



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

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Congenital heart disease (CHD) is the most common birth anomaly. With advances in repair and palliation of these complex lesions, more and more patients are surviving and are discharged from the hospital to return to their families. Patients with CHD have complex health care needs that often must be provided for or coordinated for by the primary care provider (PCP) and medical home. This policy statement aims to provide the PCP with general guidelines for the care of the child with congenital heart defects and outlines anticipated problems, serving as a repository of current knowledge in a practical, readily accessible format. A timeline approach is used, emphasizing the role of the PCP and medical home in the management of patients with CHD in their various life stages.

## abstract

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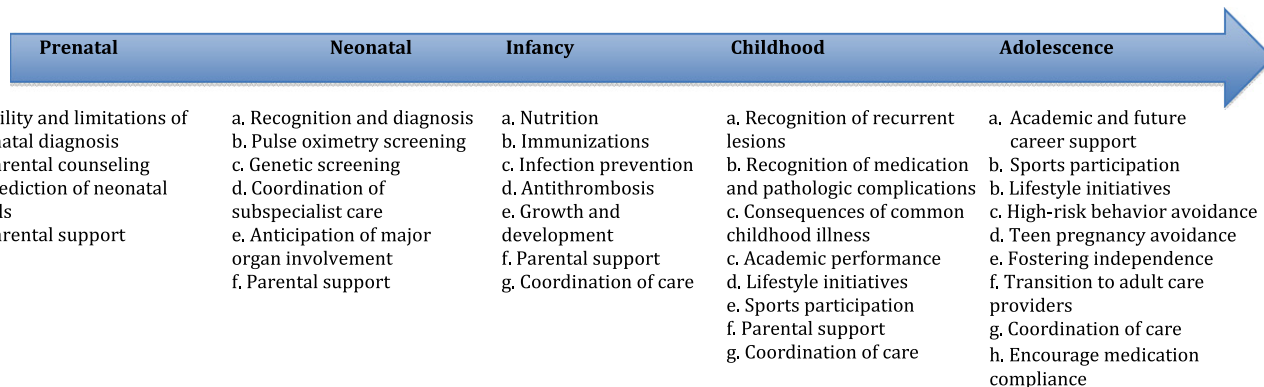
Drs Berger and Richerson contributed to the refinement of the design, the writing of specific portions of the manuscript (neonatal and cardiopulmonary resuscitation recommendations and infection, childhood, immunizations, growth, and development, respectively), and the critical review of the manuscript; Dr Bhatt contributed to the refinement of the concept and design, the writing of specific portions of the manuscript (adolescence, transition, and exercise), and the critical review of the manuscript; Dr Morrow contributed to the refinement of the concept, design, and outline of the paper and to the critical review of manuscript; Drs Freed and Beekman contributed to the refinement of the concept, design, and outline of the paper; to the editing of specific portions (exercise and exercise and final recommendations, respectively), and to the critical review of manuscript; Dr Lantin-Hermoso contributed to the original idea, concept, and design of the paper; to the outline of the topics, to the presentation of the proposal to the American Academy of Pediatrics (AAP), to ensuring all deadlines and requirements were met, to writing, to the compilation of other authors' contributions, to editing, to the critical review of the manuscript, and to the response to the AAP reviewers; and all authors approved the final manuscript as submitted.

Primary care providers (PCPs; physicians, physician assistants, and nurse practitioners) working in medical homes (MHs) are tasked with providing and coordinating the multiple health care needs of patients with congenital heart disease (CHD). With the goals of improving patient outcomes and influencing the care of these children, the American Academy of Pediatrics (AAP) and the American College of Cardiology (ACC) assembled a team of experts, including representation from the AAP Committee on Practice and Ambulatory Medicine, to review current literature and to develop this policy statement. A representative from the AAP's Family Partnership Network was requested to review this document. The role of the PCP and family engagement across the life span is emphasized.

## BACKGROUND

Advances in treatment and palliation of CHD result in more patients surviving and being discharged from the hospital with their families.

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**FIGURE 1**  
Targets of PCP engagement across the life span.

