

The care of infant with Prader-Willi-Syndrome

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Overview

- Situation of infant with PWS
- Holistic treatment concept
- Nutrition
- Physiotherapy
- Early intervention
- Endocrine aspects
- Orthopedic problems
- Additional health issues

Situation of infant with PWS

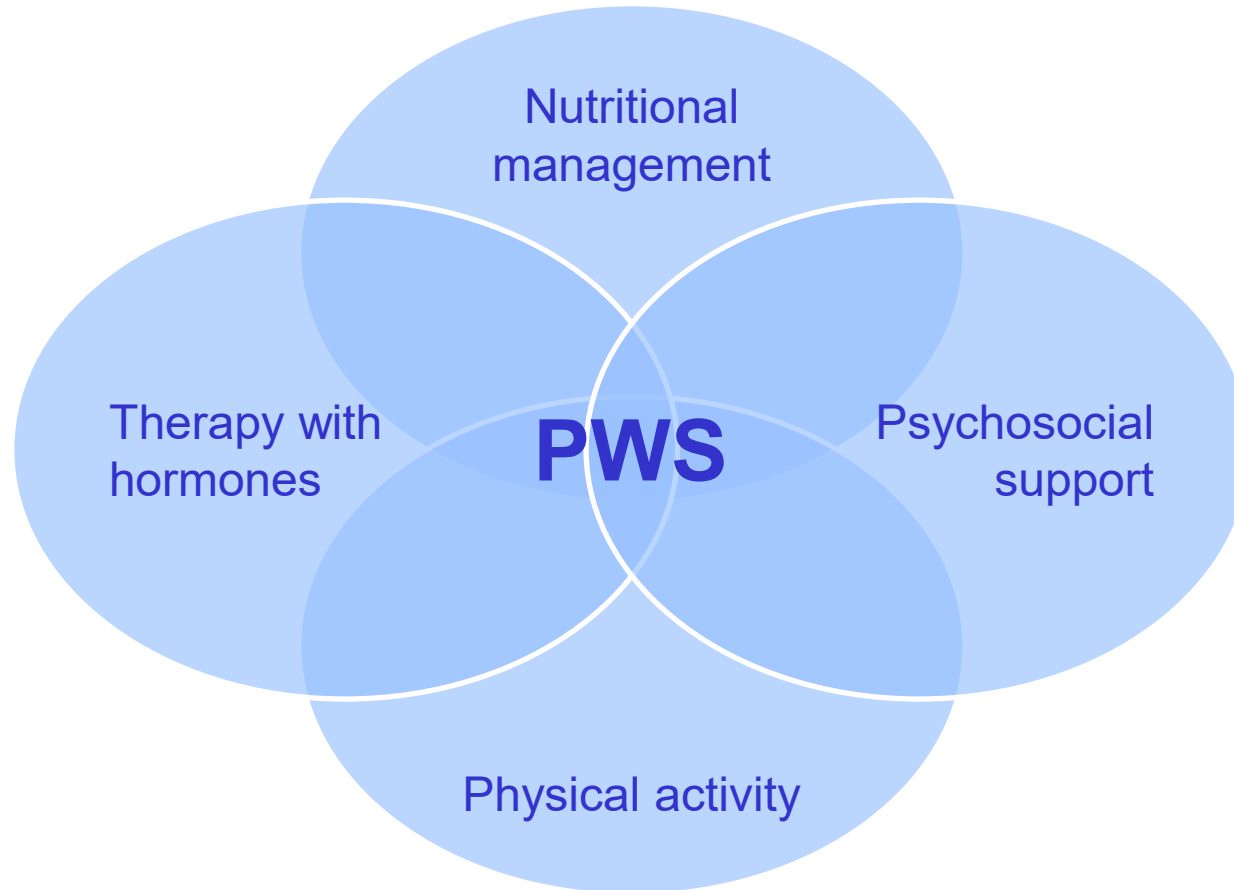
Situation:

- Breech presentation, low birth weight,
- Premature delivery
- Adoption problems after delivery
- Breathing problems
- Feeding difficulties, underweight
- Distinctive muscle hypotonia, Hypokinesia
- Failure to thrive

Support:

- Breathing stimulation and aid (CPAP, BIPAP)
- Secure sleep position, Monitoring if available
- Intensive physiotherapy
- Nutrition training
- Tube feeding
- Assistive Devices
- Supportive measures
- Occupational therapy

