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CLINICAL RESEARCH

Characteristics and outcomes of heart failure-related hospitalization in adults with congenital heart disease

Caractéristiques et résultats des hospitalisations liées à une insuffisance cardiaque chez le patient adulte avec une cardiopathie congénitale

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KEYWORDS
Heart failure;
Congenital heart defect;
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Arrhythmia

Summary
Background. — Heart failure (HF) is the main cause of death in adult congenital heart disease (ACHD).
Aims. — We aimed to characterize HF-related hospitalization of patients with ACHD, and to determine HF risk factors and prognosis in this population.

Abbreviations: ACHD, adult congenital heart disease; BNP, B-type natriuretic peptide; CHD, congenital heart disease; CI, confidence interval; HF, heart failure; PAH, pulmonary arterial hypertension.

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Methods. — We prospectively included 471 patients with ACHD admitted to our unit over 24 months. Clinical and biological data and HF management were recorded. Major cardiovascular events were recorded for ACHD with HF.
Results. — HF was the main reason for hospitalization in 13% of cases (76/583 hospitalizations). Patients with HF were significantly older (median age 44 ± 14 years vs. 37 ± 15 years; $P < 0.01$), with more complex congenital heart disease ($P = 0.04$). In the multivariable analysis, pulmonary arterial hypertension (odds ratio [OR] 6.2, 95% confidence interval [CI] 3.5—10.7), history of HF (OR 9.8, 95% CI 5.7—16.8) and history of atrial arrhythmia (OR 3.6, 95% CI 2.2—5.9) were significant risk factors for HF-related admissions ($P < 0.001$). The mean hospital stay of patients with HF was longer (12.2 vs. 6.9 days; $P < 0.01$), and 25% of patients required intensive care. Overall, 11/55 (20%) patients with HF died, 10/55 (18%) were readmitted for HF, and 6/55 (11%) had heart transplantation during the median follow-up of 18 months (95% CI 14—20 months). The risk of cardiovascular events was 19-fold higher after HF-related hospitalization.
Conclusions. — HF is emerging as a leading cause of morbidity and mortality in the ACHD population. Earlier diagnosis and more active management are required to improve outcomes of HF in ACHD.

MOTS CLÉS
Insuffisance cardiaque ;
Cardiopathie congénitale ;
Hypertension artérielle pulmonaire ;
Arythmie

Résumé
Contexte. — l'insuffisance cardiaque (IC) est la principale cause de décès chez les patients adultes atteints d'une cardiopathie congénitale (ACC).
Objectifs. — Nous avons cherché à caractériser les hospitalisations en rapport avec l'IC du patient ACC et à déterminer les facteurs de risque et le pronostic de l'IC dans cette population.
Méthodes. — Au total, 471 patients ACC admis dans notre unité sur une période de 24 mois ont été prospectivement inclus. Les données cliniques et biologiques, ainsi que la prise en charge de l'IC ont été recueillies. La survenue d'événements cardiovasculaires majeurs après IC au cours du suivi a été également relevée.
Résultats. — l'IC était le motif d'hospitalisation principal dans 13 % des cas (76/583 hospitalisations). Les patients IC étaient significativement plus âgés (âge médian 44 ± 14 vs 37 ± 15 ans ; $p < 0{,}01$), avec des cardiopathies congénitales plus complexes ($p = 0{,}04$). En analyse multivariée, l'hypertension artérielle pulmonaire (OR 6,2, IC 95 % 3,5-10,7), les antécédents d'insuffisance cardiaque (OR 9,8, IC 95 % 5,7—16,8) et d'arythmie auriculaire (OR 3,6 IC 95 % 2,2—5,9) étaient des facteurs de risque significatifs d'hospitalisation pour IC ($p < 0{,}001$). La durée moyenne de séjour des patients IC était plus longue (12,2 jours contre 6,9 jours ; $p < 0{,}01$) et 25 % des patients ont nécessité une prise en charge en soins intensifs. Vingt pourcent (11/55 patients IC) sont décédés, 18 % (10/55) ont été ré-hospitalisés pour IC, et 11 % (6/55) ont eu une transplantation cardiaque sur un suivi médian de 18 mois (IC 95 % 14—20 mois). Le risque d'événements cardiovasculaires était 19 fois plus élevé après l'hospitalisation liée à l'IC.
Conclusions. — L'IC devient l'une des principales causes de morbidité et de mortalité dans la population ACC. Un diagnostic précoce et une gestion plus active de l'IC dans cette population sont nécessaires pour améliorer les résultats.

