Need For Palliative, Corrective, and Second-Corrective Surgery in Adult Patients With Congenital Cardiopathy Classified According to Disease Complexity

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ABSTRACT

The prevalence of adult congenital heart disease has increased with advances in endovascular surgery and treatments. It is therefore essential to determine the need for surgery in these patients throughout their lives.

Based on a multi-center database on Grown-up congenital heart disease (GUCH), an observational study was performed to assess the need for palliative, corrective, and second-corrective surgery, in patients with adult congenital heart disease. Patients were classified as having MILD, MODERATE, or SEVERE anomalies. The association between need for surgery and severity of cardiopathy was analyzed using Kaplan Meier analysis.

Study population: 714 patients; age: 32 years ± 11.5; females: 55.3%. Open surgery 52.8%; Palliative surgery 15.3%; Corrective surgery 57.7%; reoperation, 11.2%. Mild cardiopathy: 26%; moderate cardiopathy: 54%; severe cardiopathy: 21.6%. None of the patients with mild, 9% of patients with moderate, and 48% of patients with severe cardiopathy needed palliative surgery before age 10 years. Patients not requiring corrective surgery before age 10-20-30-40 years: mild cardiopathy: 90%-84%-75%-70%, moderate cardiopathy: 50%-37%-30%-27%, severe cardiopathy: 52%-34%-29%-29%. Patients not requiring reoperation before age 10-20-30-40 years: mild cardiopathy: 92%-92%-92%-92%, moderate cardiopathy: 85%-81%-77%-75% and severe cardiopathy: 85%-70%-61%-61%, respectively. Half the patients with severe and moderate cardiopathy required corrective surgery in their first decade, and 70% required corrective surgery in their third decade of life.

The need for palliative and corrective reoperation was significantly higher in patients with severe congenital heart disease.

INTRODUCTION

As a result of advances in pediatric cardiology, cardiovascular surgery, and cardiac procedures in Congenital Heart Disease (CHD) in the last decade, 85% of children born with CHD survive to adulthood (1-2). This progress in therapeutics is expected to result in a greater number of adults than children with CHD (1). The growing population of adults with congenital heart disease (GUCH) poses a challenge to the cardiologist, who must acquire skills in pediatric and adult cardiology in order to provide comprehensive health care to patients who must face the consequences of successful therapy later in life, at an age that was previously unheard of for such patients.

These patients constitute a new clinical population that requires careful management, taking into account their congenital disease, the strategy employed to correct the disease/defect, and the complex course of the disease associated with the prolongation of the patient's life span. Such an approach is a challenge to the scientific community, in view of the scant knowledge and available evidence (3-6). The increase in survival is associated with the availability of multiple treatment options, according to the complexity of the disease.

GUCH disease includes a heterogeneous group of pathologies that require management by a team of specialists involving interventionists, expert image analysts, and adult cardiologists and surgeons, thus enabling a comprehensive approach to the congenital disease at different stages throughout the patient's life. The use of international nomenclature for diseases (D) and multi-center databases allows identifying the most frequent pathologies according to diagnosis, severity, need for surgery, and mortality, among other data (7). Such tools contribute to understanding this new group of patients.

