



Neurodevelopmental Issues and CHD

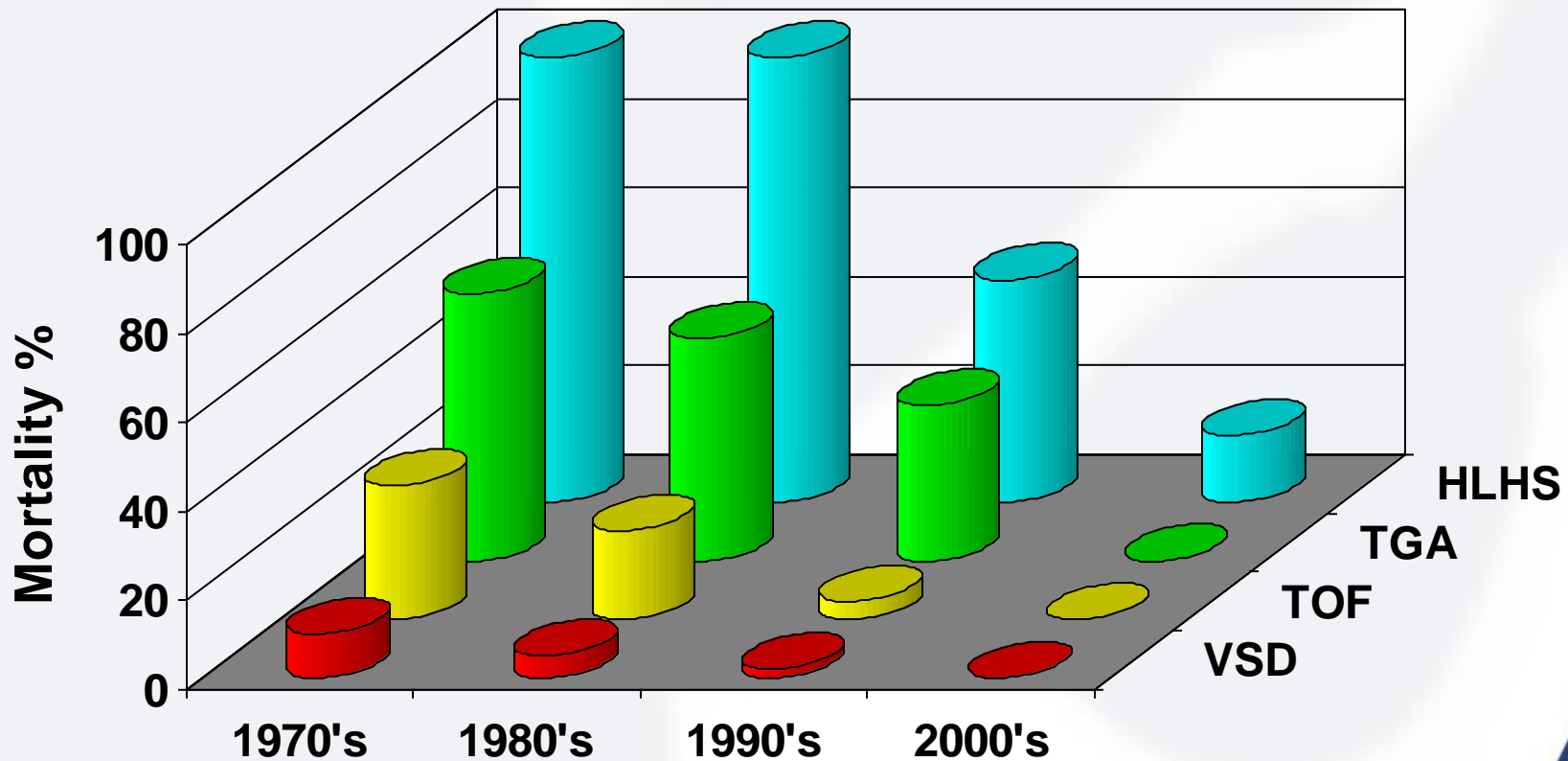
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A program of The Mended Hearts, Inc.

