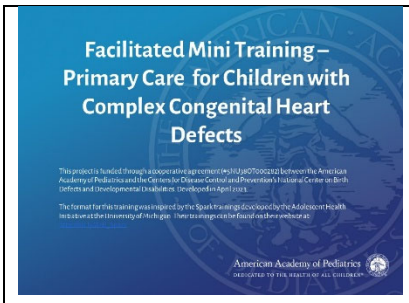
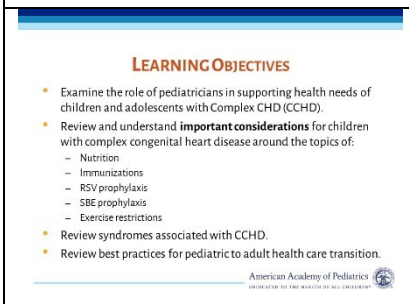
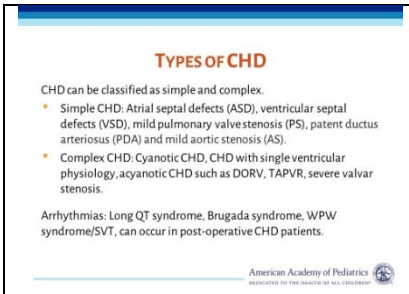
Facilitated Training – Speaker's Notes
"Primary Care for Children with Complex Congenital Heart Defects"

The American Academy of Pediatrics (AAP) has developed this **Primary Care for Children with Complex Congenital Heart Defects** resource to help pediatricians and other medical home team members care for children with complex congenital heart defects (CCHDs). The training resource examines different types of important wellness considerations, associated syndromes for patients with CCHD as well as best practices for adult transition and what role pediatricians can play. This training is intended to be facilitated by members of the practice team to encourage discussion in a short 20-25-minute presentation. This training consists of presenter slides, facilitator speaker's notes, and a case study. These materials can be presented anywhere from staff meetings to professional development opportunities.

This resource was funded through a cooperative agreement (#5NU38OT000282) between the AAP and the Centers for Disease Control and Prevention's National Center on Birth Defects and Developmental Disabilities.

The format for this training was inspired by the Spark trainings developed by the Adolescent Health Initiative at the University of Michigan. Their trainings can be found on their website at: http://bit.ly/AHI_Spark.

	**Slide 1** Today we are going to do a facilitated mini training. The idea is to encourage discussion around this topic. In this training we will explore important considerations for wellness care as it relates to **Primary care for children with Complex Congenital Heart Defects**. Before we begin our training, let's take a moment to define reproductive health. **Can anyone specify what elements might be included in care for children with complex congenital heart defects? When you think of the term "Complex Congenital Defects" what comes to mind?** **<wait for responses/discuss responses>**
	**Slide 2** Children with complex congenital heart conditions need all of the usual preventative healthcare and anticipatory guidance that children without these conditions need. However, due to their intensive needs for critical care support, surgeries and often long admissions in the first several years of life, it can be challenging for the primary care provider (PCP) to keep the child up to date with preventative care. This training will aim to highlight the role of pediatricians in supporting health needs of children and adolescents with Complex CHD (CCHD), review special considerations that apply to this population including- nutrition, vaccines/Respiratory syncytial virus (RSV) prophylaxis, Subacute Bacterial Endocarditis (SBE) prophylaxis and sports/activities. Syndromes associated with CCHD and best practices for pediatric to adult health care transition will also be reviewed.



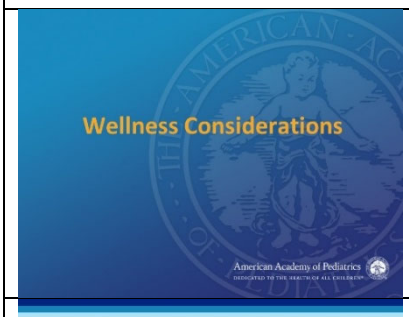
Slide 3

Congenital heart defects represent a broad spectrum of conditions ranging from simple defects with excellent prognosis to complex heart defects that require multiple procedures and have uncertain outcomes. Some of the common simple congenital heart defects are atrial septal defects (ASD), ventricular septal defects (VSD), pulmonary valve stenosis (PS), patent ductus arteriosus (PDA), and aortic stenosis (AS).

