"A vital educational resource for patients, their families, and health care professionals alike."

—BRIAN SNYDER, MD, PhD

Gillette Children's Healthcare Series

SCOLIOSIS

Congenital, Neuromuscular, Syndromic, and Other Nonidiopathic Types

Understanding and managing the condition: A practical guide for families

Tenner J. Guillaume, MD
Walter H. Truong, MD
Danielle Harding, PA-C
Lily Collison, MA, MSc
Cheryl Tveit, RN, MSN, CNML

Editors
Elizabeth R. Boyer, PhD
Tom F. Novacheck, MD

GILLETTE CHILDREN'S

Praise for *Scoliosis—Congenital, Neuromuscular, Syndromic, and Other Nonidiopathic Types*

"A vital educational resource for patients, their families, and health care professionals alike, emphasizing evidence-based best practice and providing guidance for exploring the literature and further research. Beyond the pragmatic medical information provided, the text underscores the critical partnerships between patients, their families and caretakers, and health care professionals in optimizing outcomes for individuals living with this lifelong condition."

BRIAN SNYDER, Maurice Mueller Professor of Orthopaedic Surgery, Harvard Medical School; Orthopaedic Surgeon, Cerebral Palsy Center, Boston Children's Hospital, US

"Another amazing masterpiece in the Gillette Children's Healthcare Series, after the earlier book on idiopathic scoliosis. It is impressive that all the queries that parents of children with scoliosis due to known causes might have and that may come up along the course of treatment are collected in a concise yet thorough source with current, speculation-free, easily readable responses."

MUHARREM YAZICI, Professor of Orthopaedics, Children's Orthopaedics and Spine Center, Ankara, Turkey; Past President European Paediatric Orthopaedic Society; Past President Scoliosis Research Society

"The team at Gillette Children's has created an outstanding second book on scoliosis. This resource skillfully combines the latest clinical research with personal stories, weaving together a rich tapestry of experiences that reflect the many diverse aspects of this condition. As a parent of a teenage daughter recently diagnosed with complex scoliosis, I understand the pressing need for accurate information and support. I often wished for a reference guide of this caliber during my search for information to help us navigate this journey. This uniquely practical guide offers clear language and step-by-step instructions, addressing the complexities of scoliosis throughout a person's life. Having access to this information ultimately empowers families like ours to make informed decisions in collaboration with multidisciplinary teams, balancing clinical insights with the evolving needs of our children. I highly recommend this impactful book for families and professionals working in the field of scoliosis. Congratulations to everyone who contributed to this invaluable work, which brings hope and clarity to families living with scoliosis."

ANN MARIE SUTTON, Parent of daughter with neuromuscular scoliosis, Ireland

"This is the premier resource for patients and parents affected by nonidiopathic scoliosis. It is a comprehensive book that outlines the science behind how scoliosis occurs and how it is treated when discovered. It is easy to read for nonmedical individuals but also detailed enough for medical professionals to use as a reference. It will help countless patients and their families to answer questions about their condition and how to navigate the many treatment options."

ROBERT H. CHO, Pediatric Orthopaedic Surgeon, Pediatric Spine Surgeon; UCLA Clinical Assistant Professor, US

"Gillette has put together another great resource for patients and families. The figures are clear and easy to understand. The team did an excellent job making this difficult subject matter accessible to all."

KEITH D. BALDWIN, Director of Orthopedic Trauma, Associate Professor, Children's Hospital of Philadelphia, US

"This book offers an invaluable resource for families navigating the complexities and emotions of scoliosis management. The comprehensive and compassionate approach presents clear explanations as well as practical strategies, making the book an essential resource for parents. The accessible writing style helps families to understand their child's condition and actively participate in their care."

DAVID P. MOORE, Consultant Orthopaedic Surgeon, Children's Health Ireland

"The experts at Gillette Children's Hospital have written a well-structured and accessible guide for families managing the complexities of nonidiopathic scoliosis. Focused on congenital, syndromic, and neuromuscular forms of the condition, this resource outlines the diagnostic process and treatment pathways—including both nonoperative and surgical approaches—with clarity and up-to-date clinical precision. Authentic stories from patients and families humanize these often challenging diagnoses. This book will empower families as they collaborate with their spine care teams on treatment decisions."

SUMEET GARG, Pediatric Orthopaedic Surgeon, Children's Hospital Colorado; Professor of Orthopaedic Surgery, University of Colorado School of Medicine, US

"The book blew me away. I cannot believe how easy it was to understand; I learned so much. I wish I had this years ago. I'm sad I didn't but so happy to have it now. I'm going to read it again! All surgeons and those associated with scoliosis should read this book."

DEIRDRE MCDONNELL, Adult with congenital scoliosis, Ireland

"I thoroughly enjoyed reading the book. I believe many families dealing with scoliosis, patients themselves, and health care team members will find it highly beneficial. The clear and helpful illustrations effectively explain scoliosis management, making it an excellent resource for understanding treatment options."

DAVID E. LEBEL, Paediatric Orthopaedic Surgeon, Head of Spine Program, Hospital for Sick Children (SickKids), Toronto; Associate Professor, Faculty of Medicine, University of Toronto, Canada

"Medical diagnoses such as scoliosis can be stressful to both patients and their families. While there is information available for health care professionals through the scientific literature, there is a lack of information available for families and other laypeople. This book is the exact resource needed for patients and families. With straightforward organization and easy language, it is a pleasure to read and quite informative. The unique combination of professional information and the family perspective on the conditions and treatment makes this a must-read for both health care professionals and patients alike. The authors and institution should be commended for such a fine contribution that will empower families and enhance the care of patients with scoliosis."

MICHAEL J. HEFFERNAN, Director, Pediatric Orthopaedic Surgery Fellowship Program, Children's Hospital Los Angeles; Associate Professor, Department of Orthopaedic Surgery, Keck School of Medicine, University of Southern California, US

"I am not only a parent of a child who has scoliosis but a medical professional as well, and this book provides priceless information for families and patients who have scoliosis. The journey of scoliosis treatment varies greatly between every person who has the condition, and this book, full of factual and practical information, highlights the different causes and treatments. My favorite parts are the patient stories that remind me how resilient all kids are!"

BECCA WURGLER, Parent of child with neuromuscular scoliosis, US

"After the success of the book Idiopathic Scoliosis, *Gillette's scoliosis experts have made another significant contribution. In this volume, they focus on some of the most challenging spine deformity cases, including congenital, syndromic, and neuromuscular scoliosis. The book is an informative and supportive resource for families and patients with nonidiopathic scoliosis. Insightful and compassionate, it provides doctors, patients, and families with a guide to understanding the condition. The authors expertly translate complex medical concepts into accessible language, offering valuable insights and practical advice. The clinical stories candidly address the associated difficulties, anxieties, and fears of the disease, offering valuable emotional support and reassurance that patients and their families are not alone. It's clear that the book was written by experienced professionals who care deeply about their scoliosis patients. It is a must-read for families navigating this challenging condition."*

FEDERICO CANAVESE, Head, Orthopaedic and Traumatology Department, IRCCS Giannina Gaslini Institute, Genoa; Professor of Pediatric Orthopaedics, University of Genoa, Italy

"This book addresses a complex topic in a thorough but accessible way. The authors explain medical concepts with clarity and empathy while offering a balanced view of diagnosis and treatment options. The integration of patients' lived experiences with up-to-date treatment frameworks makes it unique and relevant as a clinical and educational tool. It is a commendable contribution to family-centered health education."

JOHN S. VORHIES, Director of Scoliosis and Spine Deformity Surgery and Research, Assistant Professor of Orthopaedic Surgery, Stanford Medicine Children's Health

"Accessible, well written, and easy to understand, this book summarizes nonidiopathic scoliosis from soup to nuts for everyone from patients and families to medical professionals caring for these patients. The text makes it clear that caring for these patients means caring for the entire patient (and family), not just the condition of scoliosis. The patient vignettes scattered throughout the book create empathy and a much better understanding of the patient experience."

BENJAMIN D. ROYE, Pediatric Orthopedic Surgeon, Morgan Stanley Children's Hospital of New York; Richard T. Arkwright–St. Giles Foundation Associate Professor, Orthopedic Surgery, Columbia University, US

SCOLIOSIS
CONGENITAL, NEUROMUSCULAR, SYNDROMIC, AND OTHER NONIDIOPATHIC TYPES

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To individuals and families whose lives are affected by these conditions, to professionals who serve our community, and to all clinicians and researchers who push the knowledge base forward, we hope the books in this Healthcare Series serve you very well.

All proceeds from the books in this series at Gillette Children's go to research.

Contents

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Series Foreword

You hold in your hands one book in the Gillette Children's Healthcare Series. This series was inspired by multiple factors.

