

## Neurocognitive Issues in Congenital Heart Disease

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Neurocognitive is a word that explains how our nervous system affects our behavior. Neuro refers to the central nervous system. Cognitive refers to how our brain acquires knowledge. It also describes how it handles things like emotions, identity, problem solving skills, and other behaviors.

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### Where and when does it all begin?

The heart forms and develops by 12 weeks gestation. The brain is far from developed at this time. In fact, the brain continues to develop for the first few years of life. The heart pumps blood that provides nutrition to the brain so that it can continue to grow. In turn, the brain provides nervous signals to control the heart.

### What other things affect brain development?

Other factors that affect brain development include biology, genes, and environment. In congenital heart disease (CHD), the heart defect can cause low levels of oxygen and/or abnormal blood flow to the brain. Heart surgery, catheterizations, and anesthesia needed to repair the heart defect also impact the brain. Children born with certain genetic syndromes have a much higher risk of cognitive impairment. Interactions between the environment and brain cells are also important to brain development.

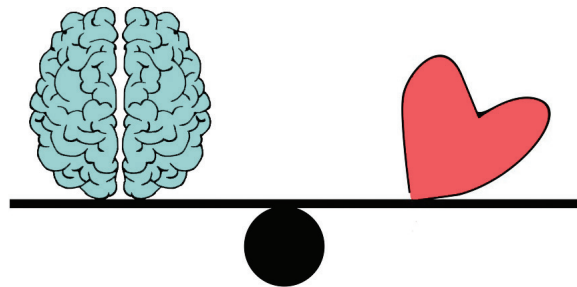
The environment includes the mother's nutritional status; exposure to alcohol, drugs and tobacco; infections; or other health conditions. Family economic status and even birth order also play a role in the growth of the brain. Brain activity, brain chemistry, and the expression of certain genes are influenced by your experiences before you were born through adulthood.

Behavioral changes occur in areas such as learning, memory, and maturation. These changes can be seen as fatigue, depression, and post-traumatic stress disease (PTSD).

CHD-related factors can affect how children with CHD develop cognitively, socially and emotionally. Illness, injury, or repeated hospitalizations that result in school absence and limited socialization or isolation can affect their development.

Litch et al, in a study in the *Journal of Thoracic Cardiovascular Surgery*, showed that the brain of full-term babies with CHD was the size of the brain of babies without CHD born at 35 weeks. This is two-thirds of the size it should be at 40 weeks. This

means that babies born with CHD are about a month behind in their brain development and are thus behind from the beginning of life. Marino et al, in a study published in *Circulation*, reported that children with more severe CHD had more severe cognitive impairment.



The American Academy of Pediatrics reviewed factors that put a child with CHD at higher risk for developmental disabilities or disorders. They found that infants who needed open heart surgery, children with cyanotic heart lesions who did not require open heart surgery as neonates or infants, and children who had CHD and other co-morbidities, such as prematurity and prolonged hospitalizations, were at a higher risk.

In children with CHD, cognitive difficulties can appear. These include shorter attention span, impulsiveness and problems with social interactions. Executive function can also be impaired. You may ask, "What is executive function?" These are skills that help us keep track of time, make plans, finish our work on time, multitask, apply previously learned information to solve other problems, analyze ideas, and look for help when we need it.

## What do we really know?

Most of what we know about neurocognitive issues in adults with CHD is from data from children. It is likely that the effects we see in children carry over into adulthood.

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We do know that neurocognitive testing in adults is most often done with tests that measure IQ. These do not tell the whole story. More detailed testing looks at areas such as concentration, reasoning, calculations, and executive function. Fewer studies have been performed using these types of tests.

IQ scores of adults with CHD range from just below average to just above average. But as the complexity of the other tests increase, patients are more likely to have difficulty completing them. These differences are subtle and can't always be interpreted from an IQ test.

Sometimes difficulties do not show up until adulthood. Brain tissue doesn't regenerate well. However, the brain has cognitive reserve. It can adapt to injury and develop new pathways to complete tasks. Cognitive reserve can get a CHD patient through school but once in the work force, he/she may encounter limits to what can be accomplished. The cumulative demand on the brain to learn and solve problems can deplete the reserve.

## What does this mean for the adult with CHD?

In school, students with CHD are more likely to receive special education and have individual learning plans and courses. A Dutch study showed that adults with CHD were less likely to have completed high school than those without CHD. The same study looked at work status. It found that patients with CHD were more likely to be working at lower level jobs and less likely to be working at scientific jobs. Madsen et al, in *Circulation* in 2014, concluded there was lower achievement at all levels of education in CHD patients than in their respective siblings.

This study also showed that CHD patients were less likely to be economically self-sufficient at age 20 and age 40, in comparison to their siblings and the general population. The ability to get insurance is challenging as well. Finally, it raised the question about how quality of life is affected.

## What can the adult with CHD do to help himself/herself?

- 1. The most important thing to know is that you are not alone. The ACHA Heart to Heart Ambassador program can connect you with trained peers who can identify with you and provide support, guidance and resources. Reach out to ACHA and ask to be matched with a Heart to Heart Ambassador.**
- 2. Talk with your ACHD provider about any educational challenges and/or employment concerns you may be having.**
- 3. If you think you are depressed, discuss this with your ACHD doctor and ask to be treated. Depression is very common in adults with CHD. There is evidence that treating depression will help cognitive function.**
- 4. Ask your provider if there are any medical treatments that may help you.**
- 5. If your provider does not ask you, be your own advocate.**
  - a. Tell him/her about learning difficulties you had in school, problems you've had on the job, or whether you have been diagnosed with ADHD or depression.**
  - b. Ask for referral for neuropsychological testing and/or vocational counseling.**

For his work on this article, ACHA thanks Stephen Crumb, DNP, AACC, of the Columbus Ohio Adult Congenital Heart (COACH) Program.