Previously unrecognized familial psychosocial stressors, racial and socioeconomic outcome disparities, other organ system involvement, complications of therapy, feeding challenges, neurodevelopmental concerns, and other special needs have been garnering more attention. Recommendations regarding newborn pulse oximetry or critical CHD (CCHD) screening and infection prevention strategies in children with CHD have been introduced or updated. Patients with CHD have complex health care needs that are to be provided for or coordinated for by their PCP. This policy statement aims to be a repository of available new information for the care of children with CHD, highlighting anticipated problems. A chronologic timeline approach (Fig 1) is used, emphasizing the role of the PCP and/or MH in the management of patients with CHD and their families in various life stages. Frequent communication among all care providers is recommended.

### General Issues Affecting Families of Children With CHD

#### Psychosocial Issues

In response to the stress of their child's serious cardiac illness, some parents report an increased incidence of emotional distress and psychosocial risk affecting optimal parenting<sup>1</sup> that may also affect the care of other children in the home. This may

**TABLE 1** Selected Advocacy/Parental Support Groups and Resources

Adult Congenital Heart Association: <a href="http://www.achaheart.org/">http://www.achaheart.org/</a>
Anthony Bates Foundation: <a href="http://www.anthonybates.org">http://www.anthonybates.org</a>
Got Transition/Center for Health Care Transition: <a href="http://www.gottransition.org/">http://www.gottransition.org/</a>
It's My Heart: <a href="http://www.childrensheartfoundation.org/publications/its-my-heart">http://www.childrensheartfoundation.org/publications/its-my-heart</a>
Little Hearts: <a href="https://www.littlehearts.org">https://www.littlehearts.org</a>
Linked By Heart: <a href="http://linked-by-heart.org/">http://linked-by-heart.org/</a>
Mended Little Hearts: <a href="http://www.mendedlittlehearts.org/">http://www.mendedlittlehearts.org/</a>
Nick of Time: <a href="http://nickoftimefoundation.org">http://nickoftimefoundation.org</a>
Parent Heart Watch: <a href="http://www.parentheartwatch.org/">www.parentheartwatch.org/</a>
Pediatric Congenital Heart Association: <a href="http://conqueringchd.org/">http://conqueringchd.org/</a>
Project ADAM: <a href="http://www.chw.org/childrens-and-the-community/resources-for-schools/cardiac-arrest-project-adam/">http://www.chw.org/childrens-and-the-community/resources-for-schools/cardiac-arrest-project-adam/</a>
Simon's Fund: <a href="http://www.simonsfund.org">http://www.simonsfund.org</a>
Sisters-By-Heart: <a href="http://www.sisters-by-heart.org/">http://www.sisters-by-heart.org/</a>
Tiny Smiles: <a href="http://www.tinysmiles.net">http://www.tinysmiles.net</a>

be exacerbated by parental guilt, financial and marital stress, challenges in parental work-life balance, and adversity because of poverty, neglect, environmental violence, or caregiver substance abuse or mental illness.

Disparate outcomes have been observed across racial and socioeconomic classes.<sup>2,3</sup> A PCP/MH with long-standing relationships with the family may be the first to recognize danger signs and be in a position to provide support and referrals to mental health professionals and groups that can provide respite care and psychological help (Table 1).

#### Need for Cardiopulmonary Resuscitation and Automated External Defibrillator Training for Qualified Caregivers

Cardiopulmonary resuscitation (CPR) and automated external defibrillator

(AED) training for qualified caregivers and family members is important. Lay CPR-AED education may be beneficial and may increase survival in out-of-hospital cardiac arrest.<sup>4</sup> Many institutions, therefore, advocate parental CPR instruction before an infant is discharged from the hospital. AED availability and CPR training before high school graduation are mandated in certain states.\*

#### Prenatal Period

CHD is often detected by fetal echocardiography, usually performed

\*For more information regarding state-specific CHD-related policies, please contact the AAP Division of State Government Affairs at [stgov@aap.org](mailto:stgov@aap.org).