Background

Owing to technical improvements in cardiac surgery and the medical management of congenital heart disease (CHD), more than 85% of the patients reach now adulthood [1]. Consequently, the prevalence of CHD in the adult population has increased rapidly and is now higher than in the paediatric population [2]. Heart failure (HF) is a serious and common complication in the long-term follow-up of adults with CHD, and is the main cause of death in these patients [3,4]. A few studies have focused on the occurrence of HF in adult CHD (ACHD) [5—7]. Although guidelines for management of HF in acquired heart diseases exist [8], there is no recommendation to prevent complications or to improve the management of HF in the CHD population. In patients with acquired heart disease, hospitalization for HF has a poor prognosis, with increased mortality and morbidity rates, despite current therapies [9]. Twonationwide studies have focused on HF in ACHD, but little is known about the characteristics and prognosis of these patients hospitalized for HF, and the risk

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factors for HF in this population have not been described in depth [7,10].

The aims of the present study were to characterize HF-related hospitalization in an ACHD population in a French cohort; and to determine the risk factors for and the prognosis of HF in patients with ACHD who are hospitalized for HF.

Methods

Study population

Patients with CHD aged 18 years or older who were hospitalized in our tertiary referral centre were prospectively included in the study, from November 2013 to October 2015. This study was approved by the institution's local ethics committee. Diagnosis, medical history, clinical events and procedure were collected from medical records. The main diagnosis was defined as the congenital heart lesion with the most severe haemodynamic anomaly, and the severity of the CHD diagnosis was assigned according to the Besthesda classification [11]. We compared patients admitted for acute HF (with or without a history of HF) with patients hospitalized for other reasons. Acute and chronic HF were defined according to European Society of Cardiology guidelines [12]. Cardiovascular risk factors (dyslipidemia, arterial hypertension, obesity [defined as body mass index $\geq$ 30.0)], diabetes mellitus and smoking) and common co-morbidities, including respiratory disease, renal dysfunction, arthritis, obesity, diabetes mellitus, cognitive dysfunction and depression, were also recorded. At admission, demographic data (age, sex, weight and height), clinical data (oxygen saturation, blood pressure, heart rate and rhythm) and biochemical data (natraemia, serum haemoglobin, creatinine and B-type natriuretic peptide [BNP] concentrations) were recorded. Treatment at admission was also reported. The systemic ventricle ejection fraction was obtained from echocardiographic and/or magnetic resonance imaging studies.

Outcomes and risk factors

Major outcomes occurring during the period following entry up to October 2016 were recorded systematically, and were defined as death, cardiac transplant or the need for a circulatory or ventricular assist device, readmission for HF, and a composite outcome, defined as the presence of at least one of those outcomes. Deaths outside the institution were based on physicians receiving a report from other healthcare providers. No patients were lost to follow-up during the 36-month study period.

We studied the predictive value of patient characteristics (sex, main defect, left-sided or right-sided cardiac lesions and history of cardiovascular complications and interventions), clinical and biological characteristics at admission and systemic ventricle ejection fraction, because these variables could potentially increase the risk of HF or were considered clinically important candidate risk factors for mortality and morbidity after HF admission; they were identified based on a review of the literature or clinical relevance.

Statistical analyses

Analyses were performed with the use of MedCalc® statistical software (MedCalc Software, Mariakerke, Belgium). Data are presented as the mean value $\pm$ standard deviation when variables are normally distributed and the median value with 95% confidence interval (CI) when they are not. Comparison between patients admitted for HF and patients admitted for other reasons was made using a two-sample *t* test or the Mann–Whitney test for quantitative data and a χ^2 test or Fisher's test for qualitative variables, when appropriate. The predictive value of patient characteristics for HF-related hospitalization was evaluated by univariate and multivariable logistic regression analysis. The multivariable logistic regression model included variables with the significance level $P < 0.2$ in the univariate analysis, after a backward selection of relevant variables, and excluding collinear variables from the model. For all analyses, a two-tailed *P* value < 0.05 was used as the criterion for statistical significance.

