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Pediatric Clinical and Epidemiological Profile of Congenital Heart Disease

Perfil Clínico e Epidemiológico Pediátrico das Cardiopatias Congênitas
Perfil Clínico y Epidemiológico Pediátrico de las Cardiopatías Congénitas

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Abstract: Introduction: Congenital heart diseases are anomalies affecting the heart structure at birth, with multifactorial etiology. The incidence of these conditions is approximately 8 per 1,000 live births and they are the second leading cause of death in this age group. The objective of this article was to summarize the clinical and epidemiological profile of patients with congenital heart disease. Methodology: This is a systematic literature review in the PubMed, Scielo, and BVS databases. Initially, 86 articles were found, and after applying exclusion criteria, 8 articles remained, of which only 3 addressed the clinical and epidemiological profile of patients with congenital heart disease and related it to some clinical outcome (discharge, surgical intervention, or death). Results: The profile found is male, infant, from metropolitan areas, with non-cyanotic heart disease, diagnosed in the postnatal period. Conclusion: This review was able to define a clinical and epidemiological profile. However, the sample of studies evaluated is small, showing a gap in the literature. Risk factors for worse outcomes were identified (cyanotic heart disease, age at surgical intervention, and number of post-surgical complications), highlighting the importance of better medical care. In this way, timely intervention and better follow-up of these patients would be possible, in an attempt to mitigate worse outcomes.

Keywords: health profile; congenital heart defects; pediatrics

Resumo: Introdução: Cardiopatias congênitas são anomalias que atingem a estrutura cardíaca ao nascer, que apresentam etiologia multifatorial.. A incidência dessas condições é cerca de 8 para cada mil nascidos vivos e correspondem à segunda causa de óbito nessa faixa etária. O objetivo desse artigo foi resumir o perfil clínico e epidemiológico dos pacientes com cardiopatia congênita. Metodologia: Trata-se de revisão sistemática de literatura nas bases de dados PubMed, Scielo e BVS. Foram encontrados inicialmente 86 artigos e após aplicar critérios de exclusão, restaram 8 artigos, os quais apenas 3 abordaram o perfil clínico e epidemiológico dos pacientes com cardiopatia congênita e relacionam com algum desfecho clínico (alta, intervenção cirúrgica ou óbito). Resultados: o perfil encontrado é masculino, lactente, procedente de regiões metropolitanas, com cardiopatias não cianóticas, diagnosticado no período pós-natal. Conclusão: A presente revisão foi capaz de delimitar um perfil clínico e epidemiológico. No entanto, a amostra de estudos avaliada é pequena, mostrando uma lacuna na literatura. Foram identificados fatores de risco para pior desfecho (cardiopatia cianótica, idade na abordagem cirúrgica e número de complicações pós-cirúrgicas), destacando-se a importância de melhor assistência médica. Dessa forma, seria possível uma intervenção oportuna e melhor seguimento desses pacientes, na tentativa de mitigação de piores desfechos.

Palavras-chave: perfil epidemiológico; cardiopatias congênitas; pediatria

Resumen: Introducción: Las cardiopatías congénitas son anomalías que afectan la estructura cardíaca al nacer y tienen etiología multifactorial. La incidencia de estas condiciones es de aproximadamente 8 por cada 1.000 nacidos vivos y representan la segunda causa de muerte en este grupo etario. El objetivo de este artículo fue resumir el perfil clínico y epidemiológico de los pacientes con cardiopatía congénita. Metodología: Se realizó una revisión sistemática de la literatura en las bases PubMed, SciELO y BVS. Inicialmente se identificaron 86 artículos y, después de aplicar los criterios de exclusión, quedaron 8, de los cuales solo 3 abordaron el perfil clínico y epidemiológico de los pacientes con cardiopatía congénita y lo relacionaron con algún resultado clínico (alta, intervención quirúrgica o muerte). Resultados: El perfil identificado fue masculino, lactante, procedente de regiones metropolitanas, con cardiopatías acianóticas y diagnóstico en el período posnatal. Conclusión: La revisión permitió delimitar un perfil clínico y epidemiológico. Sin embargo, la muestra de estudios evaluados fue pequeña, lo que evidencia una laguna en la literatura. Se identificaron factores de riesgo de peores resultados (cardiopatía cianótica, edad al momento de la intervención quirúrgica y número de complicaciones posquirúrgicas), destacándose la importancia de una mejor atención médica. Así sería posible intervenir oportunamente y realizar un mejor seguimiento para reducir los resultados desfavorables.

