International Journal of Cardiology Congenital Heart Disease 20 (2025) 100578

Contents lists available at ScienceDirect

International Journal of Cardiology Congenital Heart Disease

journal homepage: www.journals.elsevier.com/international-journal-of-cardiology-congenital-heart-disease

Temporal change in the age at time of death in adults with congenital heart disease

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A B S T R A C T

Background: Among patients with congenital heart disease (CHD), life expectancy vary by CHD diagnosis, age of the population, and study era, making it difficult to make comparisons between studies. In this study, we aimed to address the knowledge gap regarding temporal change in age at the time of death over time across all CHD diagnoses using a large cohort of adults with CHD with longitudinal follow-up.

Methods: We studied adults with CHD that received care at Mayo Clinic from January 1, 2003 to December 31, 2023.

Results: We identified 9144 adults with CHD that received care within the study period. Of 9144 patients, 1046 (11 %) died, yielding incidence of death of 8.7 per 1000 patient-years. The incidence of death differed by CHD severity groups (7.1 versus 10.3 versus 19.3 per 1000 patient-years, $p < 0.001$, for simple versus moderate versus complex CHD, respectively). Similarly, the age at the time of death differed by CHD severity groups (66 ± 17 versus 57 ± 16 versus 43 ± 19 years, $p < 0.001$, for simple versus moderate versus complex CHD, respectively). The age at the time of death was significantly higher in the late era compared to the early era (58 ± 18 versus 50 ± 17 years $p < 0.001$), and this difference was consistent across CHD severity groups.

Conclusions: There was a temporal increase in the age at the time of death in adults with CHD, and this was consistent across CHD severity groups. The older age at the time of death suggests improved longevity in this population.

There has been significant improvements in the surgical and medical management of congenital heart disease (CHD) leading to an improved long-term survival of patients born with CHD, and ongoing increase in the population of adults with CHD [1–3]. Despite these improvements, adults with CHD still have a lower life expectancy compared to the general population [1–3]. Among patients with CHD, life expectancy vary by CHD diagnosis, age of the population, and study era, making it difficult to make comparisons between studies [1–3]. Most of the available data about longevity in adults with CHD were derived from population-based studies and administrative databases that studied outcomes across all age groups and also lack granular data regarding specific CHD diagnoses [1–3]. A prior study that was based exclusively on adults with CHD addressed these limitations, but did not assess temporal change in longevity among adults with CHD over time [4]. In this study, we aimed to address the knowledge gap regarding temporal change in age at the time of death over time across all CHD diagnoses using a large cohort of adults with CHD with longitudinal follow-up.

We reviewed the Mayo Adult Congenital Heart Disease (MACHD) Registry and identified all adults with CHD that received care at Mayo Clinic from January 1, 2003 to December 31, 2023. The Mayo Clinic institutional review board approved this study, and there are no conflicts of interest. Death was ascertained by review of medical records and confirmed using the Accurint database, which is an institutionally approval mortality database. We calculated the duration of follow-up from the first clinic visit (baseline encounter) to the date of death, last follow-up, or December 31, 2023, to determine death incidence. The age of death was stratified by era (early era [2003–2013] versus late era [2014–2023]), sex, and CHD severity groups (simple, moderate, and complex CHD).

We identified 9144 adults with CHD that received care within the study period. The mean age at baseline encounter was 38 ± 16 years, and 4484 (49 %) were males. Table 1 shows the CHD diagnoses and CHD severity groups. Of 9144 patients, 1046 (11 %) died, yielding incidence of death of 8.7 per 1000 patient-years. The incidence of death differed by CHD severity groups (7.1 versus 10.3 versus 19.3 per 1000 patient-years, $p < 0.001$, for simple versus moderate versus complex CHD, respectively). Similarly, the age at the time of death differed by CHD severity groups (66 ± 17 versus 57 ± 16 versus 43 ± 19 years, $p < 0.001$, for simple versus moderate versus complex CHD, respectively). The age at the time of death was significantly higher in the late era

Abbreviations: CHD, Congenital heart disease.

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https://doi.org/10.1016/j.ijcchd.2025.100578
Received 28 February 2025; Received in revised form 11 March 2025; Accepted 12 March 2025
Available online 13 March 2025
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Table 1
All-cause mortality stratified by CHD diagnosis.

