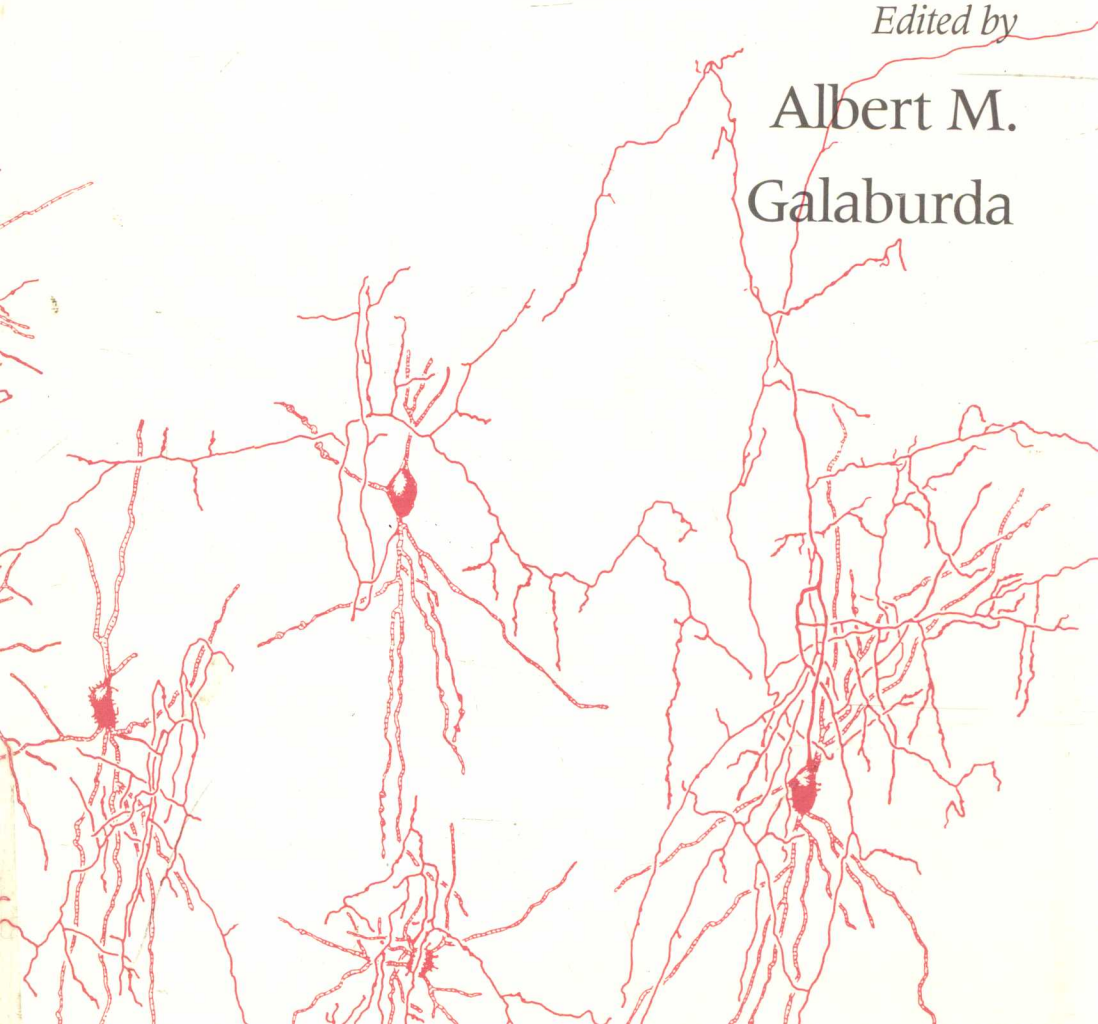
Dyslexia and Development

Neurobiological Aspects of Extra-Ordinary Brains

Edited by

Albert M. Galaburda



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江苏工业学院图书馆
藏书章

Harvard University Press
Cambridge, Massachusetts
London, England 1993

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Printed in the United States of America
10 9 8 7 6 5 4 3 2 1

This book is printed on acid-free paper, and its binding materials have been chosen for strength and durability.