Palabras clave: perfil epidemiológico; cardiopatías congénitas; pediatría

Introduction

Congenital heart diseases are structural abnormalities of the heart or its vessels that are present at birth and lead to significant or potentially significant functional consequences (SILVA, 2022). Their incidence ranges from 8 to 10 per 1,000 live births, and approximately one fifth of affected children under one year of age will die from this cause, making it the second leading cause of death in this age group (SOARES, 2020).

These malformations have a multifactorial etiology that includes both genetic and non-genetic factors. During cell division, changes may occur in the inherited genome and generate genetic abnormalities that result in cardiac malformations, as observed in Turner syndrome and trisomies 21, 18, and 13 (FERREIRA, 2024). Among non-genetic factors, environmental exposures, the use of medications or illicit substances by the pregnant woman, maternal diseases (gestational diabetes and lupus), and infections during pregnancy, such as rubella or syphilis, should be highlighted (SOARES, 2020).

This type of condition may be classified according to hypoxia, for example. Under this classification, cyanotic congenital heart diseases (with cyanosis) are characterized by insufficient blood oxygenation resulting from structural abnormalities that cause venous blood to bypass adequate pulmonary oxygenation (through a right-to-left shunt or obstruction of pulmonary blood flow) (VARDASCA, 2025). Acyanotic congenital heart diseases, in turn, are

structural abnormalities that do not impair blood oxygenation and do not produce clinical signs of cyanosis. The main congenital heart diseases of each type are presented in Chart 1 below.

Chart 1 - Classification of the main congenital heart diseases according to the presence of hypoxia.

Cyanotic congenital heart diseases	**Acyanotic congenital heart diseases**
Tetralogy of Fallot (TOF)	Atrial septal defect (ASD)
Pulmonary atresia with intact ventricular septum (PA/IVS)	Ventricular septal defect (VSD)
Critical pulmonary stenosis	Complete atrioventricular septal defect (CAVSD)
Tricuspid atresia	Partial atrioventricular septal defect (PAVSD)
Severe neonatal Ebstein anomaly	Aortic stenosis
	Patent ductus arteriosus (PDA)
	Coarctation of the aorta (CoA)

Source: prepared by the authors, 2025.

Congenital heart diseases may also be classified according to severity. Critical heart diseases are those requiring surgical treatment or catheterization during the first year of life and are screened by pulse oximetry before hospital discharge after birth (MS, 2021). Non-critical, or simple, heart diseases can be managed clinically or do not require a surgical approach during the neonatal period (SILVA, 2022). This distinction is important for appropriate postnatal therapeutic planning.

Ideally, these congenital abnormalities should be diagnosed prenatally by fetal echocardiography so that delivery can take place at a facility with a neonatal cardiology center capable of providing optimal support from the first minutes of extrauterine life (JÚNIOR, 2023). However, this is not always possible because of limited access to health services and complementary tests. A thorough examination should therefore be performed after birth to assess the presence of a heart murmur, cyanosis, tachypnea, and arrhythmias (SILVA, 2022). In addition, all newborns above 34 weeks of gestation should undergo cardiac neonatal screening by pulse oximetry of the right upper limb and one lower limb between 24 and 48 hours of life. If any measurement is below 95% or the difference between measurements is 4% or greater, the test should be repeated after one hour. If the abnormality persists, the patient

should undergo echocardiography, and discharge should be postponed until the diagnosis is clarified. This screening strategy seeks early identification before hospital discharge, thereby minimizing morbidity and mortality associated with late diagnosis (MS, 2021).

Once the diagnosis has been established, the next step is therapeutic planning. For patients with non-critical heart disease, pharmacological treatment may be used to control symptoms of heart failure or pulmonary hypertension, together with clinical monitoring and guidance for parents regarding warning signs (CORDOVIL, 2024). For patients who require surgery, several techniques are available, including defect repair, reconstruction of cardiac structures, and even heart transplantation in severe, refractory cases (MAXIMILIANO, 2024).

Thus, identifying the clinical and epidemiological profile of pediatric patients with congenital heart disease enables health services, particularly the Brazilian Unified Health System (SUS), to direct resources toward early diagnosis and improved therapeutic management in pursuit of better outcomes, since patients treated late have poorer quality of life because of disease progression (NOVELLETO, 2025).

