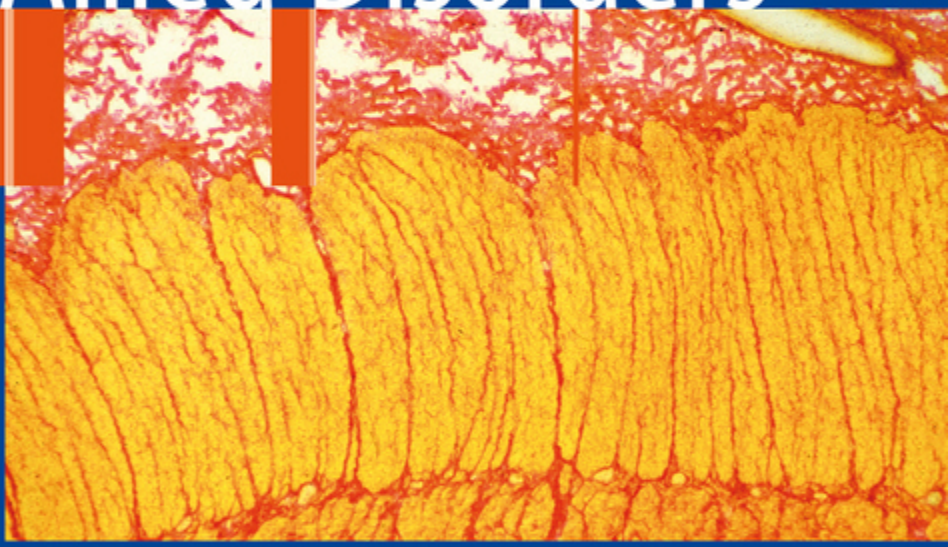
Alexander M. Holschneider
Prem Puri
Editors

Hirschsprung's Disease and Allied Disorders



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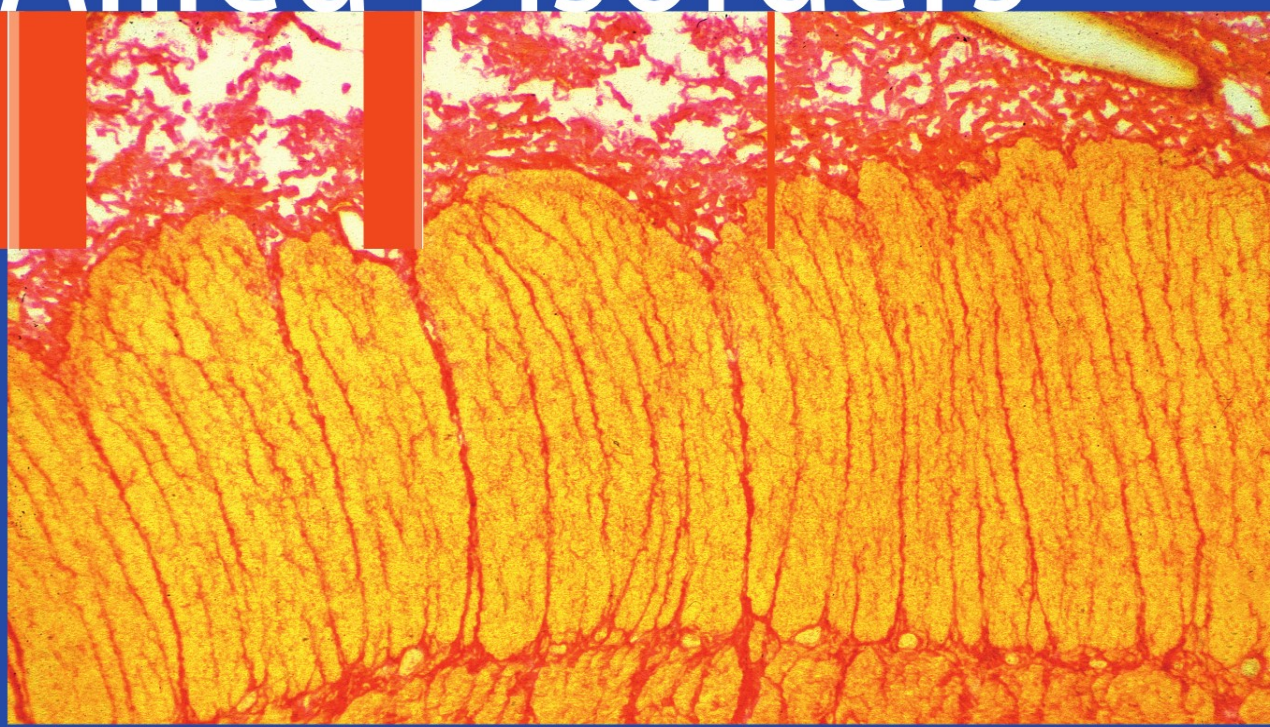

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A. M. Holschneider · P. Puri (Eds.)

