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RESEARCH

Survival and factors associated with mortality among infants with anorectal malformation: a population-based study from a middle-income country

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Abstract

Limited data on the survival of anorectal malformation (ARM) patients from lower- and middle-income countries is available. This retrospective population-based study from the State of Johor, Malaysia, determines the incidence, mortality rate, and survival of ARM patients and factors associated with mortality. Kaplan–Meier survival analysis was used to estimate the survival of ARM patients at 1, 5, and 10 years. In addition, multivariate Cox regression analysis was used to analyze mortality-related factors. There were 175 ARM patients among 803,850 live births, giving an overall ARM incidence of 2.2 (95% confidence interval [CI], 1.9 to 2.5) per 10,000 live births. The male-to-female ratio was 1.5:1. There were 122 (69%) non-isolated ARM, of which 41 were Trisomy-21 and 34 had VACTERL association. Seventy-three (42%) had congenital heart disease (CHD), with 38 severe and 35 non-severe CHD. Overall, 33 (19%) patients died, with a median age of death of 5.7 months (interquartile range (IQR) 25 days to 11.2 months). The overall estimated 1-, 5-, and 10-year survival rate for ARM patients was 82% (95% CI, 76–89%), 77% (95% CI, 70–84%), and 77% (95% CI, 70–84%), respectively. Univariate analysis shows that non-isolated ARM, VACTERL association, and severe CHD were associated with mortality. However, only severe CHD is the independent factor associated with mortality, with a hazard ratio of 4.0 (95% CI, 1.9–8.4).

Conclusion: CHD is common among ARM patients, and one in five ARM patients had a severe cardiac defect, significantly affecting their survival.

What is Known:
- *VACTERL association and congenital heart disease are common in patient with anorectal malformation.*
- *Low birth weight and prematurity are associated with a lower rate of survival.*

What is New:
- *Congenital heart disease is common in ARM patients in a middle-income country.*
- *Severe congenital heart disease plays a significant role in the survival of patients with an anorectal malformation in lower- and middle-income countries.*

Keywords Anorectal malformation · Congenital heart disease · Middle-income country · Mortality · Survival

Abbreviations

ARM	Anorectal malformation
CHD	Congenital heart disease

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CI	Confidence interval
IQR	Interquartile range
VACTERL	Vertebral defects, anal atresia, cardiac defect, tracheoesophageal fistula, renal anomalies, and limb abnormalities

Introduction

Congenital anorectal malformation (ARM) comprises a broad spectrum of abnormalities of the anus and anal cavity, with a reported prevalence of 2 to 5 per 10,000 live births [1–3]. It is the most common gastrointestinal malformation and can occur in isolation or associated with other malformations or syndromes [2]. In addition to Trisomy-21 and VACTERL association, ARM is also known to be associated with congenital heart disease (CHD). However, the reported prevalence of CHD among ARM patients is highly variable, ranging from as low as 9% to as high as 50% [4–12]. Furthermore, there is a conflicting report on the most common cardiac lesion in ARM patients, with ventricular septal defect [8, 13], atrial septal defect [4, 12], and tetralogy of Fallot [6] described as frequent lesions.

In addition, the mortality and survival of ARM patients vary greatly depending on the economic status of the countries. For example, the reported mortality in high-income countries ranges from 1.7 to 2.9% [14, 15]. In comparison, a higher mortality rate was observed in the lower- and middle-income countries, with a reported rate ranging from 12 to 29% [14–16]. Additionally, the prognosis of ARM patients was also affected by various factors such as the number of associated anomalies, prematurity, low birth weight, delayed presentation, and primary perforation [1, 2, 11]. However, studies on the effect of CHD on the mortality and survival of ARM patients are limited [16].

Furthermore, a population-based study from lower- and middle-income countries is lacking. Therefore, with a lack of resources and expertise, we postulate that the survival of ARM patients is lower than in high-income countries, and CHD plays a significant role in the survival of ARM patients. Therefore, this study aims to assess the prevalence and pattern of CHD and its effect on the overall outcome of ARM patients.

Materials and methods

This retrospective cohort study includes all individuals with ARM who were born in Johor, Malaysia, between January 2006 and December 2019. Malaysia is classified as a middle-income nation. There was an estimated population of 3.4 million in the State of Johor, with an annual live births of 50,000 per year. All cases of ARM and CHD were referred to Hospital Sultanah Aminah, a tertiary government hospital that provides pediatric cardiology and surgical services for the State of Johor, Malaysia [17].