It started with Lily Collison writing the first book in the series, *Spastic Diplegia–Bilateral Cerebral Palsy*. Lily has a background in medical science and is the parent of a now adult son who has spastic diplegia. Lily was convincing at the time about the value of such a book, and with the publication of that book in 2020, Gillette Children's became one of the first children's hospitals in the world to set up its own publishing arm—Gillette Children's Healthcare Press. *Spastic Diplegia–Bilateral Cerebral Palsy* received very positive reviews from both families and professionals and achieved strong sales. Unsolicited requests came in from diverse organizations across the globe for translation rights, and feedback from families told us there was a demand for books relevant to other conditions.

We listened.

We were convinced of the value of expanding from one book into a series to reflect Gillette Children's strong commitment to worldwide education. In 2021, Lily joined the press as Program Director, and very quickly, Gillette Children's formed teams to write the Healthcare Series. The series includes, in order of publication:

- *Craniosynostosis*
- *Idiopathic Scoliosis*
- *Spastic Hemiplegia—Unilateral Cerebral Palsy*
- *Spastic Quadriplegia—Bilateral Cerebral Palsy*
- *Spastic Diplegia—Bilateral Cerebral Palsy, second edition*
- *Epilepsy*
- *Scoliosis—Congenital, Neuromuscular, Syndromic, and Other Nonidiopathic Types*
- *Spina Bifida*
- *Osteogenesis Imperfecta*

The books address each condition detailing both the medical and human story.

Mac Keith Press, long-time publisher of books on disability and the journal *Developmental Medicine and Child Neurology*, is co-publishing this series with Gillette Children's Healthcare Press.

Families and professionals working well together is key to best management of any condition. The parent is the expert of their child while the professional is the expert of the condition. These books underscore the importance of that family and professional partnership. For each title in the series, medical professionals at Gillette Children's have led the writing, and families contributed the lived experience.

These books have been written in the United States with an international lens and citing international research. However, there isn't always strong evidence to create consensus in medicine, so others may take a different view.

We hope you find the book you hold in your hands to be of great value. We collectively strive to optimize outcomes for children, adolescents, and adults living with these childhood-acquired and largely lifelong conditions.

Dr. Tom F. Novacheck

Series Introduction

The Healthcare Series seeks to optimize outcomes for those who live with childhood-acquired physical and/or neurological conditions. The conditions addressed in this series of books are complex and often have many associated challenges. Although the books focus on the biomedical aspects of each condition, we endeavor to address each condition as holistically as possible. Since the majority of people with these conditions have them for life, the life course is addressed including transition and aging issues.

Who are these books for?

These books are written for an international audience. They are primarily written for parents of young children, but also for adolescents and adults who have the condition. They are written for members of multidisciplinary teams and researchers. Finally, they are written for others, including extended family members, teachers, and students taking courses in the fields of medicine, allied health care, and education.

A worldview

The books in the series focus on evidence-based best practice, which we acknowledge is not available everywhere. It is mostly available in high-income countries (at least in urban areas, though even there, not always), but many families live away from centers of good care.

We also acknowledge that the majority of people with disabilities live in low- and middle-income countries. Improving the lives of all those with disabilities across the globe is an important goal. Developing scalable, affordable interventions is a crucial step toward achieving this. Nonetheless, the best interventions will fail if we do not first address the social determinants of health—the economic, social, and environmental

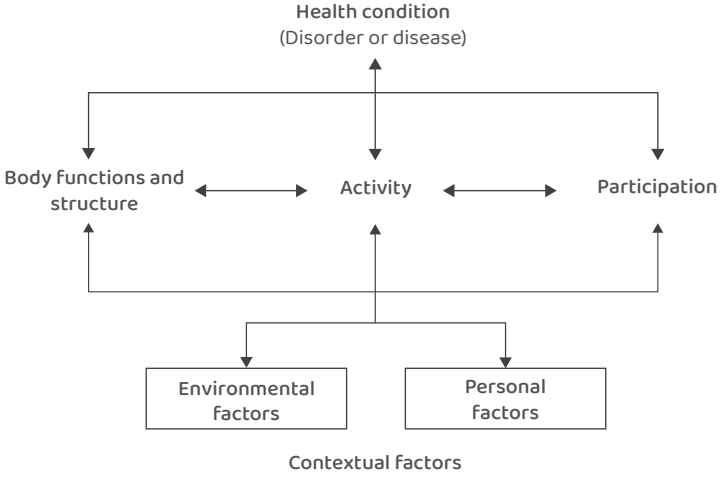
conditions in which people live that shape their overall health and well-being.

No family reading these books should ever feel they have failed their child. We all struggle to do our best for our children within the limitations of our various resources and situations. Indeed, the advocacy role these books may play may help families and professionals lobby in unison for best care.

International Classification of Functioning, Disability and Health

The writing of the series of books has been informed by the International Classification of Functioning, Disability and Health (ICF).[1] The framework explains the impact of a health condition at different levels and how those levels are interconnected. It tells us to look at the full picture—to look at the person with a disability in their life situation.

The framework shows that every human being can experience a decrease in health and thereby experience some disability. It is not something that happens only to a minority of people. The ICF thus "mainstreams" disability and recognizes it as a widespread human experience.



International Classification of Functioning, Disability and Health (ICF). Reproduced with kind permission from WHO.

In health care, there has been a shift away from focusing almost exclusively on correcting issues that cause the individual's functional problems to focusing also on the individual's activity and participation. These books embrace maximizing participation for all people living with disability.

The Family

For simplicity, throughout the series we refer to "parents" and "children"; we acknowledge, however, that family structures vary. "Parent" is used as a generic term that includes grandparents, relatives, and carers (caregivers) who are raising a child. Throughout the series, we refer to male and female as the biologic sex assigned at birth. We acknowledge that this does not equate to gender identity or sexual orientation, and we respect the individuality of each person. Throughout the series we have included both "person with disability" and "disabled person," recognizing that both terms are used.

Caring for a child with a disability can be challenging and overwhelming. Having a strong social support system in place can make a difference. For the parent, balancing the needs of the child with a disability with the needs of siblings—while also meeting employment demands, nurturing a relationship with a significant other, and caring for aging parents—can sometimes feel like an enormous juggling act. Siblings may feel neglected or overlooked because of the increased attention given to the disabled child. It is crucial for parents to allocate time and resources to ensure that siblings feel valued and included in the family dynamics. Engaging siblings in the care and support of the disabled child can help foster a sense of unity and empathy within the family.

A particular challenge for a child and adolescent who has a disability, and their parent, is balancing school attendance (for both academic and social purposes) with clinical appointments and surgery. Appointments outside of school hours are encouraged. School is important because the cognitive and social abilities developed there help maximize employment opportunities when employment is a realistic goal. Indeed, technology has eliminated barriers and created opportunities that did not exist even 10 years ago.

Parents also need to find a way to prioritize self-care. Neglecting their own well-being can have detrimental effects on their mental and physical health. Think of the safety advice on an airplane: you are told that you must put on your own oxygen mask before putting on your child's. It's the same when caring for a child with a disability; parents need to take care of themselves in order to effectively care for their child *and* family. Friends, support groups, or mental health professionals can provide an outlet for parents to express their emotions, gain valuable insights, and find solace in knowing that they are not alone in their journey.

For those of you reading this book who have the condition, we hope this book gives you insights into its many nuances and complexities, acknowledges you as an expert in your own care, and provides a road map and framework for you to advocate for your needs.

Last words

This series of books seeks to be an invaluable educational resource. All proceeds from the series at Gillette Children's go to research.

Chapter 1

Scoliosis

1.1

Introduction

You have brains in your head. You have feet in your shoes.
You can steer yourself any direction you choose.
Dr. Seuss

If you were to look up the definition of scoliosis (pronounced SKOL-ee-oh-sis, with the capital letters showing the emphasis on that syllable), you would find many sources defining it as a sideways curvature of the spine. This is generally correct, but a more accurate definition of scoliosis is a condition in which there is an atypical three-dimensional curvature and rotation of the spine.[2]

There are many types of scoliosis with varied causes. The term "idiopathic" is defined as "relating to a disease of unknown cause," and it is used in the context of many medical conditions for which the cause is unclear or unknown. This book focuses on nonidiopathic scoliosis. Nonidiopathic scoliosis is scoliosis that develops from a *known* cause and is the least common type of scoliosis, accounting for an estimated 20 percent of all scoliosis cases.[3]

Nonidiopathic scoliosis includes:

- **Congenital scoliosis:** scoliosis caused by errors in vertebral development
- **Neuromuscular scoliosis:** scoliosis secondary to (or as a result of) a primary neuromuscular condition, which is a condition that impacts the nervous and/or muscular systems
- **Syndromic scoliosis:** scoliosis secondary to (or as a result of) a primary syndrome, which is a group of symptoms that consistently occur together
- **Other nonidiopathic scoliosis:** scoliosis due to other *known* causes

This book explains scoliosis and its management for each of the four types above. The term "nonidiopathic scoliosis" is used throughout this book as a collective and shorter term for the four types. Conditions associated with scoliosis, particularly if severe, may affect many body systems, and the management is often complex and requires a multidisciplinary team. It is beyond the scope of this book to explain aspects of management other than scoliosis.

