Adults with congenital heart disease: The experience of the Department of Cardiology Kairouan Tunisia

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Summary

Introduction : Advances in diagnosis, surgery and percutaneous interventions in the care of children born with heart defects have generated a growing population of adult survivors and spawned a new subspecialty of cardiology: adult congenital heart disease.

Objective: This study aimed to assess the demographics, comorbidities, and health care use of adults with congenital heart disease (ACHD) attending department of cardiology ibn el jazzar hospital kairouan Tunisia.

Materials and Methods: This is a retrospective, observational, and descriptive study over a period of 5 years (from 2018 to 2022), including adult patients aged 18 and above with congenital heart diseases. Data were collected from echocardiographic records.

Results: Our study included 22 cases. Patient mean age was 37±15 years and 16 were female. Dyspnea was the most frequent symptom.6 of the patients had complex forms of CHD and 10 had moderate abnormalities. The main congenital heart diseases were atrioventricular canal defect (3/12 and left-sided obstructive lesions (4/22). The main complications were rhythm and conduction disorders, pulmonary arterial hypertension and infective endocarditis (13.6%).

Conclusion: We clarified the clinical characteristics of adults with CHD referred to our department of cardiology. The diagnostic and therapeutic management of these patients requires specialized human skills and well-equipped technical facilities within specialized functional units. Adults with congenital heart diseases represent a highly emerging population in our country.

Keywords

Congenital heart disease – Adult – Prognosis – Treatment-Rhythm disorders

Résumé

Mots-clés

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INTRODUCTION

Congenital heart diseases (CHD) represent the most common congenital malformations, affecting approximately 1% of births[1] .Today, thanks to advancements in prenatal screening, pediatric cardiology, cardiac surgery, catheterization, and cardiac resuscitation worldwide, life expectancy has significantly improved, allowing 85-90% of children with CHD to survive into adulthood[2]. Thus, the population of adults with congenital heart disease now represents two-thirds of the total population of congenital heart diseases, a figure that continues to steadily increase. The overall prevalence of congenital heart diseases in adults ranges between 3/1000 and 6/1000 and varies from one country to another. However, it is still underestimated globally due to a lack of comprehensive databases, especially in developing countries [1.2]

In Tunisia, as in most regions of the world, there is currently no national registry for this emerging population, making it difficult to specify the epidemiological characteristics of this adult group and their care needs in our Tunisian context.

Our contribution in this context involves conducting a retrospective study, including all the patients followed in the "Cardiology" department at "CHU-Ibn Jazzar" in Kairouan. Our department is a polyvalent center witch deals with many varieties of cardiac diseases in adults (ischemic, heart failure, cardiomyopathy, etc.) but also congenital heart diseases.

This study aims to describe the epidemiological and clinical characteristics of this particular population, specifying the different types of heart diseases and the main follow-up challenges for each type of heart disease.

MATERIALS & METHODS

This is a retrospective study including all cases of adult CHD patients collected in the cardiology department at CHU Ibn Jazzar Kairouan over a period of 3 years. Data collection was performed using patient medical records, hospitalization registers, follow-up echocardiography reports, and telephone records.

Statistical analysis was performed using SPSS 20.0. Quantitative variables were presented as mean ±standard deviation or median (interquartile). Qualitative variables were presented as effectives and percentages.

RESULTS

We recorded 22 patients aged between 20 and 65 years old with an average age of 37 years. The majority of our study population (46%) falls within the age range of 21 to 30 years. 16 were female. Left-sided obstructive lesions were discovered at an early age compared to other types of cardiopathies, with an average age of discovery of 28.6 years. Right-sided obstructions are better tolerated, with an average age of discovery at 50 years.

The diagnosis of cardiopathy was made in 9 women of childbearing age, with 3 being pregnant at the time of diagnosis. Dyspnea was the most frequent presenting symptom in one-third of cases, accompanied by a cardiac murmur. Women were more affected for all types of cardiopathies. All pathologies with a left-sided obstruction were in females.

General symptoms included cyanosis and digital clubbing noted in 2 patients, and fever in 2 patients. Physical examination revealed a cardiac murmur in 7 cases and cardiac arrhythmia in 3 patients. No facial dysmorphia or thoracic deformity was observed. Table I summarizes the clinical data of the studied population.

The electrocardiogram showed a sinus rhythm in 20 patients; right bundle branch block (RBBB) in 5 patients; supraventricular extrasystole in 3 cases; high-degree atrioventricular block (AVB) in 3 cases (two cases of AVB 2:1 and one case of complete AVB), and left ventricular hypertrophy (LVH) observed in 3 cases.