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Hirschsprung´s Disease and Allied Disorders

Third Edition

With 318 Figures and 49 Tables

Springer

A.M. Holschneider, MD
Immenzaun 6a
51429 Bergisch Gladbach
Germany
and
Former Head of The Children's Hospital of Cologne
Amsterdamerstraße 59
50735 Cologne
Germany

P. Puri, MS, FRCS
Children's Research Centre
Our Lady's Hospital for Sick Children
Crumlin, Dublin 12
Republic of Ireland

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Foreword

Drs. Holschneider and Puri have again given me the honor of writing the foreword to this magnificent new edition of their book.

This book will continue to be recognized as the most comprehensive and well-documented text ever written on this subject. This new edition expands the horizons of our knowledge of difficult and challenging conditions such as Hirschsprung's disease.

Dr. Grosfeld, a prestigious professor of pediatric surgery, was invited to write on the historical perspective of Hirschsprung's disease, and he has done so with a characteristically masterful style.

The chapter on the pathophysiology of Hirschsprung's disease is now written by Dr. Puri and Dr. Montedonico.

Dr. Moore has written a very interesting chapter on congenital anomalies and genetic associations in Hirschsprung's disease. The chapter on radiological diagnosis is now written by Dr. Kelleher.

This edition of the book characteristically continues to expand upon the genetic basis of the condition. Dr. Puri has been working in this particular area in the laboratory for many years, and we all grateful for his efforts and his contribution.

The chapter on immunohistochemical studies written by Dr. Rolle and and Dr. Puri summarizes the very exciting advances in this type of diagnosis.

An additional chapter by Dr. Milla on adynamic bowel syndrome expands our knowledge on the spectrum of motility disorders of the bowel and urinary tract.

Finally, Dr. Somme and Dr. Langer have written an additional chapter on the transanal pull-through procedure for the treatment of Hirschsprung's disease. There is no question that this new therapeutic approach represents a very important contribution to the treatment of this condition.

Again, we applaud the efforts of the editors in selecting a group of talented experts and innovators to contribute to what is still the best book on the subject.

Alberto Peña, MD

Preface

Hirschsprung's disease is one of the most important and most fascinating diseases in paediatric surgery. Our understanding of Hirschsprung's disease is developing rapidly, not only in relation to its pathophysiology and the development of new surgical techniques, but especially in relation to new genetic findings. A first comprehensive description of the pathophysiology, clinical symptoms, diagnosis and therapy of Hirschsprung's disease was outlined in 1970 by Theodor Ehrenpreis, Professor of Pediatric Surgery at the Karolinska Institute, Stockholm, Sweden, in a booklet entitled "Hirschsprung's Disease". The booklet of 176 pages was dedicated to Harald Hirschsprung (1830–1916) of Copenhagen, Denmark, and to Ovar Swenson of Chicago, Illinois, USA, the two pioneers in the study of Hirschsprung's disease. Harald Hirschsprung was a paediatrician, and Ovar Swenson a paediatric surgeon, who performed the first successful resection of an aganglionic bowel segment. That first book, published by Yearbook Medical Publishers, mainly discussed questions of postoperative continence based on the results of a large series of patients treated successfully at the Karolinska Institute.

In 1978 Ehrenpreis permitted one of the editors of the present edition to prepare an update of his internationally recognized book. Therefore, in 1982, a new book on Hirschsprung's disease by Alexander Holschneider was published by Hippokrates (Thieme-Stratton) with a foreword by Th. Ehrenpreis. It was a multiauthored textbook with particular prominence given to the results of an international clinical research study of the postoperative results in Hirschsprung's disease, undertaken from 1976 to 1978 by the author himself and a technical assistant, with special regard to the underlying surgical techniques. The follow-up studies were performed with the help of the Volkswagen Foundation in 16 paediatric surgical departments in Europe and the United States over a period of 3 years. The most interesting and unique aspect of this study was the fact that all clinical and electromanometrical investigations were performed by the same research team, independent of the staff of the individual hospital. As a result of this study concept, a most objective comparison of the results of Swenson's, Soave's, Duhamel's and Rehbein's techniques was achieved.

