Behrooz A. Akbarnia
Muharrem Yazici
George H. Thompson
Editors

The Growing Spine

Management of Spinal Disorders in Young Children

Second Edition

Springer

The Growing Spine

Behrooz A. Akbarnia • Muharrem Yazici
George H. Thompson
Editors

The Growing Spine

Management of Spinal Disorders in Young Children

Second Edition

Springer

Editors
Behrooz A. Akbarnia
San Diego Center for Spinal Disorders
Medical Group, Inc.
La Jolla, CA
USA

Muharrem Yazici
Hacettepe University Department
of Orthopedics and Traumatology
Sihhiye
Ankara
Turkey

George H. Thompson
Rainbow Babies and Children's Hospital
Cleveland, OH
USA

Editorial Assistant
Pat Kostial, RN, BSN
San Diego Spine Foundation
San Diego, CA, USA

ISBN 978-3-662-48283-4 ISBN 978-3-662-48284-1 (eBook)
DOI 10.1007/978-3-662-48284-1

Library of Congress Control Number: 2015954105

Springer Heidelberg New York Dordrecht London

Printed on acid-free paper

Springer-Verlag GmbH Berlin Heidelberg is part of Springer Science+Business Media (www.springer.com)

As with our first edition, the driving force behind the second edition of The Growing Spine textbook is our **patients and their families**. This book would mean nothing without their need for our care and expertise. We therefore dedicate it to our patients for their trust and participation and for giving us the opportunity to learn from experience and to share the knowledge we have gained with others. It is our sincere hope that this book will continue to inspire those who care for young children with early-onset scoliosis and thoracic deformities, leading to better outcomes and improved quality of life.

Much has changed over the past 5 years, and we believe this second edition textbook reflects our better understanding of the issues and advances in the field. We owe a great deal of gratitude to many who helped us reach this level of understanding, including our specialty societies: the Scoliosis Research Society (SRS), the Pediatric Orthopaedic Society of North America (POSNA), the European Pediatric Orthopaedic Society (EPOS), and the American Academy of Orthopaedic Surgeons (AAOS). These organizations endorsed the International Congress on Early Onset Scoliosis (ICEOS) meeting, supporting both the educational effort during their meetings and the formation of a task force for pediatric devices where their collaboration with regulatory agencies and the industry has enabled an environment for innovation and change. The ability to bring diverse groups together to achieve the common goal of improving our patients' quality of life has led to an enriching experience, one which we have attempted to share on the pages of this book.

Many thanks to our colleagues in North America and around the world at both the Growing Spine Study Group (GSSG) and the Children Spine Study Group (CSSG) (including investigators and study coordinators), as well as to the faculty and participants at ICEOS. These important groups supported the creation of a forum of exchange addressing issues surrounding very young children with spine and thoracic deformities and thereby advancing the research and knowledge in this field. Thanks also to the related specialties showing increased interest and participation in teaching at

ICEOS, including pulmonology, cardiology, anesthesiology, radiology, and genetics. The list is too long to mention all specialties, but suffice it to say that the help, expertise, and guidance of our specialist colleagues have been invaluable. We particularly wish to thank the Growing Spine Foundation (GSF) board and staff, who have supported our work with assistance, financial support, and guidance over the years.

This book would not be in your hands without our dedicated, world-expert authors. We are grateful to them for their contributions and for their invaluable efforts to prepare excellent chapters and to respond to multiple queries and in a timely manner.

Last, but not least, we are grateful to our associates and staff for putting up with us throughout the preparation of this book; we give them all special thanks and are deeply indebted to them for their support.

And now, most importantly, a special note from each of us to our respective families:

Behrooz Akbarnia, MD*:* My deepest gratitude, as always, to my wife Nasrin, for her continuing support, love, and encouragement throughout the completion of the second edition. Thanks also to my three children and their spouses and, last but by no means least, to my four wonderful grandchildren—Simia, Kian, Leila, and Luca—the last two born after the publication of the first edition. They were always a source of inspiration and energy for me to continue the work on this book in spite of the time it took away from them.

Muharrem Yazici, MD*:* The greatest of thanks should go to my family Ruya, Yildiz Naz, and Mehmed Emir. I have stolen time from them, and although I have, on more than one occasion, chosen academia over personal life, they have never complained and always provided their love and support for me.

George H. Thompson, MD*:* As with the first edition, my thanks and appreciation to my wife, Janice, for her support and encouragement in completing the second edition. She and our children and six grandchildren were deprived of my attention during the editing of this edition. They never complained as they understood the importance of completing this project due to the improved care for others that it will provide.

Foreword 1

The growth of the spine demonstrates symmetry in all dimensions to reach a harmonious balance not only for volume and mass (cosmetics) but also for function (motion) during growth as well as when growth is completed. This is especially important for the development of the thoracic cage and subsequent pulmonary function.

Spinal pathology from any etiology can affect growth and development in different ways. For example, if the disorder affects a very limited area, such as a localized congenital malformation (i.e., unbalanced hemivertebra or unsegmented bar), a limited operative procedure (i.e., hemivertebra excision) may have significantly different results. Localized congenital deformities may result in a significant deformity with growth, while a limited procedure may correct a deformity and allow adequate development of the spine. In other situations, the etiology may affect a larger part, if not the entire spine such as in neuromuscular, syndromic, or idiopathic deformities (infantile idiopathic scoliosis). For such situations, it must be remembered that any surgical procedure on a still growing spine will create a disturbance in the growth. Sometimes the spine may not be well balanced in three dimensions and may not be reversible following an extensive early fusion.

The other point is that scoliosis is mainly a horizontal (coronal) plane deformity where the most important mechanism is spinal torsion or rotation. Until now, surgically speaking, we have developed devices able to correct the collapsing spine with elongation or distraction rods or by growth modulation devices with a compression mechanism used anteriorly on the apical vertebral bodies on the convex side. But as yet we do not have true derotation devices.

This is why, until now, the best three-dimensional correction for a spinal deformity in a growing child, especially in the thoracic area without congenital malformations, is by repeated or serial casts and bracing (more or less mixed over time). These are used until appropriate definitive posterior spinal fusion and segmental spinal instrumentation can be performed close to the end of growth.

Despite that, some situations require surgical treatment during growth, particularly early onset or at a young age, and this book will give some proposals and even some solutions for such cases. But the main goal of this book is to advise the reader to evaluate carefully and have a clear understanding of the pathophysiology, natural history, and treatment options before treating an "early-onset scoliosis" case. We need to think about the consequences of

immediate and delayed treatment, either nonoperative or mostly operative, with its subsequent and sometimes irreversible dangers or complications. Remember that operating on a growing spine can result in repeated surgeries with exponential complication risks. Perform surgery only when truly necessary.

