

## Adult Congenital Heart Disease Units: A Need for Attention in Congenital Heart Diseases Around the World

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Congenital heart disease (CHD) is the most frequent malformation. In the most recent data has a prevalence of thirteen cases per 1,000 children and six cases per 1,000 adults [1]. Nowadays more adults are living with CHD than children, especially in developed countries, and over 85% fall into the categories of moderate and high complexity defects according to the 2001 Bethesda classification [2]. Great strides in pediatric cardiovascular surgery have increased the number of survivors. Surgical and survival successes have turned the pediatric patient with CHD into an adult postoperative CHD patient. The adult CHD (ACHD) population is growing at a rate of 5-6% per year, and it is estimated that there will be 11% more per year in 2030 [3]. Currently estimate is that there are 3,000 ACHDs per million inhabitants [4]. Extrapolating these results, it is calculated that there are approximately 261,000 ACHDs in Central America, and more than 1.8 million in South America. In the United States (US), in 2010, 1.4 million ACHD were recorded vs one million children with CHD. Furthermore, the European scene shows a population of 2.3 million adults with CHD vs 1.9 million children [5]. The global population was 7.3 billion in 2016, there are estimate 4.4 billion adults. ACHD has a prevalence of 0.15-0.3%. Therefore, it is estimated that 15 to 20 million ACHD are found throughout the world; however 30-60% are lost to follow-up, due lack of guided transfer, insufficient availability of ACHD programs, inadequate insurance coverage, deficient education of patients and caregivers regarding ACHD, inadequate resources for patients with cognitive or psychosocial

impairment, lack of comprehensive case management, and different needs for evaluation and management.

The current ACHD population is derived from two groups [6]:

- 1- Newly diagnosed, they are 10-15% of cases, and are mostly simple or medium complexity CHDs, according to the 2001 Bethesda classification.
- 2- ACHDs who underwent surgery in childhood or adolescence, these make up 85-90% of cases, they are medium and high complexity CHD, and this is the group which will grow the most over the next 10-20 years.

The new ACHD population needs super-specialized healthcare, especially those having undergone surgery for complex defects in childhood, or who never had surgery. Should be centralized, and they require solutions to healthcare transition problems, knowledge of when to investigate each anomaly, they should be examined in non-pediatric settings. It is recommended that they be concentrated in adult congenital heart disease units (ACHDUs) with the necessary infrastructure and sufficient technological resources, and especially duly trained and skilled personnel.

In 1959, Canada was the first country to develop the world's first ACHDU, subsequently, the United Kingdom established the first ACHDU in Europe in 1964. More than 50 years have elapsed for Europe to have the highest number of ACHDUs per 10 million inhabitants (ACHDU/population ratio 3.6). However in the rest of the world ACHDU/population ratio is: North America (1.7), Oceania (1.5), South America (0.4), Asia (0.3) and Africa (0.1) [7].

The European and North American recommendations standards for ACHDU functioning have been described in the respective ACHD guidelines [8,9], and these are:

### **1-Staff Members**

- Adult/pediatric cardiologist with ACHD certification, at least 2
- ACHD imaging specialist (echocardiography, cardiac resonance, tomography), at least 2
- Congenital interventional cardiologist, at least 2
- CHD surgeon, at least 2
- Anesthesiologist with CHD experience, at least 2
- Invasive electro physiologist with ACHD experience, at least 1
- Psychologist, at least 1
- Social worker, at least 1
- Cardiovascular pathologist, at least 1

**2-Diagnostic Equipment:** electrocardiogram, continuous electrocardiographic monitoring (Holter), ambulatory blood pressure or event monitoring, ergometry and cardiopulmonary stress test, echocardiography, conventional radiology, computerized axial tomography (CT) and magnetic resonance (MR), diagnostic and

treatment catheterization laboratory, diagnostic and interventional (mapping systems) electrophysiology laboratory, pacemaker/defibrillator implantation and follow-up. Finally CHD heart surgery program.

The ACHDU should provide healthcare while maintaining a close relationship with the pediatric cardiology unit and/or a cardiovascular surgery service with an active CHD program. It should treat a critical mass of patients and carry out a minimum number of procedures to be effective and maintain quality and an adequate training level.

The training and certification in ACHD is really necessary, a training period of 24 months is recommended to complete subspecialty in ACHD. For pediatric and adult cardiologist these 24 months should include 18 months in a specialized ACHDU, and six months in general adult cardiology (coronary care unit, heart failure, arrhythmia, and outpatient clinic) for pediatric cardiologist trainees, and six months of pediatric cardiology (inpatient and clinic experience) for adult cardiology trainees [10]. There are three levels of training, in which the objectives are:

**Level 1:** basic training in problems affecting ACHD patients; this enables the individual to make decisions regarding when to refer patients to specialized healthcare, not to provide care for moderately or highly complex patients.

**Level 2:** additional training in ACDH to acquire experience in managing the whole spectrum of patients. The training ranges from six months to one year.

**Level 3:** a specialist completely trained in ACHD, with the necessary degree of knowledge to take on commitments in clinical and academic research. Requires at least two years of training.

Levels 2 and 3 can only be obtained at a center with an ACDHU.

Nowadays there is a very limited number of specialists in ACHD worldwide, and at the same time ACHDUs is very low in the most developing countries. According to the Adult Congenital Heart Association (<https://www.achaheart.org>), data available in 2018, in US there are currently 113 ACHD program [11], but only 30 accredited ACHD comprehensive care centers, and approximately 308 ACHD cardiologist certified by the board. With the current ACHD population in USA (1.4 million) the relationship ACHD cardiologist/patient is very low (0.1: 4,220). In my expert opinion is really necessary to create more centers for pediatric cardiovascular surgery in developing countries, while simultaneously fostering transition programs for adolescents to ACHD care. Also training cardiologist in ACHD, and a sufficient number of ACHDUs equipped for each country's needs. Adults with CHD are already a public health problem, and the treatment needs are pressing.

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## Conflicts of Interests

I have no conflicts of interest to declare.

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