However, as our understanding of Hirschsprung's disease and associated motility disorders of the gut increased, a second edition of this book was published in 2000, this time by Harwood Academic Publishers, part of the Gordon and Breach Publishing Group. The title of this new book was changed to "Hirschsprung's Disease and Allied Disorders", because we included other enteric plexus disorders and smooth muscle disorders of the gut. The editors of this again multiauthored edition were Alexander Holschneider and Prem Puri. The book was divided into three parts: Physiology and Pathophysiology, Clinical Aspects, and Treatment and Results. As well as discussion of normal colonic motor function and the pathophysiology of classical Hirschsprung's disease, the book included special chapters on the development of the enteric nervous system, the functional anatomy of the enteric nervous system, animal models of aganglionosis, the molecular genetics of Hirschsprung's disease and the RET protein in human fetal development and in Hirschsprung's disease. New areas of special interest included intestinal neuronal dysplasia, particular forms of intestinal neuronal malformations, enterocolitis, megacystis-microcolon-intestinal hypoperistalsis syndrome, degenerative hollow visceral myopathy mimicking Hirschsprung's disease, and newer diagnostic techniques such as special neuronal markers, electron microscopy and anal sphincter achalasia. This second edition was the most comprehensive book ever published on Hirschsprung's disease and allied disorders.

With the passage of time, our understanding of enteric plexus disorders has exploded. Ehrenpreis in his preface of 1970 cited the President of the Swedish Nobel Prize Committee who stated that there are more scientists living today than during all past centuries. After having reviewed the recent literature on Hirschsprung's disease and allied disorders we are convinced that this is even more relevant today. Therefore, a new edition of Hirschsprung's disease and allied disorders was realized with the help of Springer. The previous chapters

"Clinical Generalities of Hirschsprung's Disease", "Disorders and Congenital Malformations associated with Hirschsprung's Disease", "Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome", "Degenerative Hollow Visceral Myopathy Mimicking Hirschsprung's Disease" and "Diagnosis of Hirschsprung's Disease and Allied Disorders" have been updated. A new separate chapter on "NAPDH-Diaphorase Histochemistry" has been introduced in the part "Diagnosis", next to the updated chapters "Histopathological Diagnosis and Differential Diagnosis of Hirschsprung's Disease", "Immunohistochemical Studies" and "Electron Microscopic Studies of Hirschsprung's Disease". For reasons of clarity, previously separated chapters such as the former chapters 5 and 6 "Molecular Genetics of Hirschsprung's Disease" and "Ret-Protein in Human Foetal Development and in Hirschsprung's Disease" have been brought together and concentrated in a new chapter. Chapter 3 "Functional Anatomy of the Enteric Nervous System" by M.D. Gershon and chapter 6 "Normal Colonic Motor Function and Relevant Structure" by J. Christensen have been reproduced. Chapter 12 "Particular Forms of Intestinal Neuronal Malformations" and chapter 14 "Megacolon in Adults" have become part of the new chapter 8 "Hirschsprung's Disease: Clinical Features" and chapter 18 "Neurocristopathies and Particular Associations with Hirschsprung's Disease". Chapter 17 "Intestinal Obstructions Mimicking Hirschsprung's Disease" has become chapter 21 "Adynamic Bowel Syndrome".

The chapters referring to the different surgical techniques have been updated too, but the concept of the previous editions, to compare the detailed description of one of the pioneer surgeons with the experience of a second author with the same technique, was given up. In the third edition of the book both parts of each chapter dealing with a specific surgical technique have been brought together to create new contributions for each of the different surgical approaches. The chapter "Laparoscopically Assisted Anorectal Pull-through" has been updated and a new chapter "Transanal Pull-through for Hirschsprung's Disease" has been introduced. Finally, the previous chapters dealing with early and late complications have also been brought together and the contribution of Teitelbaum and Coran on long-term results and quality of life has been updated.

The new edition is again a multiauthored book, and we have to thank all the internationally well-known authors and coauthors for their excellent and sophisticated contributions. It is their interest, help and effort that has again made possible the drawing together in one volume of the collective wisdom of many of the leading experts in Hirschsprung's disease and related disorders. Their contributions to this volume again provide a step forward in the elucidation of the genetic basis, and the correct diagnosis and treatment of this interesting disease and its allied disorders.