Background of Scientific Statement



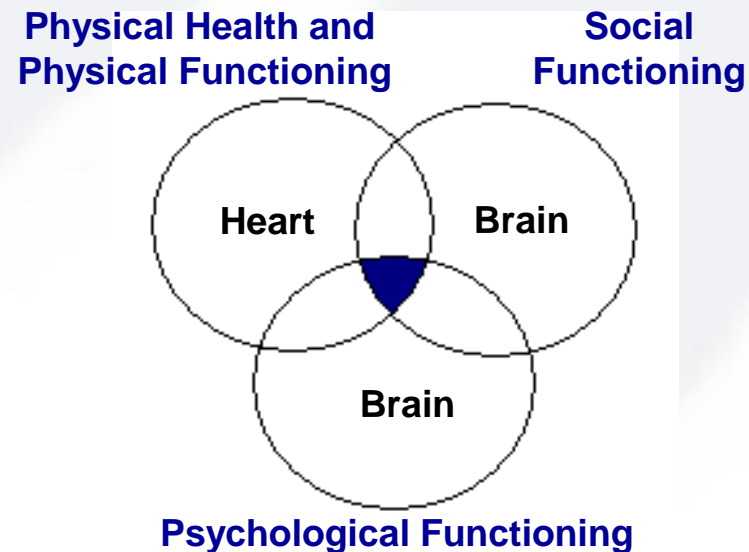
Unpublished Data - Cincinnati Children's Hospital Medical Center 2010

A look at quality of life

- As mortality decreases, we begin to look at quality of life
 - Physical
 - Psychosocial
 - **Neurodevelopmental**



- The goal is to have children with congenital heart disease function well in situational contexts (family, school, peer, etc.) and derive satisfaction from doing so.



Ware 1984,
Aaronson 1988

AHA Scientific Statement

Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management

A Scientific Statement From the American Heart Association

This statement has been approved by the American Academy of Pediatrics.

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Background—The goal of this statement was to review the available literature on surveillance, screening, evaluation, and management strategies and put forward a scientific statement that would comprehensively review the literature and create recommendations to optimize neurodevelopmental outcome in the pediatric congenital heart disease (CHD) population.

Methods and Results—A writing group appointed by the American Heart Association and American Academy of Pediatrics reviewed the available literature addressing developmental disorder and disability and developmental delay in the CHD population, with specific attention given to surveillance, screening, evaluation, and management strategies. MEDLINE and Google Scholar database searches from 1966 to 2011 were performed for English-language articles cross-referencing CHD with pertinent search terms. The reference lists of identified articles were also searched. The American College of Cardiology/American Heart Association classification of recommendations and levels of evidence for practice guidelines were used. A management algorithm was devised that stratified children with CHD on the basis of established risk factors. For those deemed to be at high risk for developmental disorder or disabilities or for developmental delay, formal, periodic developmental and medical evaluations are recommended. A CHD algorithm for surveillance, screening, evaluation, reevaluation, and management of developmental disorder or disability has been constructed to serve as a supplement to the 2006 American Academy of Pediatrics statement on developmental surveillance and screening. The proposed algorithm is designed to be carried out within the context of the medical home. This scientific statement is meant for medical providers within the medical home who care for patients with CHD.

Conclusions—Children with CHD are at increased risk of developmental disorder or disabilities or developmental delay. Periodic developmental surveillance, screening, evaluation, and reevaluation throughout childhood may enhance identification of significant deficits, allowing for appropriate therapies and education to enhance later academic, behavioral, psychosocial, and adaptive functioning. (*Circulation*. 2012;126:00-00.)

Marino et al, *Circulation* 2012

Marino et al, *Pediatrics* 2012



Scientific Statement

“The goal of this statement was to review the available literature on surveillance, screening, evaluation and management strategies and put forth a scientific statement that would comprehensively review the literature and create recommendations to optimize neurodevelopmental outcome in the pediatric congenital heart disease (CHD) population.”

