Misunderstandings about developmental dyslexia: a historical overview

Pietro Smirni,1 Luigi Vetri,2 Eliana Misuraea,3 Marco Cappadonna,4 Francesca Felicia Operto,5 Grazia Maria Giovanna Pastorino,5,6 Rosa Marotta7

1Department of Educational Sciences, University of Catania, Catania; 2Department of Health Promotion, Maternal and Child Health, Internal Medicine and Specialist of Excellence “G. D’Alessandro”, University of Palermo; 3Geriatric Unit, Department of Internal Medicine and Geriatrics, University of Palermo; 4Department of Clinical and Experimental Medicine, University of Messina; 5Child and Adolescent Neuropsychiatry Unit, Department of Medicine, Surgery and Dentistry, University of Salerno; 6Sleep Lab for Developmental Age, Clinic of Child and Adolescent Neuropsychiatry, Department of Mental Health, Physical and Preventive Medicine, University of Campania “Luigi Vanvitelli”, Napoli; 7Department of Medical and Surgical Science, University “Magna Graecia”, Catanzaro, Italy

Abstract

Developmental dyslexia is a reading disorder unrelated to intellectual disability, inadequate teaching systems or poor motivation for schooling. The first attempts to understand such difficulty of learning to read, connected the problem to a primary ‘visual defect’. Since then, several models have been developed. In the last decades, autopsy and histopathological studies on the brain of developmental dyslexics provided neuroanatomical evidence of structural and morphological differences between the normal and dyslexic brains. Furthermore, neuroimaging studies allowed to understand the neural systems of reading and dyslexia. According to more recent studies, developmental dyslexia appears as a language-related neurodevelopmental disorder with a deficit in phonological decoding and visuo-spatial organization of the language code. Therefore, dyslexia is viewed as a multi-componental and complex disorder. Consequently, rehabilitation should be aimed at both the recovery of linguistic decoding processes and the improvement of visuo-spatial and attentional processes. This brief overview should be a valuable tool for a deeper understanding of dyslexic disorder. Literature searches in Medline, PsycINFO, EMBASE, Scopus, PubMed, Web of Science identified one hundred articles focusing attention on how this disorder has been considered over the years.

Introduction

Misunderstandings about dyslexia have fairly distant historical roots, and although several clinical and research studies have contributed, over the years, to understanding dyslexia, still today there are many outstanding questions and developmental dyslexia, in clinical practice, continues to be an overused diagnosis.1,2 However, research and studies agree on what developmental dyslexia is not.3 It is not the difficulty to learn reading associated to intellectual disability, or to sensory deficits (visual, auditory) that impair the grapheme-phoneme conversion, or to education and cultural deprivation, or to psychopathological conditions that can modulate the relational, psychological or emotional problems of the child.4,5 Each of these clinical, psychological or social conditions can give rise to learning reading difficulties. Nevertheless, these conditions are generally associated with wider learning or cognitive disabilities. A child with poor cognitive resources may show difficulties or delays in learning reading due to abstraction difficulty in decoding alphabetical abstract symbols. This learning disability cannot be considered as dyslexia.6,7,8 However, the long historical journey of clinical and experimental research allowed the researchers to reach some certainties: 1. Reading disorders may occur despite a good intelligence, a good verbal comprehension and production, the lack of visual or auditory disorders, sufficient motivation and environmental opportunities to learn.5 2. Dyslexic subjects are able to correctly perceive both real objects and their visual representations.1,9 3. However, they find difficulty in correctly read the written names of the same objects and can revert, omit, substitute written letters or words, showing difficulty in decoding the symbolic written language system, regardless their visual system or general intelligence.3,9 4. Visual defects ‘per se’ do not seem to explain the origin of a specific learning difficulty to read.10,11

Materials and Methods

The aim of the current study was to collect an overview of the references reporting a brief history of reading disorder, referred to as developmental dyslexia, over the years. The practical implication may be to better understand such reading disorder and improve the quality of treatment programs for dyslexic subjects. One hundred articles were analysed, including case report, review and meta-analysis. Pre-1990 articles have been excluded from the analysis, except for some to describe the historical excursus of the theoretical models on developmental dyslexia. Further exclusion criteria were the studies that did not add historical interest to the topic. A total of fifty-seven articles have been included in the current review using as search criteria keywords such as: neureodevelopmental disorders, reading disorder,
From a ‘visual defect’ to a multi-component disorder

Over a hundred years ago, assuming that an intelligent teenager had to learn to read, a British school physician could not understand why a smart and intelligent young boy, 14-year-old, had learning reading problems. Therefore, the school doctor diagnosed his young patient’s reading disorder as a visual defect suggesting a sort of therapeutic ‘eyes training’.

