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Original Article

Epidemiology and Outcomes of Patients with Adult Congenital Heart Disease in Queensland Intensive Care Units: A Multicentre Retrospective Observational Study

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Objectives: To describe the epidemiology, clinical characteristics, and outcomes of adult congenital heart disease (ACHD) patients admitted to intensive care units (ICUs) across Queensland, Australia.
Design: A multicenter, retrospective cohort study.
Setting: Twelve adult ICUs across Queensland, including tertiary referral and regional centers, from January 1, 2015, to December 31, 2021.
Participants: Adults (≥18 years) with ACHD.
Interventions: No interventions.
Measurements and Main Results: ACHD cases were stratified by lesion complexity and admission type (medical vs surgical). Outcomes included ICU and hospital length of stay and 30-day and 1-year mortality.

Of 89,184 ICU admissions, 1,870 (2.1%) involved ACHD. The most common diagnoses were valvular (57.9%) and septal (31.0%) malformations. Lesion complexity was classified as simple (1,543/1,870; 82.5%), moderate (220/1,870; 11.8%), and complex (60/1,870; 3.2%). Medical patients (253/1,870; 13.5%) had greater illness severity, more frequent use of renal replacement therapy and ECMO, and longer ICU (3 [2-6] *v* 2 [2-6] days; $p < 0.001$) and hospital length of stay: 18 [10-33] *v* 8 [6-13] days; $p < 0.001$) when compared to surgical patients. Mortality was significantly higher in medical admissions (30 day: 34/253: 13.4%; 1 year: 50/253: 19.8%) than in surgical (30 day: 20/1,617: 1.2%; 1 year: 42/1617: 2.6%; $p < 0.001$). One-year mortality was also higher in patients with complex lesions (11/60; 18.3%) versus simple (67/1,543; 4.3%).
Conclusions: ACHD patients are an uncommon but important ICU population. Outcomes vary significantly by admission type and lesion complexity. Emergency and medical admissions are associated with disproportionately high mortality compared to elective surgical admissions and should prompt early escalation of care.

Key Words: congenital heart disease; intensive care; critical care; epidemiology; adult congenital heart disease; heart defects, congenital

Introduction

Congenital heart disease (CHD) affects 9 per 1,000 live births.[1] Due to advances in care, over 85% of individuals with CHD in high-income countries now survive into adulthood,[2] contributing to a growing population with adult congenital heart disease (ACHD). In Australia, an estimated 26,000-32,000 adults were living with ACHD in 2011, increasing by approximately 5% annually.[3] One cohort estimated that up to 16% of these patients may require intensive care unit (ICU) admission by age 40.[4]

As this population grows older, the need for ICU care has become increasingly recognized, particularly for patients with complex anatomy, perioperative risk, or complications such as arrhythmias or heart failure.[5-8] However, the broader ICU epidemiology and outcomes of ACHD patients remain incompletely understood.

Two recent North American studies have contributed significantly to the field. Keane et al.[9] and Burchill et al.[10] analyzed adult ACHD admissions to dedicated cardiac intensive care units (CICUs). These cohorts identified high rates of medical admissions for heart failure, atrial arrhythmias, and sepsis, with in-hospital mortality rates of 12.7% and 8.7%, respectively. However, both studies were limited to cardiac ICUs, which in the North American setting typically exclude postoperative and elective surgical patients, who are managed in separate cardiothoracic or surgical ICUs.

In contrast, Australian ICUs are general in structure and include all types of ACHD admissions, medical and surgical, providing a broader view of ICU burden and outcomes in this population.

Existing ICU-focused studies outside of these North American datasets remain small, single-center, or restricted to specific complications such as acute kidney injury or unplanned ICU admissions.[11,12] Thus, the generalizable epidemiology and outcomes of ACHD patients across a statewide ICU network have not been well described.

We therefore conducted a multicentre retrospective cohort study of ACHD patients admitted to ICUs across Queensland, Australia. Our objectives were to describe patient demographics, comorbidities, interventions, and outcomes. These findings aim to support better-informed ICU management strategies for this complex group.

Methods

Study Design and Population

We conducted a multicenter retrospective cohort study using data from 12 intensivist-led ICUs across Queensland.

The state of Queensland spans 1.7 million km^2, has a population of 5.5 million, and includes 106 public hospitals.[13,14] The participating ICUs include all publicly funded cardiothoracic care centers in Queensland and account for approximately 75% of the state's adult ICU beds.[15] The 12 sites include five tertiary, four regional, and three outer metropolitan hospitals. The study period spans from January 1, 2015, to December 31, 2021. Adult patients (≥18 years) with ACHD admitted to any of the 12 ICUs during the study period were included.

