Reduction in Infant Cardiac Deaths in US States Implementing Policies to Screen Newborns for Critical Congenital Heart Disease

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What is "Critical" Congenital Heart Disease (CCHD)?

▪ Specific heart defects that require prompt detection to avoid crises

- Coarctation of the aorta
- Double-outlet right ventricle
- Ebstein anomaly
- Hypoplastic left heart syndrome
- Interrupted aortic arch
- Pulmonary atresia
- Single ventricle
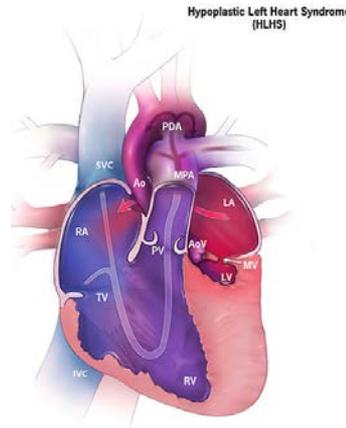
- Tetralogy of Fallot
- Total anomalous pulmonary venous connection
- D-transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus
- Critical cyanotic lesions not otherwise specified

Hypoplastic Left Heart Syndrome (HLHS)

Oster ME, Aucott SW, Glidewell J, Hackell J, Kochilas L, Martin GR, Phillipi J, Pinto NM, Saarinen A, Sontag M, Kemper AR. Lessons learned from newborn screening for critical congenital heart defects. *Pediatrics.* 2016;137(5):e20154573.

CCHD and Newborn Screening

- CCHD: 12 specific defects that are serious and often fatal
  - 2 per 1000 births
  - 300-400 infant deaths per year in US
- Majority associated with low blood oxygen because of impaired circulation
- DHHS added CCHD to Recommended Uniform Screening Panel in 2011
  - States set newborn screening panels by legislation, rules & regulations
  - Point-of-care screening uses pulse oximeters to detect low blood oxygen by shining light through skin

Objective

- To estimate the effect of state CCHD newborn screening policies on infant deaths from congenital heart disease in the United States

Study Design (1)

- Difference-in-difference analysis of impact of state screening policies on numbers of early infant deaths caused by CCHD or other cardiac causes

- This method compares changes in outcomes following the introduction of a policy in groups with the policy and without the policy

  - Can be used to assess outcomes of policies adopted at different times by state or local governments

  - The method assumes similar pre-policy trends in outcomes and attributes different post-introduction trends, after controlling for state-specific factors, to policy

Study Design (2)

- Data source -- Period Linked Birth-Infant Death Data files from National Center for Health Statistics , 2007-2013
    - Births through June 30, 2013 linked to infant deaths through December 31, 2013
    - Causes of death from death certificates classified using ICD-10 codes
- Two outcome measures – counts of infant deaths from 24 hours to under 6 months due to CCHD or due to other/unspecified congenital heart disease (CHD)
- Data grouped by state of birth and month-year
    - Numbers of deaths in infants born in a state during a month when a CCHD screening policy was in place at the beginning of the month

Study Design (3)

- State screening policies
  - Mandate implemented
    - Reviewed state legislation and websites to confirm initial dates on which providers were required to screen newborns
  - Non-mandatory
    - Mandate adopted but not yet implemented
    - Voluntary screening policy

- Poisson regression models of numbers of deaths to birth cohort
  - Log of number of births in month in state
  - Adjusted for state factors

CCHD Screening Policies as of June 1, 2013

- Birth months through June 2013 included to allow for 6 months of death records

- States first adopted CCHD screening policies in mid-2011
  - 8 states implemented mandates by June 1, 2013
    - 2 states implemented mandates during August 2011-January 2012
    - 6 implemented mandates during July 1, 2012-June 1, 2013
  - 13 states adopted but did not yet implement mandates by June 1, 2013
  - 5 other states adopted voluntary screening policies by June 1, 2013

States That Implemented Screening Mandates by June 1, 2013

State	Date adopted	Date implemented
Connecticut	May 2012	January 1, 2013
Delaware	May 1, 2013	May 1, 2013
Indiana	May 2011	January 1, 2012
Maryland	May 19, 2011	September 1, 2012
New Hampshire	June 2012	August 11, 2012
New Jersey	June 2, 2011	August 31, 2011
Tennessee	March 1, 2012	May 31, 2013
West Virginia	April 5, 2012	September 1, 2012

Changes in Infant Deaths from CCHD or Other CHD During January 1, 2007-June 1, 2013

Characteristic	All states	States with no policy implemented	States with mandatory policy			States with voluntary policy	
			Before enactment	Between enactment and implementation	After implementation	Before enactment	After enactment
CCHD deaths[a] per 100,000 births	9.8	9.8	8.3	8.0	4.2	10.6	10.6
Other CHD deaths[a] per 100,000 births	13.5	13.0	12.0	11.9	8.0	14.9	15.3

a. Deaths from 24 hours to <6 months of age

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Key Findings – Effects of Screening Policies

- Relative reductions in CCHD and Other CHD deaths compared to births in months with no state policy, adjusted for state factors and time trends
  - Mandatory screening
    - CCHD deaths to age 6 months fell by one-third (33.4%)
    - Other CHD deaths fell by one-fifth (21.4%)
    - Both changes were statistically significant
  - Non-mandatory screening
    - No reductions in CCHD or other CHD deaths (<5% difference, not statistically significant)

Extrapolation to Universal CCHD Screening in US

- Potential reduction in annual deaths for US as a whole
  - Recognized CCHD deaths: 120 (95% confidence interval (CI): 38–181)/year
  - Other CHD deaths: 117 (95% CI: 38-185)/year
    - Most of those deaths had an ICD-10 code for unspecific CHD and may represent undiagnosed or unrecorded CCHD deaths

Discussion

- A US cost-effectiveness analysis in 2013 concluded that CCHD screening would likely be cost-effective

  - Approximately $40,000 per life-year saved

  - Assumed that screening 4 million infants would avoid just 20 infant deaths per year

- Implications

  - If universal screening avoids 120 infant deaths per year (or more), universal CCHD screening is even more likely to be cost-effective

Peterson C, Grosse SD, Oster ME, Olney RS, Cassell CH. A cost-effectiveness analysis of routine screening to detect critical congenital heart disease among U.S. newborns. *Pediatrics*. 2013;132:e595-603.

Limitations and Next Steps

- Small numbers of state birth-months exposed to state mandates

- We used the most recent data that were made available to us. The 2014 linked birth-death file has been requested and additional analyses will be undertaken when those data are received

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Sensitivity Analyses

- Placebo tests
  - No changes in other leading causes of infant deaths associated with CCHD screening policies
    - Sudden infant death syndrome (SIDS), sepsis, maternal complications, preterm or low birthweight
- Tests of non-parallel trends hypothesis
  - Regression analysis excluding birth-months following implementation of mandatory screening – interaction of screening mandate and time
    - Mandatory screening * time: -0.001 (95% CI: -0.008 to 0.006)