This study was approved by the Medical Research and Ethics Committee, Ministry of Health, Malaysia (NMRR 21–1881-61,323). A waived informed consent was given due to the study's retrospective nature. All procedures performed in this study followed the institutional and national research committee's clinical standards, the 1964 Declaration of Helsinki, and its later amendments or comparable ethical standards.

ARM was diagnosed based on clinical features such as imperforate anus, presence of fistula, ectopic anus, or cloacal anomaly and was classified according to the Krickenbeck classification [18]. When a patient in Johor is diagnosed with ARM or requires surgical intervention, a 2d-echocardiogram is performed. The results are then documented in the Pediatric Cardiology Clinical Information System, regardless of whether they are normal or abnormal. In addition, other congenital malformations were recorded and divided into four groups as described by Cuschieri [3]: (1) syndromes of known cause, (2) recognized syndromes and sequences of unknown etiology, (3) VACTERL association, and (4) multiple congenital anomalies. Patients with three or more defects (vertebral, anal atresia, cardiac septal defects, esophageal atresia or tracheoesophageal fistula, renal anomalies, and radial limb defects) were considered to have the VACTERL association [19]. ARM was divided into two groups, those with associated other malformations or syndromes as non-isolated ARM [2] and, conversely, isolated ARM if there was no association with any syndrome or malformation.

CHD was defined as a significant structural abnormality of the heart or the great intrathoracic vessel that was of actual or potential significance [20]. All CHD was confirmed with two-dimensional echocardiography. Patients with patent foramen ovale, mild branch pulmonary stenosis, isolated dextrocardia, isolated bilateral superior vena cava, isolated right arch, and spontaneous closure PDA in 3 months (for a term infant) and 6 months (for premature) were not considered as CHD [21]. In patients with multiple cardiac defects, the primary lesion that required the first intervention or was hemodynamic significant was regarded as the primary defect. The severity of CHD was divided into mild, moderate, and severe [22].

Data were retrieved from the Pediatric Cardiology Clinical Information System, a clinical database for acquired and congenital heart disease in the State of Johor, and have been described in detail in our previous publication [21, 23]. It was developed in 2006 and contains all the demography, clinical, echocardiography, and outcome of patients who had pediatric cardiology consultation in the Pediatric Cardiology Unit. It was maintained in Sultanah Aminah Hospital and was updated regularly. For this study, data collected included demographic data (gestational age, maternal

gestational diabetes, syndrome, birth weight, sex, and ethnicity), ARM data (type and associated with other malformation), CHD data (type and severity), and finally the outcome at the last follow-up (alive or dead). Prematurity was defined as infants born before 37 completed weeks of gestation. All patients had a complete clinical examination by the medical and pediatric surgical team. This included a two-dimensional echocardiogram for screening of CHD. Active screening for other congenital anomalies associated with ARM or syndrome was also conducted. Similar to CHD, further ARM management was done according to the severity of the disease.

The primary outcome measured was the survival of the infant. Mortality (all causes of death) was verified with the National Registry Department, Malaysia. Secondary outcomes measured included deaths related to cardiac issues, infections, or pneumonia.

Statistical analysis

Statistical analysis was performed using Statistical Package for Social Science (SPSS) version 23 (IBM Corp., Armonk, NY, USA). Groups were compared using Student's *t*-test for normally distributed continuous data and a non-parametric test for non-normally distributed continuous data. Pearson's chi-square test for categorical variables. A two-tailed test of significance were used, and p value < 0.05 was considered statistically significant. The incidence of ARM was calculated as the sum of newly diagnosed ARM divided by total live births in the State of Johor and expressed as per 10,000 live births.

Kaplan–Meier analysis was used to estimate survival at 1, 5, and 10 years. Log-rank test was used to compare the difference between the group.

Univariate Cox proportional hazards regression was used to identify the unadjusted effect of age, sex, ethnicity, gestational age, birth weight, birth year, VACTERL association, Trisomy-21, and CHD severity on mortality. Variables with p values < 0.1 in the univariate analyses were entered into the Cox proportional hazards regression to identify the independent risk factors associated with death. A hazard ratio was considered significant if the 95% confidence interval (CI) excluded one. In our study, we use data imputation to address missing data. The type of missing data is missing at random. We do data imputation by replacing each missing value with the mean of the observed values for that variable.