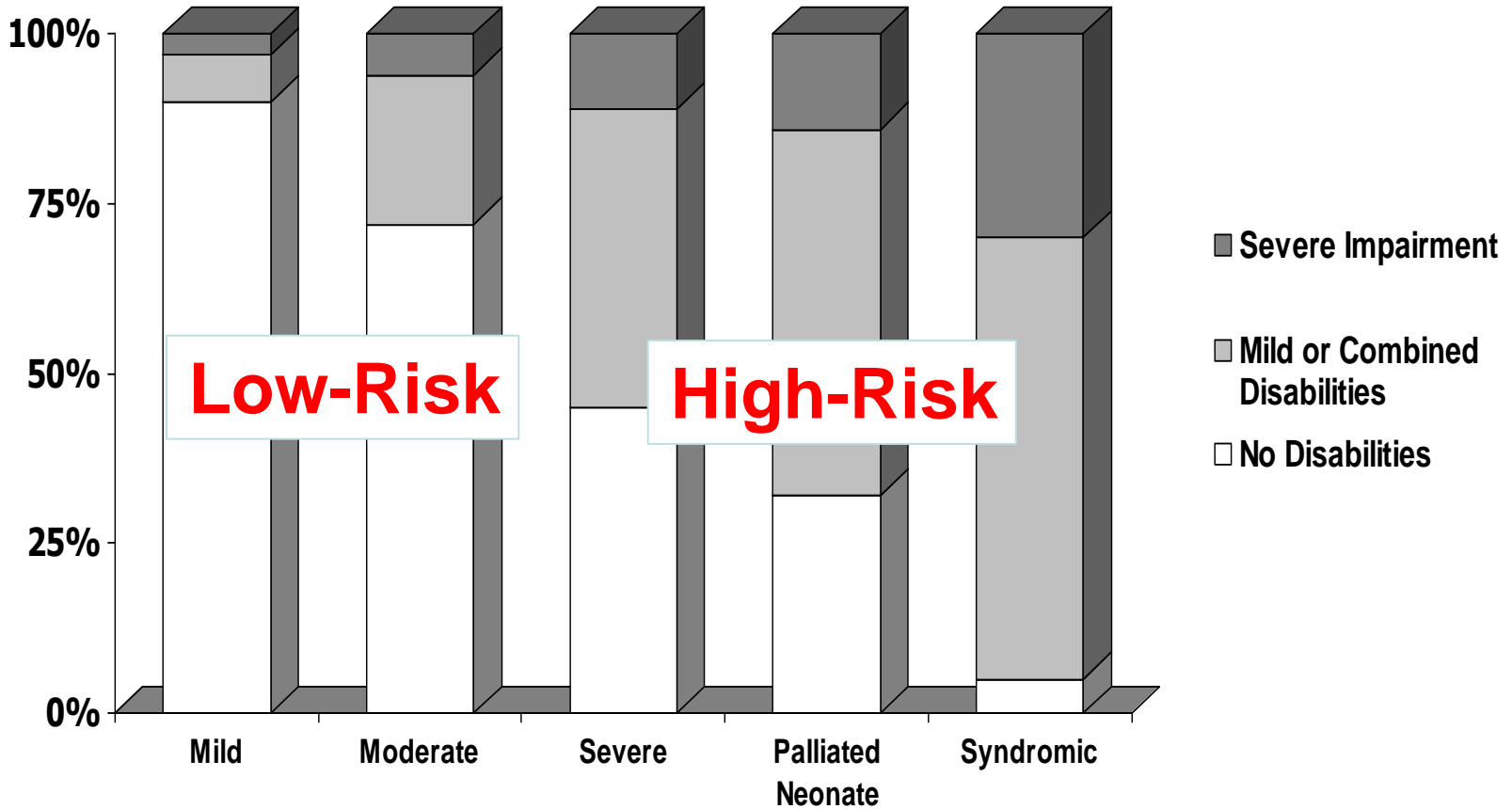


Definitions



- **“Surveillance”** – the process of recognizing children who may be at risk for developmental delay
- **“Screening”** – the use of standardized tools to identify and refine the risk recognized from surveillance
- **“Evaluation”** – a complex process aimed at identifying specific developmental disorders or disabilities that are affecting a child

Who is at risk?



