Coma and Disorders of Consciousness



Second Edition

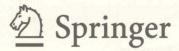
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Coma and Disorders of Consciousness

Second Edition



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Coma and Disorders of Consciousness

To medical teams and families we see every day and who inspire us.

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Foreword I

Consciousness is synonymous with human existence. Rene Descartes' bold proclamation, "Cogito ergo sum" (I think therefore I am), elegantly captures this premise. The clear inference here is that self-doubt about one's own existence establishes proof of one's existence. We exist because we know we exist. But how does one come to know that someone else is aware of his existence? Without direct access to knowledge of the self, it is impossible to prove (or disprove) the awareness of another. In normal consciousness, this problem is obviated by the multitude of behavioral expressions of conscious awareness manifested by living beings nearly every waking moment. Words, gestures, and actions, the "stuff" of consciousness, provide compelling evidence of the inner life of another. A small but significant percentage (5–10%) of those who sustain severe acquired brain injury experience prolonged disturbance in consciousness. Most will eventually recover at least basic capacity for self and environmental awareness, but this may not occur for many months and, in some cases, years. During this period of severely altered consciousness, one's existence may be stripped of the usual harbingers of an active inner life. Sensory, motor, language, perceptual, and drive systems may all be compromised in the aftermath of severe brain injury. Consequently, the repertoire of behaviors available to signal retention of conscious awareness may be dramatically narrowed or lost altogether. This predicament presents one of humankind's greatest existential dilemmas—is consciousness lost, or simply no longer apparent? This question is at the heart of Coma and Disorders of Consciousness edited by Caroline Schnakers and Steven Laureys. Both researchers are responsible for many seminal papers in this rapidly advancing field. Together, they have assembled an outstanding list of authors and have compiled a comprehensive volume that aptly depicts the state of the science in assessment and treatment of patients with disorders of consciousness (DOC). The book opens with a discussion of the complexities involved in behavioral approaches to assessment. Despite the challenges outlined, behavioral methods remain the "gold standard" in diagnostic assessment. The second chapter on prognosis reviews recently published long-term outcome studies, which have shed new light on the potential for meaningful late recovery potential in a substantial minority of persons with DOC. The next three chapters review novel functional viii Foreword I

neuroimaging and electrophysiologic approaches to assessment designed primarily to identify neurophysiologic signatures of consciousness in persons who lack behavioral expressions of self or environmental awareness. The issue of caregiver burden is explored in chapter six from a multidimensional perspective that considers the interpersonal and subjective impact of long-term caretaking. There are three chapters dedicated to the ubiquitous secondary sequelae of severe brain injury: spasticity, swallowing disorders, and sleep disturbance. The authors describe the neural systems underpinning these disorders and thoughtfully discuss their relation to impaired consciousness. Chapters 10-12 provide a review of treatments that aim to promote recovery by stimulating preserved brain circuitry. Interventions range from relatively low-cost, readily accessible procedures such as sensory stimulation and off-label drugs to highly specialized noninvasive and invasive electrical stimulation techniques. The book concludes with a provocative look at ethical dilemmas, states of consciousness associated with near-death experiences, and, finally, the future of coma science. Readers of Coma and Disorders of Consciousness will come away with a wealth of new knowledge about the science of consciousness and a profound sense of wonder in its majesty.

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Foreword II

During the last 30 years, I have had the opportunity to observe the evolution of knowledge as related to assessment and management of persons with disorders of consciousness (DOC). There has been much written, much debated, and much learned during that period of time. The neuroscientific understanding of DOC has matured significantly from basing opinions predominantly on neurological dogma to having evidence-based data in many areas to guide recommendations germane to assessment, prognosis, and treatment of this challenging population of patients.

As someone who had the pleasure of working with both Drs. Laurevs and Schnakers, I was honored to be asked to write a foreword for Coma and Disorders of Consciousness, a timely and important contribution to the medical literature in this challenging area of medicine. The neurological spectrum that is subsumed under DOC is diverse, complex, and mystifying—this has served as a nidus for many researchers to attempt to define, explore, and better understand the nature of this condition, the essence of which is rooted in the concept of consciousness. But what truly defines consciousness in terms of the degree or breadth of conscious awareness in a given individual? Must conscious awareness include both awareness of self and environment? Should we assume someone is unaware because either we cannot show that they are aware or because they cannot tell us they are aware? How should our ability to prognosticate outcomes impact on clinical service provision including withdrawal or withholding of care if such is being considered? How should pain be assessed and treated in such individuals and what mechanisms are responsible for and differentially delineate the perception of pain versus the more complex phenomenon of suffering? Should the idea of treating someone to allow them to potentially become somewhat more aware be a worthwhile outcome if they were to remain severely disabled and dependent on others for care? This volume will help those engaged with this patient population, such as the treating clinician, family member, or advocate, to explore and stimulate improved practice and to further research in this area of neuroscience.