Data Sources

Data were extracted from electronic medical records and ICU clinical databases. Demographics, diagnoses, illness severity scores, and outcomes were sourced from the Australia and New Zealand Intensive Care Society (ANZICS) Centre for Outcome and Resource Evaluation (CORE) Adult Patient Database.[16] Diagnoses were coded using the 10th Revision of the International Classification of Diseases (ICD-10).[17] Long-term mortality was obtained from the Queensland Births, Deaths, and Marriages Registry.[18] Clinical data and interventions (e.g., mechanical ventilation, vasoactive medications, and renal replacement therapy) were retrieved from the clinical information system eCritical MetaVision (iMDsoft, Boston, MA, USA).

Cohort Definition and Stratification

ACHD patients were identified using ICD-10-AM codes Q20–Q26 (Supplementary Table 1). Lesions were classified as simple, moderate, or complex based on the 2018 American Heart Association / American College of Cardiology guidelines.[19] Where classification was undefined, consensus review was used. Patients with heterogeneous lesion complexity were labeled as "diverse" and excluded from subgroup analyses.

We stratified by age (≤ 40 and > 40 years), consistent with prior studies,[11] to account for the increasing prevalence of acquired disease with age. A separate subgroup analysis was performed for the bicuspid aortic valve (BAV), anticipated to represent a large portion of the cohort. ICU admissions were categorized by admission type.

Variables Analyzed

We analyzed demographics, admission characteristics, illness severity, interventions, and outcomes. Key variables included age, sex, admission type, organ support (ventilation, vasoactive agents, renal replacement therapy), ICU and hospital length of stay, and mortality at multiple time points (ICU, hospital, 30 days, 90 days, and 1 year).

We also assessed Australian and New Zealand Risk of Death (ANZROD) risk factors, derived from Acute Physiology and Chronic Health Evaluation (APACHE) II, which identify patients with severe chronic conditions such as New York Heart Association (NYHA) class IV heart failure, end-stage renal disease, advanced malignancy, and immunosuppression.

Statistical Analysis

Descriptive statistics were used. Categorical variables are presented as counts and percentages; continuous variables as means (standard deviations; SD) or medians (IQRs) depending on the distribution. Group comparisons used the Wilcoxon rank-sum test (non-parametric) or Student's t-test (parametric) and the chi-square or Fisher's exact test for categorical variables. A two-tailed p value <0.05 was considered significant.

Ethical Considerations

Ethical approval was obtained from the Metro South Hospital and Health Service Human Research Ethics Committee (HREC/2022/QMS/82024).

Results

Prevalence and Demographics

Of 89,184 individual patients admitted to the ICU during the study period, 1,870 (2.1%) had ACHD. The mean age was 53.8 years (SD 16.5), with 35% (656/1,870) being female. The most common APACHE II risk factor was cardiac disease (99/1,870; 5.3%). Full demographic and comorbidity data are in Tables 1 and 2.

ACHD Diagnoses and Lesion Complexity

The most common congenital diagnoses were aortic or mitral valve malformations (1,082/1,870; 57.9%) and cardiac septal defects (580/1,870; 31.0%). BAV was the most frequent specific lesion, present in 1,031/1,870 (55.1%). The patient numbers in each ICD-10-AM category are presented in Table 3.

Most had simple lesions (1,543/1,870; 82.5%), followed by moderate (220/1,870; 11.8%) and complex (60/1,870; 3.2%). In addition, a smaller number of patients (47/1,870; 2.5%) had diverse lesions and were excluded from sub-analyses. The proportion of lesion complexity remained stable over time (Fig. 1; Supplementary Table 2).

ICU Admissions: Sources and Types

Most ICU admissions originated from the operating theatre (1,614/1,870; 86.3%), followed by wards (141/1,870; 7.5%), emergency departments (62/1,870; 3.3%), and interhospital transfers direct to ICU (27/1,870; 1.4%). Elective admissions were most common (1,510/1,870; 80.8%), particularly in patients with simple lesions (1,267/1,543; 82.1%). Emergency admissions were more frequent in those with moderate (41/220; 18.6%) or complex (27/60; 45.0%) lesions. Medical admissions (253/1,870; 13.5%) were more frequent among moderate (28/220; 12.7%) or complex lesions (19/60; 31.7%). Post cardiac arrest cases accounted for 51 of 1,870 (2.7%) admissions and trauma for 9 of 1,870 (0.5%) admissions.

