

Operational Policy Update



TO:

Birthing Facilities, Pediatricians, Pediatric Cardiologists

FROM:

Marian Swinker, MD, MPH, Commissioner

Bureau for Public Health

Anne Williams, RN, BSN, MS-HCA, Director Office of Maternal, Child and Family Health

RE:

Critical Congenital Heart Disease

ISSUE DATE:

July 16, 2012

EFFECTIVE DATE: September 1, 2012

On March 10, 2012, the WV State Legislature passed House Bill 4327 establishing the expectation that all infants born in a licensed health care facility be screened for Critical Congenital Heart Disease (CCHD) using pulse oximetry. Screening for CCHD is also recommended by the U.S. Department of Health and Human Services Secretary's Advisory Committee on Heritable Disorders and Genetic Diseases.

Pulse oximetry must routinely be performed on all infants at or greater than 24 hours after birth and before discharge from the hospital. In West Virginia, there is no parental opt out for newborn screening. The Birth Score Office located within WVU Health Systems will collect the screen results for surveillance purposes on the Birth Score card. Birthing facility nursery staff will mark the Birth Score Card with pulse oximetry results of "Pass," "Fail," or "Not Done." New Birth Score cards have been ordered and should be delivered to all birthing facilities by September 1, 2012. The Office of Maternal, Child and Family Health will act as a liaison with the birthing facilities and medical community to assure screening capacity and to provide educational resources as needed.

The universal screen for CCHD is an essential public health service and maximizes the State's opportunity for early identification of newborn disorders and subsequent medical intervention. Information and links regarding CCHD can be found on the newborn screening website at www.wvdhhr.org/nbms/. Enclosed you will find a copy of H.B. 4327, the U.S. Department of Health and Human Services recommendations, and the American Academy of Pediatrics and Center for Disease Control's recommended algorithm for review and reference. Under separate cover, you will receive laminated algorithms for display in the newborn nursery.

Questions about this policy can be directed to the Office of Maternal, Child and Family Health, Newborn Screening Program, at (304) 558-5388 or toll free at 1-800-642-8522.

Attachments

CC:

Mary S. Boyd, M.D., AAP WV Chapter President Birth Score Office

> Bureau for Public Health Office of Maternal, Child and Family Health 350 Capitol Street, Room 427 Charleston, West Virginia 25301-3714

Toll-Free (In WV): 1-800-642-8522 Fax: (304) 558-2183 Phone: (304) 558-5388

ENROLLED

COMMITTEE SUBSTITUTE

for

H. B. 4327

(By Delegates Hatfield, D. Poling, Brown and Staggers)

[Passed March 10, 2012; in effect ninety days from passage.]

AN ACT to amend the Code of West Virginia, 1931, as amended, by adding thereto a new article, designated §16-44-1 and §16-44-2, all relating to requiring pulse eximetry testing for newborns; setting forth legislative findings; authorizing the Commissioner of the Bureau of Public Health to require testing; providing timing requirements for testing; and requiring the commissioner to adopt procedural and legislative rules.

Be it enacted by the Legislature of West Virginia:

That the Code of West Virginia, 1931, as amended, be amended by adding thereto a new article, designated §16-44-1 and §16-44-2, all to read as follows:

ARTICLE 44. THE PULSE OXIMETRY NEWBORN TESTING ACT.

§16-44-1. Legislative findings.

The Legislature finds and declares that:

- (1) Congenital heart defects are structural abnormalities of the heart that are present at birth; congenital heart defects range in severity from simple problems such as holes between chambers of the heart, to severe malformations, such as the complete absence of one or more chambers or valves; some critical congenital heart defects can cause severe and life-threatening symptoms which require intervention within the first days of life;
- (2) According to the United States Secretary of Health and Human Services' Advisory Committee on Heritable Disorders in Newborns and Children, congenital heart disease affects approximately seven to nine of every thousand live births in the United States and Europe; the federal Centers for Disease Control and Prevention states that congenital heart defects are the leading cause of infant death due to birth defects;
- (3) Current methods for detecting congenital heart defects generally include prenatal ultrascund screening and repeated clinical examinations; while prenatal ultrasound screenings can detect some major congenital heart defects, these screenings, alone, identify less than half of all congenital heart defect cases, and critical congenital heart defect cases are often missed during routine clinical exams performed prior to a newborn's discharge from a birthing facility;
- (4) Pulse oximetry is a noninvasive test that estimates the percentage of hemoglobin in blood that is saturated with oxygen; when performed on a newborn when the baby is twenty-four to forty-eight hours of age, or as late as possible if the baby is to be discharged from the hospital before he or she is twenty-four hours of age, pulse oximetry screening is often more effective at detecting critical, life-threatening congenital heart defects which otherwise go undetected by current screening methods; newborns with abnormal pulse oximetry results require immediate confirmatory testing and Intervention; and
- (5) Many newborn lives could potentially be saved by earlier detection and treatment of congenital heart defects if birthing facilities in the state were required to perform this simple, nonlinvasive newborn screening in conjunction with current congenital heart defect screening methods.

