, phenylketonuria, exposure to Rubella, Coxsackie or Parvovirus infection, maternal Ro/La antibodies)

3. Familial factors:
    - Family history of congenital heart diseases in the first-degree relative
    - Genetic syndromes

B) Postnatal

Globally, Pulse Oxymetry Screening (POS) postnatally is used currently as the method to screen ductal dependent congenital heart disease (Yogen et al., 2014). They reported POS was feasible, cost-effective, well-tolerated, simple, and acceptable to parents and clinical staff. These findings are substantial enough to meet the criteria for universal screening (Yogen et al., 2014).

2.3 CLINICAL PRESENTATION OF CONGENITAL HEART DISEASE

The signs and symptoms of congenital heart disease may be non-specific in infants or bigger children. The symptoms includes symptoms of cardiac failure, recurrent infection, failure to thrive, unwell infant, feeble or absent pulses (brachial or femoral), persistent cyanosis in the absence of respiratory distress or cyanotic episodes, low oxygen saturation (<95%), heart murmur, presence of dysmorphic features or other congenital anomalies, sudden death, presence of arrhythmias and heart failure.

2.4 PSYCHOSOCIAL IMPACTS OF CONGENITAL HEART DISEASE AMONG CHILDREN

The improvement in medical and surgical treatment over the last decades has led to a dramatic increase in survival rates of patients with congenital heart disease. This resulted in the emergence of a growing population of children with congenital heart disease with or without surgical correction. Medical professionals managing these patients also have to deal concomitantly with their daily life issues.

Physical activity is a major issue in children with congenital heart disease that has a significant impact on their daily activity as participation in sport and exercise capacity has a direct link with cardiac status and a variety of risks, such as arrhythmias or syncope. The majority of the parent limit their child from engaging in physical activity. Nowadays, it is widely recognized that physical activity has long-term beneficial effects not only on the quality of life but also on long-term morbidity and overall mortality (Piepoli, Davos, Francis, 2004). Various studies were done to prove that exercise capacity differs significantly across the spectrum in congenital heart disease. It is important that the patient and their family are informed that exercise restriction is very rarely indicated. Obviously, safety is a major concern when prescribing exercise and athletic activities for children with cardiac disease, especially those with hypertrophic cardiomyopathy, coronary artery anomalies, Marfan syndrome and aortic valve disease, which have been associated with sudden cardiac death.

The issue regarding traveling was frequently propped up and physicians are asked by patients with chronic heart disease, whether they could travel safely. Questions usually relate to commercial aircraft travel or visiting locations at higher altitude.

Patients with cyanotic congenital heart disease, like healthy people, experience a fall in arterial oxygen saturation of about 8% from baseline but tolerate this well without supplemental oxygen; they maintain adequate tissue oxygen delivery due to the chronic rightward shift in their oxyhemoglobin dissociation curve and to secondary erythrocytosis (Broberg, Uebing, Cuomo, Thein, Papadopoulos, & Gatzoulis, 2007). In these Broberg et al. (2007) study, patients with Eisenmenger syndrome report traveling frequently and safely on commercial airlines.

The other problem faced by children with congenital heart disease is neuropsychological function including low intelligence, lack of attention or inadequate motor skill. In the study of Uzark, Jones, Burwinkle, & Varni, (2003), the mean verbal and performance IQ scores were in the high 80s and approximately 40% had scores less than 85, a full standard deviation below the population mean. Nevertheless, in some studies, the mean IQ score did not differ significantly from that of the general population. Clinical significant score on the hyperactivity scale was observed in the study by Shillingford, Glanzman, Ittenbach, Clancy, Gaynor, & Wernovsky, (2008) using the Behavior Assessment System for Children that was 3 to 4 times more frequent among the congenital heart disease patients than in the standardization sample.

There was an impact of missing school or being unable to attend fulltime because of treatment, outpatient appointments and fatigue (Birks, Sloper, Lewin & Parsons, 2007). Another study by Larcombe, (1995) described the children with other chronic conditions has also shown that prolonged periods of absence from school raised the concerns about keeping up with work and maintaining relationships with peers.

In the study done by Van Rijen, Utens, Roos-Hesselink, Meijboom, Van Domburg, Roelandt, & Verhulst, (2003), there was the high proportion of patients with a history of special education. The main reason for attendance of special education includes frequent hospitalization. Additionally, the highest educational level completed was more often a lower level compared to the normal group (Van Rijen et al., 2003).