Holistic PWS-treatment concept

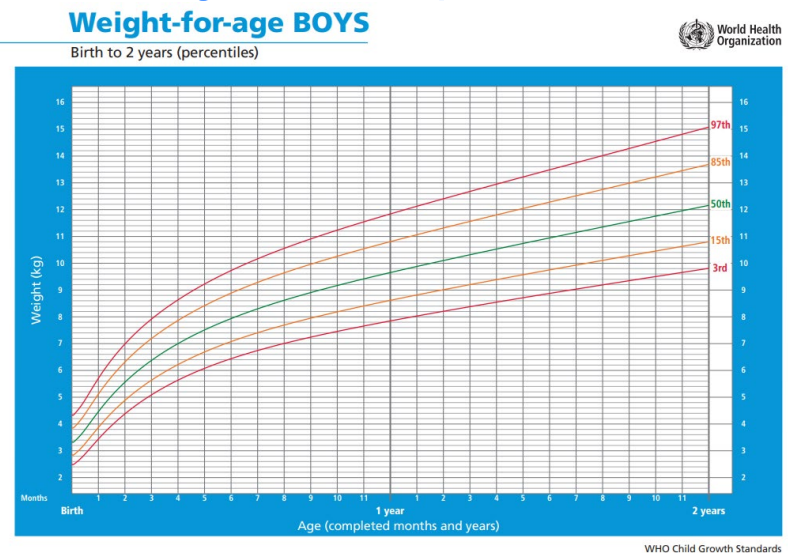


Nutrition starts with physiotherapy

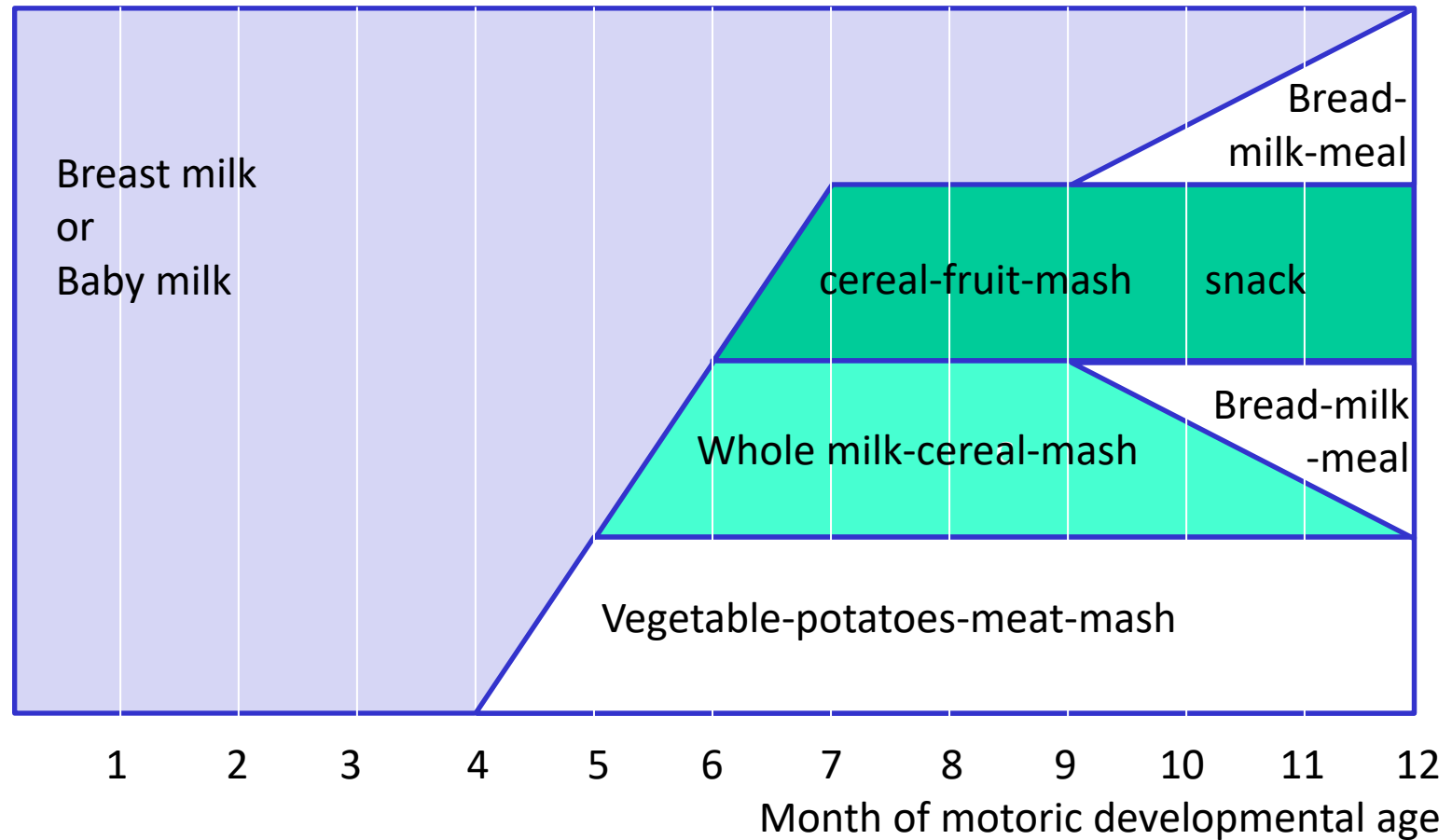
- Muscles are not ill in PWS, but muscle mass is smaller
- increased muscle tone means increase muscles by physiotherapy
- Special feeding training for example Castillo Morales
- Stimulate the baby and take a special feeding position
- Use special baby bottle teats
- Sometimes tube feeding is necessary