Library of Congress Cataloging in Publication Data

Dyslexia and development : neurobiological aspects of extra-ordinary brains / edited by Albert M. Galaburda.
p. cm.
Includes bibliographical references and index.
ISBN 0-674-21940-6 (alk. paper)
1. Dyslexia—Pathogenesis—Congresses. 2. Dyslexia—Pathophysiology—Congresses. I. Galaburda, Albert M., 1948–
[DNLM: 1. Brain—physiopathology. 2. Dyslexia—physiopathology. WL 340.6 D998]
RC564.A56 1992
616.85'53—dc20
DNLM/DLC
for Library of Congress
92-49952
CIP

Foreword

The problem of developmental dyslexia came to light as interest arose in child neuropsychiatry, because of its intrinsic characteristics as well as its relationship to problems of right/left orientation, learning disorders, and personality deficits. In Barcelona, Eulalia Torras dedicated a monograph based on her doctoral thesis to the problem, and in 1964 a distinguished student of aphasiology, MacDonald Critchley, published a monograph on developmental dyslexia, which had significant influence. Later, with the contributions of Hier and colleagues and Galaburda and Kemper, the neurobiological substrate became the chief interest after the description of cytoarchitectonic abnormalities of the brain in dyslexic children, including a deviation from the standard pattern of brain asymmetry. Even now, though, it is not known whether abnormalities are present in all dyslexics, and it remains difficult to relate innate factors to educational and developmental problems of personality development.

On the basis of work by Patricia Goldman, Norman Geschwind suggested the possibility that, together with deficits related to the affected area, certain brain lesions could result in "superior functions" in the unaffected hemisphere. He commented that "such a mechanism may explain the superior right hemisphere capacities exhibited by dyslexic children." With regard to subjects with hyperlexia, who despite their apparent precociousness in reading aloud

show mediocre reading comprehension, Geschwind was of the opinion that "one could postulate that the area implicated in the learning of graphemic-phonemic conversions was appropriately developed, but its connection to other brain regions was deficient."

Another issue of interest regarding developmental reading disorders has to do with their eventual similarity, or lack thereof, to acquired reading disorders in the adult. Elizabeth Warrington has referred to acquired disorders as "dyslexias," but they are more commonly called "alexias" in the United States and in the European continent. In 1895, Déjerine and Vialet made a fundamental contribution to our understanding of acquired reading disorders by singling out the condition called "pure word blindness," or agnosic alexia without agraphia. In 1958, Alajouanine, Lhermitte, and de Ribaucourt-Ducarne insisted on the differentiation between agnosic and aphasic alexias. At that stage, four types of alexias were recognized: (1) pure, or agnosic, alexia without agraphia; (2) aphasic alexia; (3) alexia with agraphia, which was also recognized by Déjerine and which corresponds to the limiting cases (complete dissociation) of Wernicke's aphasia type III of Roch-Lecours and Lhermitte; and (4) spatial alexia, linked to the syndrome of left hemineglect, which results from right-hemisphere damage.

This classification of acquired alexias in the adult, accepted over the past twenty years, has recently been replaced by one based on a deeper understanding of the reading problems themselves. The previous classification was based more on the context in which the deficit occurred than on the intrinsic characteristics of the deficit. The difference between the two will be more deeply understood if one takes into consideration the strategies used by patients to carry out "residual" reading activities and the characteristics of the paralexias—semantic, morphologic, derivative, and others—produced. That which the patient does badly—the manner in which errors are committed and the "paralexic substitutions"—has more significance to the new classification than that which he or she *cannot* do. An example is the new view of aphasia, in which the presence of stereotypies, agrammatisms, and paraphasias is more illuminating than the initial mutism that follows some strokes.

The present classification of acquired alexias begins with the analysis of paralexias as carried out in 1973 by John Marshall and Freda Newcombe. In an illustrative schematic diagram, Marshall examines

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three positive mental routes to reading: (1) the phonologic route, (2) the direct (holistic, gestalt) route, and (3) the lexical route, in which morphologic decomposition relies on the visual-gestalt or holistic form of the word, which is segmented into its morphometric constituents (basic form, prefixes, suffixes).

Recently, Jordi Peña from our group has proposed the following functional classification of alexias.

