

cambridge.org/cty

Original Article

Cite this article: Miller R, Martens T, Jodhka U, Tran J, Lion R, and Bock MJ (2022) Effects of universal critical CHD screening of neonates at a mid-sized California congenital cardiac surgery centre. *Cardiology in the Young* 32: 236–243. doi: 10.1017/S1047951121001797

Received: 3 March 2021 Revised: 4 April 2021 Accepted: 12 April 2021

First published online: 24 May 2021

Keywords

Congenital; cardiac; screen; newborn

Author for correspondence:

R. Miller, Department of Pediatrics, Loma Linda University Children's Hospital, 11175 Campus Street C/O Coleman Pavilion A1121, Loma Linda, CA, 92350, USA. Tel: +1 (949) 235-6460. E-mail: romiller@llu.edu

Effects of universal critical CHD screening of neonates at a mid-sized California congenital cardiac surgery centre

Robin Miller¹, Timothy Martens², Upinder Jodhka³, Jade Tran³, Richard Lion⁴ and Matthew J Bock³

¹Department of Pediatrics, Loma Linda University Children's Hospital, 11234 Anderson St, Loma Linda, CA 92354, USA; ²Department of Cardiothoracic Surgery, Loma Linda University Children's Hospital, 11234 Anderson St, Loma Linda, CA 92354, USA; ³Division of Cardiology, Loma Linda University Children's Hospital, 11234 Anderson St, Loma Linda, CA 92354, USA and ⁴Division of Critical Care Medicine, Loma Linda University Children's Hospital, 11234 Anderson St, Loma Linda, CA 92354, USA

Abstract

Introduction: CHD affects over 1 million children in the United States. Studies show decreased mortality from CHD with newborn cardiac screening. California began a screening programme on 1 July, 2013. We evaluated the effect of mandatory screening on surgical outcomes at Loma Linda University Children's Hospital since 1 July, 2013. Methods: We evaluated all infants having congenital heart surgery at Loma Linda University Children's Hospital between 1 July, 2013 and 31 December, 2018. Primary target diagnoses include hypoplastic left heart syndrome, pulmonary atresia with intact ventricular septum, tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great arteries, tricuspid atresia, and truncus arteriosus. Secondary target diagnoses include aortic coarctation, double outlet right ventricle, Ebstein anomaly, interrupted aortic arch, and single ventricle. Patients were stratified by timing of diagnosis (pre-screen, screen positive, and screen negative). Primary end points were post-operative length of stay, operative mortality, absolute mortality, and actuarial survival. Results: The cohort included 274 infants. Of these, 79% were diagnosed prior to screening (46% prenatally). Only 38% of those screened were positive, with 13% of the cohort having a "missed diagnosis." Conclusions: Primary targets were more likely to be diagnosed by screening (53%), while secondary targets were unlikely to be diagnosed by screening (10%) (p = 0.004). Outcomes such as length of stay, operative mortality, and actuarial survival were not different based on timing of diagnosis (p > 0.05). Despite late diagnosis, those not diagnosed until after screening did not have adverse outcomes.

CHD is a condition that affects about one million children in the United States. A simple test involving placing a pulse oximeter and measuring pre- and post-ductal oxygen saturations can be easily implemented in the newborn nursery to screen for CHD. Some studies have shown a decrease in mortality from CHD with the implementation of newborn cardiac screening. In fact, an observational study published in the Journal of the American Medical Association in 2017 found that states that had mandated newborn cardiac screening had seen up to a 33% decrease in death rate due to critical CHD compared to states without mandatory screening. ² California implemented such a newborn screening programme on 1 July, 2013.³ As of 2018, all 50 states had enacted similar bills. 4 Current CHD screening recommendations are to follow the American Academy of Pediatrics algorithm outlined by Kemper et al⁵ (Fig 1). This algorithm calls for screening to do be done at or after 24 hours of life. One pulse oximeter is placed on the right hand, and another on either foot. The screen is considered passed if the patient has 95% or greater oxygen saturation in the right hand or in the foot, with 3% or less difference between the hand and foot readings. If the patient has an oxygen saturation of less than 90% in either the hand or the foot, the screen is considered failed and the patient should be further evaluated and an echocardiogram should be performed. If the patient has an oxygen saturation between 90 and 95% in either the hand or the foot or greater than 3% difference in oxygen saturation between the hand and foot pulse oximetry, they should be re-screened after 1 hour up to two times before this test is considered a failed screen. A modification to this algorithm reducing re-screening was published in 20,186 (Fig 2). It should be noted that the cardiac screen is to be used in conjunction with prenatal ultrasounds as well as clinical observation within the first year of life. In this study, we will evaluate the efficacy of newborn cardiac screening on surgical outcomes of the CHD programme at Loma Linda University Children's Hospital.