Accordingly, this article presents a systematic literature review whose general objective was to summarize the clinical and epidemiological profile of pediatric patients with congenital heart disease, highlight prenatal risk factors, and discuss a diagnostic flowchart that considers the resources available within the SUS.

Methodology

This is a qualitative systematic literature review. Electronic searches were conducted in PubMed, SciELO, and BVS. The review included descriptive and retrospective studies based on document analysis, as well as literature reviews in Portuguese, English, and Spanish, published from 2003 through June 2024, addressing the clinical and epidemiological profile of pediatric patients with congenital heart disease.

The literature search was conducted systematically in PubMed, SciELO, and BVS by independent researchers using the Health Sciences Descriptors shown in Chart 2. The Parsifal tool was then used to select articles according to the PICOD framework established in Chart 3.

The study was initially registered on the PROSPERO platform under the title “Pediatric clinical and epidemiological profile of congenital heart disease - a systematic review,” with registration number CRD420251104460.

Chart 2. Descriptors according to DeCS.

Health Sciences Descriptors (DeCS/MeSH), as defined at https://decs.bvsalud.org/	**Portuguese: "Cardiopatias Congênitas" AND "Perfil Epidemiológico" OR "Perfil clínico"**
	English: "Heart Defects, Congenital" AND "Health Profile" OR "Epidemiological Profile"
	Spanish: "Cardiopatías Congénitas" AND "Perfil de Salud" OR "Perfil Epidemiológico"

Source: prepared by the authors, 2025.

Chart 3. PICOD.

PICOD	**Participants: pediatric patients with congenital heart disease**
	Intervention: risk factors for congenital heart disease
	Comparison: healthy patients
	Outcome: discharge, surgical intervention, or death
	Design: descriptive and retrospective studies based on document analysis and literature reviews in Portuguese, English, and Spanish, published from 2003 through June 2024 and retrieved from PubMed, SciELO, and BVS

Source: prepared by the authors, 2025.

Eligibility Criteria

The review included descriptive and retrospective studies based on document analysis, as well as literature reviews in Portuguese, English, and Spanish, published from 2003 through June 2024, that addressed the clinical and epidemiological profile of pediatric patients with congenital heart disease, considering the descriptors listed in Chart 2 and similar terms available in DeCS.

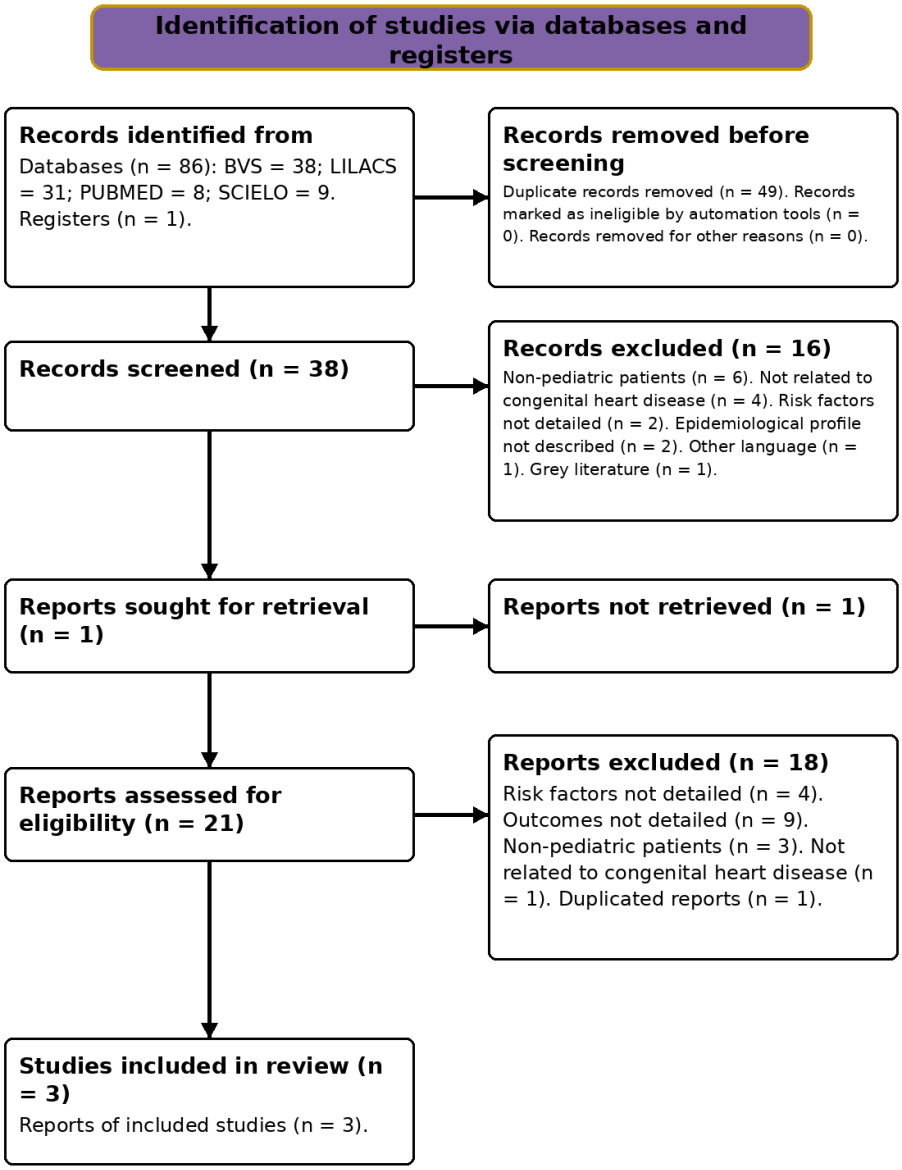
Studies involving non-pediatric patients, studies unrelated to congenital heart disease, studies that did not detail risk factors or describe an epidemiological profile, and studies published in languages other than Portuguese, English, or Spanish were excluded. Duplicate studies and studies for which full-text access was not available were also excluded.

