HEALTH RELATED QUALITY OF LIFE IN CHILDREN AGED 2-18 YEARS WITH CONGENITAL HEART DISEASE AT MUHIMBILI NATIONAL HOSPITAL IN DAR ES SALAAM, TANZANIA

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CERTIFICATION

The undersigned certify that they have read and hereby recommend for acceptance for a dissertation entitled: Health Related Quality of Life in children aged 2 to 18 years with Congenital heart disease at Muhimbili National Hospital in Dar es Salaam, Tanzania in partial fulfillment of the requirements for the degree of masters of medicine (Paediatrics and Child health) of the Muhimbili University of Health and Allied Science (MUHAS), Dar es Salaam

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DECLARATION AND COPYRIGHT

I,**Kessy Charles Shija**, declare that this dissertation is my own original work and that it has not been presented and will not be presented to any other University for similar or any other degree award.

Signature…….Date……

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DEDICATION

To my mother Lyabakola Kaduluma and my late father Charles Shija Kabadi, the foundation and support you gave me are appreciated. May God Bless you.

ABSTRACT

BACKGROUND

Congenital Heart disease (CHD) is a prevalent condition worldwide. In Tanzania approximately 7/1000 live births are born with CHD and are among the top ten chronic diseases. Due to its chronicity it affects the quality of life in various ways. The health related quality of life (HRQL) of children with CHD has not been explored in Tanzania. This study is going to demonstrate the HRQL in children with different CHD lesions. It is also going to explore the difference in quality of life among children who had cardiac surgery and compare with those not yet have had cardiac surgery.

OBJECTIVE

To determine the health related quality of life of children aged 2-18 years with congenital heart diseases attending cardiac clinic at Muhimbili National Hospital in Dar es salaam, Tanzania.

METHODOLOGY

Descriptive cross sectional study was done to determine the health related quality of life in children 2-18 years with CHD. Disease specific pedsQL cardiac module was used to collect the health related quality of life. Parents' and childrens' socio demographic features were obtained by using a specifically designed questionnaire. Data entry was done by using Epi Info and transferred to SPSS version 17 for analysis. A p- value of <0.05 in the ANOVA test when more than three groups were compared on the pedsQL^{TM} 3.0 cardiac module mean scores and a student T test for two groups. For statistical significant difference in ANOVA test a further Bonferroni alpha post hoc test with adjusted p- value of 0.0167 was applied to detect the difference among groups.

RESULTS

A total of 107 children aged 2-18 years with CHD were recruited by convenient sampling of which fifty seven (53.3%) were female.VSD 34.6% was the

commonest CHD. Eight percent of children were found to have co morbid conditions including; Down syndrome, epilepsy and speech disorder.

The overall mean scores in all the risk factors assessed were below the cut off score 69.7 of HRQL meaning poor HRQL. There was a significant difference in physical functioning domain of HRQL in the parent report stratified by disease severity (group 1,2 and 3). Multiple comparison test (Bonferroni adjustment) revealed significant difference between group 1 and 3 (mean score of 72 ± 14 against 59 ± 18 and p value 0.0008). Moreover significant difference was also noted in the cognitive domain between group 1 and 2 in the child report (mean scores and standard deviation of 86 ± 15 against 46 ± 4 with a p value of 0.00126) Children with cardiac surgery had better physical functioning compared to those without cardiac surgery with mean scores of 71 ±15 against 64± 18 and a p value of 0.03.Poor physical appearance was noted in children who had cardiac surgery compared to those not yet have had cardiac surgery, their mean scores and standard deviations being 82 ± 20 against98 ±4 with a p value of 0.011. In the multivariate analysis both cardiac surgery and CHD lesion were the predictors of poor physical functioning, beta coefficient of -0.29 with p value 0.00 and -0.42 , p value of 0.00 respectively.

CONCLUSION

From the findings of this study it may be concluded that children with CHD lesion had poor HRQL. The impact of CHD on the HRQL of the children is significant particularly in the domain of physical functioning and cognitive development. With increased access to cardiac surgery in this country more children will survive into adulthood, however CHD will take its toll on quality of life particularly in the areas of cognitive development and physical functioning.

RECOMMENDATION

A prospective cohort study is needed to explore the HRQL before cardiac surgery and after surgery, more emphasis on cognitive development in order to establish a temporal relationship.

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LIST OF ABBREVIATIONS

AS	AORTIC STENOSIS
ASD	ATRIAL SEPTAL DEFECT
AVSD	ATRIOVENTRICULAR SEPTAL DEFECT
CHD	CONGENITAL HEART DEFECT
CXR	CHEST X RAY
DHCA	DEEP HYOTHERMIC CIRCULATORY ARREST
DS	DOWN SYNDROME
ECG	ELECTROCARDIOGRAM
HLH	HYPOPLASTIC LEFT HEART
HLHS	HYPOPLASTIC LEFT HEART SYNDROME
HRH	HYPOPLASTIC RIGHT HEART
HRQL	HEALTH RELATED QUALITY OF LIFE
MD	DOCTOR OF MEDICINE
MoHSW	MINISTRY of HEALTH AND SOCIAL WELLFARE
MUHAS	MUHIMBILI UNIVERSITY OF HEALTH AND ALLIED SCIENCES
OHS	OPEN HEART SURGERY
OPD	OUT PATIENT DEPARTMENT
PedsQL	PEDIATRIC QUALITY OF LIFE
PDA	PATENT DUCTUS ARTERIOSUS
PS	PULMONARY STENOSIS
SD	STANDARD DEVIATION
SF-36	SHORT FORM 36
SD	STANDARD DEVIATION

TA	TOTAL ANOMALY,TRICUSPID ATRESIA, TRUNCUS ARTERIOSUS
TGA	TRANSPOSITION OF GREAT ARTERY
TOF	TETRALOGY OF FALLOT
UDSM	UNIVERSITY OF DAR ES SALAAM
USA	UNITED STATES OF AMERICA
VSD	VENTRICULAR SEPTAL DEFECT
WHO	WORLD HEALTH ORGANISATION

CHAPTER ONE

INTRODUCTION AND LITERATURE REVIEW

Definition of health related quality of life

The term "health-related quality of life (HRQOL)" refers to the physical, psychological, and social domains of health, which are influenced by a person's experiences, beliefs, expectation and perceptions[1]It is a reflection of the way patients perceive and react to their health status and to other non medical aspects of their lives.[2, 3]The World Health Organization (WHO) defines health as "a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity". [4]

Quality of life Measurement tools

The ability to cope with limitations and disability to a great extent affect a person's perception of health and satisfaction with life. Therefore two people with the same health status may have very different qualities of life.[5]Assessment of HRQL is a difficult process because it is dynamic and varies with ages, locations, beliefs and disease type.

This diversity in perception has lead to development of specific measurements by researchers. Several instruments exist which are used to measure quality of life in various disease conditions some of them are disease specific. For cardiac disease assessment tool the existing tools includes; Short Forms-36 (SF-36) and paediatrics Quality of Life Trade Mark (pedsQL^{TM}) cardiac modules.

The pedsQL^{TM}cardiac module is specific for cardiac conditions.[1, 6] It is designed to measure HRQL in children and adolescents aged 2-18 year and their parents. It was developed from expert suggestions not from children's own perceptions. The design of the tool is good especially for specific condition like cardiac diseases. It enables the investigators to use objective (clinical) assessment together with a patient's feeling (subjective). This helps to reduce over/under estimation of patients' perceptions and feelings. The development, refinement and linguistic

validation have been done in a number of European countries and other countries. A number of papers have been published in many languages. The tool has been used in studies with congenital heart diseases in several developed countries and also can be applied in Tanzania. It is simple to administer and compute, with few missing subjects and has been validated in 30,000 children universally. It has six dimensions namely heart problems (physical functioning or symptoms) and treatment II for those with heart treatment, physical appearance, treatment anxiety, cognitive problems and communication[6].

It measures perception of quality of life for parents of children with CHD and the children themselves. Cognitive development and communication assessment is done for children above 8 years as those under 8 years lack the abstract thinking and language skills necessary. Children 8-18 years are able to respond to questions on the study instrument while for those below 8 years parents are assessed.

Health related quality of life and associated factors in children with CHD

Survival from CHD is not always equivalent to high quality of life.[7]Globally HRQL in children with CHD is an emerging concern, largely because children now survive longer due to improved modern surgical interventions. [8]But their survival does not assure good quality of life.

Studies have revealed that children with CHD experience complex difficulties in physical, psychological (emotional, behavioral, and social) functions [5] and cognitive development.[7] They are unable to perform important social roles, have reduced school performance, as well as anxiety and depression.[7] They have the feeling of being different from peers, encounter problems in obtaining employment and insurance when they reach adulthood.[9, 10] In the course of their illness, they may also experience symptoms of the disease and restrictions in the level of activity resulting in a poorer prognosis.[9-11]If these abnormalities are not diagnosed and managed early it can lead into significant psychological morbidity in later life.[9]

A range of factors when coupled together hasten poor HRQL in children with CHD and include; socio demographic characteristics of the patients or parents/guardians (age, sex, socio demographic features of the parent and occupation), severity of cardiac defect and cardiac surgical interventions.[7, 12]

Socio demographic characteristics

Physical appearance, the changes in body images(finger clubbing, surgical scar, forward protrusion of the chest and delay in physical growth(i.e. height and weight)of children with CHD can be perceived in a different way by the society.[7]The child and parent may not be happy by the way the child appears or the delayed growth especially when compared to other peers. It is known that, major physical illnesses usually have an impact on the psychosocial well-being of any individual.[9]The majority of children with CHD have physical abnormalities. The abnormalities may be interpreted wrongly by the parents/guardian, society or even the patients themselves. Good information from health care workers can help to reduce misperceptions. If they are not well informed, patients and their families can experience some degree of psychological disturbance and therefore poor quality of life. The understanding capacity of parents has a big role to play in the care of a child with CHD. Parents of low socioeconomic status and low level of education have little knowledge on early health seeking behavior. They have tendency of delaying in looking for health services. This delay in seeking health services may worsen the child's condition resulting in poor HRQL.[7, 12]

Other socio demographic factors associated with impaired HRQL in children with CHD includes; type of sex and age of a child, acceptability of the disease, as well as personality features.[7, 13]

It has been found that children of young age are at a higher risk of poor HRQL[13, 14] compared to older age. Children of young age and their families might have no clear understanding of the causes and the outcome of their disease condition. Little understanding about the disease and its consequences, affects the child

psychologically. They suffer stress related to symptoms of the disease, the treatment and its outcomes. There is slight good awareness of the condition for older children because the patients and their parents themselves are used to the situation and therefore develop psychological adjustment which helps to reduce the stress associated with children's condition. They have adequate knowledge how to handle the situation and have accepted it. Therefore, acceptance of the condition depends on the understanding capacity of the patients and their families. Parents and families who do not accept the condition at the end experience a more stressful life than those who understand and accept the situation.

Gender is also reported to be associated with poor HRQL but findings are mixed. Some studies have found male children to be at higher risk of anxiety and behavioral disorders while others report do not. Female children were found to have poor HRQL compared to their male counterpart.[13] But male children with CHD may have psychological disturbance because of more community exposure. In many cultures boys start interacting earlier than girls. The physical restrictions due to the disease result into being isolated by peers. This separation affects the child psychologically and may lead to anxiety and behavioral disorders.