Similarly, a few years later in 1917, Hinshelwood, an Edinburgh ophthalmologist, considered dyslexia as a form of ‘congenital blindness for words’ and carefully distinguished the reading disorder from other clinical conditions in which total or partial inability to read may occur, such as poor intelligence, sensory difficulties, specific hearing impairment. Moreover, he hypothesized that the ‘purity of symptoms’ should be the crucial feature of dyslexia and, as for acquired blindness for words in adults, he postulated that the biological basis of the disorder was agenesis or developmental defect of the left angular gyrus.

In the first decade of the twentieth century, it was accepted as undoubted that dyslexia was for childhood the analogue of the ‘acquired blindness for the words’ of the adult. In contrast, on the basis of reading and spelling mistakes of dyslexics, Ortón viewed dyslexia as a failure in establishing cerebral dominance and accounted it as a language disorder. However, as the difficulty of learning to read has become more widely recognized as such, inconsistent data have emerged on its neurological basis, its nature and causes as well as on its existence as a clinical entity.

In the past, some authors denied dyslexia as a specific clinical entity ascribing it to factors such as teaching methods, inadequate family influences, emotional factors and primary visual defects. Then, two different frameworks emerged. The first considered the visuo-perceptual defect as the core of dyslexia and the other associated it with social and relational factors, such as poor education or emotional and psychological problems. Moreover, the distinction between developmental dyslexia and other categories of reading defects disappeared. As a consequence, a large body of studies collected conflicting data. These conceptual divergences persisted for a long time until the diagnosis of dyslexia was considered absurd and even a ‘mystique of learning difficulties’ was hypothesized. According to some authors most of children classified as dyslexic would rather be the result of inappropriate reading teaching methods. These extreme assumptions raise the problem of the diagnostic criteria to be used. The criteria were often different among the various researchers who considered subjects of different clinical populations as belonging to a single group of dyslexics. The most widespread criterion proved to be grossly inadequate: the child who has a reading level of two or more years lower compared to the attended class is dyslexic. This criterion does not take into account the intellectual level, the class (the first grade or the third grade), the cultural background of the quality of the school. The simple two-years criterion leads to overdiagnosis, while more rigid criteria risk false negatives, diagnosing as non-dyslexic subjects who are instead dyslexic. However, even when restrictive criteria were used, dyslexics differed from one another in terms of neurological, behavioral or psychometric performance. Therefore, different classifications were elaborated. A first one differentiated a ‘pure dyslexia’ from a ‘dyslexia plus’, depending on whether it was accompanied by ‘soft’ neurological signs. 2. A second one, considering the different patterns of reading errors, identifies three different dyslexic groups: the dysphonetic group, unable to integrate symbols with their sounds, the dyseidetic group, unable to perceive letters and whole words as configurations, and a mixed group with a combination of both dysphonetic and dyseidetic deficiencies.

3. A third classification considers the cognitive profile of dyslexics highlighted by the performances in verbal and non-verbal tests. It therefore seemed increasingly evident that dyslexia could be a multicomponent disorder, rather than a disorder resulting from a single visuo-spatial, visuo-motor or phonological problem. Therefore, the roots of developmental dyslexia move from the eyes to the brain. In the following years, a large number of researches has shown that the phenomenology of dyslexia manifests itself in a multifaceted modality with difficulties both in the visuo-spatial and in the phonological processing.

