

Scoliosis in WAGR Spectrum

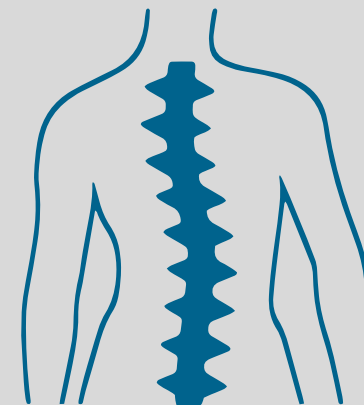
*Scoliosis occurs in about 20% of people with WAGR Spectrum**

Scoliosis is a condition where the spine is not straight, but curves sideways into a “c” or “S” shape.

Diagnosis Doctors can find scoliosis by looking at the back or through an X-ray

Treatment Depends on how curved the spine is and how fast it is getting worse

- **Observation** if the curve is small, the doctor may just watch it over time to see if it changes
- **Bracing** if the curve is getting worse and the child is still growing, wearing a brace can help stop the curve from worsening. A brace doesn't make the curve go away but can help it from getting worse
- **Surgery** if the curve is large or keeps getting worse, surgery may be needed to straighten the spine and stop the curve from worsening



Special Considerations

- A specialized healthcare team is needed for individuals with behavioral or sensory challenges who require surgery and/or bracing. This team can help with preparation, anxiety, and recovery.
- **Propofol**, a medication often used in general anesthesia, may increase the risk of acute pancreatitis in WAGR patients who also have elevated triglycerides. ** Discuss this risk with your child's doctor before any surgery

By working together, families and doctors can create a treatment plan that meets the child's needs, keeps them safe, and helps manage scoliosis effectively

Scoliosis in WAGR Spectrum: Information for Pediatricians

Higher risk Children with WAGR spectrum have a 20% risk of developing scoliosis vs 2-3% in the typical population

Early and Frequent Monitoring Regular screening should begin early. Include spine checks as part of routine physical exams, especially during periods of rapid growth. If scoliosis is suspected, timely referral to a specialist is important to manage the condition

Co-Occurring Conditions Abnormal muscle tone (high or low) is common in WAGR spectrum, which can affect when and how scoliosis develops, and how it is treated

Communication and Behavioral Considerations Children with WAGR spectrum may have developmental delays, communication challenges, or behavioral differences--making it difficult to express discomfort, understand instructions, or cooperate with treatment. Work closely with the family to meet the child's individual needs, such as providing extra support for wearing a brace, preparing for surgery, and coping with post-surgical recovery

Long-Term Management Scoliosis in children with WAGR spectrum may require long-term management, with regular check-ups even after growth has stopped. Pediatricians can help parents coordinate ongoing care and ensure that complications or progression are addressed promptly

It is advisable to monitor amylase, lipase, and triglycerides in patients with WAGR spectrum, and to avoid the administration of propofol in the setting of baseline abnormalities in these laboratory values.

*Duffy KA, Trout KL, Gunckle JM, Krantz SM, Morris J, Kalish JM. Results From the WAGR Syndrome Patient Registry: Characterization of WAGR Spectrum and Recommendations for Care Management. *Front Pediatr.* 2021 Dec 14;9:733018. doi: 10.3389/fped.2021.733018. PMID: 34970513; PMCID: PMC8712693.

**Acute Pancreatitis after Propofol Administration in a Child with WAGR Syndrome. Danley KM, Henderson WA, Ibrahim T, Hadigan CM, Han JC. North American Society for Pediatric Gastroenterology, Hepatology and Nutrition Annual Meeting, National Harbor, MD, November 2009. *Unpublished findings presented as preliminary data in abstract format at a medical conference.*