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On the Relations among Linguistic Subsystems in Typically Developing
Children and Children with Neurodevelopmental Disorders

A dissertation submitted in partial satisfaction of the
requirements for the degree Doctor of Philosophy

in

Language and Communicative Disorders

by

Lara Rosalin Polse

Committee in charge:

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2013

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Chair

University of California, San Diego

San Diego State University

2013

DEDICATION

For Dillon, my partner in life, and for Robin, Rick, Annie, Tim,
and Louis. This would not have been possible without your
constant love and support.

EPIGRAPH

`But "glory" doesn't mean "a nice knock-down argument,"" Alice objected.

`When I use a word,' Humpty Dumpty said in rather a scornful tone, `it means just what I choose it to mean – neither more nor less.'

`The question is,' said Alice, `whether you can make words mean so many different things.'

`The question is,' said Humpty Dumpty, `which is to be master – that's all.'

Alice was too much puzzled to say anything, so after a minute Humpty Dumpty began again. `They've a temper, some of them – particularly verbs, they're the proudest – adjectives you can do anything with, but not verbs – however, I can manage the whole of them! Impenetrability! That's what I say!'

`Would you tell me, please,' said Alice `what that means?'

`Now you talk like a reasonable child,' said Humpty Dumpty, looking very much pleased. `I meant by "impenetrability" that we've had enough of that subject, and it would be just as well if you'd mention what you mean to do next, as I suppose you don't mean to stop here all the rest of your life.'

`That's a great deal to make one word mean,' Alice said in a thoughtful tone.

`When I make a word do a lot of work like that,' said Humpty Dumpty, `I always pay it extra.'

– Lewis Carroll (1832-1898)
Through the Looking Glass

TABLE OF CONTENTS

Signature Page	iii
Dedication	iv
Epigraph	v
Table of Contents	vi
List of Figures	x
List of Tables	xii
Acknowledgements	xiii
Vita	xvii
Abstract of the Dissertation	xxi
Introduction	1
References	7
Chapter 1: Orthographic and Semantic Processing in Young Readers	8
Abstract	8
Introduction	8
Experiment 1	18
Methods	18
Participants	18
Materials	19
Procedure	21
Experimental Conditions	22
Results	23
Accuracy Results	23
Reaction Time Results	25
Within Grade, Between Condition Follow-up Paired Comparisons of Efficiency	26
Results from Orthographic and Semantic Conditions	27
Discussion	28
Experiment 2	32

Introduction.....	32
Methods.....	33
Participants	33
Procedure and Materials	33
Results.....	33
Accuracy Results	33
Reaction Time Results.....	34
Results from Baseline and Orthographic Conditions between Experiments 1 and 2	34
Discussion	35
General Discussion	37
Conclusion.....	42
Appendix A	43
References	44
Acknowledgements	48
Chapter 2: Single Word Reading in Children with Williams Syndrome and High Functioning Children with Autism: A Study of Contrasts.....	49
Abstract	49
Introduction	49
Cognitive Phenotypes of WS and HFA	50
Language Profiles of WS and HFA	52
Language in WS	52
Visuospatial Processing in WS.....	56
Language in HFA	59
Visuospatial Processing in HFA.....	61
Reading in WS and HFA.....	62
Reading in WS.....	63
Reading in HFA.....	65
Methods	68
Participants.....	68
Standardized Testing Battery	69
Experimental Reading Measure	69
Materials and Procedure	69
Experimental Conditions.....	71

Results	72
Standardized Testing.....	72
Experimental Reading Measure	72
Discussion	75
Appendix A:	81
References	83
Acknowledgements	92
Chapter 3: Relations Among Linguistic Subsystems in High Functioning Children with Autism and Typically Developing Children	93
Abstract	93
Introduction	93
Methods	103
Participants.....	103
Procedure and Measures	104
Results	108
Between Group Comparisons	108
Correlations Within Groups.....	110
Discussion	112
Conclusions	123
References	124
Acknowledgements	130
Chapter 4: A Multi-Level, Multimodal Language and Structural Imaging Investigation: Exploring the Brain-Behavior Relationship in Two Children with Perinatal Stroke	131
Abstract	131
Introduction	132
Methods	138
Participants.....	138
Case L Description: Left Hemisphere Lesion	139
Case R Description: Right Hemisphere Lesion.....	139
Multimodal Structural Magnetic Resonance Imaging	141
Image Acquisition	141
Image Preprocessing, Reconstruction, and Segmentation.....	142
Materials	144
Testing Battery	144

Phonological Processing Assessments.....	145
Speaking	145
Writing.....	146
Reading.....	146
Lexico-semantic Processing Assessments	146
Speaking	146
Writing.....	147
Reading.....	147
Syntactic Processing Assessments	148
Speaking	148
Writing.....	149
Reading.....	149
Narrative Processing Assessments.....	149
Speaking	149
Writing.....	151
Reading.....	151
Behavioral Analysis	152
Brain Analysis.....	152
Results and Discussion	153
Conclusions	166
Appendix A	167
References	169
Acknowledgements	176
Chapter 5: General Discussion and Conclusions	177
Appendix A	186

LIST OF FIGURES

Figure 1.1: Displays the four experimental conditions. Each box represents the screen for one trial. Condition A contains an orthographic match and an unrelated foil, condition B contains an orthographic match and an orthographic neighbor as a foil, condition C contains a semantic match, and an unrelated foil, and condition D contains a semantic match and an orthographic neighbor as a foil.....	20
Figure 1.2: Displays efficiency scores (reaction time/accuracy) by grade for all conditions. Results are displayed with standard error bars.....	26
Figure 1.3: Displays the mean reaction time to Orthographic minus Baseline (hatched) and Semantic minus Baseline (solid) for correct trials only in Experiment 1. Results are displayed with standard error bars.	28
Figure 1.4: Compares the mean Reaction Times to correct trials only in the Orthographic condition between Experiment 1 (stimuli presented in the English orthography) and Experiment 2 (stimuli presented in a false font). Results are displayed for the first through fourth grade with standard error bars.	36
Figure 2.1: Displays the four experimental conditions. Each box represents the screen for one trial. Condition A contains an orthographic match and an unrelated foil, condition B contains an orthographic match and an orthographic neighbor as a foil, condition C contains a semantic match, and an unrelated foil, and condition D contains a semantic match and an orthographic neighbor as a foil.....	71
Figure 2.2: Accuracy results from all four conditions. Results displayed with standard error bars (TD n = 27, HFA n = 19, WS n = 10).	74
Figure 3.1: Displays the raw scores in the spoken (left) and written (right) modalities for (from bottom to top): phonological, lexico-semantic, and syntactic processing. Narrative composition was assessed in the spoken domain only. Within each plot, the TD group is on the left and the HFA group is on the right. Since raw scores are reported, the y-axis represents the scale for each respective measure, and thus cannot be compared between measures. Asterisks mark significant between-groups comparisons (at the $p < .05$ level).	109
Figure 3.2: Displays the proportion of morphological errors, complex syntax, and evaluation out of the total number of propositions in the child's narrative. Scores are expressed as means with standard error bars for the TD (white bars) and HFA (gray bars) groups.	110
Figure 3.3: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for TD (top) and HFA (bottom). The width of the line represents the correlation value, and the grey-scale represents the p -value.	111
Figure 4.1: Shows T1-weighted, inversion recovery spoiled gradient echo (IR-SPGR) MRI scans in the axial (top left) coronal (top center) and sagittal (top right) planes, and the three-dimensional cortical surface reconstruction of the right (bottom left) and left (bottom right) hemispheres for Case L. Case L presents with a left temporal-parietal lesion (9;0 at time of scan).	140

Figure 4.2: shows T1-weighted, inversion recovery spoiled gradient echo (IR-SPGR) MRI scans in the axial (top left) coronal (top center) and sagittal (top right) planes, and the three-dimensional cortical surface reconstruction of the right (bottom left) and left (bottom right) hemispheres for Case R. Case R presents with a right frontal porencephalic lesion (10;11 at time of scan).	141
Figure 4.3: Displays the <i>z</i> -scores (relative to the 15 typically developing children) for Case L (left) and Case R (right) on our index metric of linguistic processing at increasing levels of complexity (phonological, lexico-semantic, syntactic, and narrative) across speaking (black bar), writing (white bar), and reading (gray bar).	155
Figure 4.4: Displays the <i>z</i> -scores (relative to the 15 typically developing children) for Case L (left) and Case R (right) on Spoken (black bar) and Written (white bar) Narrative components.	156
Figure 4.5: Displays <i>z</i> -scores of the composite index (mean FA, MD, and T2N) of the white matter fiber tracts for the two children with perinatal stroke relative to the TD group from superior (top), left hemisphere sagittal (middle), and right hemisphere sagittal (bottom) views of Case L (left) and Case R (right). Warmer colors represent negative <i>z</i> -score values and cooler colors represent positive <i>z</i> -score values.	161
Figure 5.1: Displays the performance (raw scores) on phonological, lexical, and sentence-level assessments in the spoken (left) and written (right) modalities, and on spoken narrative composition (top). Scores are presented from Typically developing children (TD), children with Williams syndrome (WS), High Functioning children with Autism (HFA), children with Perinatal stroke as a group (PS), children with Right Hemisphere lesions (RHL) and children with Left Hemisphere Lesions (LHL).	186
Figure 5.2: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for WS. The width of the line represents the correlation value, and the grey-scale represents the <i>p</i> -value.	187
Figure 5.3: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for the Perinatal stroke group combined. The width of the line represents the correlation value, and the grey-scale represents the <i>p</i> -value.	188
Figure 5.4: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for children with Right Hemisphere lesions only. The width of the line represents the correlation value, and the grey-scale represents the <i>p</i> -value.	189
Figure 5.5: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for children with Left Hemisphere lesions only. The width of the line represents the correlation value, and the grey-scale represents the <i>p</i> -value.	190

LIST OF TABLES

Table 1.1: Example of Stimuli Counterbalancing Across Items and Participants	21
Table 1.2: Experiment 1 Accuracy Results.....	24
Table 1.3: Experiment 1 Mean Reaction Time Results (in seconds) to Correct Trials Only	25
Table 1.4: Examples of Stimuli for Experiment 2	32
Table 1.5: Experiment 2 Accuracy Results.....	34
Table 1.6: Experiment 2 Efficiency Score Results (Reaction Time/Accuracy)	34
Table 1.7: Orthographic Facilitation for Real Words (Experiment 1) Relative to False Fonts (Experiment 2), Correct Trials Only	35
Table 1.8: All Stimuli	43
Table 2.1: Standardized Testing Results.....	81
Table 3.1: Age and Standardized Testing Results for TD and HFA ¹	104
Table 3.2: Examples and Coding System for the Personal Narrative Measure	107
Table 4.1: Assessment Information for each Evaluated Level of Linguistic Processing	145
Table 4.2: Transcribed Excerpts from Spoken and Written Narratives	156
Table 4.3: Mean and Standard Deviations from the Typically Developing Group, as well as z-scores from Case 1 and Case 2 for each Individual Behavioral Measure	167

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- Polse, L., Erhart, M., Ignacio, M., Reilly, J., Trauner, D., Dale, A., Halgren, E., Brown, T. (2012). Shared and distinct language performance in two children with non-overlapping early focal brain lesions. *Procedia: Social and Behavioral Sciences*.
- Polse, L. & Reilly, J. (2012). Orthographic and semantic processing in young readers. *Journal of Research in Reading*. doi:10.1111/j.1467-9817.2012.01544.x
- Boudewyn, M. Gordon, P. Long, D. Polse, L. & Swaab, T. (2012). Does discourse congruence influence spoken language comprehension before lexical association? Evidence from event-related potentials. *Language and Cognitive Processes*, 27(5), 698-733. doi:10.1080/01690965.2011.577980.

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- Reilly, J. & Polse, L. Spoken and Written Narratives in Children with Language Impairment and Children with High Functioning Autism: *Invited Speakers at the Narrative, Intervention, and Literacy conference* (September 2012), Paris, France
- Polse, L., Reilly, J., & Bellugi, U. Orthographic and semantic processing in autism and Williams syndrome. *Poster for American Speech-Language Hearing Association Annual Convention* (November 2011), San Diego, CA.
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- Kacinik, N.A., Polse, L.R., Shatnawi, S., Swaab, T.Y., Baynes, K. Left and right hemisphere contributions to the comprehension of figurative and literal language. *Poster presented at the Cognitive Neuroscience Society meeting* (2006), San Francisco, CA.

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ABSTRACT OF THE DISSERTATION

On the Relations among Linguistic Subsystems in Typically Developing
Children and Children with Neurodevelopmental Disorders

by

Lara Rosalin Polse

Doctor of Philosophy in Language and Communicative Disorders

University of California, San Diego, 2013
San Diego State University, 2013

Judy Reilly, Chair

Language is a complex multifaceted system, and as we use spoken and written language we simultaneously recruit an array of interrelated linguistic subsystems. While these subsystems have been studied extensively during language acquisition, we know little about the organization and relations among these components in the school-age years. In the four investigations of this dissertation, I use classically defined components of language (phonological, lexico-semantic, and syntactic) as well as components of reading (orthographic and semantic) as a tool to explore the relations amongst elements that comprise the language system in school aged typically developing children (TD) and children with neurodevelopmental disorders (aged 7-12). Investigating the composition of the language system in children with neurodevelopmental disorders that affect language will not only help to create more targeted interventions for these

children, but will also provide a unique window through which to better understand the underlying structure and organization of language in TD children.

Chapter 1 investigates the development of and relation between orthographic (a visuospatial-based skill) and semantic (a language-based skill) components of single word reading in typically developing children in the first through fourth grade. Chapter 2 explores these processes in High Functioning children with Autism (HFA) and children with Williams syndrome (WS); children with HFA and WS show opposite strengths and weaknesses in visuospatial and language cognitive domains, and neither population develops reading typically. Chapter 3 extends these results to include spoken and written language at increasing levels of linguistic complexity, and explores how these components relate to one another in HFA and TD, and how they are related to the ability to organize and produce a spoken personal narrative. Chapter 4 considers the underlying neural correlates of linguistic components in a case study format with two children with unilateral Perinatal stroke (one right, one left hemisphere) using multimodal structural imaging techniques alongside a detailed analysis of language performance. Together, results from these investigations suggest that assessing the associations among linguistic subsystems provides valuable information, which will add to our understanding of the mechanisms underlying language impairment in children with neurodevelopmental disorders.

INTRODUCTION

Much like riding a bicycle, using language involves simultaneous control over and operation of multiple systems. To reach your destination successfully on a two-wheeled vehicle, one must organize the pedaling, balance, steering, shifting, and braking, all of which involve motor and cognitive systems such as attention, vision, motor planning, and others. To communicate successfully through spoken or written language, one must organize the phonological or orthographic, semantic, and syntactic subsystems, all of which also involve underlying cognitive and motor systems such as attention, auditory (or visual) memory, motor planning, etc. Unlike a bicycle, however, which can take you around town, or through beautiful Burgundy countryside, spoken and written language can transport you through time and space, and to faraway and imaginary places such as Alice's Wonderland. How do children organize and recruit these linguistic subsystems into such a powerful tool—language—that has the remarkable capacity to evoke images and ideas such as an anthropomorphic egg, which has never been seen before? What are the relations among these components of language (e.g. phonology, lexico-semantics, syntax) during development? In the four investigations of this dissertation I will consider the development of and relations among linguistic subsystems in the context of spoken and written language in school-aged typically developing children and children with neurodevelopmental disorders.

Studying children with neurodevelopmental disorders that impact language not only provides a foundation of knowledge for basing clinical interventions, but also offers a unique window through which we can better understand typical development. Through identification of language components that are not functioning normally, we can gain insight as to how these processes come together to support typical language development. Additionally, while many of these children come into the world at a biological disadvantage (perinatal lesions, genetic

abnormalities), often they show remarkable neural and behavioral adaptation, leading to functional linguistic processing, which attests to the brain's plasticity to utilize alternate routes to support language. However, *language* is a broad, dynamic, complex system to study. Furthermore, every individual uses language differently—chooses different words to represent thoughts, and different sentence structures to emphasize (or de-emphasize) parts of a story—so how can we study this unwieldy system? One method that has been used for centuries is to cut language up into component parts. Throughout these pages, I will refer to subsystems, components, and levels of language (e.g. phonological, lexico-semantic, syntactic, narrative). Although I acknowledge that apportioning language in this manner is somewhat arbitrary, it provides a useful tool for discussing language at multiple degrees of complexity, and more importantly, for basing hypotheses regarding the interrelations between linguistic subsystems, which combine to create the complex, dynamic system of language. As Thelen and Smith (1996) note in their Dynamic Systems approach, “If real language understanding and real visual experience are a mix of informationally encapsulated devices and general purpose mechanisms, we must do more than draw sharp lines between components. We must explain how they mix together to make a whole” (pg. 35). With this goal, in the four investigations of this dissertation, I use classically defined linguistic components of language (phonological, lexico-semantic, and syntactic) as well as components of reading (orthographic and semantic) as a tool to explore the relations amongst the components that comprise the language system in typically developing children and children with neurodevelopmental disorders.

Chapter 1 considers how pre-linguistic, visuospatial processing skills and semantic language skills are integrated during reading acquisition and development. Reading necessarily requires orthographic processing, a largely visuospatial task that allows a child to recognize the orthographic shape of the word; and semantic processes, which allow access to the word's meaning. Without either of these components, reading would be impossible. *How do these*

components develop? What is their relationship with one another during typical reading acquisition and development? I address these questions in a cross-sectional investigation with children in the first through fourth grade using a new single word reading measure that contrasts efficiency of orthographic and semantic processing. Since the ability to read is arguably the single most important skill a child learns in elementary school, understanding how children acquire the components involved in this process is profoundly important to society. From a practical standpoint, the ability to read is fundamental to a fulfilling life on many levels: It is required to maintain most jobs, it is necessary to act as an informed member of society, and it is essential for communication with others. From a more poetic perspective, “Once you learn to read you will be forever free” (Frederick Douglass). Understanding the developmental trajectory and interactions between orthographic and semantic processing during reading acquisition and development will not only inform reading curricula for typically developing children, but will also help to create more targeted interventions for children when reading does not develop normally or at all.

Chapter 2 extends the results from Chapter 1 to explore these processes in two populations of children who do not develop reading in the typical manner. High Functioning children with Autism and children with Williams syndrome show opposite strengths and weaknesses in their visuospatial and language processing profiles. High Functioning children with Autism demonstrate typical (or even enhanced) visuospatial processing. They perform extremely well on visual search paradigms, which require identification of a specific item or pattern amid an array of distractors, as well as on block design tasks, which require re-creating a design with a number of individual blocks. However, children with autism show relatively poor language skills, most notably in social and pragmatic language, but deficits also emerge in semantic processing. Children with Williams syndrome show a complementary cognitive profile, with relative strength in the language domain and relative weakness in visuospatial processing. Initially, the complex syntax, unusual and sophisticated vocabulary, and apparent ease with which

individuals with Williams syndrome use language led researchers to declare that language was “spared” from general cognitive impairment. Twenty years of research has since revealed that language in Williams syndrome is much more complex than originally described, and is laden with many errors and atypical aspects. Nevertheless, language does indeed seem to be a strength relative to other cognitive domains. For example, children and adults with Williams syndrome often know more words than their intellectual age would predict, and they show evidence of rich semantic representations of lexical items. Since reading necessarily involves orthographic processing (a visuospatial skill) and semantic processing (a language based skill), *How are deficits observed in the visuospatial and/or language profiles of children with neurodevelopmental disorders reflected in reading performance in the school-age years? How does this reading profile compare to typical reading development?* As both populations of children show reading impairments, a better understanding of the development of reading components in these populations will help to create more targeted reading interventions for High Functioning children with Autism and children with Williams syndrome. Although Williams syndrome is a very rare genetic disorder, Autism is ubiquitous, and each year the estimated rate of affected children increases. The most current estimate from the Centers for Disease Control and Prevention is 1 in 88 children (Baio, 2012). Twenty to thirty percent of these children will be categorized as High Functioning, and will have the intellectual capacity to be contributing members of a community. Therefore, research-based clinical reading interventions, especially for these High Functioning children with Autism, are critically important not only for these individuals, but for society.

In Chapter 3, I extend these results to investigate language in High Functioning children with Autism and typically developing children at increasing levels of complexity: at phonological, lexico-semantic, and syntactic levels in both speaking and reading. I will consider how these subsystems of language relate to one another, as well as how they relate to one’s ability

to organize and produce a spoken narrative. There are different frameworks for understanding the language deficits observed in High Functioning children with Autism. Some are rooted in social aspects of language such as perspective taking and empathizing; others are cognitive processing accounts, which propose a more systemic impairment that impacts language. One such processing account suggests that children with autism have difficulty integrating components into a cohesive whole. That is, while the parts themselves are unimpaired, putting them together causes disruption. According to this view, we would expect language at the more isolated levels (perhaps phonology and lexico-semantics) to be better than more integrative components (for example syntactic and narrative). However, investigating language at multiple isolated levels of complexity does not necessarily tell us about the structure of the linguistic system as a whole.

How do language components relate to one another in typically developing children? What can investigating the relations between linguistic subsystems tell us about language in High Functioning children with Autism? David Crystal (1987) proposes that our interest in levels and components of language may have caused us to ignore what might be the more central issue in treating language impairments: the relations between the levels. In this investigation we look at how language subsystems relate and function independently as well as within a narrative context. A better understanding of the composition of the language system in High Functioning children with Autism will have clinical implications both in terms of diagnostic procedures and language interventions.

Chapter 4 considers how components of language are related to the structural characteristics of the brain. I use a case-study approach as an initial step to identify behavioral language measures and structural neuroimaging measures which will provide a foundation for a larger group investigation that will consider relations between brain structure and language behavior at the same detailed level of analysis implemented here. With this aim, in Chapter 4 I assess the subsystems of language discussed in previous chapters in two children with unilateral

perinatal focal lesions (one in the right hemisphere and one in the left hemisphere). While this investigation is preliminary, there are insights that can be gleaned using a case study approach which are inaccessible in a group study format (e.g. Caramazza & McCloskey, 1988).

Historically, case studies of individuals with focal brain injuries have identified dissociations between an impaired behavior and a lesioned brain region as a means to isolate regions of the brain that are required for a specific cognitive function. On the other hand, patient studies can also determine what the brain *can* accomplish without a specific region. In children who experience strokes very early in development (in the pre or perinatal period), the impact of the lesion on language and cognition is attenuated relative to adults (Bates et al., 2001). This difference is especially apparent in children and adults with left hemisphere lesions. While a late-acquired left hemisphere lesion can be devastating to the language system in adulthood, children with perinatal left hemisphere strokes often show typical conversational language, and indeed very few cognitive or linguistic differences have been noted between children with left and children with right perinatal lesions. Thus, this population provides a unique opportunity to study the plasticity of the human brain, and the *developmental process* of language acquisition.

Importantly, however, since no two lesions are alike, group studies (while they have the advantage of generalizing results to a broader population), cannot take into account individual variance in lesion characteristics (size, extent, type of tissue damage, state of the non-lesioned hemisphere). Using a case study approach, however, it is possible to complete a detailed linguistic profile for each individual, which includes multiple language components, contexts, and modalities of language, as well as a detailed characterization of the structure of the individual's brain using multimodal structural neuroimaging methods. Unlike many investigations of children with perinatal stroke, here I will provide a characterization not only of the lesion and the lesioned hemisphere, but will also consider the state of the non-lesioned hemisphere. In sum, the goals of this final investigation are three-fold: First, to consider regions of the brain that are *necessary* for

a specific linguistic process. Second, to investigate, in detail, neuroplasticity for language, and the components that comprise language. Third, to examine the characteristics of the *whole brain* alongside a detailed linguistic profile for both children with perinatal stroke, as a means to explore the relation between—and base hypothesis about—brain structure and language behavior at a detailed level of analysis.

Together, the investigations that comprise this dissertation will explore how components of language develop, relate, and function in typically developing children and in children with neurodevelopmental disorders (Williams syndrome, High Functioning Autism, and Perinatal Stroke). This is a vast question, and, somewhat ironically, one that is itself comprised of multiple components and subquestions which must be integrated to gain a comprehensive understanding of this broad topic. While it is my hope that this dissertation will add a component piece to this immense body of research, like Lewis Carroll's *Through the Looking Glass*, the investigations presented here will evoke many questions. In the concluding chapter of this dissertation I will explore some of these outstanding questions and avenues for future research.

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CHAPTER 1: ORTHOGRAPHIC AND SEMANTIC PROCESSING IN YOUNG READERS

ABSTRACT

This investigation examined orthographic and semantic processing during reading acquisition. Children in first through fourth grade were presented with a target word and two response alternatives, and were asked to identify the semantic match. Words were presented in four conditions: an exact match and unrelated foil (STONE – STONE – EARS), an exact match and an orthographic neighbor foil (STONE – STONE – STOVE), a synonym match and an unrelated foil (STONE – ROCK – EARS), and a synonym match and an orthographic neighbor foil (STONE – ROCK – STOVE). Accuracy and reaction time results suggest that orthographic and semantic processing follow a two-step acquisition pattern. First, the orthographic component of reading develops quickly, however, forming strong conceptual links from orthographic to semantic representations follows a protracted trajectory, which matures between the third and fourth grade. These results are consistent with research that suggests younger children rely on more concrete, perceptual systems and then transition to more flexible, abstract cognition.

INTRODUCTION

Learning to read is a difficult task, and arguably the most important in a child's elementary school years. Children must learn the alphabetic symbols and grapheme-phoneme correspondences to decode a word, and also must assign meaning to each deciphered word. While deciphering the orthographic symbols can be completed using lower-level visuo-perceptual systems, forming abstract, semantic representations from a written word requires higher-level, more conceptual processing. This suggests the involvement of multiple systems: those required for the perceptual visuo-spatial orthographic processing, and those required for more abstracted semantic processing of a word. The present experiment investigates the developmental profile of orthographic and semantic processing in young readers of English from the first to fourth grade.

Component models of reading propose that skilled reading requires the mastery and integration of distinct cognitive processes (e.g. Gough & Tunmer, 1986; Kendeou, van den Broek, White, & Lynch, 2009; Lonigan, Burgess, & Anthony, 2000; Whitehurst & Lonigan, 1998). Furthermore, many such models suggest that reading is the product of successful integration of multiple components comprised of different *levels* of cognitive processing. For example, skilled reading can be conceptualized as the product of successful integration of a lower-level perceptual component and a higher-level conceptual component. While nearly all descriptions of reading acquisition are in accord that a conceptual or semantic processing system is required to extract meaning from text, they differ as to the lower-level, more perceptual component. The vast majority of recent research focuses on how *phonological* processes interact with comprehension, and thus the importance of phonological awareness and decoding skills to successful reading acquisition has been well described (e.g. Anthony, Williams, McDonald, & Francis, 2007; Frost, Madsbjerg, Niersøe, Olofsson, & Sørensen, 2005; Goswami & Bryant, 1990; Hulme, Goetz, Gooch, Adams, & Snowling, 2007; Muter, Hulme, Snowling, & Taylor, 1998; Muter, Hulme, Snowling, & Stevenson, 2004; Perfetti & Zhang, 1995; Wagner & Torgesen, 1987). Comparatively little is known, however, about the contribution of orthographic processing to reading comprehension during literacy acquisition and development (Nation, 2009).

In adult reading, the path from orthography to meaning has been, and continues to be, strongly debated (see Harm & Seidenberg, 2004). Both rule-based and connectionist models seek to explain how skilled readers use orthographic information to extract meaning from a written word. The Dual Coding Route theory (Coltheart, Rastle, Perry, Langdon, & Ziegler, 2001) is a rule-based computational model of reading that provides a framework for understanding how phonological and orthographic processing interact with semantic processing. The Dual Coding Route proposes that there are two separate routes that can be used to comprehend a written word. The decision of which route to use is based on properties of the word itself. When the word is

familiar and frequent, or contains irregular mappings from orthography to phonology, readers utilize a “direct route,” and access a semantic representation directly from an orthographic representation. When words are less familiar or more difficult, readers utilize a phonologically mediated route, in which they activate first the phonological information and subsequently access a semantic representation from the phonology (orthography to phonology to semantics). The phonological route can be compared to “sounding out” a word, as comprehension is thought to come from the sound, rather than directly from the written (orthographic) representation. Contrasting with this view, are connectionist or cascaded models (e.g. Plaut, McClelland, Seidenberg, & Patterson, 1996), which suggest that comprehension of a written word is accomplished by an increasing sensitivity to the statistical structure that exists between phonological, orthographic, and semantic representations. One such model, the Triangle Model, is introduced below.

The Triangle model (Plaut et al., 1996; Harm & Seidenberg, 2004) is a connectionist model, which suggests that there is both a pathway from orthography to phonology *and* a pathway from semantics back to orthography and phonology. It is assumed that these pathways operate in tandem, and that comprehension is achieved via graded representations of the pathways working simultaneously. The weights attached to each of the pathways, however, may shift as a function of the word context. As such, the Triangle Model indicates that semantics is involved in the orthographic decoding of *all* items, not only the decoding of high frequency words, or words with inconsistent orthographic to phonological mappings. While these two approaches to understanding reading comprehension have been well studied in adult reading (for a review of orthographic processing in adults, see Grainger (2008)), we know very little about the processes involved in creating efficient form to meaning mappings during reading acquisition (Castles & Nation, 2008; Nation, 2009).

One possible explanation for the scarcity of research into orthographic processing during reading development, is that orthographic and phonological processes are closely related, making it difficult to study the influence of orthographic and phonological processes independently (Cunningham, Perry, & Stanovich, 2001). This is especially true during reading acquisition, when children learn to link a visual code with a phonological representation. According to Jorm and Share (1983), successful phonological decoding of words is coupled with the full orthographic representation of the word, resulting in “positive learning trials,” in which each successful decoding of a new word provides an opportunity to acquire word-specific orthographic information about that word. In this view, phonological decoding is constantly reinforcing the orthographic representation, leading to a merged phonological and orthographic representation of each known word. This compound phonological-orthographic representation likely contributes to efficient word recognition (Ehri & Wilce, 1980; Perfetti, 1992), and makes it difficult to disentangle the independent contributions of orthographic and phonological components to the development of skilled reading.

Although it is clear that orthographic and phonological processes are closely related during the reading acquisition time window, orthographic processes are not entirely dependent on phonological processes (Burt, 2006; Reitsma, 1983; Stanovich & West, 1989), and orthographic and phonological processes have been shown to exert unequal influence on the reading process (Bekebrede, Leij, Plakas, Share, & Morfidi, 2010). Indeed recent research suggests that differences in orthographic processing account for unique variance in reading proficiency, which cannot be explained by differences in phonological processing alone (e.g. Badian, 2001; Castles, Holmes, Neath, & Kinoshita, 2003; Treiman & Cassar, 1997). Such research suggests that understanding the processes involved in efficient orthographic processing may contribute to a better understanding of reading development as a whole (Aghababian & Nazir, 2000). In one such experiment, Pratarelli Perry, and Galloway (1994) investigated the visual encoding of words in

children compared to adults in order to determine whether or not children were less skilled in automatic lexical access. Nine and ten-year-old children made lexical decisions on content and function words following a brief (50ms) prime word that was either graphemically related (e.g. dog - DOG) or unrelated (dog - BRICK). They found that while adults were faster in all conditions, the facilitation priming effect was the same in adults and children, suggesting that graphemic visual encoding was similar between groups. While these results shed light on the visual encoding of graphemic information, they do not necessarily extend to orthographic similarity. Furthermore, the children in the young reader group were already nine and ten years old, and arguably not beginning readers.

A study by Nation and Cocksey (2009) extends these results to investigate *orthographic* encoding in a group of younger children. In this experiment, young children were asked to make categorical judgments about visually presented words, some of which contained an embedded word that was related to the category (e.g. *hip* is embedded in *ship* and is a category member of *body parts*). Children in this investigation were slower to reject *ship* as a member of the category *body part* in comparison to the category *animal* because *ship* contains an embedded category member. Furthermore, it was discovered that this inhibition effect remained even when the pronunciation of the embedded word differed from the carrier word (e.g. *crow* does not share pronunciation with its carrier *crown*, though orthographically it is an embedded category member of *animal*). These results strongly suggest that even children as young as seven years of age activate meaning directly from the orthography of the word.

Further evidence for the early role orthographic processing plays in reading acquisition comes from a study by Zecker (1991). In this study, children ages seven to eleven participated in an auditory rhyme detection task. Within the rhyme trials, half of the targets were orthographically similar to the cue (e.g. BUM – GUM) and half were orthographically dissimilar (e.g. THUMB – GUM). Results demonstrated that typically developing children, as young as

seven years of age, showed a facilitation effect for detecting the auditory rhyme when the orthography was similar, and that the orthographic facilitation became more robust in older age-groups. These results also suggest that children automatically activate the orthographic representation of an auditory word quite early, but that the orthographic representation continues to become stronger with development.

There also exists some evidence that purely orthographic representations may precede phonological representations in the early stages of reading acquisition (Rayner, 1988). Such evidence has its foundations in the finding that orthographic processing is more strongly correlated with earlier-acquired, visuo-perceptual performance than later-acquired, unobservable, abstract processing (such as phonological or semantic processing) (Boets, Wouters, van Wieringen, De Smedt, & Ghesquière, 2008). If the cognitive skills that support orthographic processing develop earlier than those which support semantic processing, it is reasonable to expect that the acquisition and development of orthographic and semantic components of reading will follow different developmental trajectories: Initially, orthographic processing (supported by lower-level visuo-spatial skills) will be more robust, and later in reading acquisition, both orthographic and semantic components will become comparably proficient. This integration of reading components results in rapid and efficient linking between orthography and meaning, which is one of the hallmarks of skilled reading (Castles & Nation, 2008).

Many Stage Models of Reading describe the process of reading acquisition as just such a transition: from a rigid reliance on concrete, observable aspects of a visual word, to more flexible, abstract representations (Marsh, Friedman, Welch, & Desberg, 1981; Mason, 1980; and see Rayner & Pollatsek, 1989 for a review). In early stages of reading development, for example, features of the orthography tend to be more salient to young readers than what the symbols represent (i.e. a phonological or semantic representation). In these early stages, children may use visual aspects of the word to remember its meaning (e.g. *look* has two eyes) (Gough, Juel, &

Griffith, 1992), and rely more heavily on the boundaries of the word (first and last letter) than the medial portions. Readers in these early stages also may confuse words that have the same initial and final letter, even when the words differ in the middle (e.g. *spoon* and *skin*) (Ehri, 2005). From this standpoint, reading acquisition can be viewed as a period when children learn to integrate the concrete observable components with more abstract components, resulting in efficient form to meaning mappings to support reading.

Additional evidence that beginning readers are more reliant on perceptual aspects comes from a study by Rayner (1988) in which children of different reading levels were asked to decide which response alternative was most like a target (e.g. *cabin*). They found that pre- and beginning readers responded most often on the basis of word shape and initial letter (e.g. responding that *cadin* is more similar to *cabin* than *kabin*), indicating that for beginning readers *most like* was synonymous with the perceptual interpretation *looks most like*, rather than a more abstract interpretation such as *sounds most like*. Following this task, children were asked which response alternative *sounded most like* the target, thereby drawing children's attention to the more abstract, phonological representation. Results demonstrated that the youngest readers continued to identify the response alternative that *looked* most like the target. Intermediate readers (eight-year-old children with three years of reading experience) initially tended to interpret *most like* to mean *looks most like*, but when asked to identify the word that *sounded most like* the target word, eight year olds changed their answer to the closest homophone. This suggests that while the concrete orthographic cues were still the most salient for eight-year-olds, they had the cognitive flexibility to recognize a different, more abstract dimension when cued to do so. Ten-year-olds (with about five years of reading experience), tended to choose the response alternative that *sounded most like* the target without direct instructions, suggesting that by ten years of age, *most like* is synonymous with a more abstract dimension *sounds most like*. Rayner's (1988) investigation provides evidence that for early readers, the observable features of the orthography are the most salient

aspect of a written word until about ten years of age, when the non-observable, phonological representation becomes the most salient.

Some evidence suggests that establishing good orthographic processing skills early in reading development is critical for later automatic word recognition and reading comprehension (Ehri, 2005). However, while measures of orthographic processing in preschool were predictive of reading comprehension in seventh grade, they were not predictive of reading performance in the first and third grades (Badian, 2001). It thus appears that establishing a strong foundation in orthographic processing is critical to later reading comprehension, but that children do not rely heavily on orthographic processing in the middle elementary school years, likely due to the development of phonological processing. A comprehensive model of reading development would therefore include the development and interaction of orthographic, phonological, and semantic processes. This is outside the scope of this paper, however, which concentrates on orthographic and semantic processes, and the connections that are formed between these two components during reading acquisition.

Some models propose a child's increase in language proficiency and vocabulary is the primary impetus for integrating orthography and semantic processes. The *lexical tuning hypothesis* (Castles, Davis, Cavalot, & Forster, 2007; Castles, Davis, & Letcher, 1999), for example, suggests that an increase in vocabulary acts as a catalyst in forming links between orthographic and semantic representations. Specifically, the lexical tuning hypothesis posits that orthographic processing is continually "tuned" as children acquire larger vocabularies. Early in reading acquisition, the orthographic processing system is coarsely tuned, and thus accepts similar (but not identical) inputs as candidates for a target word. Young readers, for example, would be likely to accept an orthographic neighbor, such as *bib* as identical to the word *bid*. As children acquire more words and become more experienced readers, however, this loose orthographic criterion becomes increasingly stringent, and children demonstrate more finely

tuned orthographic processing. Thus, the lexical tuning hypothesis emphasizes the interconnected nature of orthographic and semantic processing during the reading acquisition time window, and highlights the necessity for further investigation of the developmental profile of both components.

