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Risk Modeling for Hospitalization and Mortality in Pediatric Cyanotic Congenital Heart Disease: An 11-year National Emergency Cohort

Siyanotik Konjenital Kalp Hastalığı Olan Çocuklarda Hastane Yatışı ve Mortalite Risk Modellemesi: Ulusal Bir Referans Merkezinden 11 Yıllık Pediyatrik Acil Servis Verisi

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Abstract

Introduction: Although the outcomes and the mortality rates in congenital heart disease have improved dramatically, children with complex cardiac anomalies remain at increased risk for both cardiac and non-cardiac morbidity and mortality. This study aimed to characterize presentations to the pediatric emergency department (PED) by patients with cyanotic congenital heart disease (CCHD) and to identify factors associated with hospitalization and mortality.

Methods: A retrospective review of children with CCHD who were admitted to PED between January 2011 and September 2022 was conducted. Demographic, clinical, and outcome data were collected. Logistic regression was performed to identify predictors of hospitalization and mortality. The Glasgow Coma scale (GCS), Emergency Severity Index, and need for emergency life-saving interventions were also evaluated.

Results: Out of 351 presentations, 56.7% were male. The most common diagnoses were upper respiratory tract infection (49.0%), acute gastroenteritis (13.1%), and lower respiratory tract infection (LRTI) (11.7%). The most common cardiac diagnoses at the time of admission were cyanotic spell (1.7%) and heart failure (1.4%). Hospitalization occurred in 12.5% of admissions, and 6.8% required pediatric intensive care unit (PICU) admission. LRTI was the leading cause of both hospitalization (40.9%) and PICU admission (41.7%). Significant predictors for hospitalization were younger

Öz

Giriş: Konjenital kalp hastalığı olan çocuklarda sağkalım oranları artmış ve mortalite belirgin azalmış olsa da, kompleks kardiyak anomalilere sahip çocuklar hem kardiyak hem de diğer morbidite ve mortalite nedenleri açısından yüksek risk taşımaya devam etmektedir. Bu çalışmanın amacı, siyanotik konjenital kalp hastalığı (SKKH) tanılı hastaların çocuk acil servise (ÇAS) başvuru özelliklerini tanımlamak, hastaneye yatış ve mortalite ile ilişkili faktörleri belirlemektir.

Yöntemler: Ocak 2011 ile Eylül 2022 tarihleri arasında ÇAS'ye başvuran SKKH tanılı çocukların verileri geriye dönük olarak incelendi. Demografik, klinik ve sonuçlara ilişkin veriler toplandı. Hastaneye yatış ve mortaliteyi öngören faktörleri belirlemek amacıyla lojistik regresyon analizi uygulandı. Glasgow Koma skalası (GKS), Aciliyet Önem İndeksi ve acil yaşam kurtarıcı müdahale gereksinimi değerlendirildi.

Bulgular: Toplam 351 başvurunun %56,7'si erkekti. En sık tanılar üst solunum yolu enfeksiyonu (%49,0), akut gastroenterit (%13,1) ve alt solunum yolu enfeksiyonu (ASYE) (%11,7) idi. Başvuru anındaki en yaygın kardiyak tanılar siyanotik spell (%1,7) ve kalp yetersizliği (%1,4) olarak saptandı. Başvuruların %12,5'i hastaneye yatışla, %6,8'i ise çocuk yoğun bakım ünitesine (ÇYBÜ) yatışla

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Abstract

age, urinary tract infection, LRTI, emergency life-saving intervention, and ventilatory support. Mortality was statistically associated with increased cyanosis, dyspnea/respiratory distress, LRTI, heart failure, GCS ≤12, emergency life-saving intervention, significant intravenous fluid resuscitation, ventilation support, and circulatory support death. Mortality was statistically associated with increased cyanosis, dyspnea/respiratory distress, LRTI, heart failure, GCS ≤12, emergency life-saving intervention, significant intravenous fluid resuscitation, ventilation support, and circulatory support.

Conclusion: Emergency physicians should be vigilant for red flags indicating increased risk of hospitalization or mortality in CCHD patients to ensure timely and effective care.