How this book is organized

This chapter covers the overall condition of scoliosis. The other chapters are organized as follows:

- Chapters 2 to 5 explain the four types of nonidiopathic scoliosis.
- Chapters 6 to 9 explain treatment.
- Chapter 10 addresses considerations of aging.
- Chapter 11 provides further reading and research.

Throughout the book, medical information is interspersed with personal lived experience. Orange boxes highlight the personal stories.

At the back of the book, you'll find a glossary of key terms. A companion website for this book is available at www.GilletteChildrensHealthcarePress.org. A QR code to access **Useful web resources** is included below.

Finally, this book addresses scoliosis that develops from a known cause; other titles in the series address the full conditions themselves. A list of other titles in the series is included in the Foreword and on the back cover.

USEFUL WEB RESOURCES

1.4

Classification of scoliosis

To live is the rarest thing in the world. Most people exist, that is all.
Oscar Wilde

The two most common factors by which scoliosis is classified are cause and age at diagnosis. Classification is useful for describing someone's scoliosis and how it may progress. It also helps medical professionals make care decisions and recommend treatment options.

By cause

There are five types of scoliosis classified by cause:

- Idiopathic
- Congenital
- Neuromuscular
- Syndromic
- Other nonidiopathic scoliosis

See Table 1.4.1 for details.

Table 1.4.1 Classification of scoliosis based on cause

TYPE	CAUSE
Idiopathic	The term "idiopathic" means there is no known cause. With idiopathic scoliosis, the spine grows in a curved and rotated fashion for unknown reasons.
Congenital	The term "congenital" means present from birth. Congenital scoliosis is caused by errors in vertebral development. The child is born with an atypical spine (atypical vertebrae and/or atypical intervertebral discs) that can cause atypical growth of the spine, resulting in scoliosis.
Neuromuscular	The term "neuromuscular" means a condition involving the nervous system and/or muscles; it includes conditions such as cerebral palsy. Because nerves and/or muscles are affected, it can prevent the body from being able to physically support a growing spine, resulting in scoliosis.
Syndromic	The term "syndromic" means a group of symptoms that consistently occur together. Syndromic scoliosis is caused by a syndrome, such as Marfan syndrome or Down syndrome. These conditions can cause connective tissue (e.g., bone, blood vessels, cartilage, ligaments, tendons) to weaken, resulting in scoliosis.
Other nonidiopathic scoliosis	Other causes include conditions such as neural axis abnormalities, which are atypical structures within the central nervous system (brain and/or spinal cord) that can impact the growth of the spine.

By age of diagnosis

Scoliosis can also be classified by the age of diagnosis. One age-based classification is early-onset scoliosis (EOS), which is defined as scoliosis that is diagnosed prior to 10 years of age, regardless of cause or type.

There are also age-based classifications specific to *idiopathic* scoliosis:

- Infantile idiopathic scoliosis (IIS)—age of diagnosis 0 to 3 years
- Juvenile idiopathic scoliosis (JIS)—age of diagnosis 4 to 9 years
- Adolescent idiopathic scoliosis (AIS)—age of diagnosis 10 to 18 years

1.5

Diagnosis of scoliosis

In three words I can sum up everything
I've learned about life: it goes on.
Robert Frost

Individuals can be diagnosed with scoliosis at any age. However, it is most commonly diagnosed between the ages of 10 and 15. Children enter their pubertal growth spurt at this age, and this rapid growth is associated with an increased risk of scoliosis curve progression (i.e., getting larger). The larger a curve becomes, the more noticeable the signs and symptoms become.

- A **sign** is what can be seen by observing the individual (e.g., a visibly curved spine).
- A **symptom** is what the individual describes as experiencing due to the condition (e.g., back pain).

This section describes the typical diagnostic journey for children and adolescents with scoliosis, from first detection to the first appointment with a spine specialist. Of course, a "typical" journey will not apply to everyone; the diagnostic journey may look very different depending on

the country the family lives in or hospital or treatment center where the individual with scoliosis is receiving care.

First detection

Typically, scoliosis is first noted by the child, parent, or the care provider. The following changes in a child's appearance are signs of scoliosis:[6]

- One shoulder being higher than the other
- A curved spine that looks like an "S" or "C" rather than a straight line down the back
- Asymmetry (unevenness) of the waist
- One shoulder blade being more noticeable than the other
- Chest shifted to one side
- Clothes fitting unevenly
- One hip being higher than the other
- Ribs more prominent on one side than the other

Figure 1.5.1 shows three of these common signs.

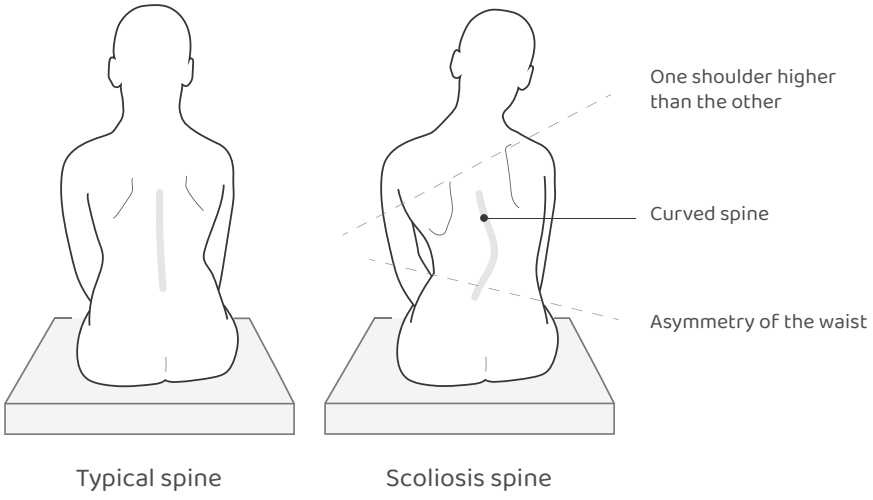


Figure 1.5.1 Three common signs of scoliosis.

Care providers may routinely check for scoliosis during checkups. They may examine the back with the individual leaning forward, either sitting or standing (called an Adams forward bend test), which can help

identify scoliosis by revealing asymmetry or signs of rotation and curvature of the spine. As shown in Figure 1.5.2, when an individual with scoliosis bends over, one side may appear higher or more prominent than the other. If the care provider suspects scoliosis, they may have X-rays taken of the spine.

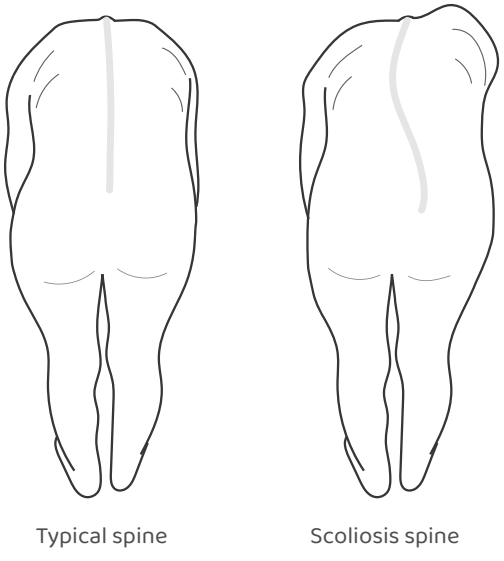


Figure 1.5.2 Adams forward bend test.

Children and adolescents who display signs and symptoms of scoliosis will be referred to a spine specialist. Their first visit may be with a spine surgeon or an advanced practice provider such as a physician assistant or a nurse practitioner.

First appointment with a spine specialist

At the first visit, there are many things the spine specialist will ask about and observe. They will verify the presence of scoliosis through X-rays and a physical examination.