Search Systematization

The process occurred in four stages, following the PRISMA flowchart shown in Figure 1. First, a search strategy using the descriptors specified in Chart 2 identified 86 articles. Duplicates were then removed, leaving 38 articles for the third stage: title and abstract screening by two independent researchers and application of the inclusion and exclusion criteria. When the two reviewers disagreed, a third reviewer was consulted. Finally, eight articles remained, but only three addressed the clinical and epidemiological profile of pediatric patients with congenital heart disease and related it to a clinical outcome (discharge, surgical intervention, or death); only these studies were used in the systematic review presented in this article.

Figure 1 - Article search flowchart - PRISMA 2020 guideline.

PRISMA 2020 flow diagram for new systematic reviews that included searches of databases and registers only



Source: Page MJ et al. BMJ 2021;372:n71. doi: 10.1136/bmj.n71.

Results

The studies selected at the end of the systematic review strategy are presented in Chart 4.

Chart 4 - Studies included in the systematic review.

Author and publication year	Study type	Objective	Results	Conclusion
Juciane R Guimarães et al. (2020)	Cross-sectional observational	To describe the clinical and epidemiological profile of patients admitted to the pediatric cardiac intensive care unit (ICU) of a tertiary hospital.	Patients aged 28 days to one year predominated (44%). Congenital heart disease (91.9%) was more prevalent than acquired heart disease (8.1%). Cardiopulmonary bypass was used in 138 surgical patients and lasted from 12 to 261 minutes. Most patients (88.9%) were discharged from the ICU, and 11.1% died. According to the RACHS-1 score, corrective cardiac surgery was performed in 75.8% of patients and palliative surgery in 24.2%.	Patients aged 28 days to one year with cyanotic congenital heart disease who undergo cardiac surgery with cardiopulmonary bypass lasting more than 120 minutes have a higher risk of death.
José Aderval Aragão et al. (2013)	Cross-sectional observational	To describe the epidemiological profile of patients with congenital heart disease who underwent surgery at Hospital do Coração.	Of the 300 patients with congenital heart disease who underwent surgery during the period, 69.7% (209) had acyanotic disease, most commonly ventricular septal defect at 21% (63). Among cyanotic diseases, tetralogy of Fallot accounted for 14% (42). Cyanotic cases accounted for 82.7% (24) of deaths and acyanotic cases for 17.3% (5).	The patients with congenital heart disease were infants, preschoolers, and school-age children, with no sex predominance. Acyanotic congenital heart diseases predominated, but most deaths occurred among patients with cyanotic heart disease.
Miriam Madhelayne P Centeno and Fabricio González (2019)	Longitudinal observational	To determine the prevalence of congenital heart disease and characterize the group of newborns with the condition at Hospital Vicente Corral Moscoso (HVCM) in Cuenca.	In the study sample (123 newborns), 83.7% of newborns with congenital heart disease were born in Cuenca; the others were transferred from other cities and provinces. A total of 97.6% were diagnosed postnatally (n = 120), 51.2% were male (n = 63), and 93.5% were mestizo. Of the mothers, 44.7% were aged 21-30 years, 27.6% were aged 10-20 years, and only 8.1% were aged 41-50 years. Classification by number of heart defects showed multiple conditions in 32.5% of cases, and cyanotic heart disease was observed in 21.1% (n = 26). By severity, 73.2% were mild and 13% very severe. Clinical resolution occurred in 43.9%; surgical treatment and catheterization were required in 13% and 4.9%, respectively. Mortality was 15.4%. Prevalence per 1,000 live births was estimated by heart defect; atrial septal defect had the highest	Using INEC data and hospital discharges of children under one year of age, prevalence for 2009-2017 was established at 2.8 nationally (CI 2.49-3.11), 2.36 in Azuay (CI 1.61-3.10), 2.76 in Cuenca (CI 1.86-3.66), and 3.77 at HVCM (CI 2.39-5.14). At HVCM, prevalence among newborns was 3.77 per 1,000 live births.

