



Behavioral and Brain Science

John L. Bradshaw and Jason B. Mattingley

# Clinical Neuropsychology

Behavioral and Brain Science

John L. Bradshaw
&
Jason B. Mattingley

DEPARTMENT OF PSYCHOLOGY MONASH UNIVERSITY CLAYTON, VICTORIA, AUSTRALIA



ACADEMIC PRESS

SAN DIEGO BOSTON NEW YORK LONDON SYDNEY TOKYO TORONTO This book is printed on acid-free paper.



#### Copyright © 1995 by ACADEMIC PRESS

#### All Rights Reserved.

No part of this publication may be reproduced or transmitted in any form or by any means, electronic or mechanical, including photocopy, recording, or any information storage and retrieval system, without permission in writing from the publisher.

Requests for permission to make copies of any part of the work should be mailed to: Permissions Department, Harcourt, Inc., 6277 Sea Harbor Drive, Orlando, Florida 32887-6777

#### Academic Press

A Harcourt Science and Technology Company 525 B Street, Suite 1900, San Diego, California 92101-4495, USA http://www.academicpress.com

#### Academic Press

Harcourt Place, 32 Jamestown Road, London NW1 7BY, UK http://www.academicpress.com

#### Library of Congress Cataloging-in-Publication Data

#### Bradshaw, John L., date.

Clinical neuropsychology: behavioral and brain science / by John L. Bradshaw, Jason B. Mattingley.

cm.

Includes bibliographical references and index.

- 1. Clinical neuropsychology. 2. Neuropsychiatric disorders.
- I. Mattingley, Jason B. II. Title.

[DNLM: 1. Neuropsychology. 2. Nervous System Diseases. 3. Mental

Disorders. WL 103.5 B812c 1995]

RC386.B72 1995

616.8-dc20

DNLM/DLC

for Library of Congress

94-49187

CIP

International Standard Book Number: 0-12-124545-4

PRINTED IN THE UNITED STATES OF AMERICA

01 02 03 04 05 06 EB 9 8 7 6 5 4 3 2

# Clinical Neuropsychology

Behavioral and Brain Science

# -Preface

If psychology is the scientific study of behavior, neuropsychology places that study firmly in the domain of its underlying neural and neuronal determinants. It has long been a truism that if one wishes to understand how a system works, a careful examination of all its potentialities for malfunction may provide unique insights. This volume will concentrate on the clinical aspects of brain malfunction, but will not ignore those experimental techniques, especially the newer ones of neuroimaging, which throw a complementary light on the general issue of brain—behavior interrelationships. While we can learn much from unfortunate experiments of nature, new minimally invasive neuroimaging techniques seem set to illuminate the essentially "mental" contributions of, for example, thought and imagination. For the first time healthy individuals can be scanned while mentally rehearsing all manner of material, and engaging in complex everyday tasks like viewing a video, composing music, or even knitting. The last, a project of our own, aims to determine those brain areas involved in complex, highly overlearned bimanually integrative skills that do not require vision.

We hope in this book to go some way toward simultaneously achieving several different aims: an account of the major neuropsychological and neurological syndromes in their own right; a description of those rare and intriguing anomalies of behavior, where relevant, which have long fascinated the general public from the reports of such gifted medical writers as Oliver Sacks and Harold Klawans; and an enhanced understanding of normal brain processes in language, speech, object recognition, attention, memory, movement control, and thought.

While psychology's historical roots lie within the twin fields of physiology and philosophy, neurology and psychiatry sprang from a common medical stem and only now, after a period of separation, are beginning once again to grow together. The driving force behind their reunion is an increasing awareness of the organicity of so many disorders. The behavioral neurologist is of course comfortable with the aphasias, agnosias, amnesias, apraxias, and other movement disorders, and the clinical neurologist treats the patient with Parkinson's disease

xiv Preface

(PD); however, the Huntington's disease (HD) patient, with very similar disabilities, may be managed by the psychiatrist, who of course also treats patients with Tourette's syndrome (TS), schizophrenia, and mood disorders. The discipline of neuropsychiatry is doing much to blur many of these old lines of demarcation.

