



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

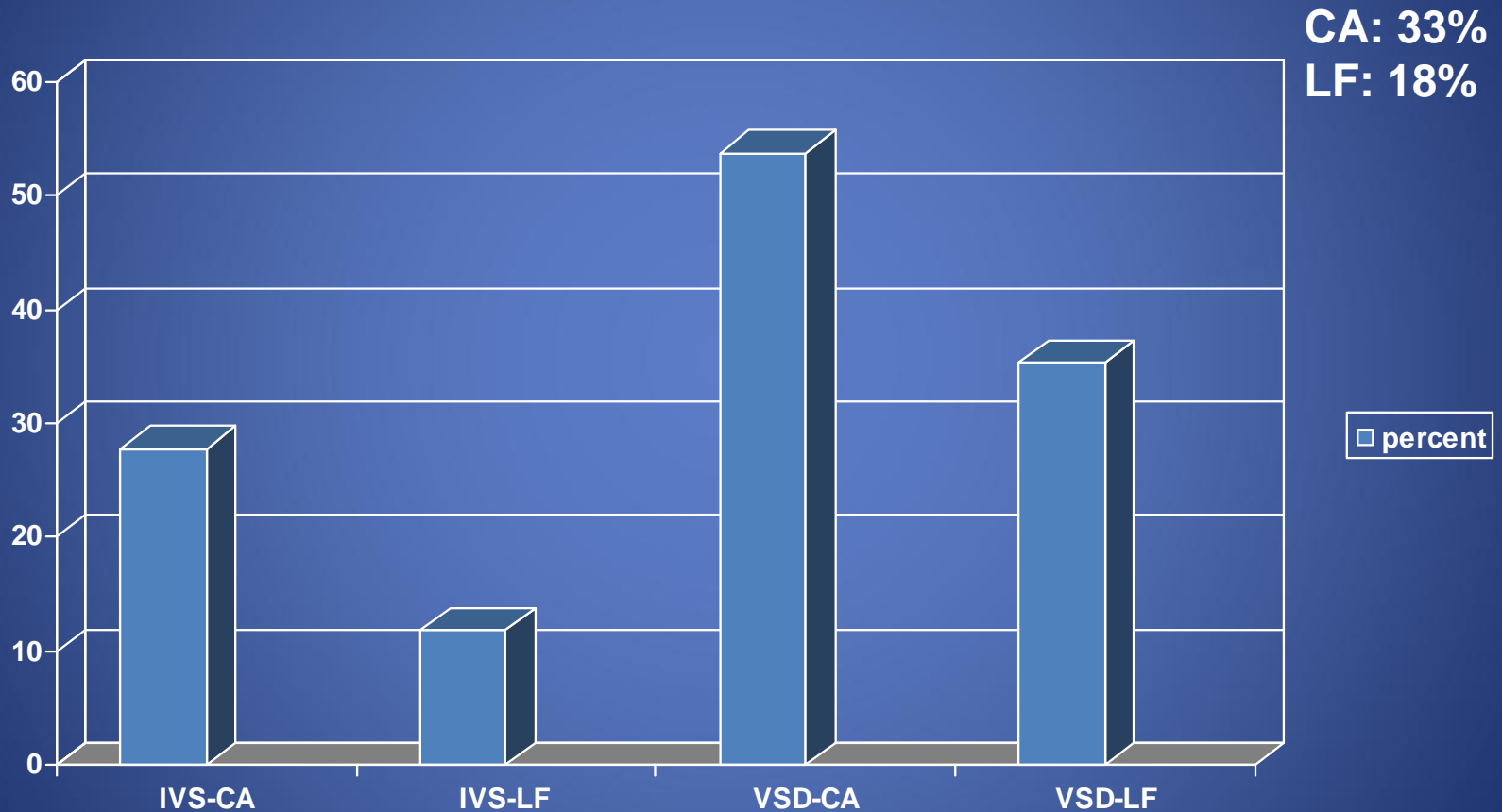
- **Speech**

- 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

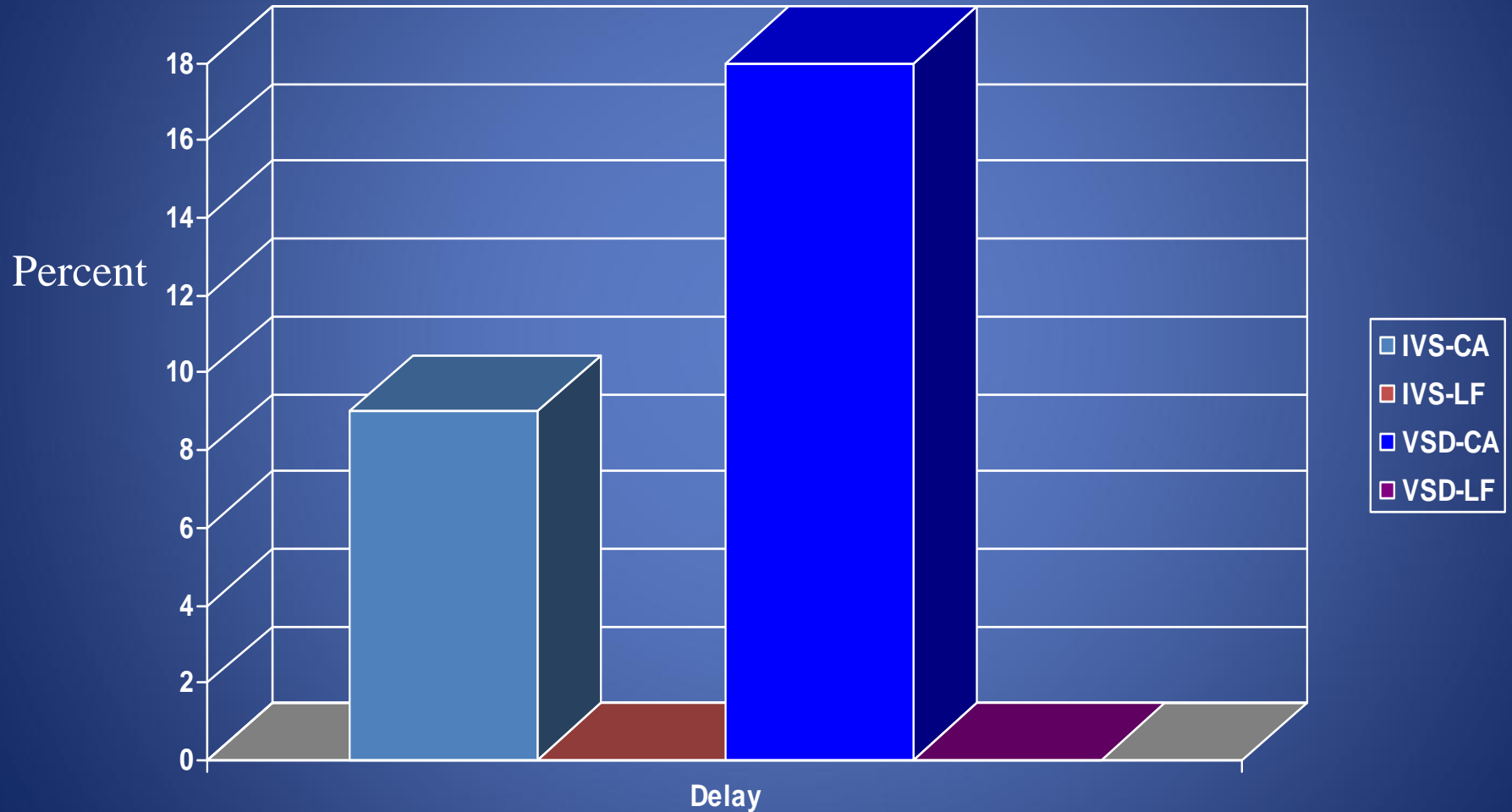
BCAS Apraxia of Speech: 4 Years



Language Concerns

- **phonological awareness (decoding)**
- **auditory analysis**
- **letter and sentence fluency**
- **expressive vocabulary**
- **pragmatic skills (connected discourse, instrumental use of language)**

