Survival outcomes for congenital heart disease from Southern Malaysia: results from a congenital heart disease registry

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ABSTRACT

Objective Limited population-based studies are available on the survival of congenital heart disease (CHD) from lower- and middle-income countries. Therefore, we evaluated the survival from birth until 15 years and associated factors for mortality.

Methods This population-based cohort study included all children with CHD registered in the Pediatric Cardiology Clinical Information System born between 2006 and 2020 in Johor, Malaysia. The mortality rate was calculated, and Cox proportional hazard regression analysis was used to determine factors associated with mortality. The Kaplan-Meier analysis was used to estimate the survival rates at 1, 5, 10 and 15 years.

Results There were 5728 patients with CHD studied, with 1543 (27%) lesions resolved spontaneously, 322 (5.6%) were treated with comfort care, 1189 (21%) required no intervention, and 2674 (47%) needed surgery or intervention. The overall mortality rate was 15%, with a median age of death of 3.7 months (IQR 0.9–9.8 months). Preoperative/intervention death was observed in 300 (11%), and 68 (3.2%) children died within 30 days of surgery or intervention. The overall estimated survival at 1, 5, 10 and 15 years was 88%, 85%, 84% and 83%, respectively. The independent factors associated with mortality were male gender, associated syndrome or extra-cardiac defect, pulmonary hypertension, antenatal diagnosis and severe lesions.

Conclusions Eight out of 10 patients with CHDs survived up to 15 years of age. However, 10% of CHDs who require intervention die before the procedure. Thus, improving congenital cardiac surgery and enhancing the overall healthcare system are crucial to improve survival.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Survival rates for children with congenital heart disease (CHD) are higher in high-income countries than in lower- and middle-income countries.

WHAT THIS STUDY ADDS

⇒ In middle-income countries, the prognosis for CHD is good, except for severe univentricular heart, non-cardiac malformations or syndromes, and pulmonary hypertension.

⇒ However, 1 out of every 10 children who require CHD intervention die before the procedure, with 1 in 2 truncus arteriosus dying before surgery.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ In addition to improving congenital cardiac surgery, enhancing the healthcare system to mitigate infections and pneumonia may decrease preoperative mortality rates and improve the survival of children with CHD.

INTRODUCTION

Congenital heart disease (CHD) is a common malformation in children, occurring in 6–9 per 1000 live births.[1] It can manifest as a severe or critical lesion that necessitates immediate intervention and poses a high risk of morbidity and mortality if not detected or treated promptly.[2] Furthermore, CHD can occur in isolation or with other anomalies, potentially leading to complications or death.

In addition to CHD severity and related medical conditions, mortality and survival rates for CHD are affected by the economic status of the country,[3–8] with high-income countries (HICs) having lower mortality and higher survival rates than lower- and middle-income countries (LMICs). Furthermore, The Global Burden of Disease Study 2017 showed a significant global disparity in CHD mortality rates.[9] HICs have shown a steady decline in CHD mortality rates, while LMICs have not made the same progress. In Malaysia, limited infrastructure and expertise pose a major challenge to paediatric cardiac services, resulting in significant deaths from critical CHD.[10] However, no study in Malaysia or other LMICs has examined the outcomes of CHD subtypes and associated mortality factors. Therefore, this study aims to investigate the overall mortality and survival of CHD and the factors associated with mortality.

METHODS

Study population

This retrospective, cohort, population-based study was conducted among children with CHD born between January 2006 and December 2020 in Johor, Malaysia. Johor, with a population of 3.4 million and a live birth of 55 000 annually, is among 14 states in Malaysia, a middle-income country. Healthcare services in Johor state are mainly provided by government hospitals, with private hospitals offering additional support. Sultanah Aminah Hospital Johor Bahru (HSAJB) is Johor's sole cardiac centre, catering to adult and paediatric cardiac services.

Due to the limited availability of specialised paediatric and congenital cardiac surgeons in Malaysia, only lesions with a lower complexity undergo interventional procedures in Johor. Major paediatric and congenital cardiac surgeries or interventions are performed at cardiac centres in Kuala Lumpur, 400 km from HSAJB, with HSAJB providing follow-up and comanagement. A dedicated paediatric palliative care team has been available since 2013, and its services are offered either at home or in the hospital, depending on the patient's needs and family preferences.

The study obtained registration with the National Medical Research Registry (ID-22-00829-3GR) and approval from the Medical Research Ethics Committee (MREC) of the Ministry of Health, Malaysia. MREC waived patient consent due to the nature of the study, which was carried out per the Helsinki Declaration guidelines.