Lost or altered function can arise from destruction (by stroke, injury, or tumor) of the brain tissue directly involved in mediating that function, as with aphasia, apraxia, or agnosia; it can also stem from loss of activation due to damage elsewhere in the system, as in PD, or from changes in neurotransmitter activity or balance, as with the affective disorders. Clearly there are more degrees of freedom for treating disorders of the last type than is the case with the first, though cell transplantation is being employed or actively considered in the treatment of Parkinson's, Huntington's, and even Alzheimer's disease (AD).

Apart from practical handbooks of neuropsychological testing procedures, textbooks of neuropsychology have tended to adopt either a cognitive or a more traditional biological emphasis. The former owes much to modern computational and information-processing theory, and more or less explicitly rejects biological substrates in its emphasis upon software and "architecture." Indeed, for related ideological reasons it tends to eschew group studies, arguing that every individual, in sickness and health, is unique. The other, more traditional, biological approach, which we have adopted, seeks to ground function within structure, and to discuss commonalities of deficit consequent upon similarities of lesion. While we admit to finding this latter approach more satisfying, due presumably to our biological biases, and suspect that it may lend itself more readily to practical applications or therapeutic interventions, we would emphasize the potential complementarity of the two traditions.

The genesis of this volume arose from our teaching (senior undergraduate and graduate) and research. At one time or another we have empirically investigated aspects of many of the syndromes described herein, and we would emphasize the reality of the two-way nexus between teaching and research; both stimulate each other in unforseen ways. A single convenient, integrative volume could not, we felt, religiously cover all primary sources, so we have selectively built upon existing major integrative reviews, where available, and fleshed them out with new, major, provocative, or intriguing material from primary sources published within the last two or three years. Similarly, rather than meticulously referencing every statement, we have tended to refer the reader to the major reviews listed as Further Readings at the close of each chapter, though occasionally, as in Chapter 6, in the absence of suitable material, we have adopted more traditional referencing procedures. The material in Further Readings has been carefully selected to provide an up-to-date, balanced, and comprehensive overview of the issues addressed. Some items in the Further Readings and not previously cited within the chapter may be of major integrative significance. The provision of a Summary and Conclusions section to each chapter aims to provide both a general survey and a balanced judgment of any controversial aspects previously discussed in detail. Not all chapters are of similar length, as their designation has been largely a matter of content. Thus controversial or unresolved issues have been treated at greater length, whereas short bridging chapters (e.g., Chapter 11) provide the necessary (largely nonclinical) background to those that follow. We have drawn together and reviewed many diverse areas of information, but if at times we may appear uncritical, it is because so much of the information is often so new and controversial that it cannot yet be properly assessed and evaluated. While we have attempted to address most major areas of relevance, and hope to have included most integrative reviews up to 1994, we note that the field is far too dynamic for us to be able to offer anything completely free of personal bias, or totally comprehensive or exhaustive.

The book is aimed at senior undergraduates, postgraduates, and researchers in the behavioral and medical sciences. We are extremely grateful to the many individuals who so readily provided us with illustrative material, and hope we have been able to acknowledge everyone appropriately. Our grateful thanks go to our partners, who are also our colleagues, Judy Bradshaw and Ada Kritikos, to our drafting artist and photographer Rosemary Williams, and to our research colleagues in psychology, neurology, and psychiatry, especially Jim Phillips, Bob Iansek, and Ed Chiu.

# Contents -

#### Preface xiii

Chapter 1 An Introduction to Clinical Neuropsychology	
Neuropsychology, Neuropsychiatry, and Behavioral Neurology 2	
Important Assumptions in Neuropsychology 3	
Conceptual Issues in Neuropsychology 6	
Cerebral Circulation and Stroke 11	
Stroke Syndromes Associated with Lesions in Major Arterial Fields	13
Neuroimaging Techniques 17	
Neural Networks 21	
Summary and Conclusions 22	
Further Reading 24	

## Chapter 2 Disorders of Speech

The Aphasias

Morphological Asymmetries in the Brain 26
Other Evidence for Evolutionary Adaptations for Language 28
Aphasia: General Considerations 31
Wernicke's Aphasia 34
Broca's Aphasia 37
Other Types of Aphasia 38
Subcortical Aphasias 42
Illustrative Case History of Paroxysmal Aphasia 45
Summary and Conclusions 47
Further Reading 49

