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Care of congenital heart disease in an adult cardiac surgery service

Atención de las cardiopatías congénitas en un servicio de cirugía cardíaca de adultos

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ABSTRACT

Introduction: congenital heart diseases (CHD) constitute the primary malformation at birth, and their diagnosis and treatment are regarded as one of the most significant triumphs of contemporary medicine. Pediatric cardiology has undergone substantial evolution, not only in diagnostic modalities but also in therapeutic approaches, thereby enabling a considerable proportion of patients to survive into adulthood. **Objective:** to ascertain the demographic and clinical characteristics, as well as the management, of adult patients with CHD who necessitated surgical repair at our institution. **Material:** we conducted a descriptive, cross-sectional study encompassing the period from 2008 to 2023, inclusive of all patients with a diagnosis of congenital heart disease who required surgical repair by our department. **Results:** a total of 128 patients were enrolled, with a slight predominance of males (53.2%). The most prevalent cardiac anomaly encountered was bicuspid aorta, accounting for 40.6% of cases, followed by atrial septal defect (18.75%). The mean age at diagnosis was 21 years, with only 1.5% of patients having a history of prior pediatric intervention. The overall mortality rate was 9.3%. **Conclusions:** the data garnered from this study indicate that delayed diagnosis of CHD remains a persistent issue, thereby underscoring the need for modifications to national public healthcare policies to ensure optimal management.

Keywords: congenital heart disease, cardiac surgery, adult.

RESUMEN

Introducción: *las cardiopatías congénitas (CC) constituyen la principal malformación presente al nacimiento, y su diagnóstico y tratamiento se consideran uno de los logros más significativos de la medicina contemporánea. La cardiología pediátrica ha experimentado una evolución sustancial, no solo en las modalidades diagnósticas, sino también en los enfoques terapéuticos, lo que ha permitido que una proporción considerable de pacientes sobreviva hasta la edad adulta.* ***Objetivo:*** *determinar las características demográficas y clínicas, así como el manejo, de los pacientes adultos con CC que requirieron reparación quirúrgica en nuestra institución.* ***Material:*** *realizamos un estudio descriptivo, transversal, que abarca el periodo comprendido entre 2008 y 2023, e incluye a todos los pacientes con diagnóstico de cardiopatía congénita que necesitaron reparación quirúrgica por parte de nuestro servicio.* ***Resultados:*** *se inscribió un total de 128 pacientes, con un ligero predominio del género masculino (53.2%). La anomalía cardíaca más prevalente fue la aorta bicúspide, que representó el 40.6% de los casos, seguida de la comunicación interatrial (18.8%). La edad media al diagnóstico fue de 21 años, y solo el 1.5% de los pacientes tenía antecedentes de intervención pediátrica previa. La tasa de mortalidad global fue del 9.3%.* ***Conclusiones:*** *los datos obtenidos en este estudio indican que persiste el problema del diagnóstico tardío de las CC, lo que subraya la necesidad de modificar las políticas nacionales de salud pública para garantizar un manejo óptimo.*

Palabras clave: *cardiopatía congénita, cirugía cardiaca, adulto.*

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Congenital heart disease (CHD) constitutes the most prevalent congenital malformation detected at birth. Historically, in the 1950s, the survival rate of children born with a cardiac anomaly was a mere 15%. However, contemporary advances have enabled over 90% of these individuals to survive into adulthood.[1] The global estimated incidence of CHD stands at 4-5 per 1,000 live births, although this metric remains undetermined in our country.[2] Among adult populations, the prevalence of CHD is approximately 0.35%, with 85-90% of these cases having a documented history of palliative surgery, corrective interventions, or other procedural measures undertaken during the pediatric stage.[3] Notwithstanding expert pediatric care, it is projected that roughly 20% of patients will necessitate surgical intervention during the initial 15 years of adulthood, with nearly half of this cohort requiring reoperation.

