PART I

Defining Dyslexia, Comorbidity, Teachers' Knowledge of Language and Reading

A Definition of Dyslexia

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This paper elaborates on the components of a working definition of developmental dyslexia. It follows the general format of a paper by Lyon published in Annals of Dyslexia *in 1995, which elaborated on a working definition proposed in 1994 (Lyon, 1995). The current definition agreed on by the work group updates and expands on the working definition from 1994.*

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CURRENT DEFINITION—2003

Dyslexia is a specific learning disability that is neurobiological in origin. It is characterized by difficulties with accurate and/or fluent word recognition and by poor spelling and decoding abilities. These difficulties typically result from a deficit in the phonological component of language that is often unexpected in relation to other cognitive abilities and the provision of effective classroom instruction. Secondary consequences may include problems in reading comprehension and reduced reading experience that can impede growth of vocabulary and background knowledge.

WORKING DEFINITION FROM 1994

Dyslexia is one of several distinct learning disabilities. It is a specific language-based disorder of constitutional origin characterized by difficulties in single word decoding, usually reflecting insufficient phonological processing. These difficulties in single word decoding are often unexpected in relation to age and other cognitive and academic abilities; they are not the result of generalized developmental disability or sensory impairment. Dyslexia is manifest by variable difficulty with different forms of language, often including, in addition to problems with reading, a conspicuous problem with acquiring proficiency in writing and spelling.

Dyslexia is a specific learning disability. This opening sentence identifies dyslexia as a specific learning disability in contrast to the more general term learning disabilities (LD). While the general LD category encompasses a wide range of disorders in listening, speaking, reading, writing, and mathematics (USOE, 1977), we continue to recommend (Fletcher et al., 2002; Lyon, 1995) that the field should discontinue the use of the broad term learning disabilities when discussing reading disabilities, and should instead discuss specific disabilities defined in terms of coherent and operational domains. From an epidemiologic perspective, reading disabilities affect at least 80 percent of the LD population and thus constitute the most prevalent type of LD (Lerner, 1989; Lyon, 1995).

As noted previously (Lyon, 1995), it is also important to recognize that many individuals with dyslexia evidence co-occurring or comorbid deficits in other cognitive and academic

areas such as attention (Shankweiler, et al., 1995; B.A. Shaywitz, Fletcher, & S.E. Shaywitz, 1994), mathematics (Fletcher & Loveland, 1986), and/or spelling and written expression (Lindamood, 1994; Moats, 1994). These observations of comorbidity do not detract from the specificity of the proposed working definition of dyslexia since the cognitive characteristics of deficits in attention and mathematics are quite different from the cognitive characteristics associated with deficits in basic reading skills (for further discussion see Lyon, 1995; Lyon, Fletcher, & Barnes, 2003).

. . . that is neurobiological in origin. This phrase recognizes the great advances in understanding the neural basis for dyslexia in the eight years since the previous definition and goes well beyond the phrase "of constitutional origin" in the 1994 definition. The neurobiological origins of dyslexia were suspected over a century ago. Thus, as early as 1891, the French neurologist Dejerine (Dejerine, 1891) suggested that a portion of the left posterior brain region is critical for reading. Beginning with Dejerine, a large body of literature on acquired inability to read (alexia) describes neuroanatomic lesions most prominently centered in the parieto-temporal area (including the angular gyrus, supramarginal gyrus, and posterior portions of the superior temporal gyrus) as a region pivotal in mapping the visual percept of the print onto the phonologic structures of the language system (Damasio & Damasio, 1983; Friedman, Ween, & Albert, 1993; Geschwind, 1965). Another posterior brain region, this more ventral in the occipito-temporal area, was also described by Dejerine (1892) as critical in reading. In the modern era, a range of neurobiological investigations using postmortem brain specimens (Galaburda, Sherman, Rosen, Aboitiz, & Geschwind, 1985), brain morphometry (Brown, et al., 2001; Eliez, et al., 2000; Filipek, 1996), and diffusion tensor MRI imaging (Klingberg, et al., 2000) supports the belief that there are differences in the temporo-parieto-occipital brain regions between dyslexic and nonimpaired readers. Perhaps the most convincing evidence for a neurobiologic basis of dyslexia comes from the now overwhelming and converging data from functional brain imaging investigations. Rather than being limited to examining the brain in an autopsy specimen or measuring the size of brain regions using static morphometric indices, functional imaging offers the possibility of examining brain function during performance of a cognitive task. In principle, functional brain imaging is quite simple. When an individual is asked to perform a discrete cognitive task, that task places processing demands on