Complex CHD may be divided into those that cause cyanosis, lesions with single ventricular physiology, and acyonotic CHD such as Double Outlet Right Ventricle (DORV), Total Anomalous Pulmonary Venous Return (TAPVR), and severe valvar stenosis. CCHD are lesions for which neonates require early surgical intervention to survive.

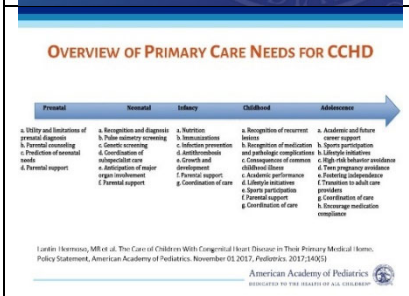
Arrhythmias also comprise a significant proportion of CHD, and in addition to long QT syndrome, Brugada syndrome, Wolf-Parkinson -Syndrome (WPW) syndrome/Supraventricular Tachycardia (SVT), can occur in post-operative CHD patients.

For the purpose of this review, the focus will be on complex CHD patients as they typically can have other associated defects and lesions requiring special attention in the medical home.



Slide 4

And so with that overview of terminology, let's start by examining the role of the medical home in the care of children with CCHD.

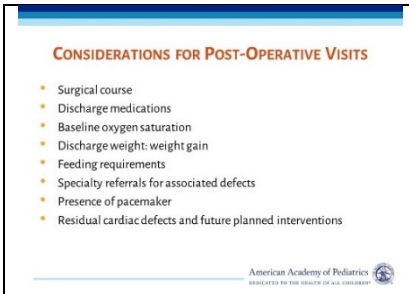


Slide 5

Has anyone had experience talking to families of children with CCHD about their care needs across the lifespan?
Did the conversation go well or not so well? What went well? What could be better?

<wait for responses/discuss responses>

Children with CCHD have complex health care needs throughout their lifespan and primary care physicians play a significant role in impacting these health care needs. This slide summarizes how focus from the medical home can shift over the course of the child's development.
Many patients with CHD are being identified prenatally and parents like to identify pediatricians well versed with these defects to have a smooth transition after discharge from the hospital. Until late childhood and early adolescence, parental support and coordinating various health care needs involving different specialists is a key role of the PCP. In late adolescence, involving the teenager in their own care and understanding the importance of lifestyle initiatives, high risk behavior avoidance and ultimately transition to adult care is at the forefront of the PCP responsibilities.



Slide 6

Is anyone familiar with post-operative considerations for patients with CCHD? If yes, which ones have you implemented?

<wait for responses/discuss responses>

In post-operative patients with CCHD, there are several important points to assess, such as their baseline oxygen saturation, discharge weight, medications, feeding, and oxygen requirements are key data points at the first office visit.

Post surgery, it is important to assess the immediate **post-operative course** as many children may have transient issues that need to be addressed such as feeding difficulties, repetition of abnormal blood work, etc.

Some of these patients may be on diuretics and consulting cardiology colleagues to determine discontinuation of these **medications** may be warranted. Such patients need close follow-up with their PCP and may require hospitalization and intravenous fluids early in their course of treatment.

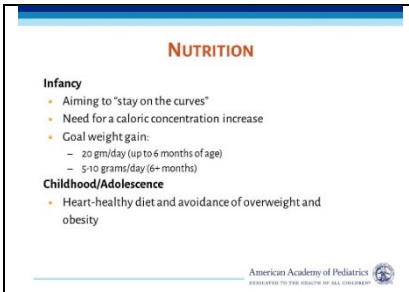
It is important to be aware of their **baseline oxygen saturations** and what their goal saturations are supposed to be as in some cases such as patients with single ventricle physiology supplemental oxygen may be detrimental to patient's health.

Assessment of **weight gain** following discharge from the hospital is important to assess feeding difficulties or need for caloric concentration.

Feeding requirements such as patients with NG feeding tubes may need referral to gastroenterology, speech therapy, physical and occupational therapy.