Keywords: Cyanotic congenital heart disease, pediatric emergency department, mortality, hospitalization

Öz

sonuçlandı. ASYE hem hastane (%40,9) hem de ÇYBÜ (%41,7) yatışlarının en sık nedeniydi. Yatış için anlamlı öngörücüler küçük yaş, idrar yolu enfeksiyonu, ASYE, acil yaşam kurtarıcı müdahale ve solunum desteği idi. Artmış siyanoz, dispne/solunum sıkıntısı, ASYE, kalp yetersizliği, GKS ≤12, acil yaşam kurtarıcı müdahale, intravenöz sıvı resüsitasyonu, solunum ve dolaşım desteği ile mortalite arasında istatistiksel olarak anlamlı ilişki vardı.

Sonuç: Acil servis hekimleri, SKKH tanılı hastalarda hastaneye yatış veya mortalite riskini artıran uyarı işaretlerine karşı dikkatli olmalı ve zamanında, etkili müdahale sağlamalıdır.

Anahtar Kelimeler: Siyanotik konjenital kalp hastalığı, çocuk acil servis, mortalite, hastaneye yatış

Introduction

Congenital heart disease (CHD) is the most common major birth defect, with an incidence ranging from 4.6 to 12.2 per 1000 live births.[1,2] CHDs are typically classified as cyanotic and acyanotic.[3,4] Cyanotic CHD (CCHD) encompasses a broad spectrum of more complex pathologies than acyanotic heart diseases and account for approximately 25% of all CHDs, with a mean incidence of 1.4 per 1000 live births.[5]

Although the incidence of CHD has remained relatively stable over time, major advances in prenatal diagnosis, pharmacological management, interventional procedures, and surgical techniques have significantly improved survival into adulthood. Consequently, presentations of CHD patients to the pediatric emergency department (PED) have increased. While some PED visits are directly related to cardiac causes, many are due to common pediatric conditions. The overlap of cardiac and non-cardiac symptoms can pose diagnostic and management challenges.[6,7]

To the best of our knowledge, most of the previous studies of CHD have focused on inpatient populations, with limited data available on the characteristics and outcomes of PED presentations, particularly for CCHD patients.[6-8] This study aims to evaluate the characteristics of PED admissions among CCHD patients and to identify the predictors of hospitalization and mortality.

Materials and Methods

This retrospective study included all patients under 18 years of age who were followed up for CCHD in the pediatric cardiology department, and who presented to the PED of our institution between January 2011 and September 2022. The PED is part of a tertiary referral center with 284 pediatric beds and a comprehensive pediatric heart center, which is one of the leading institutions in our country. It provides care for a wide range of diagnoses and performs advanced interventions including the use of mechanical support devices and heart transplantation.

Patients under 18 years of age who had a prior diagnosis of CCHD or who were newly diagnosed with CCHD at the time of admission were included in the study. The patients with acyanotic CHD or missing data were excluded from the study. Demographic data, presenting complaints, and final diagnoses were evaluated. The Glasgow Coma Scale (GCS) was used to assess the status of the patients at admission to the emergency service, categorized as mild[9-12], intermediate[13-15], or severe (≤8). The Emergency Severity Index triage system was used to define emergency life-saving interventions.[9] Life-saving interventions refer to urgent medical procedures and treatments such as airway and breathing support (e.g., positive pressure ventilation, intubation, surgical airway), electrical therapy (defibrillation, cardioversion), emergency procedures (e.g., needle decompression, pericardiocentesis), hemodynamic stabilization [e.g., significant intravenous (IV) fluid resuscitation, blood transfusion], and administration of critical medications (e.g., adenosine, epinephrine, atropine).[9] The durations of PED observation, hospitalization, and pediatric intensive care unit (PICU) stay were evaluated. Additionally, clinical features potentially influencing hospitalization and mortality were evaluated.

The study protocol was approved by the Ethics Committee of Ankara University Faculty of Medicine (approval date: April 5, 2023; approval number: İ03-164-23).

Statistical Analysis

Descriptive statistics were presented as mean ± standard deviation for normally distributed variables, median (Q1- Q3) for non-normally distributed variables, and n (%) for categorical variables. Categorical variables were analyzed using Pearson's chi-square test or Fisher's exact test, as appropriate.

Logistic regression (univariable and multivariable) was performed to identify predictors of hospitalization and mortality. First, univariable logistic regression models were used to examine the independent effects of each variable. Variables found to be significant in univariable analyses were included in a multivariable logistic regression model to assess their adjusted effects.