§16-44-2. Pulse oximetry screening required; definition; rules.

- (a) The Commissioner of the Bureau for Public Health shall require each birthing facility licensed by the Department of Health and Human Resources to perform a pulse oximetry screening on every newborn in its care, when the baby is twenty-four to forty-eight hours of age, or as late as possible if the baby is to be discharged from the hospital before he or she is twenty-four hours of age.
- (b) As used in this article, "birthing facility" means an inpatient or ambulatory health care facility licensed by the Department of Health and Human Resources that provides birthing and newborn care services.
- (c) The commissioner shall adopt procedural rules and propose legislative rules for legislative approval, in accordance with the provisions of article three, chapter twenty-nine-a of this code, that are necessary to carry out the purposes of this article.



Screening for Critical Congenital Heart Defects

Newborn screening using pulse oximetry can identify some infants with critical congenital heart defects (CCHDs, which also are known collectively in some instances as critical congenital heart disease). CCHDs are structural heart defects that often are associated with hypoxia among infants during the Report (MMWR) newborn period. Infants with CCHDs are at risk for significant morbidity or mortality early in life because of closing of the ductus arteriosus or other physiologic changes. While several defects could be considered CCHDs, in the context of newborn screening using pulse oximetry, seven defects are classified as CCHD: hypoplastic left heart syndrome, pulmonary atresia (with intact septum), tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great arteries, tricuspid atresia, and truncus arteriosus. These seven CCHDs represent about 17-31% of all congenital heart disease. All of these defects require some type of intervention-often involving a surgical procedure—soon after birth.

Without screening, some newborns with CCHDs might be missed because the signs of CCHD might not be evident before an infant is discharged from the hospital after birth. Other heart [Listen] [View Transcript 📆] defects might be considered secondary screening targets. Some

of these heart defects can be just as severe as the primary screening targets and also require intervention soon after birth. However, newborn screening using pulse oximetry may not detect these heart defects as consistently as the seven disorders listed as primary targets. These secondary targets include aortic arch atresia or hypoplasia, interrupted aortic arch, coarctation of the aorta, double-outlet right ventricle, Ebstein anomaly, pulmonary stenosis, atrioventricular septal defect, ventricular septal defect, and single ventricle defects (other than hypoplastic left heart syndrome and tricuspid atresia).

Morbidity and Mortality Weekly **Podcasts**



Healthy Little Hearts

"A Minute of Health with CDC" [Listen] [View Transcript 📜]

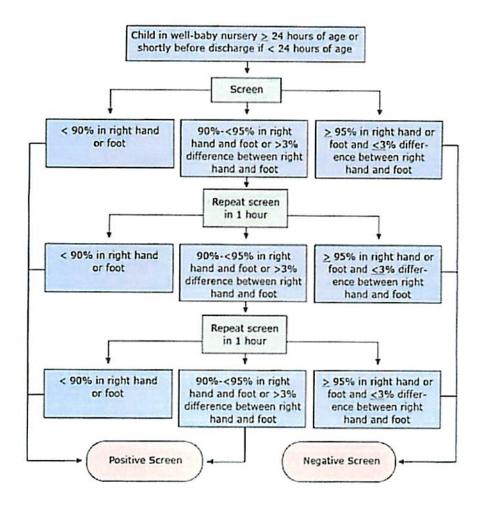
"A Cup of Health with CDC"

Methods and Screening Algorithm²

Current recommendations focus on screening infants in the well-baby nursery and in intermediate care nurseries or other units in which discharge from the hospital is common during an infant's first week of life. A pulse oximeter is used to measure the percentage of hemoglobin in the blood that is saturated with oxygen.

The following algorithm has been developed to show the steps in screening:2

[Text description of chart]



PLEASE NOTE: Percentages refer to oxygen saturation as measured by pulse oximeter.