Data source and case ascertainment

The data were retrieved from the Paediatric Cardiology Clinical Information System, a clinical registry for acquired and CHD in Johor.[11 12] CHD was defined as "a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance"[1] and was confirmed by 2D echocardiogram or other imaging modalities. CHD was classified into severe and non-severe,[13] and severe CHD was further divided into univentricular and biventricular heart (online supplemental file 1).

Asymptomatic children diagnosed with patent foramen ovale, mild pulmonary stenosis (PS), isolated right arch, mitral valve prolapse, bicuspid aortic valve, isolated dextrocardia and bilateral superior vena cava were excluded from this study.[11] Furthermore, premature infants with patent ductus arteriosus (PDA) spontaneously closed before or at 6 months of life and term infants with closure before or at 3 months were excluded.

The primary outcome measured was mortality. All mortality (all causes of death) was verified with the National Registration Department, Malaysia.

Statistical analysis

We used the Statistical Package for Social Science (SPSS) V.23 by IBM in Armonk, New York, USA, for statistical analysis. The Student's t-test was used for normally distributed continuous data, while a non-parametric test was used for non-normally distributed continuous data. For categorical variables, we used Pearson's χ^2 test. A p value of less than 0.05 was deemed statistically significant.

Kaplan-Meier analysis was used to estimate survival at 1, 5, 10 and 15 years. A log-rank test was used to compare the differences between the groups. Cox proportional hazards regression was used to identify the independent risk factors associated with death. Variables included in the study were sex, ethnicity, birth year, associated syndrome or non-cardiac malformation, pulmonary hypertension (PHT), the timing of diagnosis and the severity of the lesion. An HR was considered significant if the 95% CI excluded one.

RESULTS

There were 860 670 live births during the 15-year study period, with 5833 CHD, giving the birth prevalence of 6.8 (95% CI 6.6 to 6.9) per 1000 live births. Of the 5833 patients, 105 (1.5%) were excluded due to non-cardiac lethal congenital malformation. Of 5728 patients, 2798 (49%) were male, 4774 (83%) were term infants, and 216 (3.8%) had antenatal diagnoses. The median age of diagnosis of 5512 infants with the postnatal diagnosis was 26 days, with an IQR of 4 days to 3.9 months (online supplemental file 2). The median follow-up age was 6.2 years (IQR 2.5–10.7 years), ranging from 0 to 17 years.

Of 5728 patients, 3778 (66%) were considered non-severe CHD, 1583 (28%) were severe with biventricular heart, and 367 (6.4%) were severe univentricular heart. Table 1 shows the primary cardiac diagnosis, severity and associated non-cardiac malformation or chromosomal anomaly. Overall, 1057 (18%) were associated with syndrome or extracardiac defect, 317 (5.5%) had PHT, and 140 (2.4%) had a family history of CHD.

Of 5728 patients, 2732 (48%) required no surgery or intervention, 2674 (47%) needed surgery or intervention, and 322 (5.6%) were managed with comfort care (figure 1). Of 322 patients with comfort care, 192 (60%) were univentricular heart (online supplemental file 2), and 81 (25%) were due to medical conditions.

Mortality

The mortality rate was 15%, with significantly lower rates from 2016 to 2020 compared with 2006 to 2015 (258/2148, 12% vs 578/3580, 16%; p<0.001). The median age of death was 3.7 months (IQR 0.9–9.8 months), with 660 (79%) occurring during the first year of life. The overall neonatal death rate was 2.6 %, ranging from 2.4% to 4.8%, and infant mortality was 11%, ranging from 8.9% to 13%. Most of the death was cardiac-related (329, 40%), whereas 160 (19%) were related to infection, and 98 (12%) were due to pneumonia.

There was a significantly higher mortality rate in males than females (391/2930, 16% vs 445/2798, 13%, p=0.006), in PHT than non-PHT (110/317, 35% vs 726/5411,13%, p<0.001), in antenatal than postnatal diagnosis (79/216, 37% vs 757/5512, 14%, p<0.001) and in non-isolated CHD (275/1067, 26% vs 561/4671, 12%, p<0.001).

Table 2 shows diagnosis-specific mortality and timing of death in relation to surgery or intervention. The highest mortality was observed in infants with hypoplastic left heart syndrome (HLHS) (92%), followed by heterotaxia (70%) and truncus arteriosus (60%). Whereas for specific CHD groups, there was a significant difference in mortality among infants with univentricular hearts compared with severe biventricular and non-severe lesions (65% vs 30% vs 3.4%, p<0.001).