Results

There were 175 ARM patients among 803,850 live births during the study period, giving an overall ARM incidence of 2.2 (95% CI, 1.9 to 2.5) per 10,000 live births.

Of 175, 105 (60%) were male, 136 (78%) were Malay ethnicity, and 60 (34%) had low birth weight (Table 1). The mean age of diagnosis was 1.5 days (ranging from 0 to 75 days). All patients were followed up at a median age of 5.1 years (IQR from 0.4 to 8.8 years).

Of 175 ARM patients, 169 (97%) were detected during newborn examination, 2 (1.1%) patients presented at birth with abdominal distension, 2 (1.1%) were suspected antenatally, and 1 (0.6%) with an abnormal route of passing stools/meconium. In addition, one patient had her ARM detected at 3 months of life during pediatric intensive care admission for pneumonia. Of the 97 available data on the subtype of ARM, 51 (29%) had no fistula, 12 (6.9%) rectovestibular, 8 (4.6%) cloacal anomalies, 8 (4.6%) rectovaginal fistula, 7 (4.0%) rectourethral, 7 (4.0%) perineal, and 4 (2.3%) rectal atresia.

One hundred twenty-two (70%) ARM patients had an underlying syndrome or associated congenital malformation. Recognized syndrome with a known cause is seen in 49 (28%), with a majority ($n = 41$) being Trisomy-21. VACTERL association was observed in 34 (19%) patients. In addition, one patient had omphalocele, exstrophy, imperforate anus, spinal bifida complexes, 2 Edward syndrome, and one each for Patau syndrome, prune-belly, Goldenhar syndrome, Wolf-Hirschhorn syndrome, chromosomes 18 and 16 disorder. Of 175 ARM patients, 6 (3.4%) were associated with neural tube defects and 2 (1.1%) with omphalocele. There was no significant difference in the proportion of non-isolated ARM between females and males (51/70 [73%] vs. 71/105 [68%], $p = 0.46$).

Seventy-three (42%) had CHD, of which 38 (52%) were severe CHD, and 35 (48%) were non-severe CHD (Table 2). The commonest lesions were ventricular septal defect ($n = 29$, 40%), followed by tetralogy of Fallot ($n = 10$, 14%). Of 73 patients with CHD, 36 (49%) required surgery, and 11 (15%) patients had their cardiac defect closed spontaneously (all were small ventricular septal defects).

Of 175 ARM patients, nine (5.1%) were treated with comfort care, of which three were due to lethal congenital malformation, four were due to complex congenital cardiac defects, and two were for severe persistent pulmonary hypertension of the newborn. Overall, 33 (19%) patients died, with a median age of death of 5.7 months (IQR 25 days to 11.2 months). Of 33 deaths, 8 (24%) were cardiac-related, 7 (21%) infection-related, 6 (12%) pneumonia, and 12 (36%) of other causes (Table 3). All the infection-related death was unrelated to ARM or surgical infection. Further analysis shows that of 73 CHD patients, 14 (19%) died, nine prior to surgery and five after 30 days of surgery.

The overall 1-, 5-, and 10-year survival rate for ARM patients was 82% (95% CI, 76–89%), 77% (95% CI, 70–84%), and 77% (95% CI, 70–84%), respectively. However, there was a significantly lower survival in ARM

Table 1 The characteristic and immediate outcome of an infant with an anorectal malformation