The performance of the multivariable logistic regression model was evaluated using receiver operating characteristic curve analysis, with model discrimination assessed by calculating the area under the curve (AUC). Additionally, sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) were computed. The results of the logistic regression analyses were reported as odds ratios with 95% confidence intervals (CI). Statistical significance was defined as $p<0.05$. All statistical analyses were performed using IBM SPSS Statistics version 30.

Results

During the study period, 67 patients with CCHD made a total of 351 visits out of 1,040,393 admissions to the PED. The median number of PED visits per patient was 2.00 [interquartile range (IQR): 1.00-7.00]. Of these patients, 56.7% were male, and the median age at presentation was 20.03 months (IQR: 8.70-67.50). The most common CCHD diagnoses were Tetralogy of Fallot (TOF) (47.8%), pulmonary atresia (PA) (11.9%), Ebstein's anomaly (EA) (6.0%), and hypoplastic left heart syndrome (HLHS) (6.0%). The distribution of CCHD diagnoses is illustrated in Figure 1.

The most common presenting complaints were cough (41.3%), fever (41.0%), and vomiting (13.4%) (Table 1).

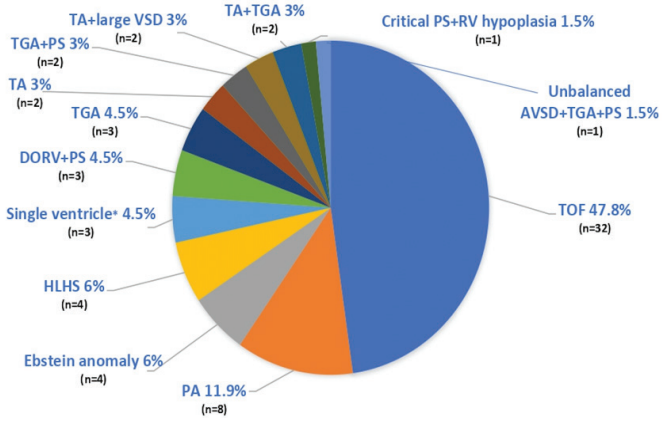

Figure 1. The distribution of diagnoses of patients cyanotic congenital heart diseases
*Single ventricle: Refers to children with anatomically single ventricle and diseases like double inlet left ventricle.
AVSD: Atrioventricular septal defect, DORV: Double outlet right ventricle, HLHS: Hypoplastic left heart syndrome, PA: Pulmonary atresia, PS: Pulmonary stenosis, RV: Right ventricle, TA: Tricuspid atresia, TGA: Transposition of the great arteries, TOF: Tetralogy of Fallot, VSD: Ventricular septal defect

Table 1. Characteristics of patients with cyanotic congenital heart disease presenting to the PED (n=351)

Characteristics	n (%)
Complaints	
Cough	145 (41.3)
Fever	144 (41.0)
Vomiting	47 (13.4)
Dyspnea/respiratory distress	44 (12.5)
Diarrhea	32 (9.1)
Increased cyanosis	30 (8.5)
Uneasiness	24 (6.8)
Sore throat	24 (6.8)
Chest pain	10 (2.8)
Eruption	10 (2.8)
Palpitation	5 (1.4)
Syncope	2 (0.6)
Seizure	2 (0.6)
Others	46 (13.1)
Visit diagnosis	
URTI	172 (49.0)
AGE	46 (13.1)
LRTI	41 (11.7)
Constipation	14 (4.0)
Otitis media	14 (4.0)
Soft tissue trauma	11 (3.1)
Urinary tract infection	7 (2.0)
Cyanotic spell	6 (1.7)
HF	5 (1.4)
Urticaria	5 (1.4)
Gastrointestinal bleeding	3 (0.9)
Cardiopulmonary arrest	2 (0.6)
Brain abscess	1 (0.3)
Seizure	1 (0.3)
Shunt dysfunction	1 (0.3)
Others	28 (8.0)
GCS	
≤8	5 (1.4)
9-12	10 (2.8)
13-15	336 (95.7)
Life-saving intervention	13 (3.7)
Significant IV fluid resuscitation	11 (3.1)
PPV	4 (1.1)
Intubation	4 (1.1)
CPR	2 (0.6)
PED observation unit admission	109 (31.0)
Hospitalization	44 (12.5)
PICU admission	24 (6.8)
Ventilation support	19 (5.4)
Circulatory support	13 (3.7)
Excitus	7 (2.0)

