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Late Diagnosis of Congenital Heart Disease - A Study Prior to Army Enlistment

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The exact incidence of congenital heart disease is unknown. The accepted incidence (6-8/1000 live birth) is based on studies of infants. A significant number of cases are diagnosed later because of lack of or missed early clinical signs. Bicuspid aortic valve and mitral valve prolapse are classic examples of these "late diagnoses".
Numerous other pathologies are also diagnosed later in life. Personal experience over many years with such cases led to a study of 52,000 young adults before admission to the army. The details of these cases were obtained from their files. Many of the positive cases were personally examined.
Cardiac abnormalities were found in 295 cases. Of these- 227 were known cases, diagnosed prior to army enlistment. Sixty eight cases were newly diagnosed. These included: Bicuspid aortic valve (13), aortic stenosis (4), aortic insufficiency (mild-7), aortic insufficiency (severe- 1), mitral valve prolapse (26), patent ductus arteriosus (2), pulmonic stenosis (10), Ebstein's anomaly (1), atrial septal defect (1), partial anomalous pulmonary venous return (1), coarctation of aorta (1) and Marfan syndrome with aortic dilataion (1). Six of these cases were sent directly to surgery and the remainder referred for follow up.
Pertinent features of these cases will be discussed.

Prevalence of Heart Lesions Related to BMI (Body Mass Index) in a Large Teenage Cohort

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Evaluation of a 22 year (birth dates from 1971-1994) cohort (113,694 from one geographical area of Israel) of army recruits between the ages of 16 and 19, revealed a statistically significant higher prevalence of diagnosed cardiac anomalies among underweight (<10th % BMI) recruits compared to normal weight recruits (10-85th %) and lower prevalence of diagnosed cardiac anomalies among overweight (85-95th %) and among obese (>95th %) recruits compared to the normal population. This trend is significant for males and females separately. The odds ratios for obese/ overweight/ normal/ and underweight are 0.66/ 0.74/ 1.0 (by definition)/ and 2.47 for males and 0.35/ 0.54/ 1.0 (by definition)/ and 1.47 for females. All P values are less than 0.05 and most less than 0.01. We will present subgroup analysis exploring the reasons for this trend.

Clinical Characteristics of Coronary Artery Disease in Adults with Congenital Heart Defects

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Background: There are now more adults with congenital heart disease than children. This aging cohort is at risk for acquired heart diseases such as coronary artery disease (CAD). The purpose of the study was to examine the clinical features of the CAD in adults with congenital heart disease.

Methods: A retrospective chart review was performed. The clinical characteristics of adults with congenital heart disease and angiographically confirmed atherosclerotic CAD were examined.

Results: One hundred and forty-one adults with CAD (69% male) were identified from a total of 12,124 patients (1%) seen in our congenital cardiac clinic. The most common cardiac diagnoses were atrial septal defect, bicuspid aortic valve disease, tetralogy of Fallot and coarctation of the aorta. We identified 7 adults with Eisenmenger physiology and CAD. The mean age of diagnosis of CAD was 56 +13 years. Twenty patients had premature CAD (14%) presenting before age 40 years. Traditional risk factors of patients with CAD were common and were present in the majority (82%) of patients. While many adults had symptoms of angina or myocardial infarction, a significant proportion (38%) were asymptomatic. The age at diagnosis in patients with coarctation of the aorta was younger than other subgroups (48+13 years). Seventy-seven percent (109/141) underwent percutaneous or surgical coronary interventions.

Conclusion: Atherosclerotic coronary artery disease may coexist with congenital heart disease. Coronary artery disease in adults with congenital heart disease typically occurs later in adulthood and in patients with traditional cardiovascular risk factors. This study highlights the need for cardiovascular risk factor screening and therapy when indicated.

