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978-0-521-82582-5 - Sudden Death in Infancy, Childhood and Adolescence, Second Edition

Roger W. Byard

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Sudden Death in Infancy, Childhood and Adolescence

This unique, comprehensive survey of virtually all aspects of sudden death in infancy and childhood will be an essential reference source for pathologists, clinicians, and lawyers who deal with such cases. Individual sections deal in detail with deaths due to inflicted and non-inflicted injuries and due to natural diseases. This new edition includes 1200 new references, 300 new illustrations, and an extensively revised chapter on sudden infant death syndrome. The intentional injury chapter has additional material on head trauma, the biomechanics of injury, neonaticide, suicide, and subtle and unusual trauma. The chapter on non-intentional injury has also been expanded to reflect more accurately its importance as a cause of death. Deaths in the first week of life are also covered. In addition, this new edition addresses the full range of natural causes of death and their pathological investigation undertaken in light of advances in our understanding of genetic susceptibility and pathophysiology.

Roger W. Byard qualified in medicine in Australia in 1978 and obtained an LMCC in Canada in 1982. He holds fellowships in anatomical pathology in Canada (FRCPC), the UK (FRCPath), and the USA (FCAP), and in family medicine with the Canadian College of Family Physicians (CCFP). He has a specific interest in sudden infant and childhood death. He has written over 270 papers in peer-reviewed journals, and 35 chapters, many of which deal with this subject. He has also presented or co-authored over 200 papers at national and international meetings. In addition to *Sudden Death in Infancy, Childhood and Adolescence* he has co-edited *Sudden Infant Death Syndrome: Problems, Progress and Possibilities* (2001), and is also currently co-editing an encyclopedia of forensic and legal medicine. He has an interest in preventive pathology and coordinates childhood

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accident prevention through the “Keeping Your Baby and Child Safe Program” in South Australia. Professor Byard has presented workshops on pediatric forensic pathology and medicine in a number of countries in Europe, North America, Africa, and Asia.

From reviews of the first edition

“This is an outstanding book for practitioners, pathologists, and researchers . . . Each chapter reads like a conference in which the pathologist holds the clues and answers questions from the clinician and other investigators from the autopsy table.”

R. L. Ariagno, *New England Journal of Medicine*

“ . . . the overall impression is of a volume based on sound scholarship and experience.”

W. R. Roche, *Lancet*

“ . . . a valuable new addition to the literature.”

S. Gould, *Archives of Disease in Childhood*

“I highly recommend this book as a unique and indispensable reference for all forensic pathologists.”

S. Dana, *American Journal of Forensic Medicine and Pathology*

“ . . . it is the book to turn to when faced with the investigation of a sudden death in infancy or childhood, or the interpretation of findings in these cases.”

J. Keeling, *Paediatric and Perinatal Epidemiology*

“The authors have produced a single, comprehensive source of information on virtually all aspects of sudden death in infants and children.”

From the foreword to the 1st edition by V. DiMaio, Editor in Chief, *American Journal of Forensic Medicine and Pathology*

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Sudden Death in Infancy, Childhood and Adolescence

Second Edition

Roger W. Byard

Specialist Forensic Pathologist, Forensic Science
Centre, Adelaide; Clinical Professor, Departments
of Pathology and Paediatrics, University of
Adelaide; Consultant Paediatric Forensic
Pathologist, Child Protection Unit, Women's &
Children's Hospital, Adelaide, Australia



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What greater pain can mortals bear than this;
to see their children die before their eyes?

Attributed to Euripides 480–406 BC

(Russell-Jones, D. L. (1985). Sudden infant death in
history and literature. *Archives of Disease in
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To Renée, Alice, and Sophie

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Foreword

It is a somewhat daunting task to provide a foreword to such an impressive work of scholarship, which now encompasses a wide spectrum of deaths from birth to early adulthood. I have watched Professor Byard's *magnum opus* develop over a number of years, as I was an examiner for his doctoral thesis from which the first edition of his textbook evolved, and I now see the second edition to be a more extensive expansion of the original work. He can reasonably be considered as the most internationally respected specialist in the realm of sudden childhood death, and this new edition will consolidate his position further.

The area of medicine in which he has chosen to work must be one of the most difficult of all, not only for its scientific and technical problems but also because it contains the interface between the highly emotive issue of death in childhood and the extremely controversial issue of child abuse. Recent years have thrown up trial after trial where fiercely fought legal battles in both criminal and civil courts have raged over allegations of child abuse and child killing. In some of these, the standard of medical evidence has left a great deal to be desired, and ignorance and prejudice have in a few instances undoubtedly led to scandalous miscarriages of justice. Although this book addresses an enormous range of medical conditions, there is no doubt that a substantial part of its utilization will be to clarify the controversies and disputes that exist over the relationship between sudden infant death syndrome (SIDS) and alleged suffocation and other forms of deliberate harm to children. Indeed, Professor Byard has

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been involved so often in such issues that his original career as a mainstream pediatric pathologist has veered towards forensic pathology, and he is one of the few who have managed to successfully bestride both specialties.