Of 2674 patients who needed surgery or intervention, preoperative death was observed in 300 (11%), with the majority (262, 87%) in severe CHD. The highest rate was observed in infants with truncus arteriosus (20/39, 51%), followed by interrupted aortic arch (8/23, 35%) and mitral atresia (7/21, 33%). Further analyses show the presence of associated syndrome or non-cardiac malformation (adjusted OR (aOR) 1.8, 95% CI 1.4 to 2.4, p<0.001), PHT (aOR 1.5, 95%, CI 1.0 to 2.2, p=0.04%) and severe CHD (aOR 3.3, 95% CI 2.3 to 4.7, p<0.001) were independent factors associated with preoperative death.

Of 2156 who had surgery or intervention, 68 (3.1%) died within and 145 (6.7%) died after 30 days of surgery or intervention. The highest mortality rate within 30 days of surgery or intervention was observed in infants with HLHS, with a 100% mortality rate. The highest mortality rates after 30 days of surgery or intervention were observed in infants with univentricular hearts, with mitral atresia and heterotaxia syndrome as the two highest rates.

Table 1 Primary cardiac diagnosis, severity, associated chromosomal anomaly and extracardiac defect in children with CHD

Primary cardiac diagnosis	Total	Severe lesion	Chromosomal anomaly	Extracardiac defect
	n (*%)	n (†%)	n (†%)	n (†%)
VSD	2176 (38.0)	234 (10.8)	265 (12.2)	61 (2.8)
Patent ductus arteriosus	988 (17.2)	156 (15.8)	200 (20.2)	37 (3.7)
Pulmonary stenosis	609 (10.6)	49 (8.0)	55 (9.0)	15 (2.5)
Atrial septal defect	321 (5.6)	4 (1.2)	73 (22.7)	9 (2.8)
Tetralogy of Fallot	299 (5.2)	299 (100.0)	53 (17.7)	14 (4.7)
D-transposition great arteries	139 (2.4)	139 (100.0)	2 (1.4)	4 (2.9)
Atrioventricular septal defect	134 (2.3)	128 (95.5)	97 (72.4)	1 (0.7)
Coarctation of aorta	112 (2.0)	82 (73.2)	18 (16.1)	9 (8.0)
Pulmonary atresia with VSD	109 (1.9)	109 (100.0)	20 (18.3)	5 (4.6)
Heterotaxia syndrome	94 (1.6)	94 (100.0)	3 (3.2)	2 (2.1)
Double outlet right ventricle	85 (1.5)	85 (100.0)	10 (11.8)	5 (5.9)
Hypoplastic left heart syndrome	77 (1.3)	77 (100.0)	2 (2.6)	5 (6.5)
Tricuspid atresia	65 (1.1)	65 (100.0)	8 (12.3)	2 (3.1)
Pulmonary atresia with intact septum	60 (1.0)	60 (100.0)	3 (5.0)	0 (0.0)
Aortic stenosis	60 (1.0)	10 (16.7)	9 (15.0)	1 (1.7)
Total anomalous pulmonary venous drainage	49 (0.9)	49 (100.0)	4 (8.2)	1 (2.0)
Truncus arteriosus	45 (0.9)	45 (100.0)	4 (8.9)	3 (6.7)
Ebstein anomaly	44 (0.8)	34 (77.3)	3 (6.8)	1 (2.3)
Mitral atresia	34 (0.6)	34 (100.0)	2 (5.9)	3 (8.8)
Double inlet left ventricle	34 (0.6)	34 (100.0)	3 (8.8)	3 (8.8)
Congenitally corrected TGA	34 (0.6)	34 (100.0)	1 (2.9)	2 (5.9)
Interrupted aortic arch	25 (0.4)	25 (100.0)	6 (24.0)	5 (20.0)
Pulmonary artery abnormality	23 (0.4)	18 (78.3)	2 (8.7)	3 (13.0)
Coronary artery fistula	16 (0.3)	3 (18.8)	1 (6.3)	0 (0)
Cor triatriatum	11 (0.2)	9 (81.8)	2 (18.2)	0 (0)
Anomalous left coronary artery from pulmonary artery	10 (0.2)	10 (100.0)	0 (0.0)	0 (0)
Aortopulmonary window	6 (0.1)	4 (66.7)	0 (0)	0 (0)
Mitral stenosis	2 (0.0)	2 (100.0)	0 (0)	0 (0)
Group diagnosis				
Severe, univentricular	367 (6.4)	367 (100)	29 (7.9)	17 (4.6)
Severe, biventricular	1583 (27.6)	1583 (100)	336 (21.2)	66 (4.2)
Non-severe	3778 (66.0)	–	494 (13.1)	115 (3.0)
All CHD	**5728 (100.0)**	**1944 (33.9)**	**859 (15.0)**	**198 (3.5)**

*Of the total number of patients with CHD.
†Of the total number of primary diagnoses.
CHD, congenital heart disease; TGA, transposition of great arteries; VSD, ventricular septal defect.

