

# Congenital Heart Disease in the United States of America: American Academy of Pediatrics Guidelines

Christopher S. Snyder, MD, FAAP  
Chair of Section of Cardiology and Cardiovascular Surgery (AAP)  
Chief of Pediatric Cardiology  
The Congenital Heart Collaborative

The Congenital Heart Collaborative



# Outline

General AAP information

SOCCS information

What we do

Education/publication

Standards

QI

Outcomes/guidelines

Advocacy

# American Academy of Pediatrics

Over 66,000 members

Almost 800 pediatric cardiologists

Advocates for the care of children

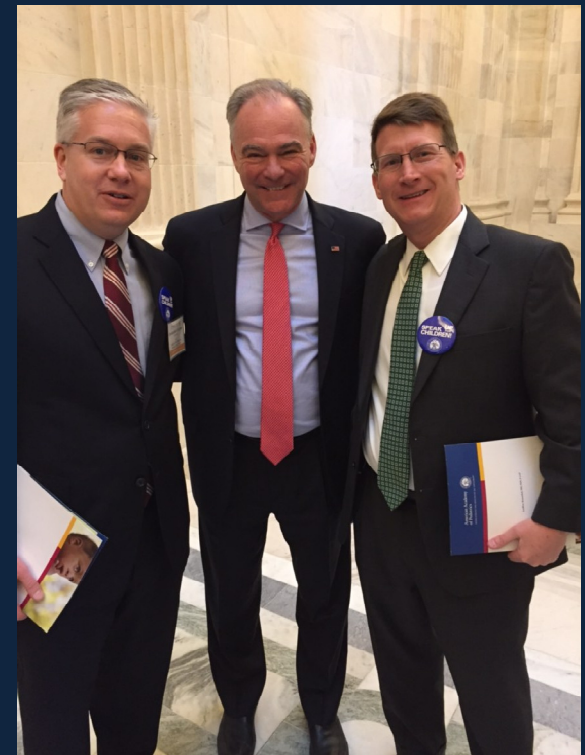
Health care

Gun laws

Suicide prevention

Pulse ox screening

Sports screening



The Congenital Heart Collaborative



# American Academy of Pediatrics

Section Of Cardiology and Cardiovascular Surgery (SOCCS) promotes the health of pediatric cardiology patients by prioritizing:

Providing optimal care

Developing research/policies

Advocating on a local and national level

Life-long education



The Congenital Heart Collaborative

 **University Hospitals**  
Case Medical Center  
Cleveland | Ohio

 **Rainbow Babies**  
& Children's Hospital

 **NATIONWIDE**  
**CHILDREN'S**  
*When your child needs a hospital, everything matters.™*

# American Academy of Pediatrics

SOCCS

Young Physician Leadership Alliance

PREP: Cardiology free for training fellows

\$35,000 Research Fellowship Award

(Children's Heart Foundation)

\$50,000 Pediatric Cardiomyopathy Research Award

(Children's Cardiomyopathy Foundation)

Mentorship

Research presentations at meeting

Interaction with "Founders"

Involvement on committees

Advocacy

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# American Academy of Pediatrics

SOCCS

## Policy and Publications

Care of Children with Congenital Heart Defects

Medical Home

Syncope in the Very Young

Prevention of Sudden Death in the Young:

What the Primary Care Physician Should Know

Genetic Basis of Common Congenital and Inherited

Heart Disease: A Guide for Pediatricians

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# American Academy of Pediatrics

## SOCCS

Advancing public policy

Congenital Heart Disease

Screening

Transparency

Public reporting

Pediatric medical devices

Educational materials

HealthyChildren.org

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# American Academy of Pediatrics SOCCS

Children's Heart Foundation Research Award

Pediatric cardiology code review

ACC Population Health Retreat

Congenital Heart Public Health Consortium

IMPAC Registry

Society of Pediatric Cardiology Training Program

The Congenital Heart Collaborative





# SOCCS Annual Program

AAP Annual Meeting, Nov. 1-3, 2018

Research presentation 55

Symposiums 4

Young Investigator Awards 3

Program directors meeting

Fellows workshop

Founder's Award Jane Newberger, MD

Publication of abstracts in CHD Journal

**2019 AAP Annual Meeting, in New Orleans**

# Background

CHDs affect affects 1% or 40,000 in the US

Critical CHD accounts for 25

No system to track CHD in the US

Over 2 million patients with CHD in US

Researchers estimated that >50% are adults

# Background

CHD is a leading cause of birth defect-associated death in infants

4.2% of neonatal deaths are due to CHD

Almost half (48%) of the deaths due to CHDs occurred before 1 year of age

# Background

Survival of infants with CHDs:  
Severity of defect  
Age at diagnosed  
How treated