PED: Pediatric emergency department, URTI: Upper respiratory tract infection, AGE: Acute gastroenteritis, LRTI: Lower respiratory tract infection, HF: Heart failure, GCS: Glasgow Coma scale, IV: Intravenous, PPV: Positive pressure ventilation, CPR: Cardiopulmonary resuscitation, PICU: Pediatric intensive care unit

Among patients presenting with cough, the majority were diagnosed with upper respiratory tract infection (URTI) (n=101, 69.7%) or lower respiratory tract infection (LRTI) (n=37, 25.5%). Fever was most frequently associated with URTI (n=99, 68.8%), followed by acute gastroenteritis (AGE) (n=17, 11.8%) and LRTI (n=12, 8.3%). Among the 44 patients who presented with respiratory distress, 23 (52.3%) were diagnosed with LRTI, 14 (31.8%) were diagnosed with URTI, 2 (4.5%) were diagnosed with heart failure (HF), and 2 (4.5%) were diagnosed with a cyanotic spell. Among the thirty patients who presented with complaints of increased cyanosis, nine (30.0%) were diagnosed with URTI, six (20.0%) with LRTI, five (16.7%) with a cyanotic spell, and two (6.7%) with HF.

The most frequent diagnoses were URTI (49.0%), AGE (13.1%), and LRTI (11.7%) (Table 1). The median age of patients with LRTI was 10.43 months (IQR: 2.00-12.00). The most common underlying CCHDs in LRTI patients were TOF (n=13, 31.7%), EA (n=12, 29.3%), and ventricular septal defect with PA (n=9, 22.0%). Among these patients, 90.2% (n=37) were observed in the PED, 43.9% (n=18) were hospitalized, and 24.4% (n=10) were transferred to the PICU. The rate of PICU admission among hospitalized LRTI cases was 55.6%.

The most common cardiac-related diagnoses were cyanotic spell (1.7%) and HF (1.4%), with median ages at admission of 4.76 months (IQR: 1.36-64.70) and 1.83 months (IQR: 0.76-10.06), respectively. Cyanotic spells occurred in 4 patients with TOF (66.7%) and 2 patients with double outlet right ventricle (DORV) and pulmonary stenosis (PS) (33.3%). HF was observed in 3 (60.0%) HLHS patients, 1 (20.0%) with transposition of the great arteries (TGA), and 1 (20.0%) with tricuspid atresia (TA) and TGA. Among patients with cyanotic spells, presenting symptoms included increased cyanosis (n=5, 83.3%), respiratory distress (n=2, 33.3%), and syncope (n=1, 16.7%). Patients with HF most frequently presented with respiratory distress (n=2, 40.0%), increased cyanosis (n=2, 40.0%), cough (n=1, 20.0%), and uneasiness (n=1, 20.0%).

A total of 109 (31.1%) visits involved observation in the PED. The most common presenting complaints leading to observation were cough (n=53, 48.6%), fever (n=40, 36.7%), respiratory distress (n=30, 27.5%), and increased cyanosis (n=16, 14.7%). The leading diagnoses in this group were LRTI (n=37, 33.9%), URTI (n=30, 27.5%), and AGE (n=17,

Table 2. Risk factors for hospitalization

Characteristics	Hospitalization		Total (n=351)	p-value
	Yes (n=44)	No (n=307)		
Complaints				
Dyspnea/respiratory distress	15 (34.1)	29 (9.4)	44 (12.5)	<0.001[b]
Increased cyanosis	12 (27.3)	18 (5.9)	30 (8.5)	<0.001[a]
Syncope	2 (4.5)	0 (0)	2 (0.6)	0.015[a]
Visit diagnosis				
URTI	0 (0)	172 (56.0)	172 (49.0)	<0.001[b]
LRTI	18 (40.9)	23 (7.5)	41 (11.7)	<0.001[b]
Urinary tract infection	3 (6.8)	4 (1.3)	7 (2.0)	0.045[a]
Cyanotic spell	3 (6.8)	3 (1.0)	6 (1.7)	0.028[a]
HF	5 (11.4)	0 (0)	5 (1.4)	<0.001[a]
Cardiopulmonary arrest	2 (4.5)	0 (0)	2 (0.6)	0.015[a]
GCS				
GCS ≤12	13 (29.5)	2 (0.7)	15 (4.3)	<0.001[a]
Life-saving intervention	10 (22.7)	3 (1.0)	13 (3.7)	<0.001[a]
Significant IV fluid resuscitation	8 (18.2)	3 (1.0)	11 (3.1)	<0.001[a]
PPV	4 (9.1)	0 (0)	4 (1.1)	<0.001[a]
Intubation	4 (9.1)	0 (0)	4 (1.1)	<0.001[a]
CPR	2 (4.5)	0 (0)	2 (0.6)	0.015[a]
Ventilation support	18 (40.9)	1 (0.3)	19 (5.4)	<0.001[a]
Circulatory support	13 (29.5)	0 (0)	13 (3.7)	<0.001[a]