particular neural systems in the brain. To meet those demands requires activation of neural systems in specific brain regions, and those changes in neural activity can be measured by techniques such as functional magnetic resonance imaging (fMRI) and magnetoencephalography (MEG). Since fMRI and MEG are noninvasive and safe, they can be used repeatedly, properties which make it ideal for studying people, especially children.

A range of neurobiological investigations from scientists around the world has documented the disruption of neural systems for reading in dyslexia that cross languages and cultures. Converging evidence using functional brain imaging in adult dyslexic readers shows a failure of left hemisphere posterior brain systems to function properly during reading (see figure 1) (Brunswick, McCrory, Price, Frith, & Frith, 1999; Helenius, Tarkiainen, Cornelissen, Hansen, & Salmelin, 1999; Horwitz,

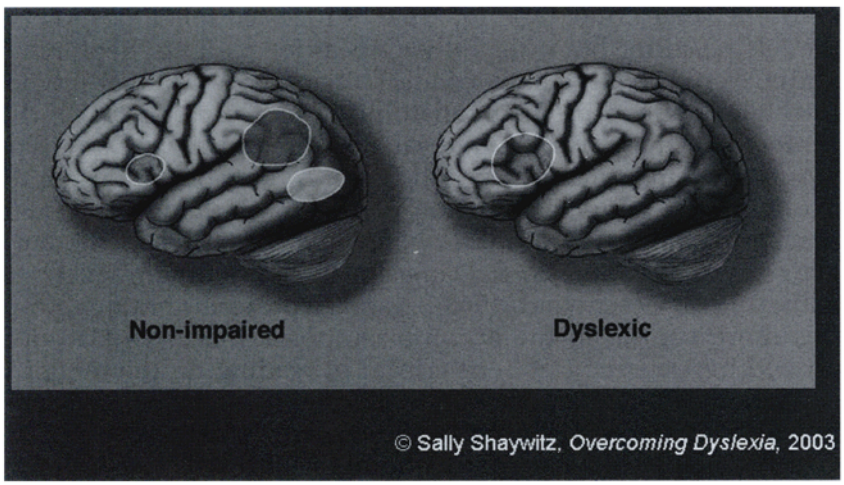

Figure 1. *Schematic of brain activation maps in nonimpaired and dyslexic readers engaged in phonological processing during the pseudoword rhyming. Nonimpaired readers activate: 1) an anterior system in the left inferior frontal region; 2) a dorsal parieto-temporal system involving angular gyrus, supramarginal gyrus, and posterior portions of the superior temporal gyrus; 3) a ventral occipito-temporal system involving portions of the middle temporal gyrus and middle occipital gyrus. In contrast, dyslexic readers demonstrate a relative underactivation in both posterior systems and an increased activation in the inferior frontal gyrus. Figure reprinted from S. Shaywitz, Overcoming Dyslexia, with permission.*