Evidence from autopsy and neuroimaging studies

Around the second half of the last century, research on dyslexia reached a new impetus from the histo-pathological and neuroimaging studies. For the first time, autopsy studies on the brain of developmental dyslexics showed a bilaterally large temporal planum, and the absence of the planum temporal asymmetry. In addition, such studies found, in the whole brain, cytoarchitectonic abnormalities, such as ectopic collections of neurons in layer one of the cortex, especially in perisylvian areas, in the thalamus lateral geniculate nucleus of the visual pathway and in the medial geniculate nucleus of the auditory pathway. Such cortical anomalies were more frequent in the areas surrounding Silvio’s fissures, in the left hemisphere, roughly corresponding to the frontal and temporal language areas.

Both reduced planum temporal asymmetry and cortical malformations suggest an anomalous brain development, in prenatal stages of corticogenesis, with abnormally high levels of surviving neurons, as a significant neuroanatomical association to developmental dyslexia.

Such abnormal neural migration affects the pattern of connectivity within and between the hemispheres, especially with the thalamus, ipsilateral and contralateral cortex and alters the development of these brain areas. Neuron ectopic collections may be associated with a developmental anomaly of the adjacent cortical layers (dysplasia).

Along with dysplasia and cortical ectopies, vascular anomalies often resemble small micro angiomas and sometimes arterio-venous malformations. Neuroradiological studies also showed a link between dyslexia and arteriovenous malformations, especially in males and in the left superior temporal areas.

Post-mortem and neuro-imaging studies provided strong neuroanatomical evidence on the structural and morphological differences between normal and dyslexic brains, giving a cogent contribution to understanding the neural and cognitive basis of developmental dyslexia as a language-based disorder.

Neural basis of reading and dyslexia

Data from brain imaging studies in
developmental dyslexia documented inter-related neural systems both in posterior and in anterior brain regions.\textsuperscript{35}

The cerebral posterior system includes two different and related structures: 1. a parieto-temporal area activates in analysis of the single word units (phonemes) and includes parts of the supramarginal gyrus in the inferior parietal lobule, parts of the superior aspect of the superior temporal gyrus and parts of the angular gyrus in the parietal lobe. 2. an occipito-temporal area, referred as the visual word-form area, activates for skilled and fluent reading.\textsuperscript{36}

The anterior system, instead, encompasses the left inferior frontal gyrus (Broca’s area) and the dorsal and ventral premotor regions and is associated with articulation and silent reading and naming.\textsuperscript{37,38}

In dyslexic readers the anterior reading system, in the left hemisphere, appeared slightly overactivated while the two posterior systems were under activated, strongly supporting the ‘neural signature of dyslexia’.\textsuperscript{35}

This neuroanatomical consistent evidence allows to understand the neural basis of reading and dyslexia as a language-based disorder supported by a basic deficit in phonological processing.\textsuperscript{35}

### Phonological model

In recent decades, on the basis of neuro-biological and neuroimaging studies, a new model of dyslexia has emphasized the role of language-phonological processing.\textsuperscript{39}

According to such a model, the core of dyslexia is a selective, specific impairment of the language-processing at the phonological level.

Learning to read implies the ability to associate spoken sounds with written letters and to reach a phonological awareness according to which written words represent the sounds of spoken words.\textsuperscript{2}

The word ‘man’ derives by the phonemes m/a/n as individual sounds combined with each other. Such word can be identified, understood and stored as the word ‘man’ after being analyzed in its individual phonemic units. In the spoken language, a genetic phonological module processes individual phonemes combining with each other into the word.

However, spoken language is a ‘natural’ system and the ‘combination’ of the phonemes is ‘automatic’. Written language, instead, is a ‘conventional’ system and reading is much more demanding and requiring a more intentional activity. The written word must be phonologically decoded and identified and, subsequently, understood by higher-level cognitive functions, such as intelligence and vocabulary.\textsuperscript{2,27,39}

Therefore, reading implies a consciously awareness of the phonological form of words. Such phonologic awareness may be lacking in dyslexic readers. Therefore, a phonological deficit in the ability to segment the written word into its underlying elements, impacts on the functionality of the higher-level linguistic processes involved in the attribution of meaning, in learning or recalling complex, new, long words, or in mechanical memorization and rapid ‘naming’.

