Research Paper

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

Medical Science

KEYWORDS : GUCH, Grown-up Congenital Heart Disease, Palliative Surgery, Corrective Surgery, Reoperation

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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 (GUTI-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: 32years ± 11.59; females: 55.3%. Open surgery 52.8%; Closed surgery 23.9%; Palliative surgery 15.3%; Corrective surgery 57.7%; reoperation, 11.2%. Mild cardiopathy: 26%; moderate cardiopathy: 52.4%; 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).

Moroever. 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 Mauvroudis, 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 (AEPC) 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 AEPC
3- The IPCCC 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, therapeutics, 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

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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 Bromptom 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.

Type of CHD	Diagnosis of CHD	Num. of cases (%)
Severe CHD		
	Single Ventricle Defects	46 (29.9)
	Transposition of Great Vessels	23 (14.9)
	Pulmonary Atresia with VSD*	15 (9.7)
	Double outlet right ventricle	12 (7.8)
	Congenitally Corrected Transposition	12 (7.8)
	VSD* with PHT†	10 (6.5)
	ASD‡ with PHT†	7 (4.5)
	Pulmonary Atresia with Intact Septum	5 (3.2)
	AV Canal with PHT†	4 (2.6)
	Others CC	20 (13.1)
	All severe CHD	154 (21.4)
Moderate CHD		
	Tetralogy of Fallot	91 (24.3)
	Aortic Valve Disease	54 (14.4)
	VSD* + other	40 (10.7)
	Coarctation of the Aorta	38 (8.6)
	ASD‡ + other	32 (8.6)
	Tricuspid Valve Disease	28 (7.5)
	Pulmonary Venous Anomalies	16 (4.3)
	Atrioventricular Canal Defect	15 (4)
	Others CC	60 (16)
	All moderate CHD	374 (52.4)
Mild CHD		
	ASD‡ isolated	86 (46.2)
	VSD* isolated	43 (23.1)
	Ductus	15 (8.1)
	Pulmonary Valve Disease	9 (4.8)
	Aortic Valve Disease	8 (4.3)
	Mitral Valve Disease	7 (3.8)
	Other Mild CHD	18 (9.7)
	All Mild CHD	186 (26.1)
All CHD		714 (100)

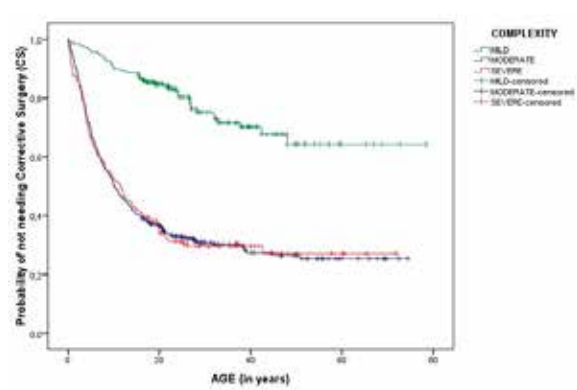
*** Ventricular Septal Defect, † Pulmonary Hypertension, ‡ Atrial Septal Defect.**

Table 2: Type of surgery performed

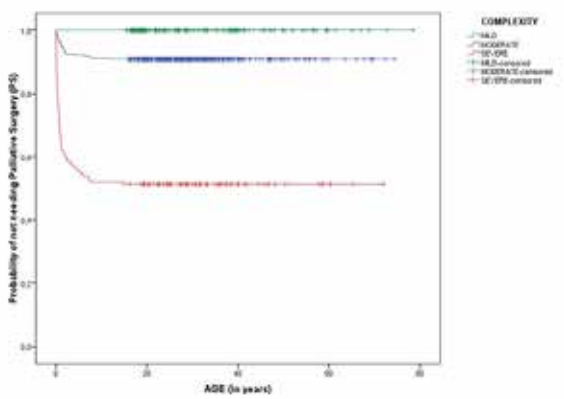
Heart Disease according to severity	Type of Surgery performed					
		Open Heart Surgery	Closed heart surgery	PS	CS	RO
Mild CHD	% CI	21 15.4 – 27.6	3.2 1.04 – 11.1	0 0	23.7 17.8 – 30.5	4.4 0.5 – 16.8
	Patients	39	6	0	44	2
Moderate CHD	% CI Patients	74 69.2 – 78.3 277	21.7 17.6 – 26.2 81	9.1 6.4 – 12.5 34	69.5 64.5 - 74 260	19.2 14.7 – 24.6 50
Severe CHD	% CI Patients	72.1 64.1 – 78.8 111	54.5 46.3 – 62.5 84	48.7 40.6 – 56.8 75	70.1 62.1 - 77 108	25.9 18.1 – 35.4 28
All CHD	% Patients	377 52.8	171 23.9	109 15.3	412 57.7	80 11.2

GRAPHS

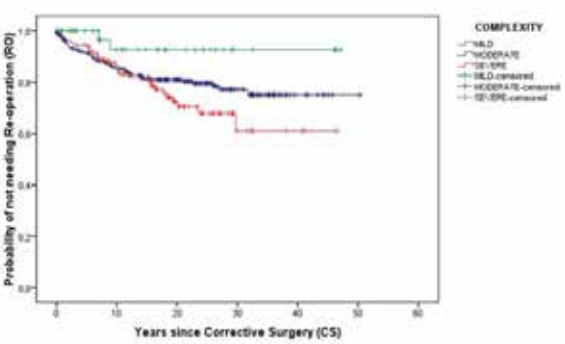
Graph 1: Probability Curve of not needing palliative surgery, according to age and complexity of congenital heart disease.



