

Screening Compliance for Critical Congenital Heart Disease in Texas Newborns



**Texas Children's
Hospital**[®]

BCM

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Pediatrics

Financial Disclosures

- None to disclose

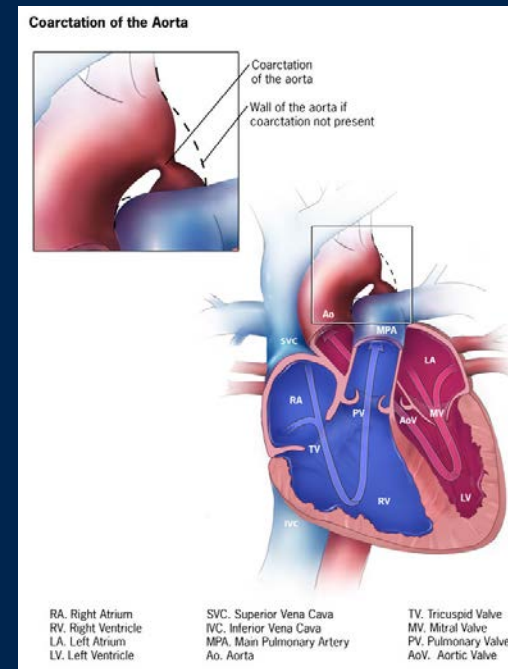
Critical Congenital Heart Disease

- Congenital Heart disease: 8-9/1000 births
- Of these: 25% are CCHD
 - life threatening and
 - require intervention within the first few days of life through early infancy

Critical Congenital Heart Disease

Affected infants appear well due to the presence of a ductus arteriosus that provides flow and oxygenation to the lungs and/or the body

Oftentimes, infants will present in **shock** after the patent ductus arteriosus has closed spontaneously a few days or weeks after discharge



CCHD screening saves lives!

- About 30% of infant deaths from CCHD occur before diagnosis
 - In CA, about 30 infants/yr died from late or missed CHD
- More than 50% died at home or EC

Table 2. Total Number of Patients in Each Group by Diagnosis

Cause of Death	No. (%) of Patients ^a		
	Study Cohort (N=898) ^b	Unknown (n=299)	Missed CCHD Diagnosis (n=152)
Aortic stenosis	31 (3.5)	19 (6.4)	12 (7.9)
Coarctation of aorta, including interrupted aortic arch	90 (10.0)	49 (16.4)	41 (27.0)
DORV and single ventricle	15 (1.7)	14 (4.7)	1 (0.7)
Hypoplastic left heart syndrome	565 (62.9)	60 (20.1)	58 (38.2)
Pulmonary atresia	30 (3.3)	22 (7.4)	8 (5.3)
Tricuspid atresia	9 (1.0)	9 (3.0)	0
TAPVR	32 (3.6)	23 (7.7)	9 (5.9)
d-Transposition of great vessels	37 (4.1)	31 (10.4)	6 (3.9)
Tetralogy of Fallot	55 (6.1)	50 (16.7)	5 (3.3)
Truncus arteriosus	34 (3.8)	22 (7.4)	12 (7.9)

Abbreviations: DORV, double outlet right ventricle; TAPVR, total anomalous pulmonary venous return.

^a Because of rounding, percentages may not total 100.

^b Study cohort indicates the 898 patients selected by the initial selection criteria specified in the "Patient Selection" subsection of the "Methods" section.

Chang et al, *Arch Pediatr Adolesc Med*, 2008

CCHD screening is effective!

- A meta-analysis of pulse ox screening for CCHD in asymptomatic newborns
 - Over 220,000 NB's
 - Overall sensitivity was 76.5%, specificity was 99.9% with a false positive rate of 0.14%
- Not all CCHDs are detected

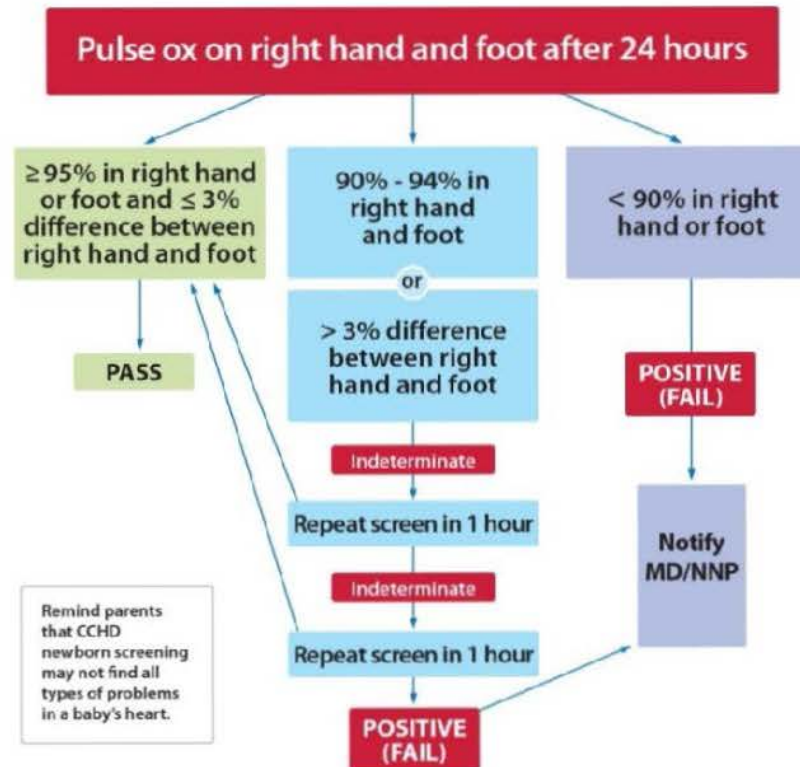
Thangaratinam, et al. Lancet, 2012;379:2459-64