Finally, this book will open your mind regarding the need for further research, which is always necessary to help the children who are depending on our care.

Paris
March 2015

Jean Dubousset

Foreword 2

This is the second edition of *The Growing Spine*, being published 4 years after the first edition in 2011. With the publication two questions are posed – why so soon, and what is different?

The field of EOS has expanded greatly with the Study Groups and the ICEOS meetings, as well as the knowledge disseminated with the first edition of *The Growing Spine*, all of which helped establish the growing spine as a recognized subspecialty in the field of spinal surgery. More physicians and clinics are concentrating on the care of these challenging problems. In addition, the SRS Growing Spine Committee Early Onset Scoliosis Consensus statement published earlier this year covers the problem well (1). The definition of EOS, "spine deformities that is present before the age of 10 years of age," is stated, and in addition the organization of EOS into diagnostic categories, evaluation, treatment goals, and treatment options is laid out. With the great expansion in knowledge and interest in the field, the editors viewed the explosion of knowledge in the field as being so rapid that the first edition is not current.

The second edition has been expanded to 57 chapters with international authors comprising 30 % of the total authorship. The chapters have been revised and updated with the addition of new chapters. The organization of the chapters into different sections is very helpful and follows the outline of the consensus statement above. There are certain chapters that still represent the definitive work of definitive experts in that particular area:

Chapter 4 on "Normal Growth of the Spine and Thoracic Cage" by Dr. Alain Dimeglio

Chapter 5 on "Normal Lung Growth and Thoracic Insufficiency Syndrome" by Dr. Gregory Redding

Chapter 16 on "Neurofibromatosis" by Dr. Alvin Crawford

Chapters 17 and 18 on "Spine Deformities in Syndromes" by Dr. Paul Sponseller

Chapter 29 on "Casting for Early-Onset Scoliosis" by Dr. James Sanders

Chapter 38 on "Traditional Growth Rods" by Drs. George Thompson and Behrooz Akbarnia

Chapter 39 on "VEPTR Expansion Thoracoplasty" by Dr. Robert Campbell

The new chapters add to the knowledge of the growing spine and are an impressive addition to the text. They expand on areas partially addressed in

the first edition, as well as add additional knowledge. Notable among these are:

Chapter 7 on "New classification System" by Dr. Michael Vitale

Chapter 15 on "Intraspinal Pathology" by Drs. Nejat Akalin and Amer Samdani

Chapter 50 on "Complications Following Distraction-Based Growing Technique" by Dr. John Emans

Chapter 52 on "Anesthetic Considerations in Growing Children and Repetitive Anesthesia" by Dr. Lena Sun

The surgical management is well covered, with chapters on "traditional surgery" as these children do not always present at a time when growth-friendly surgery can be performed. The coverage of the techniques for growth-friendly surgery is extensive with many choices presented, both proven and innovative.

The most important consideration is the outcome of the treatment as pointed out by Dr. Vitale in Chapter 55 assessing the radiographic, pulmonary, and HRQOL results. It is important to remember that the critical time is at the end of growth and later when the children become adults.

It is an honor and pleasure to be asked to write the foreword to the second edition of *The Growing Spine*, and you have in your hands the latest comprehensive knowledge in this growing unique field.

1. Skaggs DL, Guillaume T, El-Hawary R, et al (2015) Early onset scoliosis consensus statement, SRS growing spine committee. 2015. Spine Deformity 3:107.

Minneapolis, MN, USA
St. Paul, MN, USA

John E. Lonstein, MD

Preface to the Second Edition

The field of spinal deformities in young children is one of the newer areas in spinal surgery literature. For a long time, early-onset spine deformities have had to endure neglect by researchers and only recently achieved priority in the scopes of spinal deformity surgeons. However, despite this late start, early-onset deformity literature has accelerated and gained popularity at a greater rate, and this early neglect has been largely negated in the appearance of an explosive number of epidemiological, clinical, and experimental studies in the past 5–10 years.

Undoubtedly, the collaborative efforts of physicians joined together under the auspices of the Growing Spine Study Group and Children's Spine Study Group and International Congress on Early Onset Scoliosis (ICEOS) meetings held annually since 2007 have had stimulating effects on this newly kindled research effort. However, we should not be remiss in mentioning the 2010 first edition of this book, which has contributed greatly to the recognition of the growing spine as an independent subspecialty in the practice of spinal surgery.

Our book, the first edition of which was eagerly perused by the spinal surgery reader, was sold out quickly and among thousands of Springer-published textbooks and soon took its place on the top rungs of the most-downloaded list of eBooks. It was even later translated into Chinese and is being translated to other languages. Both due to this quick depletion of printed books and the rapid evolution of information in this field, preparations for the second edition began only 3 years after its initial publication, a time period that can be considered quite short for scientific medical textbooks. The edition you hold in your hands today boasts extensive reviews of all of its chapters and includes all-new developments that have found their place in the EOS literature recently in the form of updates and in all-new chapters. The editorial process was performed even more diligently, with close attention to the congruity of sections written by authors across the globe to achieve an integrated text. As distinct as this book is by presenting the classical and emerging ideas on the growing spine, it is also unique in that it boasts exceptional international contributions.

The dissemination of scientific knowledge is in constant flux. As soon as a research project is turned into written word, it is doomed to become outdated. Given time, doubtless the information that has found its way onto the pages of this book will suffer that very same fate. However, we, the editors, are confident in our belief that our book will provide current and satisfying

answers to basic questions posed by clinicians and academicians interested in the field.

Although all three editors have reviewed all chapters meticulously, mistakes overlooked are inevitable in a text of this scope. All accolades for the book belong with the authors. We are grateful to them for their invaluable and diligent contributions. All errors and omissions, however, are the responsibility of the editors. For these, we beg your forgiveness and understanding. Please do not hesitate to share with us any and all errors and omissions that you note in this book, thereby allowing us to correct them in future editions.

Finally, without the hard work of our dear friend Pat Kostial, RN, BSN, the publication of this book would not have been possible. There are no words to convey our gratitude to Ms. Kostial for her indispensable help. With Michael D. Sova's transparent, collaborative work style and diligence, who worked with us for this edition, obstacles disappeared and impossibilities only took a little time. We consider ourselves very lucky for having had the opportunity to work with such a team. It would also not have been possible to print these sentences without the professional approach of Springer, which is doubtless among the world leaders in the publication of scientific medical literature.

La Jolla, CA, USA — Behrooz A. Akbarnia, MD
Ankara, Turkey — Muharrem Yazici, MD
Cleveland, OH, USA — George H. Thompson, MD

Preface to the First Edition

Early onset scoliosis (EOS) is a major topic in pediatric spine deformity today. These challenging deformities occur in almost all differential diagnostic categories.

