THE STATUS OF CRITICAL CONGENITAL HEART DEFECT INFANT DEATHS AFTER PULSE-OXIMETRY SCREENING MANDATES

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ABSTRACT

In 2013, 29.5% of infants born with non-syndromic critical congenital heart defects were undiagnosed through at least 3 days of life, meaning that these infants with severely low blood-oxygen levels, known as birth asphyxia, were left without needed treatment until they were diagnosed. Previous research has shown that newborn deaths caused by CCHDs dropped by 33% in 8 states after they implemented mandatory pulse-oximetry screenings in the first 24 - 48 hours of life. However, estimates of the potential effects of the expansion of mandatory pulse-oximetry screening throughout all 50 states are unknown, as are the potential effects of mandating other CCHD-detecting measures, such as fetal echocardiograms. This study utilizes data from the *Center for Disease Control* on infant deaths due to CCHDs to estimate the true effect of the expansion of this mandatory measure in the US. Building from a previous study conducted between 2007 and 2013 (1), this study expands to include data from 2007 - 2019. During this period, results indicated a decline in the early infant death rate due to CCHDs associated with nationwide mandatory screening policy implementations. Concurring with the aforementioned study, mandatory screening was associated with a reduction in CCHD infant deaths.

Keywords: policy research; public policy; cardiology; screening; quality improvement

INTRODUCTION

Immense changes in health policy for the improvement of congenital heart defect (CHD) detection have occurred during the past decade. CHDs are any heart defects present at birth that are a result of the improper formation of the heart. More specifically, a critical congenital heart defect (CCHD) is classified as a severe CHD incompatible with life which requires surgical or procedural intervention within the first year of life. With almost 1% of all people being born with a CHD and 4.2% of all deaths in the first 27 days of life attributable to CHDs (1), effective detection and treatment of these conditions are imperative in saving lives.

Prior to 2011, there were no policies in the United States (U.S.) that mandated CHD screenings in newborns. Many CHD and CCHD cases have gone undetected by healthcare providers, possibly resulting in long-term negative health outcomes and, in some cases, newborn death due to these conditions (2). The impact of this issue has led to the implementation of policies in all 50 states and D.C. requiring healthcare providers to administer pulse-oximetry tests to newborns within 24 - 72 hours of life (3). These tests measure the level of oxygen carried by red blood cells in a patient's bloodstream, which allows healthcare providers to detect

possible defects in a newborn's ability to effectively move oxygen throughout the body. Low oxygen saturation in the blood is an indicator of a potential CHD and can be used as an indicator in determining if a patient requires further testing (4).

A previous study examined 8 states that had implemented mandatory pulse-oximetry screenings for all newborns between August 1, 2011, and June 1, 2013. The scholars found that the implementation of these mandates was associated with a 33.4% decrease in infant deaths due to CCHDs (5). Building from Abouk et al. findings, this study explores the association of CCHD-related infant deaths in the U.S. with the time period before any state-level mandates were implemented (2007 – 2011) and the time period during which the state-level mandates were implemented (2012 – 2019). We hypothesize that after states implemented testing mandates, there is a decline in CCHD deaths when compared against the time period prior.

METHODS

Study Design

A time-series analysis was posited to evaluate CCHD and CCHD-related deaths in the U.S. between January 1, 2007 and December 31, 2019. Aggregate state-level data from the Centers for Disease Control and Prevention (CDC) WONDER online database were used (6). Since all data related to this study were gathered using retrospective, publicly accessible means, no human subjects or non-anonymous data were utilized.

Data

The CDC's WONDER database provided total live births, CCHD live births, and CCHD infant deaths (<1 year of age) from 2007 to 2019. Mirroring the Abouk et al. study, 2007 was the initial year and 2019 was the ending year based on the availability of data at the time this study was completed. This data, produced by the National Center for Health Statistics, was measured at state-level and aggregated to a national level. The CDC suppresses data points when there are 10 or fewer deaths in a given state/year. With only 8 states (CA, FL, MI, NY, NC, OH, TX, VA) reaching at least ten infant deaths related to CCHDs in every year of the study, there were limitations in analyzing the state-level data. Therefore, an aggregation of state-level data to the national level and regional level was used to prevent gaps resulting from suppressed information.