Survival

The estimated survival rate for CHD declines rapidly within the first few years of life, with estimated survival rates at 1, 5, 10 and 15 years are 88%, 85%, 84% and 83%, respectively (figure 2A). The survival rates are significantly lower in patients with univentricular hearts than in patients with severe biventricular heart or non-severe lesions, with 15-year survival rates of 30%, 69% and 96%, respectively (figure 2B). Further analysis shows that only 18% of patients with comfort care survive up to 15 years (figure 2C). Meanwhile, the 15-year survival for patients having antenatal diagnosis, PHT, extracardiac defect or syndrome, antenatal diagnosis, born before 2016 and male gender were 54%, 62%, 72%, 82% and 83%, respectively (online supplemental file 3).

Table 3 and online supplemental file 4 show the diagnosis-specific survival of CHD in this cohort. More than 92% of ventricular septal defect (VSD), PDA, atrial septal defect (ASD) and PS survived up to 15 years of age. However, less than 50% of truncus arteriosus, heterotaxia syndrome and HLHS reached 15 years of age. Further analysis shows that atrioventricular septal defect (AVSD) has the lowest survival rate among left-to-right shunts. On the other hand, truncus arteriosus and coarctation of the aorta have the lowest survival rates for cyanotic and obstructive lesions, respectively. The best survival rates among univentricular hearts were observed in patients with tricuspid atresia and double inlet left ventricle.

Factors associated with mortality

The independent factors associated with mortality were male gender (adjusted HR (aHR) 1.15, 95% CI 1.01 to 1.32), associated syndrome or extracardiac defect (aHR 1.95, 95% CI 1.67 to 2.28), PHT (aHR 1.55, 95% CI 1.25 to 1.93), antenatal diagnosis (aHR 1.55, 95% CI 1.22 to 1.98), severe univentricular (aHR 31.13, 95% CI 25.01 to 38.74) and severe biventricular heart (aHR 8.51, 95% CI 6.98 to 10.36).

DISCUSSION

This is the first population-based study in Malaysia and LMICs that described the outcome and survival of CHD. This study

