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Congenital Heart Disease Resource Services: Early identification

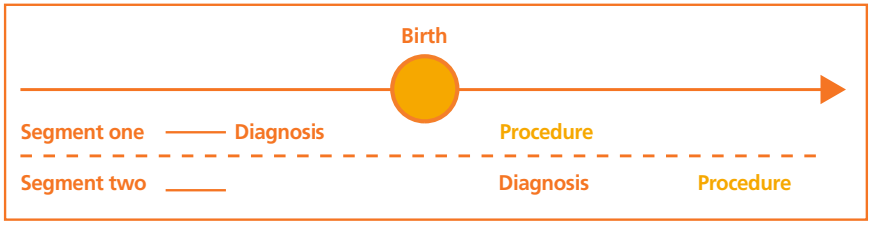
Early identification of congenital heart disease (CHD) cases is key to helping members receive Centers of Excellence (COE) care. Through early identification, case managers can proactively provide members with education to help them plan and make informed decisions before it is too late to consider quality and location.

CHD cases that can be identified early

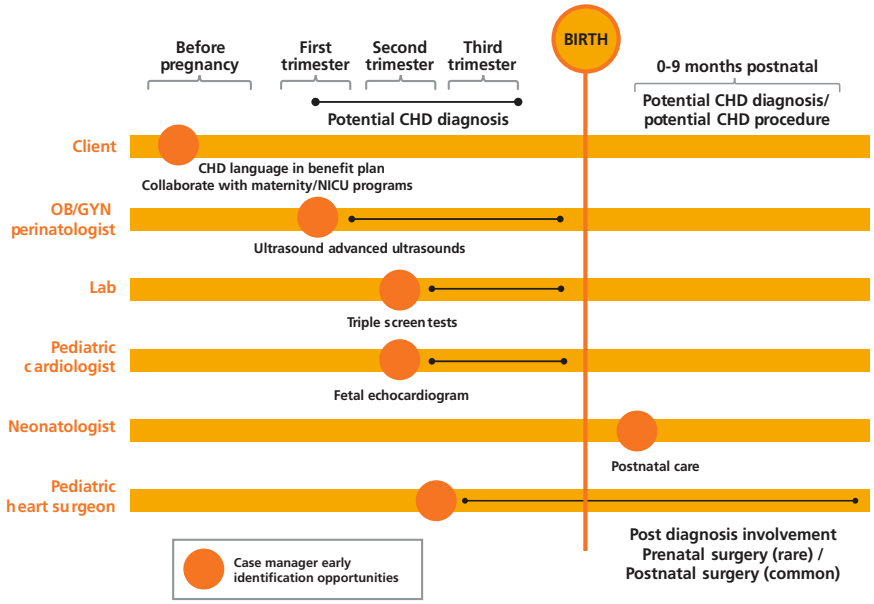
Two categories within the population of CHD cases in which complex procedures are performed in the first year of life can be identified early:

- **Segment one:** complex cases diagnosed in utero
- **Segment two:** complex cases diagnosed shortly after birth that do not undergo surgery immediately

A case identified after birth for which surgery must occur immediately is less likely to move to a different facility due to the baby's fragile state of health. Of these scenarios, early identification in utero provides the best opportunity for COE education impact.



Early Identification Timeline



Tips for early identification:

- Add CHD language to a benefit plan for parents who are diligent about checking their benefits and understand coverage during pregnancy.
- Partner with maternity management to identify high-risk pregnancies where the risk for CHD is higher.
- Work with NICU case management, which can identify babies born with CHD.
- Mine claims for ultrasounds and fetal echocardiograms tagged with CHD-related codes.

Newborn screenings for critical CHD

About one in four babies born with a heart defect has critical CHD. Babies with critical CHD need surgery or other procedures in the first year of life[1]. Screening for critical CHDs can help identify some babies with a critical CHD before they go home from the birth hospital. Newborn screening for critical CHDs is a tool that works with prenatal diagnosis and physical exams after birth to improve detection of critical CHDs. This detection allows babies to be treated early and may prevent disability or death early in life.

Importance of newborn screenings: CHD case identification

Almost all of the 50 states have passed a law or regulation that will require hospitals to screen newborns for critical CHDs[2]. Many hospitals in the remaining states choose to screen newborns, even though their state does not require it. Case managers can look for results from these screenings in order to help them identify CHD cases.

The CDC estimates that, each year, about 875 more newborns with a critical CHD could be identified at birth hospitals using pulse oximetry, a test that measures the oxygen level in a baby's blood. But, an equal number (880 babies) might still be missed each year in the United States, most of whom will have critical CHD types that are less likely to be detected through pulse oximetry newborn screening[3]. The number of babies with an undiagnosed critical CHD that might be found using pulse oximetry depends on the level of nursery care and the type of critical CHD, among other factors.