Castles and colleagues (2007) found evidence for the lexical tuning hypothesis using a masked priming longitudinal design. In this investigation, adults and children (at grade three and then again at grade five) made lexical decisions about words following a masked prime that was either a substitution of the target word (e.g. rlay : PLAY), a transposition of the target word (e.g. lpay : PLAY), or orthographically unrelated to the target word (e.g. meit : PLAY). This investigation found that while adults showed little or no facilitation from non-word primes that differed from the target by one letter, the third grade children demonstrated large facilitation effects. At fifth grade, the facilitation effects were diminished. The authors interpret these results to suggest that word recognition processes change from broadly to finely tuned. Since children demonstrated facilitation from a non-word prime that differed by one letter from a target (and adults did not) in the younger grades, this suggests that the orthographic word recognition mechanism accepted the orthographic neighbor as the target, thus priming the lexical decision. In adulthood, the non-word orthographic neighbor did not prime the target word and resulted in little or no facilitation of the meaning, presumably because more real word orthographic neighbors exist in the lexicon (e.g. bid, bib, bit, big, bin).

Currently, there are few studies that address the development of the orthographic and semantic processing systems in relation to one another in the same task within the reading acquisition window (studies that have addressed this question include Castles et al., 2007; Nation & Cocksey, 2009; Pratarelli et al., 1994; Zecker, 1991, summarized above). It is not clear, therefore, whether orthographic and semantic processes mature concurrently during reading acquisition, or whether these processes follow different developmental trajectories, leading to

strengths and weaknesses in orthographic and semantic processing at different stages during reading acquisition.

The current experiment differs from other investigations of how and when orthography influences reading comprehension in that it uses a synonym detection task to probe comprehension of meaning (rather than lexical decision (Castles et al., 2007), category judgment (Nation & Cocksey, 2009), or rhyme detection (Zecker, 1991)), and directly pits similar semantics (synonyms) with similar orthographic forms (orthographic neighbors) within a single experiment, and within subjects. Using this novel experimental design, children's accuracy and reaction times to identify semantically and orthographically similar pairs are compared. This experiment allows a glimpse into the development of the orthographic and semantic processing systems, and provides insight into how and when these two systems function together to support reading. This investigation explores the possibility that there exists a transition from an orthography first strategy (such as would be expected by the phonologically mediated pathway of the Dual Coding Route (Coltheart et al., 2001) and linear models of reading (e.g. Morton, 1969)), to a more semantically influenced comprehension strategy, where the meaning of the word influences orthographic and phonological processes (as would be expected by more cascaded/connectionist model such as the Triangle Model) (Harm & Seidenberg, 2004; Plaut et al., 1996).

Children in the first through fourth grade were asked to make semantic similarity judgments regarding a target word (e.g. STONE) when there was either an orthographic match with an unrelated foil (e.g. STONE and EARS), an orthographic match with an orthographic neighbor as a foil (e.g. STONE and STOVE), a semantic match with an unrelated foil (e.g. ROCK and EARS), or a semantic match with an orthographic neighbor as a foil (e.g. ROCK and STOVE). It was predicted that accuracy and reaction time results would support the hypothesis that the orthographic code is established quite early, but due to weaker associative links joining

perceptual, orthographic representations with conceptual, semantic representations, the younger children in the reading acquisition time window will be less efficient to map this orthographic representation onto a semantic representation. This hypothesis would be supported if younger children demonstrate faster, more accurate processing of conditions requiring orthographic processing (Orthographic condition); slower, though accurate performance on conditions probing semantic processing (Semantic condition); and less accurate, but rapid performance on the condition directly comparing orthographic and semantic processing (Orthographic-semantic condition). Older readers, by contrast, are expected to demonstrate similar reaction time and accuracy to Orthographic and Semantic conditions and higher accuracy on the Orthographic-semantic condition. Such a pattern of results would suggest that children transition from more perceptually based (orthographic) to more abstract (semantic) representations of written words during reading acquisition.

EXPERIMENT 1

METHODS

Participants

Eighty typically developing children from San Diego public schools participated in Experiment 1: Twenty first graders (12 female, 8 male; M age = 6.96 years; SD = .29), 20 second graders (11 female, 9 male; M age = 8.10 years; SD = .28), 20 third graders (11 female, 9 male; M age = 8.99 years; SD = .39), and 20 fourth graders (15 female, 5 male; M age = 9.98 years; SD = .44). All children were participating in English curriculum at school and demonstrated conversational competence in English (via teacher report, as well as an informal conversational assessment administered by the experimenter). Two first grade participants were excluded from analyses based on an inability to read single words in English.

Materials

Twenty-six sets of word triads were designed to be appropriate for young elementary school children (see Appendix A). Each triad contained a “target” word, a “match” word (the correct choice for that particular trial), and a “foil” word (the incorrect choice for that particular trial). Word-pairs designated as “match” and “foil” were normed on 100 undergraduate students from San Diego State University, who rated the semantic similarity of word-pairs on a Likert scale (1 = extremely different meanings and 7 = extremely similar meanings). Only those pairs that were rated as an average of 5.5 and above ($M = 6.06$; $SD = .33$) were used as a “match,” and only those that were rated as an average of 1.5 and below ($M = 1.35$; $SD = .83$) were used as a “foil.”

Target, match, and foil stimuli were matched in word frequency based on the Hyperspace Analogue to Language (HAL) corpus norms (Lund & Burgess, 1996), which consists of approximately 131 million words gathered across 3,000 Usenet newsgroups during February 1995 (Balota et al., 2007). Since participants chose between the match and the foil in each trial, it was important that the match and foil not differ in terms of word frequency, as that would bias participants toward one response alternative over another. Paired t -tests reveal that there were no significant differences between the frequency of the match and foil, according to the HAL frequency corpus $t(25) = .326$, $p > .1$. Match, Foil, and Target words were all between 3 and 6 letters, and match and foil words were not significantly different in length (number of letters) $t(25) = 1.9$, $p > .05$.

All three words (“target,” “match,” and “foil”) were presented simultaneously on a touch screen monitor (Dell 15-inch Touch-screen Flat Panel). Words were presented in all capital letters (size 40 Arial font) and for salience were presented within white rectangles on a black screen. The “target” word was centered at the top of the screen, and the “match” and “foil” were

presented beneath the “target” at the left and right sides of the screen (see Figure 1.1 for a schematic of study design).

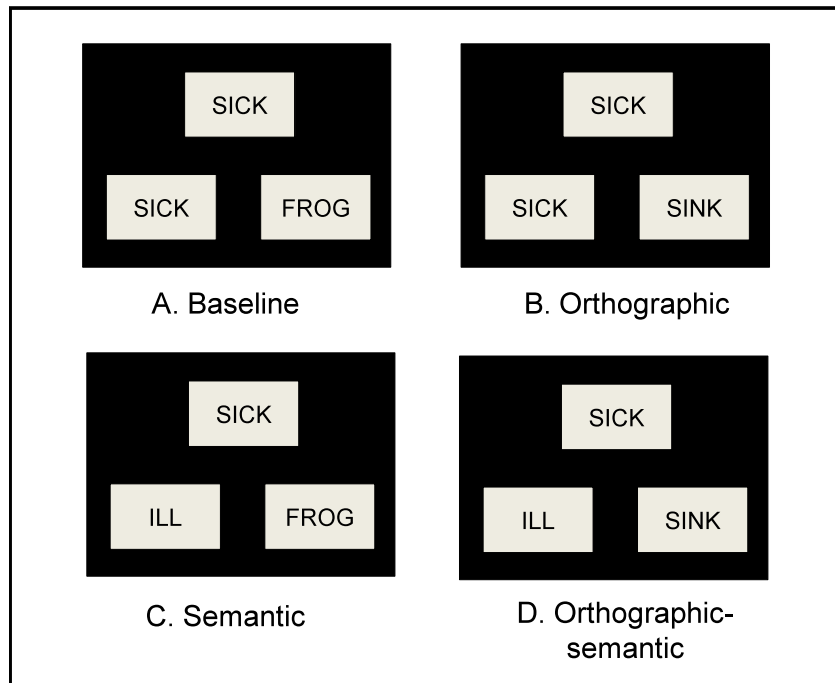


Figure 1.1: Displays the four experimental conditions. Each box represents the screen for one trial. Condition A contains an orthographic match and an unrelated foil, condition B contains an orthographic match and an orthographic neighbor as a foil, condition C contains a semantic match, and an unrelated foil, and condition D contains a semantic match and an orthographic neighbor as a foil.

The side of the “match” on the visual display was pseudorandomized such that each participant saw an equal number of “match” and “foil” words on the right and left sides of the screen, but the location of the “match” was not predictable. Each child responded to 26 trials that met norming criteria (participant average rating of 5.5 and above for synonyms and average of below 1.5 and below for foils). Four lists were created containing 6 trials of two conditions and 7 trials of the remaining two conditions, which then were counterbalanced across participants so that an equal number of each condition was presented across participants. Thus, each participant saw each stimulus word in only one of the four conditions, and participants did not see any words

repeated more than once to avoid repetition effects (for examples of stimuli and counterbalancing, see Table 1.1 and Appendix A, Table 1.8).

Table 1.1: Example of Stimuli Counterbalancing Across Items and Participants

		Student 1	Student 2	Student 3	Student 4
Baseline	Target	SICK	BOAT	SMALL	STONE
	Match	SICK	BOAT	SMALL	STONE
	Foil	FROG	GAME	APPLE	EARS
Orthographic	Target	BOAT	SICK	STONE	SMALL
	Match	BOAT	SICK	STONE	SMALL
	Foil	BOOT	SINK	STOVE	SMELL
Semantic	Target	SMALL	STONE	SICK	BOAT
	Match	LITTLE	ROCK	ILL	SHIP
	Foil	APPLE	EARS	FROG	GAME
Orthographic-semantic	Target	STONE	SMALL	BOAT	SICK
	Match	ROCK	LITTLE	SHIP	ILL
	Foil	STOVE	SMELL	BOOT	SINK

Procedure

Participants were tested individually in a quiet room at their school. In order to ensure that all participants within a grade cohort were at the same point in their academic curricula, all testing took place in the months of April and May (within the final months of the academic year). Children were seated in front of a touch screen monitor and were instructed to use one finger to touch the word that “means the same thing” as the “target” word. Children were informed that for this experiment, *means the same thing* could be either the exact same word (e.g. BOAT-BOAT), or a synonym of that word (e.g. BOAT-SHIP). All participants completed a 10-trial practice block with accuracy feedback to ensure understanding of the procedure. Participants were not given feedback during the experimental trials. Although accuracy was emphasized, children were asked to answer as quickly as possible. The stimuli remained on the screen until the child made his or her response by touching one of the response words. In order to decrease variability

associated with fatigue and distractions, the experiment was self-paced. After each response, an interim screen appeared, instructing the child to touch the screen when he/she was ready to proceed with the next trial. During the practice block, children were taught to rest between trials (when the picture was on the screen) rather than during experimental trials (when the words were on the screen).

Experimental Conditions

Four conditions were presented: Baseline, Orthographic, Semantic, and Orthographic-semantic (Refer to Appendix A for full list of stimuli).

In the Baseline condition, the “match” was the same word as the “target” (orthographically identical), and the “foil” was unrelated both orthographically and semantically (e.g. LADY (target); LADY (match); WATER (foil)). This condition required participants to execute a perceptual matching task, and it measured the time the child took to match the “word-shape” (see Rayner, 1988) and make a physical response (touching a word on the screen). Crucially, in the Baseline condition, no semantic processing or finely tuned orthographic processing was required.

In the Orthographic condition, the “match” was also orthographically identical to the “target” but in this condition the “foil” was an orthographic neighbor of the “target;” it differed from the target word by only one letter (e.g. LADY (target); LADY (match); LAZY (foil)). Thus, this condition required the child to execute a very finely tuned orthographic discrimination task (instead of simply matching word-shape) to determine that the “target” and the “foil” were indeed different words. However, this Orthographic condition did not demand semantic processing.

In the Semantic condition, the “match” was a synonym of the “target,” and the “foil” was unrelated both semantically and orthographically (e.g. LADY (target); WOMAN (match); WATER (foil)). In this condition the participant was required to process the meaning of the word, but was not presented with a perceptual, orthographic foil.

In the Orthographic-semantic condition, the “match” was a synonym of the “target” and the “foil” was an orthographic neighbor of the “target” (e.g. LADY (target); WOMAN (match); LAZY (foil)). In this condition, semantic processing and orthographic processing were in direct competition because it required participants both to perform a perceptual orthographic discrimination task to determine that the “target” and the “foil” were different words, and to access their semantic lexicon in order to determine that the “match” was semantically (though not perceptually) related to the “target.”

RESULTS

Data were analyzed using two 4 (Grade) x 4 (Condition) repeated measures ANOVAs, one with the proportion correct as the dependent variable and one using mean (untrimmed) reaction times normalized by accuracy (Efficiency scores) as the dependent variable. Ulrich and Miller (1994) argue that trimming extreme reaction times introduces considerable bias, as truncating data can eliminate data points that may be extreme, but are not spurious. In the present experiment, the data were collected by the first author, who ensured that long reaction times were indeed due to longer processing times, rather than equipment failure, interruption, or other spurious event unrelated to the experimental task. As such, similar to other investigations of reading development (e.g. Nation & Snowling, 1998, 1999), as well as recent developmental cognitive neuroscience experiments (e.g. Clarke, Taylor, & Tyler, 2011), the present investigation does not remove outliers. The variability introduced by including possible outliers (especially in the younger grades) will be reflected upon within the discussion section.

Accuracy Results

Accuracy results yielded a significant main effect of Grade, $F(3,17) = 9.64, p < .01, \eta^2 = 0.63$ and a significant main effect of Condition $F(3,17) = 58.00, p < .001, \eta^2 = 0.91$. There was also a marginally significant Grade x Condition interaction $F(9,11) = 2.82, p = .054, \eta^2 = 0.70$ (See Table 1.2).

Table 1.2: Experiment 1 Accuracy Results

		Grade			
		1	2	3	4
Baseline	Mean	.96	1.00	.99	1.00
	SD	.08	.00	.03	.00
Orthographic	Mean	.85	.95	.97	.99
	SD	.22	.09	.09	.03
Semantic	Mean	.79	.96	.98	.97
	SD	.22	.07	.06	.07
Orthographic-Semantic	Mean	.43	.75	.87	.91
	SD	.41	.22	.13	.10

Paired samples *t*-tests of accuracy reveal that in the first grade children responded to the Baseline condition with significantly higher accuracy than the Orthographic condition $t(19) = 2.43, p < .05$ and responded to the Semantic condition with significantly higher accuracy than the Orthographic-semantic condition $t(19) = 5.77, p < .001$. Although there was no observed difference in accuracy between the Orthographic and Semantic conditions in the first grade ($t(19) = 1.49, p > .05$), large differences between these conditions existed in reaction time results, which are reported below (See Table 1.3). Paired samples *t*-tests of accuracy in the second grade participants demonstrated a similar pattern: Children responded to the Baseline condition significantly more accurately than the Orthographic condition $t(19) = 2.09, p = .05$ and to the Semantic condition significantly more accurately than the Orthographic-semantic condition $t(19) = 4.08, p < .01$. There was no difference in accuracy between the Orthographic and Semantic conditions in the second grade $t(19) = .23, p > .05$. In the third grade, children responded with significantly higher accuracy to the Semantic compared to the Orthographic-semantic condition $t(19) = 3.40, p < .005$, but there was no observed difference in accuracy between the Baseline and Orthographic conditions $t(19) = 1.03, p > .1$ or between the Orthographic and Semantic conditions in the third grade $t(19) = .34, p > .1$. In the fourth grade, there was a marginally significant difference between Semantic and Orthographic-semantic conditions (such that

children responded to the Semantic condition more accurately than the Orthographic-semantic condition), $t(19) = 1.93, p = .069$, but no differences were observed between Baseline and Orthographic $t(19) = 1.00, p > .1$, or Orthographic and Semantic conditions $t(10) = 1.28, p > .1$. While these results describe the response outcome (correct or incorrect), they do not provide a representation of processing speed or efficiency, which are reported below.

Table 1.3: Experiment 1 Mean Reaction Time Results (in seconds) to Correct Trials Only

		Grade			
		1	2	3	4
Baseline	Mean	2.70	2.13	1.92	1.80
	SD	0.87	0.50	0.43	0.43
Orthographic	Mean	3.12	2.63	2.12	2.04
	SD	1.10	0.93	0.56	1.05
Semantic	Mean	7.28	3.63	2.52	2.11
	SD	6.35	1.97	1.19	0.56
Orthographic-Semantic	Mean	6.52	3.71	2.62	2.48
	SD	7.31	1.29	0.73	1.09

Reaction Time Results

Because children in the first grade performed with such low accuracy on the Orthographic-semantic condition, reporting statistics on correct trials only did not accurately reflect the sample. Additionally, due to a speed-accuracy trade off in the Orthographic-semantic condition, reporting statistics on reaction time results alone also did not accurately depict the data. To account for this speed-accuracy trade off, analyses were performed on the mean reaction time for each participant for each condition divided by the mean accuracy for each participant in each condition. These Efficiency Score analyses (Christie & Klein, 1995; Townsend & Ashby, 1983) yielded a significant main effect of Grade $F(3,17) = 8.38, p < .01, \eta^2 = 0.60$, and a significant main effect of Condition $F(3,17) = 15.93, p < .001, \eta^2 = 0.74$. A significant Grade x Condition interaction $F(9,11) = 6.98, p < .01, \eta^2 = 0.85$ was also observed (see Figure 1.2).

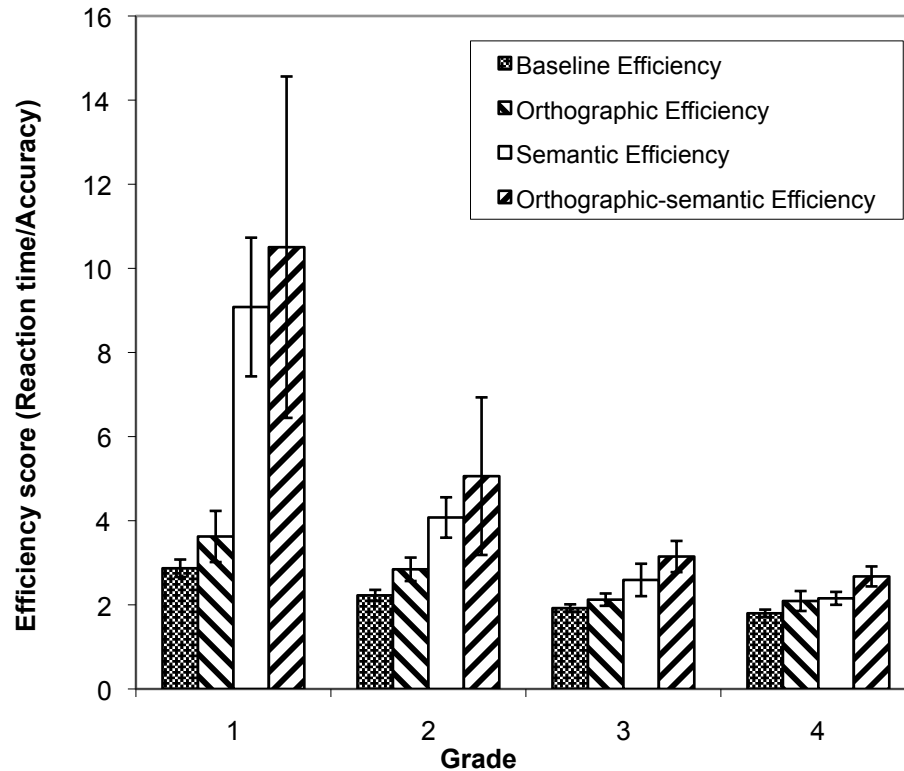


Figure 1.2: Displays efficiency scores (reaction time/accuracy) by grade for all conditions. Results are displayed with standard error bars.

*Within Grade, Between Condition Follow-up
Paired Comparisons of Efficiency*

Follow-up paired samples *t*-tests within grade and across condition reveal that in the first grade responses to the Baseline condition were significantly more efficient than the Orthographic condition, responses to the Orthographic condition were significantly more efficient than the Semantic condition, and there was no significant difference in response efficiency between the Semantic and Orthographic-semantic conditions $t(19) = 2.17, p < .05$; $t(19) = 3.37, p < .005$; $t(19) = 11.28, p > .1$ respectively. The same pattern was true for the second grade: Responses to the Baseline condition were significantly more efficient than to the Orthographic condition, $t(19) = 3.31, p < .005$, responses to the Orthographic condition were significantly more efficient than to the Semantic condition $t(19) = 2.28, p < .05$, and there were no observed differences between the Semantic and Orthographic-semantic conditions in the second grade $t(19) = 1.49, p > .1$. In the

third grade, responses to the Baseline condition were more efficient than to the Orthographic condition $t(19) = 2.37, p = .05$ and responses to the Orthographic condition were more efficient than to the Semantic condition (with marginal significance) $t(19) = 1.81, p = .08$. No differences were observed between Semantic and Orthographic-semantic processing efficiency in the third grade $t(19) = 1.62, p > .1$. In the fourth grade no differences were observed between the Baseline and Orthographic conditions $t(19) = 1.53, p > .1$ or between the Orthographic and Semantic conditions $t(19) = .42, p > .5$, but a significant difference was observed between the Semantic and Orthographic-semantic conditions, such that fourth graders responded to the Semantic condition with more efficiency than the Orthographic-semantic condition $t(19) = 2.46, p < .05$.

Results from Orthographic and Semantic Conditions

In order to control for the possibility that younger children were just slower to respond overall (i.e. differences are not due to the orthographic and semantic experimental manipulation), the mean reaction time for the Baseline condition was subtracted from the mean reaction time for the Orthographic and Semantic conditions for correct trials only. This subtraction provided a measure of orthographic and semantic processes isolated from other, more general, task demands. These differences (Orthographic – Baseline and Semantic – Baseline) were then compared within grades to achieve a measure of the fluency of orthographic and semantic processing between grades. Even after controlling for general task demands (subtracting the Baseline condition from the Orthographic and Semantic conditions), the pattern of orthographic and semantic processing results for children in the first through fourth grades remained the same: Paired t -tests revealed orthographic processing speed to be significantly faster than semantic processing speed (Orthographic – Baseline compared to Semantic – Baseline respectively) in the first grade $t(19) = 3.20, p < .005$; second grade, $t(19) = 2.22, p < .05$; marginally in the third grade $t(19) = 2.01, p =$

.059; and in the fourth grade there was no significant difference between orthographic and semantic processing speed $t(19) = .10, p > .05$ (See Figure 1.3).

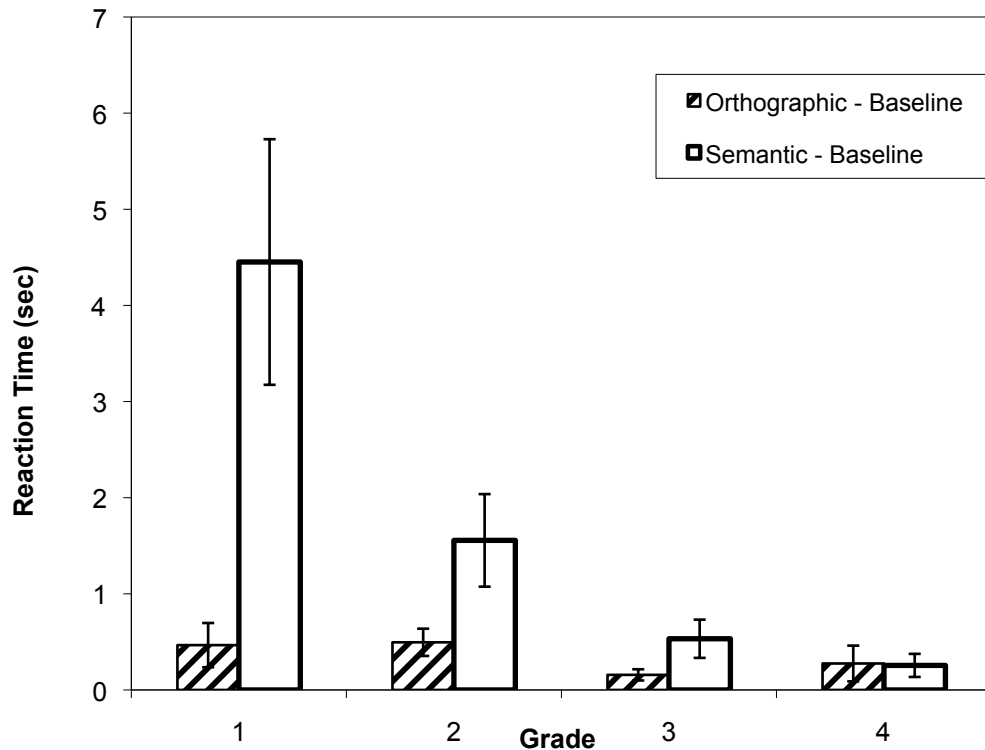


Figure 1.3: Displays the mean reaction time to Orthographic minus Baseline (hatched) and Semantic minus Baseline (solid) for correct trials only in Experiment 1. Results are displayed with standard error bars.

DISCUSSION

Accuracy analyses revealed that older children were more accurate than younger children overall, but also that younger children were relatively less accurate than older children to identify the semantic match (e.g. SHIP for BOAT), when in the presence of an orthographic foil (e.g. BOOT). In this condition (Orthographic-semantic), the children in the first grade chose the orthographic foil more often than the correct synonym match (and more often than would be predicted by chance), suggesting that they accepted the orthographic neighbor as the target word. One question that may arise from such a result is whether these young children knew the meaning of the synonym match, or whether they were guessing between an unknown word, and a word

that was perceptually similar. Importantly, the Semantic condition acted as a control for the Orthographic-semantic condition. In the Semantic condition, children (between subjects) saw the same target and match word, but the foil was unrelated both orthographically and semantically to the target word. Therefore, if children did not know the synonym match, they would have performed at chance on the Semantic condition. This was not the case. While children in the first grade performed with only 43% accuracy on the Orthographic-semantic condition, their performance on the Semantic condition was significantly higher (79% correct), suggesting that it was indeed the presence of the orthographic neighbor which caused young readers to err, rather than the possibility that the children did not know the meaning of the synonym match in the Orthographic-semantic condition.

These results support lexical tuning hypothesis (Castles et al., 2007), which suggests that younger children have more “coarsely-tuned” orthographic processing, and thus were more likely to accept an orthographic neighbor as a target word. This is apparent from the accuracy and efficiency results of the Baseline relative to Orthographic condition in the younger compared to older participants. Results demonstrate that responses to the Baseline condition (no orthographic foil) were more *accurate* relative to the Orthographic condition (orthographic neighbor foil) in the first and second grade, but not the third and fourth grade. Responses were also more *efficient* in the Baseline condition relative to the Orthographic condition in the first, second, and third grade but not in the fourth grade. Thus, both accuracy and efficiency results indicate that the younger groups accepted the orthographic neighbor as the target more often than an orthographically unrelated foil, suggesting a gradual tuning of orthographic processing. However, according to the lexical tuning hypothesis, it is vocabulary growth that drives the fine-tuning of the orthographic processing system. It is an assumption of the current experiment that the children in the older grades did indeed have larger vocabularies, as no specific standardized vocabulary measures were taken, and vocabulary size was therefore not specifically correlated

with performance on the experimental conditions. Because all children in the study were typically developing, participated in age-appropriate school curricula, and had no history of learning or developmental delay, however, it seems reasonable to assume that vocabulary size and age were closely correlated in this sample of elementary school children, as is the case for typically developing children in general.

Efficiency scores revealed that younger children performed well on the conditions requiring orthographic processing (Baseline and Orthographic), but less efficiently on conditions that required semantic processing. Importantly, there was a qualitative difference between young participants' performance on the Semantic condition (synonym match without an orthographically related foil) and the Orthographic-semantic condition (synonym match and orthographic neighbor foil). First graders, on average, responded slowly with high accuracy to the Semantic condition, but quickly and with low accuracy to the Orthographic-semantic condition. These two conditions both require the selection of a synonym match, and differ only in terms of the foil word, specifically the presence or absence of an orthographic neighbor. This result suggests that while children in the youngest age group could indeed identify the synonym match correctly, these children relied too heavily on a coarsely-tuned orthographic processing system, which prompted them to incorrectly respond that the orthographic neighbor was the target word.

Children in the first grade were highly variable in their reaction time to identify the target. Since the first author collected all the data individually, however, it can be verified that even the longest latencies were due to processing time. Because it appears that the variability in this youngest age group is an important hallmark of reading development, we have chosen not to remove data points that may be considered outliers (Ulrich & Miller, 1994). However, we note that only one data point falls more than three standard deviations above the mean (a first grader's response to the Orthographic-semantic condition), and no reaction times (in any of the grades) were less than one second, a time-window which may be considered to be an uninterpretable fast

response time for the task at hand. Additionally, even the children who took a very long time to process the written stimuli demonstrated a greater than chance performance on the Semantic condition. We believe that the high variability of the first grade readers in the conditions requiring semantic processing is attributable to the transition from an orthographically-based to semantically-based reading strategy. While some of the first graders have begun to activate some meaning directly from the text, others continue to sound out the word, resulting in much longer reaction time, less efficient reading, and consequently increased variability within this age-group. The results from the youngest group of participants can be explained within linear models of reading such as the phonologically mediated pathway of the Dual Route, which suggest that the pathway from decoding to comprehension occurs in a more serial manner (e.g. Morton, 1969; and see Lupker, 2008 for a review of linear and cascaded models). Results from the older participants, however, suggest a more cascaded or connectionist route from orthographic to semantic representations, such as the triangle model. Results from older participants are discussed below.

In contrast to the youngest group of readers, children in the second, third, and fourth grade demonstrated the slowest reaction times (and least efficient processing) to the Orthographic-semantic condition. In these older grades, when the semantic information from the orthographic neighbor (foil) was in competition with the semantic information from the synonym match, children showed slower reaction times, indicating that some semantic processing occurred *before* completion of the orthographic processing. This interference effect suggests that by second grade, the route from orthography to meaning was more cascaded (e.g. Coltheart, Rastle, Perry, Langdon, & Ziegler, 2001; Forster & Hector, 2002).

One outstanding question, however, is whether the orthographic processing investigated in Experiment 1 is truly a learned component specific to reading, or whether children were simply matching symbols in the Baseline and Orthographic conditions. Experiment 2 is thus a control experiment designed to address the possibility that children were using a symbol search strategy

to complete to the Orthographic and Baseline conditions, rather than utilizing learned orthographic processes specific to reading.

EXPERIMENT 2

INTRODUCTION

Experiment 2 is an experimental control condition, which tests the hypothesis: Is the early-established orthographic representation evidenced in Experiment 1 specific to reading in a learned orthography, or would children demonstrate the same pattern of results when matching any complex visual stimuli? To address this possible confound, the same stimuli from the Baseline and Orthographic conditions in Experiment 1 were presented in a false font (unfamiliar letter-like visual stimuli that do not contain any meaning) in Experiment 2 (for examples see Table 1.4). If the efficient orthographic processing observed in Experiment 1 did *not* index the acquisition of a component of reading, but rather reflected a symbol matching strategy, then matching any set of word-like complex visual stimuli should evoke a similar pattern of accuracy and reaction time results to Experiment 1 in the conditions that do not require semantic processing. If, on the other hand, the results from Experiment 1 *did* reflect a component specific to reading acquisition (namely orthographic processing), we would expect orthographic processing in Experiment 2 (false font) to be processed less efficiently than that observed in Experiment 1 (real words).

Table 1.4: Examples of Stimuli for Experiment 2

		Condition	
		Baseline	Orthographic
Experiment 1	Target	SMALL	SMALL
	Match	SMALL	SMALL
	Foil	APPLE	SMELL
Experiment 2	Target	⌘⌘⌘⌘⌘	⌘⌘⌘⌘⌘
	Match	⌘⌘⌘⌘⌘	⌘⌘⌘⌘⌘
	Foil	⌘⌘⌘⌘⌘	⌘⌘⌘⌘⌘

METHODS

Participants

Thirty-two typically developing children from San Diego public schools participated in Experiment 2: Eight first graders (4 female, 4 male; M age = 7.06 years; SD = .37), 8 second graders (5 female, 3 male; M age = 7.97 years; SD = .31), 8 third graders (3 female, 5 male; M age = 8.77 years; SD = .56), and 8 fourth graders (4 female, 4 male; M age = 9.87 years; SD = .27).

Procedure and Materials

The same experimental paradigm from Experiment 1 was employed in Experiment 2, with the exception that all stimuli were presented in a false font (Microsoft Word Wingdings 3). Wingdings 3 is a false font based on the English alphabet (one symbol stands for one letter), but the “letters” do not resemble letters from the Roman alphabet. Because each false font symbol represents one English letter, the same words from Experiment 1 were presented, but in this non-meaningful font. Thus, Word length and proportion of similarity between the Baseline and Orthographic conditions remained the same across Experiments 1 and 2. Specifically, the Orthographic foil differed from the Orthographic target by only one symbol, and consequently required more finely tuned discrimination to distinguish the target from the foil (see Table 1.4). In the Baseline condition, by contrast, the foil was non-similar to the target, and thus required more coarsely tuned discrimination. Since false fonts carry no semantic information, only the conditions that did not require processing of meaning were presented in Experiment 2 (Baseline and Orthographic).

RESULTS

Accuracy Results

Data from Experiment 2 were analyzed using two 4 (Grade) x 2 (Condition) repeated measures ANOVAs, one with the proportion correct as dependent variable and one using mean (untrimmed) reaction time normalized by accuracy (Efficiency scores). Accuracy results yielded a

significant effect of Condition $F(1,7) = 11.95, p < .05, \eta^2 = 0.63$, but no significant main effect of Grade $F(3,5) = 4.03, p > .05, \eta^2 = 0.71$; and no significant Grade x Condition interaction $F(3,5) = 3.69, p > .05, \eta^2 = 0.69$ (See Table 1.5).

Table 1.5: Experiment 2 Accuracy Results

		Grade			
		1	2	3	4
Baseline	Mean	.99	.99	1.00	1.00
	SD	.03	.03	.00	.00
Orthographic	Mean	.83	.91	.99	1.00
	SD	.19	.09	.03	.00

Reaction Time Results

Efficiency score results (Reaction time/Accuracy) also yielded a significant main effect of Condition $F(1,7) = 215.97, p < .001, \eta^2 = 0.97$, such that children responded to the Baseline condition more efficiently than the Orthographic condition (in which the symbols differed by only one letter), but also did not yield a significant main effect of Grade $F(3,5) = 1.11, p > .1, \eta^2 = 0.40$, or any significant interactions (Table 1.6).

Table 1.6: Experiment 2 Efficiency Score Results (Reaction Time/Accuracy)

		Grade			
		1	2	3	4
Baseline	Mean	3.09	3.63	3.33	3.21
	SD	0.70	0.96	0.99	0.77
Orthographic	Mean	4.10	4.60	5.31	5.08
	SD	1.26	1.07	1.31	1.49

Results from Baseline and Orthographic Conditions between Experiments 1 and 2

The amount of facilitation between Orthographic and Baseline conditions was also computed (mean Orthographic RT- Baseline RT to correct trials only) for Experiment 2, and was compared to that of Experiment 1 using non-parametric Independent Samples Mann-Whitney U tests. While the null hypothesis could not be rejected for children in grades 1 and 2, the null

hypothesis was rejected in grades 3 and 4, suggesting that processing speed in grades 3 and 4 differed between Experiments 1 and 2 (See Table 1.7).

Table 1.7: Orthographic Facilitation for Real Words (Experiment 1) Relative to False Fonts (Experiment 2), Correct Trials Only

		Grade			
		1	2	3	4
Experiment 1	Orthographic (O) – Baseline (B)	0.42	0.5	0.2	0.24
Experiment 2	Orthographic (O) – Baseline (B)	1.01	0.97	1.98	1.87
Facilitation (seconds)	E2 (O – B) – E1 (O – B)	0.59	0.47	1.78	1.63

DISCUSSION

The pattern of results from Experiment 2 differs from that observed in Experiment 1. In Experiment 1, words were presented in English, and results demonstrated a main effect of both Grade and Condition. Experiment 2, which presented stimuli in a false font, demonstrated a main effect of Condition, but not Grade. This result indicates that processing the false font did not improve with age, and thus strongly suggests that the improvement in orthographic processing in Experiment 1 was due to improved *reading* and not improved symbol matching, a skill which becomes qualitatively different from reading as children develop skills to recognize words directly from the orthography. There was also some evidence from Experiment 2 that early reading is similar to symbol matching, but becomes progressively dissimilar in the older grades. That is, children in first and second grade responded with more similar speed and accuracy to words and false fonts compared to children in the older grades (See Figure 1.4). This result may be due to the fact that younger children use a letter-by-letter strategy to read words (more consistent with linear models of reading), and thus for these young children, matching symbols in Experiment 2 may not have been a drastically different task from reading words in Experiment 1.

For the older children who likely no longer utilize a letter-by-letter strategy for reading words, the false font condition of Experiment 2 was more difficult, and consequently was answered with longer reaction times.

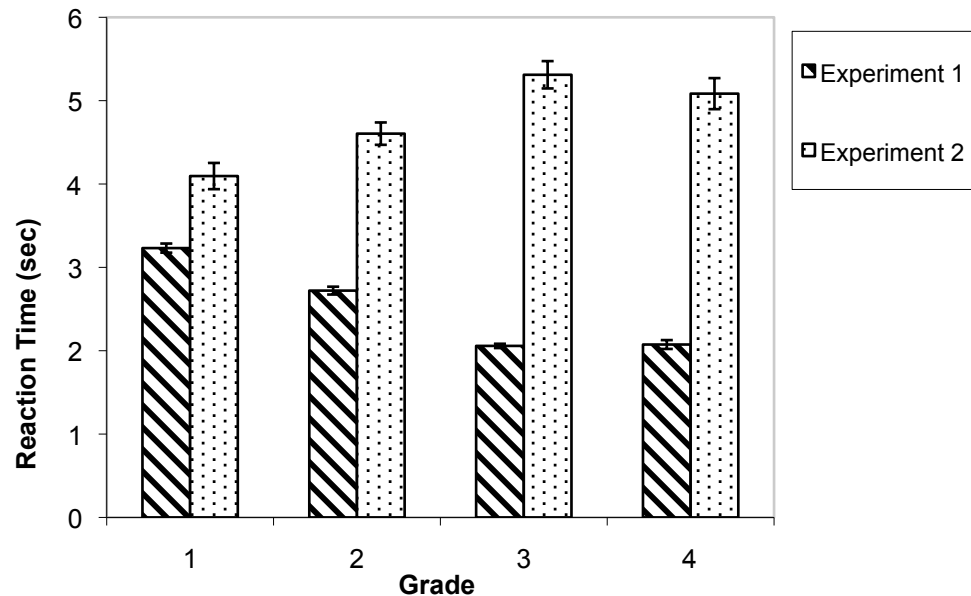


Figure 1.4: Compares the mean Reaction Times to correct trials only in the Orthographic condition between Experiment 1 (stimuli presented in the English orthography) and Experiment 2 (stimuli presented in a false font). Results are displayed for the first through fourth grade with standard error bars.