© The Author(s), 2021. Published by Cambridge University Press.



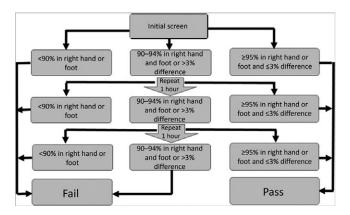


Figure 1. Original critical CHD screening algorithm (Kemper, 2011).⁵

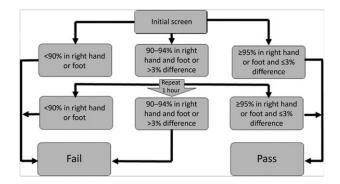


Figure 2. Modified critical CHD screening algorithm (Diller, 2018).⁶

Purpose

We aim to evaluate the influence of a mandatory, state-wide newborn cardiac screening programme on outcomes of congenital heart surgery at Loma Linda University Children's Hospital since 1 July, 2013.

Hypothesis

We hypothesise that infants with critical CHD requiring surgery within the first year of life that are not diagnosed prior to or at the neonatal cardiac screen have worse short-term outcomes and long-term survival, compared to those with diagnosis prior to or at the neonatal cardiac screen.

Methods

In this study, we evaluated infants (<1 year of age) who had congenital heart surgery at Loma Linda University Children's Hospital between 1 July, 2013 and 31 December, 2018. Only patients with primary or secondary target diagnoses (Table 1) were included in the analysis. Primary lesions included hypoplastic left heart syndrome, pulmonary atresia with an intact ventricular septum, tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great arteries, tricuspid atresia, and truncus arteriosus. Secondary lesions included coarctation of the aorta/arch hypoplasia, double outlet right ventricle, Ebstein anomaly, interrupted aortic arch, and other single ventricle lesions (such

as double inlet left ventricle, atrioventricular canal defect, and single left ventricle). Lesions were chosen based on recommendations from the United States Secretary of Health and Human Services Secretary's Advisory Committee on Heritable Disorders in Newborns. Patients with the most common cardiac lesion identified by screening [total anomalous pulmonary venous return] underwent subanalysis comparison to a historical cohort (1 January, 2008–31 December, 2012) of total anomalous pulmonary venous return patients, prior to mandatory screening, in order to determine if a reduction in mortality occurred in that group.

Demographic and clinical outcome data were collected on each patient (Table 2). The primary end points were post-operative length of stay, operative mortality, absolute mortality, and actuarial survival. The patients were stratified for analysis by method and timing of cardiac diagnosis. Categorical data are presented as number (#) and percentage (%), while continuous data are presented as median and interquartile range. The chi-square test was used to assess differences in categorical variables. The Kruskal–Wallis test was used to assess differences in continuous variables. Kaplan–Meier survival analysis with log-rank test comparison of the study groups was performed as well. R: A Language and Environment for Statistical Computing 3.4.2 (R Foundation for Statistical Computing, Vienna, Austria, 2017) was used to perform statistical calculations.

Results

Cohort

Two hundred eighty-one (281) infants (less than 1 year of age) with critical CHD undergoing cardiac surgery at Loma Linda University Children's Hospital between 1 July, 13 and 31 December, 18 were identified. Seven had incomplete information for the primary discriminator (method of diagnosis) and were excluded, leaving 274 patients which comprised the study cohort.

Method of diagnosis

Two hundred sixteen patients (79%) were diagnosed before screening, either prenatally or postnatally before screening was performed (Table 3). One hundred twenty-seven patients (46%) were diagnosed prenatally. Eighty-nine patients (33%) were diagnosed clinically after birth, prior to being screened for critical CHD.

The remaining 58 patients (21%) were screened and were either diagnosed at the time of screening or later. Twenty-two patients (8% of all patients; 38% of the patients who were screened) were diagnosed by screening, while 36 patients (13% of all patients, 62% of those who were screened) tested negative and were not diagnosed until after screening. The sensitivity of pulse oximetry screening to detect critical CHD, not accounting for those who did not undergo surgery, was 38%.