	N	Age at baseline (y)	Death N (%)	Death (1000 PY)	Age at time of death				
					All (y)	Male (y)	Female (y)	Early era (y)	Late era (y)
TOF	1227	37 ± 14	143 (12 %)	11.7	54 ± 16	55 ± 16	53 ± 16	50 ± 16*	57 ± 15*
Ebstein	863	39 ± 16	72 (8 %)	8.3	60 ± 18	58 ± 18	62 ± 17	55 ± 20*	66 ± 13*
Pulm stenosis	466	40 ± 17	36 (8 %)	7.7	64 ± 18	63 ± 21	64 ± 17	66 ± 15	63 ± 19
PA-IVS	59	26 ± 7	5 (9 %)	8.5	31 ± 6	29 ± 8	35 ± 6	39 ± 7	34
DCRV	56	45 ± 18	5 (9 %)	8.9	79 ± 6	79 ± 7	80 ± 5	79 ± 6	–
Truncus	62	27 ± 10	10 (16 %)	18.3	38 ± 7	41 ± 8	35 ± 5	39 ± 4	38 ± 9
TGA-ASO	124	22 ± 5	0	–	–	–	–	–	–
TGA-Rastelli	38	31 ± 9	10 (26 %)	10.4	37 ± 8	36 ± 9	38 ± 5	35 ± 7*	44 ± 2*
DORV-Rastelli	58	29 ± 11	6 (10 %)	11.9	37 ± 14	32 ± 6	41 ± 15	34 ± 13	39 ± 18
COA	985	37 ± 16	111 (12 %)	8.9	57 ± 19	57 ± 20	58 ± 18	56 ± 20	58 ± 19
Mitral stenosis	19	26 ± 9	2 (11 %)	12.9	49 ± 4	46	52	49 ± 2	–
Sub AS	223	38 ± 14	17 (8 %)	7.6	54 ± 16	56 ± 17	58 ± 18	49 ± 13*	64 ± 16*
AS	962	36 ± 12	95 (10 %)	9.2	50 ± 12	52 ± 12	46 ± 12	47 ± 10	51 ± 13
Supra AS	22	27 ± 9	1 (5 %)	6.2	–	59	–	–	–
PAPVR	587	47 ± 18	62 (11 %)	6.9	68 ± 16	68 ± 15	68 ± 14	65 ± 15	71 ± 14
ASD	973	47 ± 18	78 (8 %)	4.3	69 ± 16	70 ± 16	68 ± 16	62 ± 15*	71 ± 16*
VSD	519	38 ± 16	40 (8 %)	4.9	60 ± 17	63 ± 14	54 ± 20	58 ± 17	61 ± 17
AV canal defect	443	36 ± 15	46 (10 %)	9.2	55 ± 16	54 ± 19	55 ± 14	48 ± 12*	58 ± 17*
Eisenmenger	137	41 ± 13	57 (42 %)	41.2	54 ± 15	50 ± 16	58 ± 14	49 ± 14*	60 ± 15*
Unrepaired SV	112	35 ± 16	44 (39 %)	39.4	47 ± 11	46 ± 11	47 ± 10	44 ± 16	47 ± 12
PDA	121	45 ± 20	13 (11 %)	2.8	65 ± 20	67 ± 18	64 ± 22	61 ± 24	72 ± 13
Cor triatriatum	39	56 ± 19	6 (15 %)	9.3	76 ± 22	74 ± 16	81 ± 11	72 ± 18	79 ± 20
cc-TGA	272	42 ± 15	48 (18 %)	17.6	57 ± 15	54 ± 15	61 ± 14	56 ± 13	58 ± 17
TGA-atrial switch	243	34 ± 9	26 (11 %)	14.7	41 ± 10	39 ± 9	42 ± 13	36 ± 11	43 ± 8
Fontan	534	27 ± 9	110 (21 %)	21.3	38 ± 11	37 ± 12	39 ± 10	37 ± 11	39 ± 10
CHD severity									
Simple	2081	43 ± 18	170 (8 %)	7.1	66 ± 17	67 ± 16	65 ± 18	60 ± 16*	67 ± 17*
Moderate	5424	38 ± 15	561 (10 %)	10.3	57 ± 16	57 ± 18	57 ± 17	53 ± 17*	59 ± 17*
complex	1639	32 ± 12	315 (19 %)	19.2	43 ± 19	42 ± 14	47 ± 16	43 ± 15*	47 ± 14*
All CHD	9144	38 ± 16	1046 (11 %)	8.7	55 ± 14	54 ± 18	55 ± 18	50 ± 17*	58 ± 18*

AV: Atrial ventricular; ASD: Atrial septal defect; AS: Aortic stenosis; CC: Congenital corrected; CHD: Congenital heart disease; COA: Coarctation of aorta; DCRV: Double chambered right ventricle; DORV: Double outlet right ventricle; PA-IVS: Pulmonary atresia with intact ventricular septum; PDA: Patent ductus arteriosus; PAPVR: Partial anomalous pulmonary venous return; TOF: Tetralogy of Fallot; TGA: Transposition of great arteries; VSD: Ventricular septal defect; SV: Single ventricle.
Data presented as mean ± standard deviation, and count (%).
* Signifies statistically significant difference in age at the time of death based on pair-wise comparison between males versus females or early versus late era.

compared to the early era (58 ± 18 versus 50 ± 17 years $p < 0.001$), and this difference was consistent across CHD severity groups (Table 1 and Fig. 1). However, there was no significant between-group difference in the age at the time of death between males and females.