MATERIALS AND METHODS

A longitudinal, retrospective, observational study was conducted. Medical records of patients with GUCH disease (GUTI-GUCH) seen at three hospitals in the city of Buenos Aires between April 2006 and April 2012 were retrieved from a single Database and analyzed. Records corresponding to 714 patients were retrieved and classified as Mild, Moderate, or Severe, according to the degree of complexity of GUCH, in keeping with the Guidelines for the Management of Adults with CHD of the AHA/ACCA, 2008 (11-12). The surgical procedure employed in each case was classified based on criteria similar to those used by Paladino et al, and each case was included in one of the following three groups, accordingly. Surgical procedures performed to improve the clinical status of the patient without restoring normal anatomy or physiology were classified as palliative surgery (PS). Interventions aimed at repairing physiology or anatomy were considered corrective or reparative surgery (CS). Interventions after a corrective surgery were classified as re-operation (RO) (13). Surgical procedures performed without previous sternotomy were arbitrarily classified as Closed Heart Surgery and those involving sternotomy were defined as Open Heart Surgery. The Kaplan Meier test was used to calculate time from birth to first PS or first CS, and from CS to first RO, and to estimate the probability of undergoing a surgical procedure (PS, CS, or RO) according to the severity of the cardiopathy. The Log Rank Test was used when considered appropriate. The obtained data were loaded on an Access-type database, and analyzed using a Pentium IV processor and SPSS17 software. Frequency distribution and percentages of categorical variables were calculated. Logistic regression analysis was used to control for confounding factors. The level of significance was set at an alpha value of 0.05, and 95% confidence intervals (CI) were calculated accordingly.

RESULTS

Mean age of patients was 32 years ± 11.59 (age range: 16.3-79.4), 55.3% (n=395) were women, and 53.2% (n=380) received treat-
ment at a public hospital. The obtained data showed that 23.9% of patients (n=171) underwent closed heart surgery and 52.8% (n=377) had open heart surgery; 15.3% (n=109) received PS, 57.7% (n=412) had CS, and 11.2% (n=80) needed RO.

According to degree of complexity, 26% of cases (n=186) were Mild, 52.4% (n=374) were Moderate, and 21.6% (n=154) were Severe. The distribution of type of surgery according to disease complexity is depicted in Table 2.

The probability of needing PS at 10 years was 0 (CI 95% 0-2.00) in the case of patients with mild CHD, 0.91 (CI 95% 6.46-12.52) in patients with moderate CHD, and 0.52 (CI 95% 40.62-56.84) in patients with severe CHD, and was significantly higher in patients with moderate CHD as compared to those with severe CHD.

No need for CS at 10, 20, 30 and 40 years of survival was observed in 90% (CI 84.28-93.57), 84% (CI 77.61-88.69), 75% (CI 67.74-80.67) and 70% (CI 62.67-76.27) of patients with mild cardiopathy; 50% (CI 44.82-55.17), 37% (CI 32.03-42.03), 30% (CI 25.40-34.91) and 27% (CI 22.38-31.58) of patients with moderate cardiopathy; and 52% (CI 43.78-60.01), 34% (26.47-41.88), 29% (CI 21.73-36.50) and 29% (CI 21.73-36.50) of patients with severe cardiopathy, respectively. The differences in need for CS were significantly different when comparing the group of patients with moderate and severe cardiopathy, with the group of patients with mild cardiopathy (Log Rank: p= 0.000) (Graph 2). The need for CS was significantly higher in the group of patients with moderate and severe cardiopathy as compared to those with mild cardiopathy (Log Rank: p= 0.000) (Graph 2).

Need for RO 15 years after CS was similar in the moderate and severe cardiopathy groups (19%) though progression of the disease was worse in patients with severe cardiopathy, with need for RO at 30 years reaching 39% of patients in this group as compared to 23% of patients in the moderate cardiopathy group.

The prognosis was better in the group of patients with mild cardiopathy, with two patients requiring RO (8%) 8 years after CS, and a similar percentage of patients needing RO at 30 years (Graph 3).

CONCLUSIONS
Half the patients with severe congenital cardiopathy underwent palliative and/or corrective surgery before the age of 10 years, and 71% of these patients had corrective or reconstructive surgery in the third decade of life.

Only 9% of patients with moderate congenital heart disease received palliative surgery during their life. Around 50% of patients with moderate or severe congenital heart disease had corrective surgery before the age of 10 years, and approximately 70% had corrective surgery at some point in their life.

Thirty-nine and twenty-three percent of patients with severe and moderate cardiopathy respectively needed reoperation 30 years after corrective surgery.