Echocardiography (ECHO) allowed the diagnosis in all patients. It showed left ventricular hypertrophy in 7 cases; pulmonary arterial hypertension (PAH) noted in 2 cases, and Bicuspid aortic valve (BAV) in 3 cases. Various other complementary examinations were performed in our study population; thoracic CT scans (4) and diagnostic cardiac catheterization (4) were the most noted exams, followed by rhythm holter monitoring (3) and transesophageal echocardiography (3); then ambulatory blood pressure monitoring (2), and finally cardiac MRI, which was performed in only one patient.

Following the cardiac explorations, we found various congenital heart diseases grouped in Table II.

	Diagnostic	Age	Sex	Clinical signs	Complications	Treatment
Cas n° 1	AVC intermediate + MR gradeIII	40 years old	female	lipothymia	*BAV 2/1 *MR grade 3	Failure of 1st treatment 2nd complete treatment + PM
Cas n° 2	AVC partial	24 years old	female	Palpitations	*Supraventricular hyperexcitability	Failure of the 1st Cure Surgical (large residual CIA)
Cas n° 3	ASD	42 years old	female	* Palpitations *Dyspnea	* Supraventricular * hyperexcitability	Waiting for percutaneous closure
Cas n° 4	VSD restrictive peri membranous	65 years old	Male	* Souffle		monitoring
Cas n° 5	supravalvular aortic stenosis (max gradient 90 mm hg)	24 years old	female	Souffle *Enceinte 26 SA	*Endocarditis with multiple embolizations	Full-term delivery Postpartum surgery
Cas n° 6	sub valvular aortic stenosis tunnel valve	44 years old	female	Dyspnea Souffle	* Endocarditis with vegetations intra-LV, on the aortic valve, on the mitral valve	Surgery with discharge under ECMO (LV dysfunction) Death on postoperative day 3
Cas n° 7	sub valvular aortic stenosis , subaortic membrane	29 years old	female	Dyspnea Stade 2, Chest pain		Waiting for surgery
Cas n° 8	Stenosing bicuspid disease (GM at 55 mm hg) Operated for childhood coarctation	26 years old	female	* Fever	Endocarditis	Death while waiting for surgery
Cas n° 9	Bicuspid + aorta dilated to 40 mm	29 years old	female	Enceinte	Aucun	Uneventful delivery/CS
Cas n° 10	Bicuspid aortic + Tight isthmic coarctation	20 years old	female		*stroke hemorrhagic (angio-MRI: intracranial aneurysm) * HP	Treatment of the aneurysm and then sent for percutaneous treatment of the coarctation.
Cas n° 11	Operated infundibular pulmonary stenosis + massive tricuspid leak, borderline RV function	40 years old	Male	Souffle Dyspnea		Indication for RVT by bioprosthesis Postoperative death
Cas n° 12	Tight valvular RP with max gradient at 80 mmhg	60 years old	female	DyspneaSouffle		Percutaneous balloon dilatation with good results.
Cas n° 13	Corrected L-TGV (double mismatch) with borderline systemic RV function, moderate tricuspid valve leak	36 years old	female	Pregnant in the 3rd trimester Slow heart rate auscultation.	Complete AVB with fine QRS and HR at 60 b/min	installation of peri-C/S systolic electrotraining probe, delivery without incident, still in BAV with HR at 60 b/min
Cas n° 14	Complex heart disease: VDDI, APSO with MAPCA in PAH severe (Eisenmenger) + Initial aorta at 50mm and probable coarctation.	28 years old	female	Pregnant 14 SA Cyanosis (SPO2 at 78%)	Currently being explored (CT + KT)	Indication for ITG refused by the patient but spontaneous abortion.
Cas n° 15	Double entry UV in Eisenmenger	57 years old	Male	Loss of consciousness + convulsive seizures SPO2=70% Ht=75%, Hb = 22g/dl		bleeding hydration because HT>65% despite hydration
Cas n° 16	large ASD in Eisenmenger	23 years old	Male	Dyspnea SPO2 = 80% Ht = 60% Hb= 18g/dl		Hydration And put under bosentant
Cas n° 17	Ebstein's disease having a tricuspid plasty at the age of 5 then a tricuspid bio prosthesis in 2019	42 years old	Male	Dyspnea and shortness of breath	Degeneration of the prosthesis which has become stenosing and leaking	Proposed for replacement of the bio prosthesis
Cas n° 19	partial AVC	41 years old	Male	Lipothymia and dyspnea	BAV Mobytz II IM grad II	Complete repair with PM
Cas n° 20	Wide patent ductus arteriosus	42 years old	female	Dyspnea and chest pain	Left heart failure with LVEF at 42%	Percutaneous canal closure
Cas n° 21	Ebstien's disease type B	32 years old	female	Palpitation	Supraventricular tachycardia on accessory pathway	Ablation of the accessory pathway by radio frequency
Cas n° 22	Corrected L_TGV (Double discordance) with banding of the pulmonary artery at the age of 20	44 years old	female	Dyspnea on exertion and palpitation	Systemic ventricle with moderately impaired function with EF estimated at 52% Ventricular and supraventricular hyperexcitability	Heart failure treatment and antiarrhythmic therapy