A screen is considered positive if (1) any oxygen saturation measure is <90% (in the initial screen or in repeat screens); (2) oxygen saturation is <95% in the right hand and foot on three measures, each separated by one hour; or (3) a >3% absolute difference exists in oxygen saturation between the right hand and foot on three measures, each separated by one hour. Any screening that is \geq 95% in the right hand or foot with a \leq 3% absolute difference in oxygen saturation between the right hand or foot is considered a negative screen and screening would end.

Any infant with a positive screen should have a diagnostic echocardiogram, which would involve an echocardiogram within the hospital or birthing center, transport to another institution for the procedure, or use of telemedicine for remote evaluation. The infant's pediatrician should be notified immediately and the infant might need to be seen by a cardiologist for follow-up.

False positives are decreased if the infant is alert, and timing CCHD screening around the time

of the newborn hearing screening improves efficiency. Pulse oximetry screening should not replace taking a complete family medical and pregnancy history and completing a physical examination, which sometimes can detect CCHD before the development of hypoxia. Pulse oximetry screening does not detect all CCHD, so it is possible for a baby with a negative screening result to still have CCHD or other congenital heart defects.

Research is needed on screening certain populations (e.g., those at high altitudes) and evaluating diagnostic strategies (e.g., telemedicine) for nurseries without onsite echocardiography.

Current Status of Recommendations

- In September 2010, the U.S. Department of Health and Human Services (HHS)
 Secretary's Advisory Committee on Heritable Disorders in Newborns and Children
 (SACHDNC) voted to add CCHD screening to the SACHDNC Recommended
 Uniform Screening Panel.
- In September 2011, HHS Secretary Sebelius approved adding CCHD to the Recommended Uniform Screening Panel and outlined specific tasks assigned to NIH, CDC, and HRSA.
- There are differences in the screening process and the number and types of conditions included in screening in each state. The status of CCHD screening in each state can be seen here &.

Economic Considerations and Costs²

Costs of pulse oximetry screening include screening equipment, supplies associated with screening (e.g., probes, adhesive wraps), and staff time needed to perform screening, track results, and communicate with parents.

- Screening has been estimated to cost \$5.00 to \$10.00 per infant.
- The time required for each screen is about 1 to 5 minutes.

Costs associated with diagnosis and follow-up of infants with out-of-range (positive) results are not included in these estimates. For example, an echocardiogram to verify an out-of-range (positive) screen could cost several hundred dollars.

Webinars

Newborn Screening Recommendations for Critical Congenital Heart Disease (CCHD): Implications for State Programs ₽

The Early Identification of Congenital Heart Disease: Information for Primary Care Providers ₹ ₽

For More Information

CDC Feature on Screening for Critical Congenital Heart Defects In the US, about 4,800 (or 11.6 per 10,000) babies born each year have one of seven critical congenital heart defects (CCHDs). Learn more about how CCHD screening potentially can identify these babies soon after birth.

Children's National Medical Center's Congenital Heart Disease Screening Program & Children's National Medical Center provides information on CCHD screening for healthcare providers and parents. Contact them to request a toolkit for implementing CCHD screening.

Congenital Heart Public Health Consortium &

Secretary's Advisory Committee on Heritable Disorders in Newborns and Children &

American Academy of Pediatrics: Endorsement of Health and Human Services Recommendation for Pulse Oximetry Screening for Critical Congenital Heart Disease [Read article ≰ ☑]

Centers for Disease Control and Prevention. Racial differences by gestational age in neonatal deaths attributable to congenital heart defects --- United States, 2003-2006. MMWR Morb Mortal Wkly Rep. 2010;59:1208-1211. [Read article]

Chang, RK, Gurvitz, M, Rodriguez S. Missed diagnosis of critical congenital heart disease. *Arch Pediatr Adolesc Med.* 2008;162:969-974. [Read article &]

Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Lloyd-Puryear M, Howell RR. Strategies for Implementing Screening for Critical Congenital Heart Disease. *Pediatrics*. 2011; 128:e1-e8. [Read article ★ ♣] CBS News Early Show interview with Alex Kemper &

Knapp, AA, Metterville, DR, Kemper, AR, Prosser, L, Perrin, JM. Evidence review: Critical congenital cyanotic heart disease, Final Draft, September 3, 2010. Prepared for the Maternal and Child Health Bureau, Health Resources and Services Administration. [Read article 😤 🖻]

Mahle, WT, Newburger, JW, Matherne, GP, Smith, FC, Hoke, TR, Koppel, R, Gidding, SS, Beekman, RH, 3rd, Grosse, SD. Role of pulse oximetry in examining newborns for congenital heart disease: A scientific statement from the AHA and AAP. *Pediatrics*. 2009;124:823-836. [Read article &]

