Emergency care of adult patients with congenital heart disease: are we prepared? Data from a tertiary center and long-term follow-up

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Abstract

A growing number of Congenital Heart Disease (CHD) patients will achieve adulthood. We aim to assess the characteristics, clinical course, and outcomes of Adult CHD (ACHD) patients attending the Emergency Department (ED) of a referral center. Seventy-five patients, 35 females (47%) were included. The mean age was 40 (±15) years. 35 patients (47%) had complex defects, and 29 patients (39%) were in NYHA class III-IV. Main symptoms were: dyspnea 22 (29 %), palpitations 20 (27%), fever 8 (11%), syncope 6 (8%), neurologic symptoms 7 (9.3%), chest pain 5 (7%). Heart failure and arrhythmias accounted for 23 (31%) and 20 (27%) of ED diagnoses. Twenty-eight patients (37%) were discharged, one patient died during observation, 32 patients (69 %) were admitted to a cardiology ward, 6 (13%) to the intensive care unit, and 8 (17%) to a non-cardiological ward. At a follow-up of 701 (554-984) days, 10 patients (13%) died, 2 (2.6%) were transplanted, 3 (4%) were listed for a heart transplant, and 30 (41 %) were re-admitted. In multivariate analysis, complex anatomy was weakly associated with readmissions. These preliminary data indicate that attendance of ACHD patients is epidemiologically relevant.

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Introduction

The growing population of Adults with Congenital Heart Disease (ACHD) is considered a major epidemiological priority in the coming years. It has been hypothesized that, due to the marked improvement in acute and post-operative care, an increasing number of CHD patients will reach adulthood and even later in life.[1,2] Therefore, we can foresee that the Emergency Department (ED) will consistently face an increase in ACHD patients with a wide range of clinical needs.[3-12] Moreover, the lack of standardized evidence-based diagnostic and therapeutic protocols might limit the effectiveness of acute treatment and eventually jeopardize the prognosis of this subset of patients.[13,14]

Congenital heart defects in adult patients were classified by the ACHD Anatomic and Physiological (ACHD AP) system. Anatomic classification includes Class I (simple), Class II (moderate complexity), and Class III (great complexity). The physiological classification (which takes into account the functional status as well as other factors, *e.g.* the presence of valvular disease, pulmonary hypertension, arrhythmias, aortic dilatation, end-organ dysfunction, and cyanosis) is divided into stages A-D (where A is the lowest grade of severity).[15] This classification is a useful tool also to stratify the risk of patients, particularly in the emergency setting where specialistic ACHD facilities might not be available. Most of the available epidemiological evidence about this clinical setting is based on large administrative datasets carrying intrinsic limitations regarding the completeness and granularity of data and lack of follow-up. We aim to collect data on the epidemiology of ACHD attending the ED in our tertiary center, focusing on clinical presentation, framework of care provided, and outcomes (death or major event occurring after first contact with ED, discharge, admission, referral to other centers).

Materials and Methods

This was a single-center, retrospective study involving Papa Giovanni XXIII Hospital in Bergamo, Italy, a tertiary hospital with over 90,000 ED visits/year and with a CHD cardiological service.

Patients who attended the ED between January 1, 2019, and August 31, 2022, with a diagnosis of congenital heart defect were audited. Demographic, clinical variables, symptoms at presentation, CHD anatomy, and physiological stage were collected. Social history was also recorded whenever available. The hospital process-related variables, including the triage code assigned (green code = minor urgency, yellow code = urgency, and red code = emergency) and diagnostic tests performed in the ED were retrieved. Both in-hospital and long-term outcomes were considered. The latter was a composite of death, transplant listing, and ED readmissions.

Statistical analysis

Continuous variables were expressed as mean ±, standard deviation, median, and interquartile ranges and compared using Wilkinson rank sum or t-test. The normality of continuous variables was assessed by visually examining the distribution histograms. Categorical variables were presented as counts and percentages and compared using appropriate x^2 or Fisher exact tests.

The p-value threshold for statistical significance was set at 0.05. The STATA 11.0 software by Stata Corp was used for the analysis.