PERIPHERAL ALEXIAS

Neglect alexia

Attentional alexia

Spelling dyslexia (Kinsbourne and Warrington). This is characterized by the ability of the patient to spell out words, letter by letter. Each letter must be named (often aloud) before the word can be identified. In this case, what is characteristic is not the type of error but the manner in which words are identified. Indeed, the name *spelling dyslexia* appears paradoxical or erroneous, in that it is precisely by spelling that reading is accomplished.

CENTRAL ALEXIAS

Surface dyslexia, characterized by paralexias derived from the inappropriate application of the rules for graphemic-phonemic conversion (Deloche and collaborators). In this syndrome there is an alteration in the direct route from the written word to the extraction of semantic meaning. There may also be a problem in orthographic knowledge. The capacity that remains reflects the use of a route mediated by graphemic–phonemic (word)–lexical (semantic) correspondences.

Phonological dyslexia (Marie-France Beauvois and J. Derousné). This syndrome is the opposite of the former, in that there is the inability to go from spelling to sound because of a failure to apply the rules of graphemic-phonemic correspondence. Instead, the direct route is used. High-frequency words are recognized more easily, whereas the ability to read nonsense words is markedly affected. Paralexic errors consist of substitutions by visually related words.

Semantic access alexia (Warrington and Shallice). The patient has difficulty in accessing meaning from the written word, as compared with the spoken word.

Deep alexia, in which there is an alteration in graphemic-phonemic conversion. The patient is unable to read nonsense words but in this case will produce semantic paralexias (e.g., "table" is read as "chair"). Reading concrete words is superior to reading abstract words, and there is marked diminution in the ability to read verbs and particles.

The anatomic localization of these syndromes remains tentative. In general, the topography of the dyslexic lesions is viewed in the context of the topography of classical aphasic syndromes. It is also possible to find similar clinical syndromes with different lesion localizations. In principle, surface dyslexia would be found in the context of Wernicke's aphasia or during some stage of the evolution of the syndrome of pure alexia. Deep dyslexia can be seen in cases of Broca's aphasia, while phonological dyslexia would be seen in association with Broca's aphasia, conduction aphasia, or Wernicke's aphasia. In such instances, specific symptom dissociations and/or neighborhood signs would help pinpoint the exact localization. Additional anatomic verification of the deficits in autopsy specimens, as well as in life through computerized tomography scans and magnetic resonance imaging, will lead to a better understanding of the anatomical bases for the various forms of alexia.

On the other hand, there is great interest in the relationship between acquired alexias and developmental reading disorders. John Marshall has pointed out that "there has been some progress in the phenomenological description of both acquired and developmental dyslexias, within a common frame." More recently, the same author debated with John Morton concerning the question of whether developmental dyslexia reflects arrest or metamorphosis of normal reading acquisition. In other words, does the anomalous reading represent reading at an earlier stage or reading that is not ever seen in normal development? In his commentary on the debate, Galaburda wrote that in order "to defend one position or the other, it is more appropriate, it seems to me, to compare the reading characteristics of adults with acquired dyslexia and those of developmental dyslexia to those of normal children learning to read, because it is

not at all clear that acquired or developmental dyslexics make solely errors of earlier stages of reading acquisition."

The work reported in this book reflects the variety of questions and approaches I have tried to present in these modest introductory comments.

Lluis Barraquer-Bordas

Preface

Plasticity is a property of the brain during development, and development lasts a lifetime. An important function of this plasticity is the adaptation of the brain to environmental demands. The brain's environment may be defined in broad terms: a gene product can be the environment of another gene; an incoming axon can cause a membrane to express a receptor on its surface; a signal from a distant endocrine gland, a hormone, can trigger a glial cell into undergoing mitosis—each of these being examples of the internal environment of brain structures. There is, moreover, the ordinary understanding of environmental influences: light coming in through the open eyes, which causes activity-dependent synaptic reorganization; or a baby's first exposure to its mother's language, which selects that language among all possible human languages to become the child's native tongue.

In these and many other ways, the environment affects the true expression of the genetic potential of the brain. Environment can be, and often is, abnormal. Brain injury is a common occurrence, and it may take place at any stage in life—before the cradle until the grave. The effect of brain injury is at least two-fold: it destroys components of the brain, the cells, connections, blood vessels (and the functions they support); and it changes the environment of the remaining "intact" brain, which leads to additional changes in brain

structure and in behavior. The changes that take place in the brain as a response to injury are probably more important than the actual loss of brain tissue, particularly when the latter is relatively minor.