The social functioning of the patient with congenital heart disease can be considered favourable. There were no significant differences were found in participation in leisure time activities, the congenital heart disease patients obtained similar involvement as compared to normal group of children. They also involved in outgoing and domestic activities (Van Rijen et al., 2003). In contrast, to another study by Utens, Verhulst, Erdman, Meijboom, Duivenvoorden, Bos, & Hess, (1994), which showed patient with congenital heart disease appeared to prefer handicrafts more often and to visit sports and watch television less often than did their normal peers group.

2.5 QUALITY OF LIFE

The quality of life can be described in various ways. World Health Organization (WHO) defined the quality of life as individual perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standard and concerns (WHO, 1995). Whereby, health is defined as ‘a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity’ (WHO, 1947).

Quality of life may be described as a child's ability to function in situational contexts of family, school, peer and derive personal satisfaction from doing so (Aaronson, 1988).

Some studies refer quality of life as life satisfaction meaning, well-being, happiness, health, functional status and adjustment (Ferrans & Powers, 1985).

Assessment of quality of life of children is more complex. The complexity of assessing quality of life is due to the developmental differences in understanding the content measured. Children with congenital heart disease are at risk of multiple other problems such as physical, psychological, social, growth and educational achievement.

2.6 FACTORS AFFECTING QUALITY OF LIFE IN CHILDREN WITH CONGENITAL HEART DISEASE

There are numbers of factors that may affect the quality of life in children with congenital heart disease depending on their stages of growth. In infancy, they are totally dependent on their parents and as they enter early childhood, they have different needs such as relationships with other children and obtaining independence or knowledge (Nousi & Christou, 2010).

Factors affecting quality of life in children with congenital heart disease are the following:

1. Delay in physical growth, including the height and the weight, which vary depending on the type and severity of the disease. Children with cyanotic congenital

heart disease have the most pronounced delay in physical growth, which can be obvious from a very young age.

2. The change in the body image in both the pre-operative and post-operative period experienced by children with congenital heart disease at all stages of physical growth especially school-going age. The large incision scar post-operatively may give rise to comments or questions in the child's environment, especially at the school.

3. Children with congenital heart disease experience anxiety and depression due to the frequent hospitalization, daily medications and the limitations imposed by the disease itself.

4. The social acceptance in the school environment is lacking. The physical impairment that these patients experience makes them unable to participate in school activities such as physical exercise. Usually, they have refrained from activities they used to enjoy before the onset of the disease, thus the feeling of loneliness, rejection, and social isolation, which makes social integration even more difficult. The overprotective behavior of the parents, who sometimes do not permit the children to take their own initiative, and at the same time reduce their ability to take care of themselves, significantly contributes to the patients' low self-esteem.

5. Children with congenital heart disease usually show poor school performance and they usually fall behind the progress of their healthy schoolmates owing to the fact that they have a long treatment process involving frequent hospital admissions and therefore, prolonged absence from school.

6. Educational level of the parents is significantly influencing their children's quality of life. The educational level of the parents is also associated with the absence or delay in seeking medical assistance, and thereby with the worsening of the children's

health. Parents with a low educational and income level tend to have difficulties in recognizing that their children need advisory support, or they ignore its importance and its consequence.

7. Maintenance of an overall good health including a balanced diet, prevention of anemia, and full vaccination against the common diseases is the factor that contributes positively to the improvement of the quality of life. Treatment of anemia is importance, especially in patients with cyanosis, which improves physical performance, their general health condition, and therefore, their quality of life.

2.7 CONCEPT OF HEALTH RELATED QUALITY OF LIFE (HRQOL)

The concept of health related quality of life and its determinants has evolved since the 1980s to encompass those aspects of overall quality of life that can be clearly shown to affect health either physical or mental (CDC, 2013).

Health related quality of life reflects the patient's perception of the impact of the illness and its treatment on their life (Nousi et al, 2010).

HRQOL is defined as a multidimensional construct composed of the patient's perception of the impact of disease and treatment on his or her functioning in a variety of aspects of life, including the physical, psychological, and social domains (Landolt, Buechel & Latal, 2008).

Health related quality of life instruments are tools used for measuring patient's functioning and general well-being. However, they may not sufficiently assess important areas that are likely to be affected specifically by a specific disease or its treatments. On the other hand, specific instrument does not permit comparisons

across conditions and treatments and rarely capture every aspect of health related quality of life important to patients. Therefore, a combined approach of measuring patient's general functioning and well-being and specific measurements may be the best option in many situations (Varni, 2002, 2004).