Results

Demographics and previous clinical history

Between January 1, 2019, and August 31, 2022, seventy-five patients, 35 females (47%) presented to the ED. The mean age was 40±15 years (range 18-57). Twenty-three (30%) and 3 (4%) patients belonged to Physiology Class C and D, respectively, and 35 patients (47 %) had excellent complexity anatomy. Twenty-nine patients (39%) had an advanced NYHA class (III-IV). The majority of the patients had one or more associated comorbidities, such as diabetes (5%), renal failure (7%), pulmonary hypertension (15%), and genetic syndromes (11%).

Past medical and surgical history included cardiac surgery and cardiac catheterization in the previous year in 66 (88%) and 16 (21%) patients, respectively.

Sixty-three (84%) patients were taking medication at the time of ED admission: 36 (48%) beta-blockers, 30 (40%) diuretics, 25 (33%) anti-arrhythmic drugs, 31 (41%) anticoagulants, 22 (29%) Acetylsalicylic Acid (ASA), 13 (17%) Ace-inhibitors, and 10 (13%) pulmonary vasodilators. Of 66 patients, 39 (59%) were employed, and 36 (54%) were married or had a stable relationship with cohabitation. Seventeen (23%) were lost to follow-up before the admission to the ED (Table 1).

Clinical presentation and in-hospital course

At presentation, dyspnea (22%), palpitations (20%), fever (8%), neurologic symptoms (7%), syncope (6%), chest pain (5%), and bleeding (3%) were the symptoms. Other reasons for admission were abdominal pain, electrolyte disorders, and delivery (Table 2).

Green, yellow, and red triage codes were assigned to 19 (25%), 49 (65%), and 7 (9%) patients, respectively (Figure 1).

Table 1. General demographics and clinical and diagnostic characteristics of the population at presentation at the Emergency Department.

Demographic	N=75
Female, N (%)	35 (47)
Age, years (mean)	40 (±15)
NYHA class III-IV, N (%)	29 (39)
Physiology class	N=75
Class A, N (%)	11 (15)
Class B, N (%)	38 (51)
Class C, N (%)	23 (30)
Class D, N (%)	3 (4)
Complexity of CHD	N=75
Great, N (%)	22 (29)
Moderate, N (%)	18 (24)
Simple, N (%)	35 (47)
Comorbidities	N=75
Diabetes, N (%)	4 (5)
Renal failure, N (%)	5 (7)
Obesity, N (%)	5 (7)
Pulmonary hypertension, N (%)	11 (15)
Genetic syndromes, N (%)	8 (11)
Medical history	N=75
Previous heart surgery, N (%)	66 (88)
Catheterization in the last year, N (%)	16 (21)
Previous arrhythmias, N (%)	49 (75)
PM/ICD, N (%)	23 (31)
Previous endocarditis, N (%)	2 (3)
Social history	N=66
Employed, N (%)	39 (59)
Student, N (%)	5 (8)
Single, N (%)	8 (12)
Married, N (%)	36 (54)
Living with parents, N (%)	22 (33)

PM, Pace-Maker; ICD, Implantable Cardioverter-Defibrillator; CHD, Congenital Heart Disease.

Table 2. Symptoms at presentation and provisional diagnosis in the Emergency Department (ED).

Symptoms	N (%)
Dyspnea, N (%)	22 (29)
Palpitations, N (%)	20 (27)
Fever, N (%)	8 (11)
Neurologic symptoms, N (%)	7 (9)
Syncope, N (%)	6 (8)
Chest pain, N (%)	5 (7)
Bleeding, N (%)	3 (4)
Other, N (%)	4 (5)
Diagnosis	N (%)
Heart failure, N (%)	23 (31)
Arrhythmias, N (%)	20 (27)
Infections (not endocarditis), N (%)	9 (12)
Neurologic disorders, N (%)	8 (11)
Bleeding, N (%)	5 (7)
Aspecific chest pain, N (%)	3 (4)
Endocarditis, N (%)	2 (3)
Hembolism, N (%)	2 (3)
Other, N (%)	3 (4)

During the evaluation in the ED, 57 (76%) patients and 22 (29%) underwent trans-thoracic echocardiogram and chest Computer Tomography (CT), respectively.

The most frequent provisional diagnoses in the ED were heart failure (31%) and arrhythmias (27%). After ED assessment, 9 (12%) patients were diagnosed with infection; among them, 2 (3%) infective endocarditis was the final diagnosis. Other diagnoses are reported in detail in Table 2.

