July 2018

On the Uptake

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Congenital Heart Disease: What do we know today?

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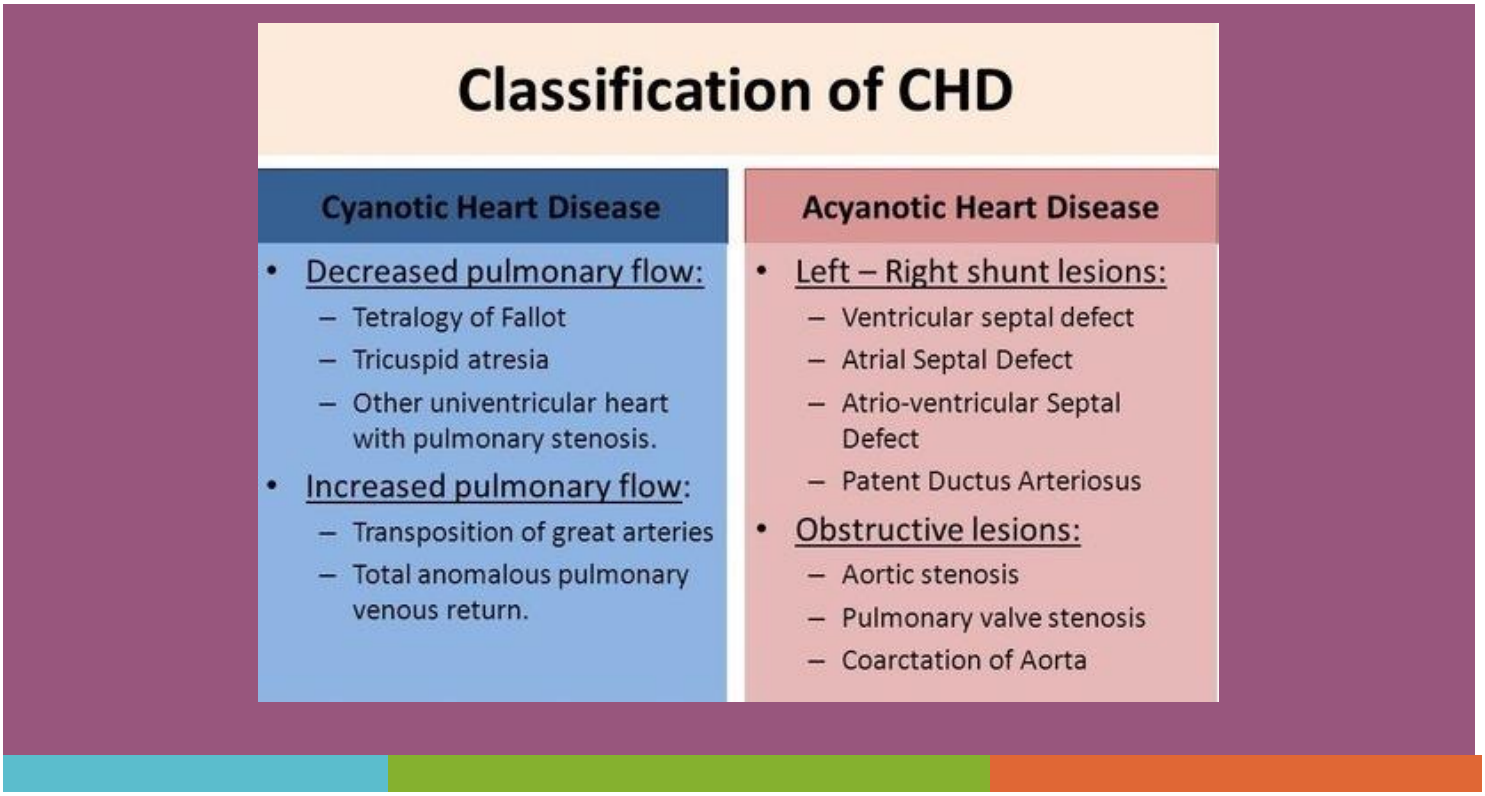
What is congenital heart disease?
Congenital heart disease (CHD) is the most common congenital malformation, accounting for about 1% of live births. The word "congenital" means existing at birth. A congenital heart defect can involve the walls or valves of the heart, or the arteries and veins near the heart.

These structural anomalies can disrupt the normal flow of blood through the heart, causing it to slow down, go in the wrong direction or to the wrong place, be blocked completely. Often, no known cause is found, but viral infections and certain inherited conditions or genetic syndromes can increase the risk of a baby to develop CHD. Fetal exposure to drugs or alcohol may also increase the risk. There are many types of CHD, ranging from simple to very complex.