Screen target and lesions

Overall, 69% (190) of patients had primary screen targets, while 31% (84) had secondary screen targets (Table 1).

Similar percentages of primary and secondary screen targets were detected prior to screening (80% primary targets versus 76% secondary targets). Of the patients that were diagnosed by screening, most (20/22, 91%) had primary lesions. Approximately half (20/38, 53%) of undiagnosed primary lesions were detected by screening. By contrast,

238 R. Miller et al.

Table 1. All infants with critical CHD lesions undergoing surgery at Loma Linda University children's hospital between 1 July, 2013 and 31 December, 2018

Screen targets and lesions									
Variable	All (n = 274)		Prior to screen (n = 216)		Screen Pos (n = 22)		Screen Neg (n = 36)		
	#	%	#	%	#	%	#	%	р
Screen target									0.004
Primary	190	0.69	152	70	20	91	18	50	
Secondary	84	0.31	64	30	2	9	18	50	
Screen lesion									<0.001
Hypoplastic left heart syndrome	38	0.14	30	14	2	9	6	17	
Pulmonary atresia/intact ventricular septum	11	0.04	9	4	0	0	2	6	
Tetralogy of Fallot	63	0.23	56	26	2	9	5	14	
Total anomalous pulmonary venous return	32	0.12	17	8	11	50	4	11	
Transposition of the great arteries	34	0.12	29	13	4	18	1	3	
Tricuspid atresia	4	0.02	4	2	0	0	0	0	
Truncus arteriosus	8	0.03	7	3	1	5	0	0	
Coarctation/arch hypoplasia	42	0.15	25	12	2	9	15	42	
Double outlet right ventricle	24	0.09	23	11	0	0	1	3	
Ebstein anomaly	0	0.00	0	0	0	0	0	0	
Interrupted aortic arch	6	0.02	5	2	0	0	1	3	
Single ventricle	12	0.04	11	5	0	0	1	3	

only two patients (2/20, 10%) with undiagnosed secondary lesions were detected by screening. Most cardiac lesions were detected prior to screening at similar rates (76%–96%) with the exceptions of total anomalous pulmonary venous return (17/32, 53%) and coarctation/ arch hypoplasia (25/42, 60%) which were under-detected prior to screening. A majority of undiagnosed total anomalous pulmonary venous return (11/15, 73%) and transposition of the great arteries (4/5, 80%) were detected by screening. A majority of undiagnosed coarctation/arch hypoplasia (15/17, 88%), hypoplastic left heart syndrome (6/8, 75%), and tetralogy of Fallot (5/7, 71%) were missed by screening. Patients missed by screening were generally born at outside hospitals before either being transferred to Loma Linda University Children's Hospital's NICU or presenting to the emergency department, so there is limited knowledge as to how they passed their newborn cardiac screen.

Demographics and outcomes

Basic demographic information (gender, race/ethnicity, pre-maturity, and birth weight) were similar between the different types of diagnoses (Table 2). Those patients with lesions not detected until after screening were older at diagnosis (1 day versus 33.5 days, p-value <0.001) and were larger (3.55 kg versus 4.071 kg, p-value 0.003) and older at surgery (11.5 days versus 48.5 days, p-value <0.001). Post-operative LOS (p-value 0.141), operative mortality (p-value 0.230), absolute mortality (p-value 0.282), and actuarial survival (log-rank test p-value 0.3) were not different based on type of diagnosis (Tables 2 and 4; Fig 3). Three deaths (3/58, 5%) occurred in patients undergoing screening (Table 4). Two patients with hypoplastic left heart syndrome who were not diagnosed by

screening died after surgery (2/36, 6%), while one patient with transposition of the great arteries diagnosed by screening died after surgery (1/22, 5%). Reoperation was more common in those diagnosed by screening (27% versus 0%, p-value 0.003).

Total anomalous pulmonary venous return subanalysis

Total anomalous pulmonary venous return was the most common lesion diagnosed by screening (11/22, 50% of all patients diagnosed by screen and; 11/15, 73% of total anomalous pulmonary venous return patients undergoing testing) (Table 1). A subanalysis was performed to determine whether a decrease in mortality occurred in patients with total anomalous pulmonary venous return undergoing surgery, compared to those who would have reached screening in the preceding era (2008–2013) without mandatory newborn critical CHD screening. There were 11 total anomalous pulmonary venous return patients in the prior era with a late diagnosis (Table 5). Similar to the primary analysis, no differences in demographics were seen, and similar trends in diagnosis age and weight and age at surgery were seen. No deaths occurred in either era in this subpopulation; therefore survival analysis could not be undertaken.