Thus, two different but integrated neu-rocognitive processes are involved in reading: the first decodes the written symbol-linguistic code and the second one decodes the visuo-spatial characteristics of the same linguistic code. An impairment in one of the two neurocognitive processes or in both results in the reading disorder referred to as developmental dyslexia. However, written language is firstly a symbol system, although it is expressed in visuo-spatial graphemes, shapes, dimensions. A single clustering of letters configures different meanings according to the spatial relation between individual letters (w/a/s is ‘was’ and s/a/w is ‘saw’). Conversely, a capital or small letter has the same value, as a linguistic sym-bolic code, regardless of its visuo-spatial features. Letters such p/b/d/q/n/a are codes whose linguistic value is closely associated with their visuo-spatial features and orientation (right/left/up/down). The organization of a visual stimulus results from the ‘meaning’ assigned to the stimulus. In reading, ‘meaning’ of the visual stimuli is linguistic in nature and the lacking integration between letters and words derives from the inability to associate linguistic (the meaning) and visual dimension of the written word.\textsuperscript{9} In addition, reversal and perceptual errors are limited to printing in their own language and are therefore lin-guistic rather than visual.\textsuperscript{9,27}

A deficit in the phonological elaboration of written language tends to slow down or compromise performance in all tasks requiring word phonological segmentation and analysis, in the decoding of complex written texts and in the processes of reading automatization, in the timed reading and comprehension of the text, in the processes of new episodic verbal learning and in the mechanical memorization of verbal materi-al (tables, dates, nomenclatures, lists of words not associated), in the rapid naming and recalling of words, in the decoding and in the functional use of words without con-text, in the use of complex, long or unfamiliar and abstract words, in the answers to multiple choice written questions, in reading and writing of foreign words.\textsuperscript{40,41}

Many ‘compensated’ adult dyslexics may learn to read and can even excel in their academic performance and gain access to higher levels of the language system with excellent achievements.\textsuperscript{42} However, this often requires high costs and resources. Moreover, it can be compromised in partic-ular contexts and in stressful situations, such as, for example, a condition of written examination or a written timed task which puts a strain on the ability to quickly make decisions on phonological data. Some timed tests reveal that decoding remains very laborious for dyslexics: their ability to identify words is not automatic and it is charac-terized by extreme slowness in making decisions based on phonological data.\textsuperscript{2}

### Compensated dyslexia and reading strategy

Analysing a written text of an exam of an Italian thirty year old compensated dyslexic, graduated in law, we highlighted relevant morphological errors concerning the name declination, the verb conjugation, using the functions (articles, prepositions, conjunctions) and the phonological distortions, omissions and substitutions (e.g. ‘la Supra Corte’ for ‘la Suprema Corte’ ‘orignarmente’ for ‘originariamente’; ‘maggiormente dibatti’ for ‘maggiormente dibattuti’; ‘ad un contratto agenzia’ for ‘ad un contratto di agenzia’; ‘l’assenza di qualsiasi rapporto impediscono’ for ‘l’assenza di qualsiasi rapporto impedisce’; ‘l’esercizio non consentono’ for ‘l’esercizio non consente’; ‘dei mezzi del reato’ for ‘dei mezzi di esecuzione del reato’; ‘i coniugi provvedeva’ for ‘i coniugi provvedevano’).

Interestingly, errors were higher in the fair copy of the text than in the rough draft. ‘La Suprema corte’, in the fair copy, for example, is ‘la Supra Corte’, (with the obvious omission of the syllable ‘ma’), while in the rough draft was properly written. The quality of errors and their frequency can be considered as residual traces of a compensated dyslexic disorder. Moreover, the greater frequency of errors in the fair copy may be associated with the prevalent use of a semantic strategy. The good reader encodes words in a flexible way either by an analytical phonological approach to individual graphemes or by a global semantic approach to the whole words, more related to the meaning of the word in the context. The familiar word ‘mother’ may be correct-
ly read with a low phonological analysis and its meaning may be semantically perceived from the context. Conversely, a less familiar, longer, or more complex word, such as ‘electroencephalogram’, requires a phonological analysis of the various graphemes. In a semantic approach, the word ‘walked’ can be read as ‘walks’ if, in context, ‘walks’ appears as a significant meaning. The dyslexic reader favors semantic strategy, strongly associated with the meaning of words and context, rather than phonological strategy associated with the analysis of individual graphemes.

