





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

ALTONAER KINDERKRANKENHAUS Incidence of spinal deformities 3

Scoliosis 15-86 % Nakamura et al. 2015

Nakamura et al, 2015

Kyphosis 8-40 %

Associated with higher BMI





- 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











Growing Rods, if bracing is not able to halt progression

Age > 10



Vertebral body tethering



Most effective scoliosis treatment for children < 5 years



Casting for very young children (< 5 years)

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At least a very efficient time buying strategy









ALTONAER KINDERKRANKENHAUS Casting for PWS scoliosis (Bosse et al, JPO 2021)

- 37 patients (34 patients with > 2 years f/u)
- All received growth hormone treatment
- Average age at casting: 32 months



Those with curves $< 50^{\circ}$ 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 scooiosis



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
- \succ 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







Magnetically controlled Growing Rods







5 year old girl with PWS, scoliosis 64° Treated with magnetically controlled growing rods

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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







22

- 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-Synndrome: 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 ²⁴





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

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Fused to T1 at age 12+5, 2 years after index surgery

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stehend FFA 3 m

ALTONAER But this is not the end of the story



4 months later

Fusion to C4

27



Developed instability C3/4 with neurologic signs







11+6 year old girl, brace not tolerated



 $6 \text{ months } 13^\circ$

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 og 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



- > Yearly spine exams
- X-ray in case of obesity
- Casting very effective for onset of scoliosis < 5 years</p>
- 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 assiciated with inncreased reisks compared to idiopathic scoliosis















Thank you