TXPOP

- Texas Pulse-Oximetry Project (TXPOP) was created to provide resources and identify strategies on how to most effectively implement CCHD screening in a diverse group of birthing centers throughout the state.
- TXPOP offers a TOOLKIT consisting of Powerpoint slides for physicians and nurses, algorithm posters, and patient education pamphlets to help birthing centers implement CCHD screening throughout Texas.

Figure 2: Texas Pulse Oximetry Project Algorithm

Critical Congenital Heart Disease Newborn Screening Algorithm



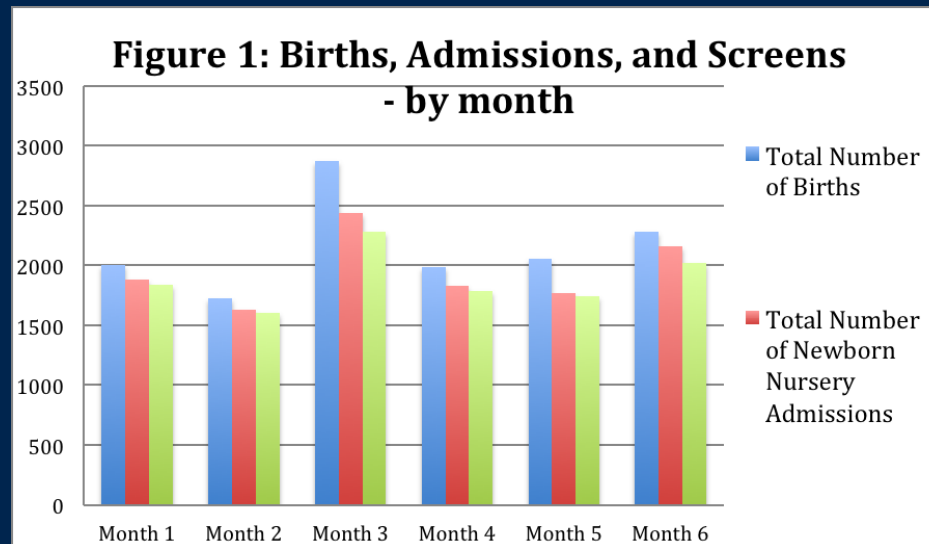
A Joint Educational Initiative of
The University of Texas Health Science Center at San Antonio/Department
of Pediatrics, Baylor College of Medicine/Department of Pediatrics and Texas
Department of State Health Services



Results

Between Feb-July 2013

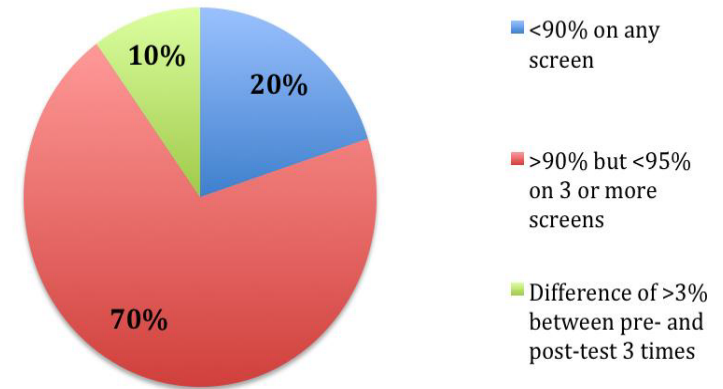
- 12,946 births in 13 participating hospitals
- 11,711 newborn nursery admissions
- 96% were screened between 24h- discharge



Results

- 11 positive screens
 - 0.08 %
 - 1 with CCHD
- 32 (0.2%) received echos
 - More echo's were ordered than positive screening tests

Figure 2: Positive Screens, by pathway



Texas Law on CCHD Screening

- As of September 1, 2014, Texas law mandated CCHD screening for all newborns
- The Texas law also mandated that all confirmed positive cases are reported to the state