An individual's personal medical history, family medical history, physical exam, and imaging results are all puzzle pieces a spine specialist will use to learn more about the scoliosis and to then determine the best treatment plan.

a) Personal medical history

Taking a person's medical history, also called a "past medical history," gives the spine specialist clues to possible causes and risk factors of the individual's scoliosis. This medical history includes the mother's experience during pregnancy as well as the individual's prior or current medical diagnoses, history of relevant symptoms such as missed developmental milestones, or previous medical procedures. It is possible that an individual, especially a young person, may present with scoliosis caused by a medical condition that has not yet been diagnosed.

b) Family medical history

Learning more about an individual's family medical history, including that of the parents, siblings, and grandparents, is also important. Certain types of scoliosis, or conditions that cause scoliosis, can have a genetic and heritable component, meaning they can run in families and can be passed down through generations.

c) Physical exam

The physical exam is another important piece of the puzzle when diagnosing scoliosis. During a physical exam, a spine specialist will conduct a nerve and skin exam and observe the individual walking, if they are able. The physical exam looks at the underlying medical condition that may impact scoliosis care as well as overall health. It also offers the spine specialist a chance to learn more about the individual's scoliosis curve. Flexibility measurements may be performed, such as bending side to side, which can indicate the stiffness or flexibility of the curve. An Adams forward bend test can provide information about the amount of vertebral rotation and curvature. A scoliometer, a specifically designed level, can be used to measure the degree of vertebral rotation, as shown in Figure 1.5.3.

During the exam, the spine specialist may take photos of the individual to document asymmetry in the hips, waistline, or shoulder height. These initial photos and measurements record the baseline values and can be used to assess curve progression and the effectiveness of future treatment.

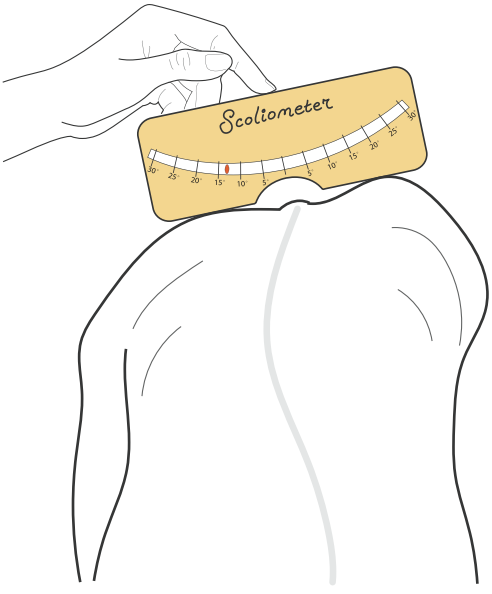


Figure 1.5.3 Adams forward bend test with a scoliometer to measure the degree of vertebral rotation.

d) Imaging results

X-rays are extremely important and useful for diagnosing scoliosis. Full-length standing or seated X-rays of the spine will be taken from the back and the side (Figure 1.5.4).

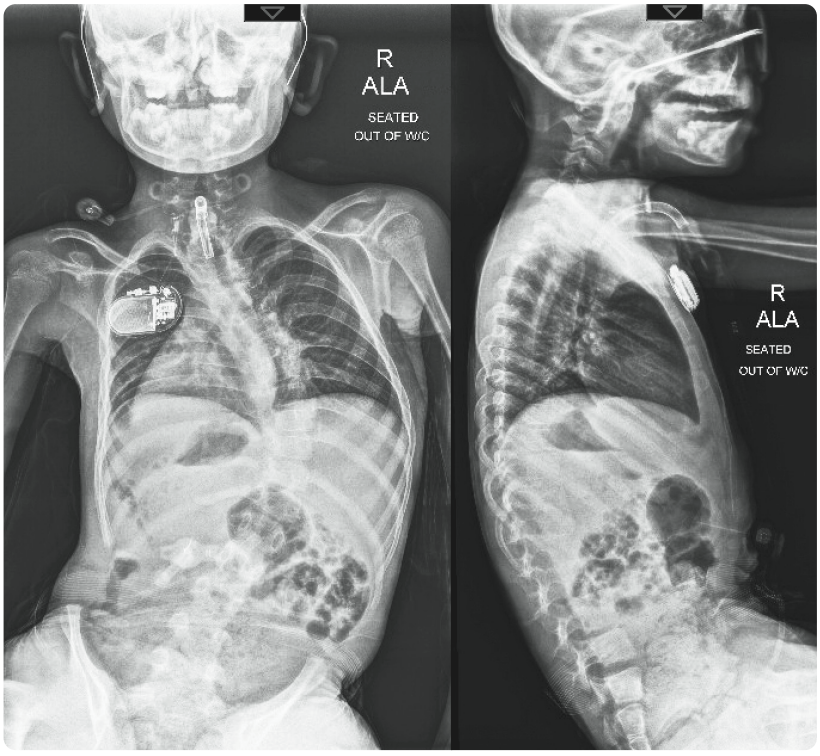


Figure 1.5.4 Scoliosis seated coronal X-ray view (left) and sagittal X-ray view (right).

These X-rays provide a two-dimensional view of the bones of the spine in the coronal and sagittal planes. At some hospitals and treatment centers, these images may be taken with technology that uses low-dose X-ray imaging and captures both the coronal and sagittal images at the same time. Associated software then allows for the production of two-dimensional images and a virtual three-dimensional reconstruction of the spine.

On the coronal plane view, the spine of a person with scoliosis looks like an "S" or "C" instead of a straight line. The individual shown in Figure 1.5.4 has a C-shaped curve in their spine. **Note:** Throughout this book, the singular term "scoliosis curve" is used, but it is important to know that an individual may have more than one scoliosis curve.

Spine specialists measure the angle of the scoliosis curve on an X-ray in the coronal plane. This measurement is called a Cobb angle (or "curve magnitude"). It is determined by measuring the angle between the two most tilted vertebrae at the upper and lower ends of a spinal curve

(Figure 1.5.5). The Cobb angle is the most commonly used measurement for quantifying the size of a spinal curve,[7] and it is the means by which a scoliosis diagnosis is made. Scoliosis is diagnosed when the Cobb angle is 10 degrees or greater. A slight curve of 1 to 9 degrees is called "spinal asymmetry" and is still considered typical (i.e., it is not considered scoliosis). The Cobb angle is measured on every X-ray over time to assess whether there is curve progression (increase in size).

Note that a very small difference of 1 degree in Cobb angle—from 9 to 10 degrees—changes the medical diagnosis from no scoliosis to scoliosis. Small differences in an individual's posture during the X-ray or in the measure of the Cobb angle can result in different angles. This is normal measurement variability, similar to when you take body temperature readings three times with the same thermometer and get three slightly different readings.

It is typical for specialists to consider a scoliosis curve to have significantly progressed only if the change in Cobb angle is greater than 5 degrees. This is considered evidence of progression as opposed to measurement variability described above.

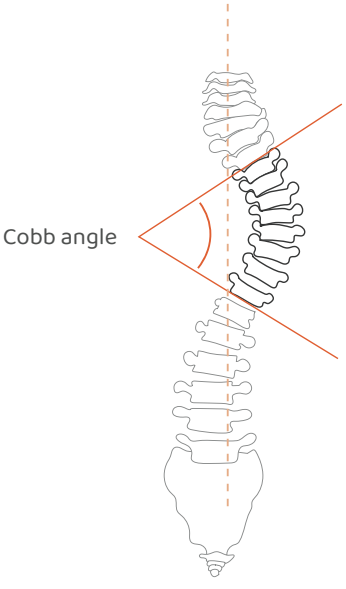


Figure 1.5.5 Cobb angle measurement.

In addition to ordering a spine X-ray, the spine specialist may order a hand X-ray. The bones in the hand have important growth plates (areas of active, new bone growth that are made of cartilage and turn to solid bone when growing is done). The hand image allows the spine specialist to estimate how much skeletal growth remains for an individual.

After the initial spine visit, the spine specialist may also order a spine MRI, which provides detailed imaging of soft tissue (i.e., not bony) internal structures in the body, such as the spinal cord and intervertebral discs. A spine MRI can be used to check for any neural axis abnormalities, which are atypical structures within the central nervous system (brain and spinal cord) that can impact the curvatures and growth of the spine.

Key points Chapter 1

- The spine consists of vertebrae (bony structures with a hole for the spinal cord) and intervertebral discs (cartilage structures that sit between vertebral bodies).
- The spine protects the spinal cord, serves as an attachment point for the ribs and supporting muscles and ligaments, supports the weight of the body, and provides points of movement for the head and torso.
- Humans have 33 vertebrae, which are commonly grouped into five regions of the spine: cervical, thoracic, lumbar, sacral, and coccygeal.
- When looking at the side view of a person, the spine has distinct curvature. Lordosis is an inward curvature (arching toward the center of the body) and kyphosis is an outward curvature (rounding away from the center of the body).
- Typical spine curvature is a slight cervical lordosis, thoracic kyphosis, lumbar lordosis, and sacral kyphosis.
- Scoliosis is a condition in which there is an atypical three-dimensional curvature and rotation of the spine. The largest change from typical spine curvature occurs in the coronal plane.
- There are five types of scoliosis classified by cause: idiopathic, congenital, neuromuscular, syndromic, and other nonidiopathic scoliosis.
- Early-onset scoliosis (EOS) is scoliosis diagnosed prior to 10 years of age.
- The presence of scoliosis is verified by a spine specialist through X-ray images and a physical exam.
- The Cobb angle is the angle between the most tilted vertebrae at the upper and lower ends of a spinal curve, as measured with X-ray images. Scoliosis is diagnosed when the Cobb angle on the coronal view is 10 degrees or greater.