Policies

State screening policy mandate and implementation dates were derived from Abouk et al. and a U.S. Department of Health and Human Services report (7). Due to time lags between the mandating of screening requirements and the universal implementation of such policies within each state, universal state-wide policy implementation dates were strictly used as the initial and end dates for the time periods compared in this study. A compilation of the policy implementation dates provided by the CDC is available (7). Figure 1 provides an illustration of these policies implemented by state overtime.

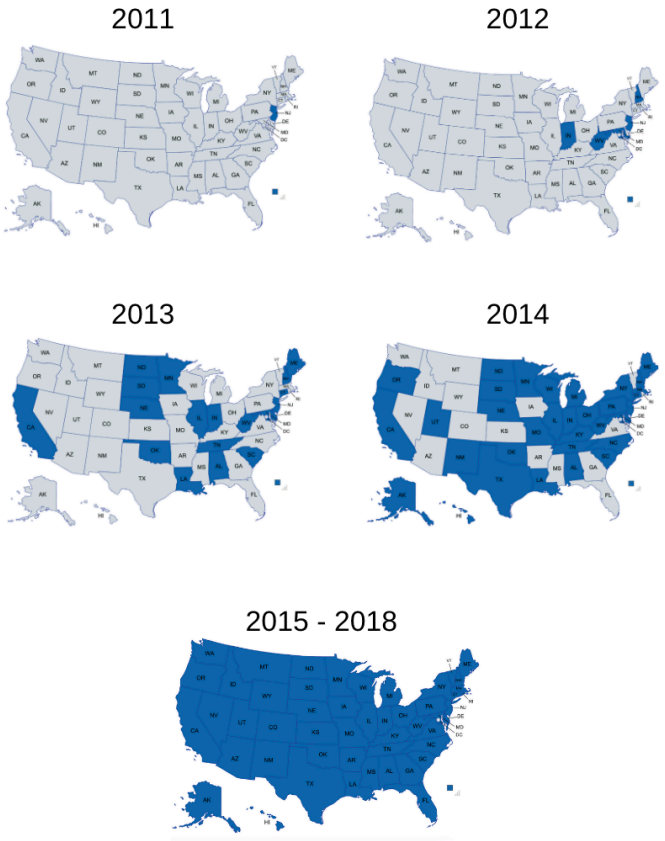

Figure 1. Infant critical congenital heart disease pulse oximetry screening state policy implementation by time period.

Outcomes Measure

Through the WONDER database, ICD-10 codes identified by the National Library of Medicine's Newborn Screening Coding and Terminology Guide as relating to CCHDs were utilized. Data for births and deaths involving infants diagnosed with conditions falling under specific ICD-10 Codes used in this study are available in Table 1.

ICD Code	Underlying Cause of Death
Q20.0	Common arterial trunk
Q20.1	Double outlet right ventricle
Q20.3	Discordant ventriculoarterial connection
Q20.4	Double inlet ventricle
Q21.3	Tetralogy of Fallot
Q22.0	Pulmonary valve atresia

Q22.4	Congenital tricuspid stenosis
Q22.5	Ebstein anomaly
Q23.4	Hypoplastic left heart syndrome
Q25.1	Coarctation of aorta
Q25.2	Atresia of aorta
Q25.4	Other congenital malformations of aorta
Q26.2	Total anomalous pulmonary venous connection

Table 1. ICD-10 codes for underlying causes of death.

Statistical Analysis

Data used in the statistical analysis were taken at a state level and aggregated by birth year and state of birth to the national level. Early infant deaths due to CCHDs were calculated for each year as mortality rate (deaths due to CCHDs per 100,000 live births) to control for variances in birth rates through the years. Data were broken into two groups - data for the years of 2007 through 2011 were included in the "pre-implementation" group, and data for the years of 2012 through 2019 were included in the "post-implementation" group. Although one state, New Jersey, had implemented a screening policy in 2011, the data for this year were included in the pre-implementation data group because the policy was not implemented until August 31, 2011, making it active for less than one-half of a full calendar year in 2011 and therefore not accurately representative of the entire year. Summary statistics relating to the dependent and independent variables are presented in Table 2. Additionally, Table 3 provides infant CCHD deaths by year and census region.