Solans, Pane, Estrada, Serra-Sutton, Berra, Herdman & Rajmil, (2008) have identified several instruments that have been used to assess the quality of life of children and adolescents in a systematic review of papers published between years 2001 and 2005, which consist of 30 generics, and 64 specific-disease instruments. The generic measures instruments include:

• WHOQOL BREF Questionnaires

• SF-36 scale questionnaires

• Child Health Questionnaire (CHQ)

• PedsQL^{TM} 4.0 Generic core Scale

Numerous disease specific instruments that have been used to evaluate patient with congenital heart disease and the examples are listed below:

• Pediatric Cardiac Quality of Life Inventory (PCQLI)

• Congenital Health Adolescent & Teenage Questionnaire (CHAT)

• Congenital Heart Disease Quality of Life Questionnaire (ConQOL)

• PedsQL^{TM} 3.0 Cardiac Module

The generic measure questionnaires assess the general health status, which quantifies the overall functional status and well-being but does not assess functional limitations

attributable to a particular disease. In contrast, a disease-specific instruments measured patient health related quality of life symptom burden, functional limitations, relevant to a specific condition. Disease specific health related quality of life measures have recently being developed for patient with congenital heart disease and are still in the process of being tested and validated for the population of different culture and language.

Studies that have used generic instruments to explore the quality of life in children with congenital heart disease include Silva, Vaz, Areias, Proenca, Viana & Areias, (2011) which used the WHOQOL-BREF questionnaire to evaluate the quality of life of adolescents and young adults with congenital heart disease. The study showed that patients with congenital heart disease had a better perception of quality of life than did the general population in the category of psychological, social relationship and environment scales. The WHOQOL-BREF instrument contains 26 questions, with the first 2 questions being general and the remaining 24 questions being divided into four different domains: physical, psychological, social relationships and the environment.

Another study that used generic measure questionnaires is the study by Lane, Lip, & Millane, (2002) which used SF-36 health survey questionnaires to assess the quality of life in adults with congenital heart disease at one general hospital in Birmingham, United Kingdom. The Short Form 36 health survey or SF-36 had 36 questions that relate to eight health status scales including physical functioning, general health, mental health, limitation of activities, social activities, emotional problems, social functioning and bodily pain. They found the poorest quality of life among adults with inoperable cyanotic congenital heart disease.

Studies that have used disease specific questionnaires include McCrindle, Williams, Mital, Clark, Russell, Klein & Eisenmann, (2007) that looked at the reliability of the Congenital Heart Adolescent or Teenage (CHAT) questionnaire in subject aged 10 to 18 years old. The questionnaire was piloted, revised, and then applied to subjects with congenital heart disease attending the pediatric cardiology outpatient clinics in seven paediatric cardiac centres in the United States and Canada. The items and domains showed excellent Cronbach's alpha and construct validity. Unfortunately, the questionnaires are self reported assessment and cover a limited age group (10 to 18 years old).

Macran, Birks, Parsons, Sloper, Hardman, Kind, & Lewin, (2006) used another cardiac disease specific questionnaire for measuring the quality of life, the Congenital Heart Disease Quality of Life Questionnaire (ConQOL). The study was a multicenter study involving 640 children from six pediatric cardiology centers in the United Kingdom. The instrument was developed to measure health related quality of life for children with congenital heart disease within the age of 8 to 16 years old. Two versions are available for the two different age range groups: the first, for children 8-11 years old, it includes the scales, symptoms, activities and relationships, and second, for teenagers 12-16 years old, which additionally include a scale on coping and control. However, this instrument only measured children but not parent proxy report. The instrument is also limited to the age group from 8 to 16 years old.

Marino, Shera, Wernovsky, Tomlinson, Aguirre, Gallagher, & Tong, (2008) conducted a study using the Pediatric Cardiac Quality of Life Inventory (PCQLI). The aimed of the study was to develop an inventory covering a wider age group children from 8 to 12 years old and adolescents aged 13 to 18 years old and versions for patients and parents (Proxy). The questionnaires were designed to discriminate

the different types of congenital and acquired heart diseases. The results of this pilot study, conducted in three cardiac clinics with 655 pairs of parents and patients in United states, showed good content validity, good internal consistency and construct validity of the instrument, with a good correlation between the components that measure the impact of the disease and the psychosocial impact. This inventory covers both children and parent proxy report but it is limited to age group 8 to 18 years old.