2.2

Types of vertebral anomalies

Every child is a different kind of flower that altogether make this world a beautiful garden.
Unknown

A method of classification of congenital scoliosis is by the type of vertebral anomaly:[8]

- **Failure of formation:** The vertebrae did not form properly and they have an atypical shape. Instead of appearing rectangular, these vertebrae appear triangular or trapezoidal on an X-ray.
- **Failure of segmentation:** The vertebrae did not separate during development and are now abnormally connected.[13]
- **Mixed anomalies:** There is a combination of failure of formation and failure of segmentation.[8]

Failure of formation

Recall from Chapter 1 that a typical vertebra has two pedicles: one on the left side and one on the right side (Figure 2.2.1). Pedicles are bony

bridges that connect the vertebral body (front of the vertebrae) to the lamina (back of the vertebrae). The two pedicles form the sides of the opening where the spinal cord passes through, also referred to as the spinal canal.

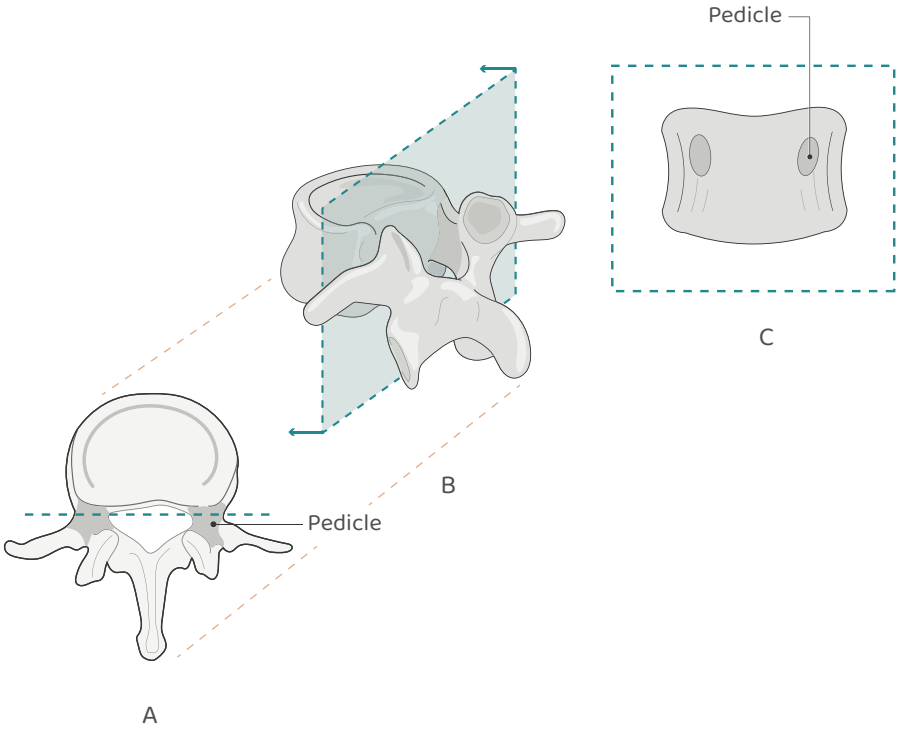


Figure 2.2.1 Vertebra showing pedicles. A: Top view. B: Side view. C: Back view: back bones removed showing the two pedicles.

Failure of formation results in vertebrae that are triangular or trapezoidal instead of rectangular This failure of formation can be incomplete or complete, depending on the number of pedicles in the atypical vertebra (Figure 2.2.2).

- **Wedge vertebrae:** Vertebrae with *incomplete* failure of formation (Figure 2.2.2 A). Both the left and right pedicles exist but one side is shorter than the other resulting in a wedge shape.
- **Hemivertebrae:** Vertebrae with *complete* failure (Figure 2.2.2 B to D). "Hemi" means half; only one pedicle exists. Hemivertebrae can be further classified by the presence or absence of intervertebral discs between adjacent vertebrae:[14]

- **Fully segmented hemivertebrae:** Intervertebral discs exist between adjacent vertebrae as indicated by the gap between adjacent vertebrae on X-ray.
- **Partially segmented hemivertebrae:** Intervertebral disc is absent between two adjacent vertebrae, which are fused to one another.
- **Unsegmented hemivertebrae:** Intervertebral discs are absent between more than two adjacent vertebrae, and the adjacent vertebrae are fused to one another on both sides.

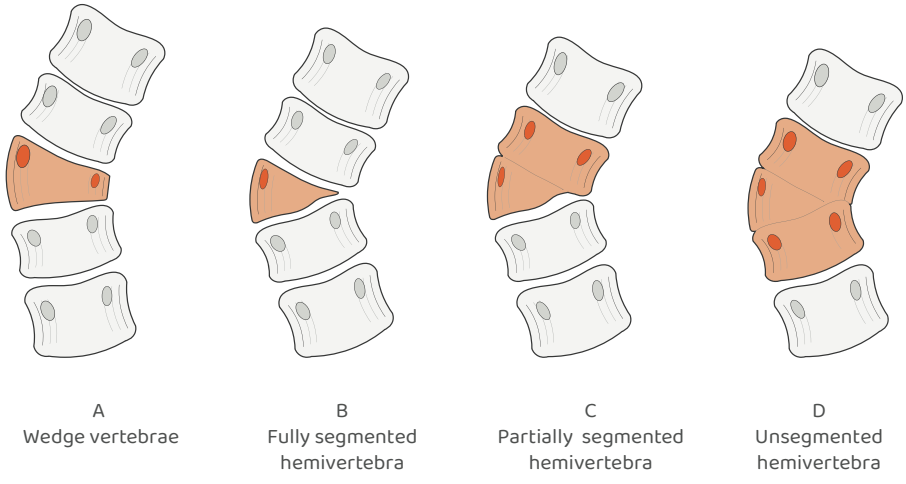


Figure 2.2.2 Failure of formation. A: Incomplete. B to D: Complete.

Failure of segmentation

Failure of segmentation can be bilateral (on both sides) or unilateral (on one side). Both are shown in Figure 2.2.3.

- Bilateral failure of segmentation results in a block vertebra.
- Unilateral failure of segmentation results in a bar connecting the vertebrae.

Key points Chapter 2

- Congenital scoliosis is characterized by errors in vertebral development.
- It is common for congenital scoliosis to be diagnosed during one of the major growth spurts of childhood.
- Congenital scoliosis accounts for 10 percent of scoliosis and occurs more frequently in females than males.
- Congenital scoliosis does not run in families.
- Congenital scoliosis can be classified as failure of formation, failure of segmentation, or a combination of both (mixed anomalies).
- When errors in vertebral development occur, there is an increased likelihood that errors also occur in the development of other organ systems.
- Anomalies in other organ systems associated with congenital scoliosis can occur in isolation or as part of a syndrome. Syndromes associated with congenital scoliosis include VACTERL association and Klippel-Feil syndrome.
- Prognosis and treatment for congenital scoliosis are largely related to the size of the curve and likelihood of progression.
- Growth potential and growth imbalance of the affected vertebra are what cause some congenital curves to progress more than others.
- If at risk for rapid progression, the scoliosis curve may be treated prophylactically (preventively).

Lisa, mother of Olivia, age 11:
PART 1

Olivia's journey started way back in the womb with her twin sister, Kempley. I had a twin-to-twin transfusion syndrome (TTTS) pregnancy, where Olivia was stealing all the nutrients from Kempley. The two were born one day shy of 27 weeks gestation. Kempley weighed only 11 oz (312 grams) and, sadly, she passed away four hours after birth.

Olivia weighed 1 lb 8 oz (680 grams) and had to fight for her life. It took the doctors three days to find a medication to stabilize her heart rate, and when they did, we got the good news that she was going to pull through. However, over those three days, Olivia lost bloodflow to all her organs, including her brain.

The ultrasound at six weeks old showed that Olivia was missing some white matter in her brain. My heart broke as the doctor tried to explain all the things that Olivia would not be able to do, including talking, walking, or any ability to function "normally." After Olivia was allowed to come home, I took the attitude that life was good. However, there were certainly still times I denied the diagnosis and what the doctors said, and I believed that Olivia was doing all the right things—all the things that a "normal" infant should do.