Year	U.S. Births	All Infant Deaths	CCHD Infant Deaths	CCHDs as % of Infant Deaths	# of States Mandating Policy
2007	4147997	29138	592	2.03%	0
2008	4132735	28059	559	1.99%	0
2009	4003587	26412	506	1.92%	0
2010	3944153	24586	453	1.84%	0
2011	3996537	23985	498	2.08%	1
2012	3943077	23629	476	2.01%	4
2013	3941783	23440	482	2.06%	14
2014	3948350	23215	464	2.00%	16
2015	3978038	23455	490	2.09%	11
2016	3970145	23161	476	2.06%	2
2017	3939295	22335	452	2.02%	1
2018	3848208	21467	372	1.73%	2

2019	3783052	20921	422	2.02%	0
2007-2019	51576957	313803	6242	1.99%	51

Table 2. Summary statistics relating to the dependent and independent variables.

Year	Northeast	Midwest	South	West
2007	65	143	210	174
2008	75	127	220	137
2009	64	123	192	127
2010	58	105	185	105
2011	61	114	199	124
2012	45	115	191	125
2013	65	114	196	107
2014	44	111	193	116
2015	50	101	224	115
2016	56	100	214	106
2017	51	113	196	92
2018	34	89	155	94
2019	48	83	184	107
2007 - 2019	716	1,438	2,559	1,529

Northeast *– Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island, Vermont, New Jersey, New York, Pennsylvania*
Midwest *– Indiana, Illinois, Michigan, Ohio, Wisconsin, Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, South Dakota*
South *– Delaware, D.C., Florida, Georgia, Maryland, North Carolina, South Carolina, Virginia, West Virginia, Alabama, Kentucky, Mississippi, Tennessee, Arkansas, Louisiana, Oklahoma, Texas*
West *– Arizona, Colorado, Idaho, New Mexico, Montana, Utah, Nevada, Wyoming, Alaska, California, Hawaii, Oregon, Washington*

Table 3. CCH deaths by census region.

A linear analysis was postulated to better understand the outcome variable of interest - the number of CCHD deaths in each given year. Additionally, average counts for the pre- and post-implementation time periods allowed for the change in in the outcome variable to be calculated. Said differently, the linear trend for annual deaths due to CCHDs in the pre-implementation group (Figure 2) was calculated and compared to the linear trend for annual national deaths due to CCHDs in the post-implementation group (Figure 3).