It is likely that the faster reaction times observed in older children to words and *not* to symbols, was also due to the fact that third and fourth grade children had learned to “chunk” words into whole words or word-parts, and thus had become less focused on the individual symbols that comprise the word when reading real words. This strategy is dependent upon having a familiarity with a word and some cascaded semantic processing of the word, two strategies that would be ineffectual in a symbol search task using unfamiliar and non-meaningful symbol strings. This different processing strategy would explain the increasingly disparate results between Experiments 1 and 2 with age, and provide further evidence that children in the younger grades are more focused on the orthographic shape of the word (a perceptual component). This explanation would further suggest that while both words and symbols begin as meaningless

marks on a page, with increasing exposure the symbols that become meaningful (i.e. words) come to be processed in a qualitatively different manner from novel symbols. In Experiment 2 we see that the younger children still process words and novel symbols similarly, whereas the older children have learned to process words in a different, more efficient manner, which does not generalize to the meaningless symbols.

GENERAL DISCUSSION

Results from the present investigation indicate that developing the orthographic and semantic components necessary for skilled reading follow different developmental trajectories during reading acquisition. Strong perceptual-level skills required to efficiently form an orthographic representation precede strong conceptual-level skills required to efficiently map the orthographic representation onto a meaning. Although even the youngest readers demonstrate the ability to process both the more perceptual-level orthographic information and the more conceptual-level semantic information, results from the present investigation suggest that these processes are not yet equally efficient. These results are in accordance with previous research that suggests that younger children rely on more concrete, perceptual systems before transitioning to more flexible, abstract cognition later in childhood (Quinn & Eimas, 1997; Rayner, 1988).

Results from Experiment 1 demonstrate a rapid increase in semantic processing efficiency in each subsequent grade. The youngest group of participants (six and seven-year-olds) demonstrated very fast, though coarsely tuned orthographic processing, and were slow to extract meaning from an orthographic representation. Children in the second grade showed fast orthographic processing (though still demonstrated some over-reliance on the perceptual, orthographic level), and also a decreased time lag to access semantic information from the text (faster access to a semantic representation). The difference in processing time between deciphering the orthographic code and accessing a semantic representation was even smaller in the third grade, and in the fourth grade there was no significant difference in reaction time

between forming an orthographic and semantic representation. This suggests that by the fourth grade, the semantic processing system is just as strong as the orthographic system. Thus, the current results from children in the second, third, and fourth grades (though not the youngest, first grade readers) are consistent with 1) Connectionist, cascaded models of reading (e.g. Harm & Seidenberg, 2004; Plaut et al., 1996), in which semantic knowledge influences the decoding and lexical retrieval processes and 2) “Direct routes” of reading comprehension, in which meaning can be accessed automatically from an orthographic representation without an intermediary phonological decoding step (Coltheart et al., 2001). These older readers were also less likely to accept an orthographic neighbor as a target word, indicating finer tuned orthographic processing, consistent with the lexical tuning hypothesis (Castles et al., 2007). The youngest readers, however, showed evidence of more coarsely tuned orthographic processing and slow lexical access, which is consistent with more linear models of reading (e.g. Morton, 1969), and the phonologically mediated route of Coltheart’s (2001) Dual Route model. These results suggest that there exists a transition from a phonologically mediated, indirect, linear route, to a more automatic, direct, or cascaded route, which involve semantic activation before orthographic processing is complete. The present results suggest that this transition occurs quite early; at around the third grade.

An alternative explanation for this finding is that the younger children lacked the cognitive control required to inhibit their initial response (the orthographic neighbor), and that the superior performance of the older children indexes the maturation of inhibitory mechanisms. While this possibility should be further explored, first graders’ performance on the Orthographic condition (orthographic match and orthographic-neighbor foil) provides some evidence against this explanation. In the Orthographic condition, children must choose between the orthographic neighbor, and an orthographic match. Children did not demonstrate difficulty inhibiting the incorrect response to the orthographic neighbor in this condition (though it has the same degree of

similarity to the target as the foil in the Orthographic-semantic condition), suggesting that it is not a lack of cognitive inhibition *in general* but rather difficulty inhibiting the perceptually similar response when it is in direct contrast with a perceptually dissimilar response option. If this is the case, the subsequent question must be *why are younger children unable to inhibit an orthographically similar response, yet able to inhibit a semantically similar response?* Children are less likely to inhibit a more automatic response, suggesting again that semantic activation is not yet automatic in young readers, and automatic orthographic processing has already emerged (making it more difficult to inhibit). In other words, inhibiting the stronger system is more difficult, thus resulting in lower accuracy for the Orthographic-semantic condition. Although discussed within a cognitive inhibition model, rather than a reading model, this interpretation is consistent with the developmental shift from perceptual to abstract cognition.

Experiment 2 was a control condition to ensure that the orthographic processing tested in Experiment 1 was specific to reading, and did not tap a symbol matching task, unrelated to reading acquisition. Experiment 2 found that matching symbol strings and matching words was more similar for the children in the younger grades than for children in the older grades, and that children in the older grades were actually slower than those in the younger age-groups to match the symbol strings. This is the opposite pattern as that observed in Experiment 1 for reading words, in which case the older participants were faster and more efficient on every condition. Results of Experiment 2 illustrate two important findings: First, that in the older groups, reading words and matching symbols appear to tap two separable processes, as younger children perform better on the symbol matching and older children perform better on the word matching. Second, results suggest that younger children may use more of a symbol-search strategy in everyday reading, as they were able to carry those skills to the new, non-meaningful font, and rapidly and accurately identify the matching string of symbols. Children in the older grades had likely learned to chunk words, or parts of words, to recognize words as a unit. Thus the letter-by-letter search

strategy was tedious and less efficient. Together results from Experiments 1 and 2 suggest that children develop orthographic processing skills very early, and create efficient links joining orthographic and semantic representations at about the third grade (around eight years of age).

Although the present investigation reports evidence that this transition occurs approximately a year later than previous studies discussed in the Introduction (namely Nation & Cocksey, 2009), the finding that automatic orthographic activation is present in early to mid elementary school years is in accordance with these previous findings. It is possible that the participant population and task may account for the slight differences observed in age. For example, in Nation and Cocksey's (2009) investigation, participants scored above average on standardized measures of reading, suggesting that this sample was slightly ahead of their peers in reading development. In addition, while the mean age of participants in Nation and Cocksey's (2009) investigation was around seven and a half years, children ranged from 6.58 to 8.75, a span of over two years, and indeed the children on the older end of this age-range were the same age as the children in the present investigation who demonstrated similar results. In view of these differences in participant demographics and age-range within groups, we believe that the results stand to corroborate, rather than contradict previous findings. Specifically, similar to the present results, investigations by Nation and Cocksey (2009), Pratarelli and colleagues (1994), and Zecker (1991), which were discussed above, all report evidence of automatic lexical access between seven and nine years of age.

The results from the present investigation add to our understanding of reading development in the elementary school years, and may have implications for reading interventions for children who struggle with reading. However, there are limitations to the applicability of the current study, which require further exploration. First of all, the results from orthographic and semantic processing are limited to the English orthography, and do not take into account orthographies that vary in the amount of grapheme to phoneme correspondence, or the amount of

information represented by each orthographic symbol (e.g. phonemic, morphological, or lexical). While it is not addressed in the current study, the complexity of a language's orthography will likely impact the profile of orthographic and semantic development during reading acquisition. More research is needed to determine whether the profile of the two components of reading will differ depending on the specific orthography studied. Future research should investigate orthographies in which each orthographic symbol carries semantic meaning, to determine whether children follow the same developmental profile (orthography to semantics) when the semantic information is embedded within orthographic characters.

A second limitation to the present study is its focus on the relationship between orthography and semantics, without directly investigating the phonological component. The reason for focusing on the orthographic-semantic contrast was both logistical and theoretical. Logistically, we did not wish to use non-word stimuli, as we were interested in probing how children access *meaning* from text, yet words that have both a homonym *and* a synonym (e.g. STAIR has both a synonym *step* and a homonym *stare*) to utilize as target words are extremely scarce. Theoretically, it was the goal of the present investigation to explore reading development within a framework that views development as a shift from cognition that is dependent on perceptual, observable features, to more abstract cognition. The study of orthography (a directly observable component) and semantics (an abstract component), allowed for the results of the present investigation to be interpreted within this concrete to abstract cognitive framework.

Lastly, the design of the present investigation is cross-sectional, which is susceptible to group differences rather than developmental differences, per se. To ensure that the differences in grade were attributable to developmental changes, a follow-up longitudinal design should be implemented.

CONCLUSION

For most adults, reading is a fairly effortless process, one in which text leaps off the page and becomes an abstract concept. This may make it difficult to appreciate that reading is the result of separable cognitive processes, which children must integrate during the course of reading acquisition. The present results highlight the fact that these processes do not develop simultaneously, but rather reflect a transition from more perceptual representations (orthographic) to more conceptual representations (semantic), a shift that occurs in the early elementary school years.

APPENDIX A

Table 1.8: All Stimuli

Target	Baseline		Orthographic		Semantic		Orthographic-Semantic	
	Match	Foil	Match	Foil	Match	Foil	Match	Foil
BIG	BIG	TAPE	BIG	BIB	LARGE	TAPE	LARGE	BIB
BOAT	BOAT	GAME	BOAT	BOOT	SHIP	GAME	SHIP	BOOT
BUCKET	BUCKET	SCHOOL	BUCKET	BUCKLE	PAIL	SHCOOL	PAIL	BUCKLE
BUG	BUG	TABLE	BUG	BUS	INSECT	TABLE	INSECT	BUS
CAP	CAP	PIG	CAP	CAT	HAT	PIG	HAT	CAT
ROAD	ROAD	LIKE	ROAD	ROAR	STREET	LIKE	STREET	ROAR
COAT	COAT	HAND	COAT	COLT	JACKET	HAND	JACKET	COLT
CUT	CUT	OWL	CUT	CUB	SNIP	OWL	SNIP	CUB
HOP	HOP	PEN	HOP	HOT	JUMP	PEN	JUMP	HOT
KID	KID	BALL	KID	KIT	CHILD	BALL	CHILD	KIT
LADY	LADY	WATER	LADY	LAZY	WOMAN	WATER	WOMAN	LAZY
LATE	LATE	SPOON	LATE	LAKE	TARDY	SPOON	TARDY	LAKE
MAD	MAD	SHEEP	MAD	MAP	ANGRY	SHEEP	ANGRY	MAP
MONEY	MONEY	STOP	MONEY	MONKEY	CASH	STOP	CASH	MONKEY
MUG	MUG	FLY	MUG	MUD	CUP	FLY	CUP	MUD
PLATE	PLATE	FIRE	PLATE	PLANE	DISH	FIRE	DISH	PLANE
RUG	RUG	BIRD	RUG	RUN	CARPET	BIRD	CARPET	RUN
SEAT	SEAT	WORM	SEAT	SEAL	CHAIR	WORM	CHAIR	SEAL
SICK	SICK	FROG	SICK	SINK	ILL	FROG	ILL	SINK
SMALL	SMALL	APPLE	SMALL	SMELL	LITTLE	APPLE	LITTLE	SMELL
STAIR	STAIR	GIRL	STAIR	STAIN	STEP	GIRL	STEP	STAIN
STONE	STONE	EARS	STONE	STOVE	ROCK	EARS	ROCK	STOVE
GUY	GUY	PILE	GUY	GUM	MAN	PILE	MAN	GUM
LOOK	LOOK	BARN	LOOK	LOCK	SEE	BARN	SEE	LOCK
SHOVE	SHOVE	BATH	SHOVE	SHORE	PUSH	BATH	PUSH	SHORE
LEAVE	LEAVE	POOL	LEAVE	LEASE	EXIT	POOL	EXIT	LEASE

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CHAPTER 2: SINGLE WORD READING IN CHILDREN WITH WILLIAMS SYNDROME AND HIGH FUNCTIONING CHILDREN WITH AUTISM: A STUDY OF CONTRASTS

ABSTRACT

Children with neurodevelopmental disorders provide a unique window into the organization underlying the acquisition and development of cognitive processes. In this investigation, we focus on the complementary cognitive-linguistic profiles of children with Williams syndrome (WS) and High Functioning children with Autism (HFA), to elucidate relations between components of single-word reading. Children with HFA demonstrate superior visuospatial processing and relatively poor language; children with WS demonstrate degraded visuospatial processing with relatively proficient language. Reading requires both visuospatial processing and language; visuospatial processes are necessary to decipher the orthography, and language is required to understand meaning. We investigate how these visuospatial-linguistic profiles are reflected in a single-word reading task. Results suggest children with WS perform with similar accuracy to typically developing children (TD) on conditions requiring semantic processing, but fall below on conditions requiring fine-level orthographic discrimination. Children with HFA, by contrast, performed similarly to TD on conditions requiring fine-level orthographic discrimination, but not on all conditions requiring semantic processing. Results indicate that established cognitive strengths and weaknesses in WS and HFA are reflected in single-word reading. These findings have clinical implications for creating more targeted reading interventions for these populations, and also reveal relations between components underlying typical reading.

INTRODUCTION

Because in typically developing children, language and reading co-develop, it is difficult to identify the independent components underlying reading acquisition and growth. The atypical

developmental trajectories of children with neurodevelopmental disorders, however, allow us to better understand the components underlying complex systems such as reading, and their relations to one another. The contrastive cognitive-linguistic profiles of children with Williams Syndrome (WS) and High Functioning children with Autism (HFA) provide a unique opportunity to probe the relationship between components underlying reading. In this investigation, we look specifically at orthographic and semantic components of reading to address the questions, *How are deficits observed in the visuospatial and/or language profiles of children with neurodevelopmental disorders reflected in reading performance in the school-age years? How does this reading profile compare to typical reading development? And lastly, What can this tell us about the relations of components underlying reading in typical development?*

Cognitive profiles of children within neurodevelopmental disorders are often characterized by relative strengths and weaknesses, which are more pronounced relative to the typical population. While some areas of processing may be spared or even enhanced, others tend to be disproportionately impaired. By investigating such contrasting strengths and weaknesses within the single-word reading domain that contrasts orthographic and semantic processing, we aim to add to our understanding of the relationship between core neurodevelopmental deficits and single-word reading performance.

COGNITIVE PHENOTYPES OF WS AND HFA

WS is a rare genetic disorder that affects approximately 1 in 7,500 to 1 in 20,000 live births, and is caused by a hemizygous deletion at chromosome 7q11.23, which includes the Elastin gene (Korenberg et al., 2000). Abnormalities in connective tissue and cardiovascular functioning, as well as mild to moderate mental retardation are consequences of this deletion. Furthermore, individuals with WS have distinctive facial features, and often project a unique, friendly, and outgoing personality (Järvinen-Pasley, Wallace, Ramus, Happé, & Heaton, 2008; Martens, Wilson, & Reutens, 2008; Mervis et al., 2000). Despite mild to moderate mental

retardation (IQs tend to be around 60 in both adults (Howlin, Davies, & Udwin, 1998) and children (Donnai & Karmiloff-Smith, 2000)), individuals with WS tend to show islands of strength in specific facets of language (for a review see Brock, 2007; Mervis & John, 2010), especially affective language (Losh, Bellugi, & Reilly, 2000; Reilly, Bernicot, Vicari, Lacroix, & Bellugi, 2005), whereas performance on non-verbal visuospatial tasks (e.g. mental rotation, block design and pattern recognition) tends to be severely impaired (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Doyle, Bellugi, Korenberg, & Graham, 2004; Farran, Jarrold, & Gathercole, 2001; Järvinen-pasley et al., 2008; Mervis, Robinson, & Pani, 1999).

Children with Autism Spectrum Disorder (ASD) demonstrate a complementary and contrastive cognitive profile: as a group they are characterized by deficits in sociability and communication (APA, 2000; WHO, 1992) relative to visuospatial processing tasks, which remain unimpaired or even enhanced (Baldassi et al., 2009; O’Riordan, 2004; O’Riordan, Plaisted, Driver, & Baron-Cohen, 2001; Samson, Mottron, Soulieres, & Zeffiro, 2012). Here we focus on a subset of children with ASD, High Functioning children with Autism (HFA). Unlike most children with ASD who have some intellectual impairment (Tager-Flusberg, 2007), the subset of children with HFA (20-30% of children diagnosed with ASD) (Gillberg & Coleman, 1992) demonstrate Full Scale IQs within the normal range (defined here as 70 or above) (for a detailed intellectual profile in HFA, see Ghaziuddin & Mountain-Kimchi, 2004). The following sections will further explore the contrasting language and visuospatial profiles observed in WS and HFA as a context for investigating the components underlying typical and atypical single-word reading. Since children with WS show specific deficits in visuospatial processing (a skill required for orthographic processing) and children with HFA show specific deficits in semantic processing (a skill required for comprehension), these populations provide an opportunity to disentangle the unique contributions of orthographic and semantic processes to successful single-word reading.

LANGUAGE PROFILES OF WS AND HFA

Language in WS

Despite their general cognitive impairment, it has been observed again and again that mature individuals with WS frequently produce sentences with complex syntax, use sophisticated and unusual vocabulary, and tell stories with increased linguistic social devices (e.g. Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Reilly, Klima & Bellugi, 1990; Udwin & Yule, 1990; von Armin & Engel, 1964). From these observations, it appears that language processing is a relative strength for this group. Studies of language performance in WS however, have provided a detailed and nuanced profile of the complex language profile in WS, which suggest that there are strengths and weaknesses within the language domain (for more comprehensive reviews, see Bellugi et al., 2000; Brock, 2007; Järvinen-pasley et al., 2008; Karmiloff-smith, Brown, Grice, & Paterson, 2003; Martens et al., 2008; Mervis et al., 2000; Mervis & John, 2010; Reilly, Lai, Bernicot, & Bellugi, 2010). Here, we provide a brief overview of formal aspects of language, including phonological, morphological, and syntactic processing, as well as semantic and discourse processing, with an emphasis on the relatively strong affective and semantic language systems.

Although some investigations have found phonological processing in WS to be appropriate to (or even exceeding) mental age (Böhning, Campbell, & Karmiloff-Smith, 2002), others have identified deficits in aspects of phonological processing. Nazzi, Paterson, and Karmiloff-Smith (2003), for example, found that toddlers with WS are severely delayed in using phonological cues to segment the speech stream, and other studies suggest that this deficit persists into the school-age years, where children with WS have difficulty with some aspects of phonological processing (such as phoneme deletion) in comparison to age and reading matched controls (Laing, Hulme, Grant, & Karmiloff-Smith, 2001). Other reports suggest that while measures of more automatic short-term phonological memory are preserved (e.g. non-word

repetition), phonological awareness tasks that require more metacognitive processes are degraded (Majerus, Barisnikov, Vuillemin, & Poncelet, 2003).

Investigations of morphological processing in WS have produced varied results. Some studies probing regular and irregular verb and noun morphology have indicated better performance on regular relative to irregular (Clahsen, Ring, Temple, 2004; Pléh, Lukács, & Racsmány, 2003), suggesting that WS individuals rely on rule-based morphological processing rather than independent lexical representations. Other studies, however, do not find such a regularity effect (Thomas et al., 2001; Zukowski, 2005). Importantly, while some of the aforementioned investigations did find evidence for enhanced processing of regular relative to irregular morphology, none of the morphological processing measures were found to be above the mental age-matched comparison group. This finding raises the question of which group is the most appropriate comparison group for investigations of WS language. To address this issue, Losh, Reilly, and Bellugi (2000) evaluated linguistic components (including morphological errors) on an oral narrative task relative to three control groups: typically developing, age-matched controls; language-matched, younger controls; and visuospatially matched, younger controls. With regard to morphological errors, this study found that children with WS consistently made more errors than all three typically developing comparison groups. Also using a narrative task, Reilly and colleagues (2004) found that a group of school age children with WS made as many errors, proportionally, as did chronologically age matched children with Language Impairment who were not mentally impaired. Together these studies suggest that productive morphology poses a challenge to children with WS.

In contrast to problems with morphology, some studies find that children with WS show better syntactic processing capabilities than their intellectual age-matched controls. On sentences such as *the horse chased the cow*, for example, which are semantically reversible (i.e. both the cow and the horse could be the chaser or the chasee), children with WS were able to identify the

picture that matched the meaning of the sentence more often than their intellectual age-matched controls (Bellugi, Bihrlé, Jernigan, Trauner, & Doherty, 1990; Bellugi, Klima, & Wang, 1996). This finding suggests that individuals with WS have some understanding of the underlying grammatical structure of the sentence, as they could not complete the task relying on semantic cues alone. Additional standardized measures of grammatical and syntactic processing have also found increased performance relative to children with Down syndrome (DNS) (Bellugi et al., 2000), who have similar intellectual impairment. In a narrative context, measuring productive syntactic performance, Losh and colleagues (2000) found that children with WS used less complex syntax relative to the typically developing control groups, but found that the frequency of complex syntax increased with age, and many of the older children with WS fell within the normal range for use of complex syntax. In sum, results from investigations of phonology, morphology, and syntax are mixed, but generally suggest that performance on these formal language facets is consistently below that of age-matched typically developing children. We now turn to semantic and discourse processing, two language areas which show a unique and complex profile in WS.

There is both anecdotal and empirical evidence that children with WS use unusual, and sometimes sophisticated vocabulary in their spontaneous language (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Udwin & Yule, 1990). As described by Bellugi, Bihrlé, Jernigan, Trauner, and Doherty (1990), when asked to list as many animals as possible, children with WS consistently named unusual animals such as *unicorn*, *tyranadon*, *brontosaurus*, and *yak*, rather than the typical, less exotic animals *dog*, *cat*, *bird*, *horse*, which are reliably named by typically developing children. Mervis and John (2008) also report that the majority of WS participants in their investigation demonstrated knowledge of concrete vocabulary that was within the low-normal range on a standardized measure of receptive vocabulary.

There is some evidence that the unique vocabulary observed in individuals with WS and may be driven by unusual or enriched semantic representations. Rossen Klima, Bellugi, Bihrlé, and Jones (1996) presented homonymous words (words with more than one meaning: one primary and one secondary) to children with WS, DNS, and typically developing fourth graders. Responses on a similarity judgment and definition task demonstrated evidence of distinctive semantic representations from TD and DNS. Specifically, when given a word triad (the homonymous word and a word related to the primary and secondary meaning) and asked to choose the two words that go together best, TD and children with DNS were both more likely to choose the homonym and the word related to the primary meaning. Children with WS, by contrast, were just as likely to identify the word related to the secondary meaning. Similarly, in a definition task, when children were asked to describe the meaning of the homonymous word, children with WS were just as likely to describe the primary meaning as typically developing children and children with DNS, but they were more likely than the other groups to also describe the less dominant, secondary meaning (Rossen et al., 1996). These findings provide evidence for unusual semantic representations in WS, but also suggest that relative to other cognitive domains, semantic processing is a relative strength.

Because of their sociability, individuals with WS are especially adept at engaging others in social interactions (Bellugi et al., 1996). Thus, due to the social context, story telling, or narrative performance, is one of the strongest language contexts for school-aged children with WS. Using a narrative assessment, in which children are asked to tell a story from a series of pictures, children with WS were found to produce narratives characterized by increased use of evaluative devices, especially social language (e.g. emotional terms and intensifiers), even when compared to typically developing children (Reilly et al., 2005). However, while children with WS are adept at using such evaluative devices in their narratives, they show deficits in more

integrative components such as story complexity and in the sequencing and global coherence of the narrative (Marini, Martelli, Gagliardi, Fabbro, & Borgatti, 2010).

It is apparent from this brief review of language processing in WS that there is considerable variability in the findings, particularly in areas of phonological and morphological processing. Nonetheless, lexico-semantic processing consistently emerges as a strength in this group, especially relative to other cognitive functions such as visuospatial processing, discussed below.

Visuospatial Processing in WS

As mentioned above, tasks requiring visuospatial processing and mental rotation are disproportionately impaired in individuals with WS relative to other cognitive functions and relative to typically developing children (Bellugi, Korenberg, & Klima, 2001; Doyle et al., 2004; Farran et al., 2001; Järvinen-pasley et al., 2008; Mervis et al., 1999). One well-established explanation for this consistent finding is hierarchical in nature, and suggests that there are two levels involved in processing complex visual stimuli: a local level, which includes processing of the pieces or segments of a visual display; and a global level, which involves processing the visual image as a whole. In typical development, children can attend to both levels of a visual display (Stiles, Reilly, Levine, Trauner, & Nass, 2012), though there are mixed findings as to whether children, like adults (Navon, 1977), are more focused on the global level. Some investigations using hierarchical form stimuli suggest that children do indeed show a strong global precedence effect (e.g. Cassia, Simion, Milani, & Umiltà, 2002; Mondloch, Geldart, Maurer, & de Schonen, 2003; Porporino, Shore, Lorocci, & Burack, 2004). Others, however, find evidence that the global processing preference is much more fragile in children, and increases in task demands (Harrison & Stiles, 2009) and degrading the global-level information (Dukette & Stiles, 1996) will readily shift a child's attention to the local level. There is consensus, however, that typically developing children have the capability to attend to both local and global levels

from an early age (Stiles et al., 2012). Unlike typically developing children, some investigations suggest that the visuospatial deficit consistently observed in children with WS can be attributed to a local processing bias, and an inability to integrate the global level. Much of the support for this hypothesis in WS comes from a drawing task, in which children with WS are asked to draw a pattern with a local and global level (e.g. a large square composed of small circles). Using this task, it was found that the children with WS were successful in drawing the local shapes (i.e. drawing the small circles), but did not orient the local level into a coherent whole (i.e. failed to organize the small circles into the shape of a square) (Bellugi, Bihrlé, Neville, Jernigan, & Doherty, 1992; Bertrand, Mervis, & Eisenberg, 1997; Rossen et al., 1996). Since drawing is a delayed skill in WS (Bertrand & Mervis, 1996), however, it is unclear from these studies whether the local processing delays are directly related to a visuospatial processing bias or to a drawing (motor) deficit.

To further investigate the nature of visuospatial processing in WS, Farran and colleagues (2001) investigated visuospatial processing in WS relative to mental age-matched individuals, using an embedded figures task, a mental rotation task, and the Block Design task from the Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999), which is a well-established hierarchical forms visuospatial task that is not confounded with drawing ability. Results from this extensive investigation demonstrated a visuospatial deficit in WS across all three tasks, but did not find evidence for a specific local-processing bias. Specifically, Farran and colleagues (2001) used a modified Block Design Task to directly examine the role of the local processing level. They presented two conditions to participants: In the first, the squares were presented contiguously (Non-segmented Condition) so that the each individual block (the local level) was less salient than the overall pattern created by the smaller blocks; In the second, the pattern was assembled with spaces between the individual blocks (Segmented Condition) so that the individual squares were emphasized. If children were already processing at a local level, this

segmentation would not change the processing strategy, and consequently would not change accuracy or response time (RT) to replicate the pattern. If, on the other hand, children were processing at a more global level, the increased salience of the smaller blocks caused by the segmentation would facilitate performance. This investigation found that even though performance in the WS group was significantly worse than the TD group in both Segmented and Non-segmented conditions, children with WS and their age-matched controls demonstrated a similar facilitative segmentation effect. This study suggests that while individuals with WS demonstrate inferior visuospatial processing relative to children of the same intellectual age, this deficit cannot be explained by a local processing bias.

Further evidence that individuals with WS do not have a specific local processing bias comes from a visual search investigation by Pani, Mervis, and Robinson (1999). In this task, individuals with WS and typical adults were asked to identify a target (e.g. the letter *T*) within an array of distracters. The distracters were manipulated into several conditions: global orientation (distracters were arranged into the shape of a contrasting letter, e.g. the letter *X*); number of distracters; and density of distracters. If participants were attending to the global level, they would demonstrate an interference effect from the contrasting letter in the global processing level. If, on the other hand, participants were focused on the local processing level, they would not show such an interference effect. Results demonstrated that individuals with WS took twice as long to identify the target as the typical group on each of the manipulations, but the participants with WS showed more interference from the global distracter than the typical group. This result again suggests that although individuals with WS show significantly delayed visual search patterns, they are indeed sensitive to the global processing level, and in fact show difficulty disengaging from this global level to focus on the local processing necessary to identify the target.

In sum, it has been observed again and again that individuals with WS are disproportionately impaired on tasks requiring visuospatial processing. Here we have reviewed investigations using drawing measures, block design measures, and visual search measures. Across tasks the WS group performs below expectation based both on chronological and mental age. However, the nature of this deficit is still unclear. There remains debate as to whether this visuospatial deficit is specific to processing the global level, the local level, or perhaps an inability to switch between processing levels (Mervis et al., 1999). Identifying the nature of this cognitive deficit will help us to better understand how this impairment in visuospatial processing may be impacting other cognitive and academic functions such as reading.

Language in HFA

The language of children with HFA is most often characterized by abnormal pragmatics (Tager-Flusberg, 1981, 1996). Specifically, children with HFA are severely limited in their conversational use of language (Loveland & Tunali, 1993), their range of speech acts (Loveland, Landry, Hughes, Hall, & McEvoy, 1988; Wetherby, 1986), their interpretation of interactions (Happé, 1994), and their narrative skills (Nuske & Bavin, 2011). However, in addition to these more apparent pragmatic difficulties, children with HFA demonstrate strengths and weaknesses in other facets of language such as phonology, morphology, and most important for the present discussion, semantics (for reviews, see Boucher, 2012; Groen et al., 2008; Tager-Flusberg, 1999, 2007).

Results from studies investigating phonological processing in HFA have been mixed; some report that phonological processing and articulation are within the normal range (Rapin & Dunn, 2003), yet others find that children with HFA show impaired phonological processing relative to typically developing children, even when IQ is controlled (Hooper, Poon, Marcus, & Fine, 2006). Children with HFA also make more articulation errors than typically developing children during expressive language tasks (Cleland, Gibbon, Peppé, O'Hare, & Rutherford, 2010;

Shriberg et al., 2001), and when repeating nonsense syllables (Kjelgaard & Tager-Flusberg, 2001). Together these results indicate degraded phonological processing, which may have a negative impact on higher-level language facets, such as semantics, pragmatics, and narratives.

Morphological processing has been described as a strength in HFA, with the majority of studies finding no significant differences in morphological errors between typically developing children and those with HFA (Howlin, 1985). One study that does report impairment in morphological processing (Eigsti & Bennetto, 2009), also reports an interaction between morphological processing and sentence length, suggesting that the differences between HFA and TD may not have been due to morphological processing per se, but rather to differences in working memory load. In sum, there is some evidence to suggest a subtle deficit in phonological processing, and more typical morphological processing. Importantly, phonological representations are closely tied to semantic representations, especially during development, when lexical representations may be more closely related to the phonological information than abstract semantic information (e.g. Cronin, 2002; Dewhurst & Robinson, 2004; Rayner, 1988). Thus, it is not surprising that semantic deficits are consistently observed in individuals with ASD.

Over four decades ago, Hermelin and O'Connor, (1967, 1970) postulated that childhood autism disorder was due to impaired semantic representations, and supported this claim with evidence from a semantic memory task. While children with autism recalled the same number of items as their typically developing peers, they did not organize the items by semantic category, suggesting weak semantic relations between words. Since this time, dozens of investigations have extended this hypothesis, and have endeavored to determine which aspects of semantic processing are impaired in children with HFA using both behavioral and neurophysiological methods (e.g. Dunn, Vaughan, Kreuzer, & Kurtzberg, 2000; Dunn & Bates, 2005; Fishman, Yam, Bellugi, Lincoln, & Mills, 2008; Henderson, Clarke, & Snowling, 2011; McCleery et al., 2010; Norbury, Griffiths, & Nation, 2010). Taken together, these investigations suggest that while

language assessments that probe ‘simple’ measures of language (such as vocabulary and cued phonological recall) tend to show typical performance in HFA, assessments of the more ‘complex’ language skills (such as identifying relations between words, understanding category membership, and integrating semantic information into a sentential context) tend to reveal deficits (Minshew & Goldstein, 1995; Williams, Goldstein, & Minshew, 2006). These results are in accordance with the Weak Central Coherence account (WCC) (Happé & Frith, 2006), which proposes that deficits in cognitive functioning can be attributed to a local processing bias, as is observed in visuospatial processing, discussed below.

Visuospatial Processing in HFA

Individuals with autism show enhanced visual processing in a myriad of different tasks including long exposure hierarchical tasks such as Block Design tasks (discussed above) (Caron, Mottron, Berthiaume, & Dawson, 2006; Shah & Frith, 1993), as well as short exposure tasks, which tap more automatic visuoperceptual processes, such as feature and target detection tasks (Jolliffe & Baron-Cohen, 1999; Jolliffe & Baron-Cohen, 2001; O’Riordan, Plaisted, Driver, & Baron-Cohen, 2001; O’Riordan, 2004). While superior perceptual processing performance in autism is a well-established finding, there remains controversy as to the mechanism underlying this enhanced cognitive skill.

The Enhanced Perceptual Functioning model (EPF) (Mottron, Dawson, Soulières, Hubert, & Burack, 2006) and the Weak Central Coherence model (WCC) (introduced above) (Happé & Frith, 2006) are two processing approaches which offer explanations for the enhanced visuospatial processing performance observed in individuals with autism. The EPF suggests that individuals with autism perform better than their typical peers on measures of visuospatial processing because they possess enhanced perceptual processing systems, which lead to superior memory for low-level visual and auditory information (for a meta analysis, see Samson et al., 2012). Importantly, this model does not assume that the enhanced local processing comes at a

cost to more integrative processing, and predicts increased local processing and *typical* global-level processing. The WCC, by contrast, proposes that the enhanced visuoperceptual processing in local-level processing tasks is a direct result of a specific global or integrative processing *deficit*. That is, the WCC views enhanced local processing as a compensatory mechanism in the context of degraded global integration. Thus, in hierarchical visuospatial processing tasks, the EPF would predict performance on the global level to be typical, and performance on the local level to be enhanced in autistic individuals relative to typically developing controls. The WCC, by contrast, would predict enhanced local-level processing but degraded global processing.

In sum, while it is clear that individuals with HFA do show enhanced low-level visuospatial processing, it is unclear whether this advantage comes at a cost to more global integrative functions, or whether visuospatial processing exists as a true island of strength in this population. As written words are themselves an example of a hierarchical visuospatial processing task—the individual letters comprising the local level and the word as a whole comprising the global level—a better understanding of how children with HFA process both levels is critical to understanding and fostering reading acquisition and development in this population.

Reading in WS and HFA

Reading requires both language and visuospatial processing. Comprehension is impossible without both the visuospatial processes required to recognize letters and words, as well as the language-based semantic processes required to understand the word's meaning. As discussed above, children with WS demonstrate impaired visuospatial processing skills, but relatively spared semantic processing skills, and children with HFA demonstrate the reverse pattern. Not surprisingly, then, reading performance in both populations is impaired, though not equally, and not in the same manner. The subsequent sections will review research on reading performance in WS and HFA, focusing on orthographic and semantic processes.

Reading in WS

Pagon, Bennett, LaVeck, Stewart, and Johnson (1987) describe reading as the strongest academic skill for individuals with WS. This assertion was prompted by the finding that nine individuals with WS (between the ages of 10 and 20) exceeded their full-scale IQ score on the reading section of a standardized achievement test. It should be noted, however, that while reading performance was indeed stronger than expected based on full-scale IQ, performance was severely impaired relative to chronological age. Howlin, Davies, and Udwin (1998) investigated reading performance in a large sample of individuals with WS using the Wechsler Objective Reading Dimensions (WORD) test (Rust, Golombok, & Trickey, 1992), and reported opposing results. In this investigation, there was no evidence for an isolated strength; rather, reading performance in WS was found to be quite low. While these mixed results provide general information regarding reading attainment levels for individuals with WS, they do not address the underlying cause(s) of reading failure, nor do they address the variability in reading achievement in WS (Laing et al., 2001).

In order to better understand the nature of the reading impairment, Barca, Bello, Volterra, and Burani (2009) examined the reading of a thirteen-year-old girl with WS with regard to lexicality, frequency, length, and contextuality. The authors found evidence for a lexicality effect (lower accuracy when reading non-words compared to words) and a frequency effect (lower accuracy when reading low frequency words compared to high frequency words), but no effect of contextuality (words that cohere and deviate from established orthographic-phonological rules were read with similar accuracy) or length (long and short words were read with similar accuracy). Since the two factors (lexicality and frequency) that influenced reading are lexical in nature (i.e. are dependent on the meaning of the word), and the two factors (contextuality and length) that did not affect reading were orthographic or phonological in nature, these findings suggest that this individual with WS accessed some of the word's meaning before performing an

orthographic-phonological translation. A possible explanation is a weakness in either orthographic or phonological processing.

According to the dual route model by Coltheart, Rastle, Perry, Langdon, and Ziegler (2001), there are two separate paths used to access meaning from a written word. For high frequency, short, or well-known words, a “direct” path is used, whereby the meaning is accessed directly from the orthography. For words that are less common, or longer, an “indirect route” or phonologically mediated route is taken; in this case, the orthography is mapped onto a phonological representation, and the meaning is derived from the phonology. In typical reading acquisition, where initially very few words are familiar, children tend to utilize the indirect route; it is not until later when children have developed a more extensive and familiar lexicon that they begin to utilize the direct route. Results from Barca and colleagues (2009), suggest that individuals with WS may develop a deviant pattern of reading, in which they draw on their relatively strong semantic system, rather than the weaker orthographic and phonological systems (Menghini, Verucci, & Vicari, 2004).