Cardiac defect

Severity of the disease is also found to be associated with impaired HRQL. Children with end-stage heart, or severe heart lesion are at increased risk of cognitive and behavior problems.[15, 16] Studies have shown that patients with severe cardiovascular diseases have poorer quality of life compared to those with mild cardiovascular diseases.[7, 9-11, 16-18]

Early diagnosis, outcome of the disease, the socio-economic and educational status of the parents are all associated with poor quality of life. An early onset illness such as CHD necessitates frequent diagnostic and therapeutic interventions. This can adversely affect the emotional balance and behavioral adaptation of children

and adolescents.[7] Mother and infant interaction start early in child development, so early diagnosis of CHD may affect that bond leading to psychosocial adverse effect to both parents and children.

Children with chromosomal abnormalities that are associated with CHD such conditions include Down syndrome (DS) (trisomy21) and deletion of chromosome 22q11 (Di George syndrome). The chromosomal abnormalities are also associated with mental deficits. Those with cyanotic CHD, the hypoxia and the syndrome itself negatively impacts on the neurodevelopment of the child.[19, 20] For acyanotic CHD, cardiac failure/collapse and repeated chest infection can impair the neurodevelopment of the child.[20] Again severe metabolic acidosis secondary to hypoxemia strengthens the negative effects of ischemia on the glial and cerebral vascular cells due to action of free radicals on membrane lipids and proteins.[20] All these factors collectively can have negative effects on the daily life of an individual leading to recurrent infections for those with heart failure, chronic effects of malnutrition, failure to thrive, frequent admissions and later on poor quality of life. Malnutrition itself in the long run can lead to failure to thrive while poor nutrition if happens during infancy may affect brain growth adversely and later recovery from surgery.[21]Therefore hypoxemia, hypoperfusion, and congestive heart failure may have significant long-term consequences in the prenatal and postnatal periods and are likely to contribute to preoperative neurologic injury.[20-22]The long term effects of these hemodynamic abnormalities are impaired motor function, inability to sustain attention, and low academic achievement.[23]

Cardiac surgery

Cardiac surgery improves both survival and quality of life even in severe cyanotic

Congenital heart disease (TOF or TGA). This has therefore resulted in havingnormal professional and peaceful family's life after correction. In one study about 90-95% of participants survived 30-35 years after surgery.[24]

Prior to the advent of surgical treatment for congenital heart malformations in the 1960s only less than 20% of children born with such lesions survived to adulthood, the majority dying in the childhood or adolescent periods.[25] The cause of death in that period was due to bleeding because the operation had to be performed while the heart is pumping blood.

The introduction of cardiopulmonary bypass and hypothermia procedures in 1950s has improved survival. Successful open heart surgery was performed in 1952 by using hypothermia.[25] This technique has helped to reduce the complications associated with bloody operation and the mortality rates have gone down significantly. In developed countries it is now reported that 85% of children reach adulthood and the majority of deaths from congenital heart disease (CHD) occur beyond 20 years of age.[26]

Following this advanced successful surgery the disease has now been transformed into a chronic condition. The chronicity of the disease has now led to emotional suffering of children with CHD and their families.[19] The parents of children with congenital heart disease (CHD) face a variety of stressors that are related to the nature of the disease and its treatment, especially when their child undergoes open heart surgery. It is reported that parents in whom the child's disease has a high impact on their family life are at increased risk for persistent low mental HRQL.[27]

Other complications related to cardiac surgery are neurodevelopment deficits.[20, 22, 23, 28]Patients post cardiac surgery may experience early and late complications. These complications can be precipitated by various factors which are classified into fixed and modifiable factors. Fixed factors include many variables specific to the individual patient, including genetic predisposition, gender, race,

socioeconomic status, and in uteral central nervous system development. Modifiable factors include not only intraoperative variables (cardiopulmonary bypass, deep hypothermic circulatory arrest, and haemodilution) but also such variables as hypoxemia, hypotension, and low cardiac output.[22]Neurodevelopment deficit largely depend on disease severity, number of hours or minutes used for operation and type of heart lesion.

Hence the growth of children with congenital heart diseases are often characterized in literatures as "abnormal" because of the demanding lifestyle changes imposed by the cardiac deficit, the frequent hospital admissions, abstention from pleasurable activities, isolation from the friendly environment.[7] Specific aetiopathogenic factors potential to cerebral injury and poor neurodevelopment in children with CHD are cardiac defect and surgery as will be explained below.[20, 23, 28]

Intraoperative risk factors

Early surgery during the neonatal period or in the first month of life has been shown to prevent cerebral complications especially cyanotic CHD like TGA. The early surgery will minimize the time of hypoxia and persistent metabolic acidosis and forestall blue spells. In comparison to acyanotic CHD no unusual risk of cerebral injury is present in the absence of severe cardiac dysfunction. However surgical corrections of the majority of CHD involve methods like cardiopulmonary bypass and deep hypothermia which reduces bloody procedure and protect cerebral structures. The hypothermia protects the brain from death by reducing blood supply, on the other hand it has potential adverse effect to the brain's normal function.[20]Cardiopulmonary bypass causes embolic complications and hypo perfusion due to low-flow cardiopulmonary while deep hypothermic circulatory arrest (DHCA) in prolonged duration more than 30 minutes has the risk for neurologic injury. Both longer duration of total circulatory arrest and occurrence of preoperative seizures are independent risk factors for poor

neurodevelopmentoutcomes at ages 1 and 4 years. A possible mechanism for post circulatory arrest brain injury is suggested by experimental reports showing that the release of excitatory neurotransmitters such as glutamate seem to be responsible for delayed cell death. Dissemination of both macro emboli and micro emboli (gaseous or particulate) may also cause brain injury.[21, 28] Therefore cardiopulmonary bypass and different methods of cardioplegia can cause developmental and cognitive functioning impairments.[19]

Post operative risk factors

It has been noted that in the immediate postoperative period, cerebral perfusion and oxygen delivery may be compromised leading to neurological abnormalities. In the post operative periods factors which lead to low cerebral blood flow includes; low cardiac output, diminished arterial partial pressure of oxygen, and altered cerebral blood flow. The consequences of these complications are hypoxic cerebral injury. Hypoxic cerebral injury is facilitated by deep hypothermic circulatory arrest (DHCA) which impairs the normal auto regulation of cerebral blood flow. Other factors which can additionally influence developmental outcome are congestive heart failure and prolonged hospitalizations which are the common complications of patients with CHD.[21, 23, 29] The seizures and/or stroke due to intraventricular hemorrhages may occur before, during or after surgery and nutritional deficiencies are among other factors in the post operative periods affecting the patient's development.[23, 29]

So the surgically repaired heart has satisfactory outcome, but this heart is not guaranteed to be anatomically normal. Patients after surgery may experience physical and psychological problems which has negative impact on the quality of life. They have risk of suboptimal developmental outcomes.[7, 19] The residual anatomical abnormalities may remain and patients may still be at risk for premature death or co morbidities, such as arrhythmias, hypertension, pulmonary, renal, and myocardial disease or coronary artery disease[16]. For example the TOF

repair has been reported to lead to variable degree of devaluation of the pulmonary outflow tract, a scar on the right ventricle, a patch repairing the ventricular sepal defect and scars on the atrium (cannulation for cardiopulmonary bypass). These sequellae expose the patients to a number of physiological complications, notably arrhythmic and sometimes haemodynamic, affecting the right ventricle.[24]

It is important to understand these complications for proper follow-up regularly by a cardiologist. Sometimes there may need second operation. In US almost 50% of the patients who have undergone surgery require additional surgery at an older age [16],which in this sense adds another risk of neurological complications and impairing the general wellbeing of patients.

Children development and health related quality of life manifestations

The neurodevelopment and cognitive functioning abnormalities may manifest as behavioral disorders such as attention deficits, hyperactivity[9], emotional liability and coordination problem.[21] Children may also show physical (the ability to coordinate skills and perform complex operations) and narrative language abnormalities.[21] Childhood is critical moments for learning, patients may experience problems such as difficulties obeying their parents, taking their medications, or attending periodic follow up which are not medical problems related to their diseases.[20] They may also have neurological deficits which include hypotonic, pyramidal findings and asymmetry of tone. There may also be features of hypertonic, decreased levels of alertness and higher incidence of feeding difficulties.[23] The manifestation depends on the stage of development of the patient.

Infancy age

At this period both mothers and children with CHD are at risk for a disturbance in relationship because of early hospitalization and cardiac surgery. Studies have shown that fetal diagnosis of CHD and post natal experience can negatively

influence the type of attachment between mother and her infant. The severity of illness does not have a direct effect on Quality of life of the infant-mother relationship.[20] The more prevalent problems are feeding difficulties and delays in reaching some motor milestones such as rolling over, crawling or walking. The motor skills delay are extremely common, most milestones are only delayed by a few months. Those with congestive heart failure commonly have increased metabolic demands resulting into failure to thrive and poor nutrition, which may affect brain growth adversely during infancy and impair recovery from surgery.[21]

Pre school age

In this age group the main delayed milestone is speech and language. The receptive language is normal but the expressive language may be delayed. In addition to difficulties with expressive language, some preschool children with complex CHD (probably less than one fourth) have continued difficulties with motor skills, including large motor (clumsiness) and fine motor (drawing, cutting) delays. However in most children the problems may improve by the time they enter school.

School age

School age children have repeated absence from school due to frequent hospitalization**.** Their main problem is Attention deficit/hyperactivity disorder (ADHD). At this age the child may experience features of depression (feeling of being abnormal, like he/she was not made to live, isolating from peers). Among children with complex CHD, the ADHD may even be more severe and is reported to affect up to one third of children. Recently, it has been observed that children have problems with “visual motor integration” and “executive planning”. Visual motor integration relates to the ability to coordinate thoughts and images into action. There is failure in these patients to coordinate their thought which has great impact in the subsequent life. School age children, have challenges in learning handwriting, seeing handwriting on the board, knowing that it's a particular letter,

and getting their hand to make the letter can be very frustrating to an otherwise bright child. The children with CHD experience difficulty with executive planning, for example in the planning process of daily activity of the school age child proceeds as follows: *"First I will get dressed, then I will go eat breakfast, and then I will pack my bag, and then head off to school".* In CHD as the number of tasks increases, it becomes increasingly difficult to coordinate. Therefore some children with CHD have a particular problem in this area, but the exact frequency of this problem is currently unknown.[29] If the child shows learning problems it is important to evaluate him or her using appropriate cognitive assessment test.