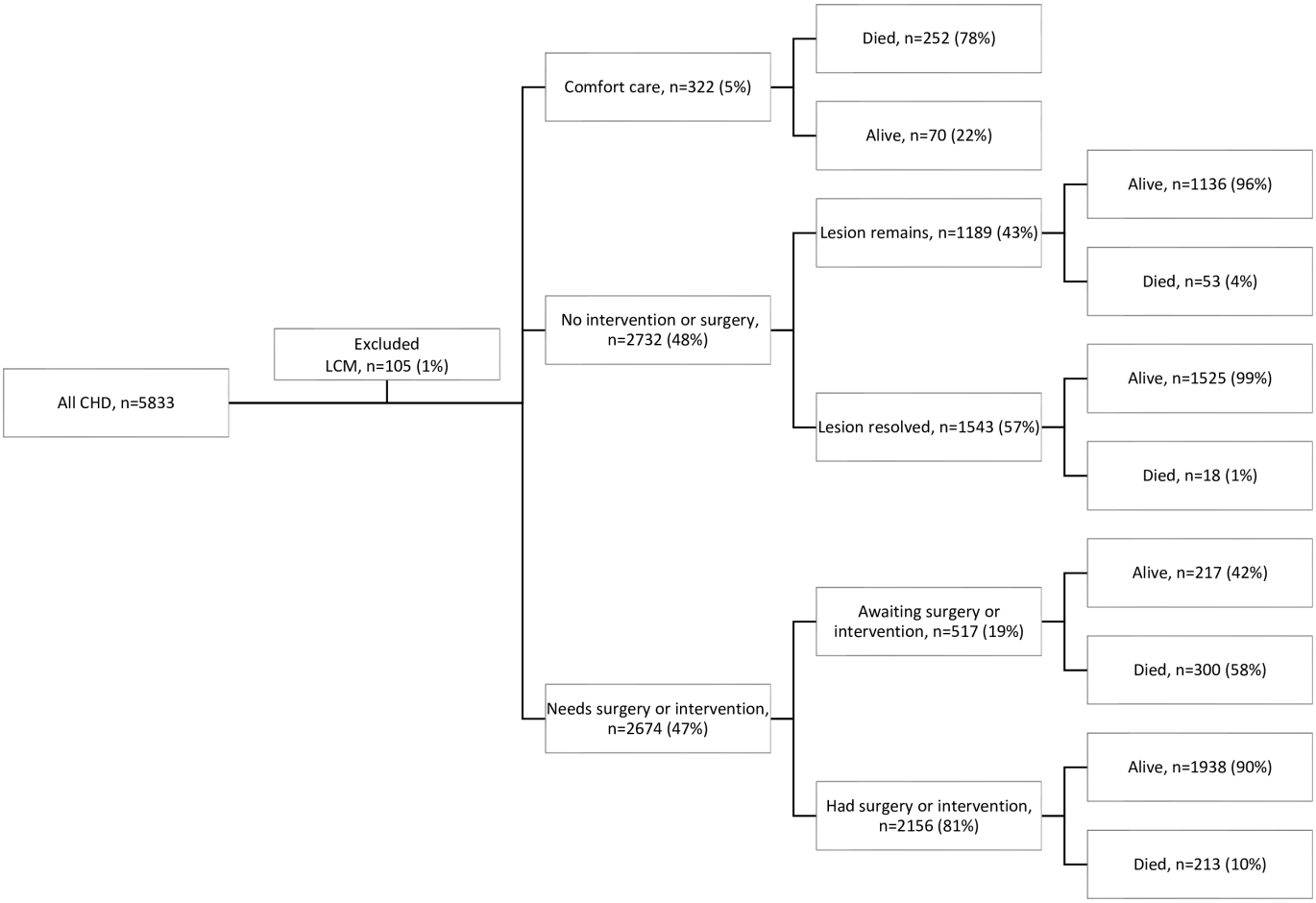

Figure 1 The outcome of the study population.

shows that survival of CHD depends largely on the severity of the lesion, with more complex lesions associated with high mortality and low survival rates. Additionally, male gender, associated non-cardiac malformation or syndrome, and PHT are significant factors that impact the survival of children with CHD in Malaysia. These factors are important to consider when evaluating the prognosis of a child with CHD.

Survival

Infant survival rates decline rapidly in the first year of life, with 1-, 5-, 10- and 15-year rates of 88%, 85%, 84% and 83%, respectively. Our study's survival rate matches that of HICs over a decade ago, which showed pooled rates of 87%, 85% and 81% for 1-, 5- and 10-year survival, respectively.[14] However, compared with a recent study in Sweden,[15] where 97% of their patients with CHD reached adulthood, our survival rate is comparatively low. Furthermore, only 60% of our children with severe CHD survive beyond 15 years, compared with 71% by Knowles *et al*[16] in the United Kingdom. Wik *et al*[17] in Norway found that 30% of those with critical CHD did not live to see their second birthday.

A lower rate is expected as our country lacks resources and expertise in congenital cardiac surgery services.[10] This is consistent with findings from other LMICs, which also report low survival rates among children with severe or critical CHD.[9]

In this study, children with left-to-right shunts (PDA, ASD, VSD and AVSD) have a higher survival rate than other types of CHD, with a 92% survival rate at 15 years. However, the survival rate for AVSD is lower than other left-to-right shunts. This is because significant deaths occur before surgery, which could be due to late surgical intervention. Therefore, to have a good outcome, a combination of early diagnosis and surgical intervention is vital.[18]

The survival rate of infants with a univentricular heart in our cohort is very low. Only 45% of them survive for 1 year, which further drops to 29% by the time they reach the age of 15. The lower survival rate could be due to the inclusion of patients with HLHS and those who received comfort care. However, this is not surprising, as univentricular heart patients have low survival in other studies.[19–21] Nonetheless, tricuspid atresia shows a comparatively better survival rate among children with univentricular hearts, with 50% surviving until age 15.