Further evidence that children with WS rely heavily on their stronger, semantic system when learning to read, comes from a report of “deep dyslexia” in another case study of a 13-year-old girl with WS (Temple, 2003). Deep dyslexia was first described by Marshall and Newcombe (1973), and is characterized, in part, by semantic errors made in single word reading tasks. For example, an individual with deep dyslexia might read the word *truck* as ‘*car*,’ demonstrating an understanding of the meaning of the word, but an inability to quickly access the phonological information to correctly read the word aloud. Such errors provide strong evidence for the use of a semantic route to reading. Temple’s (2003) case study reports that the words correctly read aloud by the individual with WS were most often high in frequency and imageability, and also that a large percentage of errors were semantic in nature. This pattern is consistent with a profile of deep dyslexia. Consequently, these results provide additional evidence that individuals with WS

may rely on the semantic system (rather than an orthographic-phonological system) when learning to read.

However, not all findings support this claim. Laing and colleagues (2001) used a paired associate reading paradigm in which real words that varied in imageability were paired with non-words that varied with respect to their phonological similarity to the word. They found that children with WS were only sensitive to the phonological manipulation, but did not show an imageability effect, unlike their typically developing peers. This result suggests that children with WS depend *less* on semantic processes relative to phonological processes. However, Mervis (2009) notes that not all studies of typically developing children have found this semantic effect using this paradigm (e.g. McKague, Pratt, & Johnston, 2001), and she highlights the importance of further research into the relative importance of semantic processing in children with WS when reading single words.

Reading in HFA

There is a general belief that reading is a particular strength for children with HFA. However, as Nation, Clarke, Wright, and Williams (2006) point out, there is little conclusive empirical evidence to support this assumption. Indeed since language delay or disorder is one of the diagnostic criteria for autism, as well as a recognized risk factor for reading impairment (Bishop & Snowling, 2004; Catts, Fey, Zhang, & Tomblin, 2001), it would be especially surprising if children with HFA demonstrated superior reading performance. One possible reason that individuals with autism are thought to show exceptional reading ability is that much of the early research investigated phonological decoding in cases of hyperlexic children with autism (e.g. Aaron, Frantz, & Manges, 1990). While these studies provide insight into decoding abilities of this subgroup, the results are not necessarily representative of the population, or of the reading process as a whole. In the following, we provide a brief review of reading in HFA with an emphasis on the apparently opposing profile to that of children with WS.

Contrary to children with WS, Children with HFA are often successful in reading words aloud, yet less successful in accessing their meaning (Burd & Kerbeshian, 1985; Frith & Snowling, 1983; Grigorenko et al., 2002; Happé, Booth, Charlton, & Hughes, 2006; Minshew, 1994; Nation et al., 2006; Newman et al., 2007; Wahlberg & Magliano, 2004). However, there are inconsistent findings with regard to reading non-words. Some find that non-words and words are read similarly (Frith & Snowling, 1983; Minshew, 1994), yet others describe reading single words as superior to reading non-words (Aaron et al., 1990; and Nation et al., 2006 in a more diverse group of children with ASD). Word-reading that is better than non-word reading suggests that the proficient decoding observed in children with autism may be supported by rote memorization of the shape of the word rather than dependence on phonological decoding. This finding of an enhanced ability to memorize the orthographic shape of a word is in accord with the enhanced visuospatial processing skills discussed above. If indeed individuals with HFA depend more on the orthographic shape of a word than typically developing children, it is likely that they utilize the direct pathway of the Dual Coding Route (Coltheart et al., 2001) (orthography to semantics), without passing through a phonological decoding process. However, it is unclear how finely tuned the orthographic processing system is, and whether or not children with HFA would be confused by orthographic neighbors of a target word.

According to the lexical tuning hypothesis (Castles, Davis, Cavalot, & Forster, 2007), children initially have very coarsely-tuned orthographic processing systems, as it is unlikely early-on that many of a word's orthographic neighbors will be included in a child's lexicon. It is thus an efficient strategy to maintain loose criteria for word identification (e.g. *big* and *bid* may be accepted as the same word, since the child knows the word *big* and not *bid*). As the size of the lexicon grows with more language and reading experience, the child must adopt more stringent word recognition criteria, which require the development of a more finely tuned orthographic processing system. It is unclear whether or not children with HFA, who have enhanced

visuospatial processing, but relatively poor lexical-semantic repertoires, will also pass through such a *lexical-tuning* process.

To test this, Speirs, Yelland, Rinehart, and Tonge (2011) used a masked priming paradigm which contrasted identical orthographic forms (e.g. blue: BLUE), similar orthographic forms (e.g. blog: BLUE), phonological primes (e.g. blew: BLUE), and orthographically dissimilar primes (e.g. sand: BLUE). They found that both typically developing children and children with HFA demonstrated a significant priming effect to the orthographically identical prime, suggesting some similarity in automatic lexical access. There was also some evidence, however, that the children with HFA were also priming to the orthographically similar prime, perhaps suggesting a more coarsely tuned lexical processing system. The authors note, however, that there is little evidence for this explanation, as the vocabularies and spoken language measures did not differ between groups. As such, it appears that while the automatic lexical processing system is strong, it may also be functioning in a qualitatively different manner, a topic that requires further exploration. In the present study, we will further investigate orthographic processing in HFA using a synonym recognition task in the context of semantically and orthographically similar response alternatives. In this way, we will extend the results of Speirs and colleagues (2011) to include a measure of semantic processing in addition to a measure of the orthographic tuning of the lexical system. Given the high proportion of children with autism who struggle with reading comprehension (often in the presence of normal word reading), Nation and colleagues (2006) note that it is important that future research pinpoint the *specific components* of reading that are impaired in HFA. In the present study, we aim to address this important question by looking at both orthographic and semantic processing of a single written word.

Since semantic processing and visual-spatial, orthographic processing are both key components of reading, unimpaired processing of both is necessary for typical reading development. However, when reading does not develop typically, as in WS and HFA, it is not

clear which component(s) is/are causing the system to fail. By investigating populations of children with contrasting cognitive-linguistic profiles (children with WS demonstrate relatively good semantic processing and poor visuospatial processing and children with HFA demonstrate good visuospatial processing and relatively poor semantic processing), and by isolating semantic and orthographic processing of a single written word, we hope to add to our understanding of reading development in WS and HFA, as well as in typical development. This fine-level analysis of single-word reading in WS and HFA will lead to more targeted reading interventions for these populations, and will also illuminate how orthographic and semantic components of reading function and interact to facilitate typical reading acquisition and development.

METHODS

PARTICIPANTS

Fifty-six children from three populations participated in Experiment 1: Nineteen children with High Functioning Autism (HFA) M age, 9.83; SD = 1.68 years, 10 children with Williams syndrome (WS) M age = 10.86; SD = 1.62 years, and 27 Typically developing children (TD) M age = 9.88; SD = 2.05 years). All participants in all groups were monolingual English speakers, with no exposure to a second language before the age of five, and all had normal (or corrected) visual and auditory acuity. Participants were excluded from the study if they had a prior medical condition that might interfere with test performance (e.g. severe closed head trauma, meningitis, brain tumor).

Children in the TD group had normal developmental and medical histories and normal neurological exams. All scored within the normal range (85 - 115) on standardized tests of intelligence, language, and academic functioning, and none had a history of chronic medication use.

Children in the HFA group met diagnostic criteria for Autism from the *Diagnostic and Statistical Manual of Mental Disorders-IV (DSM-IV)* (APA, 2000) and from the *Autism*

Diagnostic Interview—Revised (ADI-R) (Lord et al., 1997), and/or the *Autism Diagnostic Observation Schedule (ADOS)* (Lord et al., 1989). All demonstrated a PIQ of 70 or above, and none had a specific underlying genetic or metabolic diagnosis (e.g. tuberous sclerosis, Fragile X syndrome, or structural brain malformations).

Children in the WS group met the diagnostic criteria specified by the American Academy of Pediatrics, Committee on Genetics, Health Care Supervision for Children with Williams Syndrome, Pediatrics, 2001, and had a confirmed diagnosis of WS by a medical geneticist, who verified the loss of one copy of the gene for Elastin in chromosome 7q11.23 by FISH test.

STANDARDIZED TESTING BATTERY

All participants received a standardized testing battery to probe intelligence, as well as language and reading at different processing levels including phonology, morphology, vocabulary, and comprehension. This testing battery included the Wechsler Abbreviated Scale of Intelligence (WASI) (Wechsler & Chen, 1999), the Clinical Evaluation of Language Fundamentals- Fourth Edition (CLEF-IV) (Semel, Wiig, & Secord, 2003), three subtests from the Woodcock Johnson Test of Achievement- Third Edition (WJ-3) (Woodcock, McGrew, & Mather, 2001), and the Expressive One Word Picture Vocabulary Test (EOWPVT) (Gardner, 1990).

EXPERIMENTAL READING MEASURE

Materials and Procedure

In this experimental single word reading measure, which was developed by Polse and Reilly (2012), children are presented with a target word and two response alternatives, and are asked to identify the semantic match. Each trial contains a match (the correct response alternative for that trial) and a foil (the incorrect response alternative). The match and foil vary in orthographic and semantic similarity to the target. Word-pairs designated as a match or a foil were normed on 100 undergraduate students who rated the semantic similarity of word-pairs on a Likert scale from 1 (extremely different meanings) to 7 (extremely similar meanings). Only those

pairs that were rated as an average of 5.5 and above ($M = 6.06$; $SD = .33$) were used as a match, and only those that were rated as an average of 1.5 and below ($M = 1.35$; $SD = .83$) were used as a foil. Target, match, and foil stimuli did not differ in length, nor did they differ in word frequency according to the Hyperspace Analogue to Language (HAL) corpus norms (Lund & Burgess, 1996). Paired t -tests reveal that there were no significant differences between the frequency of the match and foil, according to the HAL frequency corpus $t(25) = .326$, $p > .1$. Match, Foil, and Target words were all between three and six letters, and match and foil were not significantly different in terms of word length (number of letters) $t(25) = 1.9$, $p > .05$.

Target, match, and foil were presented together on a touch screen monitor. The target word was centered at the top of the screen and the foil and match were presented side-by-side below the target. For salience, all words were presented in black font within white rectangles on a black screen. Words were presented in capital letters in size 40 Arial Font (See Figure 2.1 for schematic of experimental paradigm). The side of the match on the visual display was pseudorandomized such that each participant saw an equal number of match and foil words on the right and left sides of the screen, but the location of the correct response alternative was not predictable. Each child responded to 26 trials that met norming criteria. Four lists were created containing six trials of two conditions and seven trials of the remaining two conditions, which then were counterbalanced across participants so that an equal number of each condition was presented across participants, and children did not see any word more than once.

Participants were tested individually at a testing center as part of a larger research project. Children were seated in front of a touch screen monitor and were instructed to use one finger to touch the word that “means the same thing” as the target word. Children were told explicitly that, *means the same thing* could be the exact same word (e.g. BOAT-BOAT), or a synonym of that word (e.g. BOAT-SHIP). All participants completed a 10-trial practice block with accuracy feedback to ensure understanding of the procedure. Participants were not given feedback during

the experimental trials. Although accuracy was emphasized, children were asked to answer as quickly as possible. The stimuli remained on the screen until the child made his or her response by touching one of the response words. To ensure that the child was attending to the task when the words were presented, children were responsible for advancing the experiment to the subsequent trial. Following each trial, an interim screen appeared, which instructed the child to touch the screen when he/she was ready to proceed.

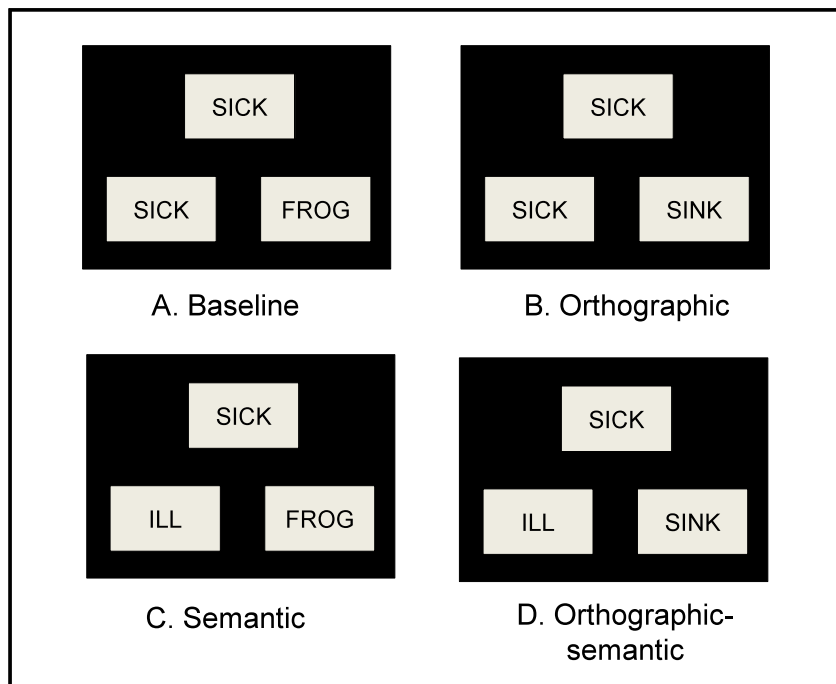


Figure 2.1: Displays the four experimental conditions. Each box represents the screen for one trial. Condition A contains an orthographic match and an unrelated foil, condition B contains an orthographic match and an orthographic neighbor as a foil, condition C contains a semantic match, and an unrelated foil, and condition D contains a semantic match and an orthographic neighbor as a foil.

Experimental Conditions

Four conditions were presented which varied in terms of orthographic and semantic similarity to the target. The following will describe the conditions in brief, but for an in depth description of the experimental conditions, see Polse and Reilly (2012).

Baseline condition: In the Baseline condition, the match is orthographically and semantically identical to the target (same word), and there is no orthographic foil (e.g. LADY

(target); LADY (match), WATER (foil)). This condition requires perceptual matching of word shape, but no fine-level orthographic processing or semantic processing.

Orthographic Condition: In the Orthographic condition, the match is orthographically and semantically identical to the target (same word), and the foil is an orthographic neighbor of the target (differs by only one letter) (e.g. LADY (target); LADY (match); LAZY (foil)). This condition requires more fine-tuned orthographic processing, but also does not require semantic processing of the word.

Semantic condition: In the Semantic condition, the match is a synonym of the target and the foil is orthographically unrelated to the target (e.g. LADY (target); WOMAN (match); WATER (foil)). This condition requires semantic processing but does not present a closely related orthographic foil.

Orthographic-semantic condition: In the Orthographic-semantic condition, the match is a synonym of the target and the foil is an orthographic-neighbor of the target (e.g. LADY (target); WOMAN (match); LAZY (foil)). In this condition, orthographic and semantic processing are in direct competition.

RESULTS

STANDARDIZED TESTING

Appendix A, Table 2.1 summarizes results from the standardized testing battery. It is clear from this table that performance from both clinical groups is below that of the typically developing children. Furthermore, performance from the WS group is consistently worse than the group with HFA. The ages of the participants, however, are not significantly different across groups.

EXPERIMENTAL READING MEASURE

Accuracy data were analyzed using a 3 (Population) x 4 (Experimental Condition) univariate ANOVA with percent correct as the dependent variable (See Figure 2). This analysis

revealed a significant main effect of Population $F(2, 212) = 26.73, p < .001, \eta^2 = 1.00$, and Condition $F(3, 212) = 4.823, p < .005, \eta^2 = .90$, and no significant Population x Condition interaction $F(6, 212) = 1.05, p > .1$. Follow-up Least Significant Difference (LSD) comparisons demonstrate that children in the TD group performed with higher accuracy than the HFA group ($p = .05$) and the WS group ($p < .005$), and that the children with HFA performed overall with higher accuracy than the children with WS ($p < .005$). However, these analyses do not allow for comparisons between populations for each of the four experimental conditions.

To derive these comparisons, we performed hypothesis-driven one-way ANOVAs for each of the four conditions. There was a main effect of Population for each condition: Baseline $F(2, 53) = 16.93, p < .005$; Orthographic $F(2, 53) = 9.20, p < .005$; Semantic $F(2, 53) = 5.35, p < .01$; Orthographic-semantic $F(2, 53) = 3.94, p < .05$. Post Hoc paired-samples t -tests are reported below.

Baseline Accuracy: No significant difference was observed between the TD and HFA groups, the TD group performed with higher accuracy than the WS group ($p < .05$), and the HFA group performed with higher accuracy than the WS group ($p < .05$). Orthographic Accuracy: No significant differences were observed between the TD and the HFA groups, the TD group performed with higher accuracy than the WS group ($p < .05$), and the HFA group performed with higher accuracy than the WS group ($p < .05$). Semantic Accuracy: The TD group performed with higher accuracy than the HFA group ($p = .08$; marginal significance), and there was no significant difference in accuracy in the TD and WS groups ($p = .10$), nor between the HFA and WS groups ($p > .1$). Orthographic-semantic Accuracy: No significant differences were observed between the TD group and the HFA group ($p > .1$), the TD and WS group ($p = .10$), nor the HFA and WS groups $p > .1$).

Lastly, to test our hypothesis that the individuals with WS would perform better on items probing semantic processing relative to orthographic processing, and that children with HFA

would perform better on items probing orthographic processing relative to semantic processing, we performed planned comparison paired t -tests within population and between conditions. These planned comparisons revealed that in the TD group there was no significant difference in accuracy between the Orthographic and Semantic conditions ($t(25) < .1$), but performance to the Orthographic-semantic condition was significantly worse than performance to the Semantic condition ($t(25) = 3.164, p < .01$). In the HFA group, the children responded with significantly higher accuracy to the Orthographic condition relative to the Semantic condition $t(18) = 2.265, p < .05$, but there was no difference between the Semantic and Orthographic-semantic conditions ($t(18) = 1.482, p > .1$). In the WS group, there was no significant difference in accuracy between the Orthographic and Semantic conditions ($t(9) = 1.4, p > .1$), nor between the Semantic and Orthographic-semantic conditions ($t(9) = 1.515, p > .1$).

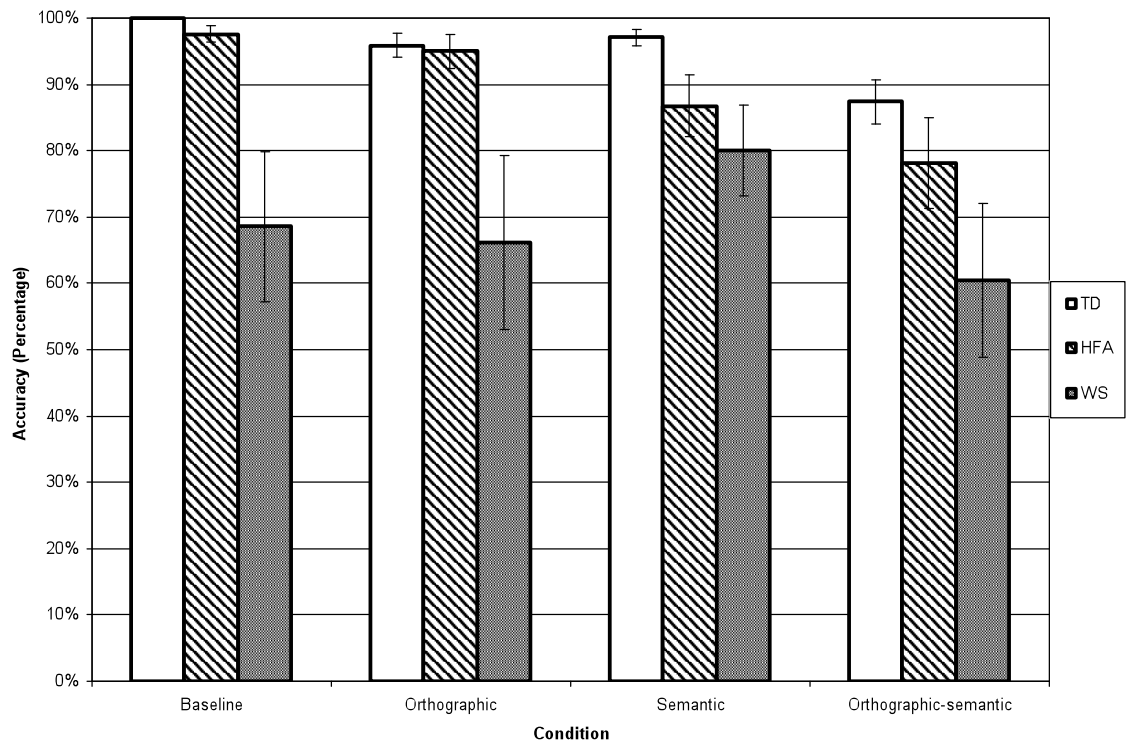


Figure 2.2: Accuracy results from all four conditions. Results displayed with standard error bars (TD $n = 27$, HFA $n = 19$, WS $n = 10$).

DISCUSSION

We now return to our first two questions posed in the Introduction. *How are deficits observed in the language and/or cognitive profiles of children with neurodevelopmental disorders reflected in reading performance in the school-age years?* and *How does this reading profile compare to typical reading development?* Evidence from the current investigation of children with WS and children with HFA suggests that predicted cognitive strengths and weaknesses are observed in single word reading, thus extending the contrastive visuospatial – linguistic cognitive profiles of WS and HFA to a reading context.

In the WS group, we found that accuracy performance was significantly worse than the TD and HFA groups on conditions requiring only orthographic processing (Baseline and Orthographic). On conditions requiring semantic processing (Semantic and Orthographic-semantic), however, the children with WS did not differ significantly from the HFA and TD groups. This result is quite striking given the intellectual impairment in WS, which makes it rare that children with WS perform on par with their chronologically age-matched peers on any measure of cognition. Thus, the finding that the semantic conditions of the present experiment do not differ from typically developing chronologically age-matched controls provides strong evidence that semantic processing is a relative strength for individuals in this population. It further suggests that children with WS may rely more heavily on semantic processing (relative to orthographic or phonological processing) when reading single words.

It should be noted, however, that there was more variability in the WS group compared to the HFA and TD groups, as well as fewer participants, due to the rarity of WS. Thus, there is the possibility that the non-significant differences may be partially driven by a smaller WS sample and large variability within the WS group. Importantly, however, with this same sample, there were significant differences between the TD and WS groups in conditions requiring orthographic processing, indicating that there is indeed a difference between orthographic and semantic

processes in WS relative to typical developing children. Because WS is such a rare genetic disorder, and acquiring large sample sizes is difficult, these results should be replicated to confirm this finding. Nonetheless we see this as initial evidence that children with WS rely more on semantic information than orthographic information in single-word reading contexts.

The heavy reliance on semantic processing that we observed in the WS population is not typical of early reading in typical development, and suggests a deviant, rather than delayed, reading profile in WS. In typical reading acquisition, there is evidence to suggest that children initially rely on the orthographic decoding system, and slightly later begin to activate meaning directly from the text (Polse & Reilly, 2012). Therefore, if children with WS performed similarly to the TD group on measures of the earlier-acquired, orthographic processing, this would have signaled a language delay. On the contrary, in the present investigation, children with WS identified a synonym (semantic processing) with similar accuracy as their TD peers, but fell below when matching closely related orthographic symbols. This pattern thus indicates an aberrant reading profile, which is more dependent on the meaning of the word than an orthographic-phonological translation. These results are in accordance with descriptions of deep dyslexia in WS (Temple, 2003).

In the HFA group, consistent with our hypotheses, we found that children with HFA performed similarly to their TD peers (and better than children with WS) on conditions requiring orthographic processing; this result suggests that the superior visuospatial performance described in other cognitive contexts extends to a reading domain. Also consistent with our hypotheses, children with HFA performed worse than the TD group on the Semantic condition, in which they were required to identify a synonym match without the presence of an orthographic foil (though this difference was marginal). When viewed within a local-global context, this result suggests that children with HFA performed better on the local level (matching the orthography) than the global level (accessing the meaning). This is consistent with both Weak Central Coherence (WCC)

(Happé & Frith, 2006) and Enhanced Perceptual Functioning (EPF) (Motttron et al., 2006) frameworks. Contrary to our predictions, however, children with HFA performed as well as the TD group on the Orthographic-semantic condition. We had predicted that children with HFA would fall below their TD peers on the Orthographic-semantic condition, as they did on the Semantic condition (with no orthographic foil). It is possible that the HFA group's proficient performance to the semantic information, even in the presence of contrasting orthographic information, suggests intact global-level processing, consistent with the EPF model (though contrasting with the WCC, which would predict degraded performance to the global level). However, this account does not explain their marginally inferior performance on the Semantic condition, and thus we feel other possible explanations for this surprising finding are warranted.

First, it is possible that the superior visuospatial performance of children with HFA allowed them to identify that the orthographic neighbor differed from the target in the Orthographic-semantic condition, and to answer correctly using a logical deduction strategy. In this case, the children with HFA may have correctly responded to the Orthographic-semantic condition trials without accessing the semantic information in the synonym. Rather, they identified the difference in the orthographic neighbor foil, and selected the alternate response option. If this explanation were correct, it would indicate that children with HFA have finely tuned orthographic processing skills in the presence of weak semantic representations. This would indicate that unlike typically developing children, the lexical tuning hypothesis (Castles et al., 2007) is not applicable to reading development in HFA. Such a conclusion suggests a deviant pattern of reading in HFA relative to TD. Specifically, the difference between typical single-word reading and single-word reading in HFA may reside in the use of semantic representations. In typical development, children use the meaning of the word to enhance orthographic decoding. In HFA, children may rely more heavily on the orthographic symbols without activating the meaning of the target word or its orthographic neighbors.

Alternatively, the lack of difference between TD and HFA in the Orthographic-semantic condition may have been driven by the decrease in accuracy performance in the TD group (relative to the Semantic condition), rather than enhanced performance on the Orthographic-semantic condition (relative to the Semantic condition) in the HFA population. That is, while it appears that the HFA group performed better on the Orthographic-semantic condition relative to the Semantic condition, accuracy from the two conditions do not differ from one another within the HFA group. On the other hand, children in the TD group performed significantly worse on the Orthographic-semantic condition compared to the Semantic condition, suggesting that the lack of difference between TD and HFA on the Orthographic-semantic condition may be driven by the decreased performance in the TD group rather than improved performance in the HFA group *per se*.

The finding that the TD group performed worse on the Orthographic-semantic condition relative to the Semantic condition is concordant with the lexical tuning hypothesis (Castles, et al., 2007), and suggests that their orthographic processing is still somewhat coarsely tuned. That is, children in the TD group activated the semantic match from the orthographic neighbor foil, and thus answered incorrectly. It is apparent from their superior performance in the Semantic condition that the children in the TD group knew the synonyms, and could answer correctly, however they were fooled by the orthographic foil.

We now step back a moment and recall that both children with WS and children with HFA struggle with reading: In general, children with HFA struggle with comprehension and children with WS struggle reading words aloud. This study provides a fine-grained analysis on the components required to read and understand a word in these two populations. However, the process of reading encompasses much more than identifying single words. Single words must be integrated into sentences, and sentences into paragraphs. Children must acquire the skills to make long-distance connections between ideas in a passage and retain that information in memory.

Thus, reading is a rich and complex process that cannot be encapsulated within a single-word paradigm design. In this study, we have dissected the complex process of reading into fundamental building blocks, and investigated orthographic and semantic processing isolated in a single word paradigm. We view identification of strengths and weaknesses at this micro-level of analysis as an initial step. In future research, we ask the question, How will strengths and weaknesses at the word level be reflected in the sentence, paragraph, and narrative levels?

In typical development, levels of language and literacy may build on one another to create complex, robust language systems that support one another. Perhaps these relationships differ in children with neurodevelopmental disorders. How will cognitive strengths and weaknesses manifest when language and literacy become more complex? In beginning with component features of a single written word, we hope to better understand which specific elements of reading are causing children with WS and HFA to fail, and to add to a better understanding of the relations between different levels of language and literacy (e.g. word, sentence, paragraph, narrative). From this knowledge, it will be possible to create more targeted reading curricula and interventions for children with HFA and WS, and to use the strengths of each population to bolster their weaknesses.

Successful reading is in and of itself a manifestation of componential integration. In typical development, however, when the components underlying reading co-develop, it is almost impossible to isolate the unique contribution each component makes to the complex process of reading. This leads us to our third and final question, What can this investigation tell us about the relations of orthographic and semantic components underlying reading in TD? The contrasting visuospatial-linguistic profiles of WS and HFA, along with their contrasting single-word reading profiles, provide a double dissociation between cognitive deficit (visuospatial: WS and semantic: HFA) and degraded task performance (orthographic: WS and synonym: HFA), which allows us to isolate the independent contributions of orthographic and semantic components of reading, and

form hypotheses as to the role of these components in typical reading development. Because these components can be isolated in children with neurodevelopmental disorders, this study suggests that orthographic and semantic processes make independent contributions to the reading process. Thus, integrating these two separable components is a necessary element in typical reading acquisition and development.

In conclusion, this study investigated how the cognitive strengths and weaknesses observed in WS and HFA are reflected in a single word reading context. Visuospatial processing is a weakness for children with WS, whereas semantic processing has been described as a relative strength. Children with HFA show enhanced visuospatial processing, but a relative deficit in semantic processing. In the present investigation, we tested orthographic and semantic components of reading, and found evidence that this general cognitive profile of WS and HFA extends to single word reading. We found that children with WS demonstrated performance similar to their typically developing peers on some trials that required semantic processing, but fell below on all items that required fine-grained orthographic processing. Children with HFA showed performance similar to their TD peers on items probing orthographic processing, but not on all semantic conditions. Overall, results from this study reveal the distinctive reading profiles of these two neurodevelopmental disorders which have implications for the design and implementation of more targeted reading interventions, and also add to our understanding of the components underlying typical reading development.

APPENDIX A

Table 2.1: Standardized Testing Results

Test	Measure	TD		HFA		WS		Main Effect <i>F</i> -stat (<i>p</i> -value)	TD-HFA ¹ sig.	TD-WS ¹ sig.	HFA-WS ¹ sig.
	Age (CA)	9.88	2.05	9.84	1.68	10.86	1.62	1.18 (<i>p</i> > .1)	ns	ns	ns
WASI ²	PIQ	105.58	11.02	91.05	21.62	65.00	4.19	26.92 (<i>p</i> < .001)	.007	.000	.000
	VIQ	116.73	13.37	93.53	18.85	73.10	10.21	33.64 (<i>p</i> < .001)	.000	.000	.003
	FSIQ	112.65	11.32	92.21	17.34	66.80	6.78	46.49 (<i>p</i> < .001)	.000	.000	.000
	Matrix Reasoning	55.00	8.81	40.44	12.15	24.5	4.06	36.33 (<i>p</i> < .001)	.000	.000	.000
	Similarities	60.58	8.10	48.50	12.04	34.10	8.06	29.00 (<i>p</i> < .001)	.000	.000	.001
	Block Design	52.04	7.74	44.33	13.81	28.80	2.53	21.73 (<i>p</i> < .001)	.032	.000	.000
	Vocabulary	59.42	8.57	40.11	13.77	28.50	7.35	38.10 (<i>p</i> < .001)	.000	.000	.020
EOWPVT ³	Vocabulary	113.08	12.76	94.95	21.63	73.20	11.62	22.44 (<i>p</i> < .001)	.002	.000	.004
CELF-IV ⁴	Core Language	106.92	9.46	77.32	21.85	59.40	15.32	38.23 (<i>p</i> < .001)	.000	.000	.017
	Expressive Language	108.04	10.04	79.94	22.15	63.20	13.38	34.16 (<i>p</i> < .001)	.000	.000	.028
	Receptive Language	106.04	10.77	81.58	21.27	60.80	12.00	33.92 (<i>p</i> < .001)	.000	.000	.003

Table 2.1 (continued)

Test	Measure	TD		HFA		WS		Main Effect <i>F</i> -stat (<i>p</i> -value)	TD-HFA ¹ sig.	TD-WS ¹ sig.	HFA-WS ¹ sig.
		Mean	SD	Mean	SD	Mean	SD				
WJ-III ⁵	Academic Skills	111.75	13.55	92.64	18.29	62	23.83	23.58 (<i>p</i> < .001)	.006	.000	.001
	Letter-word ID	111.81	11.03	98.21	16.27	77.00	17.39	22.09 (<i>p</i> < .001)	.008	.000	.001
	Reading Fluency	107.92	11.41	96.65	17.61	69.44	20.75	20.85 (<i>p</i> < .001)	.070	.000	.000
	Word Attack	107.62	9.72	96.37	11.14	81.70	13.06	21.44 (<i>p</i> < .001)	.004	.000	.003
	Spelling Sounds	107.31	11.44	97.05	13.34	75.80	27.13	14.28 (<i>p</i> < .001)	.111	.000	.004

¹ Bonferroni corrected

² Wechsler Abbreviated Scale of Intelligence (Wechsler & Chen, 1999)

³ Expressive One Word Picture Vocabulary Test-Revised (Gardner, 1990)

⁴ Clinical Evaluation of Language Fundamentals- Fourth Edition (Semel, Wiig, & Secord, 2003)

⁵ Woodcock Johnson Test of Academic Achievement- Third Edition (Woodcock, McGrew, & Mather, 2001)

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CHAPTER 3: RELATIONS AMONG LINGUISTIC SUBSYSTEMS IN HIGH FUNCTIONING CHILDREN WITH AUTISM AND TYPICALLY DEVELOPING CHILDREN

ABSTRACT

In this investigation we consider performance on and relations amongst classically defined linguistic components (phonology, lexico-semantics, and syntax) in spoken and written modalities, and their relation to narrative composition in typically developing children (TD) and High Functioning children with Autism (HFA). While performance on the majority of the linguistic subsystems did not differ between groups, children with HFA performed worse than their typically developing peers in their ability to relate a personal narrative. Correlations results indicate that in the typically developing group, almost all of the linguistic components in both spoken and written modalities were strongly correlated with one another, suggesting a highly integrated and reinforcing language system. The HFA group, by contrast, showed very few significant correlations, and those components that were correlated mostly included the spoken lexico-semantic level. Our results are consistent with a weak integration account of HFA, and suggest that language interventions for these children should target complex, functional aspects of language such as conversation and narratives rather than isolated lower-level linguistic components such as phonological and lexico-semantic processing.

INTRODUCTION

Language is a multifaceted, complex system, and as we use language we simultaneously recruit an array of interrelated subsystems. Linguists, psycholinguists, speech-language pathologists, and others often describe these subsystems as existing in a hierarchical organization comprised of multiple *levels*. Since language use is in and of itself a manifestation of the interrelatedness of linguistic components, drawing boundaries between levels is somewhat arbitrary, and models with varying numbers of levels have been posited ranging from low to high

specificity (e.g. phonology-grammar-semantics versus phonetics-phonology-morphology-morphosyntax-syntax-semantics-discourse-pragmatics). Nonetheless, defining language components has been and continues to be a functional instrument with which to study and discuss language at multiple degrees of complexity. As David Crystal (1987) notes, “But there is nonetheless no doubting the fruitfulness of the basic insight underlying the notion of level—the recognition of simultaneously-operating dimensions of structural organization capable of being analysed in independent terms from those used elsewhere in language study” (pg. 8). Here we use classically defined linguistic components (phonology, lexico-semantics, and syntax) as a tool to investigate not only the levels themselves, but also the relations amongst such levels in both typical and atypical language development. In their Dynamic Systems approach, Thelen and Smith (1994) suggest that each cognitive (or linguistic for the purpose of this discussion) act is a snapshot of the relations amongst multiple dynamic subsystems (which all have their own dynamic relations with other subsystems), and their interaction with the environment at a single point in time. They propose that, “The power source of human cognitive development is not in the separate modules but in their mutual interactions” (Thelen & Smith, 1996, pg. 37). Better understanding the relations between levels of *linguistic complexity* may act as such a “power source” for explaining not only typical development, but also atypical language development. In fact, Crystal (1987) proposed that our preoccupation with language levels and subsystems may have led us to ignore what could be the more central issue in treating and diagnosing language disorders: the interactions between levels. One consistent finding both in research and in clinical language assessments, is the high degree of variability in performance within individual subjects with language disorders. Often, such individual variance is attributed to fatigue, vulnerable linguistic representations, or attention deficits, but it could be the case that what manifests as inconsistent performance at one or multiple linguistic levels actually stems from an integrative deficit amongst linguistic subsystems. From this theoretical perspective, diagnosing a language

disorder may not include labeling a lexico-semantic deficit or a syntactic deficit, but rather would include identifying how lexico-semantic and syntactic processing interact and associate as an individual attempts to use language at increasing levels of complexity. In the present investigation, we raise the questions, *How are the linguistic subsystems related in typical development? What is their role in discourse performance?* And, *What can investigating the relations between linguistic subsystems tell us about atypical language development?*

In typical development, infants begin to babble at about six months of age, and while initially they can discriminate between phonemes that are non-specific to their native language, within the first year of life, the infant can only differentiate between the phonemes that occur in the ambient language (e.g. Werker, Gilbert, Humphrey, & Tees, 1981; Werker & Tees, 2002). This perceptual narrowing of the phonological system is one of the first indications that the infant is acquiring language-specific patterns. At about 9-10 months of age, she responds to words, and around the first year begins to label items with consistent, phonological forms, demonstrating the initial signs of linking a phonological form with a semantic meaning: the word level (Fenson et al., 1993; Fenson, Dale, Reznick, Bates, Thal, & Pethick, 1994). During this first year and a half, the child's expressive vocabulary grows at a steady pace, adding approximately one to three new words per week, until around 18 months (when the child has about 20-40 words in her expressive vocabulary), at which point there is a "vocabulary spurt," where a toddler's productive vocabulary dramatically increases, and she produces approximately 8-10 new words per week. Shortly after this vocabulary spurt, at 20-24 months, toddlers begin linking words together to convey a broader meaning. Although these "sentences" of a two-year-old are telegraphic in nature, the word order typically conforms to the grammatical rules of the ambient language. For example, a child is more likely to use the phrase *'nother cookie* to mean *I want another cookie* than *cookie 'nother*, which is a non-canonical word order in English. This example suggests some immature syntactic knowledge following the acquisition of a quantity of single words. Following

the onset of grammatical word combinations (the sentence-level), three year-olds begin to combine individual predicates into complex sentences, and then in the preschool years, they begin to produce short narratives (For references on first language acquisition, see: Barrett, 1996; Benedict, 1979; Bloom, 1993; Brown, 1973; Fenson, 1993; Fenson et al., 1994; Fletcher & MacWhinney, 1996; Goodluck, 1991; Nelson, 1973; Radford, 1996). During a biographical interview, a four-year-old girl told the following story (Reilly, 1999, unpublished data): “*Um, one day I was at this riding lesson and this horse was being cocky. And he put his head down and I falled down but I did not get hurt because my helmet was on.*” While some aspects of her story are simple, it contains many story elements, including a setting (one day, at a riding lesson), a conflict and response (the horse was being cocky and [the narrator] fell), causal elements (I did not get *hurt because my helmet was on*). Although her story would not be mistaken for the story of an older child or adult, perhaps due to errors in morphology (e.g. falled), this four-year-old child has successfully recruited the linguistic elements to organize, formulate, and articulate a personal narrative. For the purposes of this investigation, we view discourse, specifically this story-telling ability, as a milestone at the summit of the hierarchy of linguistic complexity, as it recruits earlier-acquired and less linguistically complex levels—phonological, lexical, and syntactic—and requires the integration of those skills.