The onset of sexual maturity and the difficulty accepting the physical aspects of their condition in operation, exacerbate the discomfort. Moreover they do not accept the limitations related to their physical status and therefore need psychotherapeutic support.[20]

Burden of Health Related Quality of Life

Health related quality of life disorders are prevalent among the survivors of CHD, and a significant percentage of children require remedial academic assistance.[21] In a meta-analysis study on the psychological and cognitive functioning in children and adolescents with CHD showed that patients with CHD exhibited internalizing (anxiety, depression and social withdrawal) and externalizing (hyperactivity, oppossional behavior and aggression) behavior problems than normal population. In this analysis some studies showed age being the risk factor, thus older patients with CHD showed more overall, internalizing, and externalizing problems as compared with a comparison group. The severity of CHD also was found to be associated with impairment of quality of life such that severe CHD exhibited lower cognitive functioning (performance intelligence) than patients with less severe CHD. For example cognitive functioning of patients with ASD was within the normative range but patients with hypoplastic left heart

syndrome (HLHS) and TGA had significantly lower cognitive functioning than normative.[16]

A study in Sweden on the outcome of open heart surgery reported a depressed mood in the majority of patients during recovery after Open heart surgery (OHS) and had a long-term effect on their well-being. In that study depressed mood occurred in 52% of the patients during recovery or 3 years after OHS showing that depression was not transient but may be permanent if not addressed.[30]

The Swiss study on Predictors of Parental Quality of Life after Child Open Heart Surgery showed that parents' mental HRQL is low in the immediate period after their child's open heart surgery with score of 52.0% >1SD below the mean .This decreased more at six months post surgery to 24.1% for the mother. They normalized at six months post surgery in other domains. Therefore the HRQOL is affected differently in different domains for individuals and some may have long term consequences but many recovers gradually not less than three months.[27]

The health related quality of life in Polish children with mitral valve prolapse found only one domain the physical was reduced in the studied individuals compared to the control (normal). Its mean score was 59.2 compared to 74.2 in the patients and the health (control) respectively.[31]

In the Netherland study among children aged 8-15 years with CHD obtained significantly lower mean scores on motor functioning, cognitive functioning, and positive emotional functioning than reference peers, reflecting an experience of poorer functioning. In these study children aged 8-11 years had lower mean scores on 5 of the 7 scales than reference peers. But a difference existed among different age groups such that 8-11 age group had a lower score on positive emotional functioning than 12- to 15-year-old CHD children.[14]

In another study on the association between early outcome, health-related quality of life and survival after elective open heart surgery reported that one year post surgery all parameters of health-related quality of life were improved, however

social and mental domain were the least in score with +5.1% in social and +5.1% in mental while general health perception was +19.4%.[32]

The USA study on quality of life in children with heart disease as perceived by children and parents self-report, reported mean PedsQL scores for children with cardiovascular disease were significantly lower than healthy child norms for physical and psychosocial functioning. The scores on psychosocial quality of life were significantly impaired by 21% of children ≥ 8 years of age. The severity of CHD also affected differently HRQOL thus children with less severe cardiovascular disease had less impairment on psychosocial quality of life compared to those with severe CHD.[17]

A Bosnia and Herzegovina study on the severity of congenital heart defects and its affect on disease-specific health related quality of life in children found that the severity of CHD is associated with poor HRQL. In this study CHD was categorized as mild, moderate and severe according to the type of heart lesion and its severity. Thus mild severity included ASD, VSD, AVSD, PDA and Aortopulmonary artery shunt. Moderate severity was those with obstructive anomalies such as obstruction of the pulmonary circulation, obstruction of the systemic circulation which included pulmonary stenosis, mitral valve stenosis, aortic stenosis and coarctation of aorta. The severe ones were those with complex anomalies with enlarged pulmonary circulation, they included TGA, total pulmonary venous connection, univentricular heart,TOF and pulmonary atresia. According to this study mild severe CHD had statistically significant, better HRQOL scores for the heart problems and treatment scales, perceived physical appearance, treatment anxiety, cognitive problems, and communication scales in comparison to the moderate and severe CHD.[18]

A study which was assessing functional limitations in young Children with Congenital Heart Defects after surgery reported functional difficulties in daily living skills in 40% of individuals and 50% had poor socialization. The domains

which were reported to be below the normal population were self care(84.3+23.8 or 84.3-23.8),mobility (77.2+30.0 or 77.2-30.0),and cognition (92.4+27.8 or 92.4-27.8).An overall quality of life was 83.8+23.4 or 83.8-23.4.Mean score for daily living skills was 84.4+17.6 or 84.4-17.6 and socialization 80.3+15.9 or 80.3-15.9.[33]

Health related quality of life comparison study in young adults with minor congenital heart disease (CHD) and health individuals of the same age group found substantial part of the CHD-patients experienced social impediments, i.e. at school (19%),during free time (15%),in choosing (13%) or performing job (9%),with medical examinations for job/insurance (19%) and taking out a life insurance policy (8%).[34]

In the article which summarized a research on neurologic injury in children with CHD and the factors that influence outcome demonstrated that neurological and cognitive outcome differ depending on the severity of the congenital heart disease. Children with severe congenital heart disease have developmental sequelae include mild problems in cognition, attention, and neuromotor functioning. The Caregivers need to be aware which patients are at highest risk.[23]

In a systematic review on psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease concluded that a significant proportion of survivors of open-heart surgery for CHD are at risk for psychological maladjustment and impaired QoL. In this review of previous studies the reviewer looked whether the type of congenital heart disease have effect on the quality of life, it was found that it did not relate to QoL in a heterogeneous diagnostic sample. However, two studies found a lower QoL in children with more complex malformations. In a sample of children with single ventricle anatomy only few cardiac-specific factors were found to be related to QoL. Also in this review study children of parents who remained unemployed due to the child's health condition or children of families with a low income experienced a lower QoL than controls. But also , adverse family relationship and

parental stress at follow up were both found to be negatively related to the psychosocial dimensions of QoL.[12]

A study in Egypt on quality of life among parents of children with congenital heart disease by using SF 36 subscales parents of children with heart disease compared to those with minor illness the HRQOL were significantly poorer in all subscales they assessed except pain subscale. This shows that a chronic illness has an impact not only to the patient but also the parent compared to minor illness. The subscales comparison between parents of children with Heart disease and the parents of children with minor illness were vitality subscale (39.66 vs. 75.81) general health (46.25 vs. 73.15) and role physical (39.53 vs. 61.81). The physical functioning (75.76 vs. 79.84) and social functioning (93.63 vs. 99.88) were the lowest although significant. Severity of illness, type of heart disease in addition to age of child, having multiple children, financial situation and presence of co morbid condition had significant impact on HRQOL of the parents.[13]

Congenital Heart Disease

Congenital heart disease, also known as congenital heart defect, is a general term for a series of faults in the structure of the heart and great vessels that is present from birth.[25, 35] May involve arteries, valves, coronary and major vessels of the heart and can be either simple or complex.

There is no universally accepted nomenclature for congenital cardiac malformations. Much of the controversy results from failure to distinguish the structural connections of the heart from the morphology and spatial relations of its components.[36]For simplicity may be classified into two-, non cyanotic and cyanotic congenital heart diseases.[25, 37] The acyanotic CHD are characterized physiological by normal amount of oxygen in arterial blood and normal skin color, whereas cyanotic are characterized by reduced oxygen in arterial blood and cyan skin color.[37]Among the cyanotic CHD are the tetralogy of fallot (TOF),Transiposition of great arteries (TGA),Tricuspid atresia (TA),total anomaly

(TA),pulmonary stenosis (PS) or atresia, hypoplastic left heart (HLH),hypoplastic right heart (HRH)and truncus atresia (TA). The acyanotic congenital heart diseases include the patent ductus ateriosus (PDA),defective septum such as atrial septal defect (ASD),ventricular septal defect (VSD) and obstructive defective of blood flow such as pulmonary stenosis (PS),aortic stenosis (AS),coarctation of aorta and ebstein anomaly.[25, 37]

Epidemiology of CHD

Congenital heart disease is a common birth defect with great variation in incidence. It varies from 4.2 to 12.3per 1000 live births.[38]The incidence of moderate to severe CHD is estimated to be 6/1000 live births, based on 62 studies from different countries (US, Europe, Australia, and China).[39, 40] It is reported that the rate rises to 19/1000 live births if the potentially serious bicuspid aortic valve is included and to 75/1000 live births if tiny muscular ventricular septum defects (VSDs) and other trivial lesions are included.[39]

Among the CHD, the VSD is the most commonly diagnosed congenital heart defect with the proportion of 30-35%, that is about one-third of all cases and it is seen almost three times as often as ASD and PDA. The next in frequency of occurrence are ASD and PDA with the incidence of 10% of all CHD. The pulmonary stenosis (PS) and coarctation of aorta have all together the incidence of 7% of all the CHD. While the aortic stenosis(AS) and TOF have incidence of 6%.Transposition of great arteries has incidence of 4% of all congenital defects.[37]

In Sub Saharan Africa the prevalence of CHD is reported to be higher than other countries. In South Africa prevalence of CHD is reported 5-7 children per 1 000 are born with a congenital heart disease.[41] In Tanzania the prevalence of CHD is unknown currently. However the previous study of 1976, the incidence of CHD was reported to be 7 per 1000 live births.[42] In 1989 prevalence of CHD among children aged 1 month to 15 years at Muhimbili Medical Centre was found to be 0.35%.[43] In that study VSD was found to be the commonest (44.6%) CHD,TOF

12%,PS 9.6%,ASD 7.2%,PDA 6.0%,AVSD 4.8% and others 12%. A one-year cumulative number of patients who underwent heart surgery at MNH in 2008-2009, children below 17 years were 57(54.3%) which included those with CHD and acquired Heart diseases. The frequency of CHD in that survey was found to be 35.2%.The PDA ranked the highest with a frequency of 17.1% of all operated cases.[44]

Management of heart disease in Tanzania

The children with CHD are referred from the regional or district hospitals of Tanzania to MNH. They are evaluated and their defects confirmed by Echocardiogram. The current capacity to perform Cardiac surgeryis limited and only few conditions of PDA and acquired heart diseases the surgeries are conducted in the country and almost all at MNH. Majority of patients who meet certain criteria are referred abroad. Referral abroad is facilitated by the MoHSW of Tanzania.

However the Government of Tanzania is now constructing a unit for open heart surgery at MNH. With this establishment we hope more patients will have access and benefit from such services.

PROBLEM STATEMENT

Congenital heart disease is a common condition globally and in Tanzania. The incidence of CHD in 1976 was 7/1000 of live births.[42] The prevalence of CHD varies from 0.35% in children of 1month-15 years in 1989[43] to 35.2% of below 17 years in 2010.[44]A total of 375 -529 children attend MNH OPD, 80-120 due to CHD , the rest are for other disease conditions). According to patients referral abroad in 2004- 20066, 105 children with CHD were scheduled for cardiac surgery abroad while as many as 708 were scheduled in 2008-2012at MNH(data from records). This increase in patient referral might be due to increase in diagnostic technology by the doctors and easy access of patients to the health facilities.

The newer diagnostic modalities and the rapid development of cardiac surgical techniques, in conjunction with advances in medical technology have improved the longevity of children and adolescents with congenital heart diseases. With continuous improvement of surgical techniques and with the infant operative mortality rates having decreased from 50% to nearly 15%, a large number of children with surgically palliated forms of congenital heart diseases are likely to survive into adulthood worldwide.[9]

Children born with CHD often require surgery, hospitalization and physical restriction in their behavior in order to prolong their life. The family is acutely aware of the possibility of death of their child. Therefore, the potential for psychological and emotional distress is prevalent.

In developed countries studies have shown that children with CHD experience impaired quality of life even after surgery. They are at higher risk to develop emotional and behavioral problems. Also have more medical fears, and more physiological anxiety than normal peers. They may also show an increased feeling of inferiority and basic anxiety and a more impulsive pattern of behavior. They also have low self-esteem, depression and are at particular risk for poor school adjustment.[7, 9]Withdrawn aggressive behavior, somatic complaints, depression

and anxiety are seen in children with congenital heart diseases.[21]Therefore HRQL of a child with CHD is an emerging concern globally.

In Tanzania research is essential in this area so as to explore 'quality of life' and design intervention to ensure the best possible quality of life psychologically and socially for children with CHD in Tanzania.

RATIONALE OF THE STUDY

No study on quality of life specifically *health related quality of life'* in children or adults with CHD has been done in Tanzania. As a pediatrician my concern is primarily regarding the HRQL of children with CHD. This study will provide information on HRQL of children with CHD both those who had cardiac surgery and those who have not yet had surgery. Further-more the study is going to compare HRQL according to cardiac defect type and its severity.

Information on HRQL will help clinicians gain a better understanding of relevant issues to consider in the care of these patients, which is essential for optimizing their clinical management, planning appropriate care, and evaluating specific interventions or therapeutic modes. There is evidence that early detection and treatment especially cardiac surgery can significantly minimize the negative impact of CHD lesion on the child's HRQL. Further- more the study is going to compare the HRQLof children who had cardiac surgery to those without cardiac surgery.