Table 1
Clinical Characteristics by Lesion Complexity*

Characteristic	Lesion Complexity			
	Simple n =1543	Moderate n = 220	Complex n = 60	Overall** N = 1870
Demographics		**n (%) or mean (SD)**		
Age (years)	56.1 (15.3)	43.2 (16.9)	32.5 (12.8)	53.8 (16.5)
Female sex	495 (32.1%)	103 (46.8%)	33 (55.0%)	656 (35.1%)
BMI (kg.m^{-2})	28.8 (5.9)	26.9 (5.9)	26.5 (7.8)	28.5 (6.0)
Type of admission		**n (%)**		
Surgical–cardiac	1267 (82.1%)	180 (81.2%)	34 (56.7%)	1512 (80.9%)
Surgical–non-cardiac	41 (2.7%)	1 (0.5%)	4 (6.7%)	45 (2.4%)
Medical	195 (12.6%)	28 (12.7%)	19 (31.7%)	253 (13.5%)
Post arrest	33 (2.1%)	11 (5.0%)	3 (5.0%)	51 (2.7%)
Post trauma	7 (0.5%)	0 (0.0%)	0 (0.0%)	9 (0.5%)
Total	1543	220	60	1870
Elective or emergency admission		**n (%)**		
Elective	1267 (82.1%)	179 (81.4%)	33 (55.0%)	1510 (80.8%)
Emergency	276 (17.9%)	41 (18.6%)	27 (45.0%)	360 (19.2%)
ICU interventions		**n (%)**		
Ventilation–invasive	1245 (80.7%)	173 (78.6%)	44 (73.3%)	1495 (79.9%)
Ventilation–NIV	15 (1.0%)	2 (0.9%)	1 (1.7%)	20 (1.1%)
Inotropes/vasopressors	1032 (66.9%)	139 (63.2%)	31 (51.7%)	1226 (65.6%)
RRT	31 (2.0%)	5 (2.3%)	2 (3.3%)	40 (2.1%)
Tracheostomy†	18 (1.8%)	0 (0.0%)	0 (0.0%)	20 (1.1%)
ECMO‡	14 (1.4%)	2 (1.5%)	3 (7.0%)	19 (1.0%)
Length of stay (days)		**Median (IQR)**		
ICU	2 (2–4)	2 (2-4)	2 (2- 4)	2 (2-4)
Hospital	9 (6-15)	8 (6-14)	9 (7-25)	9 (6-15)
Mortality		**n (%)**		
ICU	31 (2.0%)	4 (1.8%)	5 (8.3%)	42 (2.3%)
Hospital	47 (3.1%)	5 (2.3%)	9 (15.0%)	63 (3.4%)
30 days	39 (2.5%)	4 (1.8%)	9 (15.0%)	54 (2.9%)
90 days	49 (3.2%)	5 (2.3%)	9 (15.0%)	65 (3.5%)
1 year	67 (4.3%)	8 (3.6%)	11 (18.3%)	88 (4.7%)

* Lesion complexity, stratified according to 2018 ACC/AHA guidelines.[19,26,27]
** Includes diverse group (n=47, not tabulated) therefore total is 1870.
† 665 missing records. Percentage is calculated using N = 1870.
‡ 667 missing records. Percentage is calculated using N = 1870.
Abbreviations: ECMO, extracorporeal membrane oxygenation; RRT, renal replacement therapy.

Among medical admissions, the most common diagnostic categories were cardiovascular (76/250; 30.4%), sepsis (36/250; 14.4%) and cardiac arrest (29/250; 11.6%). Specific medical diagnoses included cardiogenic shock (27/250; 10.8%), arrhythmia (28/250; 11.2%), sepsis with shock (23/250; 9.2%), and sepsis without shock (13/250; 5.2%).

Among surgical admissions, cardiac surgery predominated, accounting for 1,146/1,620 (70.7%) of cases. The most frequent procedure was isolated valvular surgery (917/1,620; 56.6%), followed by combined CABG and valve surgery (167/1,620; 10.3%).

Representative diagnoses are presented in Supplementary Tables 3a and 3b.

Hospital and ICU Outcomes

Almost all patients (1,853/1,870; 99.1%) received care in tertiary ICUs. Most (1,559/1,870; 83.3%) were admitted from home (either via an elective admission pathway or an emergency presentation from the community), while 272/1,870 (14.6%) were transferred from another hospital (but not necessarily directly admitted to the ICU).

Median ICU LOS was 2 days (IQR 2-4), and hospital LOS was 9 days (IQR 6-15). ICU stays exceeded 7 days in only 165 of 1,870 (8.8%) of admissions; stays over 14 days occurred in 71 of 1,870 (3.8%) admissions. ICU and hospital LOS were similar across lesion complexity groups.

Interventions and Illness Severity

Interventions and illness severity are listed in Table 5**.** Most patients require organ support during their ICU stay. Invasive ventilation was used in 79.9%, vasoactive medications in 65.6%, and renal replacement therapy in 2.1%. Extracorporeal membrane oxygenation (ECMO) (1.0%) and tracheostomy (1.1%) were infrequent.

Illness severity scores indicated moderate acute illness overall. The median APACHE II score was 13 (IQR 9-16) and the Sequential Organ Failure Assessment (SOFA) score was 5 (IQR 4-7). The median ANZROD was 0.8% (IQR 0.5%-2.1%).