Rumsey, & Donohue, 1998; Paulesu, et al., 2001; Rumsey, et al., 1992; Rumsey, et al., 1997; Salmelin, Service, Kiesila, Uutela, & Salonen, 1996; S. E. Shaywitz, et al., 2003; S. E. Shaywitz, et al., 1998; Simos, Breier, Fletcher, Bergman, & Papanicolaou, 2000), as well as during nonreading visual processing tasks (Demb, Boynton, & Heeger, 1998; Eden, et al., 1996). Anterior systems, especially involving regions around the inferior frontal gyrus, have also been implicated in reading, both in reports of individuals with brain lesions (Benson, 1977) as well as functional brain imaging studies (Brunswick, et al., 1999; Corina, et al., 2001; Georgiewa, et al., 1999; Gross-Glenn, et al., 1991; Paulesu, et al., 1996; Rumsey, et al., 1997; S. E. Shaywitz, et al., 1998). This neurobiological evidence of dysfunction in left hemisphere posterior reading circuits is already present in reading disabled children and cannot be ascribed simply to a lifetime of poor reading (Seki, et al., 2001; B. A. Shaywitz, et al., 2002; Simos, et al., 2000; Temple, et al., 2001).

These data allow neuroscientists and clinicians to use a working model of the neural systems for reading based on the historic work of Dejerine and a more modern theory by Gordon Logan (see Figure 2). Logan (1988, 1997) proposed two systems critical in the development of skilled, automatic processing. One involves word analysis, operating on individual units of words such as phonemes, requiring attentional resources and processing relatively slowly. The second system operating on the whole word (word form), is an obligatory system that does not require attention and processes very rapidly. Converging evidence from a number of lines of investigation (see above) indicates that Logan's word analysis system is localized within the parieto-temporal region while the automatic, rapidly responding system is localized within the occipito-temporal area, functioning as a visual word form area (Cohen, et al., 2000; Cohen, et al., 2002; Dehaene, Le Clec'H, Poline, Le Bihan, & Cohen, 2002; Dehaene, et al., 2001; McCandliss, Cohen, & Dehaene, 2003; Moore & Price, 1999). The visual word form area appears to respond preferentially to rapidly presented stimuli (Price, Moore, & Frackowiak, 1996) and is engaged even when the word has not been consciously perceived (Dehaene, et al., 2001). It is this occipito-temporal system that appears to predominate when a reader has become skilled, and has bound together as a unit the orthographic, phonologic, and semantic features of the word (figure 2).

It is characterized by difficulties with accurate and/or fluent word recognition and by poor spelling and decoding abilities. This phrase replaces the 1994 definition that referred

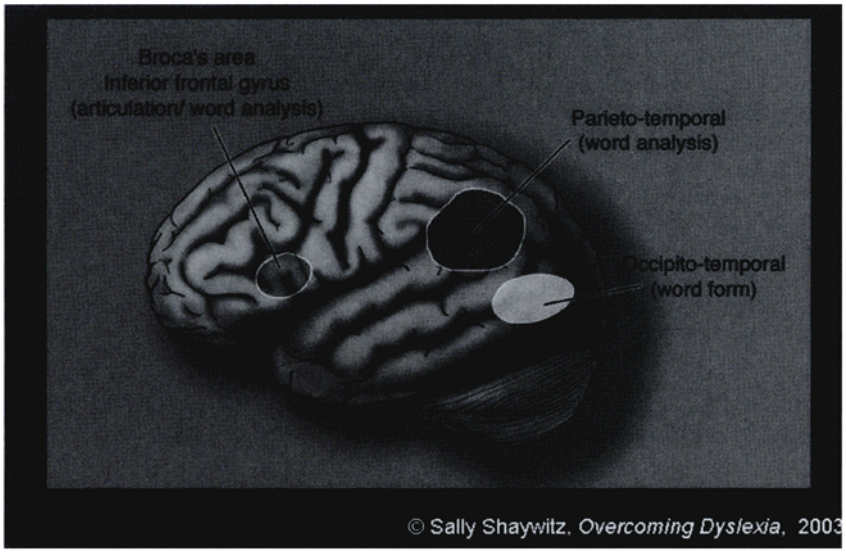

Figure 2. *Neural systems for reading. Converging evidence indicates three important systems in reading, all primarily in the left hemisphere. These include an anterior system and two posterior systems: 1) anterior system in the left inferior frontal region; 2) parieto-temporal system involving posterior portions of the superior temporal gyrus, the supramarginal gyrus, and the angular gyrus; and 3) occipito-temporal system involving portions of the middle and inferior temporal gyrus, middle and inferior occipital gyrus, and the fusiform gyrus. Figure reprinted from S. Shaywitz, Overcoming Dyslexia, with permission.*