At age one, Olivia was officially diagnosed with having spastic quadriplegic cerebral palsy (where all four limbs are affected, and the primary movement disorder is spasticity). Even though I knew the diagnosis was coming, it still didn't make it easy to hear, but I vowed I would not let it determine Olivia's fate in life. Since that day, life has been a journey. I don't know how many "ologists" Olivia has, but we pretty much see all of them. Having a specialty health care facility in our state with amazing doctors and nurses has been a godsend. They have given me so many tools to help Olivia have the most mainstream life possible. That's not to say that Olivia hasn't faced obstacles in the road, but we have learned to take detours or pave new roads to get to where we need to go. We live life with a "go with the flow" outlook.

Since her diagnosis of cerebral palsy, Olivia has had five major surgeries in her 11 years of life, including a double hip realignment when she was just three-and-a-half-years old. At her six-month follow-up, her

orthopedic surgeon determined that even with the surgery, she required an intrathecal baclofen pump to help control tone and spasticity. This medicine was supposed to control tone in the lower extremities, but it also affected the tone in her torso, and as Olivia grew, so did her need for higher doses. I began to notice that Olivia was leaning more on her right side while sitting in her wheelchair, which required making seating adjustments and adding a neck rest to help her not lean as much.

When Olivia was about six, her rib cage started to look a little uneven, but the pulmonologist wasn't concerned because lungs grow with and work around the rib cage. With that reassurance, I didn't think much of it and didn't question why it was growing that way.

Olivia, age nine.

During winter 2019, Olivia had a significant growth spurt in her torso and a big curve of her spine was detected. After X-rays, she was officially diagnosed with scoliosis, with a 29 degree curvature.

The doctor was very good at explaining why Olivia's scoliosis occurred, as well as why the rib cage formed the way it did (because of the scoliosis). He explained that she would likely need to wear a brace to slow the progression until she was old enough to have a spinal fusion. This was scary stuff to hear.

> *Olivia's story continues in Chapter 9.*

Key points Chapter 3

- Neuromuscular scoliosis is a type of scoliosis that occurs secondary to (or as a result of) a primary neuromuscular condition, a condition that impacts the nervous and/or muscular systems.
- Scoliosis affects approximately 40 to 90 percent of individuals with neuromuscular conditions.
- The incidence of scoliosis and the size of the scoliosis curves increases with the severity of the neuromuscular condition.
- Neuromuscular scoliosis curves are often C-shaped.
- Neuromuscular conditions commonly associated with scoliosis include:
  - Cerebral palsy
  - Spinal muscular atrophy
  - Spina bifida
  - Duchenne muscular dystrophy
  - Arthrogryposis multiplex congenita
- Individuals with neuromuscular conditions often have global kyphosis in addition to their scoliosis.
- Neuromuscular scoliosis curves are more likely to progress in adulthood.

Key points Chapter 4

- Syndromic scoliosis is a type of scoliosis that occurs secondary to (or as a result of) a primary syndrome.
- A syndrome is a group of symptoms that consistently occur together.
- Syndromic scoliosis is often associated with syndromes that affect connective tissue (e.g., bone, blood, cartilage). This can result in the spine becoming curved and twisted as it grows.
- Syndromes commonly associated with scoliosis include:
  - Rett syndrome
  - Autism spectrum disorder
  - Neurofibromatosis
  - Down syndrome
  - Ehlers-Danlos syndrome
  - Prader-Willi syndrome
- Conditions arising from other chromosomal abnormalities do not always have an identified syndrome. Advances in medical science will likely lead to the identification of further syndromes in the future.

Key points Chapter 5

- Other types of nonidiopathic scoliosis exist that do not fit into the four established types (idiopathic, congenital, neuromuscular, and syndromic). "Other" includes scoliosis associated with neural axis abnormalities.
- Neural axis abnormalities are atypical structures within the central nervous system (brain and spinal cord) that can impact the growth of the spine.
- Neural axis abnormalities commonly associated with scoliosis include:
  - Syrinx: a fluid-filled area that forms in the spinal cord, caused by a condition termed syringomyelia, which is an atypical widening of the central canal in the spinal cord.
  - Chiari malformation type I: a condition in which brain tissue extends into the spinal canal through the opening in the base of the skull.
  - Tethered cord syndrome: a neurologic condition where connective tissue is attached to the spinal cord which "tethers" it, usually to the bottom of the spine, and inhibits the movement of the spinal cord within the spine.
- Research indicates that 24 percent of individuals with early-onset scoliosis (EOS—scoliosis diagnosed prior to 10 years of age) have a neural axis abnormality, with a syrinx or tethered cord being the two most common.

6.3

Treatment options

The aim of medicine is to prevent disease and prolong life;
the ideal of medicine is to eliminate the need of a physician.
William J. Mayo

Treatment options for scoliosis can range from nonsurgical methods, such as observation with repeat X-rays and clinical exams, to surgical methods, such as spinal fusion.

A summary of current, general treatment options and goals for nonidiopathic scoliosis at Gillette Children's is shown in Table 6.3.1 and illustrated in Figure 6.3.1. These treatment options may vary at other hospitals and treatment centers. It is also worth noting that clinical practice continually evolves.

It is important to note that for most individuals, scoliosis cannot be "cured" (i.e., achieving a Cobb angle less than 10 degrees) without surgery.

Table 6.3.1 General treatment options and goals for nonidiopathic scoliosis at Gillette Children's

TREATMENT OPTIONS	GOALS
Observation: Regular spine X-rays and clinical exams with a spine specialist	Monitor scoliosis curve for possible progression
Bracing: A removable spine brace that applies corrective forces to the spine	Slow scoliosis curve progression Prevent or delay surgery
Casting: A full torso cast (hardened plaster or fiberglass that must be cut off to remove) that applies corrective forces to the spine	Slow scoliosis curve progression Prevent or delay surgery
Surgery: Spinal fusion (the most common type of surgery), defined as fusing (joining together) two or more vertebrae; screws and metal rods are used to hold the spine in the straightened position and facilitate fusion between bones	Slow or stop scoliosis curve progression Improve the scoliosis curve (decrease the Cobb angle) Improve spinal balance

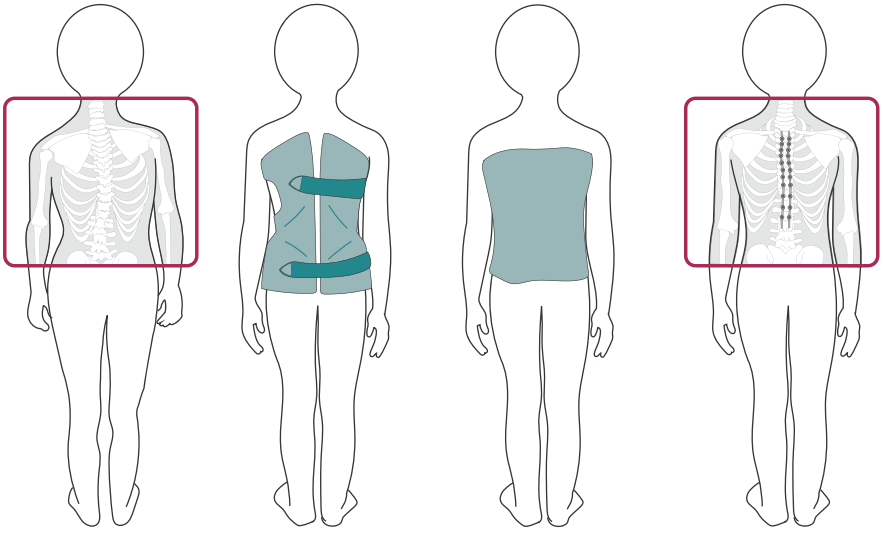


Figure 6.3.1 General treatment options for nonidiopathic scoliosis.

As the child grows, the scoliosis progression may slow or increase, resulting in treatment plans changing. It is common for an individual to undergo multiple treatment types over the course of their childhood and adolescence. For example, an individual may be observed for a period of time, then prescribed bracing treatment if their curve progresses.