Highest risk in



- Newborns or infants who had cardiopulmonary bypass during surgery (either cyanotic or acyanotic CHD types)
- Children born cyanotic even who had no open-heart surgeries as an infant.
- *Cyanotic means low blood oxygen—child may appear blue in the lips, nails or skin.*

There is more risk for children with

- CHD and ANY of the following:
 - Prematurity (< 37 weeks)
 - Developmental delay recognized in infancy
 - Suspected genetic abnormality and/or syndrome
 - History of mechanical support (ECMO and/or VAD)
 - Heart transplantation
 - CPR at any point
 - Prolonged hospitalization (>2 weeks in the hospital)
 - Perioperative seizures related to CHD surgery
 - Significant abnormalities on neuroimaging and/or microcephaly





What causes this risk?

Neurodevelopmental Definitions



- Developmental Disability (DD): The existence of a limitation or disability with the cognitive functions of the brain or the way the brain affects emotion, behavior and learning or a physical limitation.
- Developmental Delay: The child's developmental, physical and/or mental skills are not the same as children of his or her age.

Children with Complex CHD have significantly higher risks of DD



- Intelligence
- Academic achievement
- Language (development, expressive and receptive)
- Visual construction and perception
- Attention
- Executive functioning
- Fine and gross motor skills
- Psychosocial maladjustment (internalizing and externalizing problems)

Even if CHD survivor is low-risk

- Continued surveillance is critical since the level of risk can change over time
- This systematic assessment for risk should be co-managed by the primary care physician and the pediatric cardiologist and other sub-specialists



Neurodevelopmental Issues in CHD Survivors



- Mild cognitive impairment - normal to slightly lower IQ and Academic Achievement
- Impaired pragmatic language
- Decreased visual construction and perception
- Poor executive functioning
- Inattention and increased impulsivity (ADHD)
- Diminished fine and gross motor skills

CHD Survivors



- Individual deficits or delays are frequently mild, but frequently multiple
- “High prevalence – low severity” picture doesn’t meet classic criteria of a “learning disability”
- Many of these children have difficulties in school, but may not qualify for special services

CHD Survivors may have psychosocial deficits as well



- Impaired social interaction and deficits in social cognition
- Impaired core communication skills – incidence of autism spectrum disorders
- Issues with behavioral and emotional functioning

Possible social issues



- “Theory of Mind” deficits – inability to read other people
 - Difficulties interpreting others’ internal states and actions
 - Difficulties identifying and describing their own internal state
- Significant emotional and interpersonal difficulties

Screening and Management



- High-risk patients should be referred directly for formal developmental and medical evaluations
- Because different types of DDs become apparent during certain developmental periods, all patients must be monitored throughout childhood and adolescence and evaluated with age-specific tools for DDs that may show up later

Screening and Management



- ***Periodic re-evaluations*** for DDs and developmental delays at 12-24 months, 3-5 years, and 11-12 years of age
- Children over 12



What can parents and families do?

Getting Evaluated and Services



- Medical home providers should interact with
 - Early intervention programs
 - Early childhood education
 - Schools
 - Child care programs
 - Other public and private community agenciesto be certain that the special needs of the child and family are addressed through the medical home
- Special Role of the Education Specialist

Evaluation



- Referral to formal developmental and medical evaluation
- Referral to early intervention services or early childhood special education services prior to confirmation of a specific developmental diagnosis
- Periodic re-evaluations for DDs and developmental delays at 12-24 months, 3-5 years, and 11-12 years of age
- Referral of young adults for higher education and/or vocational counseling

“Helping Kids Reach Their Full Potential”