Non-critical CHD survival

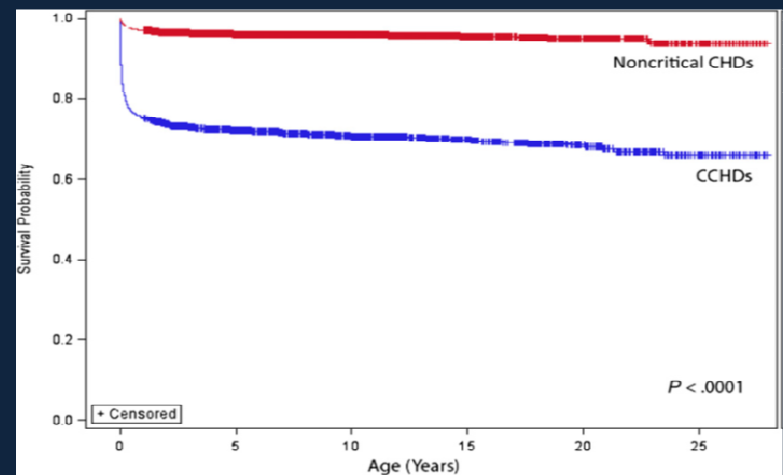
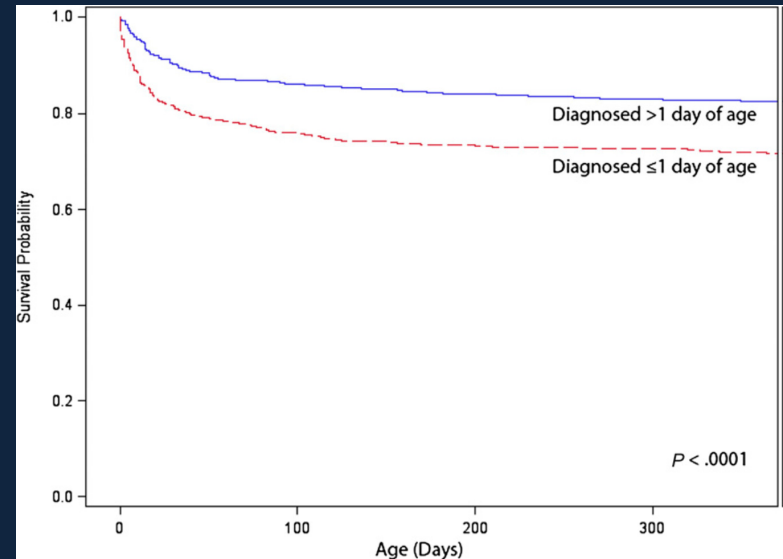
97% one year

95% 18 years

Critical CHD

75% one year

69% 18 years



# Background

In US, hospital costs for CHDs in 2004 was \$1.4 billion

These cost skyrocketed to \$1.9 billion in 2011

For insured patient in US, the “out of pocket: expenses exceed \$23,000

None of this accounts for affect on the GDP, lifestyle changes, emotional stress, and inability to return to work