Experience with Valve Sparing Aortic Root Replacement in Children and Young Adults

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Objective: Valve sparing aortic root replacement has been used successfully in adult patients with ascending aorta aneurysms, typically associated with connective tissue disorders. In children there is limited experience with this surgical technique, even though, it may prevent aortic valve replacement. We retrospectively reviewed our experience with this procedure in children.
Methods: 14 patients underwent valve sparing aortic root replacement from May 2003 to January 2011. Aged 5 to 28 years (mean 16 years). 5 patients had Marfan's syndrome with one patient presenting with acute type A aortic dissection. 3 patients had a previous Ross procedure, 4 patients with a single ventricle had a Fontan circulation, one patient had repair of pulmonary atresia and ventricular septal defect and one patient had a previous arterial switch operation. The preoperative root dimension ranged from 38 to 60 mm (mean 45). 6 patients developed more than 2+ aortic valve insufficiency
Results: There were no hospital deaths. Median postoperative ICU stay was 2.5 days and hospital stay was 5.5 days. One patient had a mild left hemiparesis that resolved. Follow up ranged from 10 months to 6.5 years (mean 3.3 years) with one sudden late death of a Marfan patient. There was one mechanical aortic valve replacement in a single ventricle patient 2 years after surgery. All 13 surviving patients are in New York Heart Association functional class I. 12 patients have no or mild aortic insufficiency.
Conclusion: Valve sparing aortic root replacement provides optimal ascending aorta reconstruction in multiple congenital defects with good medium term results. Longer follow-up will be needed for the aortic valve function.

Hybrid Procedures in the Treatment of Congenital Heart Disease

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The hybrid procedure, combining surgical and interventional catheterization procedures has been proposed as an alternative for initial palliation of patients with complex congenital heart disease. This approach to management has been introduced to minimize exposure to cardiopulmonary bypass, and improve outcomes for these infants, especially for those, whom the surgical procedure is very high risk.

We would like to present Safra/Sheba experience with the hybrid procedure.

The hybrid procedure was performed in the catheterization laboratory in four neonates. Via median sternotomy, both branch pulmonary arteries were banded with external band of 3.5 mm tube, and an interventional cardiologist inserted a delivery sheath under fluoroscopic guidance to deploy a stent through the arteriotomy in the ductus arteriosus to ensure long-term patency.

In this manner, antegrade flow to the descending aorta and retrograde flow to the aortic arch and cerebral and coronary vessels can be maintained. All procedures underwent successfully.

Preterm infants or those judged to be unsuitable for a prolonged cardiopulmonary bypass are selected for the hybrid pathway.

We conclude, The Hybrid Procedure - surgical bilateral pulmonary artery banding and interventional catheterization with ductal stenting, can be good alternative in selected patients with Hypoplastic left heart syndrome, who are considered high risk for standard Norwood-Sano surgery.

Palliative Arterial Switch Operation: A Review Of Fifteen Cases

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Palliative Arterial Switch Operation: A review of fifteen cases.

Background: This study is an examination of our unit's experience with palliative switch in univentricular and potentially biventricular hearts with transposition of great arteries. These patients were divided into three groups based on their physiology.

(a) Single ventricle physiology (8). All patients had univentricular hearts, transposition of great arteries, and systemic outflow tract obstruction (SOTO). Arterial switch is being recognized as an alternative to Norwood operation and modifications thereof. An arterial switch in these patients effectively converts the SOTO to pulmonary stenosis and thereby restricting pulmonary blood flow. There were 2 early deaths. Five patients have undergone Bidirectional Glenn. The advantage over the conventional Norwood procedure and its modifications is that it provides a valved ventriculo-pulmonary artery connection and thereby prevents diastolic run off.

(b) Borderline biventricular physiology (4). These patients had transposition of great arteries, ventricle septal defect and hypoplastic Right ventricles. They underwent arterial switch operation with pulmonary artery banding without ventricular septation thus retaining the option of a one and half or biventricular repair if the RV grows. There was 1 hospital death, 1 underwent one and half ventricle repair and the remaining two are awaiting subsequent management.

(c) Biventricular physiology (3). These patients had transposition of great arteries and multiple ventricular septal defects which could not have been closed successfully without a mutilating ventriculotomy. The arterial switch with pulmonary artery band converts the transposition physiology to a VSD physiology which allows effective banding without increasing the cyanosis. All patients made uneventful recovery. One underwent subsequent VSD closure and pulmonary artery plasty. The remaining two are asymptomatic and awaiting corrective surgery.