simply to "difficulties in single word decoding." The new definition expands on this phrase, referring specifically to difficulties with accurate word recognition (identifying real words) and to decoding abilities (pronouncing pseudowords). It also recognizes poor spelling as a characteristic of dyslexia. "Spelling is intimately related to reading not only because sounds are being linked to letters but because words are being encoded - literally put into a code instead of merely being deciphered or decoded" (S. Shaywitz, 2003, p. 191). Perhaps the most important change in this portion of the definition is the recognition that what characterizes dyslexic individuals, particularly dyslexic adolescents and adults, is the inability to read fluently. Fluency is the ability to read text quickly, accurately, and with good understanding (Report of the National Reading Panel, 2000; Wolf, Bowers, & Biddle, 2001) and is the hallmark of a skilled reader.

Data indicate that readers who are dyslexic can improve in reading words more accurately as they mature, but continue to lack fluency in their reading, which results in effortful, slow reading (Lefly & Pennington, 1991; S. Shaywitz, 2003).

These difficulties typically result from a deficit in the phonological component of language. While theories of dyslexia have been proposed that are based on the visual system (Stein & Walsh, 1997), and other factors such as temporal processing of stimuli within these systems (Talcott, et al., 2000; Tallal, 2000), there is now a strong consensus among investigators in the field that the central difficulty in dyslexia reflects a deficit within the language system (see Ramus, et al., 2003 for an up-to-date review of the theories of dyslexia). Investigators have long known that speech enables its users to create an indefinitely large number of words by combining and permuting a small number of phonologic segments, the consonants and vowels that serve as the natural constituents of the biologic specialization for language. An alphabetic transcription (reading) brings this same ability to readers, but only as they connect its arbitrary characters (letters) to the phonologic segments they represent. Making that connection requires an awareness that all words can be decomposed into phonologic segments. It is this awareness that allows the reader to connect the letter strings (the orthography) to the corresponding units of speech (phonologic constituents) they represent. The awareness that all words can be decomposed into these basic elements of language (phonemes) allows the reader to decipher the reading code. In order to read, a child has to develop the insight that spoken words can be pulled apart into phonemes and that the letters in a written word represent these sounds. As numerous studies have shown, however, such awareness is largely missing in dyslexic children and adults (Bruck, 1992; Fletcher, et al., 1994; Liberman & Shankweiler, 1991). Results from large and well-studied populations with reading disability confirm that in young school-age children (Fletcher, et al., 1994; Stanovich & Siegel, 1994) as well as in adolescents (S. E. Shaywitz, et al., 1999), a deficit in phonology represents the most robust and specific (Morris, et al., 1998) correlate of reading disability. Such findings form the basis for the most successful and evidence-based interventions designed to improve reading (Report of the National Reading Panel, 2000).

That is often unexpected in relation to other cognitive abilities and the provision of effective classroom instruction. This statement generated a great deal of discussion within the committee. On the one hand, the committee recognized that the

notion of an unexpected difficulty in learning to read is basic to almost all definitions of dyslexia including the 1995 definition (Lyon, 1995; Orton, 1937). On the other hand, while preserving the concept of unexpected underachievement, the committee did not want to embrace the idea that basic deficits in decoding and word recognition must be significantly lower than IQ as specified in typical discrepancy formulae. In fact, "There is an emerging consensus among researchers and clinicians that the dependence on a discrepancy between IQ and reading achievement for a diagnosis of dyslexia has outlived its usefulness . . ." (S. Shaywitz, 2003, p. 137). Rather, the data suggest that "unexpectedness" should be assessed via comparisons of reading age with chronological age and/or by comparing reading ability to educational level and professional level of attainment (S. Shaywitz, 2003, p. 133). A major concern with relying on a discrepancy formula is that this all too often results in a delay in identification of a reading problem, and this delay in identification results in a delay in the provision of effective reading instruction. The reader is referred to Fletcher et al. (2002) and Lyon, et al. (2003) for a more detailed review of this issue.