Author and publication year	Study type	Objective	Results	Conclusion
			prevalence at 1.26, followed by patent ductus arteriosus at 1.11.	

Source: prepared by the authors, 2025.

Guimarães and Guimarães (2020) conducted a study in Salvador, Bahia, analyzing 307 patients admitted to a pediatric cardiac intensive care unit (ICU) between 2013 and 2014. The study was set in an intensive hospital-care context and provided more detailed information on clinical and technical variables. Most patients had congenital heart disease (91.9%); among acyanotic cases, ventricular septal defect (24.5%) and atrioventricular septal defect (19.9%) predominated, while tetralogy of Fallot (30%) predominated among cyanotic cases. In-hospital mortality was 11.1%. Statistical analyses identified age below one year, cyanotic congenital heart disease, and cardiopulmonary bypass lasting more than 120 minutes as the principal factors associated with death. The study also used the RACHS-1 score, a surgical risk stratification tool that confirmed the association between greater procedural complexity and increased mortality. Another relevant finding concerned the immediate causes of death: cardiogenic shock accounted for 61.8% of cases and septic shock for 35.5%, demonstrating the importance of infectious complications in in-hospital mortality.

Aragão et al. (2013) conducted a retrospective study in Aracaju, Sergipe, involving 300 patients under 18 years of age who underwent cardiac surgery for congenital heart disease between 2005 and 2010. This hospital-based surgical study highlighted the prevalence of acyanotic heart diseases (69.7%), especially ventricular septal defect, while tetralogy of Fallot was the most frequent cyanotic condition. Its main contribution was to show that mortality was disproportionately higher among patients with cyanotic heart disease (82.7% of deaths). Deaths occurred mainly during surgery or within the first 24 postoperative hours, suggesting both clinical fragility and structural limitations in hospital support. The study also provided information on associated conditions, such as Down syndrome (4% of cases), which is often related to atrioventricular septal defects.

Paucar, Orbe, and Patino (2018), in a study conducted in Ecuador, offered a different perspective by analyzing 123 neonates diagnosed with congenital heart disease at a referral hospital between 2009 and 2017. The study focused on the perinatal and neonatal periods, allowing maternal epidemiological and prenatal-care factors to be correlated with the

occurrence and severity of congenital heart disease. The prevalence was 3.77 per 1,000 live births, higher than the Ecuadorian national average. Atrial septal defect (1.26 per 1,000) and patent ductus arteriosus (1.11 per 1,000) were the most common conditions. Approximately one third of the neonates had multiple cardiac malformations, and 21.1% had cyanotic heart disease. A particularly relevant diagnostic finding was that 97.6% of cases were identified only after birth, revealing major failures in prenatal screening. Mortality reached 15.4% and was concentrated among the most complex cases. Regarding maternal factors, young mothers predominated (44.7% were aged 21-30 years), but adolescent pregnancies also represented a substantial proportion (27.6%), highlighting social and demographic factors as additional determinants of neonatal mortality.

Statistical Analyses

This stage sought to integrate and quantitatively explore data from the selected studies, enabling a more precise assessment of outcomes related to congenital heart disease. The subsection is therefore organized into two complementary parts: the first examines pooled mortality by combining and synthesizing individual results into an overall picture; the second estimates measures of association, specifically relative risk and odds ratio, to clarify the magnitude of the relationship between congenital heart disease and the different types of mortality identified.