Awareness of the impact of severe CHD on cognitive development will inform educators of the vulnerability to delays in thinking affecting their academic performance. This delay in cognitive development is partly due to chronic hypoxia for those with cyanotic CHD. It can also be due to constant interruptions in education and emotional distress which hinder them from active interaction with their environment. Therefore teachers must be aware so as to provide a good learning environment and if possible make special programs available for these children.

Study Hypothesis

1 The HRQL of children with CHD in Tanzania is poor because of frequent hospitalization, symptoms related to the disease and frequent separation from the peers due to the illness.

2 Poorer HRQL in children with severe cardiac defect as compared to children with mild (simple) defect due to the complexity of the defect results in more symptoms, impairment in neurodevelopment and more admissions than simple CHD

OBJECTIVES

Broad Objectives

To determine the health related quality of life of children aged 2-18 years with congenital heart diseases attending cardiac clinic at Muhimbili National Hospital

Specific Objectives

1: To determine the relationship between social demographic characteristics of children with CHD lesion (sex and age of the child; occupation and education of the parent/guardian) and their HRQL

2: To determine the impact of cardiac surgery on HRQL

3: To determine the impact of type of heart lesion on HRQOL

CHAPTER TWO

METHODOLOGY

Study design

A descriptive cross sectional study was conducted to determine the health related quality of life in children 2-18 years with CHD.

Study area

The study was carried out at MNH in Dar es Salaam at the paediatric cardiac clinic. Muhimbili National Hospital is the national referral and tertiary level facility.. The hospitals in Dar e s Salaam and Tanzania as a whole refer children with CHD to MNH in patient or outpatient services. The hospital runs a paediatric cardiac clinic on a weekly basis by the two paediatric cardiologists. Out of 375-520 children seen at the OPD paediatric clinic on average 80-120 children are diagnosed with CHD per month. About 2-5 are new referrals and 5 post cardiac surgery patients are seen at each weekly clinic. Currently, cardiac surgical conditions which are operated at the hospital are PDA and acquired heart diseases. Due to limited facilities for cardiac surgery, most of patients with complex CHD are referred abroad after confirmatory diagnosis and stabilization. In 2008-2012, about 708 children with CHD were evaluated and planned for referral abroad (data from MNH records).

Study population

The study involved parents and children of 2-18 years with congenital heart disease attending the paediatric cardiac clinic at MNH who fulfilled the inclusion criteria.

Inclusion criteria were:

- All children2-18 years with confirmed CHD by echocardiogram.

- Those with cardiac surgery at least 3 months post surgery at time of recruitment. This was to allow for enough time for physiological adjustment post surgery,
- Parents/guardian who consented to participated in the study.
- Children who assented to participate in the study.

The age of participants chosen for the study was 2-18 years because the evaluation tool used to assess HRQL was designed for this age range.

Study duration

This study was carried out over a period of 11 months from April 2011 to March 2012.

Sample size

The sample size was calculated using the formula below

$n=(1.96\sigma/d)^2$

Whereas

n is the sample size estimated

d is the degree of precision

σ is the standard deviation

For this study d of 3% was used

The σ varies according to age groups[17]

σ 1 – 13.21 for 5-7 years

σ 2- 15.37 for 8-12 years

σ 3- 13.51 for 13-18 years

Sample sizes n1,n2 and n3 were calculated for the various age groups and the mean n was taken.

N was 84 with adjustment of 20% of no response the final calculated sample size was 100.

A total of 107 participants were enrolled.

Sampling technique

The sampling technique was convenient sampling. Children aged 2-18 years were recruited after signed written informed consent was obtained from their parents or guardians. For the children who were more than 8 years assent was also obtained. The sampling was aimed at recruitment of children with or without cardiac surgery at a ratio of 1:1.

Initially the patients were assessed at each clinic day to check for eligibility to participate in the study. The patients who fulfilled the inclusion criteria were approached individually with their parent/guardian.

Research tools

pedsQL™ 3.0 cardiac module

We used PedsQL™ 3.0 cardiac module to obtain participant's information about HRQL. The PedsQL 3.0 Cardiac Module is a multidimensional instrument having 5 domains related to; symptoms (7 items), perceived physical appearance (3 items), treatment anxiety (4 items), cognitive problems (5 items), and communication (3 items) for parent proxy report and children 8 to 18 years. Additional domain of treatment II (5 items) assess compliance for children who were receiving cardiac medicine. The tool is brief, very easy to use, has minimal missing data and includes both parent and child reports.

The interview was conducted to both parents/guardian and children. But only to children above 8 years interview was done to them. For children less than 8 years whose communication ability and cognitive development are less developed interviews were done only to their parents.

A 5-point Likert scale was used across child self-report for ages 8 to 18 years and parent proxy report (0, never a problem; 1, almost never a problem; 2, sometimes a problem; 3, often a problem; 4, almost always a problem). Then the items were reverse-scored and linearly transformed to a scale from 0 to 100 points i.e. 0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0. Higher score means the better the QoL.

Before using the tool user agreement was signed with MAPI Research Institute, France, through email.

Special designed questionnaire

The parent and child socio-demographic characteristics including age and sex of the child, level of education and occupation of the parents/guardian were also obtained and entered in a special designed structured questionnaire.

Procedures

For each patient, history was taken and thorough physical examination was performed. Physical findings were compared with echocardiography findings. For challenging patients cardiologist was consulted and when necessary a repeat echocardiography was done.

The patient's file was used to obtain details of type of CHD according to echocardiography findings and to verify that a child has had cardiac surgery or not. For those with cardiac surgery information regarding the surgery was obtained to determine whether it was definitive treatment or not. Names of participants and their respective hospital registration numbers were listed in a checklist in order to avoid double recruitment.

The Patients' types of CHD were classified into three groups according to cardiac defect severity and hemodynamics. These groups were;

1. group 1-left to right shunt (VSD,ASD,AV canal defect, PDA, Mitral prolapse),
2. group 2- obstructive abnormalities (Aortic stenosis and PS)
3. group 3 – complex congenital heart (TOF,TGA, single ventricle, truncus arteriosus etc).

Grading of HRQOL

In its development of the tool, the accepted cut off scores meaning poor quality of life by using pedQL generic instrument were 69.7 for child self report and 65.4 for parental proxy report in comparison with general population.[47] There is no published data that grade the quality of life into severe, moderate and mild. In most of studies where the cardiac module tool was used, the disease was categorized into mild, moderate and severe based on lesion severity and not quality of life scores.

Data management and analysis

Data management

Data were entered in Epi Info for storage and transferred to SPSS for analysis. Data cleaning was done weekly to minimize errors such as missing data.

Data analysis

Data analysis was carried out using SPSS version 13. The descriptions of socio-demographic characteristics were expressed in frequencies. The HRQOL domains were expressed in means and standard deviations and compared to groups of CHD classified according to hemodynamic differences. The subscale scores for the patients and the parents were compared across the groups in the study using univariate statistical methods. The student t test was applied for two groups and one way ANOVA was used for more than two groups. Multivariate analysis was used to detect the association. A p value of <0.05 was taken as a statistically significant.

Ethical issues and research clearance Ethical clearance

Ethical clearance to conduct the study was obtained from Muhimbili University and Allied Sciences Ethical Review Board. Permission to conduct the study was obtained from MNH administration.

Ethical consideration

Consent and assent for children to participate in the study were obtained from the eligible study candidates and parents. The parents /caretakers of the participants were told about the purpose of the study and requested to participate in the study. It was made clear that to participate in the study is voluntary and that those who will not respond or reject to participate in the study no outward consequences would happen upon their decisions. They were also assured of confidentiality and anonymity in that the questionnaire would not have any participant's name, only numbers were used. They were informed of the benefits and risks of the study and that potential harmful risk would be minimized.

Study limitations

The tool (pedsQL^{TM} 3.0 cardiac module) used in this study has not been applied in countries with limited resource settings such as sub-Saharan Africa, except the generic one which is not disease specific was used in South Africa. The tool has been designed and 26 normalized among Western populations.

The study being a descriptive cross sectional study cannot explain a causal relationship hence a better study design would have been prospective cohort study. This was not possible due to time and financial constrains.

CHAPTER THREE

RESULTS

Participant's socio demographic characteristics

Basic characteristics of children

A total of 107 children aged 2-18 years with CHD and their parents/guardians were recruited. The mean age in years and standard deviation (SD) was 6 ± 3. The majority of patients (73%) were below 8 years of age and females made up 53.3%.VSD was found to be the most commonest CHD (35%), followed by TOF (19.6%), PDA (14%) and complex CHD(14%) (Table1). Amongst the study population 9 (8%) had co morbid conditions including HIV, Down syndrome, epilepsy and speech problems.

Table 1(a) Basic characteristics of children

Age and sex of children	**Sex**		**Total**	**Percent**
Age	**Male**	**Female**		
2-7	36 (72%)	42 (73.7%)	78	72.9
8-18	14 (28%)	15 (26.3%)	29	27.1
Total	**50 (46.7%)**	**57(53.3%)**	107	100
Types of CHD	**CARDIAC SURGERY**	**NO CARDIAC SURGERY**		
VSD	16	21	37	34.6
TOF	15	6	21	19.6
PDA	7	8	15	14.0
Complex CHD	9	6	15	14.0
PS	5	2	7	6.5
AV canal defect	2	2	4	3.7
ASD	1	3	4	3.7
Mitralvalve prolapse	2	1	3	2.8
AVS	0	1	1	0.9
Total	**57(53.3%)**	**50(46.7%)**	**107**	**100**

N=107, 57

Complex included

2 patients -complex CHD in their diagnosis

2 patients -Tricuspid atresia + TGA and VSD

2 patients - TGA + Truncusarteriosus

1 patient - TOF + tricuspid atresia

1 patient - VSD + ASD, PDA, hypoplastic LV and double outlet RV

2 patients -dextrocardia

2 patients –truncus arteriosus + VSD

1 patient - total anomalies and

2 patients -single ventricle

Socio demographic characteristics of parents/guardian

Majority (74.8%) of the parents/guardian had primary education. About30% of the parents/guardians had no permanent jobs (Table 2).

Table 1(b)Socio demographic characteristics of parents/guardian

	Number	**Percent**
Education		
Primary	80	74.8
Secondary education	17	15.9
College/university	3	2.8
No formal education	7	6.5
Total	107	100
Occupation		
Formal employment	13	12.1
Petty trade	30	28.0
Peasant	25	23.4
Laborer	32	29.9
Business	7	6.5
Total	100	100

N=107,

Participants socio demographic characteristics compared with mean scores pedsQL cardiac module

Mean scores pedsQL cardiac module by sex of children

The total mean scores are below the cutoff point both in the parent and child reports. It is also low both in the male and female category. But females scored higher in the e treatment anxiety domain (mean and sd) 97± 11 than in male children (mean and sd) 90 ±11 in the parent/guardian respondents. This difference was statistically significant with a p value of 0.036. The difference in the other domains was not statistically significant (Table 2).