Assessment for associated defects such as VACTERL anomalies and may need **additional services such as referral** to other specialists.
For patients with presence of **pacemakers or other devices**, it is important to notate MRI compatibility in their chart and educate the families about it.

In addition, it is vital to be aware of **future planned surgical interventions** and timing of these interventions as it would dictate timing of routine immunization. Understanding that **residual cardiac defects** such as 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.



Slide 7

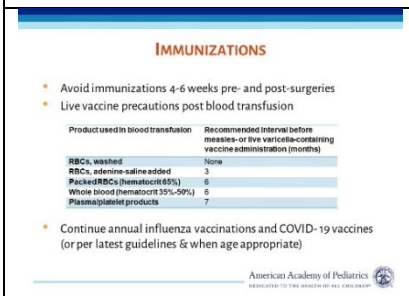
The aim is to stay on the growth curve in infants with heart disease. As those with CHD typically have increased basal metabolic demands due to inefficiency of oxygenation and increased cardiac output, it is important to be vigilant for growth failure in this patient population.

Typically, term neonates require about 100-120 kcal/kg/day of intake for adequate weight gain in infancy, for children with CCHD 120-150 Kcal/kg/day may be needed for growth. It may be difficult for a child with a shunting lesion to keep up with the 20-50% increase needed to accommodate the inefficiency of their circulation. For example, breast milk will almost certainly need to be supplemented (to up to 30 kcal/oz), and nasogastric tube or gastrostomy tube may be needed if the baby is unable to thrive without it due to increased work of breathing and metabolic demands.

Goal weight gain 20 gm/day (up to 6 months of age) is recommended. Thereafter, 5-10 grams/day in infancy should be adequate for growth. Look to your cardiology team for guidance when needed. After the initial cardiac surgeries are completed and as these infants progress through childhood and adolescence, it is important to aim for a heart healthy diet and avoidance of overweight and obesity.

Do these nutritional considerations align with what you already recommend for patients with CCHD?

<wait for responses/discuss responses>



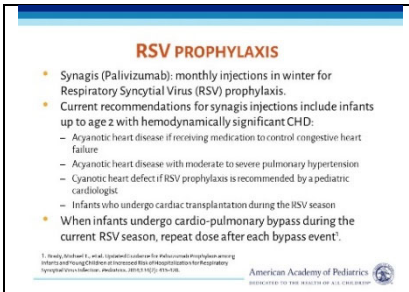
Slide 8

As pediatricians we are aware of the importance and benefits of immunization. This is exemplified in patients with CCHD due to the additional risk of hospitalization with preventable illnesses in this patient population. In CCHD, the typical schedule of childhood vaccines will probably be interrupted by necessary cardiac surgeries. Most cardiac surgical centers avoid vaccinations 4-6 weeks prior to and following surgery. It is vital to discuss this with your patients' pediatric cardiologist as different centers may have different policies.

There are certain precautions that need to be maintained post blood transfusion based on the type of blood product for live vaccines as outlined in the table shown here. Recommended intervals are extrapolated from an estimated half-life of 30 days for passively acquired antibody and an observed interference with the immune response to measles vaccine and live varicella vaccines. Live vaccines should also be avoided in 22q11 deletion patients until cleared by Immunology.

It is important to continue annual influenza vaccinations and COVID-19 vaccines throughout childhood (or per latest guidelines & when age appropriate) for these children with complex CHD.

What type of immunization challenges have you seen in your practice, when it comes to patients with CCHD? <wait for responses/discuss responses>



Slide 9

Regarding RSV (**Respiratory Syncytial Virus)** prevention, Palivizumab injections can be given monthly in the winter season for 5 injections. Be aware of the latest guidelines as they change yearly and by geographical location. Patients may need to be assessed on a case-by-case basis.

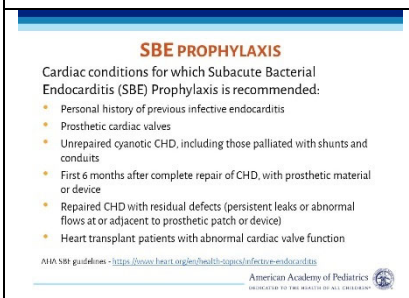
Current recommendations for infants that qualify for Palivizumab include those with hemodynamically significant CHD as outlined here.