Table 2
Additional Clinical Characteristics of ACHD Patients in the ICU

Characteristic	n (%)
APACHE-II risk factors	
Cardiac disease	99 (5.3%)
Respiratory disease	87 (4.6%)
Immune suppression	57 (3.1%)
Diabetes	32 (1.7%)
Cirrhosis	16 (0.9%)
Hepatic failure	0 (0%)
Metastatic cancer	17 (0.9%)
Hematological malignancy	5 (0.3%)
End-stage renal disease	10 (0.5%)
AIDS	1 (0.1%)
Frailty category	
None or minimal (CFS ≤4)	796 (42.6%)
Mild to moderate (CFS 5 and 6)	54 (2.8%)
Severe (CFS ≥ 7)	3 (0.2%)
Not recorded	1017 (54.4%)
ICU source of admission	
Operating theatre	1614 (86.3%)
Ward	141 (7.5%)
Emergency department	62 (3.3%)
Other hospital	27 (1.5%)
Not recorded	26 (1.4%)
Hospital source of admission	
Home	1559 (83.3%)
Other hospital	272 (14.6%)
High care facility	2 (0.1%)
Not recorded	37 (2.0%)
ICU type	
Tertiary	1853 (99.1%)
Metropolitan	9 (0.5%)
Regional	8 (0.4%)
Discharge destination	
Home	1636 (87.4%)
Other hospital−acute	76 (4.1%)
Chronic care	60 (3.2%)
Rehabilitation	27 (1.4%)
Other hospital−ICU	6 (0.3%)
Mental health	1 (0.1%)
Not recorded	1 (0.1%)
Died	63 (3.4%)

Abbreviations: CFS, Clinical Frailty Scale; ICU, intensive care unit.

APACHE III scores were highest in post cardiac arrest patients (median 74, IQR 41-95) and the lowest in elective surgical patients (median 44, IQR 35-54; $p < 0.001$). Medical patients (58, IQR 43-74) and emergency admissions (56, IQR 41.5-73) had higher illness severity compared to elective surgical cases, while post-trauma patients had moderate severity (43, IQR 32-58). Lesion complexity did not significantly impact APACHE III scores.

Clinical Frailty Scale (CFS) was documented only in around 45 % of the cohort (853/1,870) with fewer than half of the overall cohort categorized as not frail (Categories: Very Fit to Vulnerable; CFS ≤4; 796/1,870; 42.6%). However, among those who did have a CFS recorded, the majority were not frail (CFS ≤4: 796/853(93.3%). Severe frailty (CFS 7) was rare (3/1,870; 0.2%).

Mortality

In-hospital mortality was 3.4% (63/1,870), with ICU mortality at 2.3% (42/1,870). Mortality at 30, 90, and 365 days was 2.9% (54/1,870), 3.5% (65/1,870), and 4.7% (88/1,870). Among the 153 patients who had a known time-to-death, median survival post-ICU was 194 days (IQR 15-1,125). Post cardiac arrest patients had the highest mortality, reaching 11/27 (40.7%) at 1 year, per Table 4.

Mortality increased with lesion complexity. ICU mortality was higher for complex (5/60; 8.3%), than for simple (31/1,543; 2.0%) or moderate (4/220; 1.8%) lesions ($p < 0.001$). This pattern persisted at 1 year with mortality highest in complex lesions (18.3%; 11/60), per Table 1.

Comparison of Medical and Surgical Admissions

Patients were categorized as medical (including cardiac arrest) or surgical (including trauma, elective, and emergency surgery) to enable subgroup comparison. Demographics were similar between the two groups; however, medical patients had significantly longer ICU and hospital lengths of stay.

Table 3
Patient Number and Lesion Complexity* by ICD Category**

ICD Category - Congenital malformations of	Simple	Moderate	Complex	Diverse	Total (n (%))
Cardiac chambers	0	0	42	10	52 (2.8%)
Cardiac septa	496	82	0	2	580 (31.0%)
PV and TV	0	19	5	0	24 (1.3%)
AV and MV	1,031	47	3	1	1,082 (57.9%)
Heart	3	37	4	28	72 (3.9%)
Great arteries	8	20	6	6	40 (2.1%)
Great veins	5	15	0	0	20 (1.1%)
Total	1543 (82.5%)	220 (11.8%)	60 (3.2%)	47 (2.5%)	1,870 (100.0%)

* Lesion complexity, stratified according to 2018 ACC/AHA guidelines.[19,26,27]
** ICD categories correspond to diagnostic codes from the 10th edition of the International Classification of Diseases (ICD-10).[17]
Abbreviations: AV, aortic valve; MV, mitral valve; PV, pulmonary valve; TV, tricuspid valve.