All values are presented as n (%).
[a]: Fisher's exact test, [b]: Pearson chi-square test, URTI: Upper respiratory tract infection, LRTI: Lower respiratory tract infection, HF: Heart failure, GCS: Glasgow Coma scale, IV: Intravenous, PPV: Positive pressure ventilation, CPR: Cardiopulmonary resuscitation

15.6%). Observation rates were significantly higher among patients with respiratory distress, increased cyanosis, URTI, LRTI, gastrointestinal bleeding, and HF (p<0.001, p=0.011, p<0.001, p<0.001, p=0.029, p=0.034, respectively). Among patients with LRTI, 90.2% (n=37) were observed in the emergency room.

A total of 44 visits (12.5%) resulted in hospitalization, occurring more frequently among infants with a mean age of 6.75 months (IQR: 1.91-11.49). The median length of hospital stay was 6.0 days (IQR: 4.0-9.75). The leading diagnoses among hospitalized patients were LRTI (40.9%), AGE (18.2%), and HF (11.4%). The hospitalization rate among patients with LRTI was 43.9%. Risk factors associated with hospitalization are presented in Table 2.

Twenty-four (6.8%) of the PED admissions required PICU care. The median PICU stay was 6.0 days (IQR: 3.0-9.5). Among these, the most common presenting symptoms were respiratory distress (n=9, 37.5%), cough (n=9, 37.5%), and increased cyanosis (n=9, 37.5%). The leading diagnoses were LRTI (n=10, 41.7%), HF (n=5, 20.8%), and cardiac arrest (n=2, 8.3%). 24.4% of patients diagnosed with LRTI were transferred to the PICU, and among those hospitalized due to LRTI, the rate of PICU stay was even higher, with a rate of 55.6%.

Additionally, the rate of admission to the PICU was significantly higher in patients with respiratory distress, increased cyanosis, fever, LRTI, cardiopulmonary arrest, and HF (p=0.001, p<0.001, p=0.003, p<0.001, p=0.004, p<0.001, respectively). Circulatory and ventilatory support were provided to 13 (54.2%) and 16 (66.7%) of the patients in the PICU, respectively.

Seven patients, with a median age of 10.13 months (IQR: 2.16-16.83), died. The causes of death were pneumonia (n=4), cardiac arrhythmia (n=2), and HF (n=1). One patient with DORV + PS and one with HLHS died due to arrhythmia. The patient who died from HF also had HLHS. As expected, HLHS had the highest associated mortality rate (50.0%). The mortality rates by underlying CHD were: HLHS (50.0%), DORV + PS (33.3%), EA (25.0%), PA (25.0%), TA with TGA (16.6%), and TOF (3.1%).

The GCS was ≥13 in 95.7% (n=336) of PED visits. Only one patient with a GCS of ≥13 died. GCS ≤12 was significantly associated with mortality (p<0.001). Emergency life-saving procedures were performed in 13 admissions (3.7%), and

Table 3. Risk factors for mortality

Characteristic	Mortality		Total (n=351)	p-value
	No (n=344)	Yes (n=7)		
Complaints				
Fever	144 (41.9)	0 (0)	144 (41.0)	0.044
Dyspnea/respiratory distress	40 (11.6)	4 (57.1)	44 (12.5)	0.006
Increased cyanosis	26 (7.6)	4 (57.1)	30 (8.5)	0.001
Visit diagnosis				
URTI	172 (50.0)	0 (0)	172 (49.0)	0.015
LRTI	38 (11.0)	3 (42.9)	41 (11.7)	0.037
HF	3 (0.9)	2 (28.6)	5 (1.4)	0.003
Cardiopulmonary arrest	0 (0)	2 (28.6)	2 (0.6)	<0.001
GCS				
GCS ≤12	9 (2.6)	6 (85.7)	15 (4.3)	<0.001
Life-saving intervention	10 (2.9)	3 (42.9)	13 (3.7)	0.001
Significant IV fluid resuscitation	8 (2.3)	3 (42.9)	11 (3.1)	0.001
PPV	2 (0.6)	2 (28.6)	4 (1.1)	0.002
Intubation	2 (0.6)	2 (28.6)	4 (1.1)	0.002
CPR	0 (0)	2 (28.6)	2 (0.6)	<0.001
PED observation unit admission	103 (29.9)	6 (85.7)	109 (31.1)	0.004
Hospitalization	37 (10.8)	7 (100.0)	44 (12.5)	<0.001
PICU admission	17 (4.9)	7 (100.0)	24 (6.8)	<0.001
Ventilation support	12 (3.5)	7 (100.0)	19 (5.4)	<0.001
Circulatory support	7 (2.0)	6 (85.7)	13 (3.7)	<0.001