at ~18 to 22 weeks' gestation. However, both false-positive and false-negative diagnoses can occur, and certain lesions may be not be detected in basic screening because some may be progressive and others may develop later in gestation.<sup>5</sup> Prenatal counseling is dependent on the gestational age, disease natural history, presence of extra cardiac anomalies, and need for fetal intervention.<sup>6</sup>

Barring obstetric complications, delivery at or close to term is preferred because of the critical development of most organ systems, particularly the fetal brain and lungs, during the last 6 weeks' gestation.<sup>7</sup> Improved outcomes are demonstrated in newborn infants with CCHD born at 39 to 40 weeks' gestation compared with those born at 37 to 38 weeks' gestation.<sup>8</sup> The best time for the family to establish relationships with primary and subspecialty care providers, and to contact support groups if desired (Table 1), is before delivery.

## Neonatal Period

### *Pulse Oximetry or CCHD Screening*

CCHD screening, added to the US Neonatal Recommended Uniform Screening Panel in 2011, may improve the timeliness of a diagnosis of CCHD that has eluded prenatal detection.<sup>9</sup> A sample protocol has been published,<sup>10</sup> but this may vary by state<sup>†</sup> or altitude. This screening targets lesions with hypoxemia, but the test is imperfect, and a neonate who "passed" might still have acyanotic, left-heart obstructive lesions.<sup>11,12</sup> Thus, symptoms such as tachypnea, an abnormally active precordium, a concerning heart murmur, or a diminution in lower

**TABLE 2** Genetic Abnormalities Associated With CHD

Common, Presently Known Chromosome Abnormalities	Select Presently Known Single Gene Defects
Cri-du-chat syndrome	Ehlers-Danlos syndrome
DiGeorge syndrome (22q11)	Ellis-van Creveld syndrome
Down syndrome (trisomy 21)	Holt-Oram syndrome
Trisomy 18 and trisomy 13	Marfan syndrome
Turner syndrome	Mucopolysaccharidoses
Wolf-Hirschhorn syndrome	Noonan syndrome
	Smith-Lemli-Opitz syndrome
	Williams syndrome

extremity pulses warrant taking a relevant history and conducting a physical examination. Depending on findings, a pediatric cardiology consultation or other diagnostic tests, such as electrocardiography or echocardiography, may be performed.

### *Genetic Screening*

When aneuploidy is suspected, children with cardiac defects will benefit from genetic evaluation to discuss prognosis and recurrence risks. Genetic testing is recommended in particular for conotruncal abnormalities, such as d-transposition, tetralogy of Fallot, truncus arteriosus, or interrupted aortic arch type B, to rule out 22q chromosome deletion (velocardiofacial or DiGeorge syndrome), because the latter may be associated with calcium metabolism, neurodevelopmental and psychiatric abnormalities, or other organ involvement. Other genetic abnormalities associated with cardiac defects are listed in Table 2.

## Infancy/Childhood

### *Care of the Patient With CHD After Hospital Discharge*

For a coordinated transition of care from the hospital to the MH, information regarding the patient's cardiac diagnosis, completed or planned interventions, residual defects, organ system involvement, and discharge physical examination findings, including weight, oxygen requirements, and baseline saturations, must be available to the

PCP. Home medications and feeding regimens, in a schedule compatible with family home routines, including formula concentration or expressed breast milk preparation and route of administration, are important to note. The need for anticoagulation and thrombosis prophylaxis for patients dependent on systemic to pulmonary artery shunts, or for those at high risk of thrombosis and stroke,<sup>13</sup> are important to communicate. The target international normalized ratio and enoxaparin levels (for patients on warfarin and enoxaparin, respectively), the reason for anticoagulation, and the duration of therapy and frequency of monitoring should ideally be outlined in discharge summaries.

Patients with heart failure, baseline cyanosis, shunt dependence for pulmonary blood flow, or single-ventricle physiology have a limited cardiopulmonary reserve and are sensitive to intravascular volume changes. These patients may decompensate rapidly during childhood respiratory or gastrointestinal infections, when respiratory function and enteral intake are impaired or fluid losses are magnified. Such patients need close follow-up with their PCP and/or MH and may require hospitalization and intravenous fluids early in their course of treatment. It would be ideal if they were identified and if recognition strategies and interventions were discussed in advance of occurrence.