All types of brain changes made as a response to injury are not possible at every stage of development. It is not known in great detail how much change is possible at any given stage, but it is likely that major reorganization of the brain's cellular architecture and connectivity becomes impossible soon after birth in the human. On the other hand, changes in local connections and in the synaptic architecture may continue much longer, perhaps throughout life. Even subtler changes, detectable only in the molecular characteristics of neurons, probably are caused by ordinary environmental changes from day to day.

It is reasonably likely that the same mechanisms of plasticity triggered by injury to the brain are also active during normal environmental fluctuation, although probably to a different extent and with different consequences. In other words, the ability of the brain to react to environmental changes was probably acquired over the course of evolution for the purpose of adaptation to gentle changes, but the violent changes associated with brain injury activate the same mechanisms of plasticity. Although the mechanisms for brain plasticity in response to ordinary environmental stimuli may be adaptive, the same mechanisms when initiated by brain injury need not always produce positive behavioral results. On the one hand—as is the case of the bone scar that is stronger than the surrounding undamaged bone—plastic repair of the brain might conceivably lead to functional strengths. On the other hand, repair after brain injury at any age could lead to anomalous circuitries that could predispose an individual to epilepsy, motor and sensory disturbances, or cognitive deficits. It is even possible to conceive that the same repair could produce strengths in some areas as well as weaknesses in others.

Evidence is presented in this volume that early brain injury and the response to injury may lead to brain malformations. Brain malformations are often associated with functional deficits. This is particularly clear for the major malformations, those that are incompatible with survival or that lead to mental retardation, disorders of motor and sensory behavior, or epilepsy. The effects of minor malformations are less well understood. Oftentimes small brain defects are missed in routine autopsies, and when they have been noticed,

they have been either ignored or thought to be trivial. But the need to consider them as potential culprits in developmental neurological disorders is illustrated by the fact that they have been found in individuals with epilepsy, learning disorders, fetal alcohol syndrome, and autism. Furthermore, the definition of a "minor" abnormality is based mainly on the appearance of brain matter after application of classical neuropathological techniques. These techniques cannot address questions about the disorganization of connections—local or distant—nor about significant distortion of the specific neuronal makeup of affected brain regions and their neighbors. Both types of change could indeed be accompanied by substantial and measurable disorders of behavior. In addition, animal models have been recently developed that imitate naturally occurring minor malformations in the human brain, and these have begun to show that "small" anatomic abnormalities may not be so minor after all.

A major impediment to the establishment of functional relevance for the minor malformations lies in the fact that their proposed effects may be subtle enough to require specialized testing during life. The early-developmental nature of the brain injury under question makes it likely that there will be compensation for the cognitive loss and that deficits will be seen only when specific cognitive strategies are assessed. In most cases it is not likely that a routine battery of tests will uncover meaningful information. The proper testing, however, is seldom available in medical records. Subtle learning or emotional disturbances of childhood are likely to be all but forgotten in the examination of adults, or they may be buried under the more dramatic events that led to death.

But this unfortunate state of affairs is likely to change, thanks partly to increased awareness among families and health care professionals, better assessment tools, including cognitive and morphologic assessments administered during life, and growing interest in the research community. It may be added that advances in the fields of neuroscience and cognitive science in the past fifteen years can now be gainfully applied to the generation of coherent theoretical perspectives and empirical approaches to the study of living humans and animal and computational models.

In 1988, I organized a conference in Florence to gather together a group of researchers and thinkers whose work I thought would be relevant to a research program aimed toward an understanding of

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learning disorders, especially developmental dyslexia. Topics discussed were language development, reading acquisition, literacy, machine models, inter-individual differences, developmental neuropathology, neurobiology of learning and memory, and more—indeed, a broad-brush approach. As predicted, many of the contributors were shy about establishing specific links between their fields of study and developmental dyslexia. Their timidity is understandable: developmental dyslexia is only informally defined in cognitive terms; there may be several unrelated types; the condition is likely to arise from a complex set of interactions between biology and culture, about which we know little; the biologic substrates of the disorder are only preliminarily described, leaving plenty of room for caution; and, last but not least of this list of problems, a well-developed cognitive neuroscience of normal reading and even language is not as yet available. This conference and related efforts to establish a cognitive neuroscience of learning disorders, however tentative, have already paid off, and they promise to continue to give results. There is growing interest among cognitivists and computational scientists in disorders of cognition, including reading—the Society for Neuroscience has an entry in its "key words" list for *dyslexia*, and the concept itself appears to have acquired tangible scientific as well as educational validity.