Infants who continue to require RSV prophylaxis after cardio-pulmonary bypass should receive an additional Palivizumab dose as soon as possible after the procedure (even if sooner than a month from the previous dose). This is because, as evidence shows, a mean decrease in palivizumab serum concentration of 58% was observed after surgical procedures that involve cardiopulmonary bypass[1].

Do these RSV Prophylaxis considerations align with what you already recommend for patients with CCHD?

<wait for responses/discuss responses>

1. Brady, Michael T., et al. "Updated Guidance for Palivizumab Prophylaxis among Infants and Young Children at Increased Risk of Hospitalization for Respiratory Syncytial Virus Infection." Pediatrics, vol. 134, no. 2, 2014, pp. 415–420

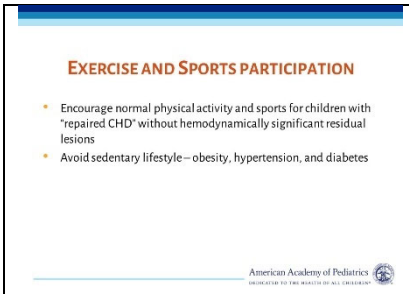


Slide 10

Subacute Bacterial Endocarditis (SBE) prophylaxis is recommended prior to gingival manipulation and other potentially bacteremic, high-risk interventions and surgeries to avoid cardiac infection. The cardiac conditions for which SBE prophylaxis is recommended are as follows: [refer to slide]

An antibiotic for prophylaxis should be administered in a single dose before invasive procedures, including dental work. If the dose of antibiotic is *inadvertently* not administered before the procedure, the dosage may be administered up to 2 hours after the procedure. Amoxicillin is the preferred choice for oral therapy because it is well absorbed in the GI tract and provides high and sustained serum concentrations. For individuals who are allergic to penicillin or amoxicillin, the use of cephalexin or another first-generation oral cephalosporin, clindamycin, azithromycin, or clarithromycin is recommended.

AHA SBE guidelines link - https://www.heart.org/en/health-topics/infective-endocarditis



Slide 11

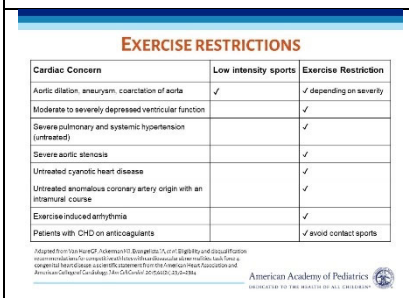
With normal biventricular function and the absence of hemodynamically significant residual lesions, most patients with repaired CHD will benefit from physical activity and conditioning and typically do not have any significant restrictions unless specifically indicated by a pediatric cardiologist.

It is important for all children with CHD to avoid a sedentary lifestyle and thus obesity, hypertension, and diabetes which can worsen their overall cardiovascular health.

Has anyone had experience talking to families and children with CCHD about exercise?

<wait for responses/discuss responses>

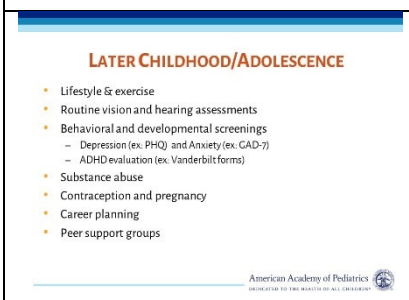
Did the conversation go well or not so well? What went well? What could be better?



Slide 12

This table goes into some concern specific exercise restrictions. As you can see - low intensity sports are recommended for the aortic dilation, and all others can do high intensity with some restrictions.

Some patients with CHD are at risk of sudden decompensation based on the type of lesion and its severity. Recommendations may vary by individual patient and should be discussed with a pediatric cardiologist. Patients with CHD on anticoagulation treatment should be advised not to participate in contact sports.