Besides the authors and coauthors, we would like to thank Mrs. Elisabeth Herschel of the Children's Hospital of Cologne, and the Children's Medical and Research Foundation, Our Lady's Children's Hospital, Dublin, for their support. Finally, we wish to thank the editorial staff of Springer, Heidelberg, Germany, particularly Ms. Gabriele Schroeder, for their interest and encouragement to publish a third edition of this book on a most important subject in paediatric surgery.

Alexander M. Holschneider
Prem Puri

Contents

List of Contributors

A.M. Alzahem
The Children's Hospital at Westmead
Westmead, Sydney
Australia

J. Amiel
Département de Génétique
Unité INSERM U-393
et Université Paris 5
Hôpital Necker-Enfants Malades, Paris
75724 Paris, Cedex 15
France

R.A. Brown
Department of Paediatric Surgery
Red Cross Children's Hospital, Klipfontein Rd.
Rondebosch
7700 Cape Town
South Africa

E. Bruder
Department of Pathology
University of Basel
Schönbeinstrasse 40
4003 Basel
Switzerland

D.T. Cass
Department of Surgical Research
The New Children's Hospital
Royal Alexandra Hospital for Children
Sydney
Australia

I. Ceccherini
Laboratorio di Genetica Molecolare
Istituto Giannina Gaslini
16148 Genova
Italy

J. Christensen
The University of Iowa
College of Medicine
Department of Internal Medicine
Iowa City, Iowa 52242
USA

A.G. Coran
University of Michigan Medical Center Head
Section of Pediatric Surgery
F3970 Mott Children's Hospital
Ann Arbor, MI 48109-0245
USA

K.E. Georgeson
Division of Pediatric Surgery
University of Alabama at Birmingham
Birmingham, AL 35294
USA

M.D. Gershon
Department of Anatomy and Cell Biology
Columbia University
College of Physicians and Surgeons
630 W 168th Street
New York, N.Y. 10032
USA

P. Griseri
Laboratorio di Genetica Molecolare
Istituto Giannina Gaslini
16148 Genova
Italy

J.L. Grosfeld
Department of Pediatric Surgery
Riley Children's Hospital
702 Barnhill Drive – Suite 2500
Indianapolis, IN 46202
USA

A.M. Holschneider
Immenzaun 6a
51429 Bergisch Gladbach
Germany
and
The Children's Hospital of Cologne
Amsterdamerstr. 59
50735 Cologne
Germany

L.H. Homrighausen
Department of Surgery
The Children's Hospital of Cologne
Amsterdamerstr. 59
50735 Cologne
Germany

V. Jasonni
Department of Pediatric Surgery
Giannina Gaslini Institute
University of Genoa
16148 Genova
Italy

J. Kelleher
Department of Radiology
Our Lady's Children's Hospital
Crumlin, Dublin 12
Republic of Ireland

H.-J. Krammer
University Hospital of Heidelberg at Mannheim
Theodor-Kutzer-Ufer 1
68135 Mannheim
Germany

M. Kunst
Department of Surgery
The Children's Hospital of Cologne
Amsterdamerstr. 59
50735 Cologne
Germany

J.C. Langer
Department of Pediatric General Surgery
Hospital for Sick Children, Toronto
Toronto, ON M5G 1X8
Canada

F. Lantieri
Laboratorio di Genetica Molecolare
Istituto Giannina Gaslini
16148 Genova
Italy
and
Dipartimento di Scienze della Salute
Sezione di Biostatistica, Università di Genova
16148 Genova
Italy

D.C. Little
Department of Surgery
Children's Mercy Hospital
Kansas City, MO 64108
USA

S. Lyonnet
Département de Génétique, Unité INSERM U-393
et Université Paris 5
Hôpital Necker-Enfants Malades
75724 Paris, Cedex 15
France

G. Martucciello
Department of Pediatric Surgery
Scientific Institut (IRCCS)
Policlinico 'San Matteo'
27100 Pavia
Italy

W. Meier-Ruge
Department of Pathology
University of Basel
Schönbeinstrasse 40
4003 Basel
Switzerland

M. Menezes
Children's Research Centre
Our Lady's Children's Hospital
Crumlin, Dublin 12
Republic of Ireland

M.L. Metzelder
Department of Pediatric Surgery
Hannover Medical School
30625 Hannover
Germany

P.J. Milla
Gastroenterology Unit
Institute of Child Health
University College London
London, WC1E 6BZ
UK

S. Montedonico
Children's Research Centre
Our Lady's Children's Hospital
Crumlin, Dublin 12
Republic of Ireland