Table 2. Birth defects commonly causing hospitalization, 2004\*

Principal diagnosis	Total number of hospital stays	Percentage of all stays for birth defects	Mean length of stay (days)	Mean costs (dollars)	Aggregate costs (dollars)	Percent male	Mean age	Percent died
<b>All cardiac and circulatory congenital anomalies</b>	46,500	33.5%	8.2	\$29,600	\$1,368,822,600	50.9%	19.8	2.6%
Cerebrovascular anomalies	3,500	2.5%	4.7	\$18,800	65,980,500	50.7%	39.4	1.0%
Tetralogy of fallot	2,500	1.8%	11.4	\$40,100	100,735,900	54.0%	2.7	2.2%
Ventricular septal defect	3,700	2.7%	8.1	\$27,900	104,435,100	50.8%	5.8	1.6%
Atrial septal defect	8,800	6.4%	3.1	\$15,000	132,584,700	40.7%	38.0	0.3%
Patent ductus arteriosus	2,500	1.8%	12.7	\$31,300	77,595,700	39.5%	3.5	1.6%
Coarctation of aorta	2,600	1.9%	8.7	\$29,300	75,488,300	63.0%	6.4	1.2%
<b>All digestive congenital anomalies</b>	25,800	18.5%	6.3	\$11,700	\$303,173,100	67.8%	7.5	0.4%
Pyloric stenosis	12,500	9.0%	2.5	\$4,100	\$51,256,200	80.8%	0.0	0.0%
Rectal and large intestine atresia/stenosis	2,100	1.5%	9.1	\$18,400	\$37,856,000	56.5%	0.6	0.6%
Meckel's diverticulum	2,400	1.7%	6.1	\$11,800	\$28,174,000	70.4%	31.7	0.2%
<b>All genitourinary congenital anomalies</b>	12,900	9.3%	4.1	\$8,900	\$114,709,700	52.0%	23.3	0.3%
Obstructive genitourinary defect	4,100	3.0%	3.3	\$7,400	\$30,548,200	59.4%	14.0	0.0%
Polycystic kidney	2,700	2.0%	4.8	\$11,900	\$32,434,300	51.7%	44.6	0.4%
<b>All nervous system congenital anomalies</b>	6,900	5.0%	7.5	\$16,200	\$112,164,200	44.8%	15.2	1.8%
<b>All other congenital anomalies</b>	47,000	33.8%	5.0	\$14,400	\$667,198,000	49.1%	19.8	0.7%
Cleft palate without cleft lip	2,900	2.1%	2.1	\$5,400	\$15,506,700	49.8%	2.4	0.0%
Congenital hip deformity	2,200	1.5%	3.6	\$12,500	\$26,930,200	28.6%	27.4	0.0%
Congenital anomalies of skull and facial bones	3,700	2.6%	5.5	\$16,800	\$61,329,500	60.9%	3.8	0.3%
Spondylolisthesis	8,000	5.7%	4.1	\$18,800	\$149,954,500	38.7%	56.3	0.0%
Cleft lip with or without cleft palate	4,900	3.6%	1.9	\$5,500	\$27,155,800	59.4%	4.2	0.1%
Pectus excavatum	1,700	1.2%	4.4	\$9,900	\$16,621,600	79.1%	14.8	0.0%
Congenital anomalies of larynx, trachea and bronchus	2,300	1.7%	7.5	\$13,900	\$32,646,500	59.5%	3.9	0.2%
Foot deformities	2,900	2.1%	1.8	\$5,900	\$16,873,900	56.0%	11.6	0.0%
<b>All birth defects</b>	<b>139,100</b>	<b>100.0%</b>	<b>6.3</b>	<b>\$18,600</b>	<b>\$2,566,067,700</b>	<b>53.3%</b>	<b>17.6</b>	<b>1.3%</b>

\*Based on principal diagnosis. Birth defects with less than 2,000 hospitalizations are not shown.

Source: AHRQ, Center for Delivery, Organization, and Markets, Healthcare Cost and Utilization Project, Nationwide Inpatient Sample, 2004.

# ACC/AHA Guidelines



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ACC/AHA Practice Guideline

Practice Guideline: Full Text

**ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)**

*Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons*

The Congenital Heart Collaborative



# AAP Guidelines for Care of Children with Congenital Heart Disease

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# AHA Guidelines for Care of Children with Congenital Heart Disease

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# ACC Guidelines for Care of Children with Congenital Heart Disease

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# AAP Guidelines for Care of Children with Congenital Heart Disease

## The Care of Children With Congenital Heart Disease in Their Primary Medical Home

M. Regina Lantin-Hermoso, MD, FAAP, FACC, <sup>a</sup> Stuart Berger, MD, FAAP, <sup>b</sup>  
Ami B. Bhatt, MD, FACC, <sup>c</sup>  
Julia E. Richerson, MD, FAAP, <sup>d</sup> Robert Morrow, MD, FAAP, <sup>e</sup> Michael D.  
Freed, MD, FAAP, FACC, <sup>f</sup>  
Robert H. Beekman III, MD, FAAP, FACC, <sup>g</sup> SECTION ON CARDIOLOGY,  
CARDIAC SURGERY