Pooled Mortality

To compare the general clinical outcomes of the studies included in this review, a chart was prepared showing the overall mortality rates observed in each population, as presented below:

Chart 1 - Overall mortality rate for each selected study

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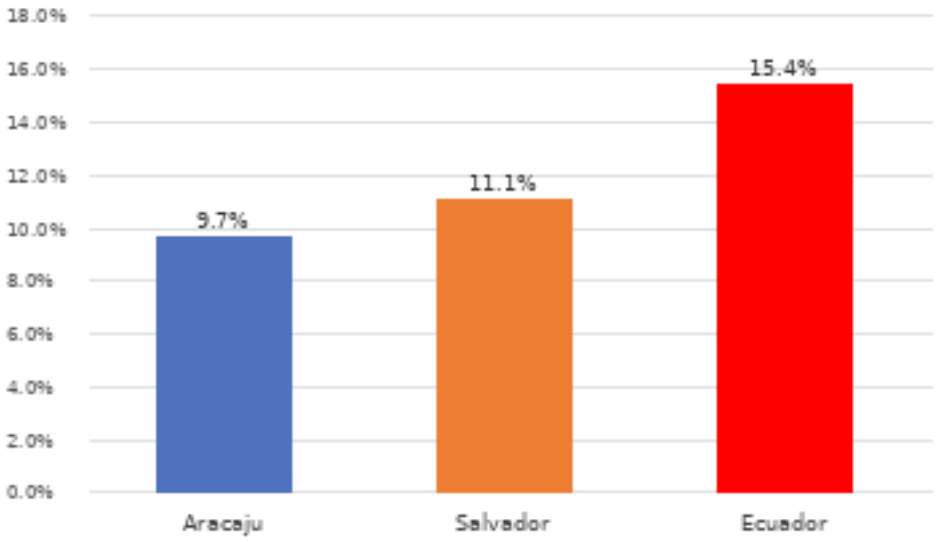

Source: prepared by the authors (2025).

Comparison of the overall mortality rates among the three studies reveals relevant variation in the impact of congenital heart disease on clinical outcomes. The Aracaju study reported the lowest mortality rate (9.7%), while Salvador reported a slightly higher value (11.1%). Ecuador had the highest percentage (15.4%), suggesting greater clinical severity or structural and care-related differences in that country's neonatal and pediatric context. Although all studies identify congenital heart disease as an important cause of death in pediatric populations, the observed contrasts may be related to differences in sample profiles, diagnostic methods, availability of therapeutic resources, timing of intervention, and quality of hospital care. The picture provided by Chart 1 therefore reinforces both the magnitude of the problem and the need for policies to expand early diagnosis and improve specialized care, particularly in more vulnerable settings.

Table 1 below presents the number of patients included in each study, the total number of deaths, study-specific mortality rates, and pooled mortality across the three locations analyzed.

Table 1 - Patients, deaths, mortality rate, and pooled mortality

Location	Number of patients	Deaths	Overall mortality rate	Pooled mortality	Confidence interval (95% CI)
Aracaju	300	29	9,7%		
Salvador	307	34	11,1%	**11,20%**	**9,1% - 13,7%**
Ecuador	123	19	15,4%		

Source: prepared by the authors (2025).

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In Aracaju, 29 of the 300 patients evaluated died, corresponding to a mortality rate of 9.7%. In the Salvador study, 34 deaths were recorded among 307 patients, representing mortality of 11.1%. In Ecuador, 19 of the 123 patients died, producing a mortality rate of 15.4%, the highest among the three studies.

When the data were pooled, overall mortality among the 730 patients evaluated reached 11.2%, with a 95% confidence interval ranging from 9.1% to 13.7%. This result indicates that, overall, approximately one in nine patients with congenital heart disease died across the different settings. Although rates varied among the studies, the values were relatively consistent, particularly in Aracaju and Salvador, while Ecuador had a higher proportion, possibly related to the neonatal profile of its sample and structural limitations in access to diagnosis and specialized treatment.

The pooled analysis thus demonstrates significant mortality among patients with congenital heart disease, highlighting the epidemiological relevance of these conditions and the need for early-intervention strategies and appropriate clinical and surgical management, especially in settings with greater structural and care-related vulnerabilities.