Key points Chapter 6

- Treatment for nonidiopathic scoliosis focuses on slowing curve progression to keep the curve as small as possible as the child grows. The larger the curve, the greater the likelihood of continued curve progression and for it to have a negative impact on the individual's health and quality of life.
- Treatment is often dictated by the estimated rate of growth and remaining growth of the patient. Skeletal maturity is currently the most accurate means of assessing these.
- Treatment for nonidiopathic scoliosis is to prevent curvature progression and address the possible resulting complications:
  - Cardiopulmonary problems
  - Pelvic obliquity
  - Back pain
- Treatment options include:
  - Observation
  - Bracing
  - Casting
  - Surgery

Key points Chapter 7

- The overall goal of nonsurgical treatment is to slow curve progression and/or to prevent or delay surgery.
- Observation for scoliosis requires regular X-rays and clinical exams with a spine specialist.
- Candidates for observation are deemed to have a lower risk of rapid scoliosis curve progression; thus, treatments such as bracing or surgery may not be necessary for their scoliosis, or at least not right away.
- Wearing a spinal orthosis, also referred to as a brace, is the most common nonsurgical treatment for progressive scoliosis curves.
- The most common type of spinal brace is the thoraco-lumbo-sacral orthosis (TLSO).
- There are two types of TLSOs: a full-time TLSO is prescribed to be worn for 18 to 23 hours per day; a nighttime hypercorrective TLSO is prescribed to be worn for 8 to 12 hours per day (i.e., worn at night only.)
- Cast treatment involves the application of plaster to the torso, molded to the child's body.
- Physical therapy may occur before, during, or after other scoliosis treatment and is considered an add-on or complement to other scoliosis treatment.

Table 8.1.1 Scoliosis surgeries

SURGERY	DESCRIPTION
Spinal fusion	Fusing (joining together) two or more vertebrae in the spine to stop curve progression and improve the angle of the curve (decrease the Cobb angle), typically performed using metal rods and screws or hooks to hold the spine in a corrected position and facilitate fusion between bones.
Growth-friendly treatment	Improving the angle of the scoliosis curve (decreasing the Cobb angle) by using metal rods and screws or hooks to stabilize the spine above and below the curve. The length of the metal rods can be expanded over time. This includes traditional growing rods and magnetically controlled growing rods (MCGRs).

This chapter also discusses halo gravity traction, which is a multistage treatment for severe spine curvatures to stretch and straighten the spine and soft tissues (skin, muscles, ligaments) *prior* to scoliosis surgery (see section 8.6).

Scoliosis surgeries can be safely conducted at appropriate facilities. Spine specialists recommend a surgery only if they believe the benefits outweigh the potential risks for the individual. There are risks with any surgery: surgical risks are things that may go wrong during or shortly after surgery while the individual is still in the hospital (in-hospital complications). Long-term complications are problems that may occur after the individual has been discharged from the hospital and may develop over the course of months or even years following surgery.

Risk management is the highest priority for hospitals and spine specialists, and many safety measures are in place to maximize safety. However, it is important for families to be aware of these possibilities and discuss them as part of the shared decision-making process.

USEFUL WEB RESOURCES

Key points Chapter 8

- The overall goals of scoliosis surgery for individuals with nonidiopathic scoliosis are to slow or stop curve progression, improve the scoliosis curve (decrease the Cobb angle), and improve spinal balance.
- Spinal fusion is the most common type of scoliosis surgery. It involves fusing (joining together) two or more vertebrae in the spine to stop curve progression and improve the angle of the curve (decrease the Cobb angle), typically performed using metal rods and screws or hooks to hold the spine in a corrected position and facilitate fusion between bones.
- Growth-friendly treatment involves using metal rods and screws or hooks to stabilize the spine above and below the curve. The length of the metal rods can be expanded over time. This includes traditional growing rods and magnetically controlled growing rods (MCGRs):
  - Traditional growing rods are manually lengthened during surgical procedures under general anesthesia.
  - MCGRs are lengthened with a magnetic remote control during routine clinic visits.
- Halo gravity traction is a multistage treatment for severe scoliosis to stretch and straighten the spine and soft tissues (skin, muscles, ligaments) prior to scoliosis surgery.

Breanna, mother of Emersynn, age six:
PART 2

Emersynn's surgery was planned for the September she was to start kindergarten. During the pre-op, her anesthesiologist, who happened to be the same anesthesiologist the girls had at three months old for an MRI, came in to meet her. It was a very special reunion!

Emersynn had an L2 hemivertebrectomy (removing the hemivertebra at the L2 level) and a spinal fusion of L2 and L3. The surgery lasted about three hours, and Blake, Ellerie, and I anxiously awaited news in the waiting room while Grandma stayed home to care for the girls' little brother, Ezra, who was two at the time. Finally, the surgeon came to tell us that it went very well and we could see her very soon. Emersynn did great through it all and was happy to see us, and she let us know she was very hungry! By that night, she was sitting up and she even took a few steps. We were surprised that we could see an immediate difference—just hours after surgery, she was already standing a little bit straighter. That progress made us feel like her road to recovery could begin and the light was already shining.

During surgery, Emersynn had a custom brace molded to fit her perfectly. She was able to get the brace on the day after surgery and spend some time getting used to it. It helped that she got to pick the design on the brace, so she was very excited to have it. Aside from occasionally getting a little sweaty, she adjusted quickly to her brace. It helped us, too, knowing that the brace kept her safe and secure in slightly chaotic situations, like a kindergarten classroom: if she happened to get bumped, the brace gave her a layer of protection.

Emersynn chose to wear her brace under her clothes with a barrier layer (a thin T-shirt) between her skin and brace. With the surgery being done in the fall, she didn't have many complaints of being too warm! Her biggest challenge was remembering not to jump, run, or play fast. She felt well enough to do all those things, but her bones needed time to heal. This did take some getting used to, especially when playing with her siblings. Having to sit out of certain activities like recess and gym class was difficult, and because it was a very mild fall season, fun things like scootering, bike riding, and playground time had to end a little earlier for Emersynn than she would have liked.

Friends and family knew to be careful with Emersynn's back, as did the staff and her peers at school. She was missed during her two weeks off school, but her teacher sent home worksheets and her classmates made her many cards of well wishes, and that made things much easier. While it was a challenge for Emersynn to take it easy after surgery, she did it and it paid off, as she was ready to have the brace removed the day before Christmas, just three months later. While she hadn't complained much about wearing her brace, and she had enjoyed her brace breaks, she was especially excited to have it off!

Once her brace was removed, some of the screws that were placed during surgery were prominent and we could see the outline of these under her skin. We weren't expecting to have those poke out so much, but Emersynn is a very petite girl, with a small frame, which likely contributed to this. As of the time of writing, four months post-op, we are keeping a close eye on them.

Emersynn still walks slightly leaning to one side. We are hopeful that with physical therapy she will continue to learn to walk straighter so there is no additional pressure to her back. Because she has learned to do everything while leaning a bit, it seems to be difficult for her to break the habit.

Today, Emersynn is thriving. She has overcome some difficult physical setbacks, starting from the very beginning being joined to her sister for the first nine months of life. She has always had the determination, even as an infant, to handle just about anything life throws her way. Whatever she puts her mind to achieving, it happens.

Emersynn with her 3D spine model.

Maria, mother of Abby, age 15:
PART 2

Unfortunately, even with our vigilance and weekly physical therapy intervention, Abby's scoliosis rapidly progressed. Also, when she was around 18 months old, she began experiencing recurrent pneumonia due to the scoliosis, which caused restrictive airway disease. We introduced a more aggressive approach to attempt to slow the progression of the scoliosis, and she was fitted for her first TLSO around age two. We chose an adorable butterfly print that matched her AFOs and tried to make the best out of the fact that she would be wearing it all the time, except when sleeping.

I am not going to sugarcoat it: with Abby's global minimal muscle tone and sensitivity to the feeling of the brace, the TLSO was extremely uncomfortable for her tiny body. It was a struggle emotionally and physically for us both, but we stuck to the plan and remained as consistent as possible to try to slow the monster that her scoliosis had become in our daily routine; it touched every aspect of her life. It caused hospitalizations, pain, fear, and great frustration.

As a parent, I found it difficult to constantly weigh the benefits versus the rewards of continuous bracing, especially not knowing what Abby's future would hold regarding her spine. It was a dance we tried to perform with as much grace and strength as we could muster. It was simply a hard reality on Abby's journey and something we expected to be a part of her life forever due to the severity and aggressive nature of her scoliosis.

Abby wearing her butterfly-print TLSO.

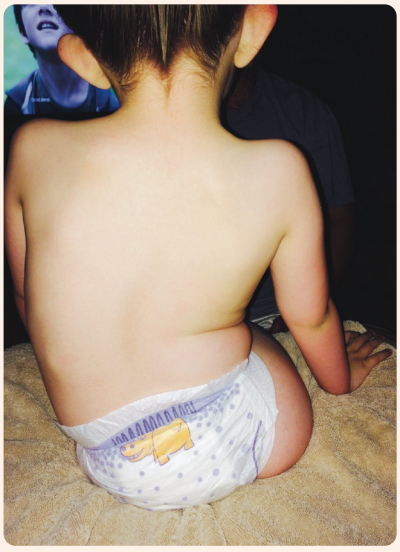
I was consumed with fears and worries for Abby. I experienced guilt that I wasn't being vigilant enough to keep her as healthy as possible and sadness that her body could never be free of the continuous bracing and pain. Her scoliosis constantly weighed heavily on my heart as her mother. Then, a bright ray of hope entered our lives! I read about a new procedure for scoliosis management using MCGRs. I promptly made an appointment at Gillette Children's where we found positivity, sincere care for Abby's condition, and expertise that gave us confidence, comfort, and most importantly, hope for Abby's future. I recall leaving that initial meeting and telling my husband I was confident everything would be okay, as we were in the best hands possible. I finally felt as though the responsibility to keep Abby's body straight and healthy no longer landed squarely on my shoulders alone. The feeling was priceless. That day was a true turning point.