Tableau 2. Distribution of CCs according to their level of complexity			
Complexity Degree	Type	Cardiopathy	N
simple CC		**small isolated CIV**	1
Moderate CC	Left_ right shunt	Large ASDOS	2
		Ostiumprimum ASD	1
		AVC (intermediateor partial)	3
		CAP LARGE TUBULAIR	1
	Right Obstacles	pulmonary valve stenosis	1
		Severe pulmonary infundibular stenosis	1
		Subvalvular or supravalvular aortic stenosis	3
	Left obstacles	Bicuspid + Coarctation of the aorta	1
		Stenosing bicuspid	1
		Bicuspid disease + Aortic aneurysm	1
Severe CC	Cyanogenic heart disease	- UV Double entry SUV in Eisenmenger	1
		- VDDI, APSO with MAPCA: VDDI in severe PAH (Eisenmenger)	1
		- Ebstein pathology	2
	others	Corrected transposition of large Vx	2

The classification adopted in our study is the anatomical classification mentioned in the 2018 ACC/AHA and 2020 ESC recommendations concerning the management of congenital heart diseases in adults. The distribution according to the severity showed a predominance of intermediate complexity congenital heart diseases (68.8%) and complex ones (27.2%). The congenital heart disease was simple in 4.5% of cases, as illustrated in Table I.

Thus, our population included 3 patients with AVC (atrioventricular canal) 1 ASD, 1 VSD, 3 non valvular aortic stenosis, 3 aortic bicuspids, 2 pulmonary stenosis, 2 Corrected L-TGV, 1 Double entry UV in Eisenmenger ,1 Complex heart disease:VDDI,APSO ,2 Ebstein's disease and 1 Wide patent ductus arteriosus.

Six patients were not proposed for surgical treatment or interventional procedures. This includes the cases of simple congenital heart disease (a small restrictive ventricular septal defect (VSD)), moderate to complex heart disease, like double discordance and aortic dilation to 40 mm in bicuspid aortic valve in a pregnant woman, which only require regular following according to specific guidelines. Finally, 3 cases were judged inoperable for Eisenmenger Syndrome, requiring symptomatic medical treatment. The remaining patients (15 cases) underwent or were proposed for surgical treatment or interventional catheterization.

One patient underwent valvular pulmonary dilation with a good result. Three patients proposed for a procedure are still awaiting (1 case of closure of the atrial septal defect; and one case each of stent dilation of coarctation of the aorta, closure of arterial duct). One patient underwent radiofrequency ablation of their accessory pathway. A patient had a pacemaker implanted for complete AVB on double discordance with L-TGV.

Six patients underwent curative surgical treatment in adulthood (3 atrioventricular canal repairs + infundibular stenosis and significant tricuspid regurgitation + subaortic stenosis + supra-aortic stenosis). One patient was operated in childhood for coarctation. Two patients are still waiting for surgery (sub valvular Rao and replacement of tricuspid bioprosthesis in Ebstein anomaly). One patient died while waiting for surgery.

Postoperative mortality in our series was noted in 2 patients. The overall reintervention rate in our case series was noted in 3 patients: 2 cases of atrioventricular canal repairs, infundibular stenosis + tricuspid regurgitation. None of our patients underwent interventional palliative treatment.

Complications noted in our patients were mainly rhythm and conduction disorders (4 cases), infective endocarditis (3 cases), pulmonary arterial hypertension with Eisenmenger syndrome (3 cases), hypertension (1 case), and a hemorrhagic stroke in a single patient.

The hospitalization rate in our series was 46%. The mortality rate in our series was noted in 3 patients.

Within the obstetric data available, pregnancy was noted in (4/15) of women of childbearing age All pregnancies were conceived without medical advice. The diagnosis of cardiopathy was suspected in 3/4 of cases by the obstetrician upon the discovery of auscultatory anomalies or cyanosis.

One of the pregnancies was complicated by miscarriage (in the patient with DORV+ APSO in Eisenmenger).

Three deliveries by cesarean section at term: the Double discordance with complete AV block and placement of a pacing lead during the procedure; the Supravalvular aortic stenosis and the Bicuspid aortic valve with aortic dilation to 40 mm.

