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Poster Session: Research in Progress:

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Biographical Sketch:
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Truman Coggins is an Associate Professor in the Department of Speech and Hearing Sciences at the University of Washington. Dr. Coggins serves as a research affiliate in the Mental Retardation and Developmental Disabilities Research Center at the University’s Center on Human Development and Disability where he heads Speech-Language Pathology.

Title:
Discourse errors as a signal of CNS damage in FASD

Content Area:
Speech-Language Pathology: Language & Learning in School-Age Children and Adolescents

Session Format:
SLP Poster Session: Research in Progress

Abstract:
Previous research (Thorne & Coggins, submitted; Thorne, Coggins, Carmichael-Olsen, & Astley, accepted) suggests that the rate of nominal reference errors (rNRE) in oral narratives identifies children with Fetal Alcohol Spectrum Disorders (FASD) when compared to their typically developing peers. To clarify the relationship between rNRE and other characteristics of FASD, correlations were computed between rNRE, physical features, and brain functioning for 16 children diagnosed with FASD. rNRE was more highly associated with physical features than brain function, and more highly associated with facial morphology than brain function and facial morphology are to each other. rNRE was shown to be highly accurate in predicting both FAS and partial FAS (pFAS) and associated physical markers of CNS damage, particularly microcephaly.
Current Research Results

Nominal Reference Errors:
In a recent examination of the diagnostic utility of narrative discourse (Thorne & Coggins, submitted), the rate of nominal reference errors (rNRE) in decontextualized oral narratives was found to accurately classify 32 school-aged children as being from one of two groups: 1) those with a previously diagnosed Fetal Alcohol Spectrum Disorder (all also exhibited social and/or behavior problems), or 2) Typical Development (FASD vs. TD: AUC = 0.90; 95% CI = 0.73 to 0.97; overall accuracy 88%; 3 false positive, 1 false negative). In this initial research, the rNRE was associated primarily with inappropriate pragmatic strategies for introducing new concepts into the narrative, with this type of error occurring 12 times as often as difficulties with successful creation of referential ties in the narrative. Children with FASD were more likely than their typically developing peers to use nominal reference strategies that presupposed knowledge from their listeners that was unavailable in the discourse context. The ability to accurately predict what information others have is a skill that has implications across many linguistic and non-linguistic activities, and has been associated with other developmental disabilities (e.g., autism).

Markers of FASD and rNRE:
Facial Phenotype—When measured precisely and categorized based on objective criteria, facial morphology (i.e., smooth philtrum, thin upper lip, short palpebral fissures) has been shown to be highly predictive of Fetal Alcohol Syndrome (FAS), and general central nervous system (CNS) damage in children with a FASD (Astley & Clarren, 2001). When used to predict which of the 16 narratives produced by children with a diagnosed FASD showed moderate to severe expression of the facial phenotype of FAS, rNRE was able to correctly classify all 6 children (AUC = 0.92; 95% CI = 0.67 to 0.99; sensitivity 100%; overall accuracy 88% with 2 false positives at best cut-point of 3.5%) making it a more sensitive marker of the facial phenotype than microcephaly (sensitivity 67%; overall accuracy 88% with two false negatives). If static encephalopathy (functional CNS damage) and microcephaly are combined to predict moderate to severe facial phenotype expression in the 16 children with a diagnosed FASD, only 5 of the 6 children are identified (sensitivity = 83%; overall accuracy = 75% with four false positives at best cut point of 3.7%). This makes rNRE a more sensitive marker of moderate to severe expression of the facial phenotype of FAS than microcephaly or functional static encephalopathy for this group of 16 children.

Functional Evidence of CNS damage (static encephalopathy)—4 of the 16 children in the sample with a diagnosed FASD were classified by an interdisciplinary team assessment as having functional evidence of static encephalopathy (defined by 2 or more standard deviations below mean on standardized functional measures in 3 or more domains). rNRE was only able to identify 2 of these 4 children (AUC = 0.56; 95% CI = 0.30 to 0.80; Significance level testing that rNRE is different than random test, p = 0.72).