<sup>†</sup>For more information regarding state-specific CHD-related policies, please contact the AAP Division of State Government Affairs at [stgov@aap.org](mailto:stgov@aap.org).

### *Nutrition and Feeding Challenges*

Adequate nutrition, necessary for growth and brain development, may be challenging in certain children with CHD. This is most apparent in infants with pulmonary overcirculation and in those who underwent stage 1 Norwood palliation for a single ventricle, who may need 120 to 150 kcal/kg per day for adequate growth. Published nutritional algorithms may serve as reference.<sup>14</sup> Some infants have limited ability for oral intake because of heart failure, suck and swallow incoordination, postoperative vocal cord injury, and airway and structural or functional neurologic abnormalities. Infants who cannot be safely orally fed may benefit from nasogastric or gastrostomy tubes. Published growth charts are available<sup>15</sup> but should be consulted while keeping in mind that some infants with heart disease may follow their own curve, and grow at a minimal rate of 5 g to 10 g per day. If growth velocity is suboptimal, fortifying the caloric density of infant formula or expressed breast milk up to 30 kcal/oz may be necessary. Potential complications in these patients may include gastroesophageal reflux, aspiration risk, osmotic diarrhea, constipation, consequences of improperly mixed formula, and in rare cases, necrotizing enterocolitis.

### *Special Immunization Needs and Infection Prevention*

Preventing, identifying, and managing infections in children with CHD are primary roles of the PCP. Current, recent, or upcoming anesthesia and surgery generally are not contraindications for immunization.<sup>16</sup> Efforts should be made to ensure vaccine administration during hospitalization or at discharge, when indicated and age-appropriate.<sup>16</sup> However, practice may vary because of concerns for a potential febrile response that may

mar the clinical picture or the ability to mount an immune response after cardiopulmonary bypass. Because children with CHD may have a lowered capacity to resist and fight infections, comprehensive routine immunizations,<sup>17</sup> including the recommended schedule for 13-valent conjugate pneumococcal vaccine, are important.<sup>‡</sup> Patients with functional asplenia should receive subsequent doses of 23-valent polysaccharide vaccine.<sup>18</sup> Some children, particularly those with heterotaxy and asplenia or nonfunctional polysplenia, will be at risk for encapsulated bacteremia, which may be prevented with daily antibiotic prophylaxis, at least until 5 years of age.<sup>19,20</sup> Recommendations for seasonal protection against respiratory syncytial virus (RSV) are available and updated regularly. Ensuring herd immunity by vaccinating close contacts, especially against pertussis and seasonal influenza, is recommended. Patients with DiGeorge syndrome would benefit from immunologic assessment of their T lymphocyte function. If a patient is found to be significantly immunocompromised, consultation with an infectious disease specialist may be considered to identify an appropriate vaccination strategy, because some patients should not receive live-virus vaccines.<sup>21</sup> Endocarditis prevention includes family education about risks and adherence to current guidelines for dental procedures involving gingival manipulation and other potentially bacteremic, high-risk interventions and surgeries. Conditions and procedures in which endocarditis prophylaxis are recommended are summarized in Table 3.<sup>22</sup> Commitment to excellent oral hygiene,<sup>23</sup> a dental home capable

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<sup>‡</sup>Please refer to Advisory Committee on Immunization Practices guidelines at [www.cdc.gov/vaccines/acip](http://www.cdc.gov/vaccines/acip).

of caring for children at high risk, application of fluoride varnish and sealants, and oral fluoride intake are important.<sup>24</sup>

### *Neurodevelopmental and Behavioral Concerns*

Some children with CHD demonstrate a higher rate of adverse neurodevelopmental outcomes<sup>25</sup> and psychological maladjustment related to low self-esteem.<sup>26</sup> This may be related to the heart disease itself, injury during earlier episodes of shock or hypoxemia, genetic issues, prematurity, prolonged hospitalization, cardiopulmonary bypass, intervention sequelae, or other factors. The PCP/MH is tasked to provide careful developmental and behavioral surveillance and screening throughout the child's life as well as ongoing assessment of academic progress. Early referrals to mental health providers and prompt treatment are important.<sup>27</sup>