Pulse oximetry screening algorithm

A failed pulse oximetry result is an indicator to suspect the baby may have CHD and will likely need CHD care. A failed result is a crucial point at which a case manager can:

- Identify a CHD case
- Provide member education in order to guide the case to a facility that will provide the best care for the condition

To learn more about the pulse oximetry screening algorithm, visit the Centers for Disease Control and Prevention website at http://www.cdc.gov/ncbddd/heartdefects/hcp.html.

Common types of complex CHD conditions

The table below provides information on some common types of complex CHDs. For a full ICD-10 and CPT code reference list, please visit the "Training and Resources" section of the myoptumhealthcomplexmedical.com client portal or contact your account manager at 1-877-801-3507.

Diagnosis	ICD-10 code	Diagnosis description
Common arterial trunk *(Also known as: persistent truncus arteriosus)*	Q20.0	The development of only one artery forming the aorta and pulmonary veins resulting in mixed blood to the coronary arteries, pulmonary arteries, and systemic circulation.
Discordant ventriculoarterial connection *(previously called transposition of the great vessels)*	Q20.3	The position of the pulmonary artery and aorta are reversed, changing the way blood circulates through the body.
Ventricular septal defect	Q21.0	An opening between the two lower chambers that causes blood to be pumped back into the lungs rather than out to the rest of the body. One of the most common congenital heart defects.
Atrial septal defect	Q21.1	An opening that exists between the heart's two upper chambers interrupting proper blood flow.
Atrioventricular canal defect *(Also known as: atrioventricular septal defect or endocardial cushion defects)*	Q21.2	Holes between the chambers of the heart; the valves that control blood flow between these chambers may not be formed correctly. Common in babies with Down Syndrome.
Tetralogy of Fallot	Q21.3	A combination of four structural heart defects causing oxygen-poor blood to flow from the heart to the rest of the body.
Tricuspid atresia	Q22.4	Tricuspid valve between the right atrium and right ventricle is not formed resulting in restricted blood flow into the lungs.
Ebstein's anomaly	Q22.5	A downward displacement of the tricuspid valve between the right-side chambers of the heart; can lead to an enlarged right atrium and congestive heart failure.
Congenital stenosis of aortic valve	Q23.0	A narrowing of the aortic valve or the aorta directly above or below the valve making it difficult to pump blood from the heart to the body.
Congenital insufficiency of aortic valve *(Also known as: bicuspid aortic valve)*	Q23.1	The aortic valve has only two flaps (cuspids) rather than three, which is normal. There may be no symptoms in childhood, but may lead to valve narrowing later in life and require repair.
Hypoplastic Left Heart Syndrome	Q23.4	Critical underdevelopment of the left side of the heart causing severe complications.
Patent ductus arteriosus	Q25.0	Defect causing abnormal blood flow between the pulmonary artery and the aorta.
Coarctation of the aorta	Q25.1	A constriction of the aorta obstructing blood flow to all the organs of the body.
Atresia of pulmonary artery	Q25.5	Defect of the pulmonary valve in which the valve opening fails to develop, leading to obstruction of blood flow from the heart to the lungs.
Stenosis of pulmonary artery	Q25.6	Narrowing at one or more points from the right ventricle to the pulmonary artery resulting in obstruction of blood flow.
Total anomalous pulmonary venous connection (TAPVC)	Q26.2	Veins from the lungs attach to the heart in abnormal positions causing oxygenated blood to enter the wrong chambers.

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Where can I go for additional CHDRS product information?

myoptumhealthcomplexmedical.com provides you with information on the product, the Centers of Excellence network, training and resources, and more.

Sources:

1. Centers for Disease Control and Prevention. Facts about congenital heart defects. Last updated December 22, 2015. Accessed December 16, 2016. http://www.cdc.gov/ncbddd/heartdefects/facts.html. Citing: Oster M, Lee K, Honein M, Colarusso T, Shin M, Correa A. Temporal trends in survival for infants with critical congenital heart defects. Pediatrics. 2013;131(5):e1502-8.
2. American Academy of Pediatrics. Newborn screening for critical congenital heart disease (CCHD) – 2016 state actions. August 9, 2016. Accessed December 16, 2016. https://www.aap.org/en-us/Documents/2016_CCHD_Newborn_Screening_State_Actions.pdf
3. Centers for Disease Control and Prevention. Screening for critical congenital heart defects. Last updated February 11, 2016. Accessed December 16, 2016. http://www.cdc.gov/ncbddd/heartdefects/screening.html. Citing: Ailes EC, Gilboa SM, Honein MA, Oster ME. Estimated Number of Infants Detected and Missed by Critical Congenital Heart Defect Screening. Pediatrics. 2015;135(6):1000-1008.

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