Table2 Mean scores pedsQL cardiac module by sex

	Male			**Female**			**95% CI**		**p value**
Domain	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**Lower**	**Upper**	
Child report									
Symptoms	14	67	(15)	15	68	(15)	-13.35636	9.75078	0.751
Treatment II	8	98	(4)	6	88	(20)	-6.06901	25.65234	0.203
*Physic appear	14	88	(17)	15	89	(18)	-14.62076	12.31838	0.862
*Treatmt anxiety	13	100	-	15	94	(16)	-3.36986	15.31386	0.200
Cognitive	14	82	(19)	15	79	(19)	-11.54653	17.25700	0.687
Communication	14	95	(11)	15	96	(15)	-10.46607	9.54892	0.926
Total scores	14	70	(9)	15	66	(11)	-3.62580	11.0980	0.307
Parent report									
Symptoms	50	67	(15)	57	68	(18)	-8.29298	4.66036	0.579
Treatment II	31	97	(7)	21	94	(16)	-3.40344	10.14445	0.322
*Physic appear	36	92	(14)	43	90	(15)	-5.121222	8.03517	0.660
*Treatmt anxiety	50	90	(24)	57	97	(11)	-14.61880	-0.51102	0.036
Cognitive	14	80	(22)	15	77	(27)	-15.56251	22.04613	0.726
Communication	14	97	(11)	15	92	(20)	-7.89716	17.33113	0.450
Total scores	50	55	(19)	57	52	(15)	-3.34520	9.809330	0.332

N=107, student t test, p <0.05

*Physic appearmeans physical appearance

*Treatmt anxiety means treatment anxiety

n-number of participants

Mean scores pedsQL cardiac modules by ages; 2-4 and 5-7 years

Lower age or higher age was not found to be the determinant of good or poor HRQL. The overall scores were below the cut off scores of 69.7 both in toddler and young children. Mean scores of physical functioning, and nonphysical functioning (treatment, physical appearance, treatment anxiety, cognitive and communication problems) were not found to be different between ages 2-4 years and 5-7 years. All the p values ofHRQL against age 2-4 and 5-7 years were not statistically different p value > 0.05 (Table 3a).

Table 3a Mean scores pedsQLcardiac modules by ages;2-4 and 5-7 years

	2-4 years			**5-7 years**			**P**
Domain	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	
Parent report							
*Symptoms	43	67	(16)	35	66	(18)	0.183
Treatment II	26	97	(13)	15	98	(9)	0.432
Physicappear	28	91	(17)	23	91	(14)	0.904
*Tretmntanxiet	43	92	(21)	35	92	(22)	0.406
Total scores	43	46	(9)	35	43	(6)	0.995

N=107,student t test

*Tretmntanxiet means treatment anxiety

*Symptoms means Heart problems/physical functioning

Mean scores pedsQL cardiac module by ages 8-12 and 13-18 years

Total mean scores in the children reports are below the cut off scores of 69.7 while in the parent reports are above that of cut off scores. Children of 8-12 years were found to have lower heart symptoms scores of (mean and sd) 64 ± 14 than those

of 13-18 years with scores of (mean and sd) 76 ± 13 in the child respondents. This was statistically significant with a p value of 0.044.

Parent respondents also reported lower heart symptoms scores (mean and sd) 67 ± 17 for children of 8-12 years compared to 13-18 years of age (mean and sd) 80 ± 10). The difference was not statistically significant a p value of 0.183. The other (non physical) domains showed no difference (Table 3b).

Table 3b Mean scores pedsQL cardiac module by age

	8-12 years			**13-18 years**			**P**
Domain	**n**	**Mean scores**	**(Sd)**	**n**	**Mean scores**	**(sd)**	
Child report							
Symptoms	21	64	(14)	8	76	(13)	0.044
Treatment II	11	92	(15)	3	100	-	0.415
*Physical appear	21	86	(19)	8	94	(11)	0.327
*Treatmt anxiety	21	96	(14)	7	100	-	0.432
Cognitive	21	81	(19)	8	79	(19)	0.756
Communication	21	96	(13)	8	93	(13)	0.644
Total scores	21	67	(10)	8	69	(8)	0.258
Parent report							
Symptoms	21	67	(17)	8	80	(10)	0.183
Treatment II	9	90	(13)	2	100	-	0.432
*Physical appear	20	89	(15)	8	94	(10)	0.904
*Treatmt anxiety	21	98	(9)	8	100	-	0.406
Cognitive	21	76	(27)	8	86	(15)	0.286
Communication	21	95	(17)	8	95	(15)	0.968
Total scores	21	76	(12)	8	80	(5)	1.00

N=107

n-number of participants

Mean score pedsQL cardiac module by occupation of the parents

The overall means scores (total scores) were below the cutoff point of 69.7 in all levels of occupation in the parent's respondents while in the child report were above the cut off scores.

In comparing the mean scores of physical functioning, treatment II, physical appearance, treatment anxiety, cognitive and communication problems to the occupational status of the parent no statistical significant difference was found. The HRQL mean scores were not associated to occupation of the parent (petty trader, peasants, employed individual and businessmen) (Table 4).

Table 4 Mean score pedsQL cardiac module by occupation of the householders

	Formal employment			Petty traders			Peasants			Laborer			Businessmen			p
Domain	**n**	**Mean scores**	**(Sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	
Child report																
Symptoms	5	70	(17)	6	61	(12)	9	73	(14)	6	64	(15)	3	64	(21)	0.58
Treatment II	2	98	(4)	1	100	-	5	96	(9)	4	88	(25)	2	95	(7)	0.90
*Physical appear	5	88	(22)	6	89	(20)	9	80	(19)	6	94	(9)	3	100	-	0.42
*Treatmt anxiety	5	93	(15)	6	91	(23)	9	100	-	6	100	-	2	100	-	0.56
Cognitive	5	74	(23)	6	75	(22)	9	87	(18)	6	80	(15)	3	82	(20)	0.71
Communication	5	97	(8)	6	87	(23)	9	95	(12)	6	100	-	3	100	-	0.51
Total scores	5	71	(17)	6	70	(13)	9	82	(8)	6	83	(6)	3	85	(7)	0.21
Parent report																
Symptoms	13	68	(15)	30	68	(19)	25	65	(18)	32	67	(15)	7	76	(12)	0.60
Treatment II	7	92	(14)	17	96	(16)	12	96	(10)	13	98	(7)	3	97	(6)	0.91
*Physical appear	10	91	(16)	23	94	(12)	18	86	(16)	22	90	(16)	6	94	(14)	0.49
*Treatmt anxiety	13	96	(12)	30	98	(9)	25	87	(28)	32	92	(20)	7	100	-	0.22
Cognitive	5	83	(27)	6	67	(26)	9	88	(18)	6	74	(21)	3	75	(43)	0.60
Communication	5	89	(26)	6	90	(24)	9	95	(13)	6	100	-	3	100	-	0.74
Total score	13	58	(19)	30	54	(14)	25	54	(21)	31	49	(14)	7	62	(19)	0.26

N=107, one way ANOVA test,

sd standard deviation ,

- all the responses were exactly the same,thus no sd ????

*Physical appear means physical appearance

*Treatment anxiety means treatment anxiety

n-number of participants

Mean score pedsQL cardiac module by level of Education of the parents

Total scores were below the cutoff point in all levels of education in the parent report while not in the child report (cut off scores for poor HRQL of 69.7).

The educational status of the parent was not found to be a determinant of HRQL of the children. The mean scores of physical functioning, treatment II, perceived physical appearance, treatment anxiety, cognitive and communication problems in parents who were primary school leavers were not statistically different from those

of no formal education or higher education (secondary and college education). All the p values were above 0.05 (table 5).

Table 5 Mean score pedsQL cardiac module by level of Education of the parent/guardian

	Primary			**secondary**			**College/university**			**No formal educ**			**p**
Domain	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	
Child report													
Symptoms	21	66	(14)	3	73	(24)	2	67	(1)	3	75	(19)	0.726
Treatment II	10	92	(16)	2	96	(4)	NA	NA	NA	2	100	-	0.735
*Physical appear	21	90	(16)	3	96	(7)	2	75	(35)	3	79	(19)	0.438
*Treatmt anxiety	20	97	(13)	3	100	-	2	83	(24)	3	100	-	0.428
Cognitive	21	81	(19)	3	80	(30)	2	66	(1)	3	88	(11)	0.663
Communication	21	94	(10)	3	94	(10)	2	100	-	3	100	-	0.865
Total scores	21	79	(10)	3	85	(18)	2	65	(9)	3	85	(11)	0.214
Parent report													
Symptoms	80	68	(17)	17	64	(16)	3	76	(2)	7	73	(14)	0.525
Treatment II	39	98	(7)	9	94	(12)	NA	NA	NA	4	83	(33)	0.056
*Physical appear	59	91	(14)	12	90	(19)	3	94	(10)	5	91	(14)	0.964
*Treatmt anxiety	80	92	(21)	17	99	(3)	3	86	(24)	7	100	-	0.329
Cognitive	21	76	(25)	3	92	(14)	2	70	(43)	3	87	(13)	0.670
Communication	21	95	(15)	3	100	-	2	71	(29)	3	100	-	0.201
Total scores	80	53	(16)	17	52	(17)	3	58	(18)	7	61	(20)	0.624

N=107, ANOVA test, NA not applicable

*Physical appear means physical appearance

*Treatmt anxiety means treatment anxiety

n-number of participants

Mean score of pedsQL cardiac module and children with cardiac surgery and those without

The total scores in the parent's respondents were below the cut off point of 69.7 both in the cardiac surgery and non cardiac surgery individuals.

In the parent responses, higher mean scores were observed in the heart problems symptoms (mean and sd) 71 ± 15 in children who had cardiac surgery than those who had not had cardiac surgery (mean and sd) 64 ± 18, with a p value 0.03.

The perceived physical appearance mean scores for those children with cardiac surgery (mean and sd) 89 ± 16 were lower compared to those who had not had cardiac surgery (mean and sd) 99 ± 3, with a p value of 0.001.

According to the children responses, the heart symptoms mean scores were lower in patients who had not yet had cardiac surgery compared to those who had with mean scores of 62 ± 17 and 71 ± 13 respectively. The difference was not statistically significant.

Physical appearance was significantly low in patients who had cardiac surgery compared to those without and their mean scores were 82 ± 20 versus 98 ± 4respectively with a p value of 0.011.

Other parameters were not statistically significant either in the parents'/guardians' and children' reports (Table 6).

Table 6 Mean score of pedsQL cardiac module by cardiac surgery

	Cardiac surgery			No cardiac surgery			95% CI		p
Domain	**n**	**Mean scores**	**(Sd)**	**n**	**Meanscores**	**(Sd)**	**Lower**	**Upper**	
Child report									
Symptoms	18	71	(13)	11	62	(17)	-3.129	19.787	0.147
Treatment II	4	95	(10)	10	94	(16)	17.114	20.114	0.864
Physical appearance	18	82	(20)	11	98	(4)	-28.639	-4.068	0.011
Treatment anxiety	18	95	(15)	11	100	-	-14.815	4.861	0.308
Cognitive problems	18	76	(20)	11	88	(14)	-26.368	1.753	0.084
communication	18	92	(16)	11	100	-	-17.401	2.338	0.129
Total scores	18	72	(19)	11	90	(6)	-22.513	-9.729	0.216
Parent report									
Symptoms	57	71	(15)	50	64	(18)	0.591	13.243	0.03
Treatment II	5	100	-	47	96	(12)	-6.956	15.681	0.443
Physical appearance	57	89	(16)	22	99	(3)	-18.214	-4.518	0.001
Treatment anxiety	57	94	(17)	50	93	(21)	-6.4425	7.962	0.835
Cognitive problems	18	74	(24)	11	85	(24)	-29.780	8.083	0.250
communication	18	91	(20)	11	100	-	-21.400	3.941	0.169
Total scores	57	52	(15)	50	55	(19)	-9.539	3.626	0.018

n=107, In subgroup analysis symptoms were shown to have improved after cardiac surgery but cognitive problems were shown to affect more those who had cardiac surgery. This was seen even in subgroup analysis of TOF, VSD and complex cardiac lesions

Mean score pedsQL cardiac module by cardiac disease severity

The types of CHD were classified into three groups according to disease severity (Table 7). Physical functioning and cognitive problem mean scores were found to be lower in children with severe diseases (groups 2&3) than those with less severe disease (group 1).