Unfortunately, each diagnosis has a different natural history, making it even more demanding. This is the first textbook on this topic. It is a compilation of the current concepts of evaluation and treatment of the various deformities of the growing spine.

We have tried to explore the normal growth of the spine and other associated organs as well as natural history of the various differential diagnostic categories and possible treatment options. It is anticipated that this textbook will need to be updated every 2–3 years in the future as concepts and treatment guidelines change. Treating the spinal deformity is not the major issue, but controlling the deformity to allow for growth of the spine and the associated organ systems, such as pulmonary, cardiac, and gastrointestinal, is the major goal. Controlling deformity allows for improved spinal growth of the involved child and the controlling associated development of these organ systems. A short trunk has an adverse effect on these organ systems. As a consequence, EOS requires a multidisciplinary care. It involves genetics, pediatrics, pulmonology, cardiology, neurology, neurosurgery, as well as orthopaedic surgery.

Treatment options for very young children are controversial. Bracing, serial Risser casts, and surgery (growth modulation and the use of distraction-based or growth-guided techniques such as growing rods) are explored in this textbook.

Preliminary treatment results have demonstrated that growth-friendly surgical techniques are effective in controlling or modulating curve progression and allowing for spinal growth. Spinal growth allows for improved capacity of the thoracic and abdominal cavities. Cosmesis is less than ideal as crankshaft remains a significant problem even in the growing rod systems. Surgical treatment complications are high, particularly infection and implant failure, especially rod breakage. Management of complications is an important aspect of the treatment of EOS. Because of the high complication rate, it is important to make the right decision regarding patient and family selection. They must be cooperative and understanding and be willing to be cooperative during the postoperative period.

Future research is important. The Growing Spine Study Group (GSSG) and other databases will hopefully guide future investigations. Only by

defining the results of treatment in a relatively large volume of children over a long period of time can the true effectiveness of each of these techniques be determined. Predicting who will worsen, improving spinal tethers to control progressive deformities and the development of self-expanding or remotely controlled devices that would obviate the need for repeated surgical procedures.

We thank our contributors who are all specialists and experts in a variety of areas involved with early onset scoliosis. We also acknowledge the contribution of the members of Growing Spine Study Group who have continuously provided the information that is the basis for a significant portion of the data presented in this book.

Special thanks for assistance in preparing and organizing this textbook are to Sarah Canale and Pooia Salari; without their assistance, the completion of this project would have been very difficult.

La Jolla, CA, USA — Behrooz A. Akbarnia, MD
Ankara, Turkey — Muharrem Yazici, MD
Cleveland, OH, USA — George H. Thompson, MD

Contents

Contributors

R. Emre Acaroglu, MD Orthopedic Spine Department, Ankara Spine Center, Ankara, Turkey

Nejat Akalan, MD, PhD Department of Neurosurgery, Hacettepe University, Ankara, Turkey

Behrooz A. Akbarnia, MD Department of Orthopaedic Surgery, Growing Spine Foundation, University of California, San Diego, San Diego, CA, USA

Ahmet Alanay, MD, PhD Department of Orthopedics and Traumatology, Acibadem University School of Medicine, Acibadem Comprehensive Spine Center, Istanbul, Turkey

Lindsay Andras, MD Department of Orthopaedic Surgery, Children's Hospital Los Angeles, Los Angeles, CA, USA

Navid Arandi, BS San Diego Spine Foundation, San Diego, CA, USA

Keith Baldwin, MD Division of Orthopaedic Surgery, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Shay Bess, MD Department of Orthopaedic Surgery, Rocky Mountain Hospital for Children, Presbyterian/St. Luke's Medical Center, Denver, CO, USA

Randal R. Betz, MD Institute for Spine and Scoliosis, Lawrenceville, NJ, USA

Laurel C. Blakemore, MD Department of Orthopedics and Rehabilitation, University of Florida School of Medicine, Gainesville, FL, USA

Oheneba Boachie-Adjei, MD, DSc Department of Orthopedic Surgery, Hospital for Special Surgery, New York, NY, USA

G. Bollini, PhD Department of Pediatric Orthopedic Service Pr Jouve, Timone Children Hospital, Marseilles, Bouches du Rhône, France

François Bonnel, PhD, MD Faculty of Medicine, University of Montpellier, Montpellier, France

Stefano Boriani, MD Department of Oncologic and Degenerative Spine Surgery, Rizzoli Institute, Bologna, Italy

Jarred Bressner, BS School of Medicine, Johns Hopkins University, Baltimore, MD, USA

Donita I. Bylski-Austrow, PhD Department of Orthopaedics, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Patrick J. Cahill, MD Section of Orthopaedic Surgery, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Robert M. Campbell, MD Division of Orthopaedic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

Federico Canavese, MD, PhD Department of Pediatric Surgery, University Hospital Estaing, Clermont Ferrand, France

Kenneth M.C. Cheung, MD, FRCS, FHKCOS, FHKAM(ORTH) Department of Orthopaedics & Traumatology, Queen Mary Hospital, Hong Kong, China

Alvin H. Crawford, MD, FACS Department of Orthopaedics, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Matthew E. Cunningham, MD, PhD Scoliosis Service, Department of Orthopaedic Surgery, Hospital for Special Surgery, New York, NY, USA

Phyllis D'Ambra, RN, BS, MPA Department of Orthopedics and Neurosurgery, Children's Hospital of Los Angeles, Burbank, CA, USA

Ozgur Dede, MD Department of Orthopaedic Surgery, Children's Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Halil Gokhan Demirkiran, PhD Department of Orthopedic and Traumatology, Hacettepe University Hospitals, Ankara, Turkey

Vincent J. Devlin, MD Center for Devices and Radiologic Health, Office of Device Evaluation, Division of Orthopedic Devices, Posterior Spine Devices Branch, Food and Drug Administration, Silver Spring, MD, USA

Alain Dimeglio, PhD, MD Faculty of Medicine, University of Montpellier, Montpellier, France

P.L. Docquier, MD, PhD Department of Orthopaedic Surgery, Saint-Luc University Hospital, Université Catholique de Louvain, Brussels, Belgium

John P. Dormans, MD, FACS Division of Orthopaedic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

Mingkui Du, MD Department of Orthopaedics, General Hospital of Chinese Armed Police Forces, Beijing, China

Ron El-Hawary, MD, MSc, FRCS (c) Department of Orthopaedics, IWK Health Centre, Halifax, NS, Canada

Hazem B. Elsebaie, MD, FRCS Department of Orthopedics, Cairo University Hospital, Giza, Egypt

John B. Emans, MD Department of Orthopedic Surgery, Children's Hospital Boston, Harvard Medical School, Boston, MA, USA

Christine L. Farnsworth, MS Division of Orthopedics, Rady Children's Hospital, San Diego, San Diego, CA, USA