#### Chapter 3 Disorders of Reading and Writing

The Alexias and the Agraphias

Alexia with Agraphia 52

Alexia without Agraphia, or Pure Alexia 53

Surface Dyslexia 54

Spelling Dyslexia 54

Deep Dyslexia 55

Phonological Dyslexia 57

Literal Alexia 57

The Agraphias 57

Preservation of Implicit Understanding in Language Processing 58

The Localization of Language Sites by Positron

Emission Tomography 60

The Localization of Language Sites by Stimulation Mapping 63

The Localization of Language Processes in Congenitally Deaf Users of Visual Signing 64

Summary and Conclusions 65

Further Reading 67

# Chapter 4 A Right-Hemisphere Contribution to Language and a Calculation Deficit

Major Left-Hemisphere Damage in Infancy 69

Subtotal Left-Hemisphere Damage in Infancy 70

Major Left-Hemisphere Damage in Adults 70

Subtotal Left-Hemisphere Damage in Adults 70

Evidence from Deep Dyslexia 71

Commissurotomy Studies 72

Right-Hemisphere Damage in Adults 74

Acalculia 76

Summary and Conclusions 80

Further Reading 81

#### Chapter 5 Disorders of Object Recognition

The Agnosias and Related Phenomena

Apperceptive Agnosia 86

Dorsal Simultanagnosia 87

Ventral Simultanagnosia 93

Associative Agnosia 94

Optic Aphasia 95

Category-Specific Deficits 98

Tactile Agnosia or Astereognosis 101

Auditory Agnosia 102

Amusia 103

Prosopagnosia 106

Color-Related Disorders 113

Disorders of Route Finding: Topographical Agnosia or Topographagnosia 114

Horologagnosia: A Recently Described Impairment of the Ability to Tell the Time 115

Disorders of Distortion and Perseveration 116

Visual or Optic Allesthesia 117

Delusional Misidentification Syndromes 117

Summary and Conclusions 119

Further Reading 122

## Chapter 6 Disorders of Spatial Cognition

Unilateral Neglect

Clinical Descriptions of Unilateral Neglect 125

Terminology and Deficits Related to Unilateral Neglect 129

Etiology of Unilateral Neglect 134

Neuroanatomical Correlates of Unilateral Neglect 134

Models of Unilateral Neglect 138

Current Issues in Unilateral Neglect 156

Recovery and Rehabilitation 174

Summary and Conclusions 183

Further Reading 186

viii Contents

#### Chapter 7 The Acallosal Syndrome

Commissurotomy Studies

The Forebrain Commissures 188

Prior Animal Studies 190

The Human Acallosal Syndrome 191

Zaidel's "Z" Lens Technique of Prolonged Visual Lateralization 193

Subcortical Transfer of Information across the Divided Forebrain? 194

Independent Processors, Wills, and Streams of Consciousness? 197

The Congenital Acallosal Syndrome 199

Case History: Alien-Hand Syndrome 200

Summary and Conclusions 203

Further Reading 204

#### Chapter 8 Disorders of Memory

The Amnesias

Working Memory 209

Regions Involved in Long-Term Memory 211

Temporal Lobe Amnesia 214

Diencephalic Amnesia and the Wernicke-Korsakoff Syndrome 215

Preserved Implicit Covert or Incidental Memory 220

Basal Forebrain Amnesia 222

An Overview of the Structures and Functions Involved in the Amnesic Syndrome 223

Case History: H. M. and the Lasting Consequences of Bilateral Medial Temporal Lobectomy 225

Summary and Conclusions 226

Further Reading 228

## Chapter 9 Dementia and Alzheimer's Disease

Causes of Dementia 232

Alzheimer's Disease: General Considerations 233

Criteria for Diagnosis and Manifestations of Alzheimer's Disease 234

Memory Dysfunction in Alzheimer's Disease 236

Language Deficits in Alzheimer's Disease 237

Other Deficits 238 Neuropathology in Alzheimer's Disease 238 The Role of  $\beta$ -Amyloid in the Etiology of Alzheimer's Disease 240 The Inheritance and Genetics of Alzheimer's Disease 242 Summary and Conclusions 246 Further Reading 248