Advances in surgical techniques, perfusion methodologies, and perioperative care have culminated in a contemporary 45-year survival rate of 85%.[4] This substantial augmentation in survival rates, coupled with the altered natural history of the condition, has enabled patients with CHD to attain adulthood, thereby transcending the exclusive purview of pediatric care. This transitional phase, however, poses novel challenges pertaining to follow-up and management. Consequently, numerous countries have instituted transition clinics, which entail multidisciplinary specialist teams and facilitate comprehensive, longitudinal care for this patient cohort.[2,5] Regrettably, in our country, such specialized clinics are scarce, being confined to a limited number of centers, thereby resulting in a dearth of CHD care provision for adult patients.

MATERIAL

Our hospital's cardiovascular surgery service was established circa the 1990s; however, the decision to implement a dedicated database was only undertaken in 2008, hence the rationale for selecting this study period. We undertook a prospective, observational study encompassing all adult patients with CHD –defined as individuals over 15 years of age– who necessitated surgical repair between 2008 and 2023. The study protocol received approval from the hospital's ethics committee. Given that the variables under scrutiny were derived from routine clinical care data, the requirement for informed consent was waived.

Variables

Routinely collected data encompassed demographic characteristics, anatomical diagnoses, previous surgical histories, surgical procedure specifics, complications, and early mortality rates.

Statistical analysis

The primary objective was to ascertain the demographic, clinical, and management characteristics of adult patients with CHD requiring surgical repair at our institution. Statistical analyses were conducted utilizing IBM SPSS Statistics version 25. Continuous variables were expressed as means or medians, whereas categorical variables were represented as percentages, accompanied by minimum values where pertinent.

RESULTS

Demographic clinical and characteristics

A total of 128 surgeries for congenital heart disease were performed in adult patients with a slight male predominance 53.2% with a mean age of 35 years. The most common rhythm on admission was sinus rhythm in 97.2% followed by atrial fibrillation in 2.8%. The mean left ventricular ejection fraction was 62% (range, 48-73%), pulmonary artery systolic pressure was 31 mmHg (range, 28-62 mmHg) and right ventricle end diastolic volume was 101 ml/m^2 (range, 70-165 ml/m^2) *(Table 1)*.

The most common heart disease was bicuspid aortic valve (40.6%) followed by atrial septal defect (18.75%). Only two patients (1.56%) had a history of cardiac surgery

Table 1: Demographic characteristics and clinical presentation.

Variable	Value
Age (years), mean [range]	35 [16-75]
Weight (kg), mean [range]	69 [41-87]
Height (cm), mean [range]	161 [156-172]
Gender, n (%)	
Male	68 (53.19)
Female	60 (46.87)
Rhythm on admission, %	
Sinus	97.2
Atrial fibrillation	2.8
Echocardiographic findings, mean [range]	
Left ventricular ejection fraction (%)	62 [48-73]
Atrial fibrillation (%)	41 [34-52]
Right ventricular end diastolic volume (ml/m^2)	101 [70-165]
Pulmonary systolic arterial pressure (mmHg)	31 [28-62]
Associated comorbidities, n (%)	
Diabetes mellitus type 2 (DM2)	12 (9.3)
Hypertension (HAS)	24 (18.75)
DM2 and HAS	16 (12.5)
DM2, HAS, and Obesity	5 (3.9)
None	71 (55.0)

Table 2: Clinical description.