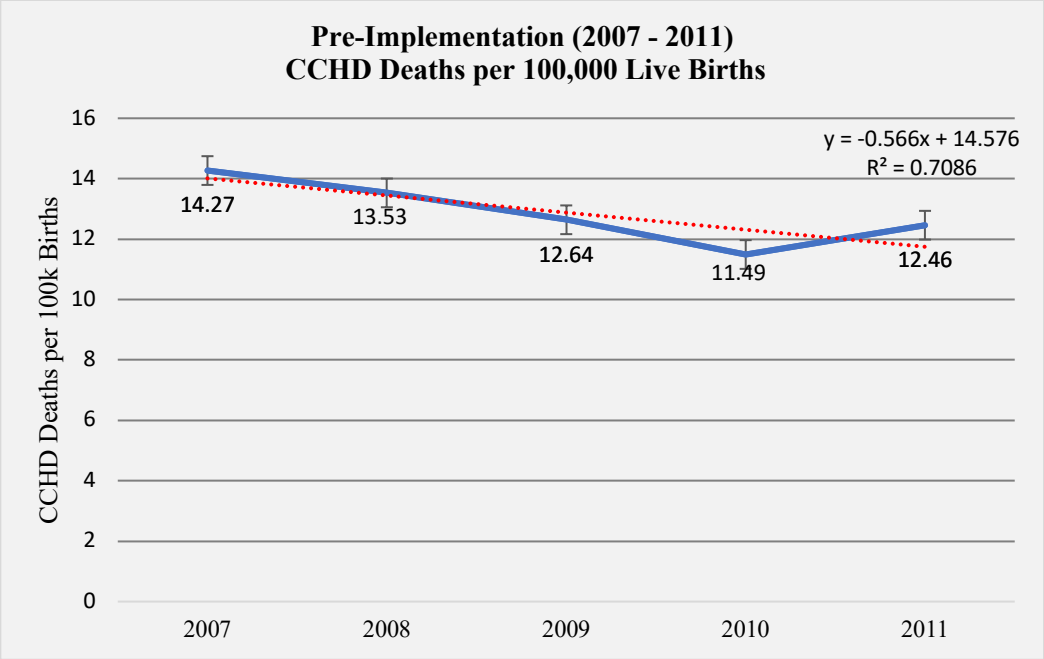

Figure 2. Trend for annual deaths due to CCHDs in the pre-implementation group

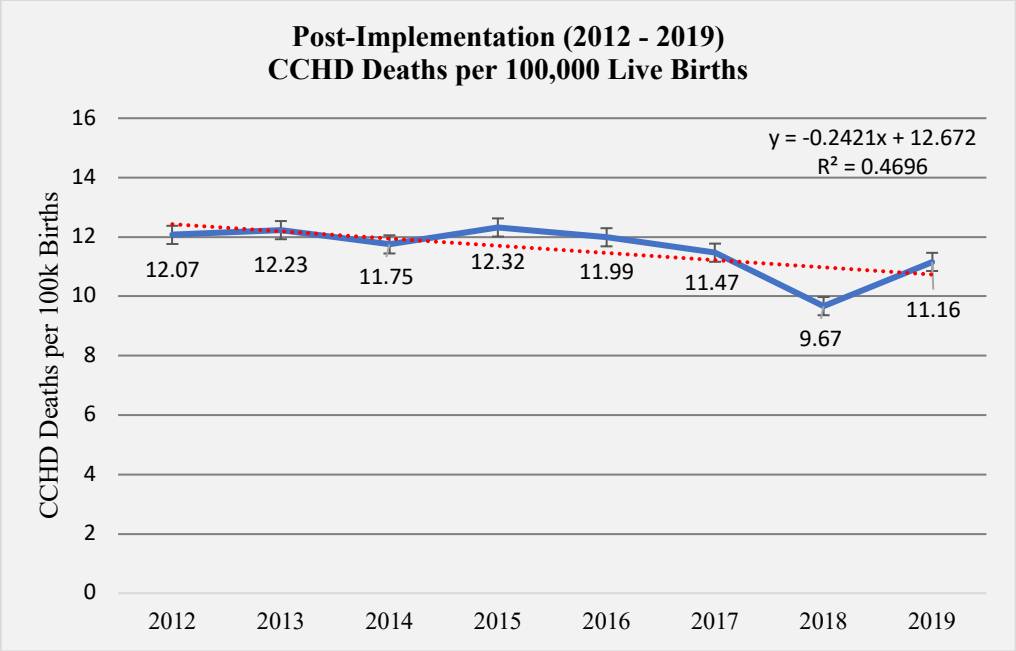

Figure 3. Trend for annual deaths due to CCHDs in the post-implementation group

RESULTS

Between August 31, 2011, to July 1, 2018, all 50 U.S. states and Washington, D.C. implemented mandatory measures requiring newborns to be screened for critical congenital heart defects through pulse-oximetry screening. From 2007 to 2011, prior to the implementation of any mandates, the mean annual count of infant deaths due to critical congenital heart defects was 522 deaths, equal to 12.89 deaths per 100,000 live births. The yearly count of deaths during this

time period trended upward at an average increase of 7.2 deaths per year, calculated to be an average yearly decrease of 0.566 deaths per 100,000. From 2012 to 2019, the mean annual count of infant deaths due to critical congenital heart defects was 454 deaths, equal to 11.58 deaths per 100,000 live births. Yearly deaths during this time period trended downward at an average decrease of 11.63 deaths per year, calculated to be an average yearly decrease of 0.242 deaths per 100,000 live births. Figure 4 demonstrates CCHD deaths per 100,000 live births from 2007 through 2019.