Variable	Total ($n=175$)		Outcome				p value
			Survivor ($n=142$)		Non-survivor ($n=33$)		
Sex							
Male	105	(60)	89	(63)	16	(48)	0.13
Female	70	(40)	53	(37)	17	(51)	
Gestational age							
Premature	34	(19)	30	(21)	4	(12)	0.33
Term	141	(81)	112	(79)	29	(88)	
Race							
Malay	136	(78)	108	(76)	28	(85)	0.36
Non-Malay	39	(22)	34	(24)	5	(15)	
Birth weight							
< 2.5 kg	60	(34)	48	(34)	12	(36)	0.78
2.5 kg and more	115	(66)	94	(66)	21	(64)	
Birth year							
2006–2010	42	(24)	32	(22)	10	(30)	0.59
2011–2015	75	(43)	63	(44)	12	(36)	
2016–2019	58	(33)	47	(33)	11	(33)	
Maternal diabetic							
No	148	(85)	122	(86)	26	(79)	0.30
Yes	27	(15)	20	(14)	7	(21)	
ARM							
Non-isolated ARM	122	(70)	92	(65)	30	(91)	0.005
Isolated ARM	53	(30)	50	(35)	3	(9.1)	
Down syndrome							
No	134	(77)	105	(74)	29	(88)	0.11
Yes	41	(23)	37	(26)	4	(12)	
VACTERL association							
No	141	(81)	121	(85)	20	(61)	0.001
Yes	34	(19)	21	(15)	13	(39)	
CHD							
No	102	(58)	90	(65)	10	(30)	< 0.001
Yes	73	(42)	50	(35)	23	(70)	
CHD severity							
Severe CHD	38	(22)	20	(14)	18	(54)	< 0.001
Non-severe CHD	35	(20)	30	(21)	5	(15)	
No CHD	102	(58)	92	(65)	10	(30)	
ARM type							
Major clinical group	163	(93)	133	(94)	30	(91)	0.70
Rare/regional variant	12	(6.9)	9	(6.3)	3	(9.1)	

(%) percentage within the outcome

ARM anorectal malformation, *CHD* congenital heart disease, *VACTERL* vertebral defects, anal atresia, cardiac defect, tracheoesophageal fistula, renal anomalies, and limb abnormalities

patients with non-isolated ARM, with survival at 1, 5, and 10 years of 77% (95% CI 69–85%), 72% (95% CI 64–81%), and 72% (95% CI, 64–81%), respectively (Fig. 1). Further analysis revealed ARM patients with VACTERL association had a lower survival rate than the non-VACTERL association, with estimated survival at 1 and 5 years of VACTERL association were 66% (95% CI, 49–83%) and 55% (95% CI, 37–77%), respectively (Fig. 2). Similarly, a lower survival was observed in ARM patients with severe CHD, with estimated survival rates at 1 and 5 years of severe CHD were 62% (95% CI, 46–78%) and 50% (95% CI, 34–67%), respectively (Fig. 3). In comparison, there

Table 2 Frequency of congenital heart disease among infants with anorectal malformation

CHD name	*N*	(%)
Ventricular septal defect	29	(40)
Tetralogy of Fallot	10	(14)
Pulmonary atresia with VSD	7	(9.6)
Atrial septal defect	6	(8.2)
Patent ductus arteriosus	6	(8.2)
Pulmonary stenosis	3	(4.1)
Total anomalous pulmonary venous drainage	3	(4.1)
Double outlet right ventricle	2	(2.7)
Hypoplastic left heart syndrome	2	(2.7)
Atrioventricular septal defect	1	(1.4)
Coarctation of aorta	1	(1.4)
D-transposition great arteries	1	(1.4)
Aortic stenosis	1	(1.4)
Interrupted aortic arch	1	(1.4)
Total	73	(100)

(%) percentage within total CHD

CHD congenital heart disease, *VSD* ventricular septal defect

Table 3 Causes of death of anorectal malformation

Cause of death	*N*	%
Infection-related	7	21
Septicemia	3	9.1
Klebsiella sepsis	1	3.0
COVID-19	1	3.0
RSV infection	1	3.0
Urosepsis	1	3.0
Pneumonia	6	18
Adenovirus pneumonia	1	3.0
Severe pneumonia	3	9.1
Aspiration pneumonia	2	6.1
Cardiac-related	8	24
Cardiac failure	5	15
Severe pulmonary hypertension	2	6.1
LPA sling causing airway obstruction	1	3.0
Others	12	42
Unknown	10	30
Hepatoblastoma	1	3.0
Leukemia	1	3.0
Total	33	100