DISCUSSION
As a result of the increasing success of pediatric heart surgery, there are a growing number of adults with congenital heart disease. In addition, there are many patients with congenital heart disease over the age of 16 years who have never undergone surgery, either because they have not needed it or because the disease went undetected. These adult patients, referred to as “grown ups with congenital heart disease”, have special needs and require specialized management in their transition from the pediatric to the adult healthcare setting (14-15).

Moreover, and increasing number of GUCH patients will need heart surgery. A long-term (12-year) study performed by Srinathan et al. showed that in spite of the number of GUCH patients needing surgery at 3, 9, and 12 years after the first procedure was similar, the percentage of patients with mild cardiopathy who required surgery dropped from 46% in the first period to 28% in the last studied period, and the percentage of patients requiring RO increased from 25% to 50% (14-15).

Adults with congenital heart disease are an increasingly complex group of patients and their treatment poses a challenge to health professionals and researchers.

A study conducted by Marelli showed that at least half of the adult patients with congenital heart disease had a complex cardiopathy; only 30% were seen by physicians specialized in adult congenital heart diseases, and less than 15% of patients seen by specialists in GUCH had severe heart disease (16).

Over the last decades, a number of European, American, and Canadian research groups have reached consensus on the need for a standard international nomenclature that will allow unifying databases on this pathology around the world (17-19).

The first projects were conducted in 1990 when the Society of Thoracic Surgeons (STS), directed by Mavroudis, and the European Association for Cardio-thoracic Surgery (EACTS), directed by Elliot, created the Congenital Cardiac Surgery Database (17-18). The need for a common language (nomenclature), standard methods for gathering and reporting data, and strategies to ensure and verify entry of data, was recognized. In 1998, both societies developed the Congenital Heart Surgery Nomenclature and Database Project, which was published as the EACTS-STS International Nomenclature and Database in 2000 (19). That same year, the Association for European Pediatric Cardiology (AEP) published their nomenclature in the European Pediatric Cardiac Code (EPCC). But in 2002, the need to join criteria led to the creation of the International Society for Nomenclature of Pediatric and Congenital Heart Disease (ISNPCH). The latter, together with the subcommittee of the Nomenclature Working Group (NWG) directed by Tchervenkov, developed the IPCCC (International Pediatric and Congenital Cardiac Code), which allowed cross-mapping the following classification systems (20-23):

1- The IPCCC derived from the EACTS-STS International Nomenclature and Database
2- The IPCC derived from the EPCC of the AEP
3- The IPCC presented in the Fyler codes of Boston Children’s Hospital and Harvard University.

The cross-map, which combines the lists of diagnoses in each of the proposed nomenclatures, was presented at the 4th World Congress of Pediatric Cardiology and Cardiac Surgery held in Buenos Aires, Argentina, in September 2005.

Our research team has joined the initiative of the ISNPCH, adopting the International Pediatric and Congenital Cardiac Code of the EACTS/STS (20-22). A number of institutions share the GUTI-GUCH database developed at the Ricardo Gutierrez Children’s Hospital. This has allowed, and will allow in the future, having up-dated information on the population of patients with grown up congenital heart disease, in order to obtain epidemiological data regarding prevalence, incidence, complexity, diagnosis, therapeutic, and clinical course of the disease. Access to available information is currently limited to subscribing institutions, which include pediatric and adult health care centers in the City of Buenos Aires. Sharing this body of information facilitates the transition from the pediatric to the adult healthcare setting. In addition to the geographic limitation of the study sample, given that it is not representative of the entire population with GUCH in Argentina, the “centralizing” effect caused by the referral of grown up congenital heart disease patients to the specialized centers that
participated in the present study, must be pointed out. Nevertheless, the data obtained in this exploratory study allow establishing the initial situation diagnosis. It is clear that the need for RO increases with patient-age, and that it increases two decades after the first surgery. Furthermore, need for surgery is strongly associated with disease complexity. (15, 24) The high percentage of patients with tetralogy of Fallot needing reoperation, with almost 50% undergoing RO 30 years after the initial surgery, is a clear example (14).