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

1.-Warnes CA: The adult with congenital heart disease: born to be bad? J. Am. Coll Cardiol. 2005, 46:1-8. | 2.-British Cardiac society. Report of the British Cardiac Society Working Party: Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart 2002, 88(suppl I): i1-14 | 3.-Casaldáliga J, Oliver JM, Subirana MT. Cardiopatías congénitas en la edad adulta. ¿Ficción o realidad? Introducción. Rev Esp Cardiol. 2009;09 (E): 1-2 | 4.-Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JI, et al: Task force 1: the changing profile of congenital heart disease in adult life. J. Am. Coll. Cardiol. 2001: 37:1170-5. | 5.-Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L: Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007, 115:163-72. | 6.-Mackie AS, Pilote L, Ionescu-Ittu R, Rahme E, Marelli AJ: Health care resource utilization in adults with congenital heart disease. Am J Cardiol. 2007, 99:839-43. | 7.-Report of the British Cardiac Society Working Party: Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart 2002, 88:1-14 | 8.-Erwin Notker Oechslin: Modelos de asistencia sanitaria en Europa y América del Norte. Rev Esp Cardiol. 2009, 09(E):3-12 | 9.-Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJ: Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. Eur Heart J. 2006, 27:1324-30. | 10.-Marelli AJ, Therrien J, Mackie AS, Ionescu-Ittu R, Pilote L: Planning the specialized care of adult congenital heart disease patients: from numbers to guidelines; an epidemiologic approach. Am Heart J. 2009, 157:1-8. | 11.-Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al: ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). Circulation. 2008, 118:2395-451. | 12- Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JL, Somerville J, Williams RG, Webb DG : Task Force 1: the changing profile of congenital heart disease in adult life. J. Am. Coll. Cardiol. 2001, 37:1170-1175 | 13-Padalino MA, Speggiorin S , Rizzoli G, Crupi G, Vida VL, Bernabei M et al: Midterm results of surgical intervention for congenital heart disease in adults: An Italian multicenter study. J Thorac Cardiovasc Surg 2007, 134:106-13 | 14.-Hickey EJ, Veldtman G, Bradley TJ, Gengsakul A, Manlhiot C, Willians W, Weeb G, McCrindle BW: Late risk of outcomes for adults with repaired tetralogy of Fallot from an inception cohort spanning four decades. European Journal of cardio-thoracic Surgery 2009, 35:156-166 | 15-Srinathan S K, Bonser RS, Sethia B, Thorne SA, Brawn W, Barron DJ: Changing practice of cardiac surgery in adult patients with congenital heart disease. Heart 2005, 91:207-12. | 16-Marelli AJ, Mackie AS, Ionescu-Ittu R, Rhame E, Pilote L: Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007, 115:163-172 | 17-Mavroudis C, Gevitz M, Ring WS, McIntosh CL, Schwartz M: The Society of Thoracic Surgeons National Heart Congenital Heart Database Report: analysis of the first harvest (1994-1997). Ann Thorac Sug. 1999,68:601-24. | 18-Jacobs JP: Software development, nomenclature schemes and mapping strategies for an international pediatric cardiac surgery database System. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2002,5:153-162. | 19.-Mavroudis C, Jacobs JP: Congenital Heart Surgery Nomenclature and Database Project: overview and minimum dataset. Ann Thorac Surg 2000, 69(4 Suppl): S1-S17. | 20.-Franklin RC, Anderson RH, Daniels O, Elliott MJ, Gewilling MH, Ghisla R, Krogmann ON, Ulmer HE, Stocker FP: Report of the Coding Committee of the Association for European Paediatric Cardiology. Cardiol Young 2002, 12:611-8. | 21.-Franklin, Jacobs JP, Tchervenkov CI, Béland M: The international nomenclature project for pediatric and congenital heart disease: bidirectional crossmap of the short lists of the EPCC and the International Congenital Heart Surgery Nomenclature and Database Project. Cardiol Young 2002, 12:431-435. | | 22.-Jacobs J et.al: Congenital Heart Surgery Databases around the world: Do we need a global database. Semin Thorac Cardiovasc surg Pediatr Card Surg Ann 2010, 13:3-19. | 23.-Franklin, Jacobs JP, Tchervenkov CI, Béland M: Update from the international working group for mapping and coding of nomenclature for paediatric and congenital heart disease. Cardiol Young 2004, 14:225-229. | 24.-Monro J: The changing state of surgery for adult congenital heart disease. Heart 2005, 91:139-140. | 25.-Lacour-Gayet F, Maruszewski B, Mavroudis C, Jacobs JP, Elliott MJ: Presentation of the International Nomenclature for Congenital Heart Surgery. The long way from nomenclature to collection of validated data at the EACTS. European Journal of Cardio-thoracic Surgery 2000, 18:128-135. | 26.-Babu-Narayan SV, Diller GP, et all: Clinical Outcomes of surgical pulmonary valve replacement after repair of Tetralogy of Fallot and potential prognostic value of preoperative cardiopulmonary exercise testing. Circulation 2014, 129:18-27. | 27.-Webb GD. Care of adults with congenital heart disease: a challenge for the new millennium. Thorac Cardiovasc Surg. 2001. 49;30-34 Webb GD et al. Care of adults with congenital heart disease: a challenge for the new millennium. Thorac Cardiovasc Surg 2001, 49:30-34 |