Variable	n (%)
Type of heart disease	
Bicuspid aortic valve	52 (40.6)
Atrial septal defect	24 (18.8)
Ventricular septal defect	20 (15.6)
Atrial and ventricular septal defects	6 (4.6)
Coarctation of the aorta	4 (3.1)
Partial anomalous pulmonary venous connection	4 (3.1)
Subvalvular aortic stenosis	4 (3.1)
Ventricular septal defect and patent ductus arteriosus	4 (3.1)
Unicuspid aortic valve	1 (0.7)
Patent ductus arteriosus	3 (2.3)
Tetralogy of Fallot	2 (1.5)
Ebstein's anomaly	2 (1.5)
Pulmonary insufficiency	2 (1.5)
History of previous heart disease	2 (1.5)
Age at diagnosis (years), mean [range]	21 [1-63]
Age at surgery (years), mean [range]	35 [16-75]
Complications	15 (11.7)
Bleeding	12 (9.3)
Severe paravalvular leak	2 (1.5)
Ischemia	1 (0.7)

in their pediatric population. *Table 2* describes the diagnosis, associated anomalies, type of previous surgical procedure, age at diagnosis and age at surgery.

Characteristics of the surgical procedures, complications and hospital stay

The mean duration of cardiopulmonary bypass time and aortic cross-clamping time were 155 minutes and 122 minutes, respectively. The most commonly used cardioplegia solution was Custodiol in 98% of cases. The mean estimated blood loss was 231 ml. Ninety six percent of patients required aminergic support with dobutamine being the primary treatment in 73%, followed by adrenaline in 18% and levosimendan in 9%.

Regarding the type of surgical procedure, 6.25% underwent minimally invasive cardiac surgery, out of which five correspondents to mechanical valve replacement in the aortic position and three patients with closure of interatrial communication.

Among the postoperative complications, the most common was bleeding in 9.3% of cases, followed by severe valvular leak in 1.56%. The mean hospital stay was seven days. In hospital survival was 90.6%. The main causes were cardiogenic shock in 6 cases, followed by hypovolemic and septic shock in three cases each. *Table 3* shows the surgical characteristics of the performed procedures.

DISCUSSION

Cardiac surgery for CHD in adult populations is becoming increasingly prevalent, with our center performing approximately eight such cases annually. The two predominant cardiac conditions treated within our cohort were bicuspid aortic valve and atrial septal defect. This aligns with existing literature, wherein bicuspid aortic valve, treated in 40.6% of cases at our institution, is recognized as the most common CHD in the general population.[6] Given its extreme heterogeneity across the clinical spectrum, bicuspid aortic valve can manifest as a subclinical entity throughout life or, at the more severe end of the spectrum, present with severe valvular dysfunction.[7] Amidst this interindividual clinical dispersion, it is well-established that complications during mid-adult life are common, conferring a substantially higher morbidity burden compared to another CHD.[6,8] It is therefore anticipated that this entity would constitute the most prevalent condition within our cohort. With regard to atrial septal defect, which accounted for 18.75% of cases, some literature posits this as the primary cardiac condition encountered in adult populations, affecting 40% of adults with CHD.[9] Analogous to bicuspid aorta, the clinical presentation of atrial septal defect is characterized by extreme variability, contingent upon factors such as defect size, vascular resistance, and comorbidities. In many instances, atrial septal defect can remain asymptomatic until adulthood, manifesting with nonspecific symptoms such as recurrent respiratory tract infections or cardiac arrhythmias, the latter being a common feature in our series. Notably,

Table 3: Characteristics of surgical procedures.

Variable	n (%)
Cardiopulmonary bypass time (minutes), mean [range]	155 [45-307]
Aortic cross clamping time (minutes), mean [range]	122 [29-180]
Cardioplegia type, %	
Custodiol	98
Del Nido	2
Surgical bleeding (ml), mean [range]	231 [30-1,370]
Excessive bleeding, %	11.7
Valve position	60 (100.0)
Mitral	3 (2.3)
Aortic	53 (41.4)
Tricuspid	2 (1.5)
Pulmonary	2 (1.5)
Type of prosthetic valve	
Mechanical	45 (75.0)
Biological	15 (25.0)
Reintervention first 24 hours postsurgical	7 (5.4)
Mortality	12 (9.3)