New in this component of the definition is the concept that the child needs to have been provided with effective classroom instruction. Documenting an individual's instructional history is critical to understanding the nature of the observed reading difficulty. For example, many children who are at risk for reading failure come from disadvantaged backgrounds where quality early childhood education and preschool experiences are less available. Thus, they frequently enter formal schooling lacking many of the essential linguistic and other early pre-reading abilities (i.e., phonological sensitivity, vocabulary, print awareness) critical for proficient reading development. If the reading instruction provided the child in the classroom is not informed by an understanding of the gaps in foundational skills and adjusted to teach the missing skills, reading failure typically occurs (see Lyon, et al., 2001 for a review of this issue). On the other hand, a number of recent studies (see Torgesen, 2000, for a review) have shown that many children identified as at risk for reading failure in kindergarten and first grade, and provided with effective instruction, developed proficient early reading skills. Indeed, Torgesen (2000) reported that effective early interventions have the capability of reducing the expected incidence of reading failure from 18 percent of the school age population to 1.4 to 5.4 percent.

But the early intervention studies summarized by Torgesen (2000) clearly indicate that none of the intervention programs

were equally effective for all of the at-risk children studied even when they were implanted intensively by well-prepared teachers. As the committee discussed these findings, a consensus emerged that the role of instructional history must be taken seriously. Specifically, the lack of response to scientifically informed instruction is one factor that differentiates severe and intractable reading deficits from reading failure resulting from inadequate instruction. Thus, the definition of developmental dyslexia and the identification of individuals with dyslexia should address and assess the quality of response to expert instruction.

Secondary consequences may include problems in reading comprehension and reduced reading experience that can impede growth of vocabulary and background knowledge. Addition of this sentence gets to the downstream consequences of phonological difficulties. This is critical in making sure that all understand how phonological difficulties lead to problems in accuracy and fluency, which may lead to problems in vocabulary and problems in background knowledge. Together, these may impact on reading and understanding connected text. It is important that we state this explicitly to put to rest the sometimes stated argument that accuracy and fluency in printed word recognition "are not really reading." Furthermore, including downstream difficulties in vocabulary and background knowledge provides the field with a causal model that can help guide comprehensive assessment efforts, for example, the assessment of phonological awareness, alphabetic principle, accuracy, fluency, and vocabulary.

SUMMARY

Substantial converging evidence relevant to the epidemiology, developmental course, neurobiology, and the cognitive and linguistic characteristics of dyslexia has accrued since the initial working definition of dyslexia from our committee was published in 1995. Likewise, since 1995, our understanding of dyslexia has been informed by a number of intervention and remediation studies that now provide the opportunity to integrate information about the nature and magnitude of response to instruction into our current conceptualizations of developmental dyslexia. The proposed 2003 definition discussed in this paper reflects our respect for the dynamic nature of the scientific process and its usefulness in extending our understanding of dyslexia.

We have revised the 1995 working definition on the basis of current converging evidence relevant to reading development, reading disabilities, and reading instruction. But the task is not complete. Our understanding of dyslexia is a work in progress and will continue to be just that. To be sure, over the next five years, our ability to bring to bear improved research methodologies and investigative modalities to the study of dyslexia literally guarantees the acquisition of new knowledge that will further modify the definition. The one constant is that this and future definitions reflect the best that science has to offer.

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