Discussion

Our study addresses the impact of a mandatory, state-wide critical CHD pulse oximetry newborn screening programme on congenital heart surgery outcomes at a mid-sized California children's hospital. We failed to find evidence that such a programme has improved surgical outcomes or survival in those

Table 2. All infants with critical CHD lesions undergoing surgery at Loma Linda University Children's Hospital between 1 July, 2013 and 31 December, 2018

		Prior to scree					Screen	_		
	_	ll (n = 274)		(n = 216	<u> </u>	(n = 2	<u> </u>	(n = 3	<u> </u>	
	= 274 #	%		#	%	#	%	#	%	р
. ,	263 56			49	24	4	18	3	9	0.118
Gender (female)	98	36	5	79	37	8	36	11	31	0.783
	273									0.70
White/Caucasian	63	23	3	51	24	6	27	6	17	
Hispanic/Latino	15	1 56	5 1	17	54	15	68	22	61	
African American	19	7		17	8	0	0	2	6	
Asian	8	3		7	3	0	0	1	3	
Other/unknown	29	11	1	23	11	1	5	5	14	
Ethnicity 2	273									0.34
Caucasian	63	23	3	51	24	6	27	6	17	
Hispanic/Latino	15:	5 57	7 1	.18	55	15	68	22	61	
Other/unknown	55	20)	46	21	1	5	8	22	
Born at Loma Linda (Y)	262 12	7 49	9 1	.23	59	3	14	1	3	<0.0
STAT category										0.03
1	24	9		17	8	1	5	6	17	
2	40	15	5	26	12	3	14	11	31	
3	19	7		18	8	0	0	1	3	
4	14	 3 5 ⁴	4 1	.21	56	14	64	13	36	
5	43	16		34	16	4	18	5	14	
Age category at surgery										<0.0
Neonate (0–30 days)	170		2 1	.43	66	15	68	12	33	
Infant (31–365 days)	10-	4 38	3	73	34	7	32	24	67	
Mortality (Y)	30			27	13	1	5	2	6	0.28
Operative death (Y)	24			 22	10	1	5	1	3	0.23
Reoperation (Y)	34			28	13	6	27	0	0	0.00
Variable	n = 274	med	IQR	med	IQR	med	IQR	med	IQR	p-valı
Gestational age (weeks)	255	39	37–39	38	37–39	39	39	39	38-40	0.02
Birth weight (kg)	248	3.025	2.6-3.4	3	2.6–3.4	3.31	2.7–3.3	3.03	2.7–3.5	0.10
Weight at surgery (kg)	273	3.5	3.0-4.3	3.43	3.0-4.1	3.55	3.3-4.0	4.071	3.5–5.0	0.00
Height at surgery (cm)	254	50.65	48-54	50	48-52.5	51	50-54	55	52–59	<0.00
Age at diagnosis (postnatal only) (d		1	1–3.25	1	1-2	1	1-2	33.5	8–79	<0.00
Age at unagnosis (postnatationty) (di	~,~, 170	17	7–60	14	7–58.5	11.5	6–43	48.5	18-88	<0.00
NICU LOS (days)	257	19	11–46	22	13-53	14	9–19.75	0	0-19	<0.00
Pre-operative LOS (days)	231									••••••
		7	2-13	7	3–18.25	4	0.3-8.8	4	2-8	0.0
Post-operative LOS (days)		9	5–20	9	5–28	8.5	4.5-11.8	7.5	5–12	0.14

240 R. Miller et al.

Table 3. All infant critical CHD surgeries at Loma Linda University Children's Hospital between 1 July, 2013 and 31 December, 2018

Diagnosis timing and screening						
	All (n = 274)					
Variable	#	%				
Diagnosis timing						
Prenatal	127	46				
Postnatal, pre-screen, pre-DC	89	33				
Postnatal, screen fail, pre-DC	22	8				
Postnatal, screen passed, pre-DC	5	2				
Postnatal, screen passed, post-DC	31	11				
Diagnosis type						
Diagnosed prior to screen	216	79				
Screened	58	21				
Diagnosed by screen	22	8 (38 of those screened)				
Missed screen diagnosis	36	13 (62 of those screened				