Tamás Fülöp Fekete, MD Spine Center, Schulthess Clinic, Zürich, Switzerland

Catherine E. Ferland, PhD Department of Orthopedics, Shriners Hospital for Children, Canada, Montreal, QC, Canada

Ivan Florentino-Pineda, MD, MBA Department of Anesthesiology and Perioperative Medicine, Children's Hospital of Georgia, Georgia Regents University, Augusta, GA, USA

Alier J. Franco, PhD Department of Intraoperative Neuromonitoring, Specialty Care, Lester, PA, USA

Luke Gauthier, MD, FRCS (c) Division of Orthopaedic Surgery, IWK Health Centre, Halifax, NS, Canada

Riccardo Ghermandi, MD Department of Oncologic and Degenerative Spine Surgery, Rizzoli Institute, Bologna, Italy

Michael P. Glotzbecker, MD Department of Orthopaedic Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Emmanouil Grigoriou, MD Division of Orthopaedic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

Manon Haché, PhD Department of Anesthesiology, Columbia University Medical Center, New York, NY, USA

Division of Pediatric Anesthesia, Columbia University Medical Center, New York, NY, USA

Daniel Haschtmann, MD Spine Center, Schulthess Clinic, Zürich, Switzerland

Daniel W.H. Ho, PhD Department of Pathology, University of Hong Kong, Hong Kong, China

M. Timothy Hresko, MD Department of Orthopaedic Surgery, Harvard Medical School, Boston Children's Hospital, Boston, MA, USA

Steven Hwang, MD Department of Neurosurgery, Tufts Medical Center, Floating Hospital for Children, Boston, MA, USA

Justin Iorio, MD Department of Orthopedic Surgery and Sports Medicine, Temple University Hospital, Philadelphia, PA, USA

Viral V. Jain, MD Department of Orthopaedics, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Ehsan Jazini, MD Department of Orthopaedics, University of Maryland Medical Center, Baltimore, MD, USA

Dezső Jeszenszky, MD, PhD Spine Center, Schulthess Clinic, Zürich, Switzerland

Charles E. Johnston, MD Department of Orthopedics, Texas Scottish Rite Hospital for Children, Dallas, TX, USA

Jean-Luc Jouve, MD, PhD Department of Pediatric Orthopedics, Timone Children Hospital, Marseille, France

Rishi Mugesh Kanna, MS, MRCS, FNB Department of Orthopaedics, Traumatology and Spine Surgery, Ganga Hospital, Coimbatore, India

Lawrence I. Karlin, MD Department of Orthopaedic Surgery, Boston Children's Hospital, Boston, MA, USA

Noriaki Kawakami, MD, DMSc Department of Orthopedic Surgery, Nagoya University, School of Medicine, Sannomaru, Naka Ward, Nagoya, Japan

Department of Orthopedics and Spine Surgery, Meijo Hospital, Sannomaru, Naka Ward, Nagoya, Japan

Pat Kostial, BSN, RN San Diego Spine Foundation, San Diego, CA, USA

Zhengda Kuang, MD Department of Orthopaedics, General Hospital of Chinese Armed Police Forces, Beijing, China

Leok-Lim Lau, MBChB, FRCS (Orth) National University Health System, University Orthopaedics, Hand and Reconstructive Microsurgery Cluster, Singapore, Singapore

Lawrence G. Lenke, MD Department of Orthopaedic Surgery, Washington University School of Medicine, St. Louis, MO, USA

Jin Lin, MD Department of Otrhopaedics, Peking Uion Medical College Hospital, Beijing, China

Breton Line, BSME Departmentof Orthopaedic Surgery, Rocky Mountain Hospital for Children, Presbyterian/St. Luke's Medical Center, Denver, CO, USA

Scott J. Luhmann, MD Department of Orthopaedic Surgery, Washington University School of Medicine, St. Louis, MO, USA

Marios G. Lykissas, MD, PhD Department of Orthopaedics, University of Ioannina, School of Medicine, Ioannina, Greece

William Mackenzie, MD Department of Orthopaedics, Nemours/Alfred I. DuPont Hospital for Children, Wilmington, DE, USA

Luke Macyszyn, MD, MA Department of Neurosurgery, University of Pennsylvania, Philadelphia, PA, USA

Richard E. McCarthy, MD Department of Orthopaedics and Neurosurgery, Arkansas Children's Hospital, University of Arkansas for Medical Sciences, Little Rock, AR, USA

Hossein Mehdian, MD, FRCS Spinal Unit, The Centre for Spinal Studies and Surgery, Queen's Medical Centre, University Hospital Nottingham, Nottingham, UK

R. Justin Mistovich, MD Division of Orthopaedic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

Gregory M. Mundis Jr , MD Department of Orthopedic Surgery, Scripps Clinic Medical Group, San Diego, CA, USA

Jozef Murar, MD Department of Orthopaedic Surgery, Northwestern University, Chicago, IL, USA

Peter O. Newton, MD Division of Orthopedics, Rady Children's Hospital, San Diego, San Diego, CA, USA

Department of Orthopaedic Surgery, University of California, San Diego, San Diego, CA, USA

Acke Ohlin, MD Department of Orthopaedic Surgery, Malmö University Hospital, Lund University, Malmö, Sweden

Jean A. Ouellet, MD Department of Pediatric Surgery, Shriners Hospital for Children, Montreal, QC, Canada

Joshua M. Pahys, MD Shriners Hospitals for Children, Philadelphia, Philadelphia, PA, USA

Howard Young Park, MD Department of Orthopaedic Surgery, University of California, Los Angeles, Santa Monica, CA, USA

Kevin Parvaresh, MD Department of Orthopaedic Surgery, University of California, San Diego, San Diego, CA, USA

Jeff Pawelek, BS San Diego Center for Spinal Disorders, San Diego, CA, USA

Francisco J. Sanchez Perez-Grueso, MD Spine Surgery Orthopedics Department, Hospital Universitario La Paz, Madrid, Spain

Teeda Pinyavat, MD Department of Anesthesiology, Columbia University Medical Center, New York, NY, USA

Connie Poe-Kochert, RN, BS, BSN, CNP Division of Pediatric Orthopaedic Surgery, Rainbow Babies and Children's Hospital, University Hospitals Case Medical Center, Cleveland, OH, USA

Ahsan Qadeer, MBBS, MD Department of Anesthesiology and Perioperative Medicine, Children's Hospital of Georgia, Georgia Regents University, Augusta, GA, USA

S. Rajasekaran, MS (Ortho), MCh, FRCS, FACS, PhD Department of Orthopaedics, Traumatology and Spine Surgery, Ganga Hospital, Coimbatore, India

Gregory J. Redding, MD Department of Pediatrics, Seattle Children's Hospital, University of Washington School of Medicine, Seattle, WA, USA