All values are presented as n (%), and all comparisons were performed using Fisher's exact test.
URTI: Upper respiratory tract infection, LRTI: Lower respiratory tract infection, HF: Heart failure, GCS: Glasgow Coma scale, IV: Intravenous, PPV: Positive pressure ventilation, CPR: Cardiopulmonary resuscitation, PED: Pediatric emergency department, PICU: Pediatric intensive care unit

3 (23.1%) of these patients died. There was a significant correlation between emergency life-saving interventions and mortality (p=0.001). Specific interventions such as positive pressure ventilation, intubation, cardiopulmonary resuscitation (CPR), and significant IV fluid resuscitation were also significantly associated with mortality. The risk factors for mortality are presented in Table 3.

Logistic regression analysis identified urinary tract infection, LRTI, emergency life-saving intervention, and respiratory support as significant predictors of hospitalization. The model demonstrated excellent discrimination, with an AUC of 0.907 (95% CI: 0.859-0.956, p<0.001), sensitivity of 45.5%, specificity of 99.3%, PPV of 90.9%, and NPV of 92.7%. Significant associations were also found between mortality and increased cyanosis, dyspnea/respiratory distress, LRTI, HF, GCS ≤12, emergency life-saving intervention, significant IV fluid resuscitation, positive pressure ventilation, intubation, and circulatory support (Table 4).

Discussion

The majority of pediatric patients present to the PED with infectious diseases, most frequently with fever, cough, sore throat, earache, abdominal pain, diarrhea, vomiting, respiratory distress, and cyanosis.[10] Children with underlying cardiac disease are exposed to similar factors as the general population, although their clinical course and management may differ significantly due to their underlying cardiac pathology. Special attention is required during differential diagnosis as symptoms of cardiac and non-cardiac conditions often overlap. There are a limited number of studies evaluating the characteristics of CCHD patients presenting to the PED.[6,7] This study aimed to identify the characteristics of PED admissions and determine the predictors of hospitalization and mortality among CCHD patients. To the best of our knowledge, this is the first study addressing this topic in our country.

Table 4. Logistic regression of clinical features for prediction of hospitalization and mortality

	Hospitalization				Mortality	
	Univariable regression model		Multivariable regression model		Univariable regression model	
Characteristic	OR (95% CI)	p-value	OR (95% CI)	p-value	OR (95% CI)	p-value
Gender, girl	1.862 (0.984-3.524)	0.056			2.554 (0.562-11.599)	0.225
Age	0.972 (0.957-0.988)	<0.001	0.987 (0.973-1.001)	0.077	0.937 (0.865-1.016)	0.114
Fever	0.563 (0.284-1.119)	0.101				
Cough	0.981 (0.516-1.865)	0.954			1.920 (0.423-8.710)	0.398
Increased cyanosis	6.021 (2.661-13.624)	<0.001			16.308 (3.463-76.788)	<0.001
Dyspnea/respiratory distress	4.958 (2.386-10.303)	<0.001			10.133 (2.188-46.929)	0.003
Uneasiness	0.997 (0.285-3.489)	0.996			2.326 (0.269-20.147)	0.443
Diarrhea	2.134 (0.863-5.278)	0.101				
Vomiting	1.263 (0.527-3.025)	0.601				
Urinary tract infection	5.543 (1.198-25.650)	0.028	9.209 (1.591-53.315)	0.013		
AGE	1.573 (0.680-3.637)	0.289				
LRTI	8.548 (4.095-17.846)	<0.001	7.131 (2.701-18.829)	<0.001	6.039 (1.302-28.015)	0.022
Cyanotic spell	7.415 (1.448-37.964)	0.016				
GCS ≤12	63.952 (13.794-296.483)	<0.001			223.333 (24.302-2052.415)	<0.001
Life-saving intervention	29.804 (7.820-113.595)	<0.001	24.822 (3.416-180.347)	0.002	25.050 (4.939-127.053)	<0.001
Significant IV fluid resuscitation	22.519 (5.716-88.719)	<0.001			31.500 (6.031-164.516)	<0.001
Ventilation support	211.846 (27.188-1650.691)	<0.001	117.893 (13.574-1023.951)	<0.001		
PPV					68.400 (7.970-586.997)	<0.001
Circulatory support					288.857 (30.585-2728.057)	<0.001
HF					45.467 (6.183-334.343)	<0.001