cyanotic pathologies were also treated within our cohort, specifically two patients presenting with Tetralogy of Fallot and Ebstein's anomaly. These conditions are predominantly observed in pediatric populations, with the majority of cases necessitating reoperation. Tetralogy of Fallot represents the most frequent cyanotic CHD in pediatric population, typically presenting within the first year of life. Survival into adulthood without corrective surgery or with palliative measures alone is exceptional.[10] In the case of our patients, neither had a history of prior corrective or palliative surgery; however, their ages at presentation were 16 and 22 years, respectively. Ebstein's anomaly, a rare condition accounting for less than 1% of all CHD cases,[11] was also encountered. The severity of this anomaly is determined by the degree of atrialization and tricuspid regurgitation, which, in turn, dictates the type of surgical intervention, symptomatology, and other clinical parameters. The optimal timing for surgical intervention remains a subject of controversy, generally being guided by a combination of factors including diminished functional class, paradoxical embolism, progressive cardiomegaly on chest radiography, and progressive right ventricular dilatation.[11,12] Within our series, these patients underwent surgery at the ages of 17 and 19 years, respectively.

In terms of complexity, approximately 50% of CHD in adult populations are classified as simple, 30-40% as moderate, and fewer than 15% as severe.[13] Notably, less than 10% of CHD cases are diagnosed after the age of 18 years.[14] The mean age at which surgical intervention was undertaken in our cohort was 21 years, with some patients having been diagnosed as early as one year of age. One of the primary challenges confronting pediatric cardiology is the phenomenon of loss to follow-up among adolescent patients, as highlighted by Liu et al.,[15] who identified four distinct domains that pose barriers to consistent follow-up care. The transition from pediatric to adult care is fraught with multiple challenges, including the absence of a seamless handoff process, the establishment of a trusting relationship, and the fostering of an appropriate balance between patient autonomy and understanding of their condition. Additional logistical challenges, such as time constraints, geographical distance, cost considerations, and the availability of specialized care, are further compounded by psychosocial factors and the inherent heterogeneity of cardiac abnormalities affecting our patient population. Patients with CHD constitute a diverse cohort with disparate priorities and needs, suggesting that a more individualized approach may be necessary to ensure adequate follow-up care. However, it can be generally stated that many of these factors are applicable to our population and may contribute to the lack of follow-up care during the pediatric stage, thereby explaining why these patients received care in adulthood. Cardiac surgery for adult patients with CHD, when performed in a referral hospital with a multidisciplinary team, is associated with low mortality rates and improved functional class. Surgical indications are primarily guided by the onset of symptoms and prognostic considerations. A thorough understanding of the patient's anatomical characteristics, previous surgical interventions, structural interventions, and their sequelae is essential for optimal patient care.[3,16] The overall mortality rate at our institution stands at 9.4%, which is comparable to rates reported in the existing literature, such as the studies by Garcia Cruz et al.[3] and Horer et al.,[17] which documented mortality rates of 12.3 and 10%, respectively. It is noteworthy that the former study exclusively encompassed complex pathologies, excluding patients with previous surgical interventions, given that prior sternotomy represents a high risk for bleeding and other surgical complications. In the study by Horer et al.,[17] the specific types of heart disease were not delineated. Within our series, we observed a predominance of patients requiring aortic valve replacement, which, as previously mentioned, constituted the primary type of CHD treated.

CONCLUSIONS

The decision to undertake cardiac surgery in adult patients with CHD represents a paradigm shift for both the clinical and surgical teams. A thorough clinical and functional assessment, coupled with the identification of comorbidities, is imperative in determining the optimal surgical approach for each individual case. In conclusion, it can be stated that late diagnosis is frequently the primary factor underlying the presentation of CHD in adult populations, thereby underscoring the necessity for physicians who are adequately trained to provide timely and effective care for these patients. The establishment of an early care system, facilitating surgical interventions during the earliest stages of life, is crucial.

Study limitations

It is acknowledged that this is a single-center study, which may have inherent limitations in terms of generalizability.

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