LPA left pulmonary artery sling, *RSV* respiratory syncytial virus

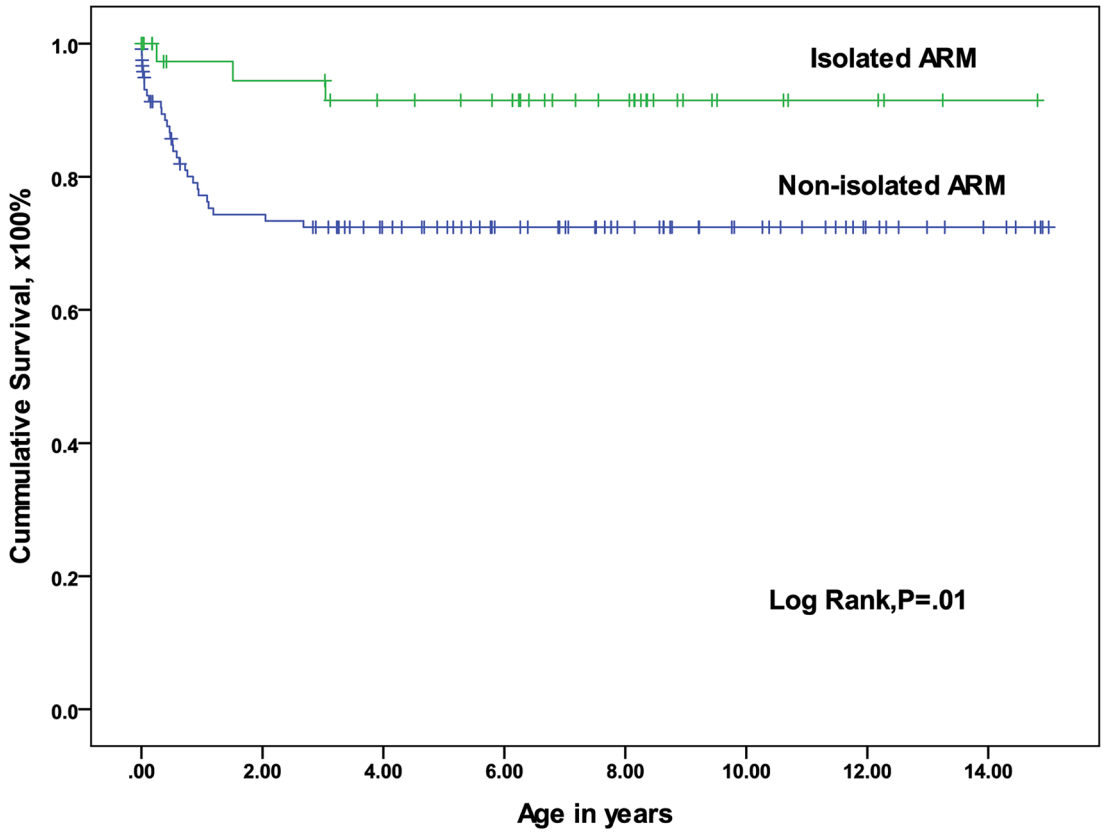

Fig. 1 Kaplan–Meier survival curves for an infant with non-isolated versus isolated anorectal malformation

