Overview of Neurodevelopmental Outcomes in Children with Congenital Heart Disease

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Neuropsychological Function

• **Intelligence**
  – generally near population average, even for recent cohorts with complex single ventricle lesions
  – data mixed for children undergoing heart transplantation (e.g., 40% scores <85)
  – ECMO pts: 50% IQ <70

• **Speech**
  – apraxia of speech
  – BCAS neuro exam: cranial nerve abnormalities, e.g., asymmetric facial movements spontaneously or in response to commands, abnormal phonation (CA group: 21%, LF group: 8%)
Speech

• oral-motor coordination and speech planning
• reduced ability to imitate oral movements and speech sounds (e.g., “stick out your tongue,” repeat “pa-ta-ka” rapidly)
• phonological deviations, cluster reductions and simplifications, omission of medial and final consonants, sequencing (transposition of syllables)
• reduced intelligibility of connected speech, especially in absence of contextual cues
• not due to abnormalities in structures
• not due to poor hearing
BCAS Apraxia of Speech: 4 Years

CA: 33%
LF: 18%
Language Concerns

- phonological awareness (decoding)
- auditory analysis
- letter and sentence fluency
- expressive vocabulary
- pragmatic skills (connected discourse, instrumental use of language)
BCAS Language Development at 2.5 Years: Expressive Language Delay (no 2-word sentences)

- IVS-CA
- IVS-LF
- VSD-CA
- VSD-LF

Percent

Delay
Visual-Spatial Skills: BCAS

- >50% could not be scored
- transplant pts: 63% deficit in visual-motor/spatial skills

age 8 years

age 16
• **Executive Functions**
  - planning, organization, developing, testing, and modifying strategies, delaying gratification, response inhibition, monitoring behavior

• **Attention**
  - clinically significant scores on parent/teacher rating scales 3-4X higher than expected for children with complex CHD

• **Motor Skills**
  - gross and fine motor delays (25-50%)
  - heart transplant and ECMO pts at particular risk
Psychosocial Outcomes

• most children with CHD achieve good outcomes
• frequency and severity of concerns depend on:
  – type of lesion (e.g., 22q11, HLHS at higher risk)
  – age at assessment (more concerns reported as children get older)
    • BCAS: as toddlers, behavior rated better than gen pop, as 8 year olds, >1/5 behavior in range of clinical concern
  – respondent (self-reports more favorable than parent, teacher, partner, even into adulthood)
Quality-of-Life

- In mixed cohort of children with CHD (Majnemer et al., 2008), functional limitations identified in 11-17% in terms of socialization, daily living skills, communication, adaptive behavior

- In BCAS, Competence (CBCL) scores of 67% of 8-year-olds with d-TGA exceeded clinical cut-off (social adaptation, activities, schoolwork) (Bellinger et al., 2009)

- 5-10 year olds with complex CHD, 49% receiving remedial aid, 15% retained in grade (Shillingford et al., 2008)

- In 6-18 year olds who underwent Fontan, 50%-8-fold greater rates of problems with anxiety, depression, attention, learning, and behavior (McCrindle et al., 2006)
Importance of Longer-Term Follow-Up

- Hovels-Gurich et al. cohort of d-TGA pts:
  - assessments at 5 years of age detected impairment in 26% of children in one or more of the following domains: motor, cognitive, academic, language and speech, neurologic status
  - assessments at age 8-14 detected impairment in 55% of children, and 6.7% showed impairments in all domains
Conclusions

- *all* CHD pts should be considered to be at increased neurodevelopmental risk

- (Some) neurodevelopmental problems appear to *predate* reparative interventions; in some respects, problems appear to increase in severity with age

- Pattern of neurodevelopmental vulnerabilities is similar across lesions (although severity might vary)

- surveillance limited to the measurement of IQ or other global indices will result in substantial rate of false negative classifications of children with CHD with regard to neurodevelopmental risk