



The Emerging Recognition of School and Behavior Problems in Children with Congenital Heart Defects (CHD)

Background

Children with complex congenital heart defects (CHD) are now surviving neonatal and infant surgery with a frequency thought to be impossible only a generation ago. While advances in medicine and surgery have allowed the ability to “mend” children born with CHD, the increasing number of survivors has created a growing population of children now entering our primary and secondary school systems.

In the United States, over 35,000 infants are born each year with CHD, and more than a third of these infants are expected to undergo temporary or corrective surgical interventions in the first year of life. An estimated one million adults are now living with a variety of congenital heart defects. This figure includes over 200,000 adults living with complex CHD, meaning a CHD severe enough to require surgery in the first months of life. There are even larger numbers of children with complex CHD. As initial survival has increased significantly, greater attention has been directed toward understanding the long term behavioral problems, academic performance and functional outcomes of this growing population.

1. What are the types of difficulties that our children have, and how frequently do they occur?

As a group, children with CHD have a higher likelihood of academic, behavioral and coordination problems compared to children without CHD. This does not mean that all children with CHD have these difficulties, but the number of children is much higher than that seen in the general population. These problems seem to be more prevalent in children with complex CHD: CHD severe enough to require surgery in the first few months of life, compared to less severe forms of CHD that either don't require surgery, can be repaired during a heart catheterization (for example, an atrial septal defect or patent ductus arteriosus), or don't require surgery until later childhood.

In infants

Problems that are more prevalent in babies with CHD include feeding difficulties (perhaps in half of all children requiring neonatal heart surgery) and delays in reaching some motor milestones such as rolling over, crawling or walking. Most newborns achieve full feedings by mouth shortly after discharge from the hospital, but approximately 25% require supplemental feedings through a feeding tube into later infancy and beyond. While delays in motor skills are extremely common, most milestones are only delayed by a few months. Many hospitals now recommend speech and physical therapy consultations after newborn heart surgery for evaluation and treatment recommendations.

In preschool children who required surgery in early infancy

For this group there is a growing recognition of delays in certain elements of speech and language. To greatly oversimplify, speech and language can be broken down into two components: receptive language (the child hears and understands words), and expressive language (getting the words out with proper grammar and pronunciation). In most of our children with CHD, receptive language is normal: if you say “point to the apple”, the child will point to the apple. However, expressive language is delayed: when you point to an apple and say “what is that?”, even though the child knows it's an apple, he or she can't seem to find the word. In addition to difficulties with expressive language, some preschool children with complex CHD (probably less than one fourth) have continued difficulties with motor skills, including large motor (clumsiness) and fine motor (drawing, cutting) delays. In most children, these problems improve by the time they enter school.





1. What are the types of difficulties that our children have, and how frequently do they occur?

Children entering primary and secondary school

As these children with complex CHD are entering primary and secondary school in larger numbers, there is a growing recognition of a combination of problems that combine to cause academic and social problems. Attention deficit/hyperactivity disorder (ADHD) is a term that may be over applied in our current society, and is an extremely complicated combination of learning and behavioral interactions. Again to greatly oversimplify, the main components are impulsiveness, easy distractibility and hyperactivity. As we look more carefully at children with complex CHD, various aspects of ADHD occur in perhaps one third to one half of children, which is perhaps two to three times higher than that seen in the general population. It is not clear if continued frustrations with speech and fine motor (handwriting) control are related to the higher incidence of ADHD—in other words, which is the chicken and which is the egg?

Two related issues are now also being seen in larger numbers of school age children with CHD:

- problems with “visuomotor integration” and “executive planning”. Visual motor integration relates to the ability to coordinate thoughts and images into action. In adults, for example, driving a car represents highly complex visual motor integration. In schoolage children, however, learning handwriting represents a particular challenge: seeing handwriting on the board, knowing that it's a particular letter, and getting your hand to make the letter can be very frustrating to an otherwise bright child. As you can see, handwriting is a common theme, and many parents have told me that their child's handwriting is “one of the worst in the class”.

- Executive planning is a feature that allows us to plan out a course of action. For example: first I will get dressed, then I will go eat breakfast, then I will pack my bag, and then head off to school. As the number of tasks increases, it becomes increasingly difficult to coordinate. Some children with CHD have a particular problem in this area, but the exact frequency of this problem is currently unknown. Finally, there are no studies as of yet to determine whether ‘higher order’ intellectual functions (such as complex mathematics or learning a second language), or artistic talents (such as painting or music) will be affected in our children with CHD.

2. What are the causes of these problems?

Although it is tempting to point at one feature as the cause of the problems I've outlined above, that would be a terrible oversimplification. It is now clear that the effects on the brain of children with CHD are multiple, and cumulative over the early years of development.

Following conception, the preliminary portions of the brain form at exactly the same time that the heart forms, in the first trimester. In most cases, whatever caused the CHD has left its mark on the heart by the end of the first trimester; complex CHD is essentially determined by the end of the 8th week of gestation. In contrast, the brain continues to develop and mature throughout pregnancy. We are just beginning to learn that the abnormal circulation caused by CHD in the fetus may be responsible for brain abnormalities present at birth—before surgery and related treatments. It is now recognized that congenital heart disease and congenital brain disease go together in many children. However, it is impossible to determine which is the chicken and which is the egg. Does the same factor or factors that cause CHD cause brain abnormalities as well? Is the brain “wired” the same way in children with CHD compared to normal? Does the abnormal fetal circulation put an abnormal fetal brain at greater risk? These are questions that are only now being investigated, as technologies such as fetal and newborn magnetic resonance imaging (MRI) become more routinely available.