In this study, 11% of patients needing surgery or intervention died before the procedure, and most deaths were due to pneumonia and infection. This rate is higher than that found by Wik *et al*.[17] High preoperative death may result from a severe presentation at diagnosis,[10] delay in diagnosis or surgery/intervention. In this study, the presence of PHT, non-isolated CHD and CHD severity was independently associated with preoperative death. However, the timing of diagnosis is not associated with preoperative death. Despite an early diagnosis of D-transposition great arteries and truncus arteriosus, significant death occurs while waiting for surgery. Hence, in addition to improving CHD services, strengthening primary and tertiary care is crucial in preventing death related to infection and pneumonia.

Table 2 Diagnosis-specific mortality and timing of death in relation to surgery or intervention

Primary diagnosis	Total patient	Mortality, N (%)	Timing of death in relation to surgery or intervention, n (%)		
			*Before	†Within 30 days	†After 30 days
VSD	2176	109 (5.0)	48/650 (7.4)	8/531 (1.5)	11/531 (2.1)
Patent ductus arteriosus	988	59 (6.0)	18/549 (3.3)	2/511 (0.4)	26/511 (5.1)
Pulmonary stenosis	609	26 (4.3)	6/70 (8.6)	2/52 (3.8)	2/52 (11.5)
Atrial septal defect	321	13 (4.0)	4/160 (2.5)	0/126 (0)	0/126 (0)
Tetralogy of Fallot	299	51 (17.1)	28/296 (9.5)	5/247 (2.0)	15/247 (6.1)
D-transposition great arteries	139	52 (37.4)	27/136 (19.9)	13/114 (11.4)	9/114 (7.9)
Atrioventricular septal defect	134	55 (41.0)	22/104 (21.1)	3/71 (4.2)	7/71 (9.8)
Coarctation of aorta	112	27 (24.1)	12/88 (13.6)	5/72 (6.9)	6/72 (8.3)
Pulmonary atresia with VSD	109	50 (45.9)	23/84 (27.4)	2/60 (3.3)	15/60 (25.0)
Heterotaxia syndrome	94	66 (70.2)	3/25 (12.0)	2/21 (9.5)	6/21 (28.6)
Double outlet right ventricle	85	25 (29.4)	9/75 (12.0)	2/54 (3.7)	6/54 (11.1)
Hypoplastic left heart syndrome	77	71 (92.2)	0/3 (0)	3/3 (100)	0/3 (0)
Tricuspid atresia	65	28 (43.1)	11/55 (20.0)	2/44 (4.5)	11/44 (25.0)
Pulmonary atresia with intact septum	60	21 (35.0)	10/59 (16.9)	6/49 (12.2)	4/49 (8.2)
Aortic stenosis	60	13 (21.7)	5/16 (31.2)	0/9 (0)	3/9 (33.3)
Total anomalous pulmonary venous drainage	49	23 (46.9)	10/48 (20.8)	4/38 (10.5)	8/38 (21.0)
Truncus arteriosus	45	27 (60.0)	20/39 (51.3)	1/18 (5.6)	1/18 (5.6)
Ebstein anomaly	44	11 (25.0)	3/27 (11.1)	0/9 (0)	0/9 (0)
Mitral atresia	34	21 (61.8)	7/21 (33.3)	0/13 (0)	4/13 (30.7)
Double inlet left ventricle	34	14 (41.2)	4/29 (13.8)	3/24 (12.5)	3/24 (12.5)
Congenitally corrected TGA	34	7 (20.6)	0/27 (0)	0/20 (0.0)	1/20 (5.0)
Interrupted aortic arch	25	14 (56.0)	8/23 (34.8)	2/15 (13.3)	2/15 (13.3)
Pulmonary artery abnormality	23	7 (30.4)	3/15 (20.0)	0/10 (0)	1/10 (10.0)
Coronary artery fistula	16	3 (18.8)	2/12 (16.7)	0/5 (0)	0/5 (0)
Cor triatriatum	11	2 (18.2)	1/10 (10.0)	1/8 (12.5)	0/8 (0)
Anomalous left coronary artery from pulmonary artery	10	3 (30.0)	1/10 (10.0)	0/9 (0)	0/9 (0)
Aortopulmonary window	6	2 (33.3)	1/4 (25.0)	0/3 (0)	0/3 (0)
Group					
Severe, univentricular	367	239 (65.1)	38/175 (21.7)	11/131 (8.4)	30/131 (22.9)
Severe, biventricular	1583	467 (29.5)	224/1458 (15.4)	53/1136 (4.7)	104/1136 (9.1)
Non-severe	3778	130 (3.4)	38/1041 (3.6)	4/884 (0.4)	11/884 (1.2)
All CHD	**5728**	**836 (14.6)**	**300/2674 (11.2)**	**68/2158 (3.1)**	**145/2158 (6.7)**

*Total patients need surgery or intervention.
†Total patients had surgery or intervention.
CHD, congenital heart disease; TGA, transposition of great arteries; VSD, ventricular septal defect.