Reller, MD, Strickland, MJ, Riehle-Colarusso, TJ, Mahle, WT, Correa, A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005. *J Pediatr*. 2008;153:807-813. [Read article &]

Related Links

- Birth Defects
- Newborn Screening

- · Hypoplastic Left Heart Syndrome
- Tetralogy of Fallot
- · Transposition of the Great Arteries
- · CDC's National Center on Birth Defects and Developmental Disabilities

References

- 2. Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Lloyd-Puryear M, Howell RR. Strategies for Implementing Screening for Critical Congenital Heart Disease. *Pediatrics*. 2011; 128:e1-e8. [Read article 🛣 🗗]

Page last reviewed: May 7, 2012 Page last updated: May 7, 2012

Content source: National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention

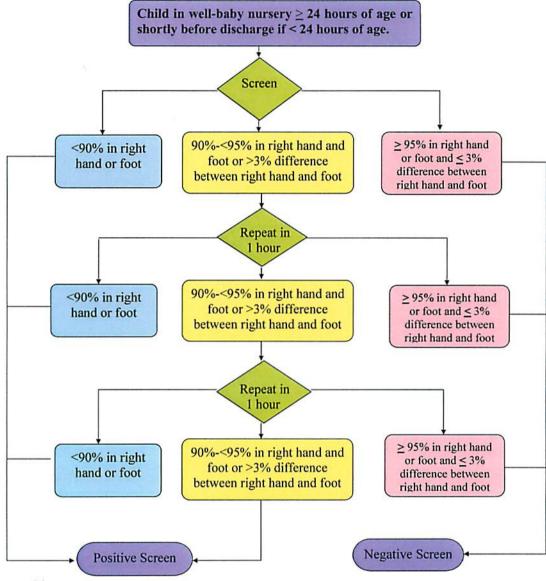
Centers for Disease Control and Prevention 1600 Clifton Rd. Atlanta, GA 30333, USA 800-CDC-INFO (800-232-4636) TTY: (888) 232-6348 - cdcinfo@cdc.gov



Positive Newborn Screening for Critical Congenital Heart Disease Referral and Follow-up Algorithm*

Screening	< 90% In Right Hand or Foot	90%-< 95% With > 3% Difference between Right Hand and Foot	90%-<95% With ≤3% Difference between Right Hand and Foot	≥ 95% With > 3% Difference between Right Hand and Foot
1 st	Notify the physician or practitioner caring for the infant for Immediate cardiology referral Admit or Transport to appropriate facility for neonatal echo and follow-up care.	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour
2 nd	Notify the physician or practitioner caring for the infant for Immediate cardiology referral Admit or Transport to appropriate facility for neonatal echo and follow-up care.	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour	Ensure patient is calm, awake and the right hand and foot with the pulse oximetry sensor are warm. Repeat screening in 1 hour
3 rd	Notify the physician or practitioner caring for the infant for Immediate cardiology referral Admit or Transport to appropriate facility for neonatal echo and follow-up care. All stal Statesies for Implementing Screening for the physician of the physic	Notify physician or practitioner caring for the infant to obtain an immediate cardiac assessment. Symptomatic: Immediate cardiology referral by physician or practitioner. Admit or Transport to appropriate facility for neonatal echo and follow-up care. Asymptomatic: Follow-up Echocardiogram Cardiology referral	Notify physician or practitioner caring for the infant to obtain an immediate cardiac assessment. Symptomatic: Immediate cardiology referral by physician or practitioner. Admit or Transport to appropriate facility for neonatal echo and follow-up care. Asymptomatic: Follow-up Echocardiogram Cardiology referral	Notify physician or practitioner caring for the infant for cardiac assessment. Symptomatic: Follow-up Echocardiogram Asymptomatic: Defer discharge for 12-24hrs. Repeat Newborn screening. or Discharge patient with Follow-up Echocardiogram Patient Education of the signs and symptoms of CCHD.

^{*}Adapted from Kemper AR et al. Strategies for Implementing Screening for Critical Congenital Heart Disease. Pediatrics. 2011; 128:e1259.



Reference
Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Lloyd-Puryear M, Howell RR.
Strategies for Implementing Screening for Critical Congenital Heart Disease. Pediatrics. 2011; 128, e1259.

A positive screen is:

- (1) any oxygen saturation measure that is <90% (in the initial screen or in repeat screens);
- (2) oxygen saturation is <95% in the right hand and foot on three measures, each separated by one hour; or
- (3) a >3% absolute difference exists in oxygen saturation between the right hand and foot on three measures, each separated by one hour.