The PedsQL^{TM} 3.0 Cardiac Module is different in contexts of having self and proxy-report as well as covering a wide age group from 2 to18 years old. Many studies in the world have used this instrument such as Uzark et al., 2008; Tahirovic, Begic, Nurkic, & Varni, 2010; Menteer, Beas, Chang, Reed & Gold, 2013.

The original English version showed excellent Cronbach's alpha and intraclass correlation coefficient. Uzark et al. (2003) determine the feasibility, reliability and validity of PedsQL^{TM} 3.0 cardiac module English version of Cincinnati Children's Hospital Medical Centre, USA. This study showed good internal consistency reliability coefficient ranging from 0.72 to 0.96.

Uzark et al. (2008) used the PedsQL 3.0 cardiac module to assess the quality of life in Children With Heart Disease in both children and parents. The result showed children with heart disease had the lower quality of life than healthy children across all age groups. Children with more severe heart disease had worse physical and psychosocial quality of life.

Tahirovic et al. (2010) conducted a study using the PedsQL^{TM} 3.0 Cardiac Module in 114 parents of children and 96 children with congenital heart disease. They found that HRQOL was poorest in children with complex congenital heart disease.

The cardiac disease specific questionnaires has been applied in congenital heart disease as well as other types of heart disease such as in the study was undertaken by Menteer et al. (2012). They evaluate health related quality of life in heart failure and post heart transplant patients using PedsQL™ 3.0 Cardiac Module. This study examined mood and health related quality of life in children with HF and compared them cross-sectionally with those of healthy control subjects and heart transplant recipients with good heart function. They found that health related quality of life is impaired in heart failure and post heart transplanted patients.

The PedsQL™ 3.0 cardiac module has been translated into various languages such as Hungarian, Brazilian, Swedish and Spanish languages. This translated PedsQL™ 3.0 Cardiac Module had been used to assess health related quality of life in the cardiac patient. Berkes, Varni, Pataki, Kardos, Kemény & Mogyorosy. (2010) investigated the psychometric properties of the Hungarian version of the PedsQL™ Generic Core Scales and Cardiac Module, they found good reliability and validity of the Hungarian translation of the PedsQL™ 4.0 Generic Core Scales and the PedsQL™ 3.0 Cardiac Module in Hungarian children with heart disease with Cronbach's Alpha exceeding 0.7.

PedsQL™ 3.0 Cardiac Module has also been translated in Brazilian version. Nascimento, Terreri, Hilario & Len. (2013) did a study regarding HRQOL of children with rheumatic heart diseases: reliability of the Brazilian version of the PedsQL™ 3.0 Cardiac Module.

The values of Cronbach's Alpha varied from 0.6 to 0.8 indicating good internal consistency and the intraclass correlation coefficient also reproducible with adequate 0.76 to 0.94 among patient and 0.76 to 0.84 among parent/caregiver.

The reliability of the Swedish version of PedsQL^{TM} 3.0 Cardiac Module was evaluated in a study involving Swedish children with congenital heart disease (Sand, Kljajic, & Sunnegardh, 2013). The result showed, Cronbach's alpha values 0.7 for both child and proxy report.

PedsQL^{TM} 3.0 Cardiac Module Spanish version was also had been published by Gonzalez-Gil, Mendoza-Soto, Alonso-Lloret, Castro-Murga, Pose-Becerra & Martin-Arribas, (2012). Even though it differs from the original version in some respects especially in school functioning and physical appearance, the internal consistency was shown to be higher than 0.7.

In Malaysia, no study has been undertaken to evaluate health related quality of life using PedsQL^{TM} 3.0 cardiac modules. Malay is the major ethnic group in Malaysia and the official language is bahasa melayu. Hence, it is important to develop a validated Malay version of PedsQL^{TM} 3.0 Cardiac Module.

The PedsQL^{TM} 3.0 Cardiac Module was chosen because of the many advantages that includes:

1. It has been validated
2. It is developmentally appropriate for our subjects
3. It is disease specific
4. It is includes both self and proxy-report and covering a wider age group for children 5-18 years old and parent-proxy age 2-18 years old
5. It can be conducted within 15 minutes
6. The questionnaires are multidimensional, including heart problem and treatment, treatment II, perceived physical appearance, treatment anxiety, cognitive problem and communication

7. These questionnaires also had minimal missing data

2.8 PedsQL™ 3.0 CARDIAC MODULE

The PedsQL instrument was designed to measure health related quality of life in children with specific disorders. This enhances measurement sensitivity of health domains relevant to the specific disorders. In contrast, a generic health related quality of life instruments, measures the overall functional health status and well being of the patient and enable comparison of the chronic health condition with healthy populations (Varni, Seid, & Kurtin, 2001).