2. What are the causes of these problems? *(continued)*

Recent studies at The Children's Hospital of Philadelphia have revealed that the brain is either structurally or functionally abnormal at birth in up to 25% of newborns with complex CHD, and appears to develop slower than in children with a structurally normal heart. In addition, the brain of a full term baby with complex CHD resembles that of a baby without CHD born 1 month prematurely. The incidence of brain abnormalities in children with less complex CHD is considerably less.

In complex CHD, so many things happen so quickly to the newborn, that it is impossible to sort out the relative contribution of many of them to long term outcomes. These factors may include, but are not limited to:

- a low oxygen level after birth
- the effects of the heart lung machine (cardiopulmonary bypass) during surgery
- the effects of poor function of the heart before and/or after surgery
- seizures and/or stroke that may occur before, during or after surgery
- nutritional deficiencies
- the effects of a long stay in the intensive care unit and hospital

In past research, much attention was paid to the way the heart lung machine was used: how long was the operation, how low was the body temperature during surgery (hypothermia), did the circulation need to be stopped completely (hypothermic circulatory arrest) during the most delicate part of surgery, etc. While these factors are certainly important to the brain and later development, we are beginning to realize that many other factors, both before and after surgery, play perhaps an even bigger role.

A considerable amount of research has been done on the use of the heart lung machine (cardiopulmonary bypass) in general and hypothermic circulatory arrest in particular. While it is beyond the scope of this review to go into all of the details, the majority of studies have shown little to no effects for short periods of cardiopulmonary bypass and/or circulatory arrest. It would be naïve to point at a particular cutoff as being 'too long'; if one chose, for example, 45 minutes, does that mean that 44 minutes is 'safe' and 46 minutes is 'dangerous'? Of course not. However; there is general agreement that shorter periods of both cardiopulmonary bypass and hypothermic circulatory arrest are preferable to longer periods, exactly how long and in what combination continues to be a subject of research and controversy. Finally, recent studies at the Cardiac Center and throughout the world have shown that longer overall stays in the intensive care unit and hospital as a newborn are associated with worse school performance years later. This most likely reflects the cumulative effects of the CHD and its treatment.

3. What are the best treatments for these problems, and are medical therapies safe in children with CHD?

Unfortunately, the research into the treatment of the problems I've outlined above is still in its infancy. It has been only in the past couple of years that the scope of the problems of handwriting difficulties, inattention, hyperactivity, school performance and behavior has been recognized. Prior to that time, there were simply not enough children old enough who survived with different forms of complex CHD for this to be fully recognized. As a result of our research in the past 15 years, we have developed a formal protocol for following newborns after hospital discharge—the NeuroCardiac Care Program. At the current time we are also recommending formal developmental testing in infancy and the preschool years to identify potential problems and institute therapies such as speech therapy, occupational and physical therapy, and to recommend changes in learning environments that facilitate learning in children with different strengths and challenges.





3. What are the best treatments for these problems, and are medical therapies safe in children with CHD?

In my opinion, there is insufficient data at the current time to comment on the safety or potential benefits of the psychotropic drugs (for example, stimulant medications for ADHD, antidepressants, etc.) in children with CHD. Many of the psychotropic medications currently marketed for children with structurally normal hearts slightly increase the risk of rhythm problems; children with CHD are possibly at increased risk for rhythm disturbances when using these medications.

It must be emphasized that no large study has determined how safe (or dangerous) these drugs are in children with CHD, or even if they work the same way as in normal children. The decision to use medications to deal with behavioral issues or ADHD must be individualized to the child, balancing the unknown risks of these medications in children with CHD against the lifelong implications of academic and social difficulties. Close follow up with the child's cardiologist is warranted when beginning any new medication.

4. How can we decrease the frequency of these problems in the future?

In my opinion, the outlook for our children with CHD remains quite optimistic. There are certainly continued challenges in improving overall quality of life, but not only are patients surviving in numbers thought impossible one or two decades ago, but serial studies show improvement in many areas of functioning compared with older children with complex CHD repaired in the past. It must be emphasized that it is quite difficult to apply to babies born in 2009 the current outcomes in current school aged children (and college students) who were diagnosed and treated in the 1980's and 1990's. This is due to improvements in the last decade—including prenatal diagnosis, post operative care, and a better understanding of cardiopulmonary bypass. A number of additional factors will ultimately contribute to the academic success of our children, including parent and teacher expectations. It is expected that babies born today will have better outcomes than those just a generation before.

The best way to improve the outcomes for future generations is a continued partnership between patients, parents, researchers, nurses and physicians. Advocacy at the government level for continued research funding by physicians, parents and patients is crucial to continue to pursue the causes and treatment of heart disease in children, as well as the secondary effects on the brain and quality of life. Philanthropic contributions play a significant role in start up funds for research as well. Finally, if families and children are willing, voluntary participation in clinical research studies remains the cornerstone of the process.

Summary of Current Findings (2009)

- In the absence of an associated brain abnormality or genetic syndrome, cognitive function (IQ, intelligence) is typically within the normal range for essentially all children with CHD
- Parental IQ and socioeconomic status are most strongly associated with IQ in CHD patients, rather than the type of CHD or its management
- Behavior problems, especially impulse control and attention difficulties, are prevalent
- Deficits in handwriting, visual motor integration and language skills are common
- Executive functioning—the ability to plan and perform complex tasks—is impaired in some children
- The association between open heart surgery and postoperative care with later cognitive, language, or behavior difficulties continues to be an area of investigation

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