On these assumptions, in the dyslexic lawyer, during a written examination, the fair copy (that is the second version) probably represents a ‘more automated’ transcription of an already known and repeatedly read text, that no longer requires a phonological analysis. In this way, morphological errors may more easily appear. Conversely, in the rough draft (that is the first version), the words still represent a new production and they may be written with a more analytic phonological analysis and adherence to their orthographic structure. Therefore, morphological errors in the compensated dyslexic lawyer should be re-interpreted as a phonological impairment and as a preferential use of semantic strategy that focuses more on the meaning than the phonological analysis of the text.

Reading and cognitive processes

For the planning of targeted interventions and individual recovery programs, the identification of the cognitive abilities underlying the dyslexic reading disorder is crucial. In addition to phonological processing, numerous studies have focused on the links between phonological recoding and other cognitive processes, in particular on attention mechanisms. The generation of phonological codes from written words is not an automatic process but demands attentional resources, particularly in the development of fluent and skilled reading. Rather, attention is a critical and overlooked cognitive dimension, for translating print into speech and achieving fluent and automatic reading. Therefore, attention may be a crucial factor in reading and in reading difficulties. For example, attention-deficit/hyperactivity disorder and dyslexia are frequently observed in the same individual.

Moreover, since the 1960s, several studies have highlighted the presence of naming-speed deficit as a second core deficit in dyslexia. Further research revealed orthographic factors, extending the double-deficit hypothesis (phonological deficits and naming-speed deficits) to the triple-deficit hypothesis. According to such hypothesis, visual processing of orthographic information is viewed as an independent factor contributing to word recognition. Researchers have also suggested further deficits in poor reading. There is a body of evidence supporting that dyslexic readers could be deficient in basic visual processes.

Genetical basis

The relationship between genetic information and dyslexia continue to be a highly complex topic. Many family aggregation studies and, even more, twin studies are compatible with a genetic etiology for dyslexia. Moreover, several studies have highlighted a strong genetic basis and an high familiarity in the pathogenesis of dyslexia. Six candidate genes have been identified and there is evidence that four of these are involved in a genetic system crucial for axon growth and neuronal migration, in the prenatal brain development, affecting the timing of early neural migration and normal connectivity patterns in the brain. Chromosomal regions have been identified where the presence of dyslexia would appear to be linked to the susceptibility of some genes, suggesting that a disturbance in the neuronal migration and a reduced activity in the left hemispheric brain regions are pathophysiological correlates of dyslexia.

It seems that genetic influences appear to be most relevant on severe dyslexia, while mild or soft dyslexia may be less influenced by genes than environment. Therefore, it will probably need to identify the forms of family dyslexia that are most associated with genetic factors and to develop more targeted interventions for people at risk from the early school years.

Conclusions and practical implications

Therefore, in recent decades, advances in genetic, neurobiological and psychological research have provided a significant contribution for understanding of a reading learning disorder difficult to interpret. However, many questions about the best definition and treatment of dyslexia are still outstanding.

As a practical implication, knowledge of the history of developmental dyslexia should allow an approach to the problems of dyslexic subjects based on research acquisitions and on the neurological basis of this developmental selective language-related disorder.

This brief historical overview of developmental dyslexia studies, from the early reports suggesting a visual defect or focusing on the reversal errors, showed that developmental dyslexia should be considered as a heterogenous neurodevelopmental language-based disorder in learning to read, in decoding letters/sounds, in accuracy and word recognition, primarily resulting from both phonological processing and visuo-spatial and visuo-attentional impairment. Therefore, rehabilitation must focus on the recovery of language and visuo-spatial and visuo-attentional processes underlying the complex and multi-component developmental dyslexia.

Moreover, a large body of studies showed that dyslexia does not compromise per se reasoning skills, problem solving ability, conceptualization, critical thinking and vocabulary. As a practical implication, knowledge of the history of developmental dyslexia should allow an approach to the problems of dyslexic subjects based on research acquisitions and on the neurological basis of this developmental selective language-related disorder.

References