The text is unique and timely on many fronts, but most importantly it provides hope where often none may have existed and awareness of advances where many would have dismissed the potential for same. The clear message of this text is that Foreword II

although the controversies associated with DOC remain partially shrouded in mystery, we are emerging into an era of better understanding that ultimately will positively impact clinical care and decision making.

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Preface

Consciousness is a word worn smooth by a million tongues. Depending upon the figure of speech chosen it is a state of being, a substance, a process, a place, an epiphenomenon, an emergent aspect of matter, or the only true reality.

George Armitage Miller

Fifty years ago, the field of disorders of consciousness was a very limited field of research. Severely brain-injured patients, who are most likely to present impaired consciousness during recovery, often died. In the 1950s, the introduction of artificial breathing changed everything. The life of these patients could be extended even in cases of severe lesions to brain areas supporting the control of vital functions. The clinician started to face patients who were alive but not reactive to their surroundings. In this context, a new field was called to emerge, the disorders of consciousness. In the 1960s, Plum and Posner defined for the first time a clinical entity called the coma. Slightly later, Jennett and Teasdale developed the well-known Glasgow Coma Scale for assessing the progress of comatose patients in intensive care units. The 1980s was characterized by the development of a new kind of treatment, the sensory stimulation programs. Finally, in the late 1990s, the emergence of neuroimaging techniques opened new opportunities to study brain reactivity in patients with severe brain injuries.

However, in spite of the medical advances and the increasing number of severely brain-injured patients, the assessment and treatment of patients recovering from coma represents a very difficult and delicate task even today. The detection of signs of consciousness is complicated by the frequent motor and cognitive limitations of these patients. Treatment options are nearly absent, leaving the clinician often with a situation of palliative rather than restorative care. Even in an experimental setting, the study of patients in a coma or related disorders of consciousness is a real challenge. These patients are easily exhausted, limiting the assessment window, and spontaneous motor reactions have to be controlled for. The development of a research environment adapted to the scientific investigation of these patients is time consuming and requests important clinical and scientific expertise. For over two decades, international research teams have been working on the scientific exploration

of disorders of consciousness, with both scientific and clinical research agendas. These research teams, bridging various medical (neurology, neurosurgery, intensive care, anesthesia, physical medicine, otorhinolaryngology) and paramedical disciplines (psychology, speech therapy, physical therapy) as well as engineering and biological disciplines, have been a major player in the development of new assessment, communication, and treatment techniques for disorders of consciousness at both behavioral and neuroimaging levels. Gathering an international crowd of experts, this version should offer readers an overview of the most recent advancements in this domain.

By focusing on both clinicians and researchers as potential readers of this book, we decided to include well-established findings about diagnostic/prognostic criteria, ethical issues, assessment techniques (i.e., behavioral scales, electrophysiological explorations, and structural/functional neuroimaging), and treatment procedures, but also techniques under development (i.e., neuromodulation) which, we hope, will stimulate ideas for future research.

In conclusion, we hope to have reached our aim by offering a comprehensive and reader-friendly book to readers both familiar or not with the difficult but intriguing field of disorders of consciousness.

We hope you enjoy reading this book.

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Chapter 1 Behavioral Assessment and Diagnosis of Disorders of Consciousness

Caroline Schnakers and Steve Majerus

Abstract Behavioral assessment is a critical step for the detection of signs of consciousness and, hence, for diagnosis of states of altered consciousness. However, because of the presence of compromising factors such as severe functional and cognitive impairment, accurate diagnosis is a challenging enterprise, leading to serious consequences on the patient's ongoing care but also on the patient's end of care. In this review, we will describe the behavioral characteristics of the main clinical entities through which severely brain-injured patients transit before fully recovering from coma, we will describe methods for assessing consciousness at the bedside, and we will discuss the existing tools that help clinicians formulating an accurate diagnosis.

Introduction

During these last years, there has been increasing fascination for the field of disorders of consciousness. Due to progress in intensive care, more and more severely brain-injured patients survive their initial brain insult, but many of these will go through various states of impaired consciousness. The prevalence of these patients is estimated at 46 per million in the United States, 14 per million in Great Britain [1], and 36 per million in Belgium [2]. Many patients will remain in a vegetative state for a more or less extended period of time before regaining some level of consciousness (minimally conscious state). Some of these cases have received important coverage

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