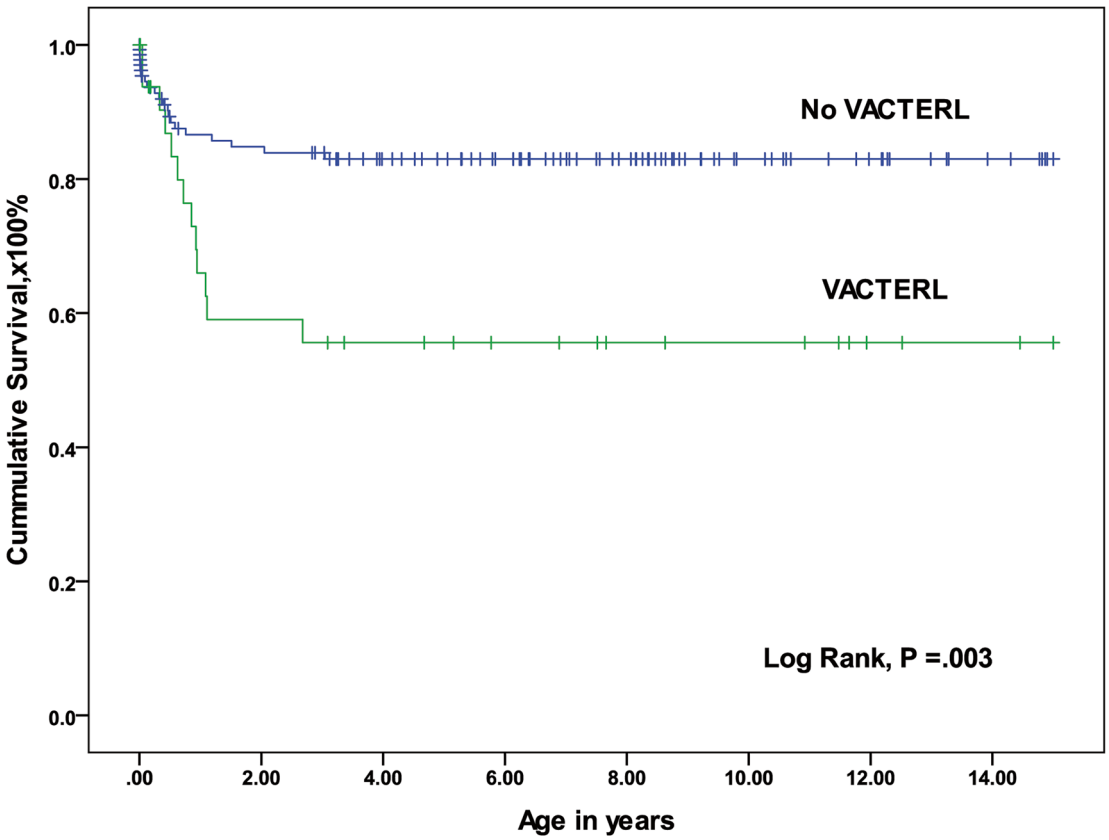

Fig. 2 Kaplan–Meier survival curves for anorectal malformation patients with and without VACTERL association

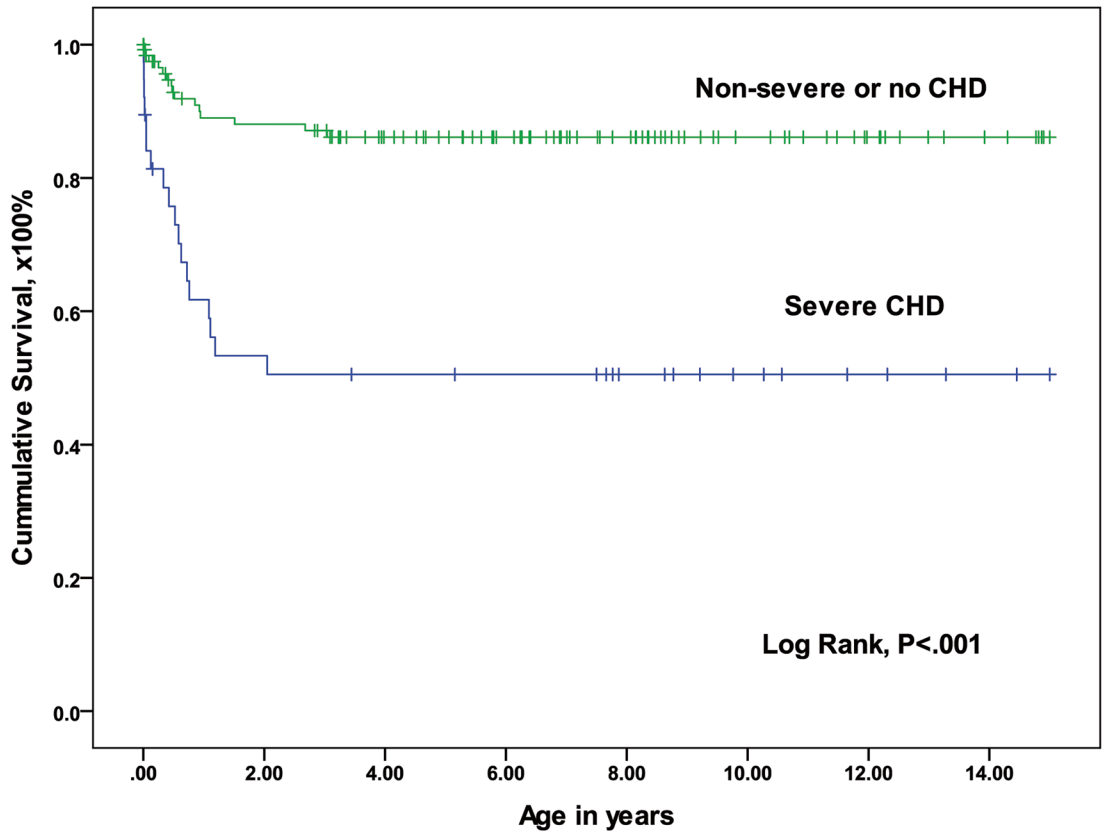

Fig. 3 Kaplan–Meier survival curves for anorectal malformation patients with severe, non-severe, and no congenital heart disease

