Scoliosis in patients with Prader-Willi Syndrome

Ralf Stücker

Altonaer Kinderkrankenhaus
University Clinic Hamburg
Orthopedic problems in PWS (West and Ballock, 2004)

- 41% Scoliosis
- 47% Flat foot
- 19% Genu valgum (Knock knee deformity)
- 10% Hip dysplasia
- 9% Osteoporosis
- 7% Patella instability
- 3% Bowlegs
- 2% Clubfoot
- 0.2% Slipped capital femoral epiphysis
Incidence of spinal deformities

- **Scoliosis** 15-86 %
  Nakamura et al, 2015

- **Kyphosis** 8-40 %
  Associated with higher BMI
PWS and scoliosis

- The overall risk of developing scoliosis before maturity is 60-70%
- 23% develop scoliosis before age 4 (infantile scoliosis)
  - related to profound hypotonia
- 15% of PWS patients will require surgery for scoliosis
- Complication rate after surgery up to 50%
- Prevalence of scoliosis seems to be unaffected by genotype, GH-treatment, age, sex or BMI (Shim et al, 2010)
Scoliosis in patients with Prader Willi-Syndrome
Odent et al, Pediatrics 2008

- Retrospective study: 145 children
- Prevalence scoliosis
  - 43,4 % at age 10,2
  - 66,7 % at maturity
Treatment strategies for scoliosis

- **Age < 5**
  - Casting
  - Bracing

- **Age 5-10**
  - Bracing
  - Growing Rods, if bracing is not able to halt progression

- **Age > 10**
  - Spine fusion
  - Vertebral body tethering
Most effective scoliosis treatment for children < 5 years

Casting for very young children (< 5 years)
2,5 years old boy, result after 3 casts

At least a very efficient time buying strategy
37 patients (34 patients with > 2 years f/u)
- All received growth hormone treatment
- Average age at casting: 32 months

- 12x cured
- 18x brace
- 4x surgery

Those with curves < 50° and age < 3 years have best prognosis
Clinical examination
Adam’s forward bend -test
Scoliosis in patients with PWS

Diagnosis can be difficult in case of obesity
Brace treatment not as effective as in idiopathic scoliosis

Brace treatment less effective in obese patients
Growth friendly surgery and serial cast correction in the treatment of early-onset scoliosis for patients with Prader-Willi-Syndrome

Oore et al, JPO 2019

- GFS: n=13 (VEPTR and TGR)
- Mean age 5,8 years
- Mean scoliosis: 76° corrected to 42° (45% correction)
- Mean Kyphosis: 59° corrected to 54° (10% correction)
- 2.2 complications per patient after surgery
- 11 of 13 patients (85%) had complications
9 years old boy with scoliosis, treated by growing rods
Magnetically controlled Growing Rods
5 year old girl with PWS, scoliosis 64°
Treated with magnetically controlled growing rods
Scoliosis > 10 years

- Brace (20-50 deg)
- Correction without fusion
  (VBT= vertebral body tethering)
- Final fusion (for curves > 50 degrees)
16 year old boy with PWS-scoliosis

Bending films
2 years after spine fusion
16 patients (3 males, 13 females)

Age at surgery: 12,3 years

9 major complications

- 4x proximal junktional kyphosis
- 2x deep infection
- 1 transient paraplegia
- 1 delayed wound healing
Hospital outcomes of scoliosis surgery in children with Prader-Willi-Syndrome: comparison with adolescent idiopathic scoliosis
McQuivey et al, Spine Deform 2021

- No difference between the incidence of major complications in PWS patients compared to AIS patients (1.7% vs 1.0%)

- Incidence of postoperative pneumonia and implant complications higher in PWS than in AIS patients
10+6 year old female with Prader-Willi-Syndrome
Treated with magnetically controlled growing rods
Development of proximal junctional kyphosis (PJK) after correction of significant kyphosis in a patient with PWS

T2-L4

4 months postop
1 year F/U, PJK was observed

1 year postop

Fused to T1 at age 12+5, 2 years after index surgery
But this is not the end of the story

4 months later

Fusion to C4
Developed instability C3/4 with neurologic signs
11+6 year old girl, brace not tolerated

Preop – 35°  Bending 17°  6 months 13°  12 months 7°
VBT
(Vertebral body tethering)

Growth Modulation with Convex Tether
Effects of 8 years of growth hormone treatment on scoliosis in children with Prader-Willi syndrome
Grootjen et al, Eur J Endocrinol 2021

- Median age: 10.8 years
- Prevalence of scoliosis was 77.7% - no difference to untreated children
- No difference in severity of scoliosis compared to untreated children
- Higher bone mineral density was associated with lower Cobb angles
Randomized controlled trial to investigate the effects of growth hormone treatment on scoliosis in children with Prader-Willi-syndrome

Van Wijngaarden et al, J Clin Endocrinol Metab 2009

- 91 children with PWS
- Median age: 4.7 years
- Similar onset of scoliosis in both groups
- No increase progression in GH treated patients

Conclusion: Scoliosis should no longer be considered a contraindication for GH treatment in children with PWS
Recommendations - Summary

- Yearly spine exams
- X-ray in case of obesity
- Casting very effective for onset of scoliosis < 5 years
- Growth friendly surgery recommended for scoliosis > 50° and > 5 years
- Halo traction before surgery when scoliosis > n70° and significant kyphosis
- Currently no experience with PWS scoliosis and VBT
- Spine fusion at maturity is not associated with increased risks compared to idiopathic scoliosis
Thank you