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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

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GILLETTE CHILDREN'S

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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.

Scoliosis is characterized by rotation (twisting) in the axial plane and sideways curvature in the coronal plane (Figure 1.3.3). While the largest change from typical spine curvature occurs in the coronal plane, scoliosis is described as three dimensional because it affects the spine in all three anatomical planes.

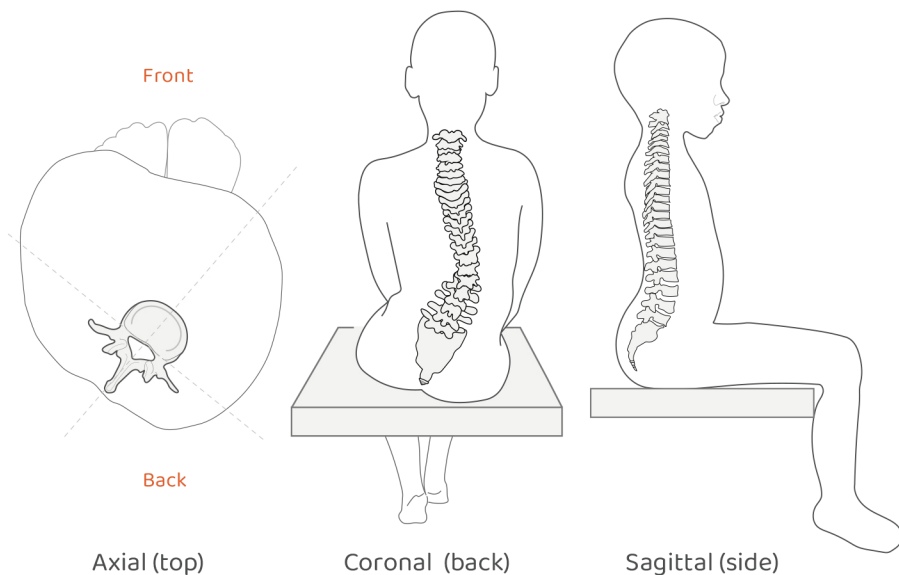


Figure 1.3.3 Spinal curvature associated with scoliosis in the axial, coronal, and sagittal planes.

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).

Chad and Becca, parents of Claire, age 13:

PART 1

Our daughter, Claire, is 13 and in the eighth grade. She is an extremely positive person, radiating optimism, happiness, and an excitement for life. She is the younger of our two children; our other daughter, Grace, is four years older.

Claire was born with spina bifida with a lesion located at L3–L4, and she is paralyzed from the waist down. When she was born, she had significant stridor (high-pitched, wheezing sound when breathing) due to vocal cord paralysis, and we were told she would need a tracheostomy.* During the surgery to place the tracheostomy, she also had a gastrostomy tube† put in since there was concern about aspiration‡ due to her airway issues.

At five days old, Claire had a Chiari decompression to address her Chiari malformation type II, a spinal cord detethering, and a lumbar shunt placed to drain a syrinx in her spinal cord. Finally, when she was a few weeks old, she had a VP shunt placed to address her hydrocephalus.

Claire has neuromuscular scoliosis, which was evident early on, but it didn't require intervention at first. When she was about eight years old, she hit a growth spurt, and it became apparent that surgery would be required. Some people might have the option to wait and see, but we knew that Claire's lung and heart function could be compromised, so we felt we needed to move past any conservative routes and have surgery to correct it. In a way, that made the decision to go ahead with surgery easy for us.

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

* A surgical procedure that creates an opening in the trachea (windpipe) to insert a tube, allowing direct airflow to the lungs when normal breathing is obstructed or impaired.

† A tube surgically inserted to carry nutrition to the stomach.

‡ Food or liquid entering the airway or lungs instead of the esophagus.

"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."

—JOHN S. VORHIES, MD, FAAOS, FAOA

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.

Other titles in the series include:

- *Craniosynostosis*
- *Idiopathic Scoliosis*
- *Spastic Hemiplegia—Unilateral Cerebral Palsy*
- *Spastic Quadriplegia—Bilateral Cerebral Palsy*
- *Spastic Diplegia—Bilateral Cerebral Palsy, second edition*
- *Epilepsy*
- *Spina Bifida*
- *Osteogenesis Imperfecta*

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