The content of this second edition is even more ambitious and comprehensive than the first. The author has extended his age range down to the moment of birth, deaths in the first week of life formerly being excluded. Young adult deaths are also considered, as there is no true temporal cut-off between some childhood conditions and those of early adulthood.

Together with Stephen Cohle, another well-known expert in the field, the first part of the book deals with trauma. The tricky problem of the nomenclature and semantics of the various categories of injury is addressed at length.

A major part of the work is then devoted to natural disease, generally dealt with in the conventional way by organ systems, though there are caveats about keeping in mind the frequency with which multisystem disorders occur.

SIDS is then considered in great detail, with further warnings about distinguishing sudden death in infancy from SIDS, which too often are confused in the minds of many people. A marked feature of the

volume is the increase in both illustrations and references, the latter now virtually providing a total capture of all relevant papers on the subject matter of the book. At the end, there are a series of appendices relating to autopsy practice, with protocols and guidelines for a variety of situations, including SIDS, non-accidental injury, metabolic disorders, infective conditions, and possible poisoning.

A foreword should not be repetitive of either the table of contents or the author's preface, but should attempt to summarize the worth of the book to the medical and also, in this case, the legal communities. There can be no doubt that this second edition, even more than the previous one, is the current international benchmark in the subject – and it is likely to remain so against any potential competitors until the next edition.

Some textbooks, like Gray's *Anatomy* and Greenfield's *Neuropathology*, sit monolithic and enduring in the annals of medicine – and in the turbulent and controversial waters of childhood deaths, Byard's book remains a similar beacon for those who seek guidance.

Professor Bernard Knight, CBE
Cardiff, UK

Preface

The second edition of this text comes at a time when there has been considerable focus by the legal and medical professions and the public on inflicted and non-inflicted injuries in infants and young children. At the same time, there have been substantial developments in the field of pediatric natural diseases, with the discovery of genetic mutations associated with a wide array of disorders. As many of these diseases and injuries may be identified for the first time at autopsy, there is a great need for accuracy in post-mortem diagnosis, with appropriate tissue sampling and investigation.

In an attempt to deal with this new information, sections of the text have been defined more clearly, with, for example, separation of sudden infant death syndrome (SIDS) from other conditions that have been grouped under the headings of unintentional trauma, intentional trauma, and natural disease. Much has occurred in the SIDS field over the past decade, with identification of many risk factors resulting in a marked reduction in incidence. However, despite continued research, it still represents a “diagnosis” in search of causal diseases due to its lack of specific pathological features and its heterogeneous etiology. New developments in the areas of non-intentional injury and homicide have also necessitated substantial revision and expansion of this chapter. Several high-profile trials have drawn attention to the complexities of inflicted pediatric trauma and the problems that may arise in attempting to clearly establish causes of death. Thus, the intentional injury chapter has added, or expanded, sections on head trauma, the biomechanics of injury,

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neonaticide, subtle and unusual trauma, death by starvation, and irresponsible testimony by medical experts. Additional new sections also deal with murder-suicides and suicide.

Extensive referencing has been a deliberate feature of the text in an attempt to provide readers with access to substantial background information to allow them to find out more about individual conditions, many of which are seen rarely in everyday practice. This second edition has over 1200 new references and 300 additional figures.

Despite the somewhat tarnished reputation of the “case report,” observational studies remain extremely useful in providing examples of unusual features of a disorder and in illustrating pathophysiological principles. Thus, wherever possible, case descriptions of particular conditions and diseases have been given.

Debate has occurred in recent years concerning the use of terminology such as “accidental” compared with “non-intentional.” While there is no doubt that many injuries in children are both non-intentional and preventable, the term “accident” has not been dispensed with completely as it is one that is familiar to readers and is distinct from inflicted injury. The chapter on non-intentional injury has also been expanded to reflect more accurately the importance of non-inflicted trauma as a cause of death in childhood and adolescence, and to demonstrate again the significant role that pathology may play in injury prevention in the community.

Deaths in the first week of life have been included in this edition, and the age range of cases has also been extended into the early twenties in several of the rarer entities, as there is often no difference in the susceptibility to, and manifestations of, disease in this age range compared with adolescence.

On a more theoretical level, we are still no closer to understanding the pathophysiological basis of many

conditions, as it is now apparent that mechanisms and mutations causing disease may be as complex and varied as phenotype. Multiple mutations or diverse injuries may have similar manifestations, and the features that we use to diagnose a condition may be coincidental to lethal processes. Unfortunately, tissues and organs have only a relatively limited range of responses to a variety of environmental insults and mutations.