Expected CCHD Cases in Texas

- Incidence of CCHD is 1.8/1,000
- About 400,000 births in Texas per year
- $400,000 \times 1.8/1,000 = 720$ expected CCHD cases in Texas per year

CCHD Reported Cases September 2014 – September 2016

CONFIRMED CCHD IN TEXAS September 2014 – September 2016			
Primary		Secondary	
Transposition of Arteries	55	Coarctation of Aorta	54
Tetralogy of Fallot	50	Double Outlet Ventricle	19
Hypoplastic Left Heart	30	Interrupted Aortic Arch	11
Total Anomalous Pulmonary	30	Ebstein Anomaly	8
Pulmonary Atresia	13	Single Ventricle	7
Tricuspid Atresia	13	Unspecified Secondary	38
Truncus Arteriosis	8		
Unspecified Primary	23		
Total Primary	222	Total Secondary	137
Total CCHD = 359			

359/720
=50% of
expected
cases

2016 TPS Fetus and Newborn Committee Report- Freedenberg, DSHS

CCHD Screening Compliance

- The objective of this project was to determine the percentage of Texas birthing centers that are in compliance with the Texas law
 - by providing universal CCHD screening
 - as well as reporting positive CCHD cases to the state
- Identify barriers to implementation
- Identify educational needs

Methods

- A list of 392 birthing centers was collected from the NICU search on the American Academy of Pediatrics website.
- A 10-question phone survey was developed and administered to the directors (or their designee) to obtain data about CCHD screening in their NICU Levels 1, 2, 3, and 4 units as well as their use of the TXPOP toolkit

Results

SCREENING COMPLIANCE FOR CCHD IN TEXAS
392 Birthing Hospitals
380 (96.9%) Responded

Newborns Level 1

Newborns Levels 2, 3, 4

hospitals (% screening)

350 (92.1%)

268 (70.5%)

75% used TXPOP toolkit

Discussion

- Infants in Level 1 are generally healthy and comprise a much more uniform patient population, for which it is likely easier to implement a uniform screening protocol
- It is often not possible to screen NICU babies at 24 hours after birth
 - These infants are more likely to be transferred, making it unclear which hospital is responsible for performing the screen
- NICU babies may have presence of other co-morbidities (e.g., congenital lung disease) that may affect the ability to interpret screening results

Questions

- What are the challenges faced in the NICUs throughout the state regarding CCHD screening?
- What are the barriers to reporting?
- Could the current TXPOP toolkit be further improved?
- Could the law be rewritten to improve screening and reporting?

Next Steps

- Ongoing evaluation of our data
- Update current findings/reports
- Identify targets for improvement
 - Such as examining policy and protocols for screening CCHD
 - capability of performing echocardiograms locally
 - awareness of the TXPOP toolkit
- Utilize QI methods to systematically approach these targets

Next Steps

Collaboration is key!



References

- Congenital Heart Defects (CHDs). (2016 March 04). Retrieved July 15, 2017, from <https://www.cdc.gov/ncbddd/heartdefects/tetralogyoffallot.html>
- Congenital Heart Defects (CHDs). (2016 September 26). Retrieved July 15, 2017, from <https://www.cdc.gov/ncbddd/heartdefects/coarctationofaorta.html>
- Ewer, A. K. (2013). Review of pulse oximetry screening for critical congenital heart defects in newborn infants. *Current Opinion in Cardiology*,28(2), 92-96. doi:10.1097/hco.0b013e32835d7e42
- Gong, A., Livingston, J., Creel, L., Ocampo, E., Mckee-Garrett, T., & Guillory, C. (2017). Texas Pulse Oximetry Project: A Multicenter Educational and Quality Improvement Project for Implementation of Critical Congenital Heart Disease Screening Using Pulse Oximetry. *American Journal of Perinatology*,34(09), 856-860. doi:10.1055/s-0037-1599214
- Guillory, C., Creel, L., Livingston, J., Mckee-Garrett, T., Fortunov, R., & Gong, A. (2017). A Multicenter Initiative for Critical Congenital Heart Disease Newborn Screening in Texas Neonatal Intensive Care Units. *American Journal of Perinatology*,34(09), 839-844. doi:10.1055/s-0037-1599053
- Reller, MD, Strickland, MJ, Riehle-Colarusso, TJ, Mahle, WT, Correa, A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005. *J Pediatr*. 2008;153:807-13.
- Texas Pediatric Society. (n.d.). Retrieved July 15, 2017, from <https://txpeds.org/txpop>
- Thangaratinam, S., Brown, K., Zamora, J., Khan, K. S., & Ewer, A. K. (2012). Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. *The Lancet*, 379(9835), 2459-2464. doi:10.1016/s0140-6736(12)60107-x

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