# Role of Pulse Oximetry in Examining Newborns for Congenital Heart Disease: A Scientific Statement from the AHA and AAP

## CONTRIBUTORS:

William T. Mahle, MD, FAHA, FAAP, Chair, Jane W. Newburger, MD, MPH, FAHA, FAAP, G. Paul Matherne, MD, FAHA, FAAP, Frank C. Smith, MD, Tracey R. Hoke, MD, FAAP, Robert Koppel, MD, FAAP, Samuel S. Gidding, MD, FAHA, FAAP, Robert H. Beekman, III, MD, FAHA, FAAP, Scott D. Grosse, PhD, on behalf of the AMERICAN HEART ASSOCIATION CONGENITAL HEART DEFECTS COMMITTEE OF THE COUNCIL ON CARDIOVASCULAR DISEASE IN THE YOUNG, COUNCIL ON CARDIOVASCULAR NURSING, AND INTERDISCIPLINARY COUNCIL ON QUALITY OF CARE AND OUTCOMES RESEARCH; AND THE AMERICAN ACADEMY OF PEDIATRICS SECTION ON CARDIOLOGY AND CARDIAC SURGERY, AND COMMITTEE ON FETUS AND NEWBORN

## KEY WORDS

diagnostic techniques and procedures, outcomes research, health policy, pulse oximetry, heart defects, congenital, tests

The findings and conclusions in this report are those of the authors and do not necessarily represent the views of the Centers for Disease Control and Prevention.

The American Heart Association and the American Academy of Pediatrics make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest.

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on November 14, 2008, and by the American Academy of Pediatrics on March 25, 2009.

The American Academy of Pediatrics requests that this document be cited as follows: Mahle WT, Newburger JW, Matherne GP, Smith FC, Hoke TR, Koppel R, Gidding SS, Beekman RH 3rd, Grosse SD, on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Interdisciplinary Council on Quality of Care and Outcomes Research; and the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery, and Committee on Fetus and Newborn. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the American Heart Association and American Academy of Pediatrics. *Pediatrics*. 2009;124(2):823–836.

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## abstract

**BACKGROUND:** The purpose of this statement is to address the state of evidence on the routine use of pulse oximetry in newborns to detect critical congenital heart disease (CCHD).

**METHODS AND RESULTS:** A writing group appointed by the American Heart Association and the American Academy of Pediatrics reviewed the available literature addressing current detection methods for CCHD, burden of missed and/or delayed diagnosis of CCHD, rationale of oximetry screening, and clinical studies of oximetry in otherwise asymptomatic newborns. MEDLINE database searches from 1966 to 2008 were done for English-language papers using the following search terms: congenital heart disease, pulse oximetry, physical examination, murmur, echocardiography, fetal echocardiography, and newborn screening. The reference lists of identified papers were also searched. Published abstracts from major pediatric scientific meetings in 2006 to 2008 were also reviewed. The American Heart Association classification of recommendations and levels of evidence for practice guidelines were used. In an analysis of pooled studies of oximetry assessment performed after 24 hours of life, the estimated sensitivity for detecting CCHD was 69.6%, and the positive predictive value was 47.0%; however, sensitivity varied dramatically among studies from 0% to 100%. False-positive screens that required further evaluation occurred in only 0.035% of infants screened after 24 hours.

**CONCLUSIONS:** Currently, CCHD is not detected in some newborns until after their hospital discharge, which results in significant morbidity and occasional mortality. Furthermore, routine pulse oximetry performed on asymptomatic newborns after 24 hours of life, but before hospital discharge, may detect CCHD. Routine pulse oximetry performed after 24 hours in hospitals that have on-site pediatric cardiovascular services incurs very low cost and risk of harm. Future studies in larger populations and across a broad range of newborn delivery systems are needed to determine whether this practice should become standard of care in the routine assessment of the neonate. *Pediatrics* 2009;124:823–836

Congenital heart disease occurs in 9 of every 1000 livebirths.<sup>1</sup> Approximately one quarter of these children will have critical congenital heart disease (CCHD), which by definition requires surgery or catheter intervention in the first year of life.<sup>2</sup> Congenital malformations are one of the



# AAP Guidelines for Care of Children with CHD

- Very large organization
- Focused on the care of children and families
- Public health officers

Desires to be inclusive

- Establishing strict guidelines can be
  - Exclusive
  - Ridgid
  - Often outdated

# Echocardiography

The AAP endorses the Intersocietal Accreditation Commission (IAC) for Echocardiography Laboratories



ICAEL accreditation then assure pediatric echocardiography laboratory standards are met

# Catheterization lab

Ensure high-quality care

A board-certified pediatric cardiologist with additional training (or experience) in catheterization and interventional procedures

Director responsible for all aspects of administration and function of the laboratory

Technicians trained in pediatric catheterization, staffing all procedures

# Quality

Promote high-quality outcomes

All pediatric cardiovascular centers should:

Participate in a regional health care network

Utilize modern information technology

Maintain adequate case volumes to achieve and demonstrate high-quality outcomes