### *Exercise and Sports Participation*

With normal biventricular function and the absence of hemodynamically significant residual lesions, most patients with repaired CHD will benefit from physical activity and conditioning (exercise prescription rather than restriction). Cardiovascular health will be enhanced in virtually all children with CHD by avoiding a sedentary lifestyle, obesity, and hypertension. Studies of rehabilitation and conditioning programs for patients with CHD have generally revealed benefits.<sup>28</sup> European recommendations support active lifestyles that include recreational and competitive sports for most patients with CHD.<sup>29</sup>

Some patients with CHD are at risk for cardiac decompensation and sudden death, depending on specific lesions and their severity.<sup>30</sup> Examples of conditions

**TABLE 3** Endocarditis Prophylaxis

Cardiac Conditions for Which Endocarditis Prophylaxis Is Recommended	Procedures for Which Antibiotic Prophylaxis Is Recommended	Procedures for Which Antibiotic Prophylaxis Is Not Recommended
Personal history of previous infective endocarditis	Dental procedures with gingival manipulation or oral mucosa perforation	Procedures on the genitourinary or gastrointestinal tract
Prosthetic cardiac valves	Invasive respiratory tract procedures involving mucosal incision (ie, tonsillectomy and adenoidectomy)	Routine injections through noninfected tissue
Unrepaired cyanotic CHD, including those palliated with shunts and conduits	Procedures on infected skin or musculoskeletal tissue	Prosthetic or orthodontic appliance placement without oral mucosa perforation
First 6 mo after complete repair of CHD, with prosthetic material or device		Lip bleeding or oral mucosa trauma
Repaired CHD with residual defects (persistent leaks or abnormal flows at or adjacent to prosthetic patch or device)		Loss of deciduous teeth
Heart transplant patients with abnormal cardiac valve function		Bronchoscopy without mucosal incision
		Piercing or tattooing through noninfected skin
		Vaginal delivery or hysterectomy

Adapted from Wilson W, Taubert KA, Gewitz M, et al; American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee; American Heart Association Council on Cardiovascular Disease in the Young; American Heart Association Council on Clinical Cardiology; American Heart Association Council on Cardiovascular Surgery and Anesthesia; Quality of Care and Outcomes Research Interdisciplinary Working Group. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation*. 2007;116(15):1736–1754 and Baltimore RS, Gewitz M, Baddour LM, et al; American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young and the Council on Cardiovascular and Stroke Nursing. Infective endocarditis in childhood: 2015 update: a scientific statement from the American Heart Association. *Circulation*. 2015;132(15):1487–1515.

at highest risk during strenuous activity include those with severe ventricular outflow obstruction, hypertrophic cardiomyopathy, congestive heart failure, coronary insufficiency, pulmonary hypertension, severe untreated systemic hypertension, Marfan syndrome and aortic dilation, exercise-induced arrhythmias, and long QT syndrome. Some other lesions of concern are listed in Table 4. Recommendations vary by individual patient, because risk may be altered with intervention and severity of residual lesions. Close collaboration between the cardiologist and the PCP is essential, and care must be taken to avoid giving the family conflicting advice.

#### Noncardiac Surgery

Up to 40% of children with CHD require noncardiac surgery by 5 years of age.<sup>31</sup> Severity of heart disease, cyanosis, pulmonary hypertension, or congestive heart failure increase the risk of perioperative morbidity. Those requiring inpatient noncardiac

**TABLE 4** Exercise Recommendations in CHD

Cardiac Concern	Low-Intensity Sports	Exercise Restriction
Aortic dilation, aneurysm	X	X (depending on severity)
Moderate to severely depressed ventricular function		X
Severe pulmonary HTN		X
Severe systemic HTN (untreated)		X
Severe aortic stenosis		X
Coarctation of the aorta	X	X (depending on severity)
Untreated cyanotic heart disease		X
Untreated anomalous coronary artery origin with an interarterial or intramural course		X
Exercise-induced arrhythmia		X

Adapted from Van Hare GF, Ackerman MJ, Evangelista JA, et al. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: task force 4: congenital heart disease: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015;66(21):2372–2384. Recommendations may vary by individual patient. HTN, hypertension.

surgery were found to have a two-fold higher mortality risk. In contrast, perioperative complications were fewer in ambulatory noncardiac surgeries.<sup>32</sup> Patients with CHD requiring noncardiac surgery benefit from careful evaluation and multidisciplinary planning, including a thorough understanding of their anatomy and physiology, with input from their PCP, cardiologist, anesthesiologist, and surgeon to minimize the risk of adverse events.