DISCUSSION

A notre connaissance, peu d'études tunisiennes se sont intéressées aux facteurs expliquant le taux faible d'atteinte du LDL c cible dans notre population tunisienne.

oivent être confirmés par des études multicentriques et un échantillon de plus grande taille avec un délai de suivi plus long.

the CHD prevalence is estimated at 6.12‰ in France [1] and Canada (Quebec) [2], 6.16‰ in the United States [2], and between 2.8‰ and 4.9‰ in Switzerland [4]. It is higher in Senegal (7.5‰) [5] and Mexico (7.8‰) [2]. The difference in prevalence between countries can be explained by the various research methodologies used and the presence or absence of national registries and comprehensive databases for this population.

Nevertheless, this sample of adults with congenital heart disease seems sufficient to provide estimates and information on the sociodemographic data of this emerging population and its specific care needs. Thus, we hope that our study will be a prelude to future studies and the creation of national registries in collaboration with different university hospitals to ensure that this growing and emerging population has appropriate access to specialized cardiac care in a safe and sustainable manner, within specialized centers.

The average age of our patients (37 years) was close to that reported by Giannakoulas et al. (38 years) in Greece [6], younger compared to Karsenty et al. in Toulouse (40 years) [7]. The highest average age was reported by Agarwal et al. in the United States (57.5 years) [8]. The most represented age group in our series was between 21 and 30 years, aligning with the results of Mbaye et al. [5] and Engelfriet et al. [9].

All types of congenital heart diseases in our patients were evidently more prevalent in females. 80% of our patients were female (F/M = 3). This result does not coincide with the literature data, where Aleman-Ortiz et al. (F/M = 1.4) [10], Engelfriet et al. [9], and Giannakoulas et al. (F/M = 1.08) [6], as well as Karsenty et al. (F/M = 1.56) in his study at Toulouse University Hospital [9], reported different gender ratios.

According to the Type of Congenital Heart Disease (CHD); Atrial Septal Defect (ASD) was consistently the most common CHD in adults among various authors. However, in our series, it represented only 9.0%. This difference in the prevalence of ASD may be explained by delayed diagnosis in our context. Many ASD patients are often pauci-symptomatic with good exercise tolerance for a long time, leading to delayed consultation (average age of ASD diagnosis in our series was 42 years). Additionally, the murmur associated with ASD is generally not intense and may escape detection during routine clinical examination by a family physician. Ventricular Septal Defect (VSD): VSD in our series represented 4.5%. In the literature, VSD accounts for 9.4% in George et al.'s study, 15.2% for Engelfriet, and 14.3% for Verheugt [11,12,13]. In Ejim et al.'s study in Nigeria, there was a predominance of VSD (31%) in their population.

The prevalence of Atrioventricular Canal (AVC) in our series (13.6%) was higher compared to literature reports. This rate is close to that noted by Mbaye et al. in Senegal (10%) but much higher than the estimate by the AHA, which ranges between 4% and 5% of all CHDs [14]. The average age of AVA diagnosis in our patients was 32 years.

Concerning the left Obstructions;: The incidence of Coarctation of the Aorta (CoA)in our series is 4.5%, aligning with Karsenty et al. (7%) and higher than Aleman-Ortiz et al. (4.8%) in Mexico, but lower than the rates reported by Engelfriet et al. (13%), Strange et al. (12%), and Verheught et al. (10%) [7.10.15].Subvalvular Aortic Obstruction (SAO) includes a range of anomalies, from a simple obstruction (membrane or fibromuscular) to a tunnel-like obstruction, sometimes associated with hypertrophic cardiomyopathy, abnormal insertion of the mitral valve, posterior displacement of the infundibular septum, Shone's syndrome, or other left ventricular outflow tract obstructions. In our series, the average age was 33 years. Supravalvular Aortic Obstruction (SAO) is a rare form of aortic narrowing. In our study, the patient was 29 years old. In our series, there were 3 cases of bicuspid aortic valves associated with other anomalies. Bicuspid aortic valve represented only about 7% of aortic stenoses operated on in Haddad et al.'s series, with patients being younger (average age of 37 ± 21

years) compared to Western populations [12.16].

Right Obstructions; for pulmonary stenosis, it represented 9.0% in our study, close to Mbaye's study (14%) but higher than those reported in most of the literature by Al-Balushi et al., Ejim et al., and Verheugt et al. [11,17,18].

Complex Heart Disease and Eisenmenger Syndrome: In our series, the prevalence of Eisenmenger syndrome (13.6%) is notably lower than that noted by Mbaye et al. and a study conducted in 2010 at the European GP Hospital in France. This prevalence is also lower than that reported in European guidelines, estimated at 3.5%. The rate of Eisenmenger syndrome is likely underestimated in our series, as many patients are followed in pneumology and internal medicine by the pulmonary hypertension unit, not all of whom are included in our study, and others are lost to follow-up or deceased.