OR: Odds ratio, CI: Confidence interval, AGE: Acute gastroenteritis, LRTI: Lower respiratory tract infection, GCS: Glasgow Coma scale, IV: Intravenous, PPV: Positive pressure ventilation, HF: Heart failure

TOF is the most common CCHD (5-10%), followed by TGA.[5] Consistent with previous studies, TOF (47.8%) was the most common CCHD followed by PA (11.9%). Similarly, Masood et al.[11] and Ilyas et al.[6] reported TOF as the most common CCHD among children admitted to PED (17.7% vs. 50%, respectively).

The most frequent presenting symptoms of CHD include respiratory distress, cyanosis, cough, feeding difficulties, fever, inadequate weight gain, and palpitations.[11-13] In our study, the most common reasons for admission were cough and fever; followed by vomiting, respiratory distress, diarrhea, and increased cyanosis, in alignment with the existing literature.

URTI is the most frequent diagnosis in the PED, followed by fever, asthma, otitis media, and viral infections.[10] Similarly, respiratory tract infections have been reported as the predominant diagnoses among children with CHD in previous studies.[14,15] This increased susceptibility, especially in children under 24 months, is attributed to their limited cardiopulmonary reserve.[7,16] Children with CHD have a threefold increased risk of developing LRTIs, which also constitutes the most common reason for hospitalization.[17] Lee et al.[14] reported an earlier onset of respiratory tract infections in patients with CCHD (median age 1.1 years). Similarly, patients with LRTI had a mean age of 10.43 months and it was the most common reason for hospitalization in our study.

The most common cardiac causes of PED admission were cyanotic spells and HF. Cyanotic spells typically occur in infants aged 2 months to 2 years with reduced pulmonary blood flow (PBF), often triggered by a decreased systemic vascular resistance and increased right-to-left shunting. This further reduction in PBF leads to worsening hypoxia and deepening cyanosis.[18] Cyanotic spells are most commonly observed in TOF, PA and DORV with PS. In our study, cyanotic spells were more frequently observed in TOF patients, with a median age of 4.7 months, consistent with expected clinical patterns.

HF results from the heart's inability to meet metabolic demands of the body due to decreased cardiac output.[19] Its incidence is estimated at 0.1-0.2% among all live births and 6-24% among children with CHD.[20-22] CHD is the leading cause of HF in children, often presenting during the first year of life. In the United States, approximately 11,000-14,000 children are hospitalized annually with HF and an overall mortality rate of 7%, compared to 0.4% in children without HF.[20-24] Lee et al.[14] reported HF as the cause of 14.7% of PED admissions among children with congenital CHD.[6,25,26] In our study, HF was the second most common cardiac diagnosis, accounting for 14.3% of cases. The leading presenting complaints were increased cyanosis, respiratory distress, and cough. The reduced incidence of HF in older children may be attributed to the effects of corrective or palliative surgical interventions.

LRTI, AGE, and HF were the most common indications for hospitalization, while LRTI, HF, and cardiac arrest predominated among those requiring PICU care. LRTI accounts for 3-18% of hospitalizations in high-income countries.[17] Pneumonia is more frequent and severe in patients with CHD and is associated with increased mortality.[27-29] PICU admission is required in 3-14% of hospitalized cases with LRTI.[17] In our study, 90.2% of CCHD patients with LRTI were observed in the PED; 43.9% were hospitalized, and 24.4% required PICU admission. Among hospitalized LRTI cases, 55.6% required PICU care, highlighting a higher burden in the CCHD population.