Results

Four hundred and seventy-one patients with ACHD were consecutively included, and 583 hospitalizations were recorded during the 24-month inclusion period months. HF was the main reason for hospitalization in 76 (13%) cases, concerning 55 (11.6%) patients with ACHD.

Characteristics of patients with HF (Table 1)

Patients admitted for HF were older and were mainly men compared with patients without HF ($P \leq 0.04$). Severe cardiac conditions, such as complex CHD, pulmonary arterial hypertension (PAH) and history of cardiovascular events (atrial arrhythmia, heart failure and pacing), were significantly associated with HF ($P \leq 0.04$). Complex CHDs were mainly represented by CHD with right ventricular outflow tract surgery, CHD associated with PAH, univentricular heart and systemic right ventricle (Fig. 1). In patients with recurrent HF (n = 25), median time between the acute HF preceding the study and the HF episode corresponding to the inclusion was 6.2 years (95% CI 1.9–13.7 years). Interestingly, patients with acute HF had significantly more co-morbidities than patients without HF ($P < 0.01$; Table 2); they were mainly represented by stroke (16.3% of patients admitted for HF).

Patient characteristics at admission

At admission, atrial arrhythmia was diagnosed in 18 (33%) patients with HF in ACHD. Among haemodynamic variables, heart rate and oxygen saturation were significantly different between patients with and without HF. Among the biological markers, concentrations of BNP, creatinine and bilirubin were significantly increased in the HF group compared with the non-HF group ($P < 0.01$) (Table 3). Ventricle function was classified as severely impaired in 36%, moderately impaired in 35% and was considered as normal or with diastolic dysfunction in 30%, using echocardiography or magnetic resonance imaging (Table 3). At admission, 38 (70%) patients with HF were already being treated with

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Table 1 Characteristics of patients: heart failure admission versus non-heart failure admission.

	HF group ($n = 55$)	No HF group ($n = 416$)	*P*
Age (years)	44 ± 14	37 ± 15	<0.0001
Men/women (*n*/*n*)	35/20	204/212	0.04
Syndrome	6 (10.9)	40 (9.6)	0.78
CHD complexity			0.04
Simple	6 (10.9)	54 (13.0)	
Moderate	21 (38.2)	221 (53.1)	
Severe	28 (50.9)	241 (33.9)	
Cardiac surgery			0.1
Palliation	13 (23.6)	123 (29.6)	
Repair	16 (29.1)	68 (16.3)	
No	26 (47.3)	225 (54.1)	
Number of cardiac surgeries			0.27
0	13 (23.6)	123 (29.6)	
1	21 (38.2)	120 (28.8)	
≥2	21 (38.2)	113 (27.2)	
CV risk factors	28 (50.9)	170 (40.8)	0.1
PAH	16 (29.1)	30 (7.2)	<0.0001
Cardiac history			
Atrial arrhythmia	34 (61.8)	118 (28.4)	<0.0001
Ventricular arrhythmia	5 (9.1)	20 (4.8)	0.2
Pacemaker	12 (21.8)	50 (12.0)	0.04
Heart failure	25 (45.5)	26 (6.3)	<0.0001
Infective endocarditis	5 (9.1)	25 (6.0)	0.4
Extracardiac medical history			
History of pregnancy	8/20 (40.0)	24/212 (11.3)	0.0004
Co-morbidities	34 (61.8)	154 (37.0)	0.0004

Data are expressed as mean ± standard deviation or number (%), unless indicated otherwise. CHD: congenital heart disease; CV: cardiovascular; HF: heart failure; PAH: pulmonary arterial hypertension.

Table 2 Co-morbidities in patients with adult congenital heart disease, with and without heart failure.

	HF group ($n = 55$)	No HF group ($n = 416$)	*P*
Respiratory disease	3 (5.5)	9 (2.2)	0.1
Sleep apnoea syndrome	0 (0)	3 (0.7)	–
History of stroke	9 (16.3)	14 (3.4)	<0.0001
Other neurological abnormalities	5 (9.1)	6 (1.4)	<0.01
Renal dysfunction	5 (9.1)	9 (2.2)	0.01
Hepatic dysfunction	2 (3.6)	8 (1.9)	0.32
Cognitive dysfunction and depression	2 (3.6)	24 (5.8)	0.7
Thyroid disturbance	8 (14.5)	43 (10.3)	0.3
Hypothyroidism	7	32	
Hyperthyroidism	1	11	
Arthritis	1 (1.8)	4 (1.0)	0.48
Obesity	3 (5.5)	15 (3.6)	0.45
Thromboembolic disease	2 (3.6)	7 (1.7)	0.3
Alcoholism	1 (1.8)	7 (1.7)	0.9

Data are expressed as number (%). HF: heart failure.