Electrocardiogram and arrhythmias

At presentation in ED, 73 (97%) patients had undergone an Electrocardiogram (ECG). The rhythm at presentation was sinus in 40 (55%) patients, atrial fibrillation or flutter in 29 (40%), ventricular tachycardia in 3 (4%), and in 1 (1%) severe bradycardia. Overall, 14 (19%) patients out of 75 required cardioversion, 6 (43%) of whom Direct Current (DC) shock.

Outcomes

Twenty-eight patients (37%) were discharged, while 46 (61%) were hospitalized, of whom 32 (69 %) were admitted to a cardiology ward and 6 (13%) to the Intensive Care Unit (ICU). Additionally, eight subjects (17%) were admitted to a non-cardiological ward, and one patient died during the observation in the ED (Figure 1). The median hospitalization duration was 10 (5-16) days. Ten patients (13%) died, one underwent transplantation, three (4%) were listed for a heart transplant, and 30 (40%) were re-admitted to the ED during a median follow-up of 701 (554-984) days. At univariate analysis, patients with more advanced NYHA class, those regularly followed at the CHD center, with a defect of great complexity, treated with diuretics, and with pulmonary hypertension were more likely to have repeated ED accesses. At multivariate analysis, great complexity anatomy showed a weak association with readmission (Table 3).

All patients who eventually died at follow-up had multiple admissions to the ED.

Discussion

Patients with CHD who have now become adults are a growing population as survival rates through successful heart surgery and post-operative care have improved significantly over the last 20 to 30 years. Perioperative mortality was comparatively low (7.1%), and a large proportion of CHD patients reached adulthood.[2] Another critical role for this achievement was the centralization of care at tertiary ACHD centers.[16] As a consequence, ED admissions of this particular population may become ever greater; in fact, as demonstrated by Agarwal *et al.,* there has been a considerable increase in ED visits among patients with ACHD during 7 years from 2006 to 2012.[1] We observed that an ECG was recorded at pre-

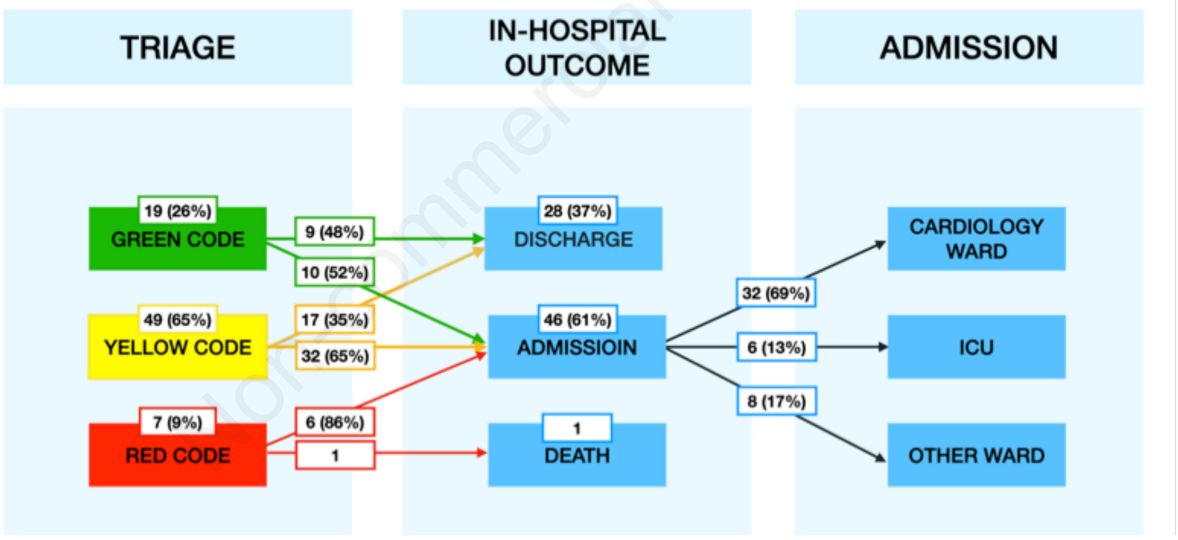

Figure 1. Figure showing access codes in the Emergency Department (ED), in-hospital outcome, and relevant departments to which patients have been admitted, if any. ICU, Intensive Care Unit.