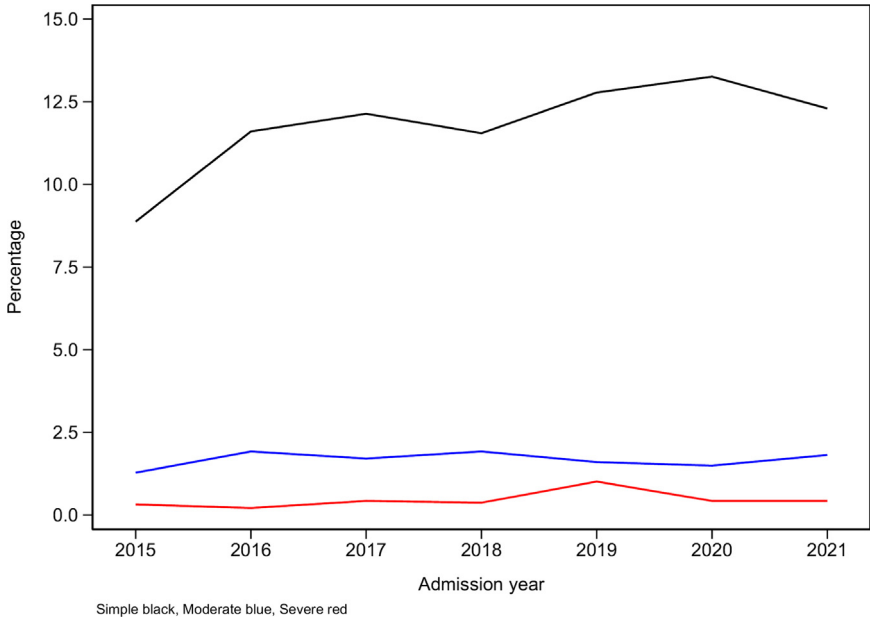

Fig. 1. Trends in lesion complexity over time. The percentage distribution by lesion complexity in ACHD ICU admissions from 2015 to 2021. Over time the distribution remained stable, with simple lesions consistently comprising the majority of cases.

Median ICU LOS was 2 days (IQR 2-3) for surgical patients versus 3 days (IQR 2-6) for medical patients ($p < 0.001$), and median hospital LOS was 8 days (IQR 6-16) *v* 18 days (IQR 10-33), respectively ($p < 0.001$).

Medical patients had significantly higher illness severity scores. The median APACHE II score was 17 (IQR 12-22) in medical patients versus 12 (IQR 9-15) in surgical patients ($p < 0.001$), and the APACHE III score was 58 (IQR 43-74) *v* 44 (IQR 35-54), respectively ($p < 0.001$). While SOFA scores were similar (5 [IQR 3-7] *v* 5 [IQR 4-7]; $p = 0.992$), predicted mortality was higher among medical patients. The ANZROD was 7.6% for medical patients (IQR 2.5-23.1%) versus 0.7% for surgical patients (IQR 0.4-1.3%; $p < 0.001$).

Medical patients were less likely to receive invasive mechanical ventilation (160/253; 63.2%) compared to surgical patients (1,335/1,617; 82.6%; $p < 0.001$), although use of inotropes or vasopressors was similar (156/253; 61.7% *v* 1,070/1,617; 66.2%; $p = 0.176$). Renal replacement therapy was more common in the medical cohort (30/253; 11.9%) compared to surgical (81/1,617; 5.0%; $p < 0.001$), as were tracheostomy (9/253; 3.5% *v* 11/1,617; 0.7%; $p < 0.001$) and ECMO (11/253; 4.3% *v* 8/1,617; 0.5%; $p < 0.001$).

Interhospital transfers were more frequent in medical patients (80/253; 31.6%) than in surgical patients (192/1,617; 11.9%; $p < 0.001$). Medical patients were also more commonly managed in regional (13/253; 5.1%) or outer metropolitan centers (8/253; 3.2%) compared to surgical patients (3/1,617; 0.2% and 1/1,617; 0.1%, respectively; $p < 0.001$ for both).

Mortality was significantly higher among medical patients across all time points: ICU mortality (27/253; 10.7% *v* 15/1,617; 0.9%), hospital mortality (38/253; 15.0% *v* 25/1,617; 1.5%), and 1-year mortality (50/253; 19.8% *v* 42/1,617; 2.6%), all $p < 0.001$. Full comparative data between the medical and surgical cohorts are provided in Table 6.

Age-based Subgroup Analysis

Patients ≤40 years were more likely to have moderate or complex lesions, while those older than 40 years predominantly had simple lesions. Despite this, mortality at all time points was similar between age groups (Supplementary Table 4).