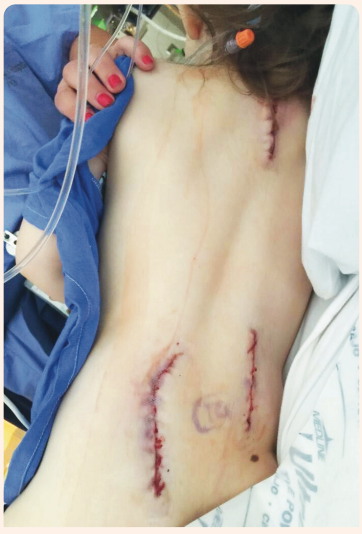
In September 2015, when she was five, Abby became one of the youngest patients at the time to have the MCGRs implanted. After having witnessed years of Abby's repeated respiratory difficulties, I firmly believe that the procedure saved her life. The rods allowed her to no longer wear any scoliosis brace, freeing her body of the weight and pain that carried. We were elated to say goodbye to all the bracing! The S-curve in her spine measured 85 degrees left lumbar curvature and 46 degrees right thoracic curvature before the rods were placed. After placement, they measured 29 degrees and 27 degrees respectively. Her body lengthened about 4 inches (10 cm) with the surgery, and her wheelchairs and all equipment had to be adjusted to accommodate her growth.

Her lungs opened and her breathing strengthened immensely, and the regular bouts of pneumonia became few and far between. It was awe inspiring, to say the least, to witness her immediate global improvement. As well, Abby's recovery from the surgery went amazingly well with no complications.

Our focus shifted to maintaining her growth. This was done first by lengthening the rods every three months to accommodate and encourage her growth externally, and then with a follow-up placement of a second set of rods in May 2018 when her first rods reached their maximum length.

The lengthening of the rods after the first surgery was done with embedded magnets and a machine placed on her back, unlike traditional rods that require internal lengthening and repeat incisions. The timing of this invention was miraculous for Abby. My intuition told me that with traditional rods she would have had a much harder road and possible serious complications with constant surgical procedures.





Abby's back before surgery (left) and immediately after surgery (right).

The second surgery (to place a new set of rods) also went well and, once again, recovery was smooth and without complications. With the regular visits to Gillette over the years, Abby developed a beautiful relationship with the entire spine care team. She thrived under their care.

Abby's latest surgery, and hopefully her last for her scoliosis, was in October 2022. It was a full spinal fusion, which improved her curvature measures to 21 degrees left lumbar and 16 degrees right thoracic. Together, the MCGRs and spinal fusion were an overall success!

Unfortunately, a couple of weeks after the fusion surgery, Abby experienced a rare complication, with a 2-inch (5 cm) section of her incision on the lower part of her spine opening. This required a second surgery to close the wound, but two days after returning home from that, the incision reopened. That resulted in a change of plans, and wound care became a huge part of Abby's daily life for the next several months.

Abby after her spinal fusion.

The relationship we developed with Abby's spine team became exponentially more important over the years. I was in constant contact with them by email and phone with photo updates of the wound, questions on changing protocols to best care for the wound, and strategies to assist Abby's comfort levels as her body healed. Abby and I were not only patient and parent, we were also active participants of her spine team and guides in her recovery. This relationship was pivotal for both Abby's healing and my strength as her primary caregiver. A scary, overwhelming period for our family was met with the utmost respect, positivity, open communication, and support.

Abby's scoliosis story will continue for the rest of her life. We will always feel a deep sense of gratitude to have found such skilled doctors and nurses to be on the journey with her. It is priceless in our special-needs world and lifts our hope to continue to shine brightly for her future.

Tethered cord syndrome

- **Detethering:** Surgical treatment of a tethered spinal cord is generally recommended when the tethered cord is causing symptoms (known as tethered cord syndrome). The procedure is called "detethering," "untethering," or "tethered cord release." The goal of detethering is to relieve tension of the spinal cord and promote increased spinal cord movement. Individuals with tethered cord syndrome and a Cobb angle of less than 35 degrees may experience improvement in their scoliosis curve progression after detethering surgery, decreasing the need for spinal fusion. Individuals with tethered cord syndrome and a Cobb angle greater than 35 degrees are likely to have curve progression despite detethering and often require spinal fusion (which may occur at the same time as or after detethering surgery).[164] A recent small research study found that detethering in young individuals was associated with a lower rate of scoliosis progression than detethering in older individuals.[165]

Jody, mother of Henley, age 12:
PART 2

When Henley was three years old in June 2015, we left five-month-old Eastyn with my parents so we could be with Henley for her tethered cord surgery, which was successful. She stayed in the ICU for two days before moving to the regular floor for three more days, and then we went home. She wore a onesie to keep the incision clean while it healed, and all summer she was unable to get dirty playing outside as she wasn't allowed to have a bath. Giving her sponge baths and washing her hair in the kitchen sink became our normal. The surgery resulted in us waiting until she was about three and a half before potty training her as we knew that would be more difficult while she was recovering and healing from this surgery.

A year after her tethered cord release, Henley started casting for scoliosis every three months. Each time, she had to be put under anesthesia, which for a parent is hard to watch, but the doctors helped us feel as comfortable as possible. Originally, the plan was to continue the casting for a full year. However, we ended up doing it for only about nine months because Henley frequently came down with different illnesses

that interrupted the planned schedule. To minimize the risks associated with anesthesia, casting was delayed as needed until she recovered. As it was with the surgery recovery, baths were not allowed with the casts, so we continued with sponge baths and washing her hair in the sink.

Once Henley turned five, she was able to start wearing a TLSO. She wore this brace 20 to 22 hours a day and had it adjusted every six months (or sometimes more frequently). This is all Henley knew as a young child. She was compliant and happy to choose a new brace design every year and a half or so.

Henley, age five, wearing her TLSO.

When she was 11, Henley qualified for a nighttime hypercorrective brace, which corrects more than a daytime TLSO but doesn't need to be worn for as many hours. She loves that it allows her to go to school brace-free! Now almost 13 years old, she is doing well, and she still goes for checkups every six months at Gillette Children's.

There have been some hard times over the years, especially with the new diagnoses that seemed to happen each time Henley got hurt or sick. She still has four curves in her spine, but all have decreased in severity due to the various treatments that have been done—the tethered cord release, casting, and bracing. We have been on an emotional roller-coaster through it all, but Henley's resilience and determination to do everything to improve her scoliosis and do what she loves have helped us all. Henley is a competitive dancer and takes seven dance classes a week. This is her ninth year dancing and she enjoys every minute of it. We are so proud of her!

If you were to ask Henley about her journey, she would say that there were definitely times she felt alone and different from her peers. When she was younger (around five years old), she would show her brace to her classmates at the beginning of the school year and tell them about it. And there were and continue to be some physical limitations that set her apart, like not being able to participate in trampoline or gymnastic activities. Henley also has anaphylactic allergies and at times feels like she has too many things going on, but by talking about them and processing it all, she is able to understand how using her voice can help others going through the same thing. She is kind, empathetic, and determined to do her best in all areas of her life.

Henley after a dance competition, age 13.

Key points Chapter 9

- General treatment options for nonidiopathic scoliosis at Gillette Children's include observation, bracing, casting, and surgery.
- While these treatment options apply to all types of nonidiopathic scoliosis, each type presents unique considerations and challenges that should be carefully evaluated.

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"I highly recommend this impactful book for families and professionals working in the field of scoliosis."

—ANN MARIE SUTTON, PARENT

"This book addresses a complex topic in a thorough but accessible way."

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Approximately 20 percent of all scoliosis cases have a *known* cause. This book explains the condition and includes both the current best-practice treatments and the lived experience of families. It is a companion book to *Idiopathic Scoliosis*, which addresses scoliosis where the cause is *unknown*.

The writing of this scoliosis book was led by Tenner J. Guillaume, MD, Walter H. Truong, MD, and Danielle Harding, PA-C, spine specialists at Gillette Children's, a world-renowned center of excellence for the treatment of brain, bone, and movement conditions. This book is part of the Gillette Children's Healthcare Series, a series of books for families who are looking for clear, comprehensive information. Health care professionals, researchers, educators, students, and extended family members will also benefit from reading it.

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