This language development profile for typically developing children suggests that language subsystems emerge sequentially in early development, and consequently it *appears* that more complex linguistic elements are constructed through mastery of the simpler, earlier-acquired components. However, this is not necessarily the case. Because development is inherently linked with time, it is tempting to view language development as also progressing in such a linear fashion. That is, it is common to imagine language development as a scatter plot with time on the x-axis and language acquisition milestones on the y-axis. If we arrange the developmental milestones in a sequential order ranging from simple to complex, in typical development we will

see a line with a sharp increase in the first two years as children acquire sounds, words, and word combinations, which asymptotes in the end of the preschool years as children have acquired sentences and narratives. While language acquisition is by no means complete at this point, it will continue at a much gentler slope, as children make subtle gains in the use of vocabulary, complex syntax, non-literal language, narratives, and in general become more sophisticated communicators. Charting these linguistic *milestones* does not necessarily capture the dynamic relations between the subsystems underlying them. In typical development, when language milestones are met on time and children show competent, even, linguistic profiles, it is difficult to disentangle the linguistic components that contribute to the attainment of each linguistic achievement. However, investigating the relations among linguistic components in *atypical* development, where children show uneven linguistic profiles, may help to 1) better understand the organization of the language system in typical development, and 2) create more sensitive and specific diagnostic tools, as well as more targeted and efficacious clinical interventions for children with language impairments.

High Functioning Children with Autism (HFA) are a subset of children diagnosed with Autism Spectrum Disorders (ASD), who have non-verbal IQs that are within the normal range (for a detailed intellectual profile, see Ghaziuddin & Mountain-Kimchi, 2004). Children with HFA make up about 20-30 percent of the children diagnosed with ASD (Tager-Flusberg, 2007), and while investigations into language performance of this HFA group have produced extremely mixed results, in general they suggest an uneven language profile, with strengths in some linguistic areas and severe deficits in others. Although most studies concur that social aspects of language, such as narratives and pragmatics, are impaired in HFA (e.g. Happé, 1994; Loveland & Tunali, 1993; Loveland, Landry, Hughes, Hall, & McEvoy, 1988; Nuske & Bavin, 2011; Tager-Flusberg, 1981, 1996; Wetherby, 1986), there is little agreement as to the functioning of individual subsystems of language (e.g. phonological, lexical, and syntactic levels), and the

mechanism(s) underlying these impairments (for reviews see Boucher, 2012; Groen, Zwiers, van der Gaag, & Buitelaar, 2008). Some investigations find that performance of children with HFA is on par with (or only slightly below) their typically developing peers at the phonological (Kjelgaard & Tager-Flusberg, 2001), lexical (Saldaña, Carreiras, & Frith, 2009), and syntactic (Tager-Flusberg & Calkins, 1990) levels. Such findings indicate that the structural levels are intact, and thus the deficits observed in higher language levels are likely due to the social constraints of language. This interpretation is consistent with Social Deficit views of autism, such as the Theory of Mind deficit (Baron-Cohen, 2000), which postulates that it is the social-communicative and perspective-taking requirements of language that have the most detrimental effect on language functioning in children with HFA.

Alternatively, some research suggests that the structural levels of language in children with HFA are not as robust as in typically developing children, and find evidence of impairment or atypical processing at the phonological (Cleland, Gibbon, Peppé, O'Hare, & Rutherford, 2010; Hooper, Poon, Marcus, & Fine, 2006; Shriberg et al., 2001), lexical (Dunn & Bates, 2005; Dunn, Vaughan, Kreuzer, & Kurtzberg, 2000; Fishman, Yam, Bellugi, Lincoln, & Mills, 2011; Henderson, Clarke, & Snowling, 2011; McCleery et al., 2010; Norbury, Griffiths, & Nation, 2010; Speirs et al., 2011), and syntactic (Eigsti, Bennetto, & Dadlani, 2007; Happé, 1997) levels. Such results are consistent with a broader processing account, which impacts all or most linguistic subsystems. One such processing account is the Weak Central Coherence hypothesis (Frith, 1989; Happé & Frith, 2006), which proposes that individuals with HFA show a processing bias toward isolated components—the local level—and a weakness in integrating components into a coherent whole. This account was originally proposed in the context of processing visual hierarchical form stimuli, in which the individual components of a visual array are arranged into a coherent pattern. For example, many Ts may be arranged into the shape of an X. In this example, the Ts comprise the local level and the X is the global level. In these investigations it is

consistently found that typical adults show a global processing bias (e.g. Navon, 1977). Some studies find that typically developing children also show a global processing bias (Mondloch, Geldart, Maurer, & De Schonen, 2003; Porporino, Shore, Iarocci, & Burack, 2004). Others, however, suggest that the global processing bias is more fragile in children (relative to adults), and increasing the task demands (Harrison & Stiles, 2009) and degrading the global level information (Dukette & Stiles, 1996) can shift a child's attention to the local level. Thus, in typically developing children, there is also some evidence of a global processing bias (though this bias may be vulnerable) (see chapter 5 of Stiles, Reilly, Levine, Trauner, & Nass, 2012 for a discussion). Children with ASD, on the other hand, are not distracted by the global-level information, and rather attend preferentially to the local level, suggesting weakness to integrate parts into a coherent whole. The weak central coherence account has since been extended to other cognitive domains such as music and pitch processing (e.g. Bonnel et al., 2003), face processing (e.g. Deruelle, Rondan, Gepner, & Tardif, 2004), motion processing (e.g. Bertone, Mottron, Jelenic, & Faubert, 2003), and recently, to language processing (Booth & Happé, 2010; Just, Cherkassky, Keller, & Minshew, 2004).

Booth and Happé (2010) administered a sentence completion task, in which they presented children and young adults (both typically developing and with ASD) with sentence stems and asked them to complete the sentence using a word or phrase. The sentences contrasted with respect to word associations (the local level) and sentence congruence (the global level). For example, *boys grow up to be men and ...* since the words *men* and *women* are closely associated lexical items, the local completion would be *women*, which is incongruous with the sentential meaning. A semantically congruous ending would be something like *... and girls grow up to be women*. Results indicated that there was variability in processing biases (toward local or global levels) within the typically developing population that was independent of IQ, but that children with HFA made more word-association errors than their typically developing, age-and-IQ-

matched peers, suggesting a local processing bias in HFA in the linguistic domain. In other words, the level of the word (and its associations) was processed preferentially over that of the sentence (and consequently the discourse congruence), perhaps due to weakness integrating the sentential components in the discourse context. Similarly, it was found that in a homograph reading task (e.g. *In her eye/dress there was a big tear*), individuals with autism showed less sensitivity to the sentential context to disambiguate the pronunciation of the target word (*tear*) relative to typically developing individuals, again suggesting a lack of integration across the sentential context (Happé, 1997). Evidence for weak integration has also been reported in a narrative comprehension context by Nuske and Bavin, (2011), who asked children with HFA and typically developing children inferencing questions following a short story. Questions were either related to the story as a whole (e.g. the story “script” or “main idea”) or to local aspects of the story (e.g. factual and detail questions). Children were also given the block design task to assess local and global visuospatial processing. Interestingly, there were no between-groups differences in accuracy to answer the detail or main idea questions. However, while performance from detail and main idea questions was significantly correlated in the TD group, they were not correlated in the HFA group, suggesting that the children with HFA (while they performed with similar accuracy) did not recruit global processing components to process the detail-related story questions and vice versa. Although patterns of behavior observed in individuals with HFA are consistent with predictions from the weak central coherence account in a number of domains, this framework does not propose a mechanism that may underlie this local processing bias.

The Cortical Underconnectivity Theory (e.g. Just et al., 2004; Just, Keller, Malave, Kana, & Varma, 2012; Kana, Keller, Cherkassky, Minshew, & Just, 2006) offers a neurophysiological framework for interpreting the integrative deficit observed in ASD across many contexts and domains. The underconnectivity theory proposes that the neural basis for the local processing bias in HFA is the lower degree of functional synchronization between anterior and posterior neural

regions. This theory was first proposed by Just and colleagues (2004), who, using an fMRI sentence comprehension paradigm identified lower levels of correspondence between activation in left inferior frontal gyrus (Broca's area) and left superior temporal gyrus (Wernicke's area) during a language task in individuals with HFA relative to typically developing controls. The authors suggest that this underconnectivity between language regions may index a neural mechanism underlying the integrative deficit observed in behavioral studies of language in individuals with autism. While this first study examined the synchrony between two regions of the brain that are within the language system, there is also evidence from other fMRI investigations that individuals with HFA show decreased connectivity between brain regions *across* cognitive systems as well. In a task that involved visual imagery in addition to language comprehension (e.g. truth judgment on sentences such as *the number eight rotated 90 degrees looks like a pair of eyeglasses*), it was found that individuals with HFA showed more activation in regions of the brain that correspond to visuospatial processes and less activation in neural regions associated with language processing. Furthermore, the regions associated with visuospatial processes and those associated with language processes demonstrated reduced synchrony relative to the typically developing participants (Kana, Keller, Cherkassky, Minshew, & Just, 2006).

There is also some evidence that it is not the underconnectivity in general that causes the behavioral deficits observed in language in HFA, but specifically underconnectivity of long-range white matter projections that cause the behavioral local processing biases and weak integration across cognitive systems observed in HFA (Belmonte et al., 2004; Lewis & Elman, 2008). Language processing, and especially complex language tasks such as relating a personal narrative, requires integration of cognitive processes that rely on networks that span the brain. To name a few, an individual must recall, organize, and plan his narrative (processes which recruit prefrontal cortical regions), must recruit the phonological, morphological, and syntactic linguistic

representations to convey his thoughts (processes which recruit at the very least the “classic language regions” in inferior frontal cortex and posterior superior temporal cortex), must plan and activate the motor pattern to articulate the linguistic representations (processes which involve premotor and primary motor cortex in posterior frontal regions), must integrate visuospatial processes to understand facial expressions and gestures of the interlocutor (processes which involve occipital and parietal cortical regions), and must attend to the perspective, emotion, and non-literal aspects of language (which involve distributed, both cortical and subcortical largely right hemisphere regions). Since these processes involve neural correlates from multiple regions across the brain, the long-range underconnectivity hypothesis would predict that narrative performance would be more disrupted than language processes that rely on more local connectivity such as phonological, lexico-semantic or even syntactic processing.

In sum, the theories presented above provide cognitive and neurological frameworks for considering the language impairments in HFA: those grounded in social deficit perspectives, such as the theory of mind account suggest that language impairment in HFA is driven primarily by difficulties in the social-communicative aspects of language such as taking multiple perspectives and empathizing with a listener. Processing accounts, such as the weak central coherence theory, propose that it is the integration of multiple components into a coherent whole that cause language impairment on complex language tasks. The underconnectivity theory hypothesizes that difficulties are due to atypical synchrony between neural regions involved in language processing, and the long-distance underconnectivity hypothesis suggests that it is long-range white matter connections, in particular, which may underlie the integrative linguistic impairment. None of these theories, on its own, however, can specifically account for 1) the mixed findings in research on language processing in HFA within and across participants, and 2) the inconsistent performance observed between linguistic subsystems. In this investigation, we evaluate language at multiple levels of linguistic complexity (phonological, lexico-semantic, and syntactic) in

written and spoken modalities, as well as spoken narrative composition in typically developing children and children with HFA in order to address questions about the associations among these linguistic subsystems, and their relation to the language system as a whole.

METHODS

PARTICIPANTS

Sixteen High Functioning children with Autism (HFA) and sixteen typically developing children (TD) aged seven to twelve (mean TD = 9.09; SD = 1.95 and mean HFA = 9.82; SD = 1.51) participated in the present study. TD and HFA groups did not differ from one another in age ($p > .2$). All participants had normal (or corrected to normal) vision and hearing, and were monolingual English speakers, with no exposure to a second language before the age of five. Children were not admitted to the study if they had a history of a medical condition that might interfere with test performance.

Children in the TD group had normal developmental and medical histories and normal neurological exams. All scored within or above the normal range on the Wechsler Abbreviated Scale of Intelligence Test (WASI) (Wechsler & Chen, 1999) (standardized Intelligence scores ranged from 89 - 129), and within or above the normal range on the Clinical Evaluation of Language Fundamentals- Fourth Edition (CELF-IV) (Semel, Wiig, & Secord, 2003) (standardized Core Language scores ranged from 94-123), and none had a history of chronic medication use.

Children in the HFA group met diagnostic criteria for Autism from the Diagnostic and Statistical Manual of Mental Disorders-IV (DSM-IV, APA, 2000) and from the Autism Diagnostic Interview—Revised (ADI-R, Lord et al., 1994), and/or the Autism Diagnostic Observation Schedule (ADOS, Lord et al., 1989). All demonstrated a Full Scale IQ of 70 or above (scores ranged from 70-128), and none had a specific underlying genetic or metabolic

diagnosis (e.g. tuberous sclerosis, Fragile X syndrome, or structural brain malformations). For results from the screening intelligence and language measures, see Table 3.1.

Table 3.1: Age and Standardized Testing Results for TD and HFA¹

Test	Measure	TD Mean (SD)	HFA Mean (SD)	<i>p-value</i>
	Age	9.09 (1.95)	9.89 (1.51)	NS
CELF-IV²	Core Language	109.5 (8.93)	82.50 (23.27)	< .001
	Expressive Language	109.5 (9.87)	85.43 (23.81)	< .005
	Receptive Language	110.31 (10.02)	82.69 (22.31)	< .001
EOWPVT³	Vocabulary	115 (8.97)	99.88 (23.10)	< .05
WASI⁴	Full Scale IQ	115.25 (11.83)	94.00 (17.17)	< .001
	Verbal IQ	121.31 (12.29)	97.88 (18.39)	< .0001
	Performance IQ	105.81 (12.54)	91.50 (20.52)	< .05

¹ All scores expressed as standard scores

² Clinical Evaluation of Language Fundamentals- Fourth Edition (Semel, Wiig, & Secord, 2003)

³ Expressive One Word Picture Vocabulary Test-Revised (Gardner, 1990)

⁴ Wechsler Abbreviated Scale of Intelligence (Wechsler & Chen, 1999)

PROCEDURE AND MEASURES

Participants were tested individually as part of a larger study. Language was assessed in speaking and reading modalities at the phonological, lexico-semantic, and syntactic levels, as well as narrative discourse in the spoken modality. Spoken phonological processing was measured using a subtest of the Phonological Abilities Test: Second Edition (PAT-2) (Muter, Hulme, & Snowling, 1997), in which children are asked to isolate the middle phoneme in a three-phoneme word. For example, *tell me the middle sound in the word CAT*. The subtest consists of ten aurally presented words. Because this test is designed to assess phonological deficits in early readers, it is normed on a group of children 4-7 years of age. Since the children in our sample are older than this age range, we report raw scores rather than normed scores. We also chose the most difficult subtest from this assessment (that which showed the most variance in the typically developing group) to avoid ceiling effects. Phonological processing in the reading domain was assessed using the “Word Attack” subtest of the Woodcock Johnson Test of Achievement- Third

Edition (WJ-3) (Woodcock, McGrew, & Mather, 2001), in which children are asked to read non-words aloud. Because the words are pseudowords, children can only use phonological decoding, and cannot rely on an orthographic reading strategy. Words on this subtest corresponded to the orthographic rules of English, for example, *plurp* and *fronkett*. Later test items are more difficult than early test items. To be statistically consistent across measures, raw scores are reported for all assessments.

Lexico-semantic processing in the reading modality was assessed using one condition from the experimental single word reading measure, Recognizing Written Synonyms (RWS), designed by Polse and Reilly (2012). In this measure, children are presented with a word and two response options that differ with regard to orthographic and semantic similarity to the target, and are asked to touch the word that is closest in meaning to the target. The orthographic-semantic condition is the most difficult condition, as orthographically similar and semantically similar words are pitted directly against one another (e.g. Target: STONE Match: ROCK Foil: STOVE). In typical development, children do not perform above chance levels on this condition until around the second or third grade (Polse & Reilly, 2012). In order to account for possible speed-accuracy trade-offs, Efficiency scores are reported (mean Reaction time/mean Accuracy). Thus, for this measure, higher scores indicate *less* efficient processing. To remain visually consistent, negative is plotted up for this measure in the box and whisker plots displayed below. In order to remain as consistent as possible across modalities, lexico-semantic processing in the speaking domain was assessed using a receptive vocabulary measure, the Peabody Picture Vocabulary Test III (PPVT-III) (Dunn, Williams, Wang, & Booklets, 1997), which requires children to touch the picture that matches the meaning of a given word (from an array of four pictures).

Sentence-level speaking was assessed using the Formulating Sentences subtest of the CELF-IV. In this assessment, children are asked to make a sentence using a specific word or phrase. Words and phrases become more difficult as the test proceeds. For example, an early item

of this subtest is, “Make a sentence using the word *children*” and a later item is, “Make a sentence using the phrase *as a consequence*.” This assessment measures productive syntactic ability, as the child must manipulate the given word or phrase into a sentential context. Sentence-level reading was evaluated using the Reading Fluency subtest of the WJ-III. In this subtest, children are given a list of sentences that are either true or false, and are asked to read them silently and circle YES or NO for as many sentences as they can within three minutes. Sentences become longer and more complex as the test goes on. For example, an early item on this subtest is *a bird can fly* and a later item is *people may use a map to find certain locations*.

Narrative-level language was assessed using a spoken Personal Narrative Task. In this task, adapted from (Berman & Verhoeven, 2002), children are asked to tell a story about a conflict. Specifically, they are given the prompt: *People disagree and they get into fights or arguments all the time. I’m sure there has been a time when you had an argument or a problem or when someone made you mad or sad. I’d like you to tell me about it. Tell me how it started, what happened, and how it all ended. Take time to think and begin when you’re ready*. This task requires children to recall, organize (in the correct temporal sequence), and produce a story, which draws on both linguistic and social processing skills. Children were videotaped and then their narratives were transcribed and coded offline using CHAT from the Child Language Data Exchange System (CHILDES; MacWhinney, 1991). Narratives were coded for the overall quality of the narrative (i.e. story elements) as well as linguistic structure within the narrative. For specifics, see Table 3.2.

Table 3.2: Examples and Coding System for the Personal Narrative Measure

Measure	Coding Scheme	Example
Narrative composition	Narratives were coded out of 4 possible points (one point each): Any setting element, Problem, Response, Resolution	<u>Setting</u> : One day, at school... <u>Problem</u> : She hit me for no reason... <u>Response</u> : So I ran to tell the teacher... <u>Resolution</u> : She made her apologize to me and then we were friends again
Length	Number of semantic propositions	When I was at school/ I was playing on the play structure/ [2 propositions]
Setting	Provided a context for the listener (out of 4 points, one point each): time, place, character, situation	<u>Time</u> : Yesterday <u>Place</u> : I was at my friend's house <u>Character</u> : I was playing with my brother <u>Situation</u> : We were at camp and we weren't supposed to have food in our cabins
Morphological Errors	Expressed as the proportion of morphological errors out of total propositions. Morphological errors were coded for errors in: Pronoun, Auxiliary verb, Verb Copula, Determiner, Preposition, Number agreement, Subject-verb agreement, and Tense	<u>Pronoun</u> : <u>him</u> went over to the teacher <u>Auxiliary</u> : it was like <u>make</u> [] me sick <u>Verb copula</u> : I [] taller than him <u>Determiner</u> : my favorite is <u>a</u> geese <u>Preposition</u> : come along <u>in</u> his plan <u>Number</u> : she had lotsa toy [] <u>Subject-verb</u> : he <u>have</u> his own gameboy <u>Tense</u> : I was so scared because <u>it's</u> Ghost Galaxy
Complex Syntax	Expressed as the proportion of complex syntax out of total propositions. Complex syntax included: Coordinates, Verb complements, Adverbial clauses, Relative clauses, Passives	<u>Coordinates</u> : he pushed me <u>and</u> I kicked him <u>Verb Complements</u> : he knew <u>that</u> h'ed <u>get in trouble</u> <u>Adverbial Clauses</u> : then she got mad <u>because</u> she didn't agree with me <u>Relative Clauses</u> : and so then a long time ago my dad gave me the same phone <u>he was</u> holding, but it couldn't call <u>Passives</u> : he kind of did got out so he <u>was</u> banned for the rest of the month
Evaluation	Expressed as the proportion of evaluative devices out of total propositions. Evaluative devices included: Taking perspective in the story, using: Mental markers, Emotional terms, Causal markers, Intensifiers	<u>Mental markers</u> : I <u>think</u> he tripped me <u>on purpose</u> <u>Emotional terms</u> : my mom got <u>mad</u> because we made a mess <u>Causal markers</u> : I went and played by myself <u>because</u> they were leaving me out <u>Intensifiers</u> : and it hurt <u>really really</u> bad!

RESULTS

BETWEEN GROUP COMPARISONS

In order to characterize performance, we present box and whisker plots of TD and HFA group performance for all measures in Figure 3.1. Since the goal of the present investigation is to look at relations between levels of linguistic complexity within participants, and not to compare each individual's performance to a group or norm, we present raw scores (rather than standard scores) even when standard scores exist. Furthermore, since we used some standard and some experimental measures (i.e. some do not have a standard score), we do not convert to normalized scores (even where they exist) in order to remain consistent when correlating across measures. Since the groups do not differ in age, this should not pose a problem for comparison within or between subjects. Independent *t*-tests of the raw scores from the individual assessments reveal that there is a significant difference between group performance (such that the children with HFA performed below the typically developing children) on the spoken phonological measure (PAT) ($t(30) = 2.21, p < .05$), a marginal difference on the spoken lexico-semantic measure (PPVT) ($t(30) = 2.03, p = .05$), and a significant difference between groups on the narrative composition score $t(30) = 2.62, p < .05$. All other between-groups comparisons for individual language elements were non-significant ($p > .1$) (Figure 3.1). Within the narrative context, groups did not differ significantly on: the length of their spoken narrative (measured as number of propositions) (TD mean = 16.125; SD = 10.21; HFA mean = 11.25; SD = 9.91; $t(30) = 1.58, p > .1$; the proportion of morphological errors per proposition ($t(30) = .967, p > .3$; or the proportion of evaluative devices per proposition $t(30) = 1.31, p > .1$. However, children with HFA used less complex syntax than the TD group $t(30) = 3.38, p < .01$ and provided a setting less often than the TD group $t(30) = 2.21, p < .05$ (see Figure 3.2).

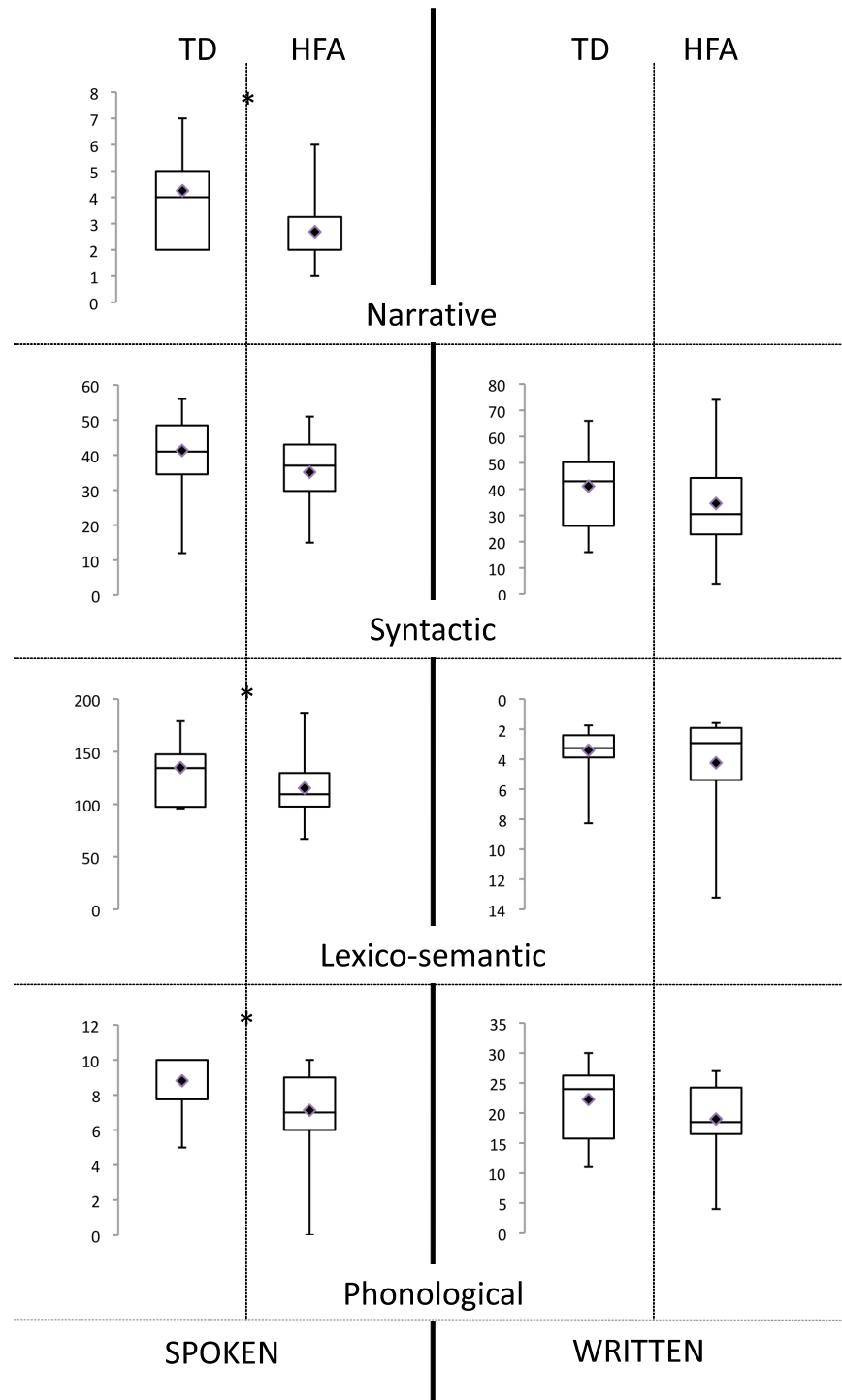


Figure 3.1: Displays the raw scores in the spoken (left) and written (right) modalities for (from bottom to top): phonological, lexico-semantic, and syntactic processing. Narrative composition was assessed in the spoken domain only. Within each plot, the TD group is on the left and the HFA group is on the right. Since raw scores are reported, the y-axis represents the scale for each respective measure, and thus cannot be compared between measures. Asterisks mark significant between-groups comparisons (at the $p < .05$ level).

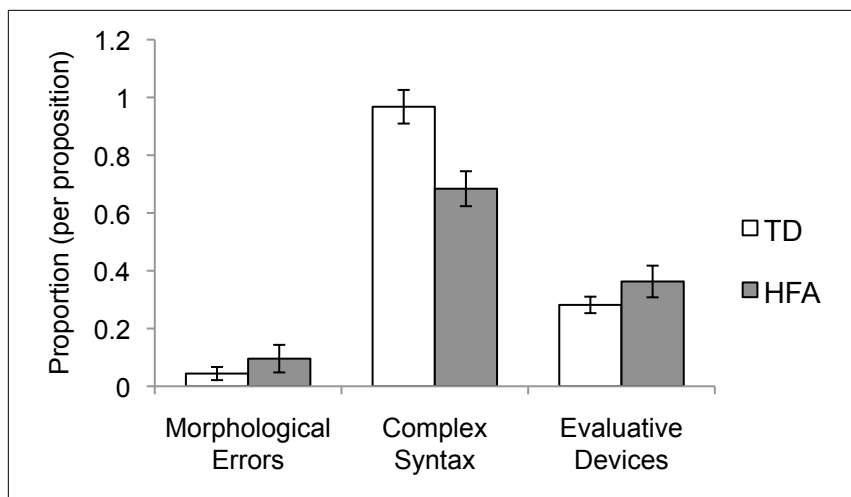


Figure 3.2: Displays the proportion of morphological errors, complex syntax, and evaluation out of the total number of propositions in the child's narrative. Scores are expressed as means with standard error bars for the TD (white bars) and HFA (gray bars) groups.

CORRELATIONS WITHIN GROUPS

To investigate the association between linguistic levels and modalities within groups, raw scores from the seven independent linguistic components were used to create two separate correlation matrices, one for the TD group and one for the HFA group. Results reveal that in the TD group, levels of linguistic complexity were tightly correlated, with 17 out of the 21 comparisons showing significant correlations (at $p < .05$) (and 20 out of 21 were significant at the $p < .1$ level). R -values ranged between .54 and .74 (See Figure 3.3). For the HFA group, only six of the 21 comparisons showed correlations that were significant at the $p < .05$ level (or at the $p < .1$ level). The R -values for the significant correlations in the HFA group showed a similar range (between .54 and .73) (See Figure 3.3). The correlation matrices will be discussed in depth in the Discussion.

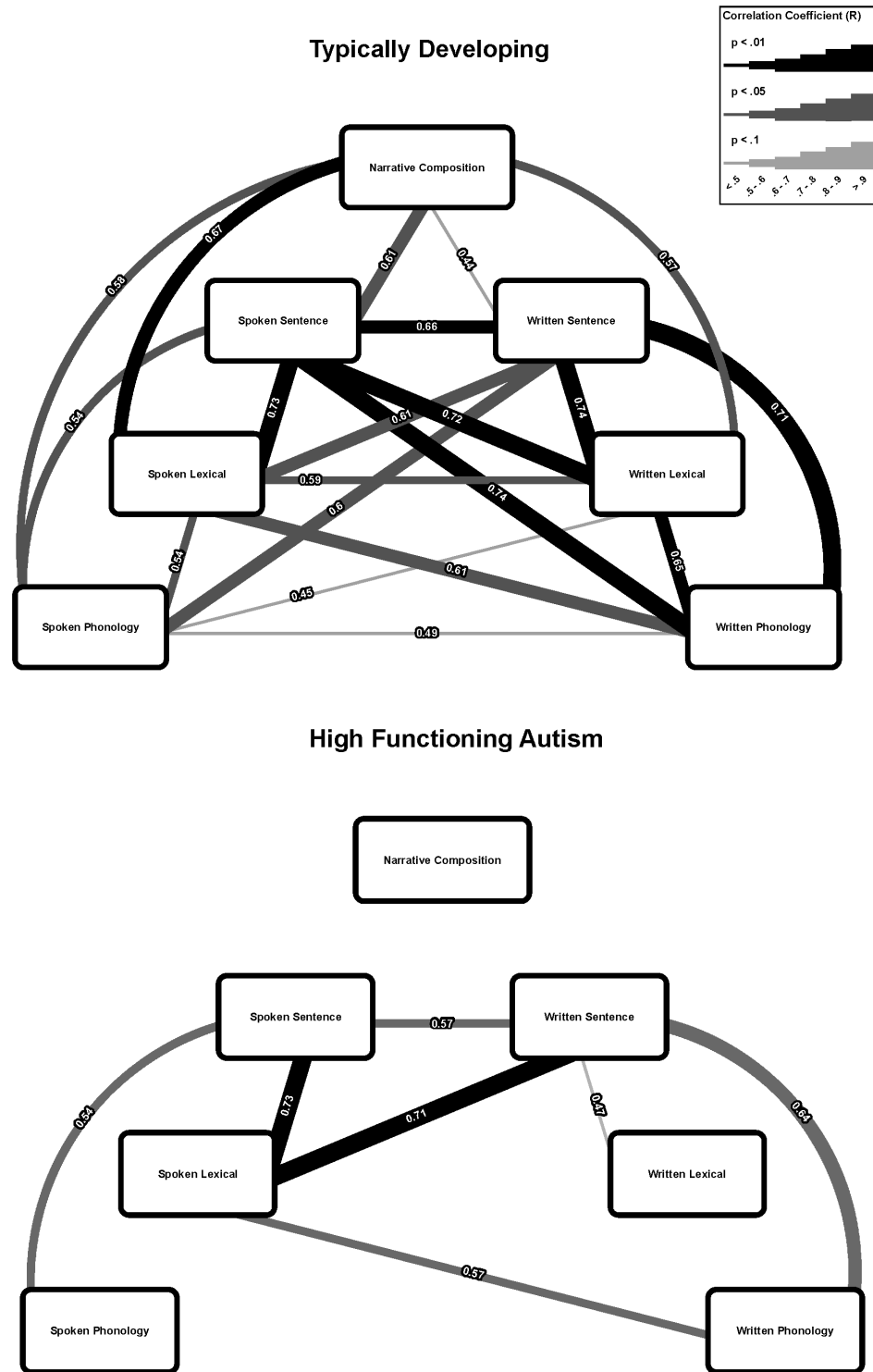


Figure 3.3: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for TD (top) and HFA (bottom). The width of the line represents the correlation value, and the grey-scale represents the p -value.

DISCUSSION

We have presented results from phonological, lexico-semantic, and syntactic levels of language in both written and spoken modalities with the aim of addressing how relations between language levels and modalities affect the linguistic system as a whole, and the ability to organize these linguistic subsystems into a cohesive narrative. We first looked at the TD and HFA performance on the individual linguistic measures. It is apparent from the box and whisker plots displayed in Figure 1 that the HFA group consistently performed below the TD group on every level of linguistic complexity. However, it is also apparent that the children with HFA do not fall dramatically below the TD group in most cases, and indeed only three of the seven linguistic subsystems were statistically significantly different between groups: spoken phonological, spoken lexico-semantic, and narrative composition. Although we had predicted that children with HFA would fall below their TD peers in their ability to compose and articulate a personal narrative, their degraded performance in the spoken phonological and lexico-semantic domains was unexpected. That is, based on previous research and processing accounts of HFA (such as the weak central coherence hypothesis, Frith, 1989; Happé & Frith, 2006), we expected the smallest units of language to be the strongest (i.e. phonology and vocabulary). We hypothesized that difficulties would emerge in the more complex, integrative levels such as syntactic and narrative. While there is some evidence for this hypothesis within the narrative context (children with HFA used less complex syntax relative to the TD group and showed few word-level, morphological errors), the isolated measures of syntactic and lexico-semantic processing did not follow this pattern. A closer look at the distributions of the individual measures (Figure 1) provides some insight into this surprising finding.

First, the phonological level was assessed using the PAT-2, which is a standardized phonological assessment normed on a group of younger children (aged 4-7). It is not surprising, then, that the typically developing children (aged 7-12) performed mostly at ceiling on this

assessment, with the median and the mode both falling at 10 (the equivalent of 100%) and the lowest score in the TD group falling at 5. The HFA group, by contrast, showed much greater spread in their distribution, with a mode of 6 and a median of 7, and some children with HFA performed with 100% accuracy and some performed with 0% accuracy. Thus, some of the difference between groups on the phonological spoken level may stem from the lack of variability in the TD group; however, the most striking element of this finding is the remarkable heterogeneity in phonological performance within the HFA group. This variability emphasizes the importance of looking across linguistic subsystems *within* individual participants when studying children with neurodevelopmental disorders.

The HFA group also performed significantly below the TD group on the lexico-semantic assessment, yet a closer look at the distribution of PPVT-III scores also suggests that this difference may be due to better-than-expected performance from the TD group rather than degraded performance from the children with HFA. To further explore this post-hoc hypothesis, raw scores were converted to standardized scores for both groups. In the TD group, children performed with an average of 111.93 (SD 8.99), which is almost a full standard deviation above the normalized mean of 100. Children in the HFA group performed with a mean standard score of 95.75 (standard deviation of 19.86), which is significantly below our TD group, but well within the typical range based on the standard norms, and well within one standard deviation of the standardized mean of 100. Thus, while the groups differ in spoken lexico-semantic processing, we cannot conclude from this finding that vocabulary is an isolated weakness in the HFA group, as their performance was within the typical range when we look at the standardized scores. It appears instead that our TD group has exceptional spoken lexico-semantic processing skills relative to the population as a whole.

Within a more naturalistic context, the Spoken Narrative assessment, a different pattern emerged, and one that was more consistent with our hypotheses. The two groups did not differ in

terms of the length of their spoken narrative, nor in the proportion of morphological errors, but the HFA group performed significantly below the TD group in their rate of complex syntax. In this context we find strong evidence for the weak central coherence hypothesis, which manifests in language performance as weak (long distance) integration of linguistic subsystems:

performance on the more local aspects of language processing (morphology) was similar in HFA and TD groups, but aspects that require integration and long-distance dependencies, such as multiple forms of complex syntax (e.g. adverbial and relative clauses) was degraded. Results from the narrative components that are more relevant to social deficit theories of autism were more mixed. First, providing a setting serves to orient the listener to the time, place, and context of the story, and necessarily requires taking the perspective of the listener (a theory of mind) to understand that he or she doesn't already know where and when the story took place. Because children with HFA provided a setting significantly less than the TD group, this result is consistent with social deficit frameworks of autism such as the theory of mind perspective (Baron-Cohen, 2000; Colle, Baron-Cohen, & Hill, 2007). Secondly, the use of evaluative devices in a personal narrative can be viewed as a social tool with which to "hook" your listener, and to help them empathize with the emotions of the characters in the story. Due to these socially-oriented goals of the evaluative devices, social deficit theories would hypothesize that children with HFA would use fewer evaluative devices than their typically developing peers. This was not the case. In fact, though the trend was statistically non-significant, children with HFA used a slightly higher rate of evaluative devices on average than typically developing children. While this finding is quite surprising from a social deficit perspective of autism, it may actually be expected from a weak central coherence account. That is, since the narrative exists in an emotional context (*"Tell me about a time when someone made you mad or sad"*), using single emotional lexical items (e.g. sad and mad) does not necessarily require integration, and indeed could be simply the local, associative response to a question (e.g. *"My brother made me mad and sad"*). Since children

performed worse than their typically developing peers on the more global narrative processing measure (the Narrative Composition), which requires more long-distance integration, and better on the evaluative measure that requires more local processing, these results (from within the narrative context) fit nicely within the weak central coherence account of HFA.