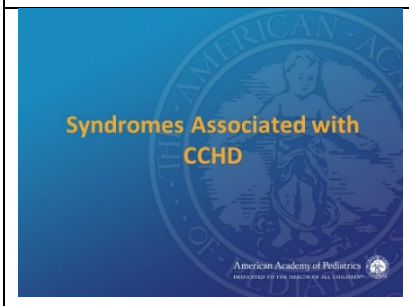


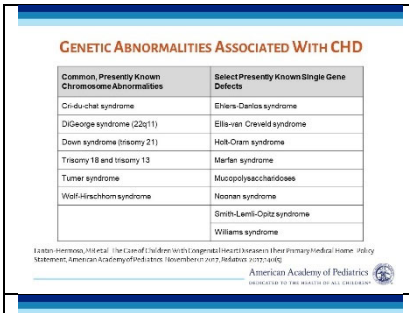
Slide 13

As discussed previously it is important to address risk factors for development of obesity early on even in patients with CCHD. It is important to introduce **exercise** prescription — "Prescription to play for 60 minutes of active physical activity per day" to avoid obesity.

Because patients with CCHD are at a high risk of hearing loss and vision abnormalities, they should undergo more frequent **hearing and vision screenings** than is routine in primary care.
Children with CCHD are also at increased risk of developmental disorders or disabilities. Periodic **developmental surveillance**, **screening**, **evaluation**, and reevaluation throughout childhood may enhance identification of significant deficits. This will allow for appropriate therapies and education to enhance academic, behavioral, psychosocial, and adaptive functioning.

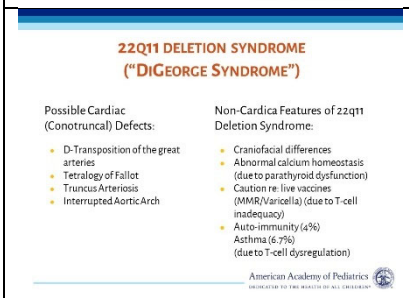
Compared to peers, children and adolescents with CHD are 5 times more likely to suffer anxiety/depression. For children with complex CHD (ex, single ventricle physiology), the risk is 7 times higher than peers.

	Slide 13 Continued… Screening for and addressing **substance abuse** and risk of teen-aged **pregnancy** (and giving information regarding appropriate contraception and safe sex practices) are important topics for discussion at adolescent primary care visits. Certain CCHDs have a very high risk of maternal morbidity and mortality. It is important to discuss **career planning** in the teens and young adult years. For CCHD, it is crucial to have health insurance coverage as an adult, as individuals with a CHD are typically followed by cardiology throughout their life. It is also important for adolescents with CHD to have **peer support groups**. Many cardiac surgical centers have specific summer camps and activities geared towards children with CCHD and the PCP can encourage adolescents to participate in these. **Which of these considerations do you already routinely apply in your practice and which do you think you could start applying?** **<wait for responses/discuss responses>**
	**Slide 14** Let's now turn our attention to identifying syndromes that may be associated with CCHD, as in these children, multiple body systems will require special attention from the PCP. **Are you aware of other types of genetic syndromes that are often associated with CHD?** **<wait for responses/discuss responses>**
	**Slide 15** Some CHDs occur in isolation, without syndromic features. In infancy, it can be very difficult to recognize facial or skeletal features that will, over time, become more apparent, and which will allow providers to recognize syndromic patterns. Thus, an infant may be recognized to have a severe cardiac malformation at birth or even prenatally, but the care team may not recognize at that early stage a relevant syndrome (which can have implications on other organ development and function, such as eyes, kidneys, and hearing) and long-term implications on development. The PCP should remain alert for the possibility of unrecognized syndromic features. The next slide briefly outlines several such syndromes for which the PCP should remain alert.



Slide 16

While it's beyond the scope of this presentation to explore these in more detail, this slide serves as a reference for some relatively common syndromes associated with CHD.

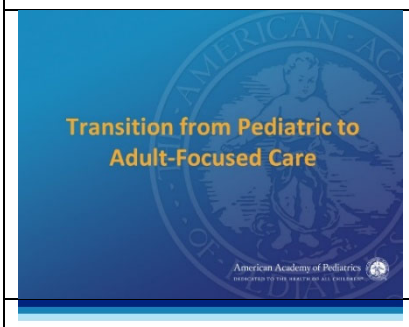


Slide 17

The chromosomal microdeletion at 22q11.2 which causes DiGeorge Syndrome (DGS) results in impaired development of the pharyngeal pouch system. Thus, patients may have developmental abnormalities of craniofacial structures, parathyroid glands, thymus and cardiac outflow tract. Since the late 1990s, patients diagnosed with outflow tract anomalies are routinely tested for 22q11.2 deletion. If not yet completed, genetic evaluation should be pursued in such patients.