Overall total scores were below the cut off point of 69.7 in the child report group 2 CHD lesions while in the parent respondents total scores were below the cut off points in all CHD lesion subgroups

In children's responses the physical functioning mean scores were lower in severe disease but the difference was not significant. The mean scores were 65±12, 60 ± 5 and 69 ± 17 in groups 3,2 and 1 respectively with a p value of 0.622.

With regard to cognitive development, children with severe cardiac disease were found to have significantly lower mean scores 75 ± 19 in group 3 and 46 ± 4 in group 2 than the less severe cardiac disease group 1(mean score and sd) 86 ± 2 with a p value of 0.008. The statistical significant difference was between group 2 and group 1 by Bonferroni alpha Post hoc test with adjusted p value of < 0.0167.

From the parent's responses, children with a less severe disease had good mean scores in the physical function compared to severe disease. The mean scores were 72 ± 14, 67 ± 17 and 59 ± 18 in group 1, 2 and 3 respectively with a p value 0.000. Group comparison found a significant difference between group 1 and 3 by Bonferroni alpha post hoc test with adjusted p value < 0.0167.

The cognitive and other domains did not show any statistical significant differences across disease severity.

Table 7Mean score pedsQL cardiac module by cardiac disease severity

	Group 1			Group 2			Group 3			P
Domain	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	**n**	**Mean scores**	**(sd)**	
Child report										
Symptoms	19	69	(17)	2	60	(5)	8	65	(12)	0.622
Treatment II	9	93	(16)	NA	NA	NA	5	96	(8)	0.695
*Physical appear	19	89	(16)	2	100	-	8	83	(21)	0.416
*Treatmt anxiety	19	97	(13)	2	100	-	8	96	(12)	0.911
Cognitive	19	86	(15)	2	46	(4)	8	75	(19)	0.008
Communication	19	96	(13)	2	91	(12)	8	93	(13)	0.796
Total scores	19	80	(11)	2	66	(2)	8	78	(11)	0.265
Parent report										
Symptoms	64	72	(14)	7	67	(17)	36	59	(18)	0.000
Treatment II	35	95	(14)	1	100	-	16	99	(4)	0.467
*Physical appear	43	94	(11)	7	88	(20)	29	87	(16)	0.075
*Treatmt anxiety	64	96	(15)	7	93	(19)	36	89	(24)	0.199
Cognitive	19	84	(23)	2	56	(25)	8	71	(24)	0.202
Communication	19	97	(13)	2	100	-	8	88	(23)	0.376
Total scores	64	56	(17)	7	51	(17)	36	50	(16)	0.161

N=107, ANOVA, P value <0.0167,Bonferroni alpha Post hoc test significant between group 1 and group 3 in the parent symptom report and between group 1 and group 2 in child cognitive problem report,

GROUP 1 left to right shunt,

GROUP 2 obstructive cardiac defect,

GROUP 3 complex CHD including TOF, TGA and TA,

*Physical appear means physical appearance,

*Treatmt anxiety means treatment anxiety.

Multivariate analysis of the predictors of HRQL

Cardiac surgery and types of congenital heart diseases are negative predictors of physical functioning in the multivariate analysis, beta coefficients of -0.29 for cardiac surgery and -0.42 types of CHD (Table 8). While cardiac surgery was found to be a positive predictor of perceived physical appearance beta coefficient of 0.32; type of congenital heart diseases was a negative predictor of that parameter, beta coefficient of -0.19. In this analysis their statistical difference value was highly significant showing, physical functioning with p values of 0.000 for cardiac and types of CHD and perceived physical appearance p value of 0.006 for cardiac surgery

Table 8 Multivariate analysis of the predictors of HRQL

	Physical functioning			**Physical appearance**			**Treatment anxiety**			**Cognitive**		
Predictors	**ß**	**t**	**P**	**ß**	**t**	**P**	**ß**	**t**	**p**	**ß**	**t**	**p**
Sex	-0.02	-0.25	0.79	0.03	0.27	0.78	0.19	1.97	0.05	0.23	1.08	0.29
Age	0.03	0.39	0.69	-0.00	-0.07	0.93	0.12	1.23	0.22	0.23	1.08	0.29
Education	0.04	0.44	0.65	-0.02	-0.24	0.80	0.06	0.62	0.53	0.03	0.15	0.87
Occupation	0.02	0.21	0.82	0.01	0.06	0.94	-0.06	-0.62	0.53	-0.12	-0.57	0.57
*G of CHD	-0.42	-4.69	0.00	-0.19	-1.74	0.08	-0.15	-1.57	0.11	-0.22	-1.02	0.31
*C.surgery	-0.29	-3.11	0.00	0.32	2.81	0.00	0.00	0.00	0.96	0.27	1.22	0.23

ß beta coefficient,

*G of CHDmeansgroup of CHD

*C.surgery means cardiac surgery

CHAPTER FOUR

DISCUSSION

Participant's socio-demographic characteristics and HRQL

Children with CHD in this study were found to have poor HRQL as the overall scores (total scores) were below the cut off scores of 69.7. [47] This was more demonstrated in the parent's responses in all of the risk factors assessed. In the children report few factors had lower total mean scores than the cutoff point. This variation between parent's report and children report might be explained by the bigger sample size in the parent's responses than in the children and also the other reason might be that the parent's responses involved all the ages in this study where some small children might have not yet had cardiac surgical interventions which improves some of the parameters as has been observed in this study.

According to male gender, children were found to have a significantly higher risk of developing treatment anxiety symptoms than female children as reported in the parent respondents. There was no difference in the children report for male and female anxiety features possibly children fear to express their problems.

This study is contrary to other previous studies. The study by Uzark et al in USA, there was no gender difference on HRQL using the same tool and age category as used in this study. [17] The difference in finding between this study and their study might be due to cultural and social issues. Uzark's study was done in USA where culturally and economically is different from the study participants of this study. Children of the Western countries are free to their parents. They are more exposed and acquire independency from children care centers earlier than African children. The exposure makes them more competent and interactive. Also advanced technologies in the west countries facilitate their learning and hence adapting very fast than African children. There is also no gender bias in western countries in terms of raising the male and female children. Therefore all of these factors might be contributing to the difference observed in these two studies.

Also Mostafa et al found female children being at risk of developing anxiety features than male. [13]Although Mostafa found female children experiencing anxiety features compared to our findings which were more in male, they used a different assessment tool SF 36 while this study used the PedsQL cardiac module tool. Probably the questions towards eliciting the anxiety problems were different to this study. But anxiety features are expected to be more in female than in male children because male children are more courageous than female. Therefore the finding in this study on anxiety problems more on male must be explained by other reasons other than gender only. These include age and CHD lesion group matching which were not considered in this study.

With regard to age, it was found that children 8-12 years old had significantly poor physical function than those of older ages 13-18 years.

In the study done by Mostafa et al exploring the relationship how age affect HRQL. They found mean score for QOL including physical functioning being the least for the lowest age category, it increased to the highest in the primary school age category, and then during the adolescence(> 12 years) it worsens again.[13] In their study they assessed Qol of parents of children with cardiac disease compared those parents of children with minor illness. They used SF-36 tool to measure different parameters of QoL different from that one used in this study. The differences in findings may be brought due to these two differences of tool and study subject where in their study included children with acquired heart diseases.

In the study by Uzark et al in USA no difference was noted between age and HRQL.[17]Using the same tool and age category as used in this study no difference was observed on physical functioning domain in their study. So difference that is seen in this study especially at ages 8-12 against 13-18 on physical functioning domain is likely to be contributed by other factors such as difference in sample size of these two groups.

According to parents' socio demographic features there were significant difference found in relation with HRQL. This study assessed parent's socio demographic

characteristics on occupation and education. On occupation, majority of parents were in low class (peasants, petty traders, unemployed and those who said are businessman). Concerning level of education of parents, the majority were primary school leavers. Higher socio economic status and higher education level of the parent are implied good quality of life for the child.[13] Families with good socio economic status and well educated provide good support to their sick children. When the child is well supported socially and psychological she/he develops well and hence a good quality of life.

In this study we found no difference in HRQL for children whose parent's occupation and level of education were better compared to those of low socio demographic characteristics (occupation and education). The reason for this could be that the assessment was just on their occupation and not their financial status. Occupation status does not always reflect economic status. Financial status may have yielded better information. The majority of participants were primary school leavers. Therefore it is difficult to draw any conclusion basing on this observation.

The findings were not consistent with other previous studies. Some studies reported socio demographic characteristics of parents (occupation and economic status) being associated with poor HRQL while others did not. Mostafa et al study in Egypt demonstrated poor HRQL in parents of children with heart diseases when their parents had low socio economic status compared to those who had good socio economic status, but education status was not associated.[13] While the study by Uzark in USA found that family socio economic status has no impact on HRQL.[17]

Health related quality of life and cardiac operation

In this study physical functioning domain was found better in children who had cardiac surgery compared to those who have not undergone cardiac surgery in the parent's respondents and not in the children report. The possible reason for

statistical significant being noted only in the parent report is due to few subjects in the children report. However Successful cardiac surgery normally improves the condition of the patients in terms of physical functioning. The symptoms which the patients had been experiencing some of them disappear after successful cardiac surgery done.

These findings are in keeping with other studies which found patients with CHD after cardiac surgery show improvement in HRQL especially physical functioning. A study by Wray et al demonstrated a significant improvement in self-perception following surgery for CHD.[46]In this study, children with CHD before cardiac surgery considered themselves as weaker, more frightened and more ill than the healthy children.

Another study which examined the association between early outcomes, health related quality of life and survival following open-heart surgery found that one year post surgery all parameters of HRQL were improved except for social and mental health status which improved later.[32] Also the Swiss study found normalization of parent's perception on HRQL after six months post surgery. In their study they looked at predictors of parental quality of life after the child open heart surgery and found low parent's mental HRQL in the immediate period and this continued improving with time.[31]

Cognitive development was found to be poor in children who had cardiac surgery compared to those without cardiac surgery but not statistically significant. Although majority of children with CHD lesions need cardiac surgery only those with severe CHD lesion get early surgical intervention in our country due to limited resources. It is likely that children with severe CHD lesion have neurodevelopment impairment due to cerebral hypoxia and other associated complications such as Heart failure, infections and frequent separations due to admissions. Prolonged surgery is also associated with poor neurodevelopment in children as it causes hypoxia due cardiopulmonary bypass. On the other hand frequent separation from interaction with peers causes delay in acquiring

neurodevelopment skills. So children who had cardiac surgery are likely to have good quality of life on neurodevelopment compared to the one who have not yet cardiac surgery. But in this study child with cardiac surgery were the one who were found to have poor cognitive development compared to those without cardiac surgery meaning that disease severity and cardiac surgery might be the contributing factors for poor cognitive development. Therefore the low cognitive development observed in this study might be multifactorial causes either being caused by the complications before cardiac surgery due to the severity of CHD lesion or the cardiac surgery itself.

This finding are consistent with other studies that cardiac surgery is associated with poor neurodevelopment [17, 18, 19, 20] because of its complication, however multiple causes play part.

There was also lower mean scores perceived physical appearance problems in children who had cardiac surgery than those who had no cardiac surgery for both parent and child respondents. Children before cardiac surgery appeared normal in their physical appearance. Cardiac surgery scar disfigures their body images. However some individuals might have disfiguring due to enlarged heart possibly is not much of their concern. Due to lack of knowledge about the scar after cardiac surgery patient's perceive as an abnormal thing and should not be there. This is perceived by parents and children as abnormal and they don't like to appear. No literature found compared children with cardiac surgery and those who had not yet had cardiac surgery against perceived physical appearance. But in studies which looked quality of life to children with CHD after surgery physical appearance was found to affect children quality of life.[17,18]

Mean scores pedsQL cardiac module and CHD disease severity

This study showed significant difference in HRQL for the physical functioning domains in children with CHD lesion (group 1,2& 3).Multiple comparison tests revealed significant difference between group 1 and 3.