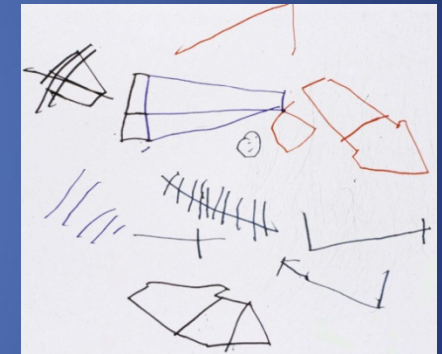
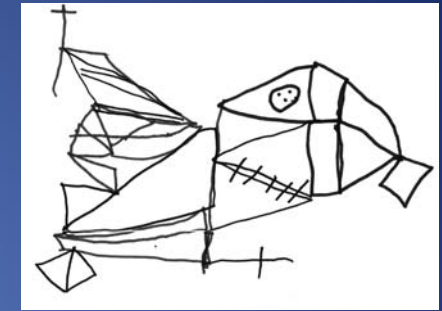
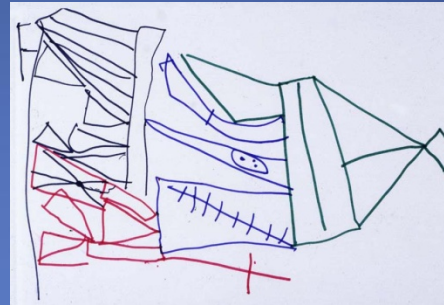
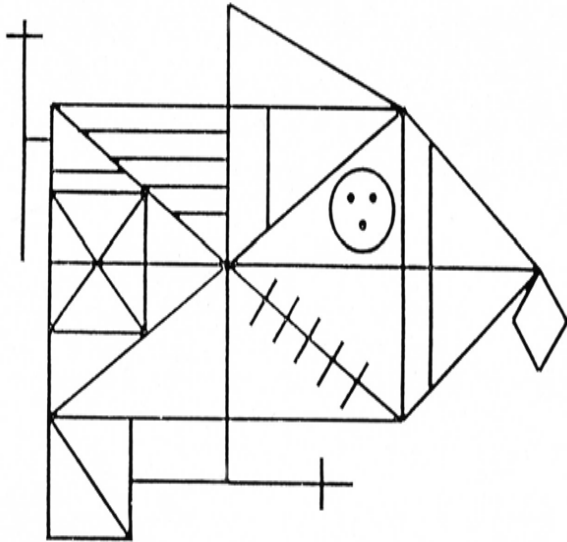
BCAS Language Development at 2.5 Years: Expressive Language Delay (no 2-word sentences)



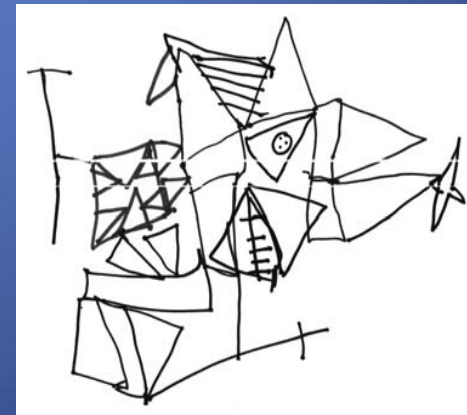
Visual-Spatial Skills: BCAS

age 8 years

REY-OSTERRIETH COMPLEX FIGURE



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



← 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