It is important to evaluate the immune function of these patients, particularly in the first 3 years of life, to determine if it is safe to give live vaccines (MMR, Varicella). These should not be given unless advised that it would be safe and effective by an Immunologist. With low T-cell number/function (CD4%), not only is immune deficiency expected, but also the risk of immune dysregulation presenting as auto-immunity or atopy increases.

In one study (Deshpande et al), among the 415 patients diagnosed with DGS, 25% reported congenital heart defects, 4.1% reported autoimmunity, and 6.7% reported asthma.



Slide 18

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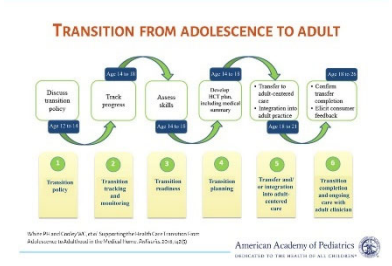
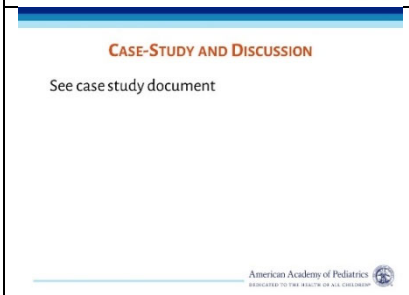
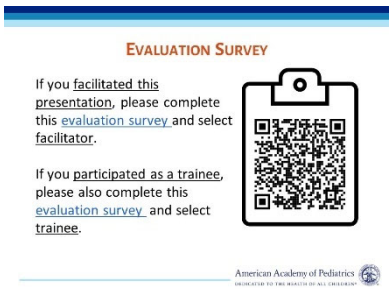
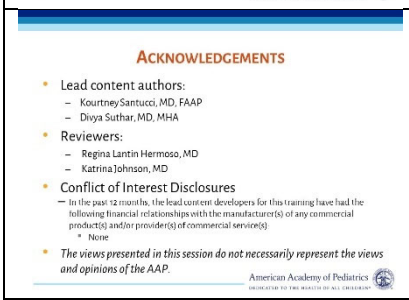
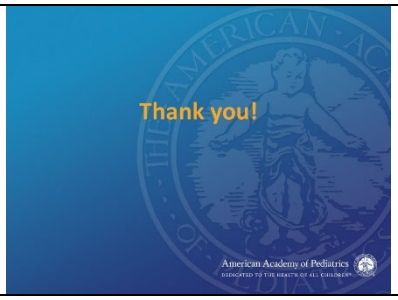


Slide 19

The 2018 report from the American Academy of Pediatrics provides new practice-based quality improvement guidance on six key elements of health care transition, and these include (1) a transition policy, (2) tracking and monitoring, (3) readiness assessment, (4) transition planning (including patient education to fill the gaps in knowledge identified by the readiness assessment), (5) transfer of care, and (6) transfer completion.

This process is typically addressed at pediatric cardiology visits, but it is important for the PCP to address any concerns that the adolescent may have regarding this, to empower patients with CHDs to successfully navigate their health care needs as adults.

	Has anyone had experience talking to families and children with CCHD about adult transition? **<wait for responses/discuss responses>** **Did the conversation go well or not so well? What went well? What could be better?** **<wait for responses/discuss responses>**
	**Slide 20**
	**Slide 21** **<call attention to the evaluation link>** **<How can [your/our] practice** improve care for Children with Complex Congenital Heart Defects? **Do you anticipate any changes to processes or workflow? What are some barriers you might encounter, and how could those barriers be mitigated?>**
	**Slide 22** **<call attention to the evaluation link>**
	**Slide 23** **<call attention to references on slides>**
	**Slide 24**
	**Slide 25** Thank you for your participation today!

For more resources view: **aap.org/CHDcare**