Table 4 Univariate and multivariate analysis of factors associated for mortality of infants with anorectal malformation

Variables		All	Mortality		Crude HR (95% CI)	*p* value	*Adjusted HR (95% CI)	*p* value
		N	*N*	(%)				
Sex	Female	70	17	(24)	1.71 (0.87–3.39)	0.122	1.82 (0.91–3.62)	0.088
	Male	105	16	(15)	Reference		Reference	
Ethnicity	Malay	136	28	(21)	1.56 (0.60–4.03)	0.362	1.41 (0.54–3.62)	0.483
	Non-Malay	39	5	(13)	Reference		Reference	
Gestation	Term	141	29	(21)	0.64 (0.22–1.81)	0.399	–	
	Premature	34	4	(12)	Reference			
Birth weight (kg)	< 2.5	60	12	(20)	1.21 (0.59–2.47)	0.591	–	
	≥ 2.5	115	21	(18)	Reference			
Birth year	2006–2010	42	10	(24)	1.04 (0.46–2.55)	0.854	–	
	2011–2015	75	12	(16)	0.86 (0.38–1.94)	0.707	–	
	2016–2019	58	11	(19)	Reference			
VACTERL	Yes	34	13	(38)	2.77 (1.38–5.58)	0.004	1.39 (0.64–3.03)	0.403
	No	141	20	(14)	Reference		Reference	
Trisomy-21	Yes	41	4	(9.8)	0.37 (0.13–1.05)	0.061	0.49 (0.16–1.48)	0.206
	No	134	29	(22)	Reference		Reference	
CHD severity	Severe	38	18	(47)	4.66 (2.34–9.27)	< 0.001	4.03 (1.93–8.42)	< 0.001
	Non-severe/no CHD	137	15	(11)	Reference		Reference	

A *p* value is considered significant if a 95% confidence interval does not include 1

HR hazard ratio, *CI* confidence interval, *CHD* congenital heart disease

*Analyzed with Cox regression analysis, corrected for sex and ethnicity

were no significant differences in the survival of ARM patients by sex, ethnicity, or gestational age. However, corrected for sex and ethnicity, multivariable Cox regression analysis shows severe CHD as the only significant factor associated with mortality with a hazard ratio of 4.0 (95% CI, 1.9–8.4) (Table 4).

Discussion

This first population-based study from lower- and middle-income countries examined the prevalence of CHD among ARM patients over a 14-year duration. In addition, it analyzed its effect on the overall survival of ARM patients. In this cohort, almost one in two ARM patients had CHD, and severe CHD was associated with poor outcomes. Previous studies showed wide variation in the prevalence of CHD among ARM patients, ranging from 9 to 50% [4–12]. The broad difference in the prevalence of CHD among ARM patients is due to the nature of the study, which involved a single center, a small sample size, and patient selection. The prevalence of CHD among ARM patients in this study (42%) is slightly higher than the population-based studies in Italy and the UK, with a rate of 36% [1, 2]. There are several possible explanations for this result. Firstly, due to the active screening for CHD with 2d-echocardiogram in all neonates with congenital anomalies. Secondly, it could be due to the inclusion of a small ventricular septal defect, which closed spontaneously. Nevertheless, our study shows that one in five ARM patients had a severe cardiac defect.

Similarly, there were contradicting reports of the most common CHD in ARM patients, with ventricular septal defect, atrial septal defect, and tetralogy of Fallot as the commonest lesion [4, 8, 12, 13]. In this study, a ventricular septal defect is the commonest cardiac defect among ARM patients and represents one-third of CHD. This result is similar to our general population, where a ventricular septal defect is the most common type of CHD [21].

The incidence of ARM in this study is within the lower range of the published population-based studies, ranging from 2 to 4 per 10,000 live births [1]. A lower incidence of ARM in this study could be due to underreporting, as some infants have died before reaching a diagnosis or were referred to other surgical centers in a nearby state.