Mortality

The overall mortality in our cohort was 15% and has significantly improved over time, correlating with Su *et al*,[22] which shows improvement in mortality in most middle-income countries. However, our mortality rate is higher than those reported in Spain,[23] Norway[7] and Taiwan,[8] with mortality of 5%, 6% and 7%, respectively. This result is not surprising given the lack of resources and expertise in our healthcare system, as well as the complexity of lesions associated with non-cardiac malformations or syndromes in our cohort.

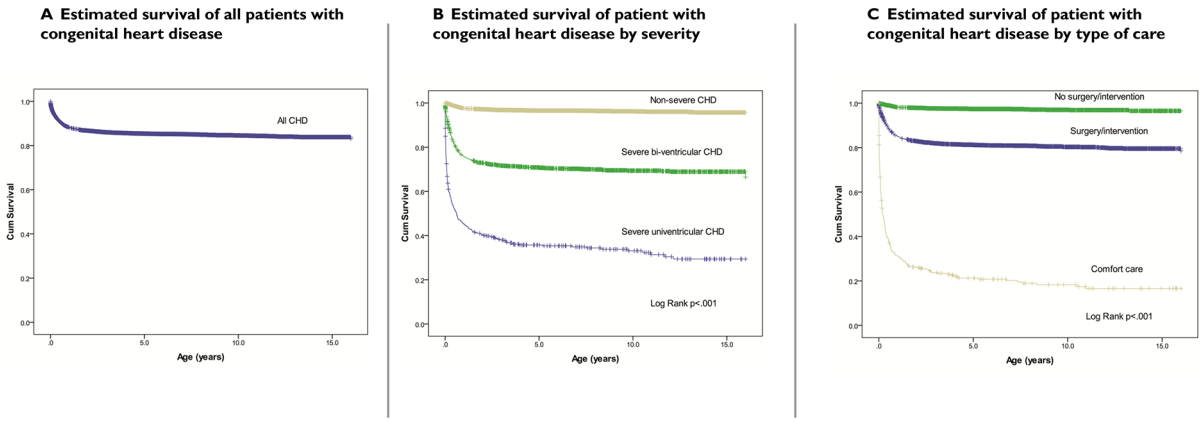

Figure 2 The estimated survival of all children with congenital heart disease (A), by severity of lesion (B) and by management (C).

Table 3 Survival estimates of children with CHD at 1 year to 15 years of age in Johor, Malaysia, 2006–2020