Julie L. Reigrut, MS Department of Engineering, K2M, Inc., Leesburg, VA, USA

Joseph E. Reynolds Research and Development, SpineForm LLC, Cincinnati, OH, USA

Amer F. Samdani, MD Department of Neurosurgery, Shriners Hospitals for Children-Philadelphia, Philadelphia, PA, USA

James O. Sanders, MD Department of Orthopedics and Rehabilitation, University of Rochester, Rochester, NY, USA

John F. Sarwark, MD Department of Orthopaedic Surgery, Northwestern University, Feinberg School of Medicine, Chicago, IL, USA

Brian P. Scannell, MD Department of Orthopaedic Surgery, Carolinas HealthCare System, Levine Children's Hospital, Charlotte, NC, USA

Dietrich Schlenzka, MD, PhD Department of Spine and Paediatric Orthopaedics, ORTON Orthopaedic Hospital, Helsinki, Finland

John Schmidt, PhD Department of Engineering, K2M, Inc., Leesburg, VA, USA

Daniel M. Schwartz, PhD NeuroMonitoring Experts, Teaneck, NJ, USA

Richard M. Schwend, MD Division of Orthopaedic Surgery, Children's Mercy Hospital, Kansas City, MO, USA

Anthony K. Sestokas, PhD Department of Intraoperative Neuromonitoring, Specialty Care, Nashville, TN, USA

Suken A. Shah, MD Department of Orthopaedic Surgery, Nemours/Alfred I. DuPont Hospital for Children, Wilmington, DE, USA

Ajoy Prasad Shetty, MS (Ortho), DNB (Ortho) Department of Orthopaedics, Traumatology and Spine Surgery, Ganga Hospital, Coimbatore, India

David L. Skaggs, MD, MMM Department of Orthopaedic Surgery, Children's Hospital Los Angeles, Los Angeles, CA, USA

John T. Smith, MD Department of Orthopaedics, University of Utah, Salt Lake City, UT, USA

Brian Snyder, MD, PhD Department of Orthopaedic Surgery, Boston Children's Hospital, Boston, MA, USA

You-Qiang Song, PhD Department of Biochemistry, The University of Hong Kong, Hong Kong, China

Paul D. Sponseller, MD, MBA Department of Orthopaedic Surgery, Johns Hopkins Bloomberg Children's Center, Baltimore, MD, USA

Tricia St. Hilaire, MPH Children's Spine Foundation, Valley Forge, PA, USA

Lena S. Sun, MD Department of Anesthesiology, Columbia University Medical Center, New York, NY, USA

George H. Thompson, MD Division of Pediatric Orthopaedics, Rainbow Babies and Children's Hospital, University Hospitals Case Medical Center, Case Western Reserve Universal, Cleveland, OH, USA

Michael To, FRCS(Edin), FHKCOS, FHKAM(ORTH) Department of Orthopaedics and Traumatology, Queen Mary Hospital, Hong Kong, China

Evan Trupia, BS Department of Orthopaedic Surgery, Columbia University Medical Center, New York, NY, USA

Vidyadhar V. Upasani, MD Division of Orthopedics, Rady Children's Hospital, San Diego, San Diego, CA, USA

Department of Orthopaedic Surgery, University of California, San Diego, San Diego, CA, USA

Michael G. Vitale, MD, MPH Division of Pediatric Orthopaedics, Department of Orthopaedic Surgery, Columbia University Medical Center, New York, NY, USA

Department of Pediatric Orthopaedic Surgery, Morgan Stanley Children's Hospital of New York Presbyterian, New York, NY, USA

Eric J. Wall, MD Department of Orthopaedics, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Guanjun Wang, MD Department of Orthopaedics, General Hospital of Chinese Armed Police Forces, Beijing, China

James G. Wright, MD, MPH, FRCS (c) Department of Surgery, Hospital for Sick Children, Toronto, ON, Canada

Burt Yaszay, MD Department of Pediatric Orthopedics, Rady Children's Hospital San Diego, San Diego, CA, USA

Muharrem Yazici, MD Department of Orthopaedics, Hacettepe University Faculty of Medicine, Cankaya, Ankara, Turkey

Qibin Ye, MD Department of Orthopaedics, General Hospital of Chinese Armed Police Forces, Beijing, China

Caglar Yilgor, MD Department of Orthopaedics and Traumatology, Kiziltepe State Hospital, Mardin, Turkey

Guney Yilmaz, MD Department of Orthopaedics and Traumatology, Hacettepe University School of Medicine, Ankara, Turkey

Xinyu Zhang, MD Department of Orthopaedics, General Hospital of Chinese Armed Police Forces, Beijing, China

Part I

General

Embryology and Anatomy: Spine/ Spinal Cord

1

Shay Bess and Breton Line

Contents

Key Points

1. Development of the spine and spinal cord begins during the third week of gestation.
2. Early development includes formation of the axes of the embryo, formation of primitive neural tissue, and notochord development.
3. The axial skeleton eventually arises from the somites, and normal vertebral and neural formation is dependent upon normal development of the paraxial mesoderm and somites.
4. Errors in the formation of the paraxial mesoderm and somites and errors in the formation of the cartilaginous precursors to the vertebrae and neural arch structures lead to congenital scoliosis and spinal dysraphism conditions, as well as abnormalities in other developing organ systems.
5. Neurocentral joints allow continued growth of the spinal canal, and secondary vertebral ossification centers persist until the third decade of life.

S. Bess, MD (✉) • B. Line, BSME
Department of Orthopaedic Surgery, Rocky Mountain Hospital for Children, Presbyterian/St. Luke's Medical Center, 2055 High Street, suite 130, Denver, CO 80205, USA
e-mail: shay_bess@hotmail.com

B.A. Akbarnia et al. (eds.), *The Growing Spine: Management of Spinal Disorders in Young Children*,
DOI 10.1007/978-3-662-48284-1_1

1.1 Early Development

The initial development of the spine begins during the third week of gestation. The embryo at this stage of development exists as a two-cell layered structure called the bilaminar germ disc. On approximately day 15, a groove forms in the midline of the germ disc called the primitive groove. The primitive groove forms an initial deepening at the cranial end of the embryo and then extends caudally and grows along the length of the germ disc forming the cranial and caudal axes of the embryo. This central deepening is termed the primitive pit, and the collection of cells that surround the primitive pit forms the primitive node (Fig. 1.1). The head of the embryo eventually forms at the primitive pit and primitive node. The entire structure (primitive pit, node, and groove) is called the primitive streak. The primitive streak establishes the embryonic longitudinal axis, giving rise to left and right sides of the embryo. Therefore, the cranial/caudal, left/right, and ventral/dorsal axes are formed during this third week of gestation.