According to the Complexity of CHD and regarding the complexity level of CHDs, the majority of lesions in our patients were moderate (68.1%), and complex (27.7%), while simple lesions accounted for only 4.5%. There is a difference in the distribution of CHDs according to their complexity in different studies:

The rate of moderate CHDs in our series was higher than that of simple CHDs, aligning with the results of Mbaye et al. in Senegal and Van Bulck et al. in 8 European countries. However, in Giannakoulas et al.'s and Karsenty et al.'s studies, the rate of simple CHDs was relatively higher than that of moderate CHDs.

The rate of complex CHDs (27%) was close to that reported in the literature by Agarwal et al. (25.2%) and G.P. Diller et al. (24.6%) [8.19]. Complex CHDs often require highly specialized care and therapeutic management with human skills and optimal technical facilities, which are not always available in our context.

In terms of symptoms, dyspnea was the most common (33.3%), which is far from the literature data. In the study by Engelfriet et al., dyspnea represents (63%), for Favelli et al. (73%) [12,20]. Other discovery circumstances were heart murmurs, palpitations, or incidental discovery. Other signs included palpitations, murmurs, and cyanosis.

ECG remains an essential examination in the assessment of congenital heart diseases in adults. It is important to obtain an initial ECG to compare it with any subsequent ECGs. However, the ECG is not sufficient for the detection of paroxysmal and nonsustained arrhythmias, hence the use of 24-hour Holter monitoring, which was performed in 18.2% of our patients, often due to the onset of new symptoms (mainly palpitations, dizziness, lightheadedness, or syncope).

TTE performed in all our patients allowed for a diagnosis in all cases. The need for another complementary examination to confirm and refine certain details was necessary in 31.8% of cases. Unlike in children, the limitations of TTE in adults are mainly reduced echogenicity in some patients and the difficulty of acoustic windows, especially the subcostal view. Transesophageal echocardiography was performed in 27% of cases, compared to 8% in the study by Mbaye et al. [5] and 6% in that of Engelfriet et al. [9]. The indications for TEE in our series mainly involved ASD proposed for interventional treatment to conduct a precise anatomical study of the defect and its edges with possible 3D analysis, and suspicions of infective endocarditis.

Diagnostic cardiac catheterization was performed in 18.2% of cases vs. 22% in Mbaye et al. [5]. Diagnostic catheterization in adult CC is an important tool for morphological and especially hemodynamic diagnosis, particularly in confirming pulmonary arterial hypertension (PAH).

Thoracic CT was requested in 18.2% of our patients, mainly in the presence of ASD in addition to catheterization, bicuspid aortic valve, aortic dilatation, and 1 case of coarctation. The rate of resorting to Angio scan in patients is not well known in the literature. Angio scan allows for a morphological study, especially of extracardiac vascular elements, and is therefore an important tool for primary diagnosis, but its role in monitoring is limited due to the risk of radiation and the use of contrast agents.

Cardiac MRI has a broad indication in the field of CC in adults, especially during follow-up. It plays a significant role in morphological study (cardiac and extracardiac vascular structures) and hemodynamic study (quantification of leaks, ventricular functions) in several CC. In our study, cardiac MRI was indicated in accordance with international recommendations in 4.5% of cases.

In our study, 24-hour Ambulatory Blood Pressure Monitoring (ABPM) was indicated during the follow-up of our patients with CoA (9.0%) to identify their blood pressure profile at rest and during physiological effort to unmask persistent or recurrent hypertension after coarctation repair. Indeed, systemic hypertension in the upper body is common in patients with unoperated coarctation and may persist in nearly one-third of patients who

have undergone successful surgical or interventional catheterization [14]. Hypertension can be noted in the absence of any hemodynamic gradient in this population, requiring regular monitoring.

The most frequent complications found in our study were Arrhythmias, Pulmonary Arterial Hypertension (PAH),Chronic Heart Failure ,Infective Endocarditis (IE),Mortality and Reintervention Issues.

The entire spectrum of arrhythmias can be encountered in adults with congenital heart disease (ACHD), with several subtypes that often coexist. Bradyarrhythmias may involve abnormalities of the sinoatrial node,atrioventricular node,or His-Purkinje system. It has been estimated that approximately 50% of patients will develop atrial tachyarrhythmia during their lifetime [21]. Ventricular arrhythmias are considered the leading cause of sudden death in several subtypes of congenital heart disease, with an overall risk that is up to 100 times higher than in age-matched controls [22]. Fortunately, the absolute risk of these fatal events remains relatively low, at about 0.1% per year. In our study, supraventricular tachyarrhythmias were noted in 13.6% of our patients, mainly represented by supraventricular hyperexcitability without sustained events. Rhythm disturbances accounted for 18% according to Engelfriet et al. Favelli et al., as well as Verheugt et al., found them in 14.8%, 16.34%, respectively [11,20].