Specific primary diagnosis*	Total number	Survival estimates (95% CI)			
		1 year	5 years	10 years	15 years
VSD	2176	96.0 (95.2 to 96.9)	94.9 (94.0 to 95.9)	94.6 (93.6 to 95.6)	94.0 (92.7 to 95.4)
Patent ductus arteriosus	988	95.7 (94.4 to 97.0)	93.9 (92.4 to 95.5)	93.4 (91.8 to 95.1)	92.8 (90.8 to 94.9)
Pulmonary stenosis	609	96.9 (95.5 to 98.3)	95.7 (94.1 to 97.4)	95.3 (93.5 to 97.2)	94.8 (92.7 to 96.9)
Atrial septal defect	321	97.4 (95.7 to 99.2)	95.7 (93.4 to 98.0)	95.7 (93.4 to 98.0)	95.7 (93.4 to 98.0)
Tetralogy of Fallot	299	91.2 (87.9 to 94.4)	83.7 (79.5 to 88.0)	81.6 (76.9 to 86.2)	81.6 (76.9 to 86.2)
D-Transposition great arteries	139	64.3 (56.3 to 72.3)	62.5 (54.3 to 70.7)	62.5 (54.3 to 70.7)	62.5 (54.3 to 70.7)
Atrioventricular septal defect	134	74.6 (67.2 to 82.0)	60.6 (52.2 to 69.0)	55.6 (46.1 to 65.1)	55.6 (46.1 to 65.1)
Coarctation of aorta	112	76.4 (68.5 to 84.3)	75.4 (67.4 to 83.5)	75.4 (67.4 to 83.5)	75.4 (67.4 to 83.5)
Pulmonary atresia with VSD	109	65.2 (56.1 to 74.2)	52.5 (42.9 to 62.0)	52.5 (42.9 to 62.0)	52.5 (42.9 to 62.0)
Heterotaxia syndrome	94	37.4 (27.5 to 47.4)	31.7 (22.0 to 41.3)	27.5 (17.5 to 37.5)	19.6 (8.0 to 31.3)
Double outlet right ventricle	86	75.1 (65.8 to 84.3)	71.2 (61.4 to 81.0)	71.2 (61.4 to 81.0)	65.7 (52.0 to 79.4)
Hypoplastic left heart syndrome	77	7.9 (1.8 to 14.0)	6.6 (1.0 to 12.2)	–	–
Tricuspid atresia	65	70.1 (58.8 to 81.4)	58.6 (46.4 to 70.8)	58.6 (46.4 to 70.8)	49.9 (34.6 to 65.2)
Aortic stenosis	60	86.7 (78.1 to 95.3)	83.2 (73.7 to 92.7)	77.2 (65.3 to 89.1)	72.7 (58.5 to 86.8)
Pulmonary atresia with intact septum	60	67.8 (55.9 to 74.2)	64.3 (52.1 to 76.6)	64.3 (52.1 to 76.6)	64.3 (52.1 to 76.6)
Total anomalous pulmonary venous drainage	49	54.4 (40.3 to 68.5)	51.9 (37.7 to 66.2)	51.9 (37.7 to 66.2)	51.9 (37.7 to 66.2)
Truncus arteriosus	45	40.0 (25.7 to 54.3)	40.0 (25.7 to 54.3)	40.0 (25.7 to 54.3)	40.0 (25.7 to 54.3)
Ebstein anomaly	44	75.0 (62.2 to 87.8)	75.0 (62.2 to 87.8)	75.0 (62.2 to 87.8)	75.0 (62.2 to 87.8)
Group diagnosis					
Severe, all	1950	70.0 (68.0 to 72.0)	64.2 (62.0 to 66.4)	62.6 (60.4 to 64.8)	61.5 (59.1 to 63.9)
Severe, univentricular	367	45.2 (40.1 to 50.3)	35.7 (30.6 to 40.8)	33.1 (28.0 to 38.2)	29.4 (23.7 to 35.1)
Severe, biventricular	1583	75.8 (73.6 to 78.0)	70.8 (68.4 to 73.2)	69.4 (67.0 to 71.8)	68.9 (66.5 to 71.3)
Non-severe	3778	97.7 (97.1 to 98.3)	96.5 (95.9 to 97.1)	96.2 (95.6 to 96.8)	95.7 (94.9 to 96.5)
All CHD	**5728**	**88.2 (87.4 to 89.0)**	**85.4 (84.4 to 86.4)**	**84.5 (83.5 to 85.5)**	**83.2 (82.2 to 84.2)**

*Selected CHD with a frequency of more than 40.
CHD, congenital heart disease; VSD, ventricular septal defect.

Identifying mortality factors helped in strategies to reduce deaths in children with CHD. This study found that male gender, syndrome/non-cardiac malformation, PHT and lesion severity were independent factors for mortality. This finding is consistent with a recent study in the USA.[24] However, our study has found that early diagnosis is not associated with good survival, which contrasts with other studies.[25 26] This may be due to a low rate of fetal diagnoses, with the majority being severe univentricular hearts associated with unfavourable outcomes.[27] Second, insufficient resources and expertise may delay surgical procedures or interventions. Hence, it is imperative to adopt a multifaceted approach (online supplemental file 5) that addresses various aspects of healthcare, infrastructure and education to enhance outcomes for CHD in our country.[28]

Limitation

Limitations in this study include the inclusion of children treated with comfort care, which may result in a lower overall survival rate for CHD. Additionally, due to the varied classifications of CHD, comparing results with other studies is difficult. Lastly, limited variables in the registry, such as additional lesion and surgical data, make it difficult to determine all mortality factors.

The study's findings may not accurately reflect CHD survival rates in Malaysia. A national registry is needed for a more accurate picture. However, the study provides valuable information for planning congenital cardiac services in Malaysia. Additionally, these results may not apply to other LMICs due to differences in healthcare settings, antenatal diagnosis and cultural/religious factors.

CONCLUSION

This population-based study revealed that the overall outcome of CHD is good except for those with severe lesions, non-cardiac malformations or syndromes, and PHT. In addition to improving congenital cardiac services, enhancing the overall healthcare system to mitigate infections and pneumonia may decrease preoperative mortality rates and improve overall survival.

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