Table 4. All infants with critical CHD lesions undergoing surgery at Loma Linda University Children's Hospital between 1 July, 2013 and 31 December, 2018

Mortality by screen lesion									
	All (n = 30)		Prior to screen (n = 27)		Screen Pos (n = 1)		Screen Neg (n = 2)		
Variable	#	%	#	%	#	%	#	%	p-value
Mortality by screen lesion									
Hypoplastic left heart syndrome (n = 38)	10	33	8	30	0	0	2	100	0.91
Pulmonary atresia/intact ventricular septum (n $=$ 11)	1	3	1	4	0	0	0	0	0.59
Tetralogy of Fallot (n = 63)	5	17	5	19	0	0	0	0	0.73
Total anomalous pulmonary venous return (n = 32)	2	7	2	7	0	0	0	0	0.49
Transposition of the great arteries (n = 34)	5	17	4	15	1	100	0	0	0.4
Tricuspid atresia (n = 4)	0	0	0	0	0	0	0	0	0.5
Truncus arteriosus (n = 8)	2	7	2	7	0	0	0	0	0.63
Coarctation/arch hypoplasia (n = 42)	1	3	1	4	0	0	0	0	0.7
Double outlet right ventricle (n = 24)	4	13	4	15	0	0	0	0	0.58
Ebstein anomaly (n = 0)	0	0	0	0	0	0	0	0	NA
Interrupted aortic arch (n = 6)	0	0	0	0	0	0	0	0	0.5
Single ventricle (n = 12)	0	0	0	0	0	0	0	0	0.5

diagnosed by screening, compared to those diagnosed after screening or initial hospital discharge. Additionally, we find only a 38% sensitivity of the screening to detect critical CHD. Finally, we find significantly lower sensitivity of screening to detect patients with hypoplastic left heart syndrome (25%), tetralogy of Fallot (29%), and coarctation/arch hypoplasia (12%) than expected.

Early studies regarding newborn pulse oximetry screening focused on testing sensitivity and specificity, feasibility, and cost-benefit analysis^{7,8}. In 2009, a joint scientific statement from the American Heart Association and American Academy of Pediatrics was published examining the role of pulse oximetry screening,⁹ which was followed by a 2011 publication providing strategies for implementing such screening.⁵ In 2011, the America Academy of Pediatrics endorsed the Health and Human Services recommendation for mandatory pulse oximetry screening.¹⁰

Since the adoption of mandatory screening by all states, few publications have assessed the impact of such screening. Abouk

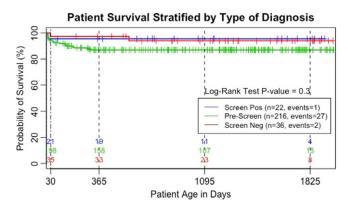


Figure 3. Kaplan-Meier survival curve for patients stratified by type of diagnosis.

et al. reviewed all live births in states with mandatory screening policies from 2007 to 2013 and found a 33.4% reduction in early infant deaths due to critical CHD over time.² More recently, Campbell and colleagues evaluated surgical outcomes over a 2-year period with mandatory screening and found no difference in outcomes or mortality, when compared to a 2-year period prior to screening.¹¹ Our study corroborates Campbell's findings. We evaluated survival in two ways. We evaluated for differences in survival between those diagnosed by screening and those with missed screens and we evaluated for a survival difference in the most common lesion diagnosed by screening (total anomalous pulmonary venous return) compared to a historical cohort (2008–2013 versus 2013-2018). We found no difference in survival between those diagnosed at screening (95% survival) versus those with missed screens (94% survival). Additionally, there were no deaths of patients with total anomalous pulmonary venous return in either study period.