This is in consistency with the study done in Poland on quality of life in children with mitral valve prolapsed which found physical functioning as the only domain being statistically poor while other domains which were assessed being less significant. This means that more impairment will be in severe diseases like in our study .

The Uzark et al study in USA which used the same method as in our study found both physical and nonphysical functioning domains being poorer in children with severe cardiac diseases than to those with less severe cardiac diseases in the child report. In the parent report significant difference was found only on physical functioning domains while nonphysical domains were less statistically significant.[17] They compared normal population and children with cardiac disease by using pedsQL cardiac module and pedsQL generic instruments. Disease severity was classified into four groups according to American Heart Association classification of cardiac disease severity.

The same findings were reported in the study of Bosnia and Herzegovina where severe cardiac diseases affect severely the HRQL than less severe diseases. The difference in this study is that all the HRQL domains including physical functioning were reduced significantly across disease severity [18].

Also the Egyptian study using SF-36 found that cardiac disease type affected differently the QoL. The domains which were assessed included; physical functioning, general health, role limitation due to physical health problems, vitality, role limitation due to emotional problems and social functioning. [13]These findings show variation between one study and another because of variation in methods of study (instrument, sample size and sampling techniques) and socio demographic characteristics differences (culture and norms).

Whereas children with severe disease (group 2&3) were having low mean scores HRQL on their physical functioning than mild disease (group 1) as were reported by parents. Only the significant differences were between group 1 and 3 not 1 and

2. The reason for group 1 and 2 not being statistically significant is because few subjects were studied in group 2 therefore inadequate to detect the difference. Also the significant difference was found in the parent respondents and not in the child respondents are due to; perceptions of parents on consequences associated with physical exertions. They think that physical exertion such as play or run worsen the child's condition. They tend to give protection by restricting to do physical works. This is why parent's respondents on physical functioning problems were demonstrated more in children with severe disease. Parents of children with mild cardiac defect have less perception on effects of physical functioning. Children with mild condition perform normally the daily work and without experiencing symptoms of physical exertion. The child's presentation gives assurance to the parents that physical exercise has no harm to the child. Children with less severe cardiac defect have less heart symptoms. Less severe CHD some of them are asymptomatic such as ASD, others present with symptoms later in childhood or adulthood. It becomes obvious from 2-4 years and mainly is due to heart failure. In this line more physical functioning reports were observed in parents of children with severe CHD than those parents of children with less severe diseases. Also no difference was observed in the children report may be due to children themselves undermining their problems. They might be doing so because they do not want to be called lazy.

The physical functioning problems in severe CHD such complex CHD is due to hypoxia, hypoxic spells and metabolic acidosis. The hypoxic spells sometimes cause fainting or loss of consciousness of the child. For those who had cardiac surgery some had palliative surgery because of the complexity of the defect. So may still experience symptoms of either, arrhythmias, heart failure or hypoxia depending on the hemodynamic abnormality they had.

Concerning cognitive development it was found that children with CHD lesion (1, 2,3) have poor cognitive development according to their disease severity. Statistically significant difference was noted in the children respondent whereas children CHD lesion severity increases, cognitive development become more impaired. After Bonferroni adjustment analysis a significant difference was found between group 1 and 2. The difference between group 1 and 2 possibly all of subjects in group 2 had poor cognitive development and because were few that is why huge difference is noted in the mean scores. This means that children with severe diseases had suboptimal neuro-development compared to those with less severe disease. The causes of this difference can be explained by the chronic hypoxia experienced during infancy period in children with severe cyanotic CHD and also cardiac operation complications which might be severe in those children who had severe CHD.[22] Children with severe diseases such as complex CHD, TOF and other severe CHD, cyanosis plus metabolic acidosis are noxious to a developing brain. The long term consequence of hypoxia is neurodevelopment deficit. Cardiac surgery has also been found to affect the brain. The determinant factor of brain injury in cardiac surgery is the duration of surgery. Children who had chronic hypoxic and long duration of cardiac surgery, majority develop cognitive disorders. Children with complex CHD could have experienced chronic hypoxia as well as complicated cardiac surgery. The end result of this is cognitive development impairment.

The mean scores cognitive problems by disease severity was not statistically significant in the parent respondents but were decreasing as the disease severity increases. The explanation for this may be parent's ignorance on assessing school performance of children or under estimation of the parents on child's cognitive development might be the explanation for that finding.

Our findings are inconsistent with other studies which found children with CHD had poor HRQL in many domains. The Uzark et al study, found both physical and nonphysical functioning domains being poorer in children with severe cardiac

diseases than to those with less severe cardiac diseases in the child report. In the parent report significant difference was found only on physical functioning domains while nonphysical domains were less statistically significant.[17] The same findings were reported in the study of Bosnia and Herzegovina where severe cardiac diseases affect severely the HRQL than less severe diseases. All the pedsQL cardiac module domains were reduced significantly in children with severe diseases [18]. In a systematic review also some studies reported lower QoL in children with complex CHD.[12] Also the Egypt study using SF-36 found that cardiac disease severity is associated with poor QoL compared to minor illness.[13] The low cognition in severe CHD we found in this study is similar as that in the Meta-Analysis where children with severe CHD exhibited lower cognitive functioning than those with mild disease [16]. The findings reported here are similar to our study because most of them examined children with CHD who had undergone cardiac surgery.

Multivariate analysis in determining the predictors of HRQL

In this study, cardiac operation and severity of CHD were found to be negative predictors of HRQL in the physical functioning domains. This means that as the disease severity increases the physical functioning lowers and that children who have not yet have cardiac surgery their physical function was lower than those with cardiac surgery. These observations were highly statistically significant showing that there is a linear relationship between these predictors and the outcome but this does not mean that they are the causes.

Therefore the study has shown that cardiac surgery and CHD lesion severity are the predictors of quality of life. However not all the pedsQL cardiac module domains were impaired. It is the physical functioning and cognitive development domains were found to be affected by severity of CHD lesion. For children who had cardiac surgery physical functioning was found to be better compared to those not operated. But poor cognitive development was observed in patient who had cardiac surgery meaning that, cardiac surgery which is an intensive procedure

might be associated with negative impact on child cognitive development as an additional effect to the already poorly developed brain of children with severe CHD lesion. Therefore early preventive measures including early cardiac surgery and follow up of these children for their cognitive development and generally their condition in total is mandatory in order to improve their health and quality of life.

CHAPTER FIVE

CONCLUSION

From the findings of this study it may be concluded that the impact of CHD on the HRQL of the child is significant particularly in the domain of physical functioning and cognitive development. It can also be concluded that those who have surgery will have a better quality of life in some domains than those who do not. Severe CHD lesions are associated with poor HRQL especially physical functioning and cognitive development. Although cardiac surgery improves the physical functioning of children with CHD lesion it is associated with poor cognitive development. However this may not be the only cause of poor cognitive development in children with CHD lesion because those with severe CHD lesions are associated with frequent illness and hypoxia and hence affecting the child cognitive development. Therefore surgical interventions have shown to prevent some of the negative impacts on child's quality of life.

RECOMMENDATIONS

There are various risk factors which affect the HRQL of children with CHD. In this study it was found that different CHD lesion affects differently the quality of life and surgical intervention is also associated with good outcome in quality of life by relieving the symptoms and other domains of quality of life. However in terms of cognitive development and physical appearance perception might have an impact although this study is inadequate to make conclusion. According to universal and Tanzanian health policy states the right of health services to one in need. As cardiac surgery has been found to bring a good outcome in all previous studies except for neurodevelopment, it is therefore important to provide surgical intervention to children with CHD lesions especially those with cyanotic CHD

according guidelines and health policy of the country. The MOHSW should make this as a priority and the surgery possibly to be done early according especially those with cyanotic CHD in order to improve the quality of life. Lastly a prospective cohort study is needed to explore in details on these risk factors of HRQL especially before cardiac surgery and after surgery, with emphasis on cognitive development in order to establish a temporal relationship.

REFERENCES

1. Macran S, Birks Y, Lewin B. ConQoL Manual; disease specific measure of health-related quality for children withcongenitalheartconditions.Availableathtt://www.cardiacrehabilitation.org.uk/congoL/conQoL_manual.pdf (2011).
2. Moons P, Deyk KV, Budts W, Geest SD. Caliber of Quality of life Assessments in Congenital Heart Disease, A plea for more Conceptual and Methodological Rigor. Arch Pediatr Adolesc Med. 2004;158:1062-9.
3. Gill TM , Feinstein AR. A critical appraisal of the quality of quality-of-life measurements. JAMA. 1994;272:619-26.
4. World Health Organization (WHO). Constitution of the World Health Organisation-Basic Documents; Forty fifth edition. 2006. Available at htt://www.who.int/governance/eb/who_constitution.en.pdf (2011).
5. Culbert EI, Ashburn DA, CullenD G, Jay AJ, William AW, Blackstone EH et al. Qality of life of Children after Repair of Transposition of Great Arteries Circulation. 2003;108:857-62.
6. Berkes A, Pataki I, Kiss M, Kemeny C, Kardos L, Varni JW . Measuring Health related quality of life in Hungarian Children with Heart Disease:Psychometric properties of the Hungarian version of Paediatric Quality of life Inventory 4.0 Generic Core scales and the cardiac Module Health and Quality of Life Outcomes. 2010;8(14) :2-12.doi:10.1186/1477-7525-8-14
7. Dionysia N, Apostolos C. Factors affecting the Quality of life in Children with Congenital Heart Disease. Health Science Journal. 2010;4(2):94-100.
8. Allen SK, Gauvreau K, Bloom BT, Jenkins KJ. Evidence based referral results in significantly reduced mortality after congenital Heart Surgery. Paediatrics. 2003;112:24-8.
9. Beena J, Francis J. Emotional and Behavioral Problems in Children and Adolescents with Congenital Heart Disease. Journal of Indian Association for Child and Adolescent Mental Health.2005 [Special article].1(4):1-5.
10. Moons P, De Geest S, Budts W. Comprehensive care for adults with congenital heart disease. Eur J Cardiovasc Nurs. 2002;1:23-8.

11. Moons P, Deyk KV, De Geest S, Gewilig M,Budts W. Is the Disease severity of Congenital Heart Disease associated with the quality of life and perceived health of adult patients? Heart. 2005;91:1193-8.
12. Beatrice L, Helfricht S, Fischer JE, Urs Bauersfeld U,Markus AL. Psychological adjustment and Quality of life in Children and Adolescents following open-heart surgery for Congenital Heart Disease;A systematic Review. BMC Paediatrics. 2009;9(6):1471-2431.
13. Mostafa AS ,Zaher R, El-Dowaty AA, Dalia EM. Quality of life among Parents of Children with Heart disease. Health and Quality of Life Outcomes. 2008;6(91):1-7.
14. Spijkerboer AW , De Koning WB, Bogers AJ, Heling AW, Verhulst FC et al. Health-related Quality of life in Children and Adolescents after invansive treatment for Congenital Heart Disease. Quality of life Research 2006 ;15(4):663-73.
15. Wray J, Radley-Smith R. Cognitive and Behavioral Functioning of Children Listed for Heart and /or Lung Transplatation American Journal of Transplantation. 2010;10(11):2527-35.
16. Petra AK, Merel K, Mulder BJ. Psychological and Cognitive functioning in Children with Congenital Heart Disease;A Meta-Analysis. Journal of Paediatric Psychology. 2006:1-15.
17. Uzark K,Jones K, Slusher J, Limbers CA, Claudia TM, Patel MM et al. Quality of life in Children with heart disease as perceived by Children and Parents. Paediatrics. 2008 ;121(5):e 1060-7.
18. Tahirovic E, Begin H, Nurkie M, Tahirovic H, Varni JW. Does the severity of Congenital Heart defects affect disease-specific Health related Quality of life in Children in Bosnia and Herzegovina? Eur J Paediatr. 2010;169:349-53.
19. Menahem S, Poulakis Z, Prior M. Children subjected to cardiac surgery for congenital heart disease, part 1; Emotional and Psychological outcomes Interact Cardio Vasc Thorac Surgery. 2009 ;19(7):600-4.
20. Deliontо L, Marpelli D, Volpeand B. Measurements of Cognitive outcomes and Quality of life in Congenital Heart Disease. Heart. 2006;92:569-74.