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Before school age

- Provide developmental assessment of infants, toddlers, and pre-schoolers to maximize school readiness
- Identify school difficulties, such as problems with reading, writing, and math
- Address speech and language difficulties experienced by infants, toddlers, pre-schoolers, and school-age children

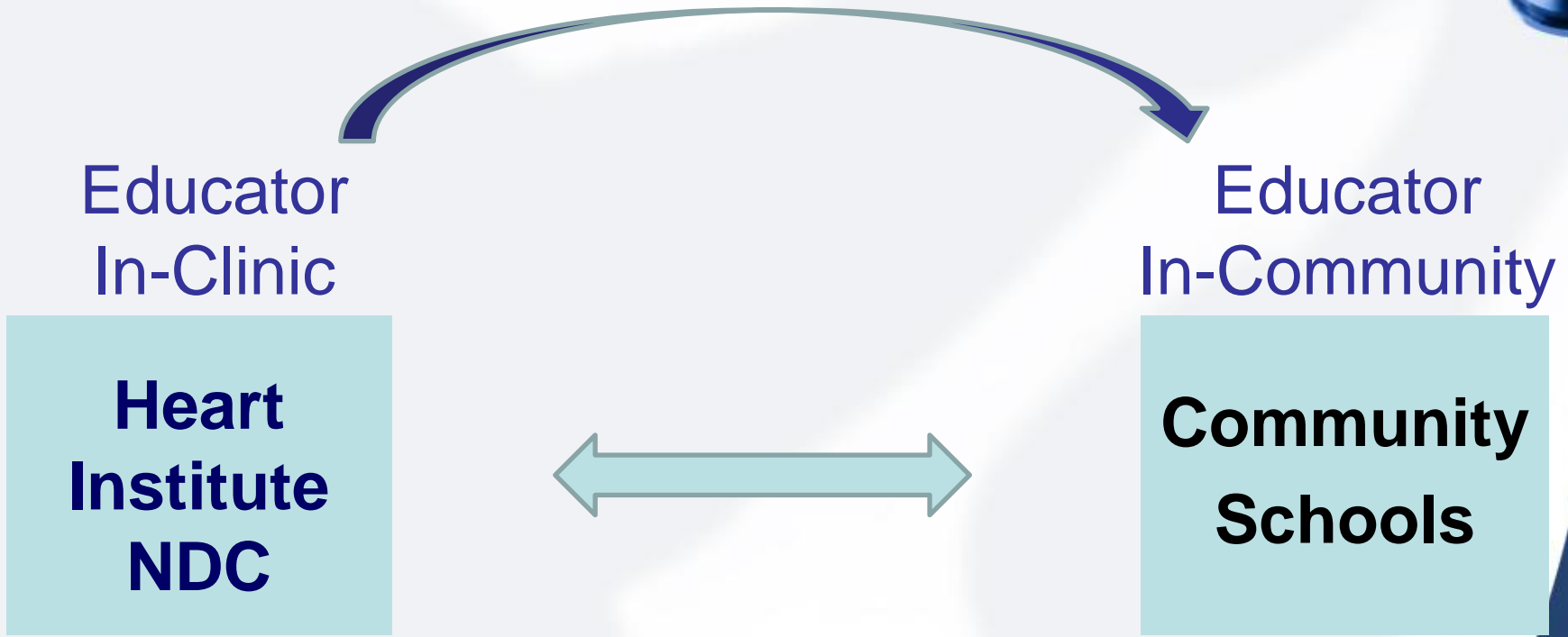


Things to look at

- Identify feeding or nutritional problems
- Address attention, behavioral, and emotional problems
- Provide intervention for difficulties with social relationships with peers and family members
- Recognize challenges with task management and organization
- Diagnose delayed fine and gross motor skills



School Intervention Program Follow Through



Lifelong Care



- **Children with moderate or great complexity CHD require life-long care**, initially by a pediatric cardiologist and later by an adult CHD specialist or cardiologist familiar with CHD.



Questions and Answers