A three-layered embryo is formed by the proliferation and migration of epiblast cells through the primitive streak (Fig. 1.2a–c). Epiblast cells invade and replace the hypoblast cell layer, forming the definitive endoderm. Migration of epiblast cells between the epiblast and endoderm layers continues, forming a third cell layer, mesoderm. Upon establishment of the mesodermal layer, the epiblast is renamed the ectoderm or ectodermal layer.

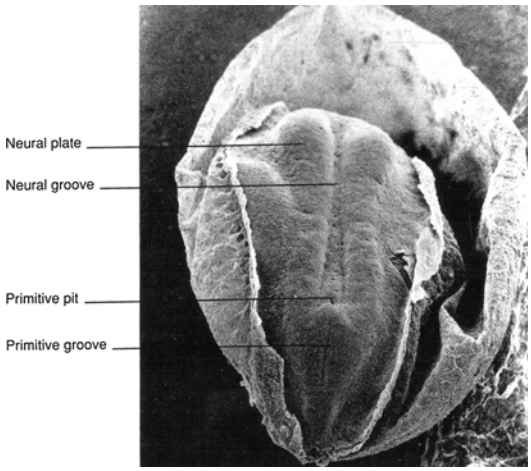

Fig. 1.1 Photomicrograph of primitive streak in the bilaminar germ disc. The primitive pit, primitive groove, and primitive node form the primitive streak. The head of the embryo will eventually form at the primitive pit and primitive node, and the entire structure (the primitive streak) establishes the embryonic longitudinal axis (Adapted from Tamarin (1983) With permission from John Wiley & Sons)

Two midline structures develop in the mesoderm: the prechordal plate and the notochordal process. The notochordal process begins as a hollow mesodermal tube and goes on to become a solid rod structure, called the notochord. The notochord induces the formation of the vertebral bodies, and subsequently, the vertebral bodies coalesce around the notochord inducing the notochord to form the nucleus pulposus (Fig. 1.3a, b).

Following the development of notochord, three distinct structures form in the mesoderm: the paraxial mesoderm, intermediate mesoderm, and lateral plate mesoderm. As pertains to the spine and spinal cord, the paraxial mesoderm, which lies adjacent to the notochord, gives rise to cell lines that form the critical structures called the somites. The somites are responsible for formation of the axial skeleton, voluntary musculature, and the skin dermis (Fig. 1.4). The intermediate mesoderm and lateral mesoderm are involved in the development of the urogenital and cardiopulmonary systems. As a consequence, defects that alter the development of the mesoderm resulting in vertebral abnormalities may also result in concurrent abnormalities in the urogenital and cardiopulmonary systems. VACTERL syndrome is an acronym with each letter representing an associated defect secondary to abnormalities in the mesodermal development including Vertebral anomalies, imperforate Anus, Cardiac abnormalities, TracheoEsophageal fistula, Renal dysplasia, and Limb malformations. Approximately 30–60 % of vertebral abnormalities diagnosed in childhood will have an additional organ system abnormality, with the genitourinary system most commonly involved. This underscores the need to evaluate for additional organ system involvement in children and infants with congenital vertebral abnormalities, including cardiac and renal ultrasound.

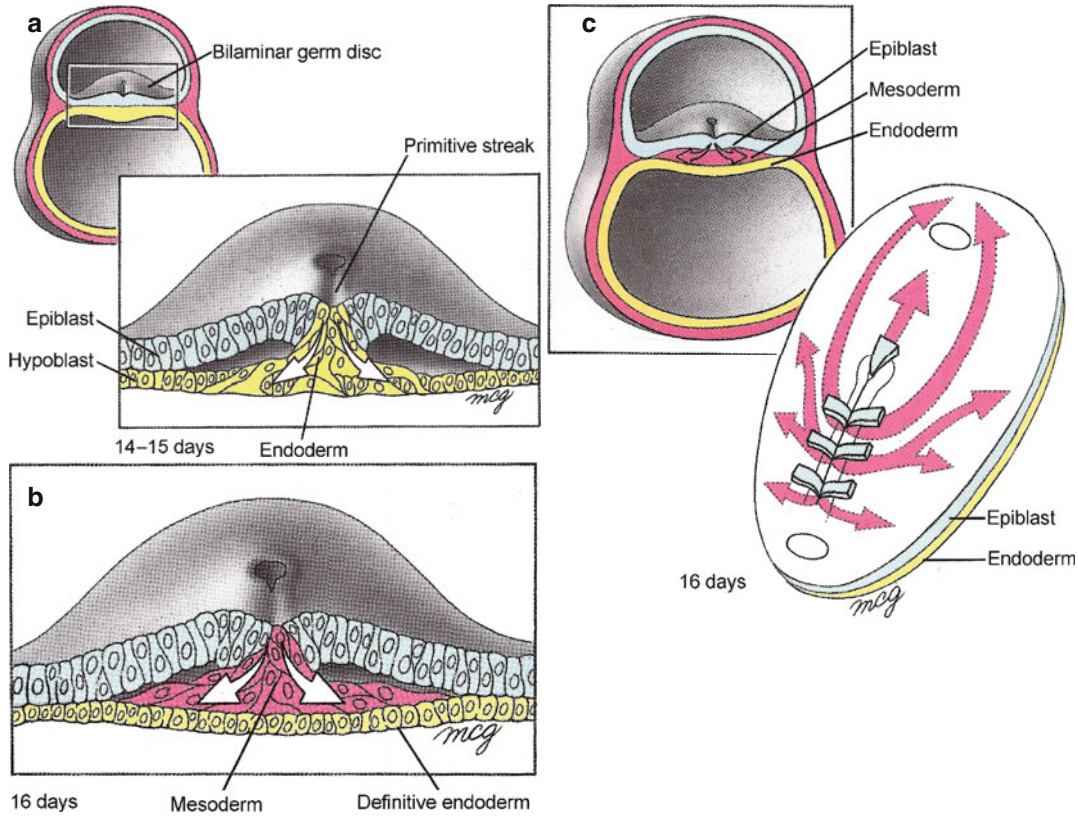

Fig. 1.2 (**a**–**c**) Proliferation and migration of epiblast cells. Epiblast cells proliferate and migrate through the primitive streak eventually forming the endoderm, mesoderm, and ectoderm; the definitive three-cell layered embryo (Adapted from Larsen (1993). With permission from Elsevier)

1.2 Somite Formation and Differentiation

The axial skeleton, voluntary muscle, and the dermis of the neck and trunk are derived from the somites. The somites emerge as paired on approximately gestational day 20, arising from the paraxial mesoderm and developing in a cranial to caudal fashion at a rate of approximately 3–4 somites per day (Fig. 1.5).Initially 42–44 somite pairs exist adjacent to the notochord. The cranial-most somite pairs eventually form the base of the skull and extend caudally to a rudimentary structure, the embryonic tail. However, caudal 5–7 somites regress, leaving a total of 37 somite pairs for development. Somite pairs 1–4 form the occiput as well as the bones of the face and inner ear. Somites 5–12 form the cervical spine (there are eight cervical somites but ultimately only seven cervical vertebrae because the first cervical somite participates in occiput formation). Somites 13–24 form the thoracic vertebrae, somites 25–29 form the lumbar vertebrae, and somites 30–34 form the sacral vertebrae. The remaining three terminal somite pairs form the coccyx and persist after regression of the terminal embryonic tail. The consecutive somite pairing on the embryo creates an anatomic template that organizes the vertebral alignment and the corresponding peripheral nervous system (PNS), which persists to maturity.