Any infant with a positive screen should have a diagnostic echocardiogram, which would involve an echocardiogram within the hospital or birthing center, transport to another institution for the procedure, or use of telemedicine for remote evaluation. The infant's provider should be notified immediately and the infant might need to be seen by a cardiologist for follow-up.

False positives are decreased if the infant is alert. Timing CCHD screening around the time of the newborn hearing screening improves efficiency. Pulse oximetry screening should not replace taking a complete family, medical and pregnancy history and completing a physical examination, which sometimes can detect CCHD before the development of hypoxia. Pulse oximetry screening does not detect all CCHD, so it is possible for a baby with a negative screening result to still have CCHD or other congenital heart defects.

A negative screen is:

Any screening that is \geq 95% in the right hand or foot with a \leq 3% absolute difference in oxygen saturation between the right hand and foot is considered a negative screen and screening would end.

WEST VIRGINIA CRITICAL CONGENITAL HEART DISEASE

POSITIVE SCREENING SUPPORT OPTIONS

	2012 WV Birth Hospitals (29 WV sites,		Option 1 Marshall Peds Cardiology	Option 2
1.	APPALACHIAN REGIONAL	South Williamson, KY	(304) 526-2399	Triage Beepe (304) 987-455
2.	BLUEFIELD REGIONAL MEDICAL CENTER	Bluefield, WV	Mohammad Iqbal, MD 276) 322-3180 or (276) 920-1382	Triage Beepe (304) 987-455
3.	CABELL HUNTINGTON HOSPITAL	Huntington, WV	Marshall Peds Cardiology (304) 987-4559	Triage Beepe (304) 987-455
١.	CAMDEN-CLARK HOSPITAL	Parkersburg, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	CITY HOSPITAL	Martinsburg, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	DAVIS MEMORIAL HOSPITAL	Elkins, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	FAIRMONT GENERAL HOSPITAL	Fairmont, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	GARRETT COUNTY HOSPITAL	Oakland, MD	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	GRANT MEMORIAL HOSPITAL	Petersburg, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
).	GREENBRIER VALLEY MED CENTER	Ronceverte, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	JEFFERSON MEM HOSPITAL	Ranson, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
2.	LOGAN GENERAL HOSPITAL	Logan, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	MONONGALIA GENERAL HOSPITAL	Morgantown, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	OHIO VALLEY MED CENTER	Wheeling, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
5.	PLEASANT VALLEY HOSPITAL	Point Pleasant, WV	Marshall Peds Cardiology (304) 987-4559	Triage Beepe (304) 987-455
	PRINCETON COMMUNITY HOSPITAL	Princeton, WV	Mohammad Iqbal, MD 276) 322-3180 or (276) 920-1382	Triage Beepe (304) 987-455
	RALEIGH GENERAL HOSPITAL	Beckley, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	REYNOLDS MEMORIAL HOSPITAL	Glendale, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
).	RUBY MEMORIAL HOSPITAL	Morgantown, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
١.	SAINT MARY'S HOSPITAL	Huntington, WV	Marshall Peds Cardiology (304) 987-4559	Triage Beepe (304) 987-455
	ST. JOSEPH'S HOSPITAL (BUCKHANNON)	Buckhannon, WV	WVÙ Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	ST. JOSEPH'S HOSPITAL (PARKERSBURG)	Parkersburg, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	STONEWALL JACKSON HOSPITAL	Weston, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
١.	SUMMERSVILLE MEM HOSPITAL	Summersville, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
i.	THOMAS MEMORIAL HOSPITAL	Charleston, WV	Farah Garmany, MD (304) 549-4973	(304) 987-455
	UNITED HOSPITAL CENTER	Clarksburg, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	WEIRTON MEDICAL CENTER	Weirton, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
	WELCH EMERGENCY HOSPITAL	Welch, WV	Mohammad Iqbal, MD (276) 322-3180 or (276) 920-1382	Triage Beepe (304) 987-455
	WHEELING HOSPITAL	Wheeling, WV	WVU Peds Cardiology (MARS) (304) 598-6000 or 1-800-982-6277	Triage Beepe (304) 987-455
).	WILLIAMSON MEMORIAL HOSPITAL	Williamson, WV	Marshall Peds Cardiology (304) 987-4559	Triage Beepe (304) 987-455
			Farah Garmany, MD	Triage Beepe

NOTE: The above offers the provider options for pediatric cardiology support. The provider is responsible for the patient and may obtain an echocardiogram through their institution or discuss the positive screening with a subspecialist of their choice. They are not required to contact the providers above.