There may be several reasons behind the failure to find a difference in survival. First, this study is only evaluating those infants who undergo cardiac surgery in the first year of life. Those infants who do not undergo surgery due to death or severe morbidities limiting surgical options are not included in this analysis. If an improvement in survival is present, it could be due to improvement in this population of patients. Additionally, recent increases in prenatal diagnoses of critical CHD (46% in our study; 78% in Campbell et al) decrease the number of newborns requiring screening, which would also decrease the overall utility of screening. Finally, in both cohorts, only a small fraction of the total cohort underwent screening (21% in our study; 4% in Campbell et al) due to the combination of prenatal diagnosis and detection due to clinical findings prior to screening. We speculate that those with symptomatic critical CHD allowing for clinical diagnosis prior to hospital discharge have worse disease and would be expected to have a higher mortality, when compared to those who remain asymptomatic through initial hospital discharge and undergo

Our study has a number of advantages over the reports by Abouk and Campbell. Like Campbell's, our study utilises a retrospective chart review and registry, as opposed to Abouk who utilised administrative data. Campbell notes the inferiority of this type of data. Our study also limits inclusion criteria to those requiring cardiac surgery prior to 1 year of age and those lesions specifically noted in the recommendations, which are further categorised into primary and secondary targets, consistent with state and

published guidelines.³ Campbell et al includes all neonates (less than 1-month-old) with certain exclusionary cardiac lesions. This study also includes patients over a 5.5-year period, compared to only 2 years in the Campbell study. These factors allow our cohort to most closely resemble the group of newborns targeted for screening, compared to other published data.

In addition to a lack of benefit in mortality, we found lower sensitivity of screening to detect target lesions compared to prior reports. The 2009 American Heart Association and American Academy of Pediatrics scientific statement provides a range of overall sensitivities of 50%-100% with a combined sensitivity of 75%. Our sensitivity of 38% is much lower. The principal lesions accounting for this low sensitivity are hypoplastic left heart syndrome (25%), tetralogy of Fallot (29%), and coarctation/arch hypoplasia (12%). Prior studies report 100% sensitivity of diagnosing hypoplastic left heart syndrome, with 69% and 53% sensitivity to detect tetralogy of Fallot and coarctation/arch hypoplasia, respectively. The two deaths in those with missed diagnoses occurred in patients with hypoplastic left heart syndrome. The significant difference in sensitivities may relate to problems with scaling results of small studies to facilitate widespread implementation. It is important to note that Loma Linda University Medical Center is a major referral institution, and many of the infants came to Loma Linda for cardiac surgery from outside hospitals with lower than average efficacy of the newborn cardiac screen. Loma Linda's clinic staff is highly sensitive at identifying infants with critical CHD; therefore, many of the infants at Loma Linda University Children's Hospital were identified prior to the newborn cardiac screen. The newborn cardiac screen was designed for institutions that do not have the same resources as a large referral centre. It should be noted that the newborn cardiac screen plays an important role in the community hospital setting, especially in places with poor prenatal care, poor access to healthcare, and in institutions without a paediatric cardiology programme. Additionally, the newborn cardiac screen is less sensitive if performed before 24 hours of life. If the screen was performed early, the primary paediatrician should consider repeating the screen at the first newborn appointment.

Conclusions

In our cohort, we found only a 38% sensitivity of mandatory newborn critical CHD screening to detect target cardiac lesions requiring surgery in the first year of life among patients who were not diagnosed prenatally. A high proportion of newborns with hypoplastic left heart syndrome, tetralogy of Fallot, and coarctation/arch hypoplasia remained undiagnosed after screening. In communities where prenatal screening is less common, the pulse oximeter screening may have a greater impact than it does at a large referral centre with more resources and more consistent prenatal screening. Despite diagnosis by screening, there were no differences in outcomes or survival compared to those missed by screening among patients who underwent cardiac surgery. Mandatory newborn critical CHD pulse oximetry screening can detect newborns with critical heart disease but should not replace traditional methods of monitoring in the fetal, newborn, and infant periods. Careful clinical examination at routine office visits, including auscultation for murmur, pulse oximetry, and palpation of pulses, remains vital in the diagnosis of infants with critical CHD.

242 R. Miller et al.

 Table 5. All infants with TAPVR undergoing CCHD screening or late diagnosis prior to surgery at LLU between 1 January, 2008 and 31 December, 2018