Structural Evidence of CNS damage (microcephaly)—4 of the 16 children in the sample with a diagnosed FASD were identified as having microcephaly, an indirect measure of
structural CNS damage indicating a high likelihood of hypoplasia of one or more brain structures (Archibald et al., 2001; McGee & Riley, 2006). rNRE was able to identify all four of these children (AUC = 0.92; 95% CI = 0.67 to 0.99; Sensitivity 100%; overall accuracy 88% with two false positives at best cut point of 3.8%).

**Combined Structural Markers, FASD Facial Phenotype plus Microcephaly**—When moderate to severe evidence of the facial phenotype of FAS is combined with either evidence of structural or functional CNS damage, a diagnosis of FAS or pFAS can be given. rNRE was able to correctly predict which narratives were produced by a child with either FAS or pFAS for all 32 of the children in this sample (AUC = 0.98; 95% CI = 0.85 to 0.99; sensitivity 100%; overall accuracy 94% with one false positive at best cut point of 3.8%). If rNRE is used to predict only those children which have microcephaly and moderate to severe expression of the facial phenotype of FAS, it correctly identifies all 5 children (AUC = 0.96; 95% CI = 0.83 to 0.99; sensitivity 100%; overall accuracy 94% with one false positive at best cut point of 3.8%).

**Summary:**
rNRE was shown to accurately predict which narratives came from which members of this group of 32 children based on:

1) an existing diagnosis of a FASD and social/behavioral problems (sensitivity 94%; overall accuracy 88%)
2) moderate to severe expression of the facial phenotype of FAS (sensitivity 100%; overall accuracy 88%)
3) microcephaly (Sensitivity 100%; overall accuracy 88%)
4) moderate to severe expression of the facial phenotype of FAS combined with microcephaly (sensitivity 100%; overall accuracy 94%)
5) Full or partial FAS (sensitivity 100%; overall accuracy 94%)

rNRE was not shown to be as sensitive to functional evidence of static encephalopathy in the sample indicating that some of the children with functional evidence of CNS damage perform differently on this task than those with microcephaly or moderate to severe expression of the facial phenotype of FAS.

**Discussion**
Given that the facial morphology of FAS has been reported to be strongly associated both with particular mid-line brain structure differences (Sowell et al., 2001), and with cognitive dysfunction (Astley & Clarren, 2001), a demonstration that a particular discourse behavior is also highly associated with that facial morphology points to its potential for use in neurolinguistic studies of the function of those mid-line brain structures. The fact that our discourse measure appears to be more highly associated with the facial morphology of FAS than general cognitive function indicates that it is a skill that can be functionally separated from other cognitive skills in school-aged children, and potentially involves structural damage in regions of the brain shown to be abnormal in FAS. Preliminary evidence that rNRE is more strongly associated with structural evidence of CNS damage, particularly microcephaly, than functional evidence of static
encephalopathy indicates that higher level discourse functions may be more vulnerable to this type of damage than other areas of cognitive functioning. A rigorous demonstration of an association between this specific discourse behavior and differences in CNS structure seen in FAS would have broad implications for its use in the identification of pragmatic language difficulties in children with or without prenatal alcohol exposure. If confirmed these findings could provide an important step towards and understanding of the neurophysiology and etiology of pragmatic language difficulties.

Note: Since 1997, the Washington State FAS Diagnostic and Prevention Network has been using a comprehensive, reproducible method for diagnosing the full spectrum of outcomes resulting from prenatal alcohol exposure. The method, called the 4-Digit Diagnostic Code (Astley, 2004), utilizes quantitative, objective measurement scales and specific case-definitions to render diagnoses across the fetal alcohol spectrum. The four digits in the Code reflect the magnitude of expression of the four key diagnostic features of FAS: (1) growth, (2) facial morphology, (3) CNS damage/dysfunction, and (4) prenatal alcohol exposure. A patient's 4-Digit Diagnostic Code is derived after a thorough evaluation by an interdisciplinary team of professionals which includes a speech-language pathologist along with a physician, a psychologist, and other professionals. Language measures contribute to the CNS score along with measures from other areas of cognitive functioning such as memory and executive function.

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References