The number of pulmonary valve replacements in patients with repaired tetralogy of Fallot increased exponentially at the Royal Brompton Hospital between 1993 and 2010, becoming more marked as of 2005 (26).

In agreement with other reports, our findings showed a progressive increase in the need for RO among moderate and severe cases, 10 years after the first corrective surgery. Unlike Srinathan et al. (15), we found that the percentage of patients with severe cardiopathy who underwent RO was higher than that of patients with moderate congenital heart disease. The difference between both studies may partly be explained by the difficulties the latter group of patients has accessing RO in our country, since limited health care resources and insufficient capacity to meet demand force healthcare providers to prioritize patients with more complex heart disease.

It is estimated that by the year 2020 there will be more adults than children with congenital heart disease (27). Thus, adult cardiologists will increasingly encounter these patients in their practice.

The paradigm has changed, and adult congenital heart disease bridges the gap between pediatric and adult cardiology. A number of research groups around the world, including our own, have embraced the challenge and are working in a specialized multidisciplinary fashion, using international nomenclature to provide quality health care to an emerging population of adults with congenital heart disease, and thus contribute to sharing experience

Conflicts of interest: The authors declare they have no conflict of interest

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Table 1: Prevalence related to the complexity of congenital heart disease.

<table>
<thead>
<tr>
<th>Type of CHD</th>
<th>Diagnosis of CHD</th>
<th>Num. of cases (%</th>
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</thead>
<tbody>
<tr>
<td>Severe CHD</td>
<td>Single Ventricle Defect</td>
<td>46 (29.9)</td>
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<td></td>
<td>Transposition of Great Vessels</td>
<td>23 (14.9)</td>
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<td></td>
<td>Pulmonary Atria with VSD</td>
<td>15 (9.7)</td>
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<td></td>
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<tr>
<td></td>
<td>Double outlet right ventricle</td>
<td>12 (7.8)</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Congenitally Corrected Transposition</td>
<td>12 (7.8)</td>
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<td></td>
<td>VSD with PHT†</td>
<td>10 (6.5)</td>
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<tr>
<td></td>
<td>ASD with PHT†</td>
<td>7 (4.5)</td>
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<tr>
<td></td>
<td>Pulmonary Atria with Intact Septum</td>
<td>5 (3.2)</td>
<td></td>
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<tr>
<td></td>
<td>AV Canal with PHT†</td>
<td>4 (2.6)</td>
<td></td>
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<tr>
<td></td>
<td>Others CC</td>
<td>20 (13.1)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>All severe CHD</td>
<td>154 (21.4)</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Moderate CHD</td>
<td>Atrioventricular Canal Defect</td>
<td>15 (4)</td>
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<td></td>
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<tr>
<td></td>
<td>Atresia</td>
<td>60 (16)</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>All moderate CHD</td>
<td>374 (52.4)</td>
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</table>

* Ventricular Septal Defect, † Pulmonary Hypertension, ‡ Atrial Septal Defect

Table 2: Type of surgery performed

<table>
<thead>
<tr>
<th>Heart Disease according to severity</th>
<th>Type of Surgery performed</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Open Heart Surgery</td>
</tr>
<tr>
<td>Mild CHD</td>
<td>%</td>
</tr>
<tr>
<td>Patients</td>
<td>%</td>
</tr>
<tr>
<td>Moderate CHD</td>
<td>%</td>
</tr>
<tr>
<td>Severe CHD</td>
<td>%</td>
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<tr>
<td>All CHD</td>
<td>%</td>
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Graph 1: Probability Curve of not needing palliative surgery, according to age and complexity of congenital heart disease.
Graph 2: Probability Curve of not needing corrective surgery, according to age and complexity of congenital heart disease.

Graph 3: Probability Curve of not needing second-corrective surgery, according to age and complexity of congenital heart disease.

REFERENCE