

ADULT CONGENITAL HEART DISEASE: A CHALLENGING POPULATION

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Summary

Congenital heart disease is a global problem, managed by interventional catheterization and surgery, hence most of these patients reach adult life. Such patients therefore require specialized care and a multidisciplinary approach. This article will briefly cover the epidemiology, types of patients, the need for specialized centers and the organization of services and care for these patients.

1. Introduction

Congenital heart diseases are abnormalities that occur as a result of a cardiac defect(s) during embryonic development of the cardiovascular system. If these defects – or others – do not lead to death of the fetus; the infant is born with a congenital heart disease. Adults in this context are defined as those who have reached the age of 16 years and older. In Egypt, from our experience, the problem is even more challenging owing to the lack of resources, insufficient public awareness and low socio-economic standards in addition to illiteracy and lack of proper patient records.

2. Epidemiology of Congenital heart disease

The prevalence of congenital heart disease (CHD) is approximately 8/1000 live births.

About 60% are diagnosed in infancy, 30% in children and 10% in adults above the age of 16 years.

Fifty years ago there were many more infants and children with CHD than in adults. Due to the great advances in early diagnosis, interventional and surgical techniques; today there are more adults with CHD. It is expected that >85% of infants born with CHD will reach adulthood.

Because of the lack of proper statistical studies and the diverse nature of the population of adults with CHD; it is difficult to put a true figure for the prevalence of this population. In the United Kingdom, it is calculated that there are at least 150,000 adults with CHD. In the USA they are about 1,000,000. In Egypt – a developing country – with a population of roughly 72,000,000 and a birth rate of 1.6% annually, the number of new CHD cases would be about 15,200 patients per year. Due to ignorance, socio-economic factors and differing surgical results, approximately 70% of those would reach adult life.

3. Types of adult patients with CHD

A. Patients diagnosed as adults:

A good number of CHD are compatible with adult survival and may not be discovered except late in life. Examples of such lesions are secundum atrial septal defects, mild forms of Ebstein's anomaly of the tricuspid valve, mild to moderate aortic and pulmonary valve stenosis. In countries like Egypt, due to the poorer socioeconomic situation and the lack of modern diagnostic facilities, this is a common occurrence.

B. Patients diagnosed in infancy and childhood:

Naturally these are the commonest group and can be divided according to whether they have received treatment or not.

- Mild lesions that only need follow-up. Bicuspid aortic valves are an example. Such patients need follow-up and endocarditis prophylaxis.
- Patients with significant or complex lesions who have had a surgical or catheter interventions. Most of these need regular follow-up at a specialized center for adult CHD.
- Patients who have neglected to seek the proper treatment especially if surgical. Again due to the parents' view of major heart surgery on their child, this is not uncommon in many parts of the world. For example a good number of adult tetralogy of Fallot patients are seen in Egypt. Eisenmenger's disease on top of a large left to right shunt is also common.

4. Congenital heart disease in adults – Unoperated survival

There are many patients with congenital heart defects that may reach adulthood. They can be divided into common and uncommon defects with expected adult survival and those with exceptional adult survival.

4.1 Common Defects with Expected Adult Survival

I Acyanotic malformations without a shunt, left-sided:

1. Bicuspid aortic valve
2. Coarctation of the aorta

II Acyanotic malformations without a shunt, right-sided:

1. Pulmonary valve stenosis

III Acyanotic malformations with left to right shunt

1. Shunt at atrial level – ASD II
2. Shunt at aortopulmonary level – Patent ductus arteriosus (PDA)

4.2 Common Defects with Exceptional Adult Survival

1. Ventricular septal defect(VSD)
2. Tetralogy of Fallot

4.3 Uncommon Defects with Expected Adult Survival

1. Situs inversus with dextrocardia
2. Congenital corrected TGA
3. Discrete subaortic stenosis
4. Ebstein's anomaly of the tricuspid valve(TV)
5. Pulmonary artery stenosis
6. Primary pulmonary hypertension
7. Partial AV septal defect
8. Sinus venosus ASD
9. Partial anomalous pulmonary venous connection (PAPVC)
10. Lutembacher's syndrome
11. Sinus of valsalva aneurysm
12. Pulmonary AV fistulae

4.4 uncommon defects with exceptional adult survival

1. Congenital obstruction to LA flow
2. Unicuspid aortic valve
3. Supravalvular AS
4. Vascular rings
5. Ventricular septal defect VSD with Double Outlet Right Ventricle (DORV) or Left ventricle (LV)
6. Anomalous origin of left coronary artery(LCA) from pulmonary trunk
7. Aortopulmonary window
8. Taussig- Bing anomaly
9. Truncus arteriosus
10. Univentricular heart
11. Complete Transposition of the great arteries TGA
12. Tricuspid atresia

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Biographical Sketch

Dr. Khalid Sorour, born in Cairo, Egypt 1950, is a medical graduate, from Cairo University. He pursued his postgraduate studies and training in Kasr El Ainy Teaching Hospital, which is the main teaching hospital for Cairo University. He was appointed in the Cardiology Department, Cairo University as Cardiology resident 1974- 1977, Assistant lecturer 1977-1983 , Lecturer 1983-1988, Assistant professor of Cardiology 1988-1993. He was appointed as Professor of Cardiology in 1993 to present. He also was a Post Doctoral Observer in Adult and Pediatric Cardiology, Baylor College of Medicine, Houston, Texas, USA 1987 and a Post Doctoral Observer in Coronary Interventions, Washington Hospital Center, Washington DC, USA 1990

In 2004-6 he became Head of the Catheter lab, Acting Head of Cardiology Dept. Benisuef Univ. 2003 to present . During his academic career he published a chapter on Rheumatic Fever and Rheumatic Heart Disease in Garson's et.al. *Science & Practice of Pediatric Cardiology*, supervised more than 30 thesis and

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Prof. Sorour is a member of the Egyptian Society of Cardiology, Society of Pediatric Cardiologists and Cardiac Surgeons and the Egyptian Hypertension society, member of the Pan Arab Congenital Heart Disease Association (PACHDA). He is also a participant in the Euro Heart Survey for Adult Congenital Heart Disease and PCI in coronary artery disease and member of the management team and head of clinical quality assurance of the Infective Endocarditis project Cairo University (HEEPF).