Relative Risk and Odds Ratio

To deepen the understanding of mortality associated with congenital heart disease, this review uses statistical measures of association, specifically relative risk (RR) and odds ratio (OR). RR compares the probability of an event, in this case death, between patients exposed to a specific condition (cyanotic heart disease) and those not exposed (acyanotic heart disease), providing a direct estimate of increased risk. OR measures the ratio between the odds of death and the odds of survival and is particularly useful in retrospective studies; it tends to overestimate the magnitude of the association when the event is not rare, but it reinforces the strength of the observed effect. Understanding these measures is essential for interpreting the clinical relevance of the mortality differences observed between types of heart disease. The results are shown in Table 2 below:

Table 2 - Mortality types, relative risk, and odds ratio

Location	Acyanotic mortality	Cyanotic mortality	**RR**	**OR**
Aracaju	2,4%	26,4%	11	14,6
Salvador	7,3%	17,6%	2,4	2,7
Ecuador	5,2%	53,8%	10,4	21,5

Source: prepared by the authors (2025).

Comparative analysis of the three studies included in this review consistently shows that mortality was higher among patients with cyanotic congenital heart disease than among those with acyanotic disease. In the Aracaju study, 26.4% of patients with cyanotic heart disease died, compared with only 2.4% of acyanotic patients. This difference produced an RR of 11.0, indicating that patients with cyanotic heart disease had an elevenfold risk of death. The calculated OR was 14.6, reinforcing the magnitude of the association. In Salvador, deaths occurred in 17.6% of cyanotic cases and 7.3% of acyanotic cases, corresponding to an RR of 2.4 and an OR of 2.7. Although the difference in this study was smaller than in Aracaju, it still confirmed the greater lethality of cyanotic forms. In Ecuador, mortality among patients with cyanotic heart disease reached 53.8%, compared with 5.2% among acyanotic patients, representing an RR of 10.4 and an OR of 21.5, the strongest association among the three studies analyzed.

Despite differences in setting and population, the convergence of results provides robust evidence that cyanotic congenital heart disease is an independent and significant risk factor for mortality. These findings reinforce the need for early diagnosis, timely referral to specialized centers, and rapid access to surgical treatment, since these measures can substantially reduce the lethality associated with these conditions.

Discussion

Despite methodological differences, the three studies share convergent elements that make it possible to outline the clinical and epidemiological profile of children hospitalized with congenital heart disease. Demographically, although sex distribution was relatively balanced, there was a slight male predominance. Neonates were the most affected age group; clinical presentation tends to be more severe in this period, requiring more frequent hospitalization and surgery. Most patients came from metropolitan areas near referral centers, although there was also a notable flow from inland areas, as shown in the Salvador study (64.8%), suggesting unequal access to diagnosis and specialized treatment.

Clinically, most diagnosed heart diseases were acyanotic, including ventricular septal defect (VSD), patent ductus arteriosus (PDA), and atrial septal defect (ASD), although cyanotic diseases were strongly associated with higher mortality and complication rates, especially tetralogy of Fallot and transposition of the great arteries (TGA). Associated conditions, such as malnutrition (affecting more than half of patients in Salvador) and genetic syndromes,

particularly Down syndrome, were also frequent and contributed to greater clinical vulnerability. Most patients required surgical treatment, often urgently, and both cardiopulmonary bypass time and the anatomical severity of the heart defect were important prognostic factors for the risk of death.

In summary, the typical patient hospitalized with congenital heart disease in the settings studied was a child from a few months to a few years of age, often diagnosed late or after birth, from a peripheral or inland area, and more frequently affected by acyanotic heart disease, but at substantial risk of complications and death when cyanotic forms, malnutrition, or concomitant malformations were present. This profile reinforces the need for early-diagnosis strategies, nutritional follow-up, and expanded access to specialized pediatric cardiology services.

Chart 5 below highlights the main comparative characteristics reported by each selected study:

Chart 5 - Main comparative characteristics of the selected studies

Characteristics	**Aracaju (Brazil)**	**Salvador (Brazil)**	**Cuenca (Ecuador)**
Setting/Period	Hospital do Coração, 2005-2010	Pediatric Cardiac ICU, 2013-2014	Hospital Vicente Corral Moscoso, 2009-2017
Sample	300 patients (<18 years, surgical)	307 patients (0-15 years)	123 neonates
Predominant age group	Infants, preschoolers, school-age children	28 days to 1 year (44%)	Neonates (at birth)
Sex	No predominance	52.4% male	51.2% male
Main acyanotic heart diseases	VSD (21%), PDA (18%), ASD (7.7%)	VSD (24.5%), AVSD (19.9%), PDA/ASD (13.2%)	ASD (32.2%), PDA (28.4%), VSD (10.9%)
Main cyanotic heart diseases	Tetralogy of Fallot (14%), TGA (6.3%)	Tetralogy of Fallot (30%), tricuspid atresia (17.6%)	Tetralogy of Fallot (0.04‰), TGA (0.15‰)
Overall mortality	9,7% (29/300)	11,1% (34/307)	15,4% (19/123)
Cyanotic disease vs mortality association	82.7% of deaths in cyanotic patients	68.8% of deaths in cyanotic patients	73.7% of deaths in cyanotic patients