#### Chapter 10 Movement Control I

Cortical Systems and Apraxia

Prefontal Cortex 252

Premotor Cortex and Supplementary Motor Area 257

Primary Motor Cortex (Motor-Sensory Cortex) 261

Parietal Cortex 263

Apraxia: A Disorder of Voluntary Gesture and Action 264

Summary and Conclusions 274

Further Reading 276

#### Chapter 11 Movement Control II

Cerebellum and Basal Ganglia

Cerebellum 278

The Basal Ganglia 282

Summary and Conclusions 288

Further Reading 288

### Chapter 12 Subcortical Movement Disorders I

Parkinson's Disease

Parkinson's Disease, Aging, and the Dopaminergic System 290

Positive and Negative Symptoms in Parkinson's Disease 294

Deficits in the Internal Generation of Programmed Activity 296

Cognitive Impairments 298

Dementia and Depression in Parkinson's Disease 301

Disease Progression 303

Management of the Disorder 303

Causes of Parkinson's Disease 306

Summary and Conclusions 308 Further Reading 310

#### Chapter 13 Subcortical Movement Disorders II

Huntington's Disease

Prevalence, Age of Onset, and Inheritance of Huntington's Disease 312

Symptomatology of Huntington's Disease 314

Cognitive Deficits in Huntington's Disease 316

The Neuropathology of Huntington's Disease 318

Summary and Conclusions 321

Further Reading 322

#### Chapter 14 Other Movement and Neuropsychiatric Disorders

Tics, Tourette's Syndrome, Obsessive-Compulsive Disorder, Autism, Affective Disorders, and Other Such Disabilities

Extrapyramidal Movement Disorders 325

Tic Disorders: General Features 329

Tourette's Syndrome: General Symptomatology 332

Hereditary and Environmental Factors in Tourette's Syndrome 335

Anatomical and Neurochemical Aspects of Tourette's Syndrome and Obsessive–Compulsive Disorder 336

Obsessive-Compulsive Disorder: General Symptomatology 338

Autism 340

Other Neuropsychiatric Deficits: Aggression and Violence 346

Affective Disorders 347

Summary and Conclusions 349

Further Reading 351

#### Chapter 15 A Disorder of Thought

Schizophrenia

One Disease or Many? 354

Course and Progression of Schizophrenia 356

Attentional Deficits 360

Feedback and Feed Forward in Schizophrenia 361

Contents

Brain Anomalies in Schizophrenia 362

Drug Treatment of Schizophrenia: The Dopamine Hypothesis 366

Schizophrenia as an Inherited Disorder 367

Environmental Factors 368

Case History 370

Summary and Conclusions 370

Further Reading 372

#### Chapter 16 Final Thoughts

The Present and Future Status of Experimental and Clinical Neuropsychology 373

References 381
Author Index 431
Subject Index 451

# An Introduction to Clinical Neuropsychology

The inscrutable arcanum of the organization of the brain . . . Attracted us irresistibly. We saw that an exact knowledge of the structure of the brain was of supreme interest for the building up of a rational psychology. To know the brain, we said, is equivalent to ascertaining the material course of thought and will.

-Santiago Ramón y Cajal, Recollections of My Life

The basic precept of neuroanatomists such as Cajal, that explanations of human behavior can develop from an understanding of the structural organization of the brain, represents an approach to brain-behavior relationships that lies at the opposite end of the spectrum to that of neuropsychology. From its narrowest interpretation, the primary goal of neuropsychology is to use detailed behavioral analyses to infer underlying structural and functional properties of the brain. Thus, the kinds of data yielded by neuroanatomical and neuropsychological approaches are likely to be quite different, even though their ultimate goals are essentially the same. Moreover, although the levels of explanation offered by the two approaches are often different, the insights provided by one may be useful in refining and constraining the interpretations of the other. In this volume, although our view of brain-behavior relationships is taken principally from a neuropsychological perspective, we shall also draw on insights obtained from neuroanatomical and neurophysiological studies. This reflects our basic philosophy that cognitive processes do not exist in a vacuum; they are determined by the inherent structure and function of the neural substrate.

In this introductory chapter, we provide an overview of some of the important conceptual and methodological issues in neuropsychology. Our intention is to familiarize the reader with the assumptions and principles upon which hypotheses about brain-behavior relationships are based, and to explain the different approaches to data acquisition and interpretation in neuropsychological research. In addition, because we place considerable emphasis on studies of brainlesioned patients, we have included sections on the cerebral vasculature and on the behavioral consequences of pathologies involving specific arterial territories. Finally, we consider two modern techniques, neuroimaging and artificial neural network modeling, both of which have had a considerable impact on contemporary models of the structural and functional organization of the brain.