Table 3. Univariate and multivariate analysis.

Variables	Univariate Odds ratio	Univariate 95% CI	Univariate p value	Multivariate Odds ratio	Multivariate 95% CI	Multivariate p value
NYHA class	2.8	1.3-6	0.009	1.8	0.7-4.8	0.2
Pulmonary hypertension	4	1.2-20	0.03	2.5	0.4-12	0.2
Diuretics	3.1	1.7-8.4	0.02	1.8	0.5-5.9	0.9
Great complexity	2	1.1-3.6	0.02	1.8	0.96-3.4	0.06
Regularly followed	0.06	0.01-0.5	0.01			

CI, Confidence Interval.

sentation in all but two of the patients included in our cohort. One of these two patients underwent a pacemaker check as a first-line investigation. This observation is consistent with most patients complaining of chest symptoms.[17] Furthermore, it is recognized that arrhythmic complications are highly prevalent in the population of CHD patients.[18] As a consequence of their arrhythmic history, a significant number of patients had a pacemaker or ICD implanted. Moreover, the therapeutic pharmacological options are usually limited to class III anti-arrhythmic agents in this population. All these factors create additional complexity to the evaluation in the ED. According to our data, overall, the management of patients with CHD in the ED contemplates specialistic images such as transthoracic echocardiogram or chest CT in a large proportion of patients (57 (76%) patients and 22 (29%), respectively).[3] Most patients were taking medications at the time of ED admission: about two-thirds were on anti-platelets or anticoagulation, and 13% were on pulmonary vasodilators.[19] These data have to be taken into consideration since such medications can have specific toxicity profiles. They may significantly affect the risk of patients with trauma or conditions requiring urgent surgery or invasive procedures.[20]

The mortality rate in our cohort (13%) is higher as compared to literature data, reflecting a higher prevalence of acute conditions differently from the standard outpatient population. Diller *et al.* reported a mortality rate of 7.7%, 524 out of 6969 adult patients treated at the Royal Brompton Hospital over 9 years of follow-up.[2] It is also worth noting that 3% of patients presented with endocarditis. Although this is an unexpectedly high prevalence, comparison with other studies is more complex since incidence expressed as cases per 1000 person-years is usually reported.[21,22] However, we can speculate that this finding might reflect both a referral bias and the specific characteristics of our cohort, which includes a significant proportion of patients with complex anatomies.

The univariate analysis indicated that patients in the most advanced NYHA class, with very complex anatomy, diuretic use, and pulmonary hypertension, were more likely to have repeated ED visits. At multivariate analysis, only great complexity anatomy showed a weak association with readmission (Table 3).

Although it has been demonstrated that patients who are lost to follow-up have a higher risk of events at follow-up, we could not confirm this observation with our data.[23] In particular, adult congenital patients regularly followed experienced more ED admissions. As mentioned before, this paradox can derive from the peculiar pattern of patient referral to our ED, which is located within a tertiary congenital heart center.

Not surprisingly, patients who eventually died at follow-up had multiple admissions to the ED.[24]

Finally, a major challenge for the aging population with complex CHD is managing end-of-life conditions. We do not have information about patients' end-of-life dispositions in the present dataset. Without a structured palliation pathway for these patients, a respectful, balanced, and shared decision between the Emergency Medicine (EM) physician and cardiologist is advisable.

Limitations

This study has several limitations. First, it is a retrospective, single-center study with a limited sample size. The small numbers did not grant enough power to perform extensive multivariate analysis. Furthermore, as our ED department is within a hospital with a long tradition of congenital surgery, a referral bias effect cannot be excluded. Finally, we cannot present comparisons of the ACHD epidemiological burden in the ED among different eras.

Conclusions

A considerable proportion of patients with CHD were admitted to the ED multiple times. Subsequently, during long-term monitoring, patients who had visited the ED displayed a noteworthy mortality rate. The principal provisional diagnoses were heart failure and arrhythmias, which translated into specific needs in terms of clinical skills. Irrespective of the presentation symptoms, most patients underwent specialistic workup, including CT and echocardiography. Nevertheless, EM physicians must play an active clinical role in managing these patients. These preliminary data indicate that attendance of ACHD patients is epidemiologically relevant and establishing shared protocols might optimize management of this peculiar population.

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