Practical aspects of nutrition

- PWS infant needs 2/3 of energy intake compared with a healthy infant of the same age
- Wake for feedings every 4 hour during the first month
- Give weight chart to the parents for self control
- Target: reach 3.-10. percentile in the weight chart
- Weigh every week and report in growth chart
- Control energy intake in accordance to weight development
- At the end of the first year feeding becomes easier and the infant grows appropriately



From milk to infant food



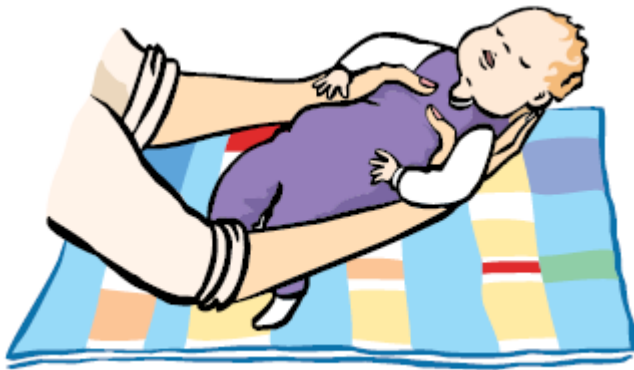
Educate family in nutrition

Target: Taking part at the family meals when able to sit at the table, maybe use a special baby seat

- Start with mashed meals when infant can control head in upright posture
- Add enough unsaturated fatty acids (rape oil)
- Be careful if children have dysphagia
- Chewing is a good training for speaking
- Give more and more solid food for learning to chew and moving the tongue (supervise chewing)
- Give piece of bread instead of cookies
- No sweet drinks
- Conditioning taste

Physiotherapy

- Voita therapy based on neurophysiological principles
- Bobath concept to support movement development
- Speech therapy - Castillo Morales



Early intervention for Infant with PWS

- First: Love your child
- Spend time with the baby
- Turning from left to right when changing nappies
- Playing in prone position when awake
- Baby massage
- Kangaroo – body contact
- Sing, speak to the baby
- Look for eye contact
- Stimulate with music, colors, lights
- Be sensitive to small changes
- Baby swim

Endocrine aspects I

Cryptorchidism

- Therapy: Operation at the end of first year
- Potentially preparation with GnRH-nose spray 3 x 2 intakes for 4 weeks or β -hCG i.m. 3 x 1500 IE
- Anaesthesia: www.orphanetanaesthesia.eu

Labial synechia

- Estrogen containing Salve locally approx. 2 weeks, repetition if necessary
- Cave: possible side effect: temporary breast growth

Endocrine aspects II

Thyroid problems

- Infants with PWS have central or peripheral hypothyroidism that's why check TSH and fT4
- Treatment with L-Thyroxin

Central adrenal insufficiency (CAI)

- Education on rare observation of CAI in PWS
- In urgent cases of clinical symptoms of CAI after check of laboratory parameters 30 mg/m² body surface area hydrocortisone in 3 parts

Treatment with growth hormone

- From age of 6 month until 98% of final height
- No growth hormone test necessary if PWS is genetically confirmed

Target:

- Improvement of body composition
- Increase of final height

Conduction of therapy:

- Dose recommendation max. 0,035 mg/kg body weight/day sc
- Dose management by IgF1, IgFBP3, BIA, longitudinal development/growth speed
- Check for side effects: metabolism, sleep apnea, heart

Prevent orthopedic problems

Prevent Scoliosis:

- No early passive sitting
- Sitting is allowed when child reaches position themselves
- First crawling than sitting
- Build up muscles at the trunk

Prevent Foot problems:

- massage, bandage, tape
- Prevent cranial deformations: change head and toe position in bed every day

Additional health issues

Problems:

- High pain threshold
- Poor thermoregulation
- Teeth erosion-loss of enamel/ severe tooth wear
- Decreased vomiting
- Daytime sleepiness