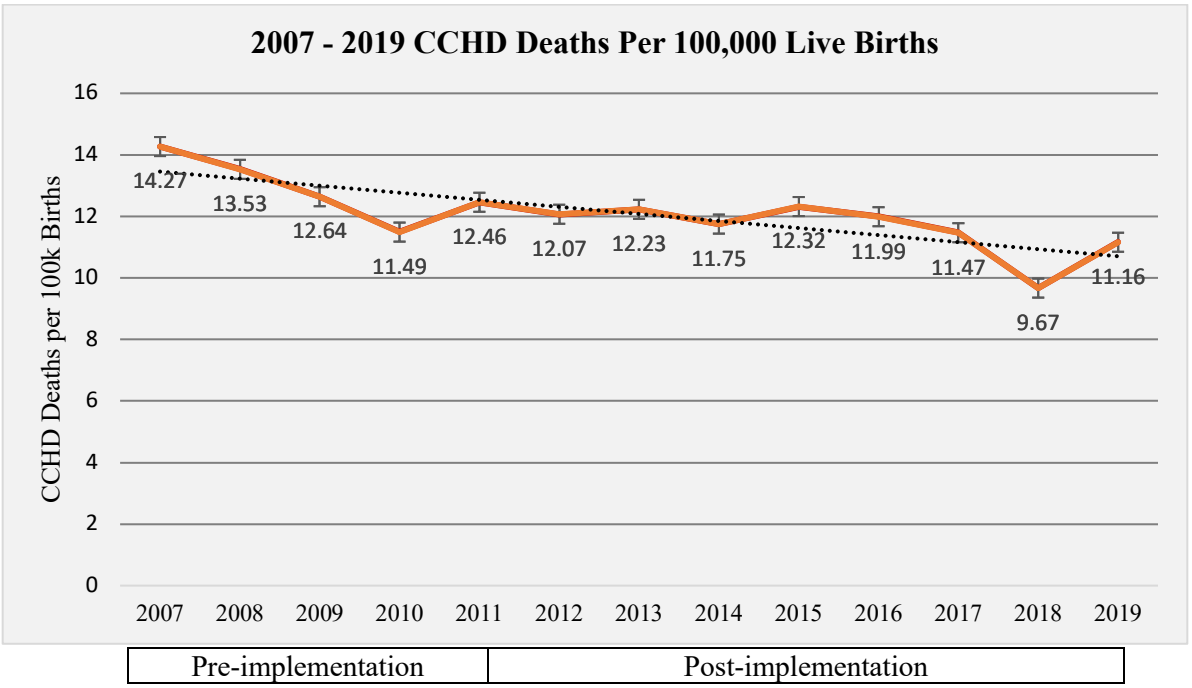

Figure 4. CCHD deaths per 100,000 live births from 2007 – 2019.

The average total of annual U.S. live births decreased between the time periods of interest, pre-implementation (2007 - 2011) and post-implementation (2012 - 2019) by 126,008. The average number of infant deaths also decreased by 3,733. With this decrease, there was also a decrease of the average number of CCHD infant deaths by 67. However, there was an increase (0.03%) in CCHD deaths as a percentage of total infant deaths. Table 4 provides the changes in pre-implementation and post-implementation CCHD infant deaths.

Time Period	Average U.S. Population	Average Amount of Infant Deaths	Average Amount of CCHD Infant Deaths	Average CCHD as a % of Infant Deaths
Pre-implementation (2007 – 2011)	4,045,002	26,436	522	1.97%
Post-implementation (2012 – 2019)	3,918,994	22,703	454	2.00%
Change	*-126,008*	*-3,733*	*-67*	*+0.03%*

Table 4. Change in pre-implementation and post-implementation CCHD infant deaths

DISCUSSION

Concurring with a past study (5), mandated pulse-oximetry screening measures are associated with infant lives saved. The implementation of these policies correlated with a decrease in infant CCHD deaths. Although the results do not include the potential lives saved from defects other than CCHDs in the process of pulse-oximetry screenings, there may be an opportunity that similar screenings for other infant morbidities could also reduce infant mortality rates. More research is encouraged to confirm similar outcomes with other mandated screening implementations and outcome measures.