Table 4
Mortality Data by Type of Admission

Admission type	Number	ICU mortality	Hospital mortality	30-day mortality*	90-day mortality	1-year mortality
Operative (cardiac)	1512	9 (0.6%)	13 (0.9%)	10 (0.7%)	15 (1.0%)	25 (1.7%)
Operative (non-cardiac)	69	0 (0.0%)	3 (4.3%)	2 (2.9%)	3 (4.3%)	5 (7.2%)
Medical	253	27 (10.7%)	38 (15.0%)	34 (13.4%)	38 (15.0%)	46 (18.2%)
Trauma	9	0 (0.0%)	0 (0.0%)	1 (11.1%)	1 (11.1%)	1 (11.1%)
Post cardiac arrest	27	6 (22.2%)	9 (33.3%)	7 (25.9%)	8 (29.6%)	11 (40.7%)
Total	1870	42	63	54	65	88

* The 30-day mortality less than hospital mortality due to late (>30 days) inpatient deaths.

Table 5
Illness Severity and ICU Interventions and Complications

Characteristic	Median (IQR), n(%)
Severity of illness	
APACHE* II	13 (9-16)
APACHE III	46 (35-57)
SOFA**	5 (4-7)
APACHE III risk of death	0.033 (0.017-0.068)
ANZROD†	0.008 (0.005-0.021)
ICU interventions	
Ventilation−invasive	1495 (79.9%)
Ventilation−non-invasive	20 (1.1%)
Inotropes/vasopressors	1226 (65.6%)
Renal Replacement Therapy	40 (2.1%)
Tracheostomy ‡	20 (1.1%)
ECMO§	19 (1.0%)

* Acute Physiology and Chronic Health Evaluation score; estimates disease severity and predicts in-hospital mortality based on physiological variables and chronic health conditions.
** Sequential Organ Failure Assessment; used to assess organ dysfunction and predict ICU outcomes.
† Australian and New Zealand Risk of Death score; a risk-adjusted mortality model developed for Australian and New Zealand ICUs, incorporating patients' demographics, diagnosis, and physiological parameters to estimate the probability of death.
‡ 665 missing data. Percentage is calculated using N = 1870.
§ 667 missing data. Percentage is calculated using N = 1870.

Bicuspid Aortic Valve Subgroup Analysis

BAV patients were older (mean 57.7 (SD 13.8) *v* 49.0 years (SD 18.3); p <0 .001) and more often male (75.8% *v* 51.6%; p < 0.001). Illness severity and lengths of stay were similar. Mortality was significantly lower in BAV patients at all time points (e.g., 30 day: 1.3% *v* 4.9%; p < 0.001). Full details are in Supplementary Table 5.

Discussion

Key Findings

This multicenter study provides a comprehensive overview of ACHD patients requiring ICU admission. ACHD accounted for 2.1% of ICU admissions and was dominated by simple lesions, particularly BAV. Most admissions were elective cardiac surgical cases with low short-term mortality, while emergency and medical admissions had significantly higher mortality. Lesion complexity was not strongly associated with ICU outcomes but was linked to higher 1-year mortality. Complexity proportions of ACHD lesions remained stable over the years, likely due to the relatively short study period, which may have limited the ability to observe any differences associated with changing pediatric management.

Medical presentations—typically cardiogenic shock, arrhythmias, cardiac arrest, or sepsis—were more frequent in patients with moderate or complex lesions. These patients had higher illness severity scores, were more likely to require renal replacement therapy, tracheostomy, and ECMO, and had higher predicted and actual mortality. These patients often present to non-tertiary centers, where timely recognition and transfer are critical. Australasian-trained intensivists, who all undergo cardiac ICU training, are well equipped to stabilize these complex cases, underscoring their pivotal role in early management and coordination of care.

ACHD ICU care is highly centralized, with 99.1% of admissions in tertiary ICUs. Patients often travel to these centers for surgery and may re-present to regional hospitals with medical problems. This highlights the need for well-resourced regional centers to manage these patients initially when they present emergently.

Comparison with Existing Literature

The 2.1% prevalence of ACHD in our ICU cohort aligns with international trends, reflecting improved survival rates of children with congenital heart disease into adulthood.[20,21] The mean patient age of 53.8 years underscores this demographic shift driven by advances in pediatric treatment.[22,23]

Our inclusion of older patients undergoing elective surgery, particularly for BAV, explains the predominance of simple lesions and older age profile compared to similar studies.[11] Prior work has also shown BAV surgery to occur at a median age of 62 years,[24] which is consistent with our BAV subgroup findings.

Elective cardiac surgery was the leading cause of ICU admission, as in other studies.[4] These patients had low ICU and hospital mortality, contrasting with the medical admissions who, though fewer in number, had more complex lesions and higher mortality, involving diagnoses such as arrhythmias, cardiogenic shock, and septic shock. This supports previous findings that unplanned ACHD ICU admissions carry a worse prognosis.

In our cohort, the most common ACHD diagnoses were congenital malformations of the aortic or mitral valves (57.9%) and cardiac septa (31.0%), aligning with prior studies indicating the high prevalence of congenital aortic valve abnormalities in ACHD populations.[20,21] BAV was the most frequently identified defect, present in 55.1% of cases, reinforcing its prominence as a major contributor to ACHD ICU admissions.[19,22]

Interestingly, patients with moderate lesions had lower ICU and in-hospital mortality than those with simple lesions.