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Table 3 Clinical and biological characteristics of patients with and without heart failure.

	HF group ($n = 55$)	No HF group ($n = 416$)	P
Clinical characteristics			
Height (cm)	169.0 (165.0–171.0)	168.0 (166.0–170.0)	0.46
Weight (kg)	62.5 (57.0–72.5)	65.0 (64.0–68.0)	0.63
Heart rate (bpm)	80.5 (75.0–85.3)	75.0 (73.0–76.0)	0.01
Systolic BP (mmHg)	119.0 (110.0–123.6)	117.0 (115.0–119.9)	0.38
Diastolic BP (mmHg)	71.5 (68.0–77.0)	71.0 (70.0–72.0)	0.43
Oxygen saturation (%)	92.5 (90.0–94.0)	97.0 (96.0–97.0)	<0.01
Biological characteristics			
Creatinine (mmol/L)	87.50 (82.0–105.6)	74.0 (72.0–76.0)	<0.01
Haemoglobin (g/dL)	14.4 (13.5–15.3)	14.1 (13.9–14.3)	0.06
Natraemia (mmol/L)	138.0 (136.0–139.0)	139.0 (139.0–139.0)	0.06
Bilirubin (mmol/L)	30.5 (25.1–43.6)	16.0 (15.0–17.9)	<0.01
BNP (pg/mL)	549.0 (343.6–715.9)	70.0 (62.0–83.2)	<0.01
Systemic ejection fraction			
EF $\geq$ 50%	16 (29.1)	345 (82.9)	<0.01
35% $\leq$ EF < 50%	19 (34.5)	50 (12.0)	<0.01
EF < 35%	20 (36.4)	21 (5.0)	<0.01

Data are expressed as median (95% confidence interval) or number (%). BP: blood pressure; BNP: B-type natriuretic peptide; EF: ejection fraction; HF: heart failure.

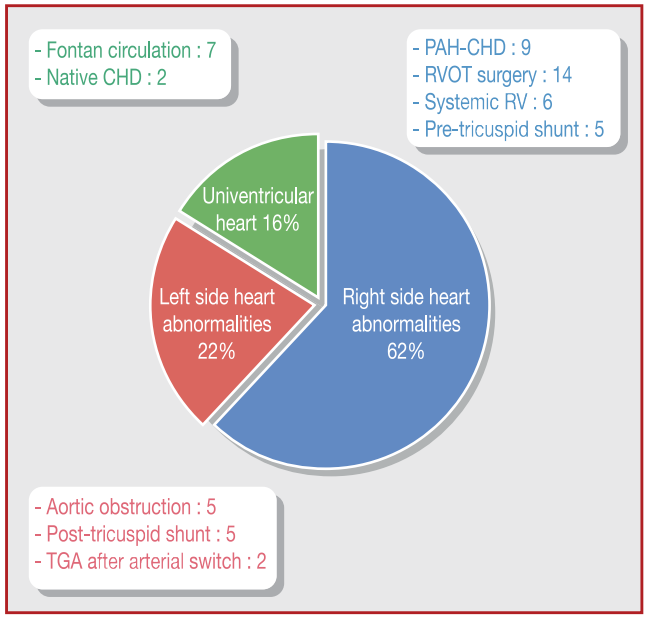

Figure 1. Distribution of congenital heart defects in patients admitted for heart failure. Congenital heart defects were clustered in three categories, according to the failing ventricle. Patients with right ventricular outflow tract (RVOT) surgery included patients with tetralogy of Fallot, double outlet right ventricle (RV), truncus arteriosus, pulmonary atresia and pulmonary stenosis. There was no patient with the Ross procedure. Patients with D-transposition of the great arteries (TGA) after atrial switch and congenitally corrected TGA were in the group with systemic RV. Aortic obstruction included patients with aortic subvalvular and supravalvular stenosis, valvular stenosis and aortic coarctation or interruption. Coronary complication with myocardial ischaemia was the aetiology of left ventricle failure in the two patients with D-TGA after arterial switch. CHD: congenital heart disease. PAH: pulmonary arterial hypertension.

diuretics, 30 (54.5%) were receiving beta-blockers, 14 (25.5%) were receiving angiotensin-converting enzyme inhibitors or angiotensin II receptor blockers and 25 (44.5%) were receiving anticoagulation therapy.