One question that is raised from these between-groups comparisons is: Why do observed patterns from the individual measures of language components in isolation differ from the pattern of individual components assessed within the narrative context (e.g. syntactic processing was degraded in the narrative context but not in the isolated measure)? A possible explanation that is consistent with the weak central coherence hypothesis is the very nature of the narrative: it is a context that requires more integration than the isolated linguistic assessments. That is, to relate a personal narrative, it is already required that the child recall, organize, and hold in memory at least a sketch of his or her story. In the isolated syntactic assessment, by contrast, the child was only responsible for constructing, holding in memory, and integrating parts of a single sentence at a time. However, there is also a social deficit account, which has equal power to explain these discrepant findings. It is possible that the more socially demanding task of telling a story to another person caused the children with HFA to perform below their typically developing peers on language components that did not show differences when assessed using isolated standardized measures. In sum, results from linguistic components in isolation and within the narrative context provide some conflicting results, which can be explained by an integrative framework, as well as by a social communication framework. Results from the correlation analyses between components within groups, however, provide further information on the underlying organization of these linguistic components in children with HFA and typically developing children, which will help to disentangle these two possible accounts.

Results from correlations between linguistic subsystems within populations reveal multiple significant correlations in the TD group, and relatively few significant correlations in the

HFA group. Starting at the smallest unit of linguistic complexity, we found that in the TD group, spoken phonology was significantly correlated with: lexico-semantic processing, syntactic processing, and narrative composition in the spoken modality, and phonological processing (marginal significance), lexico-semantic processing (marginal significance), and syntactic processing in the written modality. Spoken lexico-semantic processing was significantly correlated with syntactic processing and narrative composition in the spoken modality and phonological, lexico-semantic and syntactic processing in the written modality. Spoken syntactic processing was significantly correlated with narrative composition in the spoken modality and with phonological, lexico-semantic, and syntactic processing in the written modality. In the written modality, written phonological processing was significantly correlated with written lexico-semantic and written syntactic levels. Written lexico-semantic processing was correlated with written syntactic and narrative composition. Written syntactic processing was also correlated with narrative composition. In the HFA group, by contrast, performance on spoken phonological measure was correlated with syntactic processing in the spoken modality only. Spoken lexico-semantic processing was significantly correlated with syntactic processing in the spoken modality, and phonological and syntactic processing in the written modality. Syntactic processing in the spoken modality was significantly correlated with syntactic processing in the written modality. In the written domain, written phonological processing was significantly correlated with written syntactic processing and written lexical-semantic processing was (marginally) correlated with written syntactic processing. Narrative composition was not significantly correlated with any other level of linguistic analysis. It is clear from these results that there is a different overall organization in the levels of association between linguistic subsystems: while linguistic subsystems are tightly associated in the TD group, they do not show the same infrastructure in the HFA group. However, there are also a number of questions raised from this finding. The first series of questions involves the specific linguistic subsystems that are related

and unrelated: We see that the vast majority of linguistic subsystems are correlated in the TD group, but what about the one relation that is not (even marginally) correlated (written phonological processing with spoken narrative)? On the other hand, why are *some* linguistic subsystems correlated in HFA? Is there any evidence that the more tightly associated subsystems are more robust than other, more “isolated” subsystems? Secondly, what are the possible cognitive and neural mechanisms for these behavioral dissociations, and associations between subsystems of language? Lastly, and most importantly, what can these patterns of association between linguistic subsystems tell us about the language system as a whole in typically developing children and children with HFA? How can identification of this profile assist in creating more targeted, efficacious clinical interventions?

Our first series of questions concerns the associations that do not conform to the overarching patterns: in TD, the subsystems that do *not* correlate with one another, and in HFA, those that do. While almost every linguistic subsystem in the spoken and written modalities was correlated with every other subsystem in the typically developing group, there was one exception: written phonological processing (assessed using the Word Attack subtest of the WJ-III) did not correlate with performance on the spoken narrative. One possible explanation for this disparate finding is that the distance between subsystems is too great. That is, if we picture the increasing levels of complexity from this investigation building on one another from 1) phonological 2) lexico-semantic 3) syntactic and 4) narrative, then according to this schema, phonological and narrative are the furthest apart in this “vertical” orientation. Also if we consider the two separate modalities (spoken and written) as existing in parallel, there is also a degree of separation in this more “horizontal” dimension. Thus, perhaps it is the separation both in level of complexity and in modality that explains the lack of association between written phonological and spoken narrative subsystems. However, if this account were correct, we would expect to see a weakening trend—decreasing correlation values as subsystems become “further” apart—leading up to the non-

significant correlation. While there is some evidence for this hypothesized explanation in terms of qualitative trends (e.g. spoken phonological seems to be more weakly correlated with narrative and syntactic levels relative to lexico-semantic levels), it is by no means a consistent pattern and thus warrants further, more direct exploration in future research. Alternatively, the lack of relation between written phonological processing and narrative may be a result of the task demands. That is, while spoken phonology is necessarily recruited to tell a story (as the narrative is a spoken task), accessing the orthographic representations is not necessarily required or automatic in childhood. It is possible that the lexico-semantic level in the written modality *did* show correlations with the narrative because full orthographic representations of words are already linked with their spoken, phonological form. Thus, when children recruit word and syntactic level spoken information to perform the narrative, it is possible that orthographic representations of words are also automatically activated. If this hypothesis were correct, these multiple subsystems would have been regularly co-activated, which would account for their highly associated relationship. Lastly, it could be that written phonological processing and spoken narrative composition *are* correlated, but the Word Attack subtest of the WJ is not a sensitive measure of phonological processing in the written modality. As is the case for each of the subsystems, here we present data from one representative measure. It is by no means certain, however, that the measures we chose to represent linguistic processing at each level of complexity is the best or the only representative measure. In order to confirm these patterns of relations between linguistic subsystems and modalities, our results should be replicated using these as well as other (standardized and experimental) measures of linguistic processing to ensure that results are not the consequence of the specific measure, but rather represent the organization and associations between elements that comprise the language system.

In the HFA group, while very few of the linguistic subsystems were significantly correlated with others, there were a handful of strong correlations, especially from the lexico-

semantic subsystem, which, in the visualization of the data presented in Figure 3, appears almost as a “hub” in the linguistic system. This finding does not indicate that vocabulary is a strength in HFA, but rather that *when* lexico-semantic processing is a strength, other linguistic subsystems such as spoken syntactic and written syntactic subsystems are also likely to be strong. On the other hand, when lexico-semantic processing is a weakness, these other subsystems are likely to show weakness. Thus, it seems that while the lexico-semantic processing level may be critically important to the language system as a whole, we cannot conclude that it is more robust than other systems, as a high correlation value could represent high or low performance. However, as performance on the lexico-semantic measure does correlate with other levels of linguistic complexity in spoken and written modalities, perhaps spoken lexico-semantic processing could act as a sensitive diagnostic tool to identify broader language impairment in this population. Importantly, while strength (or weakness) at the spoken lexico-semantic level associates with syntactic processing (in spoken and written modalities), it does not extend to narrative performance. Thus, it is likely that while children with HFA can integrate single word lexico-semantic information into syntactic contexts (based on their tightly associated relationship), they show less association (and more impairment) at the narrative level, suggesting a lack of long-distance integration. Also interesting to note is that spoken lexico-semantic processing was *not* correlated with written lexico-semantic processing in the HFA group, as it was in the TD group. This may have implications regarding reading strategies in children with HFA. It is possible that children with HFA rely less on phonological information when reading a single word, and are more focused on the orthographic shape of the word. This finding is consistent with research that indicates superior visuospatial processing in HFA (e.g. Caron, Mottron, Berthiaume, & Dawson, 2006; O’Riordan, Plaisted, Driver, & Baron-Cohen, 2001; O’Riordan, 2004; Shah & Frith, 1993), and indeed there is some evidence that children with HFA show a processing bias toward the orthographic information when reading single words (Polse, Bellugi, & Reilly, submitted).

In our second series of questions, we ask, what are the possible cognitive and neural mechanisms underlying these correlations, and lack thereof? Unlike the results from the between groups analyses, which were consistent with both social deficit and weak integration accounts of HFA, the results from the correlation analyses provide strong support for integration perspectives such as the weak central coherence account. Specifically, there is some level of correlation between language elements in children with HFA (especially between spoken lexico-semantic processing and syntactic processing), suggesting that those with strong lexico-semantic processing have the capacity to integrate this skill into a more complex, syntactic context. There are also limits to this integration, however, as there were no significant correlations with narrative composition performance, and children with HFA performed significantly below their TD peers on the ability to tell a personal narrative. Thus, we do not find evidence for a *lack* of coherence or integration; rather, we find evidence for an overall *weakness* in associations across the linguistic system, which suggests that the individual linguistic elements are not strengthening and reinforcing one another at increasing levels of complexity in children with HFA, as they are in typical development. On the other hand, if the differences between groups could be explained solely through a social deficit theory, we would expect to see similar associations between linguistic components, which are then disrupted when placed in a social context. Instead, we find strong evidence that there is a different underlying organization of components that comprise the language system, a finding which is more consistent with an overall processing account.

As this was a behavioral investigation, we can only speculate as to the underlying neurological or neurophysiological causes of the observed weak integration across linguistic subsystems. However, the cortical underconnectivity and long-range cortical underconnectivity accounts provide neurologically based frames of reference for interpreting our results, and foundations upon which to build hypotheses. First, weak associations in behavioral performance across linguistic elements may be the manifestation of any number of neural irregularities,

including irregularities in structural anatomy (e.g. aberrant myelination patterns) and/or irregularities in function (e.g. asynchronous patterns of neural activation in different regions of cortex). Since children with HFA showed typical performance on many of the tests for the isolated linguistic elements, yet significantly reduced processing in the narrative (which requires integration of the lower-level subsystems), our results fit nicely with both the underconnectivity and the long-distance underconnectivity hypotheses. That is, children with HFA showed fewer significant correlations between linguistic subsystems throughout the language system, which may reflect less co-activation between the subsystems and consequently less synchrony across language regions, which is consistent with the underconnectivity theory. However, children with HFA only showed dramatically decreased *performance* on the narrative composition measure, which likely recruits more long-range, interhemispheric neural regions. This suggests that perhaps the white matter structural integrity required to form the long-distance connections involved in recalling, organizing, and producing a personal narrative may be weak or impaired, consistent with the hypothesis that decreased myelination of the long-range white matter tracts may negatively impact performance.

Lastly, what can this investigation tell us about the language system as a whole, and how can it inform clinical interventions? At the most fundamental level, this investigation suggests that the organization of the language system in typically developing children and children with HFA differs. And importantly, it does not differ simply because children with HFA perform worse (which would be an uninteresting finding based on the diagnosis of a neurodevelopmental disorder involving communication), but rather, regardless of between group differences in performance (which are few), the relations between the elements that comprise the language system within individuals do not show the same associations in children with HFA as they do in typically developing children.

These differences in relations among linguistic elements also have clinical relevance for language diagnoses and interventions in children with HFA. We find two pieces of corroborative evidence for Crystal's (1987) assertion that looking at the interactions between levels of language may serve as a valuable diagnostic tool. First, we find evidence for high variability both within subjects (different performance on the same linguistic component in the different assessment contexts) as well as between subjects (the HFA group consistently showed more variable distributions across measures relative to the TD group). Thus, as Crystal (1987) hypothesized, perhaps it is not that syntactic processing is vulnerable or variable, for example, but rather that the relations *between* syntactic processing and narrative processing are vulnerable or weak, which may explain the inconsistent findings within individuals. Secondly, we find that the performance in an integrative context differs from performance in isolated assessment environments. This finding suggests that when an individual is called on to integrate phonological, lexico-semantic, and syntactic components together into a narrative, points of weakness are revealed. This may also help to explain some of the contradictory findings in the literature on language processing in children with HFA. For example, some investigations find phonological processing to be a relative strength, and others find an increased level of articulatory errors in the speech of children with HFA, suggesting fragility in the phonological system. It is likely that these contradictory results come from different contexts of assessment: receptive phonological processing in isolation may be quite good, but decay in an expressive discourse context, which requires recruitment and integration of multiple linguistic elements. These results suggest that clinical evaluation of language disorders should assess linguistic structures in isolation as well as in an integrative environment. Identifying and diagnosing disorders based on integrating two or more subsystems may also have implications for how to treat these specific integrative impairments.

Based on patterns of typical language acquisition as well as reductionist logic, often there is an expectation that the smallest units of linguistic complexity provide a platform for more

complex linguistic structures; if there is weakness at one of the “lower” levels, the “higher” levels cannot function properly. This type of logic has led to intervention strategies that focus on the low-level perceptual properties of language (e.g. Fast Forward, Tallal et al., 1996), which operate based on the assumption that treating low-level perceptual processes will generate to higher-level language skills. While this investigation provides support for this notion in the typically developing group (strength or weakness at one level correlates closely with almost all other levels), we do not find evidence for this “building up language” model in the HFA group. That is, the smallest units of processing, while lower than the TD group, did not show severe impairment; performance at the syntactic levels did not differ from the TD group, but narrative performance, the most naturalistic, and arguably most important language skill, was impaired. Furthermore, with the exception of the lexico-semantic subsystem, linguistic elements were not correlated with one another in the HFA group, which implies that there is no reason to believe that intervention at one isolated level (e.g. phonology) will generate to any other linguistic subsystem. Consequently, these results provide little support for focusing language interventions for children with HFA on isolated linguistic elements (such as phonological awareness, rapid naming, or grammatical or syntactic structures), and instead suggest treating language in communicative, functional contexts—such as narratives and conversations—which require the recruitment and integration of multiple subsystems.

CONCLUSIONS

In conclusion, we investigated language at multiple levels of complexity (phonological, lexico-semantic, and syntactic) in spoken and written modalities, as well as spoken narrative composition in typically developing children and High Functioning children with Autism. While results on the isolated linguistic elements did not differ dramatically across groups, performance on the narrative composition measure was significantly impaired in children with HFA relative to their typically developing peers. There were also striking differences in the patterns of relations

between linguistic subsystems in the typically developing group relative to the group with HFA. In typically developing children, performance on almost all of the linguistic subsystems was significantly correlated with one another. In the HFA group, by contrast, there were very few significant correlations. Our results are consistent with the weak central coherence and underconnectivity hypotheses, and suggest that since linguistic elements are not correlated in HFA, language interventions should focus on the language system as a whole rather than on isolated linguistic elements.

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CHAPTER 4: A MULTI-LEVEL, MULTIMODAL LANGUAGE AND STRUCTURAL IMAGING INVESTIGATION: EXPLORING THE BRAIN-BEHAVIOR RELATIONSHIP IN TWO CHILDREN WITH PERINATAL STROKE

ABSTRACT

Children who experience unilateral strokes in the perinatal period have been found to show almost typical conversational language in childhood and no side-of-lesion differences, attesting to the brain's remarkable plasticity to reorganize to support linguistic processes. However, standardized measures of isolated language structures tend to show impairment, perhaps revealing fragility of the language system. While the superiority of language use over language structure in children with perinatal stroke is a consistent finding, the vast majority of research on language processing in children with perinatal stroke reports increased variance in the perinatal stroke group relative to typically developing children. This increased variance in behavioral performance may stem from differences in the lesion characteristics, as well as the state of the non-lesioned hemisphere. Group investigations have the ability to generalize results to a broader population, but they cannot use this individual variability to better understand the relations between the unique characteristics of the brain and an individual's unique language profile. In this case study, we present a detailed account of language processing at multiple levels of linguistic complexity in speaking, reading, and writing modalities, alongside detailed quantitative multimodal imaging measures of white matter microstructure in both hemispheres in two school-aged children with unilateral perinatal stroke, one in the left hemisphere (affecting temporal-parietal perisylvian regions) and one in the right hemisphere (affecting anterior perisylvian regions). Results indicate that the two children with perinatal stroke both perform better on unstructured, language-use measures relative to standardized measures of language structure, however we also identify unique linguistic impairment. We provide evidence that assessing the non-lesioned hemisphere is critical to understanding language behavior, as white

matter structural integrity in the child with the right hemisphere lesion was significantly reduced in both hemispheres, whereas only the left hemisphere showed reduced white matter integrity in the child with the left hemisphere lesion. We relate our findings to theories of neuroplasticity, hemispheric specialization, and developmental cognitive neuroscience.

INTRODUCTION

It has long been observed that relative to adults with late-acquired unilateral stroke, children with early occurring, perinatal stroke (acquired in the third trimester up until one month postnatally) show attenuated effects of the lesion, often resulting in nearly typical cognitive functioning (Bates et al., 2001; Kempler, Van Lancker, Marchman, & Bates, 1999). This difference is especially striking when comparing language performance in children and adults with left hemisphere focal lesions. Often, adults with late-acquired left-hemisphere lesions demonstrate dramatically impaired language at almost every level of linguistic processing, including word-finding and naming difficulties, impaired verbal fluency, and reduced comprehension, especially of syntactically non-canonical sentence structures (e.g. Blumstein, Goodglass, Statlender, & Biber, 1983; Tatemichi et al., 1994). Adults with right hemisphere lesions also show impairments in language processing, but these deficits tend to be more subtle, often affecting social and pragmatic language relatively more than linguistic structure (Blonder, Bowers, & Heilman, 1991; Bryan, 1988; Foldi, 1987; Gardner, Brownell, Wapner, & Micheow, 1983). In striking contrast, following an initial language delay (Bates, Thal, Trauner, & Nass, 1997; Marchman, Miller, & Bates, 1991; Thal et al., 1991), children with left hemisphere perinatal stroke often show conversational language that is comparable to their typically developing peers by about five-to-seven years of age (Bates et al., 2001; Reilly, Losh, Bellugi, & Wulfeck, 2004; Reilly, Bates, & Marchman, 1998; Vicari et al., 2000), and most investigations of young pre-school and school-aged children with perinatal stroke find no side-of-lesion

differences in language performance (Bates et al., 2001; Chapman, Max, Gamino, McGlothlin, & Cliff, 2003; Demir, Levine, & Goldin-Meadow, 2010; Reilly et al., 1998; 2004).

Although the competent conversational language of children with perinatal stroke attests to the brain's remarkable ability to utilize alternative neural circuitry to support language following an early insult, there is evidence that this alternate organization is less robust than the typical organization. Specifically, while children with perinatal stroke perform similarly to their typically developing peers in conversational language, they show decreased performance on more restrictive tasks that probe language components in isolation (Ballantyne, Spilkin, Hesselink, & Trauner, 2008; Ballantyne, Spilkin, & Trauner, 2007). Even in these restrictive testing environments, however, which elucidate language vulnerability in children with perinatal stroke, children with both left and right hemisphere lesions perform poorly, and indeed these investigations also consistently find no side-of-lesion differences. It is not until after the school-age years (in pre-adolescence and adolescence), that side-of-lesion differences are noted in a narrative context. In this context, both children with right and children with left hemisphere lesions perform worse than their typically developing peers, but the children with left hemisphere lesions demonstrate the most severe deficits in morphology and use of complex and diverse syntax relative to children with right hemisphere lesions and typically developing children (Reilly, Wasserman, & Appelbaum, 2012). Language performance from children with right and children with left perinatal stroke also provides a window into the brain's early organization and extent of hemispheric specialization for language. On one side of this debate, Hemispheric Specialization/Innate Predetermination Theory (e.g. Chomsky, 1975; Fodor, 1983; Pinker, 2007) minimizes the role of development and suggests that language is innately and irreversibly constrained to the left hemisphere. This theory would hypothesize that typical language function could never exist following a left hemisphere lesion, no matter how early in development the injury took place. In contrast, Equipotentiality Theory, proposed by Lenneberg (1967),

emphasizes the role of development and proposes that initially both hemispheres are equally suited to support language functions. Thus, this view would hypothesize that following an early lesion, the intact hemisphere would have the capacity to assume language functions, resulting in normal language.

Thus far in the research, there is strong evidence for neural plasticity for language from children with left hemisphere perinatal stroke. Interestingly, however, there is also evidence that there are limits to neural plasticity. Taken together, previous research suggests that children with right and children with left perinatal stroke show an initial delay in lexical acquisition and vocabulary growth, followed by subsequent development in the school-age years where conversational language is comparable to their typically developing peers (though performance on standardized measures of language components is below norms). In adolescence, children with right and left perinatal stroke demonstrate narrative performance that is below their typically developing peers, but the adolescents with left hemisphere lesions show the most impaired performance relative to the other two groups. This general profile is based on cross sectional investigations of language performance in groups of children with right and children with left hemisphere perinatal stroke compared to typically developing children, and thus we can only infer that this pattern would be observed developmentally in the same individual over time. It is also important to note that each of these studies on language performance in children with perinatal stroke notes greater variability in language performance in perinatal stroke groups relative to typically developing groups (Stiles, Reilly, Levine, Trauner, & Nass, 2012). This increased variance has implications both theoretically and practically.

Theoretically, neuroimaging research on language in the developing brain suggests that in comparison to adults, children use additional neural regions to accomplish the same language tasks, and this distributed organization becomes more tuned and localized with age. Lexico-semantic processing, for example, appears to become more strongly left lateralized and more

reliant on anterior cortical regions later in childhood and in adolescence (e.g. Balsamo et al., 2002; Booth et al., 2003; Brown et al., 2005; Gaillard et al., 2001; Gaillard et al., 2003; Holland et al., 2001; Schlaggar, Brown, Lugar, Visscher, & Miezin, 2002; Schmithorst, Holland, & Plante, 2007; Szaflarski, Holland, Schmithorst, & Byars, 2006). Investigations that consider both developmental age *and* linguistic proficiency, however, suggest that this localization process is not merely dependent on age, but rather is highly interdependent on the language development itself; as linguistic systems become more robust, the neural substrates supporting language processes become more specialized and presumably more efficient (e.g. Brown et al., 2005; Mills, Coffey-Corina, & Neville, 1993; Schlaggar et al., 2002, add others). Since children with perinatal stroke do not have access to the same neural tissue as typically developing children, it is likely that during this developmental process, they must co-opt alternative neural substrates to support language. Some investigations suggest that the remaining healthy neural tissue in the left hemisphere takes on language functions (Liegeois et al., 2004), while others have found homologous right hemisphere regions support language processing (Guzzetta et al., 2008; Staudt, 2002). Although it remains a matter of debate whether the reorganization for language in children with left hemisphere lesions involves use of contralateral homologous brain regions (Jacola et al., 2006; Staudt, 2002; Tillema et al., 2008) or of ipsilateral surrounding regions (Raja Beharelle et al., 2010), it is likely that any alternate neural organization manifests behaviorally as increased linguistic variability. Group investigations have the benefit of achieving results that can be generalized to a population, but these pooled or averaged effects mask individual variability, which may add to our understanding of neural substrates that have the capacity to support language.

On the practical side, there are many ways to assess language performance, and different types of language assessments often illustrate a distinct aspect of the language profile. As mentioned above, highly constrained language assessments that evaluate linguistic components in

isolation consistently find school-aged children with perinatal stroke to perform significantly below their typically developing peers; such assessments would conclude that “language” is impaired. On the other hand, less restrictive, conversational language assessments reliably demonstrate competent language in conversational and narrative contexts, and would conclude that “language” is functional. These seemingly opposing results demonstrate the importance of using multiple levels of linguistic analysis when assessing language performance, especially in children with neurodevelopmental disorders. However, since results from restrictive and open-ended language assessments are often from different investigations, it is impossible to explore relations between language components in isolation (e.g. phonological and syntactic processing) and the language system as a whole (e.g. conversational and narrative performance) within an individual. Analyzing language at multiple levels of complexity (phonological, lexical, syntactic, and narrative levels) in two children with unilateral perinatal stroke relative to a group of typically developing children will enable us to address and pose questions regarding the relations between these levels (and their underlying neural correlates) within individual children.

Furthermore, most investigations of language in children with perinatal stroke focus on receptive and/or expressive spoken language, but in the older school-age years, written language becomes increasingly important, raising questions as to how the language system will support written language both receptively (i.e. reading) and expressively (i.e. writing). We have seen from previous research that language processing in school-aged children with perinatal stroke is proficient but vulnerable. When these children acquire a new modality of linguistic communication (i.e. reading and writing), will we observe a similar profile? Alternatively, will the fragility of the language system be insufficient as a platform for written language? In this case, we would expect more dramatic impairments in the written relative to the spoken modality. Investigating language at increasing levels of complexity (e.g. phonological, morphological, lexical, syntactic, narrative) within the same participants across speaking, reading, and writing

modalities, will allow us to begin to address questions about the relations between levels of linguistic complexity and language modalities in individuals with perinatal stroke.

Thus far we have focused our discussion on the importance of implementing a detailed and comprehensive linguistic assessment to better characterize language behavior in children with perinatal stroke. We now turn our attention to the importance of applying that same depth of analysis to better characterize the brains of these children. Differences in size, shape, and location of the lesion may account for much of the between-subjects variability reported in investigations of children with perinatal stroke, but participants are often grouped together based on simplistic, broad lesion categories such as anterior versus posterior and right versus left. Because each lesion is unique, it is impractical in a group study format to attempt to create subgroups based on highly specified lesion characteristics, as attempting to match for multiple characteristics (lesion size, location, extent of damage to cortical/subcortical structures) significantly reduces statistical power. In a case study format, however, it is possible to map and characterize the lesion in detail, which can then be discussed in relation to that individual's language behavior. With this aim, here we implement three-dimensional multimodal structural neuroimaging techniques which allow us to more precisely characterize the location, extent, and tissue types involved in the lesion. Combining the detailed lesion analysis with an exhaustive linguistic assessment on a case-by-case basis will provide an opportunity to explore the brain behavior relationship, as well as the extent and limits to neural plasticity.

Another source of variability in this population that has been relatively unexplored is the condition of the intact hemisphere. Often it is assumed that the intact hemisphere is relatively unaffected by the contralateral lesion, and behavioral performance is considered in relation to the lesioned hemisphere alone. It is also common to look at linguistic performance in only children with left hemisphere lesions (relative to typically developing children), which has the implicit assumption that neuroplasticity for language is chiefly seen when the lesion occurs in the left

hemisphere. The functional neuroimaging studies discussed above, however, reveal that the right hemisphere plays a large role in language processing early on. Thus, disruption to neural tissue in the right hemisphere will very likely impact the development of neural correlates for language.

Here we implement multimodal structural imaging to directly compare the tissue properties of the white matter fiber tracts within *both* hemispheres (in one child with a left hemisphere lesion and one child with a right hemisphere lesion) as a means to explore whether the structural integrity of the presumably *intact hemisphere* can account for additional variance in language performance.

In summary, in this case study of two school-aged females with perinatal stroke, we present detailed linguistic profiles in speaking, reading, and writing modalities, alongside detailed quantitative multimodal imaging measures of white matter microstructure in order to further explore the relationship between language behavior and the brain. Specifically, these two children with perinatal stroke allow us to consider the relations between: 1. Language modalities (speaking, reading, and writing); 2. Levels of linguistic complexity (phonological, lexico-semantic, syntactic, narrative); and 3. Neuroanatomical characteristics (lesioned and intact hemispheres) and language performance.

METHODS

PARTICIPANTS

Two female participants (9;0 and 10;11) with perinatal stroke, and 15 typically developing (TD) children (aged 7-12, mean age = 9.48; SD = 1.87) participated in the present experiment. The same 15 TD children were included for both behavioral and neuroanatomical analyses. All participants were screened using the following measures: Parent history questionnaire, standardized cognitive and intelligence tests (described below), and a neurological exam. All children in the TD group had normal developmental and medical histories and normal neurological exams, and all scored within the normal range (85 - 115) on standardized tests of intelligence (mean Full Scale IQ from the Wechsler Abbreviated Scale of Intelligence (WASI)

(Wechsler & Chen, 1999) = 114.66; SD = 11.59), language, and academic functioning, and none had a history of chronic medication use.

Case L Description: Left Hemisphere Lesion

Case L is a female child with encephalomalacia in the left temporal-parietal lobe measuring approximately 3.2 by 2.9 by 2.5 cm (see Figure 4.1). At the time of the present investigation, Case L was 9;0. The MRI scan was read for clinical purposes by a board-certified pediatric neuroradiologist and revealed evidence of compensatory enlargement of the left ventricle and thinning of the corpus callosum, and atrophy of the left cerebral peduncle, pons, and medulla from Wallerian degeneration. The WASI revealed an IQ of 72, which is slightly below the lower “normal” range and two standard deviations below average. Results from other cognitive and language measures will be discussed in detail in subsequent sections. Case L was a full-term infant with normal birth weight, who had some fetal distress during delivery, but no resuscitation was required. She did not experience any problems in the newborn period, and was diagnosed at 7 months of age when a mild hemiparesis was noted. Her developmental milestones were slightly delayed, and she sat at 9 months and walked independently at 18 months. Case L developed partial complex seizures beginning at age 4 ½ years, and has had 4 seizures total, the last around age 8 years. The neurological exam at 8 ½ years of age, revealed a mild right inferior quadrant visual field defect on confrontation examination; a moderate right hemiparesis with arm more affected than leg; and mild cortical sensory deficits on the right side with impaired proprioception and graphesthesia. She is currently receiving physical, occupational, and speech/language therapy.

Case R Description: Right Hemisphere Lesion

Case R is a female child, aged 10;11 at the time of testing, who was found to have a large, right hemisphere frontal porencephalic cyst, which communicates with the frontal horn of the lateral ventricle, affecting anterior perisylvian regions. The lesion measures approximately

3.18 by 3.61 by 3.69 cm (see Figure 4.2). The clinical neuroradiological assessment revealed areas of gliosis, which are seen in the periventricular white matter bilaterally and surrounding the cyst, and a diffuse thinning of the corpus callosum. Case R demonstrates an IQ (from the WASI) of 73, which is very similar to Case L, and also falls slightly below the low-normal range. Other cognitive and linguistic assessments will be discussed below. Case R was born prematurely (33.5 weeks) and had a right intraventricular and parenchymal hemorrhage that resulted in the porencephaly. Developmental milestones were reached within the normal times, and she walked at 15 months. The child developed seizures at the age of 5 ½ years and had several seizures per year until the age of 12. Neurological examination at the age of 11 years revealed no visual field defect, mild left hemiparesis with some use of her left hand and no significant leg involvement; and no sensory impairments were detected on the exam. Case R is also currently receiving physical, occupational, and speech/language therapy.

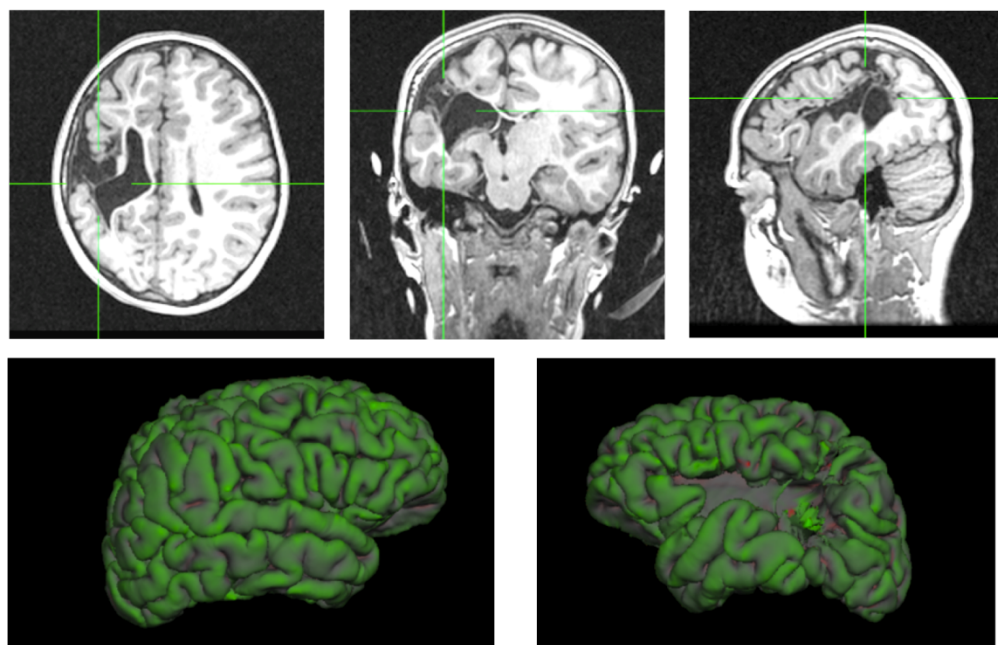


Figure 4.1: Shows T1-weighted, inversion recovery spoiled gradient echo (IR-SPGR) MRI scans in the axial (top left) coronal (top center) and sagittal (top right) planes, and the three-dimensional cortical surface reconstruction of the right (bottom left) and left (bottom right) hemispheres for Case L. Case L presents with a left temporal-parietal lesion (9;0 at time of scan).

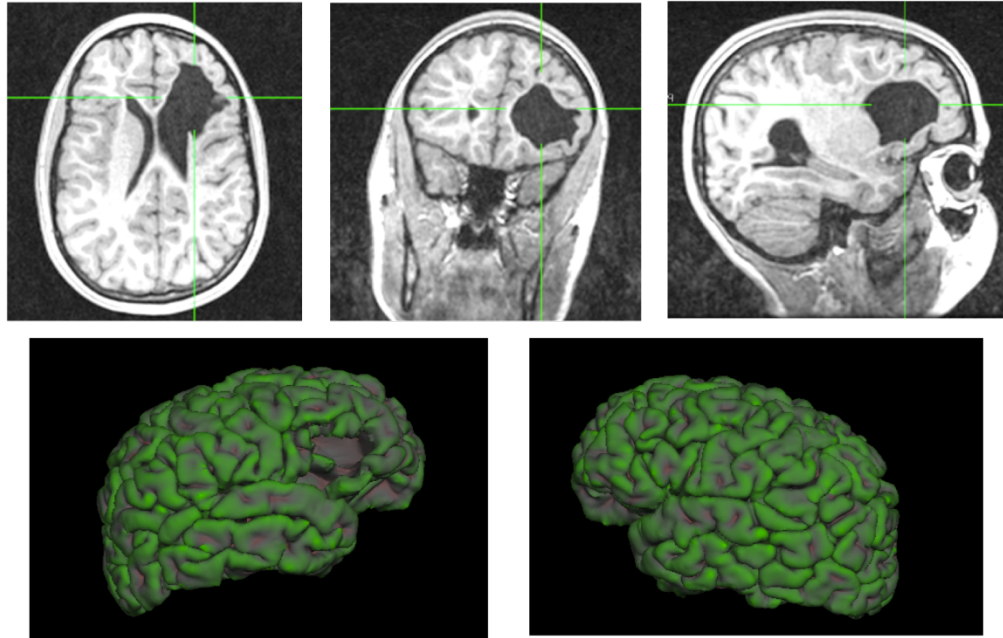


Figure 4.2: shows T1-weighted, inversion recovery spoiled gradient echo (IR-SPGR) MRI scans in the axial (top left) coronal (top center) and sagittal (top right) planes, and the three-dimensional cortical surface reconstruction of the right (bottom left) and left (bottom right) hemispheres for Case R. Case R presents with a right frontal porencephalic lesion (10;11 at time of scan).

MULTIMODAL STRUCTURAL MAGNETIC RESONANCE IMAGING

Image Acquisition

All research participants underwent multimodal structural magnetic resonance imaging (MRI). MRI data were obtained on a 1.5 Tesla GE Signa HDx 14.0M5 Twin Speed scanner (GE Healthcare, Waukesha, WI) using an eight-channel phased array head coil. High resolution anatomical data were acquired with a single 3D inversion recovery spoiled gradient echo (IR-SPGR) T1-weighted volume, acquired with pulse sequence parameters optimized for maximum gray/white matter contrast (TE=3.9 ms, TR=8.7 ms, TI=270 ms, flip angle=8°, TD=750 ms, bandwidth=±15.63 kHz, FOV=24 cm, matrix=192×192, voxel size=1.25×1.25×1.2 mm). Diffusion weighted imaging (DWI) data for white matter tissue property quantification were acquired using single-shot echo planar imaging with isotropic 2.5 mm voxels (matrix size = 96x96, field of view = 24 cm, 47 axial slices) covering the entire cerebrum, cerebellum, and

brainstem without gaps. Three volumes were acquired with 51 diffusion gradient directions using b-values of 600, 800, and 1,000 mm²/s, each with an additional b = 0 volume. For use in nonlinear B0 distortion correction, two additional volumes were acquired with one b = 0 volume and a single diffusion direction (b = 800 mm²/s), with either forward or reverse phase-encode polarity.

For T1-weighted volumes, real-time, prospective motion tracking and correction (PROMO) was implemented using spiral navigator scans and an extended Kalman filter (EKF) algorithm (Gelb, 1974), as described previously for use in 3D pulse sequences (White et al., 2010). PROMO has been shown to significantly reduce head motion artifacts, improve the reliability of quantitative morphological measurements, and increase the radiological clinical utility of MRI data collected in children (Brown et al., 2010; Kuperman et al., 2011).