As the embryo develops, the somites separate into subdivisions. Accordingly, the ultimate tissue structure that develops from each somite is produced from the respective somite

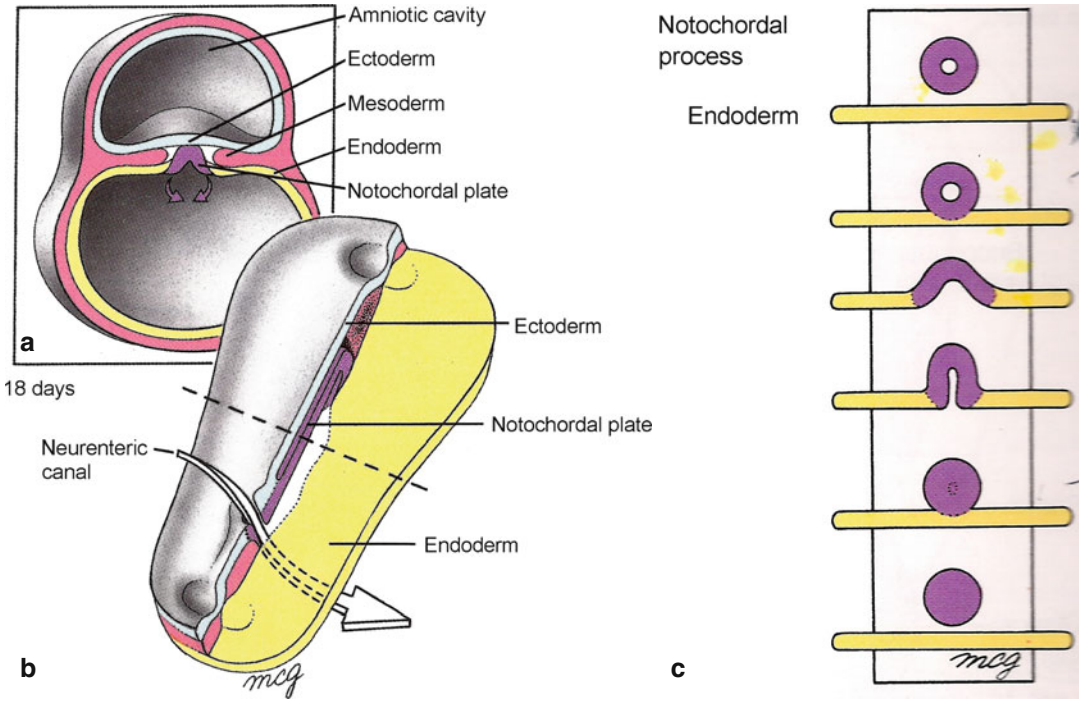

Fig. 1.3 (**a**–**c**) Formation of the notochordal process and notochord. The hollow notochordal process forms within the mesoderm and goes on to form the solid notochord. The notochord induces vertebral body formation and eventually becomes the nucleus pulposus (Adapted from Larsen (1993). With permission from Elsevier)

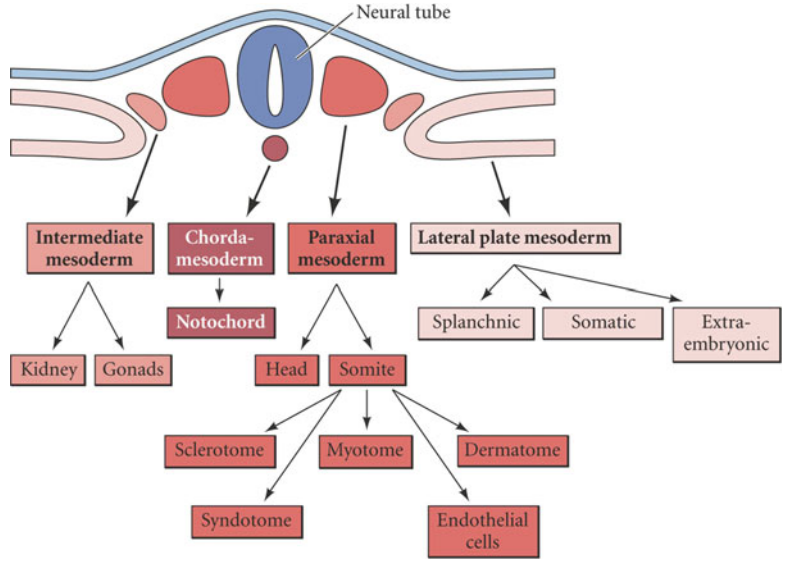

Fig. 1.4 Paraxial mesoderm, intermediate mesoderm, and lateral plate mesoderm formation, location and eventual structures (Adapted from Larsen (1993). With permission from Elsevier)

subdivision. The first somite subdivision that appear are the sclerotomes. The sclerotomes ultimately give rise to the bony spinal column. Sclerotomes are formed when a hollow central cavity forms within the somite. This cavity develops in the medial region of the somite adjacent to the midline notochord and neural tube. The central cavity fills with cells, termed