The overall mortality of ARM varied, with a higher rate in the lower- and middle-income countries than in higher-income countries [14, 15]. A slightly higher rate of mortality was observed in this study compared to the 12% reported by Wright et al. [14]. A higher rate of mortality could be due to the inclusion of those who were treated with comfort

care secondary to lethal congenital malformation as well as hypoplastic left heart syndrome in our cohort. Another reason was that notable death was related to infection in this study, which is still a common scenario in other lower- and middle-income countries [24].

The overall estimated survival rate of ARM patients at 1 year was 82% and reached a plateau at 3 years with a rate of 77%, with a lower survival rate in non-isolated ARM. Our survival rate is lower than a recent study from high-income countries by Ford et al. [1] from the UK and Cassina et al. [2] from Italy. In these two high-income countries, the survival rate of non-isolated ARM patients ranged from 87 to 89% compared to 72% in our cohort.

There were many contributing factors associated with poor survival. Previous studies have shown that low birth weight, prematurity, late presentation, and a higher number of congenital anomalies as significant factors associated with mortality [1, 2, 11]. However, this study shows that severe CHD plays a significant role in ARM survival. Almost half of the ARM patients with severe CHD did not survive by 5 years of age and were five times more likely to die than non-severe CHD or non-CHD. This result is not surprising, being a lower- and middle-income country lacking resources and expertise, especially in dealing with a non-isolated ARM with severe CHD. Limited resources and expertise in pediatric congenital cardiac services in our country have significantly affected the outcome of CHD children. Previous studies in our country showed that only 6% of severe CHD were detected in utero, and 36% were diagnosed after 1 month of life [21]. In addition to late diagnosis, long waiting time for cardiac surgery is also a common scenario, leading to significant death. Almost 20% of critical CHD died while waiting for surgery, and 10% died within 30 days of surgery or intervention [25]. Furthermore, as in other low–middle-income countries, infection and pneumonia remain a major challenge, of which about 11% died due to infection and pneumonia after surgical correction [25]. Therefore, in relation to Sustainable Development Goal 3.2, "end preventable deaths in neonates and children under five by 2030, overall infection control and prompt access to congenital cardiac surgery are much needed to improve the survival of ARM patients." Early detection and surgical intervention of severe CHD are pivotal to achieving this goal.

Strength of the study

The major strength of this study includes a population study over 14 years with an early cardiac evaluation with a 2d-echocardiogram. This allows early diagnosis of severe CHD, allowing a preoperative plan and subsequent management.

Limitations

There were a few limitations of the study. Firstly, as in other studies using clinical registry [2], missing data in the subtype of ARM and a small number of other congenital anomalies in this cohort prevent us from analyzing the effect of these variables on the overall survival of ARM patients. Secondly, we may miss some early and undiagnosed neonatal deaths, which may have underestimated the prevalence of CHD among ARM patients. Finally, the lack of surgical ARM data in the registry prohibits us from analyzing the roles of the surgical variable on the overall survival of the ARM in our cohort.

Conclusion

This study explored the prevalence of CHD and its effect on the overall survival of ARM patients in lower- and middle-income countries. The present study shows that CHD is common among ARM patients, and severe CHD is associated with a lower survival rate. Hence, improved overall medical care, particularly for those with severe CHD in lower- and middle-income countries, is needed for better ARM survival.

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Authors' contributions All authors contributed to the study conception and design. AS and NLMS performed material preparation and data collection. MNMB performed the analysis. MNMB and NZ wrote the first draft of the manuscript, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Availability of data and material The data that support the findings of this study are available from the corresponding author upon reasonable request.

Code availability Not applicable.

Declarations

Ethical approval All procedures performed in studies involving human participants followed the clinical standards of the institutional and/or national research committee and the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. The study was approved by the Medical Research and Ethics Committee, Ministry of Health, Malaysia (National Medical Research Register (NMMR-21-1881-81323)).

Consent to participate Medical Research and Ethics Committee, Ministry of Health, Malaysia, waived the written informed consent.

Consent for publication Not applicable.

Competing interests All the authors declare that they have no conflict of interest. This work is original and has not been published or presented elsewhere, nor is it currently under consideration for publication elsewhere.

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