Where does that leave us? On a positive note, it would seem that we are on the threshold of discovering mechanisms for many childhood conditions that will lead us to improve diagnoses and to develop screening tests and treatment regimes. Given the rarity of many of these conditions, thorough post-mortems may be essential in identifying cases and enabling further studies to be undertaken.

Pediatric forensic pathology represents a developing discipline that is attempting to apply established forensic techniques to pediatric cases. The step from pediatric hospital to forensic mortuary is not, however, an easy one – conditions are rare and often quite complex, autopsy findings are subtle, and diagnoses may have considerable ramifications beyond the confines of pathology departments. Although the significance of certain findings remains uncertain, this will not stop the need for opinions to be formulated, substantiated, and defended in that most public of forums, the courtroom. Despite these problems, it is hoped that this text may provide some background information and guidance to assist pathologists, clinicians, and lawyers who find themselves dealing with the sometimes uneasy and changing interface of law, pathology, and pediatrics.

Roger W. Byard

Adelaide, Australia

2003

Preface to the first edition

Although there is a considerable body of literature on sudden adult death, there has been less interest in comprehensively classifying rare and diverse causes of sudden death in children. For example, publications dealing with sudden infant death tend to concentrate on sudden infant death syndrome (SIDS), and older children and adolescents are often included in the same series as adults. As well, some series have specifically excluded children who were hospitalized or under one year of age.

This book represents an attempt to redress the perceived deficit in the literature by gathering together in a single text the range of diseases, malformations, and conditions that can cause sudden, and often unexpected, death in both infants and children. Due to the unique nature of the neonatal period, perinatal deaths have not been included, and the age range of cases is generally between one week and 19 years of age.

It is our goal to provide a comprehensive, system-by-system review of a wide range of entities, including both “common” and more arcane disorders, along with illustrations of the conditions and back-up references for further reading. More attention has been paid to some of the rarer conditions that are unique to childhood than to some of the more common conditions found at all ages, as these are described well in general texts. For example, idiopathic arterial calcinosis has deliberately been dealt with in much greater depth than bacterial pneumonia, although the latter is far more common.

While the book has been divided into chapters based loosely on organ systems, a number of

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conditions may involve multiple systems. In these cases, the most detailed descriptions are to be found in the chapters that seem to deal best with the underlying pathological process that leads to sudden death.

As well as a general text, this book is also intended for use as an autopsy manual for pathologists confronted with an infant or child who has died unexpectedly, by providing protocols and checklists to serve as reminders of more obscure disorders that may otherwise be overlooked in a busy autopsy room.

Unfortunately, the number of disorders that may cause sudden death in childhood is quite vast, ranging from congenital abnormalities of early life to the acquired disorders of early adulthood. While we have attempted to cover as many conditions as

possible within the limitations of this short text, some will not have been included, due either to deliberate exclusion or to oversight on the authors' part. As well, due to the professional background and experience of the authors, the text tends to be ethnocentric, with a concentration on diseases and conditions that are found predominantly in Western countries rather than in other areas of the world. We apologize for any omissions. Given these imperfections, we hope that the text will be of practical and theoretical use to physicians and students who are engaged in the practice and study of pediatric medicine and pathology.

RWB, Adelaide, Australia

SDC, Grand Rapids, USA

March 1994

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I would like to acknowledge and thank pathologists and colleagues at the Forensic Science Centre and the Women's and Children's Hospital, Adelaide, specifically Dr Terry Donald, Dr Ross James, Dr John Gilbert, Dr Hilton Kobus, and Mr David Eitzen, for their continued and much appreciated support. Professor Peter Blumbergs, Dr Tony Bourne, Dr Terry Donald, Det. S/C Rick Fielder, Professor Yee Khong, Dr Jill Lipsett, Dr Lynette Moore, Dr Lloyd Morris, Dr Rebecca Scroop (Adelaide, Australia), Dr Joyce deJong (Grand Rapids, USA), and Professor Cenk Büyükkunal (Istanbul, Turkey) are also thanked for contributing photographs and artwork. The permission of Mr Wayne Chivell, South Australian State Coroner, to publish details of cases is also acknowledged gratefully, as is the help of his staff. Dr John Gilbert, Roland Hermanis and the South Australian Police (SAPOL) Physical Evidence and Photographic Sections are also thanked for photographic work.

The extensive update in references was only possible due to the excellent work of Ms Bet Witton, Librarian, Department of Administrative and Information Services, Adelaide. Finally, this text would not have been possible without the continued support and untiring editorial and indexing work of Ms Renée Amyot.

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