Medical and surgical management of HF in ACHD

The mean hospital stay of patients with HF in ACHD was significantly longer (12.2 days vs. 6.9 days; $P < 0.01$). Management in the intensive care unit was required for 19/77 (25%) for 1–45 days (median 1.5 days). During the 36-month study period, four patients needed inotropic treatment, none were supported by circulatory or ventricular assist devices and eight patients were listed for heart transplantation (six were transplanted; two were still awaiting a heart transplant). A surgical or percutaneous intervention to correct haemodynamic lesions was performed in five patients. Moreover, two patients underwent atrial arrhythmia ablation, one underwent ventricular tachycardia ablation and two underwent cardiac resynchronization therapy.

Risk factors and prognostic value for HF-related admissions

Risk factors for HF-related admission are shown in Table 4. History of HF and PAH associated with CHD had the strongest predictive value for acute HF in ACHD. Patients with a history of HF had an almost 10-fold higher risk of HF recurrence, and patients with PAH had a 6-fold higher risk of acute HF ($P < 0.001$). In the multivariable logistic regression analysis, including variables with a significance level $P < 0.2$ in the univariate analysis, and after backward selection and exclusion of variables with collinearity (complex CHD, stroke and co-morbidities), PAH, history of HF and history of atrial

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Table 4 Univariable analysis of heart failure-related hospitalization risk factors.

Variables	Odds ratio	95% CI	*P*
Age (years)	1.0	1.0—1.0	<0.001
Age ranges			
15 years ≤ age < 25 years	0.5	0.1—1.6	0.22
25 years ≤ age < 35 years	0.6	0.3—1.14	0.10
35 years ≤ age < 45 years	2.3	1.0—5.0	0.04
45 years ≤ age < 55 years	2.5	1.0—6.2	0.05
55 years ≤ age < 65 years	2.7	0.9—7.9	0.07
65 years ≥ age	2.5	0.9—7.3	0.09
Male sex	1.8	1.2—3.1	0.02
Complex CHD (Bethesda classification)	3.1	1.5—6.1	0.001
PAH	6.2	3.5—10.7	<0.001
History of HF	9.8	5.7—16.8	<0.001
Pacemaker	1.6	0.9—3.1	0.1
History of atrial arrhythmia	3.6	2.2—5.9	<0.001
History of stroke	3.0	1.6—5.7	<0.001
CV risk factors	1.5	0.9—2.4	0.12
Co-morbidity	2.4	1.5—3.9	<0.001
History of pregnancy	0.7	0.3—1.4	0.3

CHD: congenital heart disease; CI: confidence interval; CV: cardiovascular; HF: heart failure; PAH: pulmonary arterial hypertension.

arrhythmia remained significant risk factors for HF-related admission ($P < 0.001$). These risk factors explained 80% of HF-related admissions in patients with HF in ACHD, whereas at least one of these factors concerned only 35% of patients without HF ($P < 0.01$).

Prognostic value of HF-related hospitalization

During the median follow-up of 18 months (95% CI 14—20 months), 11 (20%) patients died, 10 (18%) were readmitted for HF and six (11%) were transplanted. The risk of these combined cardiovascular events (recurrence of HF, death or transplantation) was 19-fold higher (95% CI 3—110) after HF-related hospitalization (Fig. 2). The probabilities for freedom from cardiovascular events at 3, 6, 12 and 24 months were 90.8%, 88.5%, 83.8% and 76.6%, respectively, in patients hospitalized for HF vs. 96.9% in patients who did not experience HF ($P < 0.01$; Fig. 2). In the HF-ACHD group, among the patient characteristics and clinical or biological variables at admission and discharge, only BNP concentration at discharge was significantly associated with readmission for HF: mean BNP at discharge was 351 ± 205 pg/mL in patients with HF in ACHD who were not readmitted, and 776 ± 288 pg/mL in patients who were readmitted for HF ($P < 0.01$).

