Study of Cardiac Diseases in the Pediatric Cardiology Clinic, Pediatric Hospital, Ain Shams University

Thesis

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List of Abbreviations

A NEUR .....A Anti neuronal antibodies

AF .....Anti failure measures

AS .....Aortic stenosis

ASD .....Atrial septal defect

ASOT .....Anti streptolysin o-titre

AVSD .....Atrioventricular septal defect

BAV .....Bicuspid aortic valve

CCVDS .....Congenital cardiovascular disease

CHD .....Congenital heart disease

CHF .....Congestive heart failure

CRP .....C-reactive protein

CXR .....Chest x-ray

DCM .....Dilated cardiomyopathy

DORV .....Double outlet right ventricle

ECG .....Electrocardiogram

ECHO .....Echocardiography

ESR .....Erythrocyte sedimentation rate

GABHS .....Group A-β hemolytic streptococci

HOCM .....Hypertrophic cardiomyopathy

LAP .....Long acting penicillin

PDA .....Patent ductus arteriosus

PS .....Pulmonary stenosis

RCM .....Restrictive cardiomyopathy

RF .....Rheumatic fever

RHD .....Rheumatic heart disease

SV .....Single ventricle

VSD .....Ventricular septal defect

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INTRODUCTION

Rheumatic fever and the more serious rheumatic heart disease have a greater impact on child health in developing countries than industrial countries. In Egypt, rheumatic heart disease is a significant health problem, with an estimated prevalence rate of 5.1 per 1000 school children.

The decrease in mortality from carditis and decrease in the symptoms of rheumatic heart disease are both related to the effective prevention of recurrence of rheumatic fever by a program of penicillin prophylaxis ***(Bassili et al., 2000).*** Although a 4 weekly prophylaxis schedule had been traditionally recommended, the World Health Organization (WHO) and the American Health Association now recommend a 3-weekly regimen for individuals living in high risk areas ***(Dajani, 1995)***. In fact, studies on the serum penicillin levels in Egyptian children have shown a drop in the serum penicillin concentration to below a therapeutic level during the third week following the injection of long acting penicillin. Consequently, a 2-weekly regimen is now implemented for Egyptian children rather than the previous 4-weekly regimen which led to a lower incidence of reoccurrence of the rheumatic activity ***(Kotby et al., 1996).*** However, compliance problems should be taken into account considering shorter treatment regimens.

Children with cardiomyopathy may present with a variety of signs or symptoms including congestive heart failure, arrhythmia, acute biochemical crisis, encephalopathy, generalized muscle weakness, dysmorphic features, or sudden death. They may as well present with cardiomyopathy as an accidental finding during evaluation of unrelated diseases. Despite improvements in medical therapy and increased availability of cardiac transplantation, cardiomyopathy remains one of the leading cardiac causes of death in children. In the majority of cases of pediatric cardiomyopathy, the cause is not identified and the progression is often grim ***(Schwartz et al., 1996).*** Studies have shown that intensive palliative treatment doesn't lead to better prognosis ***(Kumar et al., 1996)***. Reduction in morbidity and mortality associated with cardiomyopathy will require better understanding of its causes and pathogenesis so that etiology-specific therapies can be implemented ***(Schwartz et al., 1996)***.

Cardiovascular malformations are the most common type of congenital malformation ***(Wren et al., 2000).*** The estimated birth prevalence of cardiac malformations is 8 per 1000 ***(Wray and Senesky, 1999).*** It accounts for the majority of deaths from congenital defects in childhood, being six times more common than chromosomal abnormalities and four times more common than neural tube defects ***(Tworetzky et al., 2001).***

Impaired central nervous system function and developmental and cognitive impairment have been widely reported in children with congenital heart disease; especially those with cyanotic lesions. Surgical intervention for congenital heart disease can result in significant increase in IQ scores, especially among older children, and more specifically in those with cyanotic lesions. However, even after surgical correction of cyanotic lesions, considerable neuro-developmental and cognitive deficits may persist ***(Wray and Sensky, 1999)***.

Aim of the work

The aim of this retrospective study is to evaluate the patients following up at the Pediatric Cardiology Clinic, Pediatric Hospital, Ain Shams University for the assessment of the frequency of etiology, effectiveness of treatment, surgical intervention and the progression of these patients.

Study and Methods

All patients following up at Pediatric Cardiology Clinic, Pediatric Hospital, Ain Shams University will be identified from records and enrolled in the study.

Data Collection

Data will be collected from medical records and will include patients' demographic, clinical, radiologic, electro-cardiographic, echocardiographic evaluation, cardiac

catheterization assessment, preoperative cardiac and non cardiac diagnoses, complications, operative data if done and follow up including clinical status and as well as therapeutic measures.

Data analysis

Data will be coded & processed on P.C. computer on Microsoft access and analyzed on SPSS program.

INTRODUCTION

Rheumatic fever (RF) and rheumatic heart disease (RHD) remain a significant cause of cardiovascular morbidity and mortality in countries around the globe ***(Kaplan, 2005).*** Despite a documented decrease in the incidence of acute rheumatic fever acute (RF) and a similar documented decrease in the prevalence of RHD in industrialized countries during the past five decades, this non-suppurative cardiovascular sequel of group A streptococcal pharyngitis remains a medical and public health problem in both industrialized and industrializing countries even at the beginning of the 21st century. The most devastating effects are on children and young adults in their most productive years ***(WHO, 2004).***

Rheumatic fever (RF), a sequel of group A β-streptococcal throat infection, occurs in untreated susceptible children and is a multisystem inflammatory disease. The clinical signs of RF are the same throughout the world. In the 1950s, Jones established the major criteria for diagnosing initial attacks of RF, which comprised polyarthritis, carditis, and chorea. These criteria were revised in 1965 and the second revision in 1992 remains useful to date ***(Dajani et al., 1992).*** They have been periodically revised by American Heart Association in collaboration with other groups ***(Ferri, 2002)***. Arthritis is the earliest and most common feature of the disease,

present in 60%–80% of patients. Carditis, the most serious manifestation of the disease, occurs a few weeks after Streptococcus pyogenes throat infection in 30%–45% of RF patients, and usually presents as pancarditis. Endocarditis is the most serious sequel and frequently leads to chronic (RHD). Valvular lesions, especially mitral and aortic regurgitation, are the most common events caused by repeated valvulitis ***(Sampaio et al., 2007).***

Congenital heart disease (CHD), cardiomyopathy, and arrhythmias are also common causes of mortality and morbidity in infants and children, particularly during the perinatal period. Cardiac malformations present at birth are an important component of pediatric cardiovascular disease and constitute a major percentage of clinically significant birth defects, with an estimated prevalence of 4 to 50 per 1000 live births. For example, it is estimated that 4 to 10 live born infants per 1000 have a cardiac malformation, 40% of which are diagnosed in the first year of life ***(Moller et al., 1993)***. The true prevalence, however, may be much higher. For example, bicuspid aortic valve, the most common cardiac malformation, is usually excluded from this estimate. Bicuspid aortic valve is associated with considerable morbidity and mortality later in life and by itself occurs in 10 to 20 per 1000 in the general population ***(Ward, 2000).*** Recent studies are finding a high degree of heritability of bicuspid aortic valve, alone and with other cardiovascular anomalies, especially left ventricular outflow