TAPVR stratified by era of diagnosis					
Demographics and outcomes					
	ERA "08-13" (n	= 11)	ERA "13-18"	(n = 15)	
Variable	#	%	#	%	р
Diagnosis timing					<0.00
Postnatal, screen fail, pre-DC	NA	NA	11	73	
Postnatal, screen passed, post-DC	NA	NA	4	27	
Pre-mature (Y/N)	0	0	3	20	0.23
Gender (female)	5	46	5	33	0.68
RACH score					0.25
1	0	0	0	0	
2	8	73	7	47	
3	0	0	2	13	
4	2	18	6	40	
5	0	0	0	0	
6	1	9	0	0	
ARISTLE basic level					0.23
1	0	0	0	0	
2	0	0	3	20	
3	3	100	12	80	
4	0	0	0	0	
STAT category					0.67
1	0	0	1	7	0.01
2	0	0	0	0	
3	0	0		0	
		91	-		
4	10		14	93	
5	1	9	0	0	
Age category at surgery					0.21
Neonate (0–30 days)	2	18	7	47	
Infant (31–365 days)	9	82	8	53	
Reoperation (Y/N)	0	0	2	13	0.11
Readmission with 30 days (Y/N)	1 Madian	9	1	7	1.00
Variable Weight at surgery (kg)	Median	IQR	Median	1QR 3.2-4.5	p
	4.9	4.4–5.6	3.6		0.03
Height at surgery (cm)	60	53-62	54	50-58	0.12
ARISTLE basic score	9	9–9	9	6.8-9	0.06
STAT score	1.9	1.9–1.9	1.9	1.9–1.9	0.30
Cardiopulmonary bypass time (minutes)	110	101–121	128	114–149	0.05
Aortic cross-clamp time (minutes)	49	45.5–57	51	37–71	0.81
Deep hypothermic cardiac arrest time (minutes)	0	0–2	0	0–0	0.06
Surgical admission age (days)	117	52–126	27	2–46	0.00
Age at surgery (days)	122	56-128	37	5.5-53	0.00
Current age (days)	3525	3078–3976	1081	680-1647	<0.00
Pre-operative length of stay (days)	3	1–5	2	0–5	0.38
Post-operative length of stay (days)	4	3–9	8	6.5–10	0.03
Total surgical admission length of stay (days)	7	6–14	11	9-15.5	0.09

Acknowledgements. None.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of interest. None.

Ethical standards. This article does not contain any studies involving human participants. All institutional ethical protocols were followed.

Limitations. Only those patients who underwent cardiac surgery are reported. Patients who died or did not undergo surgery in the infant period were not included. No conclusions can be drawn about the screening test as a whole (false positives/negatives, specificity), since only a subgroup of those tested were included (i.e. those who underwent cardiac surgery).

References

- Gilboa SM, Devine OJ, Kucik JE, et al. Congenital heart defects in the United States: estimating the magnitude of the affected population in 2010. Circulation 2016; 134: 101–109.
- Abouk R, Grosse SD, Ailes EC, Oster ME. Association of US state implementation of newborn screening policies for critical congenital heart disease with early infant cardiac deaths. JAMA 2017; 318: 2111–2118.
- AB-1731. Newborn screening program: critical congenital heart disease, Assembly Bill No. 1731. (2011–2012).

- Glidewell J, Grosse SD, Riehle-Colarusso T, et al. Actions in support of newborn screening for critical congenital heart disease — United States, 2011– 2018. MMWR Morb Mortal Wkly Rep 2019; 68: 107–111. DOI: http://dx. doi.org/10.15585/mmwr.mm6805a3.
- Kemper AR, Mahle WT, Martin GR, et al. Strategies for implementing screening for critical congenital heart disease. Pediatrics 2011; 128: e1259–e1267.
- Diller CL, Kelleman MS, Kupke KG, Quary SC, Kochilas LK, Oster ME. A modified algorithm for critical congenital heart disease screening using pulse oximetry. Pediatrics 2018; 141: e20174065.
- Hoke TR, Donohue PK, Bawa PK, et al. Oxygen saturation as a screening test for critical congenital heart disease: a preliminary study. Pediatr Cardiol 2002; 23: 403–409.
- Koppel RI, Druschel CM, Carter T, et al. Effectiveness of pulse oximetry screening for congenital heart disease in asymptomatic newborns. Pediatr 2003; 111: 451–455.
- Mahle WT, Newburger JW, Matherne GP, et al. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Pediatr 2009; 124: 823–836.
- Mahle WT, Martin GR, Beekman RH, et al. Endorsement of health and human services recommendation for pulse oximetry screening for critical congenital heart disease. Pediatr 2012; 129: 190–192.
- 11. Campbell MJ, Quarshie WO, Faerber J, Goldberg DJ, Mascio CE, Blinder JJ. Pulse oximetry screening has not changed timing of diagnosis or mortality of critical congenital heart disease. Pediatr Cardiol 2020; 41: 1–6.