In our study, respiratory distress, increased cyanosis, cyanotic spells, and a GCS score of ≤12 were identified as significant predictors of hospitalization. As with all critically ill patients, timely and effective interventions are essential for children with cyanotic CCHD presenting to the emergency department. Emergency life-saving interventions were also among the most significant parameters associated with hospitalization. PICU management commonly involved both invasive and non-invasive mechanical ventilatory support, circulatory support with vasopressors, IV fluid resuscitation, and blood product transfusions. Our findings showed that 6.8% of CCHD patients required PICU admission, and those admitted frequently needed high levels of support, including mechanical ventilation (66.7%) and circulatory support (54.2%).

CHD accounts for approximately 12.4% of all childhood deaths.[29] Advances in diagnosis and interventional therapies have significantly improved survival; nonetheless, HLHS continues to carry the highest mortality. Recent studies show that 96% of infants with CHD who survive their first year remain alive at age[16], shifting CHD-related mortality into adulthood.[30,31] Globally, reductions in CHD mortality vary according to socioeconomic status, ranging from 34.5% in low-income settings to 64% in high-income countries.[32] As CHD-related mortality shifts into adulthood, the leading causes of death also evolve, with arrhythmia and HF being the most common.[30] Lee et al.[14] reported respiratory tract infection (5.8%) as the leading cause of mortality in CHD patients. Similarly, in our study, pneumonia was the most common cause of death, followed by arrhythmia and HF. Several clinical parameters—such as increased cyanosis, emergency life-saving interventions, intravenous (IV) fluid resuscitation, respiratory and circulatory support, and GCS score—were significantly associated with mortality. GCS at the time of admission has been reported as a strong predictor of mortality.[33,34] In our study, 40% of patients with a GCS score ≤12 died. Our study showed that patients with CCHD should be managed more carefully than the healthy population, even when they have a higher GCS score, because of the increased risk of sudden deterioration. Additionally, early and effective emergency

life-saving interventions are essential in preventing mortality in patients with CCHD. Health-care access and quality are another important factor for prognosis. In our country, the national health insurance system provides full coverage for all children under the age of 18; however, low family income can still pose a barrier to accessing health care, particularly for those living in rural areas. Furthermore, parental education levels may affect caregivers' ability to recognize early signs and symptoms of illness, leading to delayed medical intervention, more severe clinical presentation at the time of hospitalization, and a poorer prognosis for affected children.

Study Limitations

This study has several limitations. First of all, this study is subject to the usual limitations of a retrospective study. Second, our center is a tertiary referral hospital with a specialized pediatric cardiology unit and PICU, which may introduce selection bias, as more complex or severe cases are preferentially referred. Third, variability in clinical assessment among different physicians may have influenced decisions regarding admission and observation. Lastly, associated genetic syndromes or extracardiac anomalies—which may affect clinical severity and outcomes—were not included in the statistical analysis.

Conclusion

Due to the complex nature of CCHD, clinicians must be vigilant regarding risk factors for hospitalization and mortality. Even patients presenting with seemingly mild symptoms may deteriorate rapidly. Early recognition and timely emergency interventions are critical. A better understanding of the demographic characteristics and predictors of poor outcomes will support the development of more effective management strategies for this vulnerable population. Further prospective, multicenter studies are warranted to validate and expand upon our findings. The predictive markers identified in this study may serve as a valuable guide for pediatricians in the emergency management of children with CCHD.

Ethics

Ethics Committee Approval: The study protocol was approved by the Ethics Committee of Ankara University Faculty of Medicine (approval date: April 5, 2023; approval number: İ03-164-23), and the study was conducted in accordance with the principles of the Declaration of Helsinki.

Informed Consent: Retrospective study.

Footnotes

Authorship Contributions

Concept: G.V., M.G.R., B.G., G.B.Y., S.A., İ.K., T.U., T.K., D.T., Design: G.V., M.G.R., B.G., G.B.Y., U.Ç., T.K., Data Collection or Processing: G.V., M.G.R., B.G., G.B.Y., U.Ç., S.A., İ.K., Analysis or Interpretation: G.V., M.G.R., İ.K., T.U., T.K., D.T., Literature Search: G.V., M.G.R., B.G., G.B.Y., U.Ç., S.A., İ.K., T.U., T.K., D.T., Writing: G.V., M.G.R., B.G., G.B.Y., U.Ç., S.A., İ.K., T.U., T.K., D.T.

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