A proportion of patients with congenital heart disease, especially significant left-to-right shunts and left heart obstructive lesions, develop pulmonary arterial hypertension (PAH). The heterogeneity of PAH forms associated with congenital heart disease is represented in the new classification of PAH, as it can be part of Group 1 (shunts causing pulmonary arterial hypertension), Group 2 (PAH secondary to left heart obstructive lesions), or Group 5 (segmental PAH) [23]. Eisenmenger syndrome is the extreme form, defined as ACHD with initially significant systemic-to-pulmonary shunt, which in turn induces severe pulmonary vascular disease.The prognosis of Eisenmenger syndrome is grim but remains better than idiopathic PAH with significant functional limitation associated with late-stage right heart failure.The incidence in our patients (13.6%) was high compared to that reported in the literature, in Favelli et al. (6%), Haddad et al. (5%), Karsenty et al. (12.6%), and Aleman-Ortiz et al. (15.58%) [10]. PAH constitutes a formidable evolving complication of left-to-right shunts, and its high incidence in our series can be explained by the delay in repairing these congenital heart diseases in our patients. This delay is due either to loss of follow-up, lack of resources, or sometimes to the delayed indication for repair by pediatricians or treating cardiologists not accustomed to managing congenital heart diseases.

Chronic heart failure is a frequent complication in the literature. The pathophysiological mechanisms vary according to the type of heart disease, but most congenital heart malformations, whether operated on or not, have a potential risk of evolving towards heart failure. It has been shown that adults with congenital heart disease tolerate a lower functional capacity with fewer symptoms than patients with acquired heart disease [24]. In our study, only one patient experienced a heart failure episode. The incidence of heart failure in Karsenty et al.'s study (27.4%) [7] was higher but remains much higher than that noted by Chan et al. in Canada (6.3%) [25]. However, it is less significant than that reported in Senegal by Mbaye et al. (30%) [5].

Infective endocarditis was observed in 13.6% of our patients, compared to 2.9% in Karsenty et al.'s study and 3% in Engelfriet et al.'s study [7.9].The incidence of IE in our context is higher than that reported in the literature. The most frequently identified potential entry point is oral-dental, emphasizing the need to educate these patients about good oral hygiene and regular visits to dentists, and possibly antibiotic prophylaxis surrounding dental care procedures in high-risk heart diseases. Despite recommendations on endocarditis prophylaxis, its prevalence in congenital heart diseases remains problematic. Its incidence is higher than the general population (11/100,000 patients-year vs. 1.5 to 6/100,000 patients-year) [26]. It affects men with ACHD twice as often as women [27]. Patients at the highest risk are those with cyanotic congenital heart disease, left heart lesions, atrioventricular canal, and surgery in the last 6 months. A recent Swedish registry found a 20 to 30 times higher risk of IE for patients with isolated ventricular septal defects compared to the general population, without significantly affecting their outcome [28]. It is important to note that adults with congenital heart disease frequently have

at-risk foreign bodies such as probes or devices and valved or non-valved tubes that often require several changes throughout their lives.

IE on CIA prostheses has been described. The incidence of IE on the Melody valve is increased compared to pulmonary homografts, but the risk factors remain to be precisely determined [29,30]. Like acquired heart diseases, the diagnosis of IE is often delayed, and IE should be considered in any patient with congenital heart

Mortalitywas noted in 13.6% of our patients, a result higher than that reported by Mbaye et al. in Senegal (1%) and close to that of Engelfriet et al. (2.5%) [5.9]. In our study, no deaths were reported in the medical records of patients hospitalized in the department. It is noteworthy that 2 out of 3 patients died in the postoperative period.

The third case of death was related to septic shock complicating a lingering endocarditis. According to recent literature data, heart failure (HF) is considered the leading cause of death in patients [1]. It is associated with significant morbidity and mortality (20%) in adults with congenital heart disease [1].

The reintervention rate was estimated at 50% in our case series, higher than that found in the literature by Karsenty et al. (30%) and Strange et al. (21%) [7.15]. Among the heart diseases requiring reintervention in our series were partial and intermediate atrioventricular canals (CAV) due to the persistence of a significant shunt. Special attention must be given to the quality of surgical procedures, especially for certain heart diseases such as CAV surgery. This is a delicate procedure that cannot be equated with routine closure surgery for atrial or ventricular septal defects. It requires specific expertise and, in our view, should only be performed in specialized congenital heart disease centers. The rational goal is to reduce the incidence of residual lesions, the rate of reinterventions, and deaths due to heart failure.