## Neuropsychology, Neuropsychiatry, and Behavioral Neurology

The discipline of neuropsychology began in the late 19th century with the patient studies of Broca, Wernicke, Lichtheim, and Hughlings Jackson. These workers adopted the principle of functional localization, in which specific cognitive functions are assumed to be subserved by discrete brain regions. Using the method of anatomoclinical correlation, these workers established associations between damage to particular brain regions and distinct manifestations of behavioral anomalies such as aphasia. The localizationist approach, which developed from Gall's original, though misguided, attempts to localize such various complex behaviors as "philoprogenetiveness" according to the topography of an individual's skull (Spurzheim, 1815; cited in Farah, 1994), became the cornerstone of the developing discipline of neuropsychology. However, whereas the early anatomoclinical studies persisted in assigning complex functions to specific brain regions, it was not until Hughlings Jackson established his famous dictum, that localizing a lesion and localizing a function are conceptually distinct, that real progress was made in elucidating the nature of human brain-behavior relationships.

Despite its relative youth, neuropsychology has already found a distinct niche within the fields of medicine and psychology. There are, however, several other disciplines that are clearly related; thus, neuropsychiatry involves "understanding the neurobiological basis, optimal assessment, natural history, and most efficacious treatment of disorders of the central nervous system with behavioral manifestations" (Cummings & Hegarty, 1994, p. 209). One subtle distinction between neuropsychology and neuropsychiatry is that the latter is often motivated by a desire to understand disordered behavior in terms of its implications for diagnosis and treatment. Similarly, the discipline of behavioral neurology (Devinsky, 1992) involves the use of data concerning the anatomy and physiology of the central nervous system (CNS) to guide interpretations of disordered behavior consequent upon neural damage. Although neuropsychology shares these aims, at least insofar as it too is concerned with patient diagnosis and management, it is ultimately concerned with elucidating the mechanisms underlying abnormal and normal behavior. Indeed, modern neuropsychology is based on data obtained not only from brain-damaged patients, but also from normal individuals; it exploits over 100 years of research in experimental psychology to help explain the patterns of disordered perceptual, cognitive, and motor processes seen in patients with neural damage.

There is also a sense in which neuropsychology itself can be further divided into distinct subspecialties. Whereas clinical neuropsychology is concerned with explaining how specific patterns of disordered human behavior may arise from disruption of particular brain processes, cognitive neuropsychology is focused primarily on explaining disordered behavior in terms of disruption to particular information-processing units, with (at best) only peripheral concern for their anatomical or physiological instantiation. Thus, the aim of cognitive neuropsychology is to understand information processing independently of the neural hardware. The reason for this approach, it has been argued (e.g., Caramazza, 1988), is that we know too little about normal brain processes and behavior to begin relating the two; this limitation becomes even more obvious when we attempt to understand the behavioral consequences of brain damage. An alternative view is that despite the obvious gaps in our knowledge, we nevertheless have a sufficiently detailed database from which to begin relating results of behavioral analyses with those obtained from studies of brain anatomy and physiology (e.g., Kosslyn, 1994). It is this latter view that we have adopted in this book; although in some cases the links we have proposed between particular behavioral anomalies and underlying brain processes remain tentative, we believe such endeavors facilitate the iterative process of developing and refining models of normal and disordered behavior.

## Important Assumptions in Neuropsychology

#### Modularity

The prevailing assumption in neuropsychology is that cognitive processes are organized into distinct processing units or *modules*. In one conceptualization, modules are hardwired, autonomous, computational mechanisms that perform highly specific functions, such as feature detection (Fodor, 1983). The outputs from several modules may be combined in the service of more complex cognitive operations, though the content of any given module is not available to consciousness. The concept of modularity, or some variant thereof, underpins most theorizing in modern neuropsychology (Shallice, 1988). It is appealing from both a computational and an evolutionary perspective; dividing a computational system into smaller subunits permits rapid, parallel processing of many different inputs, and it permits changes in specific subunits without necessarily affecting the operations of others. In addition, local damage may impair only a subset of specialized functions, rather than causing a global reduction in processing efficiency.