Image Preprocessing, Reconstruction, and Segmentation

Image files in DICOM format were transferred to a Linux workstation for viewing, rating, and automated cortical reconstruction. The T1-weighted volumes were rigid-body registered and realigned into a common stereotactic space. Heterogeneities in image intensity were corrected online using GE's calibration normalization procedure as well as offline using the FreeSurfer software suite (version 3.0.5; <http://surfer.nmr.mgh.harvard.edu>). Gradient coil nonlinear warping was corrected using tools developed through the Biomedical Informatics Research Network on morphometry. Three-dimensional reconstruction of the cortical surface and delineation and segmentation of the white matter and pial surfaces was conducted using well-documented, histologically validated algorithms (Dale, Fischl, & Sereno, 1999a, 1999b; Fischl, Salat, & Dale, 2002). This automated procedure involves the segmentation of gray and white matter, the tessellation of the gray/white matter boundary, the inflation of the folded, tessellated surface, and smoothing of the cortex with an automated correction for topological defects. Once

these surfaces are delineated, any of several parcellation schemes can be applied, which automatically assign anatomical labels to each vertex on the cortical surface for every individual within the standardized surface-based atlas space (e.g. Desikan et al., 2006; Destrieux, Fischl, Dale, & Halgren, 2010). Here, for anatomical simplicity the cortical surface was delimited into 74 distinct cortical regions of interest per hemisphere based on the sulcal and gyral boundaries, as has been used previously in several studies measuring cortical surface area, thickness, and volume down to the age of three years (Brown & Jernigan, 2012; Brown et al., 2012; Fjell et al., 2012; Walhovd et al., 2012).

Preprocessing of the diffusion-weighted images was performed according to previously described procedures (Hagler et al., 2009). In brief, preprocessing of diffusion images included the correction of subject motion, correction for image distortion in the diffusion weighted volumes due to eddy currents created by the gradient coils (Jovicich et al., 2006), magnetic susceptibility artifacts, and differences in intensity scaling. Images were resampled to a higher resolution using linear interpolation (1.875 mm³ isotropic voxels).

White matter tracts were identified and segmented based on a probabilistic atlas with information about the location and orientation of a comprehensive set of cerebral fiber tracts. The fiber atlas was previously constructed by manually generating a full set of tracts in 21 subjects individually. In each subject, diffusion tensors were derived from the DTI dataset and three eigenvectors and corresponding eigenvalues were calculated from the tensor matrix. FA, ADC, and TDC were each calculated based on these eigenvalues. With the use of DTI Studio software (Laboratory of Brain Anatomical MRI, Johns Hopkins Medical Institute) and eigenvector maps, a deterministic fiber tracking method generated streamlines from each voxel. Manually drawn ROIs identified streamlines that connected known anatomical sites and formed known tracts. For each subject, the T1-weighted dataset was registered to a common space and the transformation matrix was applied to the fiber streamline maps for coregistration and to produce an average fiber

streamline map. The average fiber streamline maps indicate the relative probability that a fiber tract is positioned in a given space. For each participant, the eigenvector maps were rotated, aligned and resampled to coregister to the atlas. The fiber orientation probability distribution at each voxel was then assessed for the likelihood that a voxel was part of a particular fiber path given the diffusion values. This automated probabilistic method has previously been described in detail (Hagler, et al., 2009). In each fiber tract, the mean fractional anisotropy (FA), mean diffusivity (MD), and mean T2-normalized signal intensity (T2N) over the extent of the white matter pathway were calculated individually for each subject. FA is a measure of the directionality of water movement within tissue and generally increases with maturation (and with increasing myelin content on fiber tracts). MD reflects the overall magnitude of water diffusion (regardless of directionality) within a specified tissue region of interest. It generally decreases with maturation within gray and white matter. T2N is associated with myelin deposition and tissue health/integrity in white matter fiber tracts (Glasser & Van Essen, 2011; Hagmann, De Vita, Bainbridge, Gunny, Kapetanakis, Chong, Cady, Gadian, & Robertson, 2009), and also generally decreases with age/maturation. White matter fibers measured in this study are short- and long-range longitudinal, crossing, and association fibers that previously have been implicated in many different aspects of language and cognitive functioning.

MATERIALS

Testing Battery

Participants were administered a battery of standardized and experimental measures to probe linguistic processing at multiple levels of linguistic complexity. To provide a more in depth exploration of communication in these individuals with left and right perinatal focal lesions, we assessed linguistic processing in speaking, writing, and reading domains at the phonological, semantic, syntactic, and narrative/discourse levels of language. For information about assessment measures for each level of linguistic processing, see Table 4.1.

Table 4.1: Assessment Information for each Evaluated Level of Linguistic Processing

Language Assessments			
	Speaking	Writing	Reading
Phonological	PAT ¹ (Rhyming; Blending, and Isolating Phonemes)	Spelling of Sounds (WJ ²)	Word Attack (WJ ²)
Lexico-semantic	EOWPVT ³		RWS ^{6*}
	WASI Vocabulary ⁴	WPV ^{5*}	Letter-word ID (WJ ²)
	WASI Similarities ⁴		
Syntactic	Recalling Sentences (CELF-4 ⁷)	Writing (WJ ²)	Reading Fluency (WJ ²)
	Formulating Sentences (CELF-4 ⁷)		
Narrative	Spoken Personal Narrative ^{8*}	Written Personal Narrative ^{8*}	—

¹ Phonological Abilities Test (Muter, Hulme, & Snowling, 1997)

² Woodcock Johnson Test of Achievement- Third Edition (Woodcock, McGrew, & Mather, 2001)

³ Expressive One Word Picture Vocabulary Test (M. F. Gardner, 1990)

⁴ Wechsler Abbreviated Scale of Intelligence (Wechsler & Chen, 1999)

⁵ Written Picture Vocabulary: Non-standardized spelling assessment. Children are asked to spell words from pictures (no spoken input).

⁶ Recognizing Written Synonyms (Polse & Reilly, 2012)

⁷ CELF-4: Clinical Evaluation of Language Fundamentals- Fourth Edition (Semel, Wiig, & Secord, 2003)

⁸ Spoken and Written Personal Narrative: Procedure adapted from Reilly et al. (1998).

* Indicates a non-standardized assessment measure

PHONOLOGICAL PROCESSING ASSESSMENTS

Speaking

Phonological Abilities Test (PAT) is a standardized phonological processing test that assesses metacognitive phonological processing. Four subtests were administered from the PAT: Rhyming Detection, Rhyming Production, Phoneme Blending, and Phoneme Isolation. The Rhyme Discrimination subtest assesses the child's ability to identify rhyming words presented in pairs. For example, the child may be presented with the words *book* and *look* and is asked to make a judgment about whether or not the two words rhyme. Ten word pairs were aurally presented, and all word pairs consisted of one or two syllables. In the Rhyme Production subtest, children listened to 10 words produced by the experimenter, and following each word were asked

to produce a rhyme. For example, a child may be given the word *cat* and asked to say a word that rhymes with *cat*. The Phoneme Isolation subtest assesses the child's ability to segment a word into its phonemic parts. The child is asked to identify either the initial (10 items), medial (10 items), or final (10 items) phoneme in a word (e.g. *What's the ending sound in the word wish?* or *Tell me the middle sound in drop*). Lastly, the Phoneme Blending test evaluates a child's ability to assemble a series of phonemes into a word. In this test, the experimenter presents a series of isolated phonemes and asks the child to make them into a word (e.g. /k/ /æ/ /t/ *can you make those sounds into a word?*) Words varied in length from two to six phonemes.

Writing

Written phonetic and phonological processing was assessed using the Spelling of Sounds subtest (subtest 20) of the Woodcock Johnson Test of Achievement- Third Edition (WJ-3) (Woodcock, McGrew, & Mather, 2001)). In this measure, children are asked to write nonsense words from dictation. This assessment probes written phonological processing devoid of semantic processing, as the words children are asked to write are non-words, and thus carry no meaning. For example, children may be asked to write the nonsense word *splunted*.

Reading

Phonological processing was assessed in the reading domain using the WJ-3 subtest 13 (Word Attack), in which children are asked to read non-words aloud. Since children are only reading non-words, this test assesses their orthographic to phonological processing skills without utilizing semantic representations. Words corresponded to the orthographic rules of English, for example, *plurp* and *fronkett*. Later test items were more difficult than early test items.

LEXICO-SEMANTIC PROCESSING ASSESSMENTS

Speaking

Expressive lexico-semantic knowledge was assessed using the Expressive One Word Picture Vocabulary Test (Gardner, 1990), which is a norm-referenced, picture-naming test.

Children are presented with a series of pictures of increasing difficulty and are asked to name the picture. Basal and ceiling scores are established. The WASI Vocabulary subtest was also administered, as a less-structured measure of vocabulary. In this subtest, children are presented with a word (no picture) and asked to define the word. For example, *Can you tell me what an alligator is?* Words become more difficult as the test goes on. The last measure of lexico-semantic processing was the Similarities subtest of the WASI, which assesses a child's semantic representations, and her ability to identify similarities across representations. In this subtest, a child is given two words and asked to identify how the two are similar. For example, *pillow* and *cloud*. Items become more abstractly related as the test progresses.

Writing

The semantic contribution to written lexico-semantic processing was evaluated using the Spelling subtest from the WJ-3. In this subtest, children are asked to write real words from dictation. In addition, to assess lexico-semantic processing in the writing domain without a phonological model, children were administered the experimental Written Picture Vocabulary test (WPV), which includes a series of 20 pictures, and children are asked to write the label for each picture without a verbal prompt from the experimenter. Earlier test items are frequent and conform to spelling conventions, whereas later items are less frequent and contain unusual phonological to orthographic mappings. For example, the first item on this test is *boat*, and the last item is *ostrich*.

Reading

Lexico-semantic processing in the reading domain was assessed using a standardized and an experimental reading paradigm. The standardized assessment was the Letter-word Identification subtest from the WJ-3. In this subtest, children are asked to read letters and words aloud, and the difficulty of the words increase as the test goes on. In contrast to the Word Attack subtest above, this test assesses the child's ability to decode real words (i.e. includes semantic

processing). We also administered a non-standardized lexico-semantic reading measure, Recognizing Written Synonyms (RWS). This measure pits orthographic and semantic processing against one another (for more specifics on this reading measure, see Polse & Reilly, 2012). Children are presented with three words on a touch screen computer monitor (a target word at the top of the screen (e.g. BOAT), and a match and a foil beneath) and are asked to identify a semantic match to the target word. The semantic match can be either a synonym or an exact word match. There are four conditions: The Baseline condition requires only coarse-level orthographic processing and does not require semantic processing (e.g. BOAT, GAME), the Orthographic condition requires fine-level orthographic processing, but no semantic processing (e.g. BOAT, BOOT), the Semantic condition requires semantic processing but only coarse-level orthographic processing (e.g. BOAT, SHIP), and the Orthographic-semantic condition requires fine-level orthographic processing as well as semantic processing (e.g. SHIP, BOOT).

SYNTACTIC PROCESSING ASSESSMENTS

Speaking

Syntactic processing was assessed using the Recalling Sentences subtest of the Clinical Evaluation of Language Fundamentals-Fourth Edition (CELF-4) (Semel et al., 2003). In this subtest, children are asked to listen to a sentence and repeat it. Sentences increase in length and syntactic complexity. For example, an early item on this measure is *the tractor was followed by the bus* and a later item is *the math teacher sorted, labeled, boxed, and delivered the calculators*. This assessment measures both receptive syntactic processing as well as syntactic memory, as the child must keep the sentence in memory before he/she repeats it. To assess syntactic processing more independent of syntactic memory, the Formulating Sentences subtest of the CELF-4 was administered. In this assessment, children are given a word or phrase and asked to make a sentence utilizing that word or phrase. Words and phrases become more difficult as the test proceeds. For example, an early item of this subtest is *make a sentence using the word "children"*

and a later item is *make a sentence using the phrase “as a consequence.”* This assessment measures productive syntactic ability, as the child must manipulate the given word or phrase into a sentential context.

Writing

Syntactic processing in the writing domain was assessed using the Writing subtest of the WJ-3 (subtest 11). Analogous to syntactic processing in the Formulating Sentences subtest in the spoken domain, children were asked to provide a written description of a picture using the provided words or phrases. For example, an early item on this test shows an illustration of a cat, and the experimenter says *this is a ...* and the child writes the appropriate word (cat). In a later item, the child is asked to *write a good sentence using the words “despite her anger.”* This subtest evaluates the child’s syntactic processing ability in the written domain.

Reading

Syntactic processing in the reading domain was assessed using the Reading Fluency subtest of the WJ-3 (subtest 2). In this subtest, children are given a list of sentences that are either true or false, and are asked to read them silently and circle YES or NO for as many sentences as they can in three minutes. Since children do not need to read these sentences aloud, this is not a measure of phonological processing, but rather assesses the child’s reading speed and comprehension. Sentences become longer and more complex in later items on the test. For example, an early item is *a bird can fly* and a later item is *people may use a map to find certain locations*. This subtest provides a measure of syntactic comprehension in the reading domain.

NARRATIVE PROCESSING ASSESSMENTS

Speaking

Telling a personal narrative requires linguistically organizing (in a temporal sequence) a series of connected events including a setting, a problem, and a solution. This draws not only on the linguistic processing skills assessed above, but also on social pragmatic skills. Thus, the

narrative portion of this assessment provides a rich measure of a child's ability to integrate linguistic and communicative components. In this assessment, adapted from Reilly and colleagues (1998), children were given the prompt: *People disagree and they get into fights or arguments all the time. I'm sure there has been a time when you had an argument or disagreement when someone hurt your feelings. I'd like you to tell me about a time when someone made you mad or made you sad. Maybe it was a friend or your brother or sister. Tell me how it started, what happened and how it all ended. Take some time to think about it and when you are ready, go ahead and start.* Children were videotaped and then their narratives were transcribed and coded offline using the CHAT from the Child Language Data Exchange System (CHILDES; MacWhinney, 1991). Narratives were coded for:

Length: The number of semantic propositions.

Morphological Errors (Presented as a proportion of errors over total Propositions):

Number agreement errors (e.g. she has lotsa toy[])

Subject-verb agreement errors (e.g. he have his own gameboy)

Complex Syntax (Presented as a proportion of complex syntax over total Propositions):

Coordinates (e.g. he pushed me and I kicked him)

Verb Complements (e.g. he knew that h'ed get in trouble)

Adverbial Clauses (e.g. Then she got mad because she didn't agree with me)

Relative Clauses (e.g. And so then a long time ago my dad gave me the same phone he was holding, but it couldn't call)

Passives (e.g. He kind of did get out so he was banned for the rest of the month)

Evaluation (Presented as a proportion of evaluative markers over total Propositions):

Mental Markers (e.g. think, wonder, decide)

Emotional Terms (e.g. sad, angry, mad)

Causal Markers (e.g. because, so, since)

Intensifiers (e.g. very, really, dreadfully)

Setting (out of 4 possible components):

Time (e.g. yesterday..., one time...)

Place (e.g. we were at school, and...)

Characters (e.g. my sister and I were playing...)

Situation (e.g. we were playing hide and seek...)

Total Narrative Composition (out of 4 possible components):

Setting: Any of the setting components

Problem: The child presented a conflict

Response: The child presented a response to the conflict

Resolution: The child communicated how the conflict ended

Writing

Following the child's Spoken Narrative, children were given two sheets of lined paper and a pen, and were told: *Now that you have told me your story, I would like you to write it down.* After children completed their written narrative, they were asked to read it back to the experimenter, and correct any errors they found. The written narratives were transcribed and coded for the same measures as the spoken narrative, in addition to Spelling Errors, which are presented as a proportion of spelling errors over total words in the written narrative.

Reading

There is no measure for Personal Narrative in the reading domain.

BEHAVIORAL ANALYSIS

In order to obtain a metric of performance for the two cases relative to the typically developing group, raw scores from all the language assessment measures were converted to *z*-scores. Because our main aim was to address language components relative to one another (and not within language levels), we then created composite scores for language levels for which there was more than one assessment administered. For example, for phonological processing in the spoken domain, we averaged the *z*-scores from the individual measures of the PAT (Rhyming, Blending, and Isolating) into one composite index score for *spoken phonological processing*. Similarly, for lexico-semantic processing in the spoken domain, we averaged *z*-scores from the EOWPVT, WASI Vocabulary, and WASI Similarities as our index score for *spoken lexico-semantic processing*. For linguistic levels with only one assessment, that score was used as the index value. For information regarding which measures contributed to these linguistic index scores, see Table 4.1, and to view results from the complete testing battery (all individual measures), see Appendix A.

BRAIN ANALYSIS

In order to create a metric of the structural integrity and tissue properties of cerebral white matter fiber tracts for the two cases relative to the typically developing group, a composite index was computed for all fiber tracts for both hemispheres in every subject. For each white matter fiber tract identified using automated tractography (e.g. left superior longitudinal fasciculus, right fornix, forceps minor), the three measures of diffusivity and signal intensity (FA, MD, and T2N) were converted to the same normalized scale and combined into an equally weighted composite index using *z*-scores. For normalization, the scales and signs of the three measures were made consistent with one another with regard to whether they reflect developmentally "immature" (low FA, high MD, high T2N) versus "mature" (high FA, low MD, low T2N) states (Brown et al., 2012b). Then, the mean distribution of values for this composite

index was calculated for the 15 individuals in the TD comparison control group. In this way, each tract for the two cases could be compared probabilistically to the distribution of structural integrity values for the TD group in standard deviation units, for both the lesioned and non-lesioned hemispheres, similar to our behavioral comparison.

RESULTS AND DISCUSSION

We have gathered detailed language and neuroanatomical results from a group of 15 typically developing children and two children with unilateral perinatal stroke (one left and one right), with the goal of considering relations between linguistic levels and modalities, as well as between behavior and the brain. Before we explore these complex issues, however, we first summarize the general trends from behavioral and imaging results from the two children with perinatal stroke relative to their typically developing peers.

Behavioral results from the two cases replicate previous findings from group investigations of children with perinatal stroke in two primary ways: First, children with perinatal stroke demonstrate functional language performance which, especially in Case L, is indicative of the brain's remarkable plasticity to utilize alternative neural circuitry to support language processes (e.g. Bates et al., 2001). Second, with a few exceptions, children with perinatal stroke performed worse on restrictive language assessments that probe isolated language components relative to measures that assessed the use of language in communicative contexts (Ballantyne et al., 2008, 2007; Reilly et al., 1998; Reilly et al., 2004). Contrary to previous group findings from younger children with perinatal stroke (which report no side-of-lesion differences), the current exhaustive linguistic assessment had the sensitivity to isolate unique strengths and weaknesses in the two participants, which differ from one another. Importantly, these differences exist amidst a large degree of overlap in overall language performance. The similarities, and then differences in language performance will be discussed in turn.

Although the two children present with non-overlapping lesions, they showed many similarities in their linguistic profiles (with a few notable exceptions, discussed below). For both children, performance on all isolated levels of linguistic processing that were assessed using standardized tests hovered around two standard deviations below the mean, or just below the normal range (See Figure 4.3). This finding is in sharp contrast to the highly distinctive language performance we would expect from two adults with a right and left hemisphere lesion, in which the adult with a left hemisphere perisylvian lesion would be likely to perform much worse on measures of structural language relative to an adult with a right hemisphere lesion. The similar language profiles from the two cases provide evidence against a strict hemispheric specialization perspective, and emphasize the distributed organization and the importance of the developmental process in the establishment of language in a child's brain. That is, if the left hemisphere were capable of supporting the development of language by itself, we would expect Case R to show language performance that was similar to her typically developing peers. However, if, as many pediatric functional neuroimaging studies of language suggest, some language functions in childhood are more bilaterally distributed, and as part of the normal developmental process require participation of the right hemisphere, we would expect degraded (though functional) performance from *both* the child with the right and the child with the left hemisphere lesion, as both have access to some (though not all) of the tissue implicated in this distributed, bilateral neural organization for language.

A second similarity in the language profiles of the two cases is that both performed better on the quasi-unstructured language assessment (telling a personal narrative) than on standardized tests of language components (see Figure 4.4). Specifically, both children told stories that were comparable in length to their typically developing peers; both included evaluative devices (such as mental verbs, intensifiers, and causal devices); and both included a setting to orient her listener to the story environment (see Table 4.2 for examples). These story elements demonstrate that the

two children with perinatal stroke are good *communicators* (i.e. have the ability to use language in a social context), even though structural language components are quite impaired. Interestingly, though performance on these language-use measures was similar to their typically developing peers overall, both children showed severe deficits in isolated linguistic elements within the narrative context. Specifically, Case R made more spelling errors than Case L and the TD group, and Case L made significantly more morphological errors than Case R and the TD group (findings from Case L are similar to Reilly et al., 2012). These isolated differences are discussed in depth below.

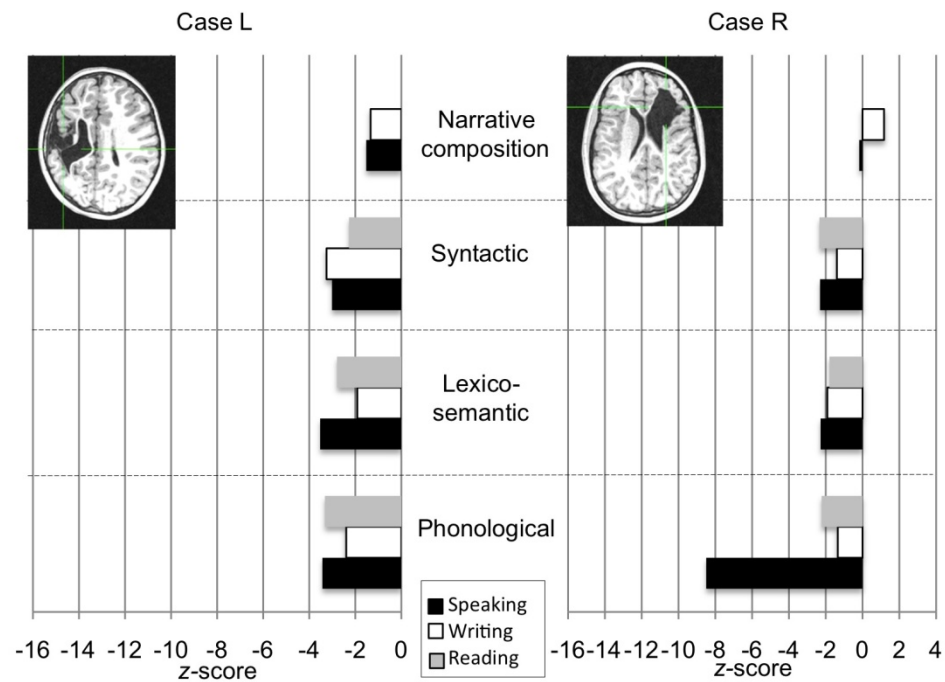


Figure 4.3: Displays the z-scores (relative to the 15 typically developing children) for Case L (left) and Case R (right) on our index metric of linguistic processing at increasing levels of complexity (phonological, lexico-semantic, syntactic, and narrative) across speaking (black bar), writing (white bar), and reading (gray bar).

First, Case R demonstrated dramatically impaired phonological processing relative to Case L and the TD group. This deficit was the most extreme in the Isolating Phonemes subtests of the PAT (in initial, medial, and final word positions), in which she did not answer a single item correctly (out of 30 opportunities). There is some evidence that this phonological processing

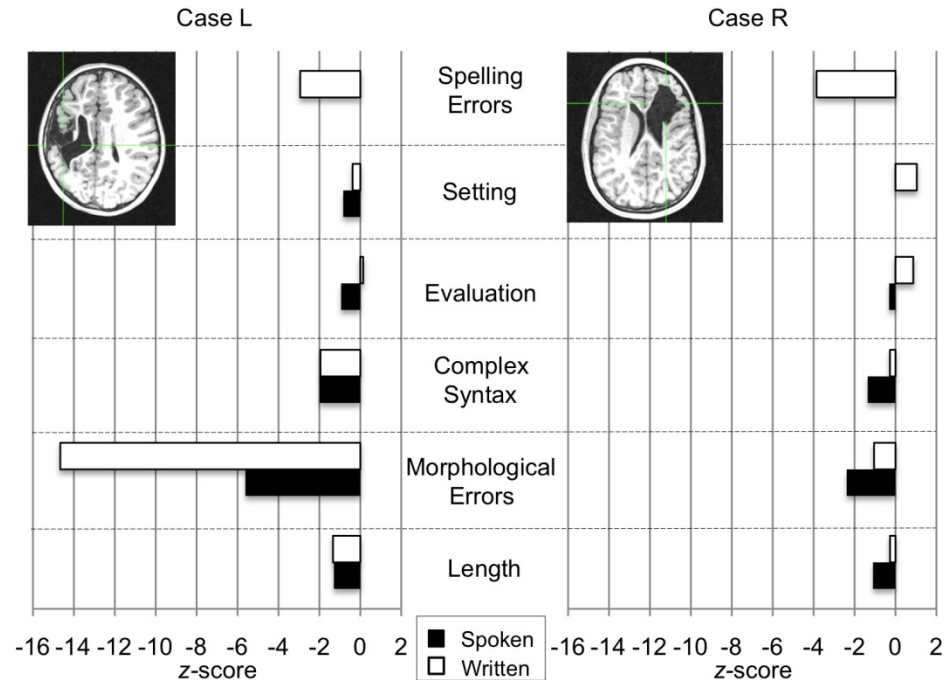


Figure 4.4: Displays the z-scores (relative to the 15 typically developing children) for Case L (left) and Case R (right) on Spoken (black bar) and Written (white bar) Narrative components.

Table 4.2: Transcribed Excerpts from Spoken and Written Narratives

Participant	Modality	Excerpt from child's personal narrative
Case L	Spoken	my sister... <u>bite</u> an'... <u>pinch</u> an'...made me bleed...and... <u>in</u> my arm
Case L	Written	My SiSter <u>bit</u> * and <u>cry</u> * and <u>bit</u> * and bich* really hard <u>make</u> me beud* in my romare*
Case R	Spoken	and then Nolin popped up and then...he...kicked me <u>on</u> no reason
Case R	Written	He kikt* me <u>on</u> the stumnik* on PrPis* for now* resun* I <u>don't</u> now* that He was goweng* to KiK* me <u>anthes</u> * tumik*

* Underlined words indicate a morphological error and asterisks indicate a spelling error.

deficit may persist across language levels and modalities. That is, Case R also demonstrated dramatically impaired performance on the single word reading measure (RWS), especially in conditions containing a foil with a similar orthographic shape. She performed with 0% accuracy on all conditions containing an orthographic foil, suggesting that the reading modality (and perhaps specifically orthographic processing) is very fragile. As mentioned above, Case R also made dramatically more spelling errors in the written narrative context than Case L and the TD

group. There is some evidence from previous research that both the impaired orthographic processing in the reading modality and the spelling errors in the writing modality may stem from degraded phonological representations in the spoken modality. There has been considerable research on phonological processing capacity and subsequent reading ability, with many investigations showing that spoken phonological awareness is correlated with reading ability in the school-age years (e.g. Anthony, Williams, McDonald, & Francis, 2007; Catts, 1993; Catts, Adlof, & Weismer, 2006; Catts, Fey, Tomblin, & Zhang, 2002; Catts, Fey, Zhang, & Tomblin, 2001; Hogan, Catts, & Little, 2005; Nation, Snowling, & Clarke, 2007). This research has been relatively less extended to the writing domain, but research that has explored early language skills and their relation to emergent literacy suggests that phonological awareness skills are also closely associated with emergent writing (e.g. Abbott, Berninger, & Fayol, 2010; Babayiğit & Stainthorp, 2011; Berninger, Abbott, Abbott, Graham, & Richards, 2002; Berninger & Abbott, 2010; Berninger et al., 1992; Defior, Gutiérrez-Palma, & Cano-Marín, 2012; Dunsmuir & Blatchford, 2004; Ehri, 2005; Ehri et al., 2001; Puranik, Lonigan, & Kim, 2011). Here, we find additional evidence that phonological skills are associated with reading and writing ability, as the deficit in spoken phonological processing appears to resonate across linguistic levels and modalities.

Case L showed much more consistently impaired performance on standardized assessments across levels and modalities (performance on all standardized measures hovered between two and four standard deviations below the TD mean). She performed significantly better on the narrative assessment overall, with one notable exception in morphological processing. Specifically, Case L made dramatically more morphological errors (in both her written and spoken narrative) than both Case R and the TD group. In our initial hypotheses, we posed two possible scenarios for the relations between spoken and written language in children with perinatal stroke. First, language in the written domain may mirror that of the spoken domain (i.e. functional but vulnerable, especially when probing isolated language components).

Alternatively, we postulated that the vulnerability of the language system might provide an inadequate platform to support written language, in which case we would expect written language to be relatively worse than spoken language. Results from Case L provide evidence for the latter hypothesis: Weak morphological representations in the spoken modality manifest as severe impairments in this more challenging and new communicative modality (writing). In sum, both children provide evidence that fragile representations in the spoken modality persist in the written modality. In Case R, degraded phonological representations manifest as increased spelling errors in the written modality; in Case L, degraded morphological representations in the spoken modality manifest as dramatically increased morphological errors in the written modality.

The finding that children with perinatal stroke perform better on quasi-unstructured language use tasks, yet struggle with individual linguistic elements is one of the most consistent findings across perinatal stroke research. Very few studies, however, address possible contextual or neurological mechanisms that may account for this consistent result. Contextually, unlike standardized tests of language, in a narrative assessment the child has the freedom to choose the words and syntactic structures to convey her thoughts, and thus is likely to choose linguistic elements with which she is the most competent. According to this view, the child is using a strategy to maximize her linguistic strengths and minimize her weaknesses. However, this strategy is only beneficial in language contexts in which the child has control of the linguistic structures she uses, perhaps accounting for her diminished performance on restrictive, standardized language measures. Furthermore, relating a personal story is one of the first (and most important) communicative functions in the life of a young child or toddler. Parents and caregivers often ask a child to tell about an event that caused a conflict, or to describe the incident that made him sad. Therefore, relating a personal story is a highly practiced language context by the middle of the school-age years. Thus, in contrast to restrictive measures of language, which require linguistic operations such as making a sentence from a given phrase (as in Formulating

Sentences) or isolating words into phonological parts (as in the PAT), the narrative exists in a highly practiced, naturalistic *language-use* context. In subsequent sections, we consider whether this distinction between performance on language use and language structure tasks is represented in the neuroanatomy of children with perinatal stroke.

First, however, we describe the general trends that emerged from our new multimodal metric of white matter structural integrity in both hemispheres for the two children with perinatal stroke (relative to TD). These analyses show, not surprisingly, significant deviations in white matter microstructure within the lesioned hemisphere for both children relative to the typically developing group (Figure 5). For Case L, the average composite tissue integrity score for her left hemisphere was more than three standard deviations below the mean of the TD group (-3.14). For Case R, this averaged score for the right hemisphere was -3.63. Interestingly, however, while the overall white matter structural integrity in the intact (right) hemisphere was within the average range in Case L (-0.16) relative to the TD group, Case R showed notably aberrant structural integrity in the left, non-lesioned hemisphere as well (-1.81). This general hemispheric difference between the two children was evident in more detail within longitudinal, crossing, and short-range association fiber tracts that were both proximal and distal to their lesion areas. For example, the forceps minor (Fmin), which connects the cerebral hemispheres via the genu (anterior) corpus callosum, showed tissue properties that were very similar to the TD group average (-0.19) in Case L, whereas Case R showed significantly abnormal microstructure here (-3.48). This result is consistent with the greater extent of anterior, proximal gray matter damage evident in Case R. In comparison, the forceps major (Fmaj), a tract that connects the hemispheres through the splenial (posterior) portion of the corpus callosum, showed microstructure properties that were comparably different from the TD group in both children (Case L = -2.30, Case R = -2.18). This likely would not have been predicted given that Case L shows greater involvement of nearby posterior perisylvian gray matter, and Case R shows no infarct posteriorly even within the

lesioned hemisphere. Similar differences were also evident for longitudinal association fibers. For example, the inferior frontal occipital fasciculus (IFO), which connects the occipital and frontal lobes, showed expected deviation from normal on the left Case L (-2.68) but showed average-range tissue properties on the right (0.06). In contrast, Case R showed notably low white matter integrity measures for the IFO within both the right (-3.56) and left (-2.31) cerebral hemispheres. This differential pattern involving the non-lesioned hemispheres of the two children becomes strikingly apparent when one standard deviation (positive or negative) is used as an arbitrary cutoff for classifying tract tissue properties. For Case L, only 3 of the 18 white matter fiber tracts on the left are within this “normal” range, whereas all 18 tracts on the right are normal by this standard. In stark contrast, for Case R, no fiber tracts on the right are within one standard deviation of the distribution for typically developing children, and only 5 of 18 tracts within the left hemisphere also meet this standard.

Based on this neuroanatomical profile, we might expect Case R (who shows decreased structural integrity in *both* hemispheres) to show the most severe linguistic impairments, as the lesion includes the right hemisphere homologue to Broca’s area, *and* structural integrity in the left hemisphere is significantly reduced. Somewhat surprisingly, however, Case R demonstrated *less* severe language impairment than Case L on most linguistic levels and modalities (except phonological processing), and most strikingly in morphological processing. Since Case L (regardless of a relatively healthy right hemisphere) demonstrated the most severe linguistic impairments across linguistic subsystems and modalities, yet Case R did not show typical language processing either, our findings provide strong support for three contentions that, taken together, do not easily fit within either equipotentiality or innate predetermination perspectives. First, there is an undeniable advantage for the development of many aspects of language processing within the left hemisphere. Second, the right hemisphere also plays an essential role in normal language development. Third, the functional brain organization that we observe for

language at any given point in time is the result of dynamic interactions between biological and psychological constraints which constitute the *developmental process* of language acquisition. Thus, our results are the most consistent with a soft hemispheric specialization or pluripotentiality perspective (Elman et al., 1997), which suggests that different regions of the cortex have the capacity to support a wide variety of processes, but with varying degrees of success.

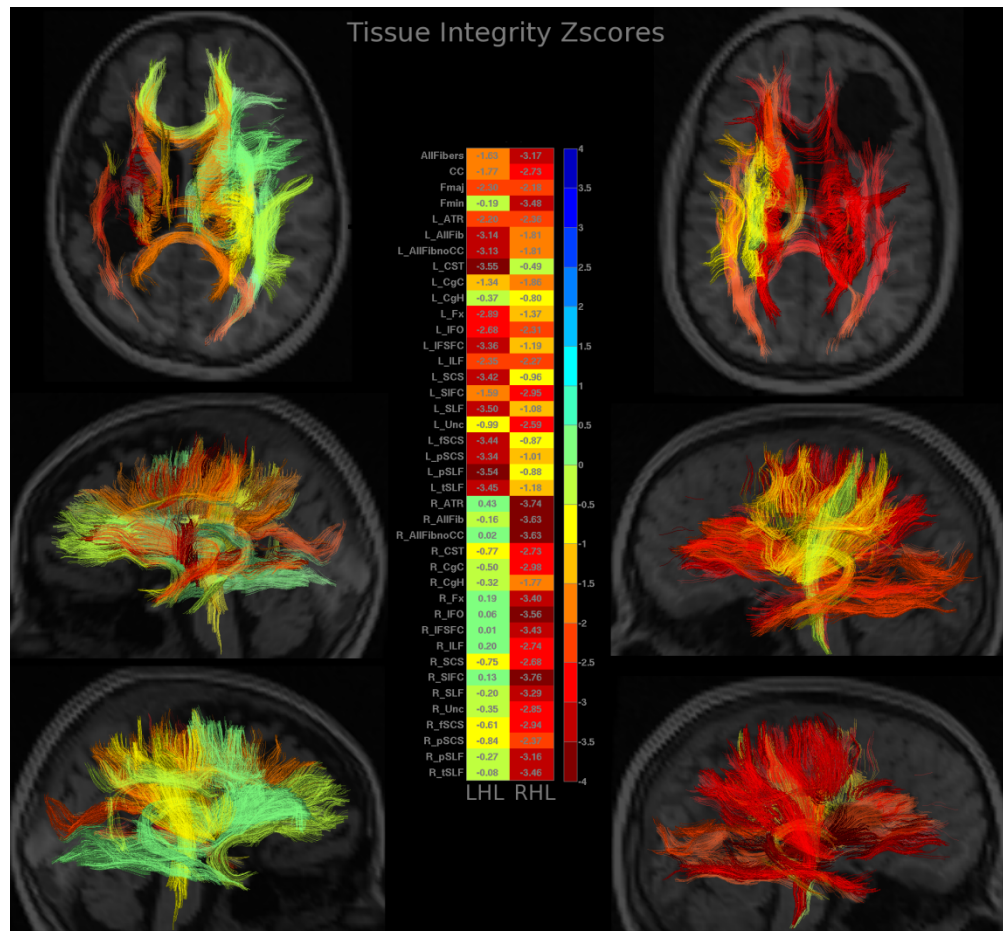


Figure 4.5: Displays z-scores of the composite index (mean FA, MD, and T2N) of the white matter fiber tracts for the two children with perinatal stroke relative to the TD group from superior (top), left hemisphere sagittal (middle), and right hemisphere sagittal (bottom) views of Case L (left) and Case R (right). Warmer colors represent negative z-score values and cooler colors represent positive z-score values.

Some questions emerge from this finding: Why do we observe reduced white matter structural integrity in the non-lesioned hemisphere only in Case R? Do the linguistic deficits observed in Case R stem primarily from the right hemisphere lesion or the lack of white matter

microstructural integrity? First, it is important to note that since Case R was a preterm infant, her risk of periventricular leukomalacia was increased, which could provide an explanation for the decreased white matter structural integrity in the non-lesioned, left hemisphere. However, if there were significant involvement of the non-lesioned left hemisphere, we would expect to see motor impairments and/or hemiparesis on the contralateral side, which is not the case; she does not present with any hemiparesis and shows no motor deficits on the right side. Furthermore, since language performance from Case R was actually better than Case L, it is unlikely that there was any significant bilateral damage, as presumably damage to white matter in the left hemisphere (in addition to a focal lesion in the right hemisphere) would have caused more severe damage relative to Case L who shows robust white matter structural integrity in the right hemisphere. In response to our second question, do the linguistic deficits observed in Case R stem primarily from the right hemisphere lesion or the lack of white matter microstructural integrity? A strong early hemispheric specialization perspective would hypothesize that the state of the left hemisphere would contribute the most to language function, whereas a more developmental model would predict that characteristics from both hemispheres are critical to building the underlying neural correlates for language. The Interactive Specialization view (Johnson & De Haan, 2011; Johnson, 2001) is one such developmental model that suggests that forming the cortical circuitry for a cognitive skill (e.g. language) involves patterns of interactions between partially active brain regions, which compete to acquire their role in cognition. Since this perspective emphasizes the role of the relationship *between* brain regions rather than specific neural substrates of language, this perspective hypothesizes that the robustness of the tracts connecting neural regions would be critically involved in language outcome.