This book came about as a result of a second meeting, which took place in Barcelona. Each chapter emphasizes one aspect of research on the neurobiology of developmental dyslexia. My goal was to gather together a selective body of research on normal and abnormal brain development and on child neurology that would focus on the discovery that abnormalities of brain development, albeit possibly minor in comparison to others, may play a significant role in the pathogenesis of learning disorders, particularly developmental dyslexia.

The major themes in ongoing research bearing on the neurobiology of dyslexia are represented in this volume: plasticity during brain development leading to variation; the effect of sex hormones on brain differentiation; developmental neuropathology, including human descriptive neuropathology and animal models; cerebral lateralization and asymmetry; vision research; genetic factors in behavioral disorders; neuropsychology of language development; and the imaging of functioning, living brains. It is anticipated that in the next

decade, "The Decade of the Brain," we will see a convergence of research findings from each of these fields and arrive at a greater understanding of the development of the brain for cognitive and emotional behaviors.

The conference in Barcelona came about as a result of the warm generosity of Emily Landau and the personal involvement and support of Caryl Frankenberger of the Fisher-Landau Foundation of New York.

The hard work and dedication of Loraine Karol, administrative assistant to our Neurological Unit at the Beth Israel Hospital, guaranteed a flawlessly organized conference, comfortable travel, and pleasant stay for all speakers and guests in Barcelona. William Baker, executive director of the National Dyslexia Research Foundation, was responsible for establishing links with dyslexia researchers in North America and Europe, for organizing the dyslexia research community in Spain around the conference, and for many of the welcome amenities. Montserrat Estilles and her able assistants were our hosts and hostesses in Barcelona. Ruth Davis, U.S. consul in Barcelona, was supportive and helped us establish contacts with Barcelona officials. Judy Sharp in Madrid carried on as hostess when some of the group continued on to the capital city.

Speakers and guests alike made the conference a success beyond all our expectations. We were fortunate to have among us, in perhaps her last public appearance, Isabelle Liberman, whose contribution to the field of dyslexia is legendary.

My colleagues Gordon Sherman and Glenn Rosen offered helpful advice during the editing stages of the book, and Eileen Moran's and Meggin Sullivan's help were invaluable during the preparation of the typescript. Finally, Angela von der Lippe, Linda Howe, Pat Pershing, and Kate Schmit of Harvard University Press demonstrated great patience and encouragement when dealing with me on the long multiauthored manuscript.

To each I offer my grateful appreciation.

Albert M. Galaburda

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Regressive Events in Early Cortical Maturation: Their Significance for the Outcome of Early Brain Damage

Barbara L. Finlay
Brad Miller

Two features of the neocortex must be taken into account if we are to understand the mechanisms of its normal and abnormal development. First, the entire neocortex has a common organizational structure: the tangential organization of layers with their characteristic cell types, inputs and outputs, and the radial organization of the cortical column. Second, the cortex is locally differentiated with identifiable cytoarchitectonic areas that differ in their total cell complement, the proportions and sizes of cells in various layers, and specific types of input and output. We need to understand the general mechanisms that produce overall cortical structure and the means by which local differentiation develops.

The basic sequence of generative events that make the cortex is well known. Cortical neurons are generated from columnar epithelial cells residing in the ventricular zone (Sidman, Miale, and Feder, 1959). After their last cell division, neuronal cells migrate out to form the cortical plate, with each wave of cells coming to reside nearer the pial surface than the previous generation; thus the cortex is said to develop in an "inside-out" sequence (Rakic, 1974; Angevine and Sidman, 1961, 1962; see McConnell, 1988, for review). Cortical cells migrate along radial glial fibers, remaining in very close apposition to the fibers until they reach their destination (Rakic, 1971, 1972, 1974). The fundamental uniform scaffolding of the

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