In the non-HF group, three (0.72%) patients died, 51 (12.2%) were readmitted for reasons other than HF (40/51 [78%] were planned admissions) and one patient was transplanted for refractory infective endocarditis despite two surgical treatments.

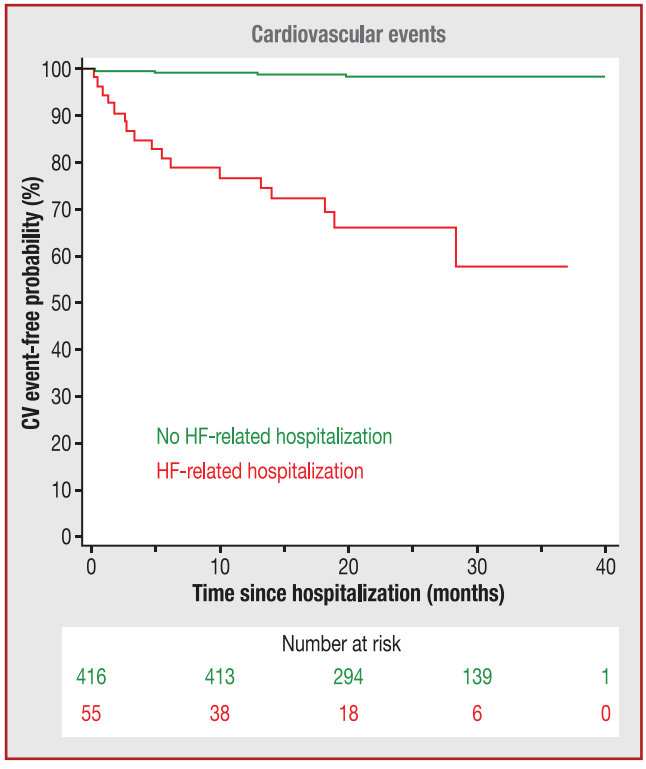

Figure 2. Risk of cardiovascular (CV) events in patients with ACHD after heart failure (HF)-related hospitalization and after no HF-related hospitalization ($P < 0.01$).

Discussion

We have evaluated the burden of HF-related hospitalization in ACHD in a single tertiary centre. Several important findings were observed. First, HF is a severe event in ACHD, associated with a high mortality rate (20%) and a 19-fold higher risk of cardiovascular events. Secondly, HF-related hospitalizations are a major cost factor in ACHD care, accounting for 13% of ACHD hospitalizations, with a

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longer stay and intensive care requirement for one in four patients. Finally, PAH, atrial arrhythmia and history of HF are important risk factors for a new HF event. These factors allow the identification of patients at major risk of HF, who require a rigorous follow-up and active management.

HF is the main cause of death in the ACHD population [13], and is associated with a high morbidity rate. The incidence for first HF admission was 1.2 per 1000 patient-years, which is substantially higher than in the general population [10]. As in other studies [14,15], we found that HF was a common cause of hospitalization of patients with ACHD. The prevalence of hospitalization for HF in the USA has been increasing since 1998 [15]. This prevalence should increase with the ageing ACHD population, which explains why patients with HF in ACHD were older in our series compared with patients without HF.

Zomer et al. found mortality was fivefold higher in patients with HF in ACHD compared with patients without HF in ACHD [7]. We also observed a high mortality rate (20%) in patients with HF in ACHD in our study. Moreover, HF-related hospitalization was associated with a strong increase in the risk of cardiovascular events (HF recurrence, death or heart transplantation). In acquired heart disease, mortality increased significantly after each HF hospitalization, and after adjusting for age, sex and major co-morbidities, the number of HF hospitalizations was a strong predictor of all-cause death [16]. From our findings, we suppose that this evolution is similar in the ACHD population. Zomer et al. reported a 1-year mortality rate of 24% after first HF admission [7]; they identified main defect, multiple defects and surgical interventions in childhood as independent risk factors of HF admission. Other studies have demonstrated that the independent factors that predict mortality are numerous and varied [7,10].