Main additional risk factors	Postoperative complications (cardiac arrest, acidosis, bleeding); structural hospital limitations	CPB >120 min, sepsis, cardiogenic shock, age <1 year	Young maternal age, lack of prenatal care, multiple heart defects
Associated Down syndrome	4%	Not reported	13,80%

Source: prepared by the authors (2025).

Regarding the other comparative characteristics, the type of heart disease was confirmed as a central determinant. Across the three studies, cyanotic heart diseases were consistently associated with higher mortality rates in surgical settings, intensive care units, and the neonatal period. This finding reinforces that the anatomical and pathophysiological severity of the heart disease is one of the main predictors of mortality.

Second, early age emerged as a critical factor. Both the Brazilian ICU study and the Ecuadorian study showed that neonates and infants have greater clinical vulnerability and an increased risk of death. This finding is consistent with established knowledge that the first year of life, particularly the neonatal period, is a phase of physiological instability requiring early diagnosis and intervention to ensure better chances of survival.

Another important point concerns hospital infrastructure and the technical aspects of procedures. Guimarães and Guimarães (2020) highlighted prolonged cardiopulmonary bypass and infectious complications as determinants of mortality. Similarly, Aragão et al. (2013) identified surgery and the immediate postoperative period as the highest-risk times, suggesting limitations in the support available. These findings indicate that outcomes are directly affected not only by the intrinsic severity of the heart disease but also by the structural and technical conditions of the care provided.

Paucar, Orbe, and Patino (2018) added the dimension of social and maternal-child determinants. The high proportion of postnatal diagnoses, together with young maternal age and lack of prenatal screening, shows that shortcomings in primary care and prenatal follow-up have significant effects on neonatal morbidity and mortality. These data reinforce the need to expand access to prenatal screening and fetal echocardiography in settings of greater socioeconomic vulnerability.

Conclusion

This systematic review synthesized relevant evidence on epidemiological factors associated with congenital heart disease and their relationship with mortality. Although only three articles met the inclusion criteria, the studies showed consistent results, particularly regarding the influence of heart disease type, age at diagnosis, and structural conditions of care as critical determinants of clinical outcomes. The recurrence of these findings in different settings reinforces the robustness of the conclusions despite the small number of available studies.

Overall, the clinical profile outlined by the studies indicates that patients hospitalized with congenital heart disease are predominantly young children, frequently diagnosed after birth and often from regions with limited access to specialized centers. Acyanotic heart diseases are more common, although cyanotic forms are strongly associated with a higher risk of complications and death. Associated conditions, such as malnutrition and genetic syndromes, especially Down syndrome, increase these patients' vulnerability. This picture reinforces the importance of early diagnosis, appropriate nutritional monitoring, and expanded access to specialized services as fundamental strategies for reducing morbidity and mortality associated with congenital heart disease.

The three studies confirm that cyanotic congenital heart diseases are strongly associated with higher mortality than acyanotic forms. This consistency across different settings and populations underscores the severity of cyanotic heart disease and highlights early diagnosis, appropriate risk stratification, and timely access to specialized treatment as fundamental strategies for reducing lethality in these patients.

One limitation of this review was the scarcity of publications meeting the established methodological criteria, which restricted the breadth of the analysis. This limitation also demonstrates the need for greater investment in epidemiological research on congenital heart disease, particularly in Latin American countries, where systematic data and multicenter studies remain scarce.

Despite this limitation, the findings make relevant contributions to clinical practice and public health. Identifying cyanotic heart disease, neonatal age, and failures in prenatal screening as central determinants of mortality reinforces the urgency of early-diagnosis strategies, improved hospital infrastructure, and stronger maternal and child health care. This study therefore not only compiles existing evidence but also creates opportunities for new investigations that can expand knowledge and support more effective care policies for patients with congenital heart disease.

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