One of the limitations of the current study is referral bias since it was based on a cohort of adults with CHD followed at a single CHD referral center, and this may affect generalizability of the results. Additionally, the database did not have data about race, ethnicity and socioeconomic class, which are known determinants of mortality.

In conclusion, the current study showed a temporal increase in the age at the time of death in adults with CHD, and this was consistent across CHD severity groups. The older age at the time of death suggests improved longevity in this population. The larger sample size of the current study allowed for stratification based on individual CHD diagnoses, thereby providing invaluable data required for patient counseling regarding life expectancy for the different CHD diagnoses in the current area. Of note, there was no difference in longevity between males versus females, which contrasts with the general population were females generally have greater longevity. Further studies required to further delineate this concept.

CRediT authorship contribution statement

Zeyad Kholeif: Writing – review & editing, Writing – original draft. **Omar Abozied:** Writing – review & editing, Writing – original draft. **Ahmed T. Abdelhalim:** Writing – review & editing, Writing – original draft. **Sara ElZalabany:** Writing – review & editing, Writing – original draft. **Amr Moustafa:** Writing – review & editing, Writing – original draft. **Ali Ali:** Writing – review & editing, Writing – original draft. **Alexander C. Egbe:** Writing – review & editing, Writing – original draft.

Disclosures

None.

Funding

Dr. Egbe is supported by National Heart, Lung, and Blood Institute (NHLBI) grants (R01 HL158517, R01 HL160761, and R01 HL162830). The MACHD Registry is supported by the Al-Bahar Research grant.

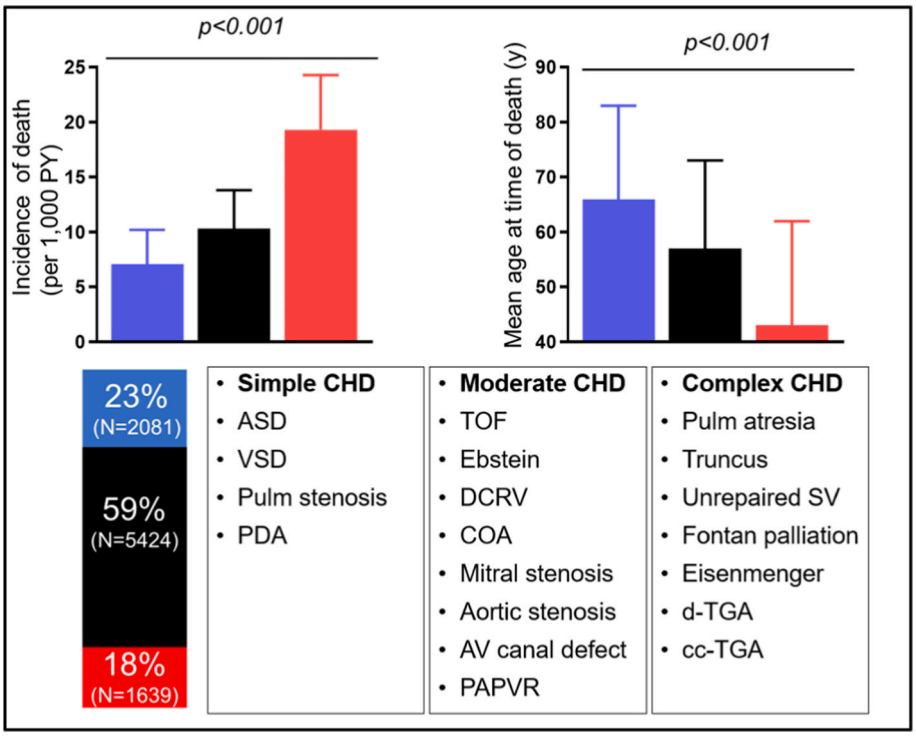

Fig. 1. All-cause mortality and congenital heart disease (CHD) severity groups. The incidence of all-cause mortality was significantly higher in patients with severe/complex CHD (red) compared to moderate CHD (black) and simple CHD (blue). Similarly, the age at the time of death was significantly lower in patients with severe/complex CHD (red) compared to moderate CHD (black) and simple CHD (blue).
The incidence of death was presented as mean and 95 % confidence interval per 1000 patient-years, while the age at the time of death was presented at mean ± standard deviation.
ASD: Atrial septal defect; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; TOF: Tetralogy of Fallot; DCRV: Double chamber right ventricle; COA: Coarctation of aorta; PA PVR: Partial anomalous pulmonary venous return; TGA: Transposition of great arteries; SV: Single ventricle.

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.

Acknowledgement

None.

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