Loss to follow-up is a common issue in this population. Studies in Canada, Germany, the United Kingdom, and the United States have shown that 21 to 76% of adolescents and young adults with congenital heart disease are either lost to follow-up or receive inappropriate follow-up from centers with insufficient experience in managing such patients [1]. Several risk factors for loss to follow-up have been identified, such as male gender, absence of previous intervention, low complexity of congenital heart disease, or lack of information on adult follow-up [1]. In our study, the rate of patients lost to follow-up or receiving inappropriate follow-up represents 20% of patients, a relatively high rate. The primary cause of loss to follow-up in our patients was a lack of information about the importance of adult follow-up, especially among patients who believe they are cured after intervention for their congenital heart disease. Other causes of inappropriate follow-up in our context were the lack of financial means for patients to travel to hospitals with a specialized adult congenital heart disease cardiologist.

Physical activity is a major problem directly impacting the quality of life and cardiac status of patients. Nowadays, it is widely accepted that physical activity is not only beneficial in the long term for quality of life but also in terms of morbidity and mortality [31]. Many studies have shown that exercise programs in adults with congenital heart disease are both safe and improve physical performance and quality of life [32–33]. Even in patients with pulmonary arterial hypertension (PAH), there is noted benefit when the physical program is carefully chosen [34,35]. However, 1/3 of adults with congenital heart disease regularly engage in moderate physical activity, and 1/3 have no physical activity at an average age of 26 years [36]. It is important to inform both the child and their family that a sports contraindication is exceptionally required, unlike the previous practices of most pediatric cardiologists who prohibited sports under the pretext of «you have a heart condition.»

Sexual activity is an important component of the patient's and partner's quality of life. According to AHA recommendations [37], sexual activity is safe for most patients with congenital heart disease. However, it is important to address this issue to prevent health problems related to risky sexual practices and unwanted and/or complicated pregnancies in complex heart conditions. Adolescent and young female adults with congenital heart disease are sexually less active than their peers; however, they are more likely to engage in risky sexual practices such as lack of contraception and the use of drugs or alcohol during intercourse [38]. Pregnancy and contraception are crucial in this population and both pose potential risks, often overlooked [39.40]. In fact, only half of women contraindicated for pregnancy

report having received this information, and about half have not been informed about contraceptives. Additionally, 28% of high-risk pregnancy patients do not use contraception during sexual intercourse [41]. Moreover, nearly half of adult female patients have contraception contraindicated with their heart condition and/or have not been informed about pregnancy risks. The congenital heart disease specialist and, in conjunction, the gynecologist can provide expertise on the type of contraception compatible with the patient's heart condition and the risks associated with pregnancy [17]. Pregnancy was noted in 56% of our women of childbearing age. This incidence is close to that of Karsenty et al. (48.9%) [7], in the General Dutch population (40%) [7], and in Engelfriet et al. in Europe (36%) [9]. 75% of pregnancies were in women with moderate congenital heart disease. All pregnancies were conceived without medical advice. Moreover, one woman was lost to follow-up and only consulted after the start of pregnancy. In 3/4 of cases, congenital heart disease was discovered during pregnancy.

One of the main objectives of our study is to analyze the various medical and non-medical problems encountered by patients with congenital heart disease in their care journey. This study has highlighted some dysfunctions at different stages, from diagnosis to follow-up, the transition problem, and the lack of specialized centers for optimal reception and follow-up in adulthood. According to the recommendations of the ACC/AHA and ESC, the management of patients with complex or intermediate-severity congenital heart disease [1] cannot be conceived by the general cardiologist alone and must be done in close collaboration with a specialist in this field (Class I C). For low-risk patients (simple congenital heart disease group) [42], it is recommended to have at least one evaluation in a regional specialized center to determine the follow-up (Class I C). The expertise of a congenital heart disease center is based on human skills (highly qualified medical and paramedical personnel for the management of adult congenital heart disease) and a technical platform meeting standards. Recommendations from scientific societies specify the organizational aspects of specialized centers, the required training standards for medical and paramedical personnel [1], with specific training in electrophysiology, imaging, catheterization, and cardiac surgery.