#### Late Childhood/Adolescence

##### Obesity

Patients with CHD are not immune to the growing trend of obesity and inactivity in North America.<sup>33,34</sup> In a study of more than 700 children with CHD, 28% were overweight and 17% had at least 1 BMI calculation indicating obesity.<sup>35</sup> The etiology of obesity is multifactorial and includes poor nutritional choices and physical inactivity from perceived handicap by the child or parent, possibly an offshoot of the “vulnerable child syndrome.”

Overweight or obese patients have a lower percent-predicted maximum oxygen consumption and a higher blood pressure response to exertion. Those who exercise experience fewer complications. Adolescents with greater risk knowledge may adopt a more favorable diet.<sup>35</sup> Physical activity and nutrition counseling are important because obesity may have unique and harmful implications in children and adolescents with CHD.

### *Risk Reduction and Transition to Adult Care*

Fostering patient engagement by providing the knowledge and skills for care participation, self-advocacy, career planning, and job suitability is an important role for the PCP. Substance abuse, teen-aged pregnancy, and information regarding appropriate contraception and safe sex practices are topics for discussion. Certain CHDs, listed in Table 5, carry a high risk of maternal morbidity and mortality.<sup>36</sup> When developmentally appropriate, giving adolescents increasing responsibility for their health and encouraging provider visits without a parent for older teenagers are important goals.

Successful transition to adult care is a multidisciplinary process that begins in the teen-aged years and is facilitated by the AAP MH initiative aiming to ensure the timely and appropriate conclusion of pediatric relationships and a shift from childhood dependence to adult responsibility.<sup>37</sup> It behooves the PCP to remind the patient, family, and other providers that a successful transition may improve outcomes. Older teenagers and young adults lost to follow-up have a higher incidence of unplanned hospital admissions via the emergency department, and gaps in cardiac care increase morbidity.<sup>38,39</sup>

### **Other Endeavors Important to Children With CHD**

#### *Electronic Health Records*

Electronic health records (EHRs) may provide accurate, up-to-date, legible,

**TABLE 5** CHD Considered High-risk for Maternal Morbidity and Mortality

1. Patients with mechanical heart valves
2. Single ventricle patients with Fontan palliation
3. Unrepaired or palliated cyanotic lesions
4. Patients with Marfan syndrome and aortic dilation
5. Severe systemic AV valve regurgitation
6. Severe left ventricular outflow tract obstruction or aortic stenosis
7. Significant pulmonary hypertension
8. Moderate to severe systemic ventricle dysfunction

Adapted from Greutmann M, Pieper PG. Pregnancy in women with congenital heart disease. *Eur Heart J*. 2015;36(37):2491–2499. AV, atrioventricular.

and accessible medical information important in the care of patients with CHD. Although cost and technology deficiencies are barriers to universal acceptance, many centers are espousing the use of EHRs in compliance with the US federal mandate that includes both incentives for implementation of EHR systems and penalties for nonadapters.<sup>40,41</sup> EHRs may prove useful to flag patients with CHD who will benefit from the recommendations described in this statement. For example, single-ventricle interstage patients may be flagged as being at a high risk for mortality and morbidity and as susceptible to dehydration and intercurrent respiratory illnesses. This is particularly important when patients present for care at new institutions. Caregivers may also be prompted to provide RSV or subacute bacterial endocarditis prophylaxis during well-child visits if an “advisory” is highlighted. EHR use facilitates growth and weight trending. Medical information portability, interoperability, and multiinstitutional sharing are helpful for continuity and test duplication avoidance. A regularly updated and accurate “cardiology coordination note” is ideal for multidisciplinary care. This note may contain contact information for care team members, the patient diagnosis, medications, baseline saturations, interventions performed, information on implants or pacemakers, the need for

infective endocarditis prophylaxis and anticoagulation, exercise restrictions if applicable, and a care plan describing the circumstances that might constitute an emergency, with specific guidance for action. This cardiology coordination note would ideally be shared among all care providers and with the MH and the family.