The PedsQL™ 3.0 Cardiac Module was designed specifically to measure HRQOL in the pediatric population with congenital heart disease. This module has a more sensitive scaling range and additional construct and items, which are specific to cardiac patients compare to the generic core module. The PedsQL™ 3.0 Cardiac Module comprised of six domains, including heart problem, treatment, treatment anxiety, cognitive problems, perceived physical appearance and communication.

2.9 CHILD SELF-REPORT VERSUS PARENT PROXY-REPORT

The self-report HRQOL is considered the gold standard in the adult. However, in pediatric populations, the accuracy of self-report is affected by several factors including cognitive function, language skills and developmental stage (Varni, Seid & Rode, 1999; Cremeens, Eiser & Blades, 2006). Pediatric HRQOL instruments must take into account the child's cognitive function and developmental stage. They also need to look at the perspective of the parental-proxy report.

The parental proxy-report of HRQOL is used to estimate child HRQOL when the child is unable or unwilling to complete the HRQOL measure such as when the child is too young, cognitively impaired, and ill or fatigued to complete the instrument. This parent proxy report is also used when young child self-report scale reliabilities < 70 standards (Varni et al., 2002, 2007c).

Information provided by proxy-reports is not in complete agreement with patient's report (Varni et al., 1999; Cremeens et al, 2006). This imperfect agreement between self-report and proxy-report, is also known as cross-informant variance. Parents' proxy-report instruments need to be used in patients who are unable to self-report such as toddlers (age 2 to 4 years), ill patients, cognitively impaired and those who are unable to provide self-report. The demonstration of cross-informant variance using previous tools indicates the need for a more reliable and valid child self-report HRQOL measurement instrument for the broadest age range possible (Varni et al, 1999, 2002).

The previous study has shown that pediatric patients as young as 5 years old can reliably and validly self-report their HRQOL when an age-appropriate measurement instrument is utilized (Varni et al, 2007a, 2007b). Pediatric patient-reported outcomes should be accepted as the standard for HRQOL measurement in pediatric clinical trials.

Consequently, a reliable and valid parent proxy-report instrument are essential when children themselves are unable to self-report. Parent proxy-report should only be the primary outcome measure when the child is too young or ill or otherwise unable to self-report (Varni et al, 2007c). Even though pediatric patient self-report is preferred;

there remains a fundamental role for parent proxy-report in pediatric clinical trials and health services research.

2.10 RATIONALE OF THE STUDY

Up to date, there is no assessment tool in Bahasa Melayu to measure HRQOL among children with heart disease. The validated tool is available is in English and other languages. Therefore, it is important to produce a translated Malay version PedsQL^{TM} 3.0 Cardiac Module that could be used for the population in Kelantan and Malaysia.

3. OBJECTIVES

3.1 GENERAL OBJECTIVE

- To produce a reliable and validated Malay version PedsQL™ 3.0 Cardiac Module.

3.2 SPECIFIC OBJECTIVES

i. To translate PedsQL™ 3.0 Cardiac Module questionnaires from English version to Malay version.

ii. To assess the reliability of PedsQL™ 3.0 Cardiac Module Malay version.

iii. To assess the validity of PedsQL™ 3.0 Cardiac Module Malay version.

3.3 HYPOTHESIS

- The Malay version of PedsQL™ 3.0 cardiac module is reliable and valid tool for assessing HRQOL in children with congenital heart disease in the Kelantanese population.

4. METHODOLOGY

4.1 RESEARCH DESIGN

This is a cross sectional study. The study was conducted at Hospital Universiti Sains Malaysia (HUSM) and Hospital Raja Perempuan Zainab II (HRPZ II), Kota Bharu Kelantan.

4.2 PARTICIPANTS

The participants were divided into 2 groups:

1. Children with congenital heart disease aged between 5-18 years old.
2. Parents or guardians of children diagnosed with congenital heart disease aged between 2-18 years old.

Children with Congenital Heart Disease were identified from admission registrations in Hospital Universiti Sains Malaysia (HUSM) and Hospital Raja Perempuan Zainab II (HRPZ II). The participants were recruited when they came for follow up in the cardiac and echocardiography clinic.

4.2.1 INCLUSION CRITERIA

1. Patients with congenital heart disease
2. Age of patients from 2-18 years old
3. Parents and guardians of children with congenital heart disease age 2-18 years old