Prognostic risk models would be helpful to identify patients with HF who might benefit from closer follow-up and earlier medical or surgical interventions. In a retrospective study, Stefanescu et al. highlighted the usefulness of the Seattle Heart Failure Model to identify patients with ACHD at high risk of poor outcome [17]. However, even in patients with acquired heart disease, prediction of mortality and HF hospitalization, using different models, remains difficult, and is only moderately successful [18]. We identified three major risk factors for HF in ACHD, which were history of HF, PAH and atrial arrhythmia. We suggest that patients with ACHD who have one of these cardiac conditions should be closely monitored with active management. According to our results, patients with complex CHD, a pacemaker or co-morbidities (i.e. stroke or renal dysfunction) are also at risk of HF and require more attention.

HF mortality in ACHD raises questions about the timing of the diagnosis of HF in this population. Patients with ACHD might be diagnosed with HF when the disease is already at an advanced stage, as they may not readily report symptoms [19]. Furthermore, aetiology, pathophysiology and triggers of impaired ventricular function in ACHD are complex and diverse. The main therapeutic strategy is medical, as recommended by guidelines, even if there are no robust clinical trials to guide clinicians on specifically managing patients with HF in ACHD [8,20]. We observed that patients with HF were insufficiently treated; only 25.5% received angiotensin-converting enzyme inhibitors or angiotensin II receptor blockers, and 54.5% received beta-blockers. Active research into HF in ACHD is required to determine the roles of patient education, new efficient medical therapy [21] and assistance devices [22].

We observed a high proportion of patients with HF with moderately to severely complex CHD in our study (87.3%), while 44% of patients had atrial septal defect in the nationwide study by Rodriguez et al. [10]. Patients with specific lesions, such as tetralogy of Fallot or Fontan circulation, are at greater risk of HF than patients with simple left-to-right shunts [5,10]. The complexity of CHD may explain why the mean duration of hospitalization for patients with HF in ACHD was longer in our study than in this published survey (7.6 days) [10]. Of note, the median hospital stay was similar in a large French observational survey (13 days), which consisted of a single-day snapshot of unplanned HF hospitalizations [16]. These findings highlight the need for medical resources with both ACHD and HF skills for the management of these patients.

Readmissions are a popular target for quality improvement, because they are costly and are thought to be largely preventable. We found an 18% rate of patient readmissions at a median follow-up of 18 months. Only BNP concentration at discharge seemed to be predictive of early HF recurrence, as has been shown for HF in acquired heart diseases [23]. The readmission rate in the HF in ACHD population seems lower than that in HF among patients with normal cardiac anatomy, where the readmission rate is ranged between 30% and 34% at 3 months, and was 56.1% at 1 year [9,24]. However, approximately half of the rehospitalizations were not related to HF [25], and the proportion of non-cardiovascular admissions was higher in those with preserved ejection fraction [26]. In ACHD, a recent study found that patients admitted with HF once were more likely to be readmitted within 12 months than patients admitted with other diagnoses [27]. The authors identified that risk factors for readmission were dependent on the type of heart defect, suggesting that individualized lesion-specific strategies are required to improve care.

Study limitations

Because this was a single-centre study, the sample of patients included may not be representative of the pattern of HF in the overall ACHD population. Nevertheless, biases arising from the use of administrative databases are avoided, such as misclassification and retrospective design associated with studies using this kind of database. A multicentre prospective registry would be useful to improve the diagnosis and management of HF in ACHD — especially HF diagnosis in ACHD, which may be tricky [20]. In this present study, we assessed acute HF defined according to the European guidelines [8]; we did not assess chronic HF in ACHD, and further studies would be necessary to characterize it.

Precipitating factors, such as non-adherence to treatment, were not assessed, and their identification could improve risk stratification and prevention strategies. However, we observed that 33% of patients hospitalized for HF had atrial arrhythmia at admission. Moreover, in acquired HF, precipitant factors were not predictors of mortality [28]. No relevant risk factor for readmission was found, because of the small size of the HF population and the short duration of

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follow-up. Longer and larger studies are needed to improve diagnosis, risk stratification and the management of patients with HF in ACHD.

Conclusions

HF is emerging as a leading cause of morbidity and mortality in the expanding ACHD population. History of HF, PAH and atrial arrhythmia are the main risk factors for HF-related admissions in patients with ACHD, and can be used to identify patients at risk. More active follow-up and management are required to improve outcomes in this vulnerable population. More evidence-based medicine, from larger and longer registries or clinical trials, is needed to determine the best clinical practice in the management of HF in patients with ACHD.

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Disclosure of interest

The authors declare that they have no competing interest.

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