21. Pidula MA, Ades AM. Neurodevelopmental implications of Congenital Heart Disease. NeoReviews. 2006;7:e363-e9.
22. Ballweg JA, Wernousky G, Gaynor JW. Neurodevelopmental outcomes following Congenital Heart surgery. Paediatr Cardiol. 2007;28(2):126-33.
23. Mahle WT. Neurologic and Cognitive outcomes in Children with Congenital Heart Disease. Current opinion in Paediatr. 2001;13:482-6.
24. Jimenez M, Espil G, Thambo JB, Choussat A. Outcomes of operated Fallot of Tetralogy. Arch Mal Coeur Vaiss. 2002 ;95(11):1112-8.
25. Available at htt/en.wikipedia.org/wiki/cardiascurgery. Accessed 2011
26. Ophe'lie L, Weissenfluh CV, Gahl B, Markus SB et al. Quality of life of grown-up congenital heart disease patients after congenital cardiac surgery. European Journal of Cardio-thoracic surgery. 2009;36:105-11.
27. Markus AL, Buechel EV, Latal B. A 6 month prospective study on predictors of parental Quality of life after Child Open Heart surgery. J Paediatr. 2011;158:37-43.
28. Menace CC, Plessi AJ, Wessel DL, Jonas RA, Newburger JW. Current incidence of acute neurologic complications after open-heart operations in Children. Ann Thorac Surg. 2002;73:1752-8.
29. Bellinger DC, Jonas RA, Rappaport LA, Wernovsky DW, Kuban KC, Barnes PD et al. Developmental and Neurologic Status of Children after Heart Surgery with Hypothermic Circulatory Arrest or Low-Flow Cardiopulmonary Bypass. N Engl J Med. 1995;332:549-55.
30. Karlsson AK, Lidell EJ . Depressed mood ovr time after open Heart surgery impacts patients well-being;a combined study. Eur J Cardiovasc Nurs. 2008 ;7(4):277-83.
31. Janiec I, Werner B, Sieminska J, Ravens-Sieberer U. Quality of life of Children with Mitral valve prolapse. Qual Life Res. 2011;20:537–541.
32. Martin LM, Halpin LS, Barnett SD, Speir AM, Hunt SL, Mukherjee NA. The association between early outcome, Health-related Qulaity of life and survival following elective open-heart surgery. J Cardiovasc Nurs. 2008 sep;23(5):432-42.

33. Limperopoulos C, Majnermer A, Shevell MI, Rosenblatt B, Rohlicek C, Tchervenkov C et al. Functional limitations in Young Children with Congenital Heart Defects After Cardiac surgery. paediatrics. 2001 dec;108(6):1325-31.

34. Fekkes M, Kamphuis RP, Ottenkamp J, Erik Verrips, Vogels T, Zwindermann KH et al. Health related Qulity of life in Young adults with minor Congenital Heart Disease. Psychology & Health. 2001;16(2):239-350.

35. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation. 1971;43:323-32.

36. Tyran MJ, Becker AE, Macartaney FJ, Jimenez MQ, Shinebourne EA, Anderson RH. Nomenclature and Classification of Congenital Heart Disease. Br Heart J. 1979 ;41(5):544-53.

37. Jacobs JP, Wernovsky G, Elliott MJ. Analysis of outcomes for congenital cardiac disease: can we do better? Cardiol Young. 2007;17(suppl 2):145-58.

38. Samanek M, Vorikova M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemia survival study. Pediatr Cardiol. 1999;20:411-17.

39. Grabitz RG, Joffers MR,Collins-Nakai RI . Congenital Heart Disease incidence in the first year of life. Am J Epidemilology. 1988;128:381-8.

40. Hoffmann JI,Kaplan S. The incidence of Congenital Heart Disease. Journal of American College of Cardiology Foundation. 2002;39:1890-900.

41. Fifth report on the provision of services for patients with Heart Disease 2002;88:iii1-iii59. Available at https://www.hst.org.za/uploads/files/chap 15_06.pdf

42. Padmini RA, D'souza NM. The incidence and pattern of Congenital Heart Disease in Tanzanian Children. East African Medical Journal 1976;53(36).

43. Kazimoto TK. Dissertation study on Cardiac disorders among Children aged 1month to 15 years at Muhimbili Medical Centre;Diagnosis, Management and course of the Disease. 1989.

44. Nyawawa ET, Ussiri E, Chillo P, Waane T, Lugazia E, Mpoki U et al. Cardiac Surgery; One year experience of Cardiac surgery at Muhimbili National Hospital, Dar es salaam, Tanzania. East and Central African Journal of Surgery. 2010;15(1):111-8.

45. Editorial and Comments CanadMAJ. 1959;81.

46. Wray J ,Sensky T. How does the intervention of cardiac surgery affect the self-perception of children with congenital heart disease? Child Care Health Dev. 1998;24(1):57-72.

47. Varni JW, Limbers C, Burwinkle TM. Literature review. Health -related Quality of Life measurement in pediatric oncology .Hearing the voice of the children. Journal of Pediatric psychology .2007 ;32(9):1152-1163

APPENDIX

Questionnaires

Socio-demographiccharacteristics

To be administered by an interviewer

Child sex Boy □ Girl □

Child age (years)…….

Child education yes attendinding school □ not attending school □

If yes, nursery□ ,primary which class □ ,secondary□university/college □

Sex of the parent/care taker

Male□ Female□

Relation to the child

Biological Parent□ Step Parent□ Grandparent□

Adoptive Parent□ Foster Parent□ Caretaker□ Other (specify)…….

Level of education of the father/mother or care taker

No formal education □ Standardseven □ Form four □ College/university

Employment status of parent/care taker

1. Formal employment □
2. Petty trader □
3. peasant □
4. Business □
5. Others mention……..

Marital status of the parent/care taker

Married □ Single □ Divorce □ Widowed □ Cohabiting □ Others……..

Family size

Three Five More than seven

Type of CHD

Information from patient file

Write the type of CHD …….

Has she/he has cardiac defect repaired?

Yes □ no □

Was the operation definitive □ palliative □

Consent form (English version)

Title: Health related quality of life in children with congenital heart diseases aged 2-18 years at Muhimbili national hospital in Dar es Salaam, Tanzania.
Id no……

Greetings! My name is Dr. Charles Kessy Shija, a resident in the department of Paediatric and Child Health. I would like to conduct a study mentioned above as necessary requirement for fulfillment of my postgraduate studies Purpose of the study

The aim of the study is to determine Health related quality of life in children with congenital heart diseases 2-18 years at Muhimbili National Hospital. Knowing the magnitude of the problem will help in improving management of these patients in broad spectrum as far as their health is concerned.

How to participate

Patients who meet the inclusion criteria will be recruited into the study. The parents and children above 5 years will be interviewed using standard questionnaire. While children below 5 years interview about their health will be obtained from their parents using questionnaire for respective age.

Risks

There is no any anticipated harm to the patient and parents in this study.

Benefits

Patients who will be found to have any problem concerning cardiac, psychological or any will be managed by the appropriate expert in that problem..

Confidentiality

Any patient's information will not be revealed to anybody except the attending doctors and parent/ patient if request.

Cost

No payment is requested from you as a fee to participate in the study.

Person to contact in questions or problems

The following you may contact incase there is a problem or you want to get clarifications.

Dr. Charles KessyShija (0713 207107) - Investigator

Prof KarimManji (0754 350630) -Professor, in Paediatrics and Child Health (MUHAS).

Dr Margaret Hogan (0784 339750)-Senior Lecturer and Psychologist (MUHAS)

I…… have read/been told of the contents of this form and understood its meaning. I agree to participate me and my child to participate in this study.

Signature…….. (Participant) Date……

Signature…….. (Researcher) Date……

Fomuyaridhaa (Swahili version)

Kichwa chahabari:Hali ya maisha ya kiafya ya watoto walio na tatizo la kuzaliwa la moyo wenye umri kati ya miaka 2-18 katika hospitali ya taifa Muhimbili ,Dar es salaam, Tanzania

Namba ya usajili…… …….

Salaaam! Mimi naitwa Dr Charles Kessy Shija, ni mwanafunzi wa udhamili Chuo Kikuu cha Sayansi za Afya.Nafanya utafiti wa hali ya maisha ya watoto walio na tatizo la kuzaliwa la moyo waliolazwa katika wodi ya watoto na wale wanaofanya kliniki ya magonjwa ya moyo katika hospitali ya Taifa ya Muhimbili.

Madhumuni ya utafiti

Kujua matatizo ya hali ya maisha ya kiafya katika watoto wenye tatizo la kuzaliwa la moyo, ninatarajia kusajili watoto 100 pamoja na wazazi wao watakaokuwa wamelazwa katika wodi ya watoto au wanahudhuria kliniki ya matatizo ya moyo wakiwa na tatizo la moyo la kuzaliwa.Utafiti huu utasaidia kugundua matatizo ya hali ya kiafya kwa watoto hawa na kuboresha zaidia fya zao.

Jinsi ya kushiriki

Kama utakubali kushiriki ,nitakuhoji maswali machache kuhusu ugonjwa wa watoto/mtoto wako,mwenye umri zaidi ya miaka2.Maswali yanayoulizwa yanayohusiana na tatizo la moyo yapo kwenye dodoso maalum.

Utunzaji wa siri

Taarifa za ugonjwa wa mtoto wako zitatunzwa kwa siri kwa kutumia isipokuwa kwa daktari anayemtibu mwanao pamoja na daktari mtafiti, wewe ukitaka kufahamu kuhusu matokeo ya mwanao utapatiwa.

Madhara /Athari

Hakuna athari au madhara yoyote yanayotegemewa kutokana na utafiti huu.

Uhuru wa kushiriki

Kushiriki kwenye utafiti huu ni hiari yako. Unaweza kujitoa wakatiwowote. Kama utachagua kutoshiriki, mtoto wako ataendelea kupata huduma hospitalini kama kawaida.

Faida ya utafiti

Ukishiriki kwenye utafiti huu,mtoto wako atachunguzwa kama ana tatizo la kiafya hasa hali ya maisha ya kiafya na atapatiwa matibabu stahili, na atapatiwa ushauri itakapohitajikakufanya hivyo.Kuna kamati ya kusimamia udhibiti waUtafiti huu.

Taarifa

Endapo unahitaji kupata maelezokuhusuhakizako au taarifa ,wasiliana na wafuatao.

Dr. Charles Kessy Shija (0713 207107) - Investigator

Prof KarimManji (0754 350630) -Professor , in Paediatrics and Child Health (MUHAS).

Dr Margaret Hogan (0784 339750)-Senior Lecturer and Psychologist (MUHAS)

Je unakubali kushiriki kwenye utafiti? (weka alama) vema ndiyo..... hapana.....

Mimi ....., nimeelezwa na nimesoma maelezo haya. Maswali yangu yamejibiwa.

Nimekubali mimi na mwanangu tushiriki kwenye utafiti huu

Sahihi ya mzazi/mlezi .....

Sahihi ya ndugu/shahidi .....

Sahihi ya Mtafiti ..... . Tarehe……