This observation should be interpreted cautiously, given the low number of deaths in the moderate lesion complexity group. Furthermore, the much smaller cohort size for the moderate group in comparison to the simple lesions group makes the mortality rates disproportionately sensitive to minor changes in absolute numbers.

No significant mortality difference was observed between younger and older ACHD patients. This may be due to a balance between higher congenital disease severity in younger patients and the increasing burden of acquired comorbidities in older patients.

Illness severity scores confirmed a wide acuity range. APACHE III scores were highest in cardiac arrest patients and lowest in elective cases. Lesion complexity did not correlate with APACHE scores. Still, it was higher in medical presentations,

Table 6
Clinical characteristics stratified by medical and surgical admissions

Characteristic	Surgical		Medical	p-Value
Number	N = 1617		N = 253	
Demographics		**n (%) or Mean (SD)**		
Age (years)	54.0 (16.5)		52.3 (16.8)	0.119
Female sex	578 (35.7%)		78 (30.8%)	0.137
BMI (kg/m)	28.6 (6.0)		27.8 (6.1)	0.060
Length of stay (days)		**Median (IQR)**		
ICU	2 (2-3)		3 (2-6)	< 0.001
Hospital	8 (6-13)		18 (10-33)	< 0.001
Severity of illness		**Median (IQR)**		
APACHE* II	12 (9-15)		17 (12-22)	< 0.001
APACHE III	44 (35-54)		58 (43-74)	< 0.001
SOFA**	5 (4-7)		5 (3-7)	0.992
ANZROD†	0.007 (0.004-0.013)		0.076 (0.025-0.231)	< 0.001
Interventions		**n (%)**		
Ventilation−invasive	1335 (82.6%)		160 (63.2%)	< 0.001
Ventilation−NIV	17 (1.1%)		3 (1.2%)	0.745
Inotropes/vasopressors	1070 (66.2%)		156 (61.7%)	0.176
RRT	81 (5.0%)		30 (11.9%)	< 0.001
Tracheostomy ‡	11 (0.7%)		9 (3.5%)	< 0.001
ECMO §	8 (0.5%)		11 (4.3%)	< 0.001
Admission source		**n (%)**		
Home	1417 (87.6%)		142 (56.1%)	< 0.001
Other hospital	192 (11.9%)		80 (31.6%)	
High care facility	1 (0.1%)		1 (0.4%)	
Not recorded	7 (0.4%)		30 (11.9%)	(N/A)
ICU type		**n (%)**		
Tertiary	1613 (99.7%)		232 (91.7%)	< 0.001
Regional	3 (0.2%)		13 (5.1%)	
Outer metropolitan	1 (0.1%)		8 (3.2%)	
Discharge destination		**n (%)**		
Died	25 (1.5%)		38 (15.1%)	< 0.001
Home	1486 (91.9%)		150 (59.3%)	
Chronic care	39 (2.4%)		21 (8.3%)	
Other ICU	3 (0.2%)		3 (1.2%)	
Other hospital (not ICU)	47 (2.9%)		29 (11.5%)	
Rehabilitation facility	16 (1.0%)		11 (4.4%)	
Mental health facility	1 (0.1%)		0 (0.0%)	
Not recorded	0 (0.0%)		1 (0.4%)	(N/A)
Mortality		**n (%)**		
ICU	15 (0.9%)		27 (10.7%)	< 0.001
Hospital	25 (1.5%)		38 (15.0%)	< 0.001
30 days	20 (1.2%)		34 (13.4%)	< 0.001
90 days	27 (1.7%)		38 (15.0%)	< 0.001
1 year	42 (2.6%)		50 (19.8%)	< 0.001

* Acute Physiology and Chronic Health Evaluation score; estimates disease severity and predicts in-hospital mortality based on physiological variables and chronic health conditions.
** Sequential Organ Failure Assessment; used to assess organ dysfunction and predict ICU outcomes.
† Australian and New Zealand Risk of Death score; a risk-adjusted mortality model developed for Australian and New Zealand ICUs, incorporating patients' demographics, diagnosis, and physiological parameters to estimate the probability of death.
‡ 665 missing data. Percentage is calculated using 1617/253 denominators.
§ 667 missing data. Percentage is calculated using 1617/253 denominators.

suggesting that acute illness severity is driven more by the nature of the presentation than by congenital anatomy.

Despite the low short-term mortality observed in our overall cohort, ACHD patients are known to experience excess mortality over the longer term compared to the general population, with chronic heart failure and sudden cardiac death being the leading causes of late mortality.[25] Our findings support the critical role of intensivists in recognizing, stabilizing, and triaging ACHD patients, especially in non-specialist centers. The diversity of ICU presentations and outcomes highlights the need for tailored risk stratification and post-ICU follow-up.