### *Advocacy and Legislation*

The AAP, state AAP chapters, the ACC, and other advocacy groups aim to minimize the impact of CHD. Through their efforts, some states have passed legislation that ensures that newborn infants are screened for CCHD, that students are trained in lifesaving CPR before high school graduation, that opportunities for physical activity to reduce obesity are available, that tobacco use be prevented, that insurance not be denied for preexisting conditions, and that public funding for CHD research be increased. Initiatives establishing pediatric patient-centered MHs are likewise underway. The ACC Adult Congenital and Pediatric Cardiology Section has advocated for congressional support of legislation important to the CHD community, such as the Advancing Care for Exceptional Kids Act of 2015, ongoing funding of the Children’s Health Insurance Program, and funding from the Centers for Disease Control and Prevention to support surveillance and research in CHD. The AAP Division of State Government Affairs provides state-specific information on CHD-related policies. Individual states may enact legislation or adopt regulations or standard-of-practice guidance.<sup>§</sup>

<sup>§</sup>For more information regarding state-specific CHD-related policies, please contact the AAP Division of State Government Affairs at [stgov@aap.org](mailto:stgov@aap.org).

## SUMMARY

Patients with CHD and their families have multiple needs. Care and support provided by the PCP/MH, as outlined in the recommendations below, are invaluable for improved outcomes throughout the patient's life span.

## Recommendations

1. Promote care coordination and communication among the family, PCP, and subspecialists at all times, but especially during the transition from the hospital to the home or from pediatric to adult care;
2. Advocate for infrastructure support for caregivers, and recognize stressors that can affect care across the life span;
3. Facilitate patient access to pediatric subspecialty care and medications;
4. Be up to date on pediatric basic and advanced life support. Encourage caregivers to undergo CPR training for patients at an increased risk of sudden death;
5. Augment a thorough newborn history and physical examination (including palpation of lower extremity pulses) with neonatal pulse oximetry to improve the likelihood of recognizing CCHD, acknowledging differing legal obligations to do so in various jurisdictions;
6. Facilitate nutritional support to encourage growth and development in infants;
7. Prescribe antibiotics for asplenia, seasonal RSV prophylaxis for high-risk patients, and influenza vaccination for eligible patients and family

members *when indicated*. Be cognizant of the risks of live-virus vaccines in patients with DiGeorge syndrome. Encourage dental hygiene and adhere to endocarditis prophylaxis regimens when applicable;

8. Anticipate other organ involvement, thrombosis risk, neurodevelopmental and learning difficulties, complications of therapy, and susceptibility to childhood illnesses;
9. Assist in promoting a lifestyle of good nutrition and physical activity in children and adolescents. In most cases, exercise prescription is appropriate;
10. Counsel patients against illicit drug, alcohol, and tobacco use, unprotected sex, and teen-aged pregnancy;
11. Foster self-reliance and facilitate smooth transitioning to adult health care; and
12. Support accuracy in EHRs and use them to flag patients who may benefit from the above recommendations and to help facilitate provider access to medical information, despite patient geographic mobility.

## IMPLEMENTATION

This document aims to reinforce best-practice recommendations as they pertain to children with CHD, for which diagnosis codes already exist. Time-based coding to value incremental work may be applied to the processes outlined in this statement.

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## ABBREVIATIONS

AAP: American Academy of Pediatrics  
ACC: American College of Cardiology  
AED: automated external defibrillator  
CCHD: critical congenital heart disease  
CHD: congenital heart disease  
CPR: cardiopulmonary resuscitation  
EHR: electronic health record  
MH: medical home  
PCP: primary care provider  
RSV: respiratory syncytial virus

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

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