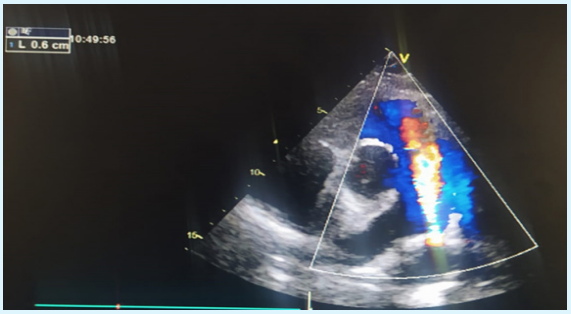

Figure 1. Ductus arteriosus 6 mm wide in a 42-year-old woman

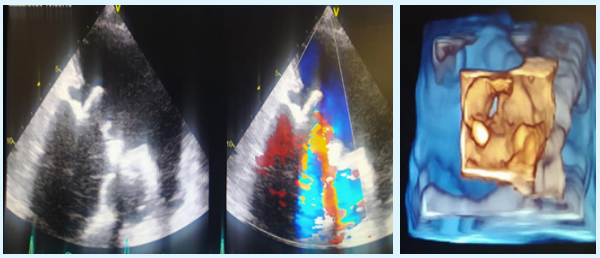

Figure 2. Ebsteine's disease with degeneration of the tricuspid prosthesis which has become stenosing and leaking

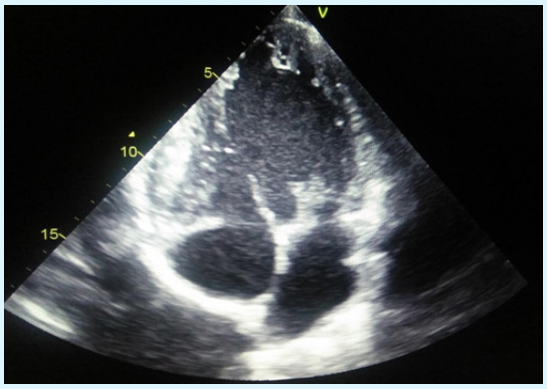

Figure 3. Double-inlet single ventricle

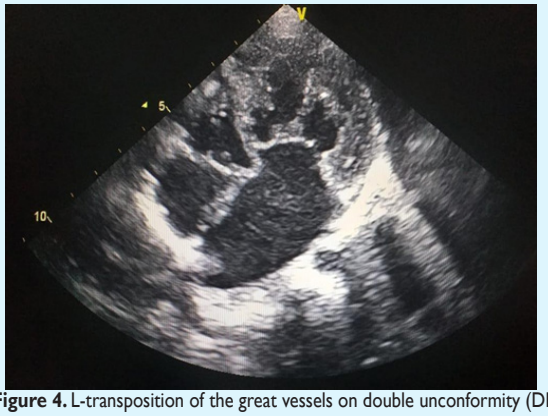

Figure 4. L-transposition of the great vessels on double unconformity (DD)

CONCLUSION

This study has allowed us to achieve the main objective. Thus, we have demonstrated that the incidence of adult congenital heart diseases is not rare, both globally and in the Tunisian context. This is an emerging population, with the prevalence of these congenital heart diseases steadily increasing, estimated at 3,000-6,000 per million inhabitants.

We have also shown that these adults exhibit residual lesions and complications, primarily hemodynamic and rhythmic, depending on the type of heart disease and corrections performed. The complications are of a cardiological nature, including rhythm or conduction disorders and pulmonary hypertension, or extra cardiological, requiring the involvement of various specialists (particularly gynecologists and obstetricians for contraception and pregnancy monitoring).

The diagnosis relies on transthoracic echocardiography as a first-line examination, requiring specific expertise in adult congenital heart diseases, with occasional recourse to transesophageal echocardiography. Invasive exploration through catheterization, and non-invasive methods using other imaging techniques (CT angiography, MRI) may be essential to complete the assessment and guide the management of these heart diseases.

Several therapeutic advances have been made in the management of adult congenital heart diseases, especially in the field of interventional catheterization in reference centers and the emergence of new therapies for pulmonary hypertension. Catheterization has, in some cases, replaced surgery with similar performance and a low morbidity rate. This has significantly contributed to improving the quality of life and prognosis of this population.

The management of patients faces numerous challenges: sometimes highly complex heart diseases, loss of follow-up during the transition from pediatrics to adulthood, and return to the healthcare system sometimes in an emergency context. The care of these adults should include genetic counseling, education, and information for patients on endocarditis prophylaxis, pregnancy and contraception types, physical activity, and socio-professional integration.

Follow-up in expert centers with multidisciplinary specialties ensures a better quality of life for these patients, especially those with complex congenital heart diseases. Through this work, we have attempted to provide an overview of congenital heart diseases in adults—a rapidly emerging population requiring specialized care. We hope that this study contributes to a comprehensive vision for managing the transition, organizing care, and improving the care of these patients, which can only be achieved within specialized units for adult congenital heart diseases.

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