Implications of Study Findings

This study highlights the heterogeneity of ACHD patients in the ICU and the need for tailored approaches to care.

Emergency and medical admissions, though fewer in number, were associated with disproportionately high mortality and should prompt early escalation and consideration for transfer to specialized centers. It would be prudent for ICU teams to develop pathways for stabilizing and managing critically ill ACHD patients (especially those with complex anatomy) in non-specialist centers, including identification of those that can be managed locally where possible, given that these patients may present to non-specialist hospitals.

Multidisciplinary care involving intensivists, cardiologists, cardiac surgeons, and specialized ACHD teams is essential, particularly for patients with complex lesions who require advanced resources and expertise. The reliance on tertiary ICUs suggests that robust regional referral networks and retrieval services are vital to ensure that ACHD patients receive appropriate care. This is especially critical in regions with widely dispersed, decentralized populations such as the state of Queensland, where this study was conducted. Strengthening regional ICU capabilities, telemedicine support, and retrieval pathways may help optimize outcomes for this highly specialized patient population.

Strengths and Limitations

This is the largest study to date examining ACHD admissions to the ICU, with near-population coverage in a large geographic region. Its broad inclusion criteria and multicenter design enhance generalizability to other high-income health systems with centralized ACHD services.

Several limitations merit consideration. First, the retrospective design and reliance on administrative coding, which may introduce misclassification. Some variables, such as frailty, were not consistently recorded, and functional outcomes beyond 1-year mortality were not assessed. Subgroup analyses, particularly for complex lesions, were limited by small sample sizes.

Importantly, we did not include physiologic classification (e.g., AHA/ACC stages A-D), which reflects key features such as exercise capacity, heart failure symptoms, cyanosis, and arrhythmia burden. Lesion complexity alone may have limited prognostic value and can obscure significant heterogeneity in clinical risk within anatomical subgroups. For example, patients with repaired tetralogy of Fallot and preserved physiology may fare better than those with Eisenmenger syndrome, despite similar or lower anatomical classification.

Additionally, while illness severity was assessed using APACHE III and SOFA scores, these tools were not developed or validated for congenital heart disease populations. They may not account for ACHD-specific factors such as baseline hypoxemia or single-ventricle physiology, which may explain some of the observed disconnect between anatomical complexity and illness severity scores or outcomes.

Future studies should prospectively evaluate high-risk subgroups and consider incorporating physiologic classification to enhance risk stratification and improve predictive accuracy in this heterogeneous population. Additional focus is needed on the role of comorbidities, immunosuppression, transplant status, and specific acute presentations such as septic shock. Long-term outcomes, including functional status and quality of life, will also be important to better understand the impact of critical illness on this growing population.

Conclusion

This multicenter study provides the most comprehensive overview to date of ACHD patients in ICU. Although they represent a small proportion of admissions, they require highly individualized care. Short-term outcomes are generally favorable, but complexity and unplanned admissions are associated with higher risk. As the ACHD population grows, ICU services must evolve to deliver coordinated, specialized, and longitudinal care for this unique group.

Data Availability Statement

Data cannot be shared publicly due to institutional ethics, privacy, and confidentiality regulations. Data released for research under Sect. 280 of the Public Health Act 2005 requires an application to the Director-General of Queensland Health (PHA@health.qld.gov.au).

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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& editing, Validation, Supervision, Methodology, Data curation, Conceptualization.

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Supplementary materials

Supplementary material associated with this article can be found in the online version at doi:10.1053/j.jvca.2025.08.049.

Appendices

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Appendix 2: Tables and Figures for the Main Manuscript

Table 1: Clinical Characteristics by Lesion Complexity*

Table 2: Additional Clinical characteristics of ACHD patients in the ICU

Table 3: Patient Number and Lesion Complexity* by ICD Category**

Table 4: Mortality Data by Type of Admission

Table 5: Illness severity, ICU interventions and complications

Table 6: Clinical characteristics stratified by medical and surgical admissions.

Figure 1: Trends in Lesion Complexity over Time

Appendix 3: Tables for the Supplement

Supplementary Table 1: ICD codes for congenital heart disease

Supplementary Table 2: Lesion Complexity over Time

Supplementary Table 3a: Diagnostic categories are stratified by lesion complexity. *Data presented as Non-Operative and Operative Diagnostic Subcategories*

Supplementary Table 3b: Diagnostic categories are stratified by lesion complexity.

Data presented as a Major Non-Operative And Operative Diagnostic Category.

Supplementary Table 4: Mortality, Complexity, and ICD Data by Age Category

Supplementary Table 5:

Bicuspid Aortic Valve *v* non-Bicuspid Aortic Valve: Demographic, Illness Severity, Length of Stay and Mortality data.

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