Many doctors classify CHD as either cyanotic or non-cyanotic. The main difference between the two groups is that cyanotic CHD causes low levels of oxygen in the blood, which can lead to cyanosis, a bluish discoloration of the skin, lips and fingernails. Babies with non-cyanotic CHD have enough oxygen in their blood; they don't display cyanosis, but may still develop complications later in life, such as high blood pressure. In both types (cyanotic and non-cyanotic), the heart



isn't pumping blood as efficiently as it should. Severe defects can cause symptoms such as heart murmur, cyanosis, fast breathing, shortness of breath, poor feeding (especially in infants because they tire easily while nursing), poor weight gain and fatigue during exercise or activity (in older children).

Many children with CHD don't need treatment, but others do. Treatment can include medicines, catheter procedures, surgery and heart transplants. The treatment depends on the type of the defect, how severe it is, and the child's age, size and general health. Two common forms of CHD accounting for 80% of surgeries required ruing the first few weeks of life are transposition of the great arteries (TGA) and hypoplastic left heart syndrome (HLHS). Survival of children with CHD has greatly improved due to medical advances. Many decades ago, only 20% of children with complex CHD reached adulthood. Today, more than 90% survive.

Brain injury is common in children with CHD

Despite improvements in intra-operative monitoring and post-surgical care, stroke, white matter injury, cerebral sinovenous thrombosis, subdural hemorrhage, and seizures remain common. With increased usage of neuroimaging in the CHD infant we are gaining a greater understanding of the factors that contribute to the developmental impairments seen later in childhood. For example, recent studies done antenatally or postnatally demonstrate that the brain of newborns with CHD is immature as in preterm infants, suggesting that their heart condition starts to affect brain development from the fetal period.

Children with CHD are at risk for neurodevelopmental impairment

Children with CHD exhibit deficits in multiple developmental domains that are not explained by focal injuries identified before and after cardiac surgery: visual-spatial skills, memory, executive functions, language, and motor functions. The rate of developmental impairments varies between 27-40% with neurological exam abnormalities in 30%.

Knowledge gaps

While there are many factors that contribute to the outcomes of children with CHD, we continue to search for more neuroprotective and neural recovery treatments that improve the life-long functioning of children with CHD. In the developing fetus we still struggle to fully appreciate the cause for the majority of non-syndromic CHD infants, nor do genetics allow us a perfect correlation with anatomic variants. With the growth of fetal medicine in this field

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Want to learn more on the topic? Suggested readings:

1. https://www.medicinenet.com/congenital_heart_disease/article.htm#congenital_heart_defects_facts
2. Wernovsky G, Licht DJ. Neurodevelopmental outcomes in children with congenital heart disease – what can we impact? Pediatr Crit Care Med 2016; 17(8 Suppl 1):S232-42.
3. Licht DJ, Brandsema J, von Rhein M, Latal B. Neurological disorders in children with heart disease. In: Swaiman's Pediatric Neurology. Principles and Practice, 6th Edition. Elsevier 2018; Chap. 157. pp. 1205-1214.

we have gained significant insight into many potential mechanisms, yet we fail to have any treatments for these fetuses, or their mothers, that may help to advance brain maturation during this period of vulnerability.

Surgical advancements have made huge strides in the survival of infants with CHD and the current treatments can be performed safely and without major injury to the infant brain in the majority of cases. What remains relatively unknown is the ideal time during which surgical treatment maximizes brain development while continually improving survival rates. Neuroprotecting agents administered during surgery, such as xenon, metformin or melatonin, may also improve outcomes, but the utility of these agents will likely require randomized controlled trials.

As the neurodevelopmental profile of the CHD infant continues to be explored, there continues to be a greater gap in the needs of the infant beyond the acute surgical periods to that of community care with physiotherapy, occupational therapy and speech and language therapy among others.

Take home points for families

- Congenital heart disease can be life-threatning. If your child certain symptoms such as shortness of breath or fast breathing, cyanosis, fast breathing, poor feeding (especially in infants), poor weight gain and fatigue during physical activity (in older children), you should seek medical attention.
- Despite increased survival, children with CHD are at risk of brain injuries and neurodevelopmental challenges. Follow-up with your child's cardiologist, pediatrician, neurologist and therapists is important.
- Ongoing research is aiming at improving long-term outcomes in children with CHD.

Take home points for clinicians

- Congenital heart disease carries high rates of morbidity and mortality. Several modifiable risk factors are under investigations in order to optimize medical and surgical care.
- The primary issue of maturation arrest rather than cellular loss highlights the potential for brain recovery from injury. The failed attempts to improve neurodevelopmental outcomes through modifications of cardiopulmonary bypass necessitate a paradigm shift towards fetal, pre- and post-operative opportunities to improve brain health in infants with CHD.
- Randomized controlled trials are needed to test the utility of certain neuroprotective agents.