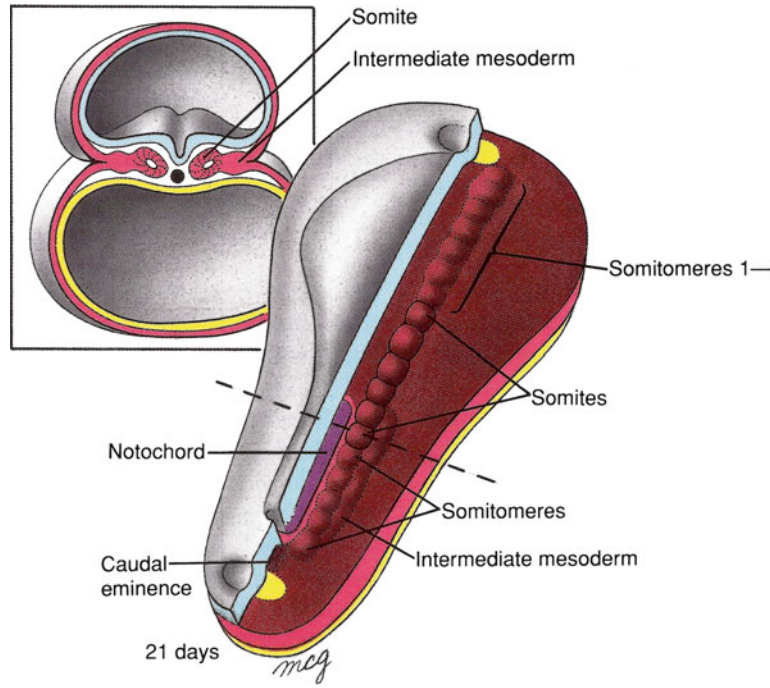

Fig. 1.5 Somite formation. The paired somites arise from the paraxial mesoderm and form the axial skeleton, voluntary muscle, and the dermis of the neck and trunk (Adapted from Larsen (1993). With permission from Elsevier)

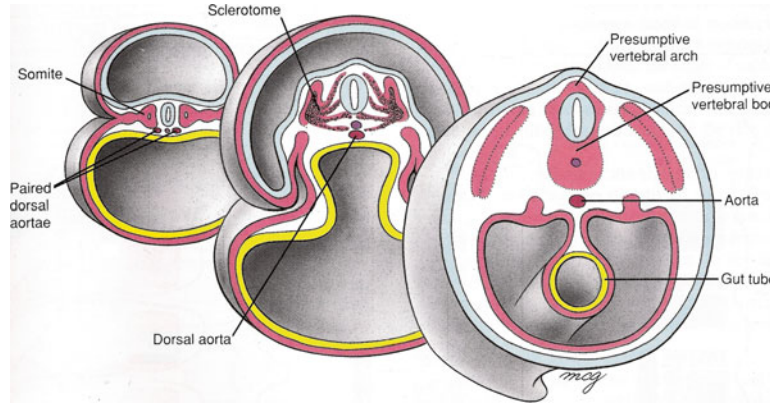

Fig. 1.6 Sclerotome formation. The central cavity within the somite fills with loose core cells and eventually ruptures. Core cells migrate toward the midline and envelop the notochord and neural tube forming a sclerotome. The ventral sclerotome forms the vertebral body, and the dorsal sclerotome becomes the vertebral arch (Adapted from Larsen (1993). With permission from Elsevier)

loose core cells, and eventually ruptures, allowing the core cells to migrate toward the midline and envelop the notochord and neural tube (Fig. 1.6). The cellular structure that eventually surrounds the notochord and neural tube is termed sclerotome. The ventral sclerotome that surrounds the notochord eventually becomes the vertebral body, and the dorsal sclerotome that envelops the neural tube eventually becomes the vertebral arch.

Normal vertebral body and vertebral arch development are dependent upon sclerotome induction by the underlying notochord and neural tube. Abnormalities in this sclerotomal-notochord induction signaling process creates spinal dysraphism, which is a spectrum of birth defects caused by failure of neural tube closure. Spina bifida is defined as incomplete closure of the neural arch leaving the underlying neural elements uncovered. The severity of spina bifida ranges from spina bifida occulta, in which the neural arch fails to completely close, to more severe conditions of spina bifida, in which the contents of the neural canal extend out of the

canal and become continuous with the overlying skin. The type and severity of spina bifida are classified by the neurological tissue that extends out of the canal, which may include the neural meninges (dura and arachnoid), as well as nerve roots. The neurological tissue that extends out of the spina bifida defect is contained within a membranous tissue called a cele. The cele is what is visible on the skin surface overlying the spina bifida defect and, as indicated above, may contain meningeal tissue, in which case, the cele is termed a meningocele. The cele may also contain neural tissue and meninges, called a meningomyelocele.

Once the sclerotomes form and become positioned adjacent to the notochord and neural tube, each sclerotome divides into a cranial and a caudal portion. This cranial and caudal division allows the spinal nerves to emerge from the neural tube and exit at their respective level (Fig. 1.7a–d). Once the sclerotome division is complete, the caudal portion of the suprajacent sclerotome merges with the cranial portion of the subjacent sclerotome. This sclerotomal merging forms the vertebral precursor. This sclerotomal division and then the subsequent re-fusion explain why there are eight cervical nerves but only seven cervical vertebrae (Fig. 1.7d). The cranial division of the first cervical somite forms a portion of the base of the occiput, while the caudal division of the first cervical somite and the cranial division of the second cervical somite form the atlas. The first cervical nerve exits above the C1 vertebra, the second cervical nerve exits between C1 and C2; this pattern persists to the C7-T1 foramen where the C8 nerve root exits. The sclerotomal cells that remain following the sclerotome division surround the notochord and form annulus fibrosis, which is the fibrous portion of the intervertebral disc. The portions of the notochord that becomes enveloped by the sclerotomal tissue form the nucleus pulposus. Then during the process of maturity, the original notochord cells of the nucleus pulposus are replaced by fibrocartilageous cells.

1.3 Central Nervous System Development

Two key structures originate in the mesoderm during early development: the notochordal process and the prechordal plate. The prechordal plate induces the overlying epiblast cell layer to form the neural plate, then the neural plate cells differentiate into neurectoderm. Once formed, the neurectoderm proliferates in a cranial to caudal fashion. The cranial portion of the neural plate is broad shaped and gives rise to the brain, while the tapered caudal region of the neural plate forms the spinal cord. The positioning of the neural plate as it develops is such that the caudal portion of the neural plate overlies the notochord and is bordered by the somite pairs. This positioning allows the caudal portion of the neural plate to become enveloped by the sclerotomes forming the spinal canal, and then the neural plate itself becomes the spinal cord (Fig. 1.8). The neural plate becomes the neural tube by a process called neurulation, in which the neural plate involutes, until the lateral edges of the folded neural plate and overlying ectoderm meet and fuse in the midline forming the tubular shape of the neural tube (Fig. 1.9).

Once the neural tube fuses in the midline, it separates from the overlying ectoderm and differentiates into three distinct layers (Fig. 1.10). The innermost cell layer of the neural tube, called the ventricular layer, lays adjacent to the lumen of the neural tube (the neural canal). The ventricular layer is comprised of neuroepithelial cells, which are the precursors to the cells that eventually comprise the CNS. The first generation of cells produced by the neuroepithelial cells are neuroblasts. Neuroblasts eventually become the neurons in the CNS. Once formed, neuroblasts migrate away from the ventricular layer to form the mantle layer. The mantle layer eventually becomes the gray matter of the CNS. The neuroblasts in the mantle layer organize into four columns during the fourth week of gestation, forming paired dorsal and ventral col-