What can the neurological profiles tell us about the difference in performance between language use and language structure? We have seen from previous functional neuroimaging research that processing language in childhood involves relatively more distributed, bilateral

brain areas relative to adults. However, there is also evidence that very early in development (12-18 months), infants utilize localized neural substrates to understand language, which are relatively more active on the left and qualitatively similar in topography (but slower to activate) than those used in adulthood (Travis et al., 2011). Importantly, these findings from children and infants are not in opposition to one another, but rather describe different developmental periods between which the definition of “language” changes dramatically. Around a child’s first birthday, “language” is comprised mostly of single words and (perhaps) comprehension of simple sentences. Appropriately, then, neuroimaging studies that investigate language at this early time-point are also restricted to comprehension of single words, and find evidence of localized neural correlates that support isolated lexico-semantic processing. In childhood, when children gain the ability to use complex and diverse linguistic structures to talk about abstract and imaginary themes, it follows that (as previous neuroimaging research has reported), “language” involves more distributed neural networks to support these complex linguistic feats. With this in mind, we consider the consistent finding that children with right and left hemisphere perinatal stroke perform better on unrestricted language-use tasks than those that probe isolated linguistic elements. It is possible that while children with perinatal stroke take advantage of the healthy tissue to form functional language-use networks, the network structures supporting isolated linguistic elements (such as those assessed with restrictive standardized measures) may develop abnormally and thus remain vulnerable when performance is highly constrained. This finding has implications not only for understanding language in children with perinatal stroke, but also for understanding typical language development and for revealing more about the complex functional organization of cognitive processes in the brain. Lateralization can be viewed as a developmental process; as children master increasingly complex language, the neural circuitry supporting these linguistic elements becomes more specialized, efficient, and eventually, more localized.

Having now presented the behavioral and neural profiles of the two case studies, we revisit the questions we posed in the Introduction. First, what can this investigation tell us about the relationship between language levels and modalities? Results from the two case studies suggest that there is a high level of correspondence across modalities; speaking, reading, and writing are similarly proficient or impaired across linguistic subsystems. The association between speaking and writing modalities is especially apparent in morphological processing of Case L, which is severely impaired in both the spoken and written narrative. There is also a degree of correspondence between the levels of linguistic analysis, in that the smaller linguistic components appear to impact the larger, more complex elements. Evidence for this “vertical” correspondence comes from phonological processing in Case R, which is reflected at the word and narrative levels (in orthographic and spelling performance respectively).

Second, what can this investigation tell us about the relationship between behavior and the brain? Most importantly, we find evidence that there is no simple one-to-one correspondence between neuroanatomy and language. Rather, our case study, which uses detailed linguistic and quantitative neuroanatomical measures, provides evidence that there exists a complex, distributed, dynamic relationship between language and its neural correlates. Up to this point, we have been focusing on the details of these children’s linguistic profiles. Now, however, it is important to step back to appreciate that both children present with large lesions involving perisylvian regions. The fact that either child (perhaps especially Case L) is using language functionally attests to the brain’s plasticity to recruit alternative neural substrates to support language functions. On the other hand, neither is using language typically, suggesting that the alternate neural organization for language is not as robust as the typical organization. One of the questions we raise in this case study involves the extent to which the characteristics of the non-lesioned hemisphere impact language outcome. In the current investigation, Case L presented with decreased white matter structural integrity in the lesioned, left hemisphere, and robust

structural integrity in the right hemisphere. Case R, by contrast, presented with decreased structural integrity in both hemispheres. From these anatomical characterizations alone, we can only make indirect hypotheses regarding the extent to which the lesion and the decreased white matter integrity of the left hemisphere affected the child's language failings (and successes!). While a functional neuroimaging paradigm would more directly address the areas of cortex involved in processing language elements, neuroimaging methods to address narrative language expressively in the spoken or written modality are still in their infancy. Thus, although this case study cannot directly address the relationship between the functional organization of the brain and language behavior, it is the first of its kind to use a fine level of detail in linguistic analysis (in multiple communicative modalities) in combination with quantitative multimodal structural imaging techniques as a tool to address brain behavior relations in children with perinatal stroke. At the very least, our findings point out the importance of assessing the tissue properties of the supposedly "unaffected" hemisphere, to contribute to the interpretation of behavior.

In closing, we feel it is important to remind the reader that these are case studies, and as such we cannot claim that results from these two individuals will generalize to any population. Furthermore, the relations we observe are not causal or developmental in nature; rather we report descriptive relations between language behavior and neural morphology from two children with right and left hemisphere perinatal stroke at a single point in time. What this study *can* contribute is a detailed, multi-level and multi-pronged analysis, impossible with a group study design, which allows us to pose questions for future research regarding the biological state of the brain and the multi-leveled language-system in children with perinatal stroke. It is our hope that these multimodal metrics of structural integrity will continue to be used for the study of individual differences in brain-behavior relationships in future research.

CONCLUSIONS

The goal of this investigation was to better understand the relationship between neuroanatomy and language function. We utilized a quantitative case study format, which compared innovative multimodal imaging metrics in combination with language analysis at multiple levels of linguistic complexity in two school-aged children with perinatal stroke against a backdrop of typically developing age-matched controls. Like previous research, we found that both children (one with a left and one with a right hemisphere lesion) used language functionally, and performed better on unrestrictive measures of language than on standardized measures of isolated linguistic elements. However, the exhaustive nature of the language analysis allowed us to identify distinctive patterns of impairment, which would not have come to light using a less acute language assessment battery. We have also introduced new methodological and theoretical perspectives to the study of children with perinatal stroke; for the first time, we present a detailed analysis of language at multiple levels of complexity alongside a characterization of the tissue properties and structural integrity of *both* hemispheres. These innovative methods allow us to consider the complex relationship between language behavior and the brain.

APPENDIX A

Table 4.3: Mean and Standard Deviations from the Typically Developing Group, as well as z-scores from Case 1 and Case 2 for each Individual Behavioral Measure

	Test	TD		Case L	Case R	Case L z-score	Case R z-score
		Mean	SD				
Speaking	PAT Rhyme Discrimination	9.93	.26	9	6	-3.61*	-15.23***
	PAT Rhyme Production	9.6	.63	0	2	-15.18***	-12.02***
	PAT Phoneme Blending	9.47	.64	10	8	.83	-2.29*
	PAT Isolating Initial Phoneme	8.80	1.21	9	0	.17	-7.29**
	PAT Isolating Final Phoneme	8.80	1.01	9	0	.20	-8.68 **
	PAT Isolating Medial Phoneme	8.88	1.61	4	0	-2.98*	-5.46**
	EOWPVT	104.13	16.86	52	56	-3.09*	-2.86*
	WASI Vocabulary	41.4	7.91	9	23	-4.10*	-2.33*
	WASI Similarities	27.6	5.19	10	19	-3.39*	-1.66
Writing	CELf Recalling Sentences	61.13	12.10	21	31	-3.32*	-2.49*
	CELf Formulating Sentences	41.73	6.92	23	27	-2.71*	-2.13*
	Phonological Spelling Sounds	28.40	4.76	17	22	-2.39*	-1.34
	Lexical WPV	11.67	4.53	3	3	-1.91	-1.91
	Syntactic WJ Writing	17.13	4.04	4	11.5	-3.25	-1.39

Table 4.3 (continued)

	Test	TD		Case L	Case R	Case L <i>z</i> -score	Case R <i>z</i> -score	
		Mean	SD					
Reading	Phonological	Word Attack	23.20	5.48	5	11	-3.32*	-2.23*
	Lexical	RWS Baseline	1	0	1	1	0	0
		RWS Orthographic	.95	.09	1	.29	.56	-7.04**
		RWS Semantic	.96	.07	.67	.33	-4.11*	-8.79**
		RWS Orthographic-semantic	.86	.19	0	0	-4.39*	-4.39*
	WJ Letter-word ID	52.13	10.38	33	30	-1.84	-2.13*	
	Syntactic	Reading Fluency	40.26	14.20	8	7	-2.27*	-2.34*
Narrative	Spoken	Length (Propositions)	18.26	10.25	5	7	-1.29	-1.20
		Morphological Errors/Proposition	.05	.10	.60	.29	-5.62*	-2.38*
		Complex Syntax/Proposition	.94	.27	.40	.57	-1.99	-1.36
		Evaluation/Proposition	.33	.14	.20	.29	-.94	-.31
		Total Setting Components (out of 4)	2	1.20	1	2	-.84	0
		Total Narrative Components (out of 4)	3.13	.74	2	3	-1.52	-.18
	Written	Length (Propositions)	11.73	6.54	3	10	-1.34	-.26
		Morphological Errors/Proposition	.06	.13	2	.2	-14.66***	-1.05
		Complex Syntax/Proposition	.76	.22	.33	.70	-1.95	-.27
		Evaluation/Proposition	.30	.23	.33	.5	.13	.87
		Total Setting Components (out of 4)	1.53	1.41	1	3	-.38	1.04
		Total Narrative Components (out of 4)	3.07	.80	2	4	-1.34	1.17
			Spelling Errors/Word	.07	.08	.29	.36	-2.94*

* $z < -2$, ** $z < -5$, *** $z < -10$

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CHAPTER 5: GENERAL DISCUSSION AND CONCLUSIONS

The investigations that comprise this dissertation have explored relations among components of language in typically developing children and children with neurodevelopmental disorders, using units of analysis ranging from small and relatively simple to large and multifaceted, and finally have considered the neural correlates underlying these subsystems of language. While each chapter adds a piece to our understanding of how these components develop and function to support language (in typical and atypical school-aged children), in this concluding chapter I aim to synthesize and interpret the results from the previous chapters within a Dynamic Systems framework, as well as to consider directions for future research.

Chapter 1 investigated orthographic and semantic components of single word reading (independently and in direct competition) in first to fourth grade children using a novel single word reading paradigm. Results from accuracy and efficiency measures of orthographic processing were similar across age groups, suggesting that orthographic processing is already a strength in the early elementary school years. Semantic processing, on the other hand, demonstrated greater efficiency at each subsequent grade. While the youngest children possessed the ability to correctly identify the synonym match in the Semantic condition (with about 80% accuracy), their performance dropped to less than chance level on the Orthographic-semantic condition, in which the foil was an orthographic neighbor of the target. This pattern of results suggests that young readers already possess robust orthographic processing skills, which must then be integrated with semantic processing during reading acquisition. What are the mechanisms that motivate this integrative process? The Dynamic Systems approach to cognition suggests that the “mechanism” underlying any behavior is the product of the interactions between multiple dynamic subsystems, as well as with the environmental context. Thus, many cognitive systems may be present simultaneously, but will not “emerge” behaviorally until the appropriate

contextual environment is present. The theory further postulates that when the subsystems self-organize, they will seek preferred behavioral modes with varying degrees of stability and instability. The system will revert to this stable state until it is perturbed enough to establish a new steady attractor state. Since typically developing first graders performed quite well on the Semantic condition (where they identified a synonym match in the presence of an unrelated foil), but failed on the same task *in the context of an orthographic-neighbor*, the Dynamic Systems standpoint suggests that orthographic processing is the steady attractor state for young readers. This theory emphasizes the importance of considering the environmental context when interpreting behavioral performance, as the response from the Orthographic-semantic condition on its own would suggest that young children fail to access semantic representations from a written word. However, in the absence of an orthographic response option (the context of the Semantic condition), the system was “pushed” to utilize the less-stable semantic processing state, which only emerged behaviorally under the proper environmental conditions. Chapter 2 allowed further exploration of this interpretation: Are the uneven cognitive profiles of children with Williams syndrome and High Functioning children with Autism reflected in orthographic and semantic processing of single word reading? Could this be a consequence of less co-activation of these dynamic systems?

Children with Williams syndrome and High Functioning children with Autism demonstrate opposing strengths and weaknesses in their visuospatial and language profiles: children with Williams syndrome have relatively strong language in comparison to visuospatial processing skills and High Functioning children with Autism show the reverse pattern. Investigating orthographic (a highly visuospatial process) and semantic (a language-based process) components of single word reading in these populations affords information not only regarding how these processes support reading in High Functioning children with Autism and children with Williams syndrome, but of how these processes relate to one another. Results from

Chapter 2 indicate that children with Williams syndrome fall significantly below the typically developing group on the conditions that require fine-level orthographic processing but not on all conditions requiring semantic processing. High Functioning children with Autism, by contrast, performed similarly to the typically developing group on all conditions requiring fine-level orthographic discrimination, but fell marginally below the typically developing group on the Semantic condition. It is important to note that throughout Chapter 2, a non-significant result is interpreted as a strength in Williams syndrome. While we feel that this is an appropriate interpretation given the moderate mental retardation associated with Williams syndrome, it prevents us from concluding that “good” language skills “tune” orthographic processing (as would be hypothesized by the Lexical Tuning hypothesis) or facilitate the integration between orthographic and semantic processing, since children with Williams syndrome performed below the other two groups on every measure of standardized testing. However, results from Chapter 3 do provide evidence that *relative* strengths and weaknesses in language and visuospatial processing *within* cognitive systems (i.e. within Williams syndrome and within High Functioning children with Autism) impact the way a child recruits the components required to process a single written word. Since Chapters 1 and 2 only considered two aspects of a relatively small linguistic element (single word reading), we cannot, from Chapter 2 results alone, address the second part of the question raised above: Could the increased stability of the attractor states, and the lack of integration between orthographic and semantic processing in these atypical populations be a consequence of less co-activation of the underlying dynamic systems? Chapter 3 provides some insight into the organization of the language system as a whole in typically developing children and High Functioning children with Autism.

Results from Chapter 3 suggest that the weak integration observed in language performance in High Functioning children with Autism may stem from weak relations between subsystems of language. The Dynamic Systems perspective postulates that each subsystem is

constantly interacting dynamically with every other system and with the environmental context, but it is also the case that some systems will be more regularly co-activated than others. Language is a context that involves automatic, simultaneous co-activation of multiple linguistic components. In expressive language, for example, producing a word requires activation of its component phonological parts, producing a sentence requires activation of its component words, producing a narrative requires activation of the sounds, words, and sentences, in addition to the systems related to the story's tone, emotion, and perhaps even motor representations from mentally simulating the actions in the story (as would be predicted by an Embodied Cognition perspective). In view of this persistent co-activation across cognitive and linguistic subsystems, it is not surprising that results from Chapter 3 reveal that in typically developing children, the subsystems of language are highly correlated. In High Functioning children with Autism, however, this was not the case. While High Functioning children with Autism showed relatively good performance at almost every level of analysis (with the exception of Narrative performance), they did not show the same interconnected structure of the language system, suggesting a different underlying organization for language—a lack of association among linguistic subsystems—consistent with the weak central coherence hypothesis. One question raised from these results is: Are weak correlations between linguistic subsystems a hallmark of neurodevelopmental disorders in general, or is this profile specific to High Functioning children with Autism? If weak associations between linguistic systems is a general marker of neurodevelopmental disorders affecting language, perhaps the profile reflects other, more general cognitive deficits which are concomitant with many neurodevelopmental disorders (e.g. intellectual impairment, attention deficits, atypical social interactions). If, on the other hand, this profile of weak relations between linguistic subsystems is unique to High Functioning children with Autism, this would provide stronger support for the Weak Central Coherence account of autism, and would have implications for language interventions for this group.

To investigate whether this weakly associated language profile is unique to High Functioning children with Autism or representative of a more general cognitive or linguistic deficit, preliminary results are presented from performance on and correlations among linguistic components in children with Williams syndrome and children with Perinatal stroke. It is important to note that the following preliminary results are from small sample sizes (Williams syndrome group $N = 13$; Perinatal Stroke group $N = 11$; (7 with Right Hemisphere Lesions and 4 with Left Hemisphere Lesions)) and thus these results should be interpreted only as trends from which to base future research. Performance results are presented in the bar graph in Appendix A, Figure 1, which displays the average performance (with standard error bars) for each population (Typically developing (TD), High Functioning Autism (HFA), Williams syndrome (WS), Perinatal stroke as a combined group (PS), children with Right Hemisphere Lesions (RHL), and children with Left Hemisphere Lesions (LHL)) at each level of assessment in the spoken and written modalities. Note that since results from the typically developing children and High Functioning children with Autism were presented and discussed in Chapter 3, the brief discussion that follows will focus on the other two populations (Williams syndrome and Perinatal Stroke).

Children with Williams syndrome and children with Perinatal stroke show similar performance to one another (below the typically developing group) on most linguistic components, except performance on the Narrative Composition measure, in which children with Perinatal stroke performed better than children with Williams syndrome, and indeed scores from the Perinatal stroke group were only slightly below the typically developing group. Children with right and children with left hemisphere strokes performed similarly to one another across linguistic levels and modalities. These preliminary findings from children with Perinatal stroke are consistent with previous research on this population, but contrast with the case study presented in Chapter 4. These differences between the case study and group results emphasize the strengths and weaknesses of a case study approach. On the one hand, the detailed level of analysis

presented in Chapter 3 allowed identification of differences in linguistic profiles which have been lost when we collapse across multiple subjects. Additionally, in order to draw inferences regarding the neural regions which are *required* and *sufficient* for specific linguistic processes, the case study approach is preferred, as no two lesions or functional organizations of the brain will be identical. On the other hand, case studies do not allow results to be extended beyond the individual, and thus provide very little clinically relevant information, and cannot add significantly to our knowledge of neural plasticity in general. Together the incongruous results from the case study and group results presented in this dissertation serve as a reminder that while children with right and children with left hemisphere Perinatal stroke do not demonstrate differences in language performance as a group, this overarching pattern does not necessarily apply to each individual.

In sum, performance measures on the language components presented above suggest broadly that children with Williams syndrome and children with Perinatal stroke perform similarly to one another (and below their typically developing peers) on every level and modality of linguistic analysis, except for the narrative composition measure, in which the children with Perinatal stroke performed similarly to the typically developing group. While these results provide a depiction of performance at each level in each modality, they do not address the relations between levels and modalities. Investigating the correlations between performance on each linguistic subsystem will provide insight into the question at hand: Are weak correlations between linguistic subsystems a hallmark of neurodevelopmental disorders in general, or is this profile specific to High Functioning children with Autism?

The visualization of the language system from children with Williams syndrome (Appendix A, Figure 2) indicates that the subsystems are much more strongly correlated relative to the High Functioning children with Autism (presented in Chapter 3). Interestingly, the spoken language side of the visualization shows much stronger correlations than the written side. This is

an especially intriguing finding given the strength of spoken over written language behaviorally in this group. It is not uncommon for individuals with Williams syndrome, who show competent conversational abilities and even sophisticated vocabulary and syntactic structures, to show very limited (or absent) reading and writing abilities. Furthermore, some research suggests that the strength in the spoken language domain may stem from strong phonological memory; that is, sensitivity to the sounds in a word or sentence rather than the meaning, *per se*. If this were the case, it would not be surprising that spoken phonological awareness is strongly correlated with every other language subsystem in the spoken modality (syntactic and narrative). This finding suggests, perhaps, that phonological representations may be critically important to language representations in Williams syndrome. If this result is replicated and extended, it may also have implications for clinicians working with this population. Recall that the strong associations between spoken phonological processing and other language components does not necessarily indicate that phonological processing is always a strength, but rather *when* it is a strength, other systems of language also tend to be strong (and vice versa). Thus, unlike in High Functioning children with Autism, the language profile of children with Williams syndrome suggests that focusing clinical interventions on the low-level processing components may be an efficacious intervention strategy. In other words, due to their tightly associated organization, improving spoken phonological processing may improve other, more complex levels of language.

The last series of visualizations (Appendix, Figures 3,4, and 5) illustrate the correlations amongst the linguistic components in children with Perinatal Stroke as a group (Figure 3), children with Right Hemisphere lesions (Figure 4) and children with Left Hemisphere lesions (Figure 5). Performance from the Perinatal stroke group combined illustrates a moderate number of moderately strong correlations, yet none of the “lower” linguistic subsystems are correlated with narrative performance. This profile falls somewhere between that of the typically developing children and the High Functioning children with Autism. Interestingly, when the groups are

separated into children with right and children with left hemisphere lesions, a very different pattern emerges. The profile of the children with right hemisphere lesions closely resembles that of the typically developing group, though not as robustly (which could be a function of the decreased sample size). The children with the left hemisphere strokes, however, show almost no correlations at all. As the children with left hemisphere strokes make up the smallest sample size by far, these results should be interpreted with caution.

These preliminary results suggest that weak coherence among components of language is not a general marker of neurodevelopmental disorders affecting language, but seems to be specific to High Functioning children with Autism (and perhaps children with left hemisphere Perinatal stroke). This finding provides some support for the Weak Central Coherence account of autism, but we are left with many questions: Does the weak central coherence cause weak correlations between linguistic subsystems, or do weak relations among linguistic subsystems cause more local-level processing? How are these associations among language components represented in the structure and function of the brain?

The questions raised from these findings on the relations among linguistic subsystems in High Functioning children with Autism, children with Williams syndrome, and children with Perinatal stroke will be the focus of my future research. My first and most immediate goal is to use the results from the Perinatal stroke case study presented in Chapter 4 to form hypotheses to test in a group study format. Secondly, I will extend the preliminary results presented here to include more participants, which will enable us to draw more concrete conclusions regarding the relations among language components in children with Williams syndrome and children with unilateral Perinatal stroke. My longer-term goals involve relating these behavioral profiles to the structural and functional characteristics of the brain: Do weak correlations among subsystems of language within a population predict lower white matter structural integrity? Can we identify differences in activation synchrony (using functional imaging methodologies with high spatial

and temporal resolution such as Magnetoencephalography) between language regions when performing comprehension tasks that involve increasing levels of linguistic complexity (e.g. phonemes, words, sentences)? Lastly, what can identification of these unique associative language profiles tell us about the efficacy of different types of language interventions? Will interventions that focus on the low-level phonological and orthographic aspects of words and sentences be more efficacious for children with Williams syndrome and children with right hemisphere Perinatal stroke than for High Functioning children with Autism and children with left hemisphere Perinatal stroke? On the other hand, will interventions that focus on the more global linguistic aspects (e.g. story structure, narrative, conversation) be more successful for High Functioning children with Autism and children with left hemisphere Perinatal stroke than for the other groups?

In closing, the investigations in this dissertation indicate that exploring the relations among subsystems of language will provide valuable information which will help to create more targeted and efficacious interventions for children with language disorders. Rather than low-level language components providing independent platforms upon which to build more complex structures, results from this dissertation suggest that typical language development involves the integration and co-activation of multiple subsystems that are in constant interaction with one another and with the environment. To return to the initial analogy, while pedaling, steering, and braking are all required skills for riding a bicycle, it is not until those skills are integrated and organized with one another and with the terrain that one can cycle through town, or *Through the Looking Glass*.

APPENDIX A

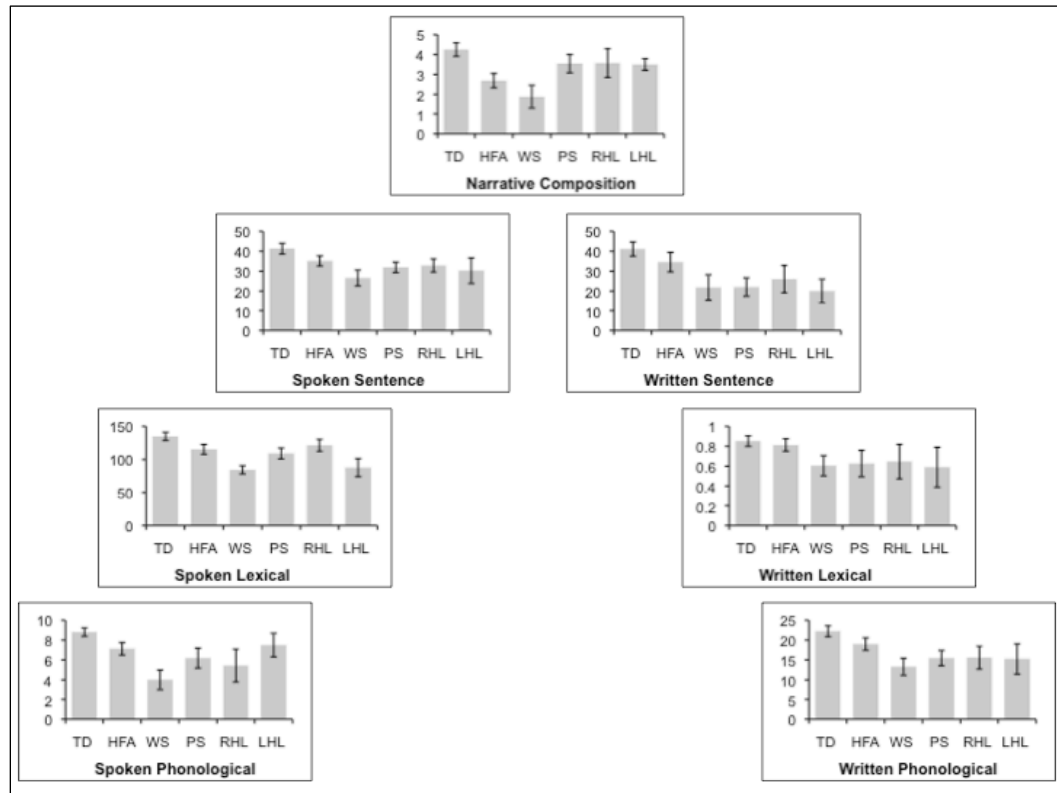


Figure 5.1: Displays the performance (raw scores) on phonological, lexical, and sentence-level assessments in the spoken (left) and written (right) modalities, and on spoken narrative composition (top). Scores are presented from Typically developing children (TD), children with Williams syndrome (WS), High Functioning children with Autism (HFA), children with Perinatal stroke as a group (PS), children with Right Hemisphere lesions (RHL) and children with Left Hemisphere Lesions (LHL).

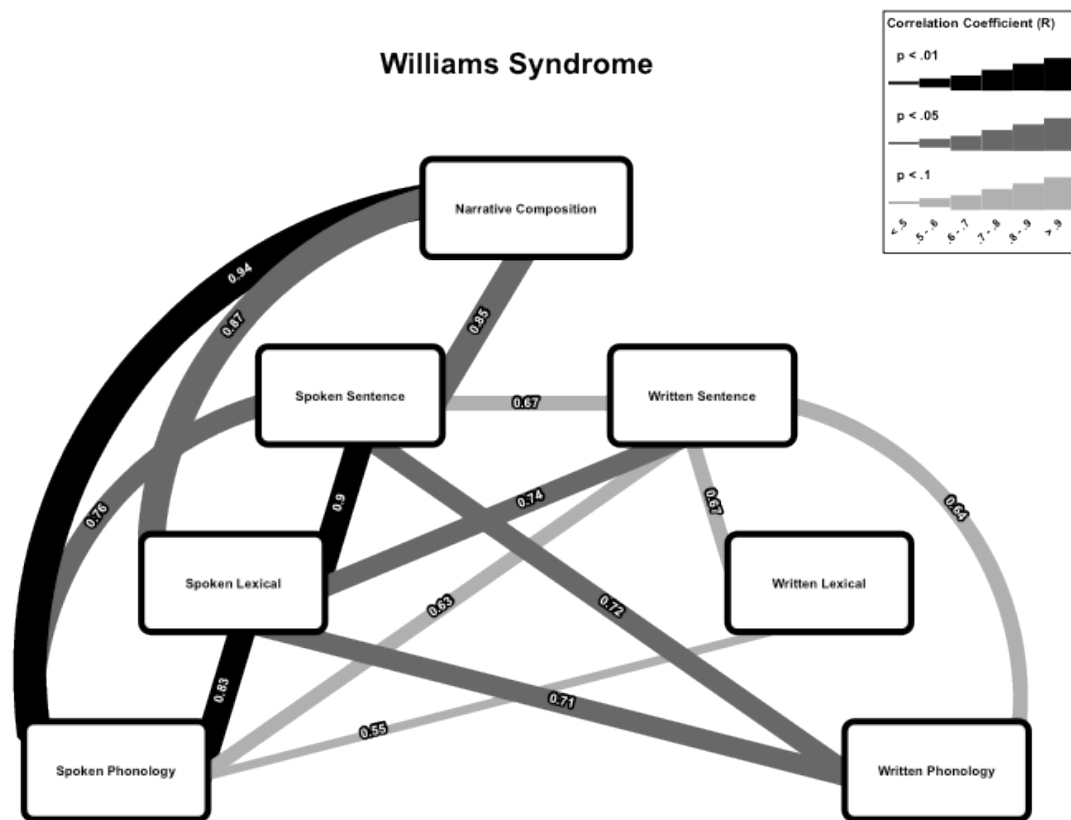


Figure 5.2: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for WS. The width of the line represents the correlation value, and the grey-scale represents the p-value.

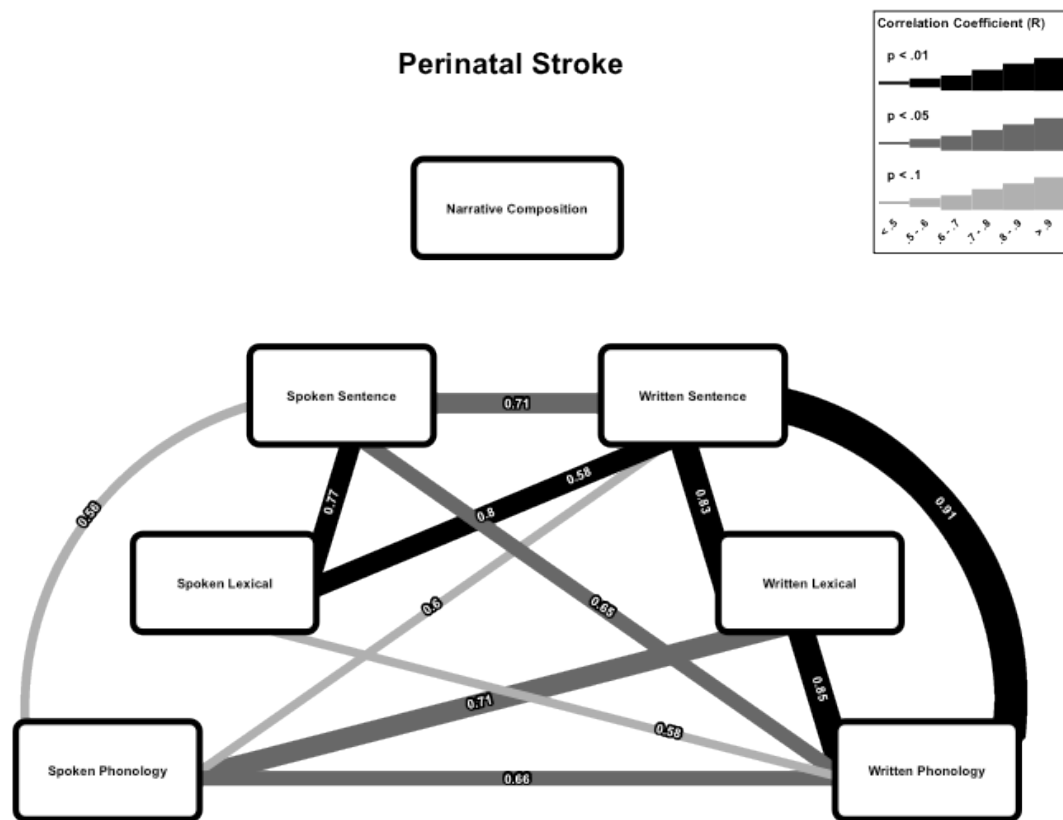


Figure 5.3: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for the Perinatal stroke group combined. The width of the line represents the correlation value, and the grey-scale represents the p -value.

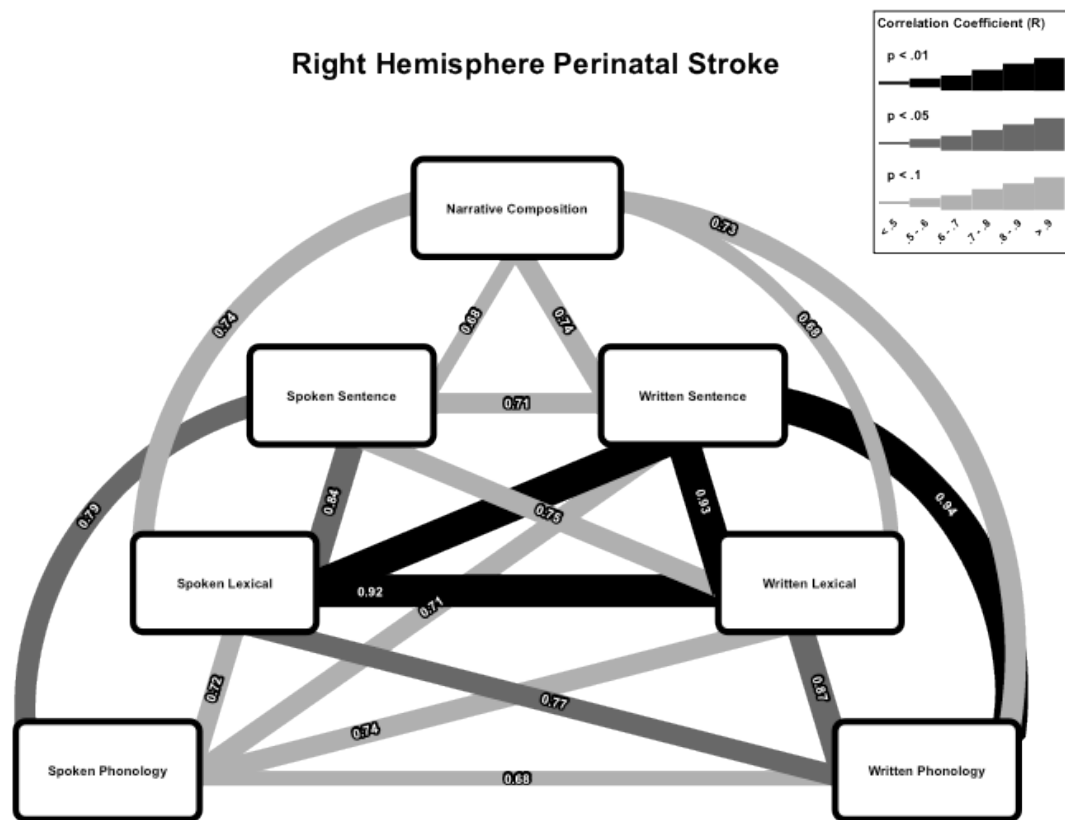


Figure 5.4: Displays the correlations among the linguistic subsystems (phonological, lexicosemantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for children with Right Hemisphere lesions only. The width of the line represents the correlation value, and the grey-scale represents the p -value.

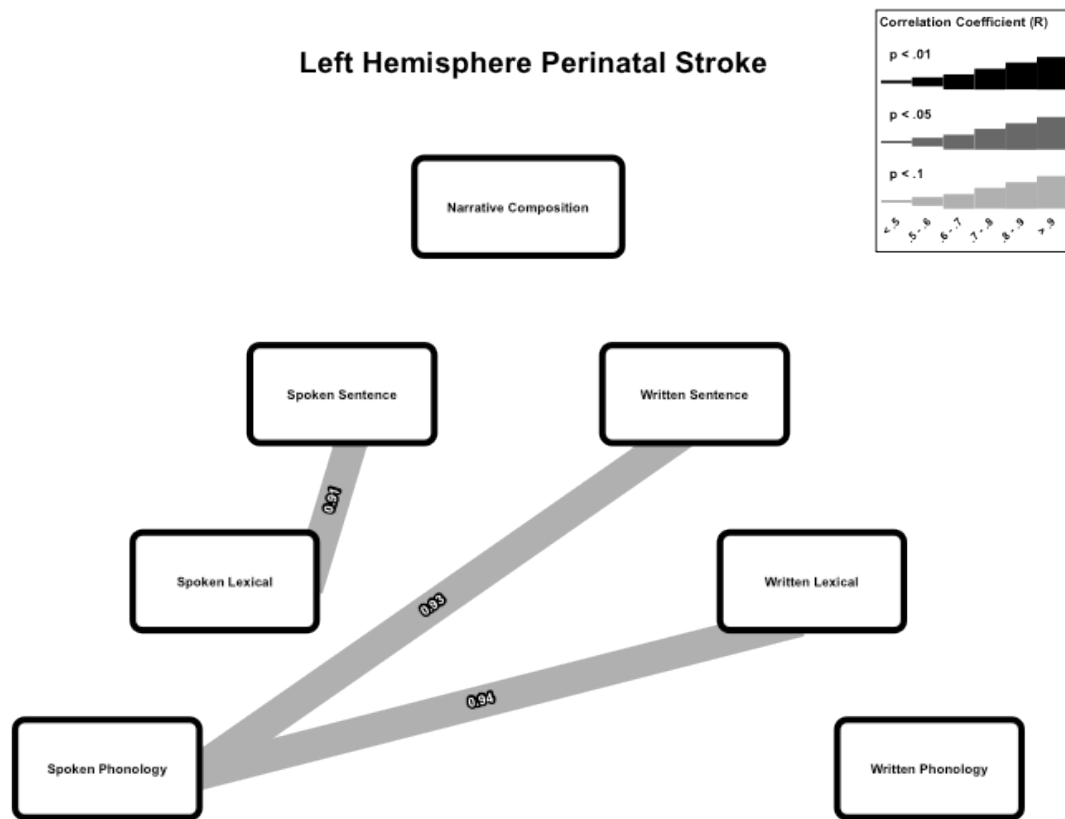


Figure 5.5: Displays the correlations among the linguistic subsystems (phonological, lexico-semantic, syntactic, and narrative) in the spoken modality (left) and written modality (right) for children with Left Hemisphere lesions only. The width of the line represents the correlation value, and the grey-scale represents the p -value.