An interesting note to be made is that, although the mean yearly CCHD deaths per 100,000 live births was lower in the 2012 - 2019 timeframe than in the 2007 - 2011 timeframe, the rate of reduction in deaths per 100,000 births was greater in the 2007-2011 period. In the 2012 - 2019 period, there was a mean yearly reduction of 0.24 CCHD deaths per 100,000, while that reduction was 0.57 CCHD deaths per 100,000 in the 2007 - 2011 period. It is difficult to determine if the reductions in the more recent time period would have occurred naturally, at a greater rate, or at a lesser rate had the testing mandates not been implemented. Nonetheless, the results posit further exploration when considering the true impact of the policy mandates.

Furthermore, the outcome measure tested in this study is limited solely to infants under one year of age. It is likely that a study inclusive of a wider age group would yield a greater decrease in deaths due to CCHDs as compared to the predicted null outcome. Early detection of CCHDs is critical in preventing further health complications due to decreased levels of oxygen in an infant's rapidly developing body. As previously mentioned, perinatal asphyxia can cause permanent brain damage or death in severe cases. Furthermore, roughly 25% of CHD's require surgical or catheter intervention in the first year of life (8). This only scratches the surface of the detrimental effects CCHD's can have on one's 'healthspan.'

With the increase in electronic health records and data reporting, data continues to better inform policy decision-making. Meanwhile, improvements in medical technology are enabling healthcare providers to improve upon existing detection measures and procedures after diagnosis. With the timespan of this study in mind, advancements in the U.S. healthcare system may play a role in proper reporting mechanisms and better quality outcomes in general.

Dodge-Khatami (2016) framed the advancements that have been made in the CCHD field by writing, "In no other field of science or medicine has so much been accomplished in so little time, with heart defects that were an unconditional death sentence 60 years ago, to the current operative survival rates of more than 96% for all defects considered together" (pgs. 110-111). A nearly unimaginable amount of progress has occurred in decreasing the CHD and CCHD mortality rates and in increasing quality of life. However, there is still room for improvement, despite the progress that has been made. The result of healthcare improvement from policy implications, such as pulse oximetry mandates, is inspiring. More research can be used as a tool to push policy proposals onto the state and national agenda. Ultimately, the hope is that these combined efforts can further improve healthcare interventions, improve care provided, and save lives.

LIMITATIONS

ICD-10 codes for CCHD's were determined based on the *International Statistical Classification of Diseases and Related Health Problems, Tenth Revision* (*ICD-10*). Classification

of deaths using ICD-10 codes is a limitation, as deaths may be due to other associated factors, such as the potential of errors in reporting with ICD-10 codes and reporting of the reason for death. In addition, despite the implementation of screening mandates, there remains the likelihood that some hospitals were already screening newborns for CCHDs or that policies were not carried out in a time-effective manner after implementation. Therefore, the results may be skewed due to the lack of individual unit observations of screening practices within each facility. Mandates do not necessarily account for the totality of actual screening practices. Confounding factors of death as an outcome of CCHDs could also be influenced based on the availability of prenatal cardiac resources if detected early, with rates of prenatal diagnoses of CCHDs being variable as well.

CONCLUSIONS

From 2011 to 2019, all states implemented mandatory policies for newborn pulse oximetry screening for CCHD. The implementation of these policies was associated with a significant decrease in infant cardiac deaths compared with states without these policies prior to 2011. This study sheds light on the power of policy effectiveness and health outcomes, in this case, on potential infant lives saved. Early detection with other non-invasive technologies could also be explored to decrease other types of infant morbidities. Further research may yield valuable information regarding the cost incurred due to the increase in pulse oximetry screenings and more accurate predictions of future lives saved.

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