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LETTER TO THE EDITOR

Adults with congenital heart disease: A growing public health problem?



Adultos con cardiopatía congénita: ¿un problema de salud pública creciente?

Dear Editor:

Congenital heart disease (CHD) is the most common defect; one out of every 100 live-born infants has CHD. Due to advances in pediatric cardiovascular surgery, today, over 85% of children with CHD survive to adulthood.¹ In Europe, there is an estimated population of 2.3 million adults with CHD (ACHD), which surpasses the pediatric population of 1.9 million.² In the United States (US), in 2010, 1.4 million ACHD were recorded vs. one million children with CHD.³ In Latin America, in 2009, Brazil calculated approximately 552,000 ACHD.⁴ Extrapolation of this data gives an estimated 1.2 million ACHD in South America. The ACHD population will continue to increase, with a 5–6% annual growth rate. The new ACHD population, known as GUCH (Grown-Up Congenital Heart Disease) will be a public health problem in the next 10–20 years, due to the rapid increase in complex CHD survivors (single ventricle heart, hypoplastic left heart), with new hybrid treatment techniques resulting in children operated on today becoming postoperative adults with CHD remnants, after-effects and complications.

The health systems in each country need to be alert to this growing problem. Many ACHD who are currently alive are a young, economically productive population. However, their health status oscillates between relapses and hospitalizations due to CHD decompensation. Hospitalization emergencies in ACHD include, in order of frequency, arrhythmia, heart failure, syncope, aortic dissection, endocarditis, thromboembolism, bleeding (noncerebral), aortic aneurysm/dissection, and sudden cardiac death⁵; and increase the health expenditures for their care. A retrospective observational cohort study in Canada, with data obtained from the Canadian Institute for Health Information (CIHI) between May 2004 and March 2014, showed that among 59,917 hospitalizations, the yearly costs increased 21.6%: from \$99.7 million CAD (95% confidence interval [CI], \$89.4–\$110.1 million) in 2004 to \$121.2 million (95% CI, \$112.8–\$129.6 million) in 2013 ($P < 0.001$). The cost was greater in adults (4.5%/year, $P < 0.001$) than in children (0.7%/year, $P = 0.006$). Adults represented 38.2% of the costs

in 2004, compared with 45.8% in 2013 ($P = 0.002$). The costs increased more in complex ACHD (7.2%, $P = 0.001$).⁶ The US analyzed ACHD healthcare costs from 2002 to 2012, finding an increase in charges (billed: USD \$543 million vs \$1.5 billion, a 178% increase; reimbursed: \$221 vs \$433 million, respectively, a 95% increase).⁷

Training specialists in ACHD are pertinent, along with structuring adult congenital heart disease units (ACHD Units). In 2015, the US identified 255 cardiology programs, with 93% having a university affiliation. Eighty-one programs were surveyed, and only one-third (29%) reported having a scholarship for ACHD cardiology training.⁸ It is clear that there is a very limited number of specialists worldwide. In 2014, the European Society of Cardiology (ESC) Working Group on Grown-up Congenital Heart Disease made recommendations for ACHD standards and organization of care, as well as for training in GUCH in Europe. A training period of 24 months was recommended to complete subspecialty training in GUCH. These 24 months should include 18 months in a specialized GUCH center and six months in general adult cardiology (including the coronary care unit, heart failure, arrhythmia, and outpatient clinic) for pediatric cardiology trainees, and six months of pediatric cardiology (including inpatient and clinic experience) for adult cardiology trainees. Alternatively, these six months could be undertaken in a specialized GUCH center with “learning experiences”.⁹ In the US, in December 2012, after a joint petition by the American Board of Pediatrics and the American Board of Internal Medicine, ACHD was formally recognized by the American Board of Medical Specialties as a subspecialty of adult and pediatric cardiology. As such, advanced training in ACHD is needed for an additional two years after completion of either pediatric cardiology or adult cardiovascular disease training.¹⁰ One ACHD Unit is needed for every 10 million inhabitants. In 2013, the number of ACHD Units per 10 million inhabitants was highest for Europe (3.6), followed by North America (1.7), Oceania (1.5), South America (0.4), Asia (0.3) and Africa (0.1).¹¹ Simple heart defects could be managed by general cardiologists; for moderate and highly complex defects, a subspecialized cardiologist is essential. Mortality in ACHD is reduced when their management adheres to the directives in the currently available guidelines.¹² In 1996, Canada became the first country to implement guidelines, followed by the ACC/AHA guidelines in 2008, the ESC guidelines for the management of grown-up congenital heart disease in 2010, and, more recently, the AHA guidelines

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for older patients (CHD > 40 years).^{13–16} The diverse problems that affect ACHD require the participation of several specialists, such as CHD cardiovascular surgeons, electrophysiologists, intensive therapists, cardiac rehabilitators, obstetrician-gynecologists, pediatric cardiologists, pulmonologists, and internists; all working as a team, coordinated by an ACHD subspecialist. The transition between pediatric and adult care is a fundamental component of management. The responsibility falls on the pediatric cardiologists interested in forming CHD Transition Units, as this is where healthy lifestyle advice, reproductive counseling, and recommendations on sports and physical activity and the role or profession a GUCH patient should choose, should be imparted. Currently, only one third of centers in Europe and the US have a formal CHD Transition Unit. Each country in America should implement its own health policies aimed at caring for GUCH patients, developing healthcare reference centers led by an expert ACHD cardiologist. A GUCH patient is a chronic patient; he or she should never be discharged from cardiology follow-up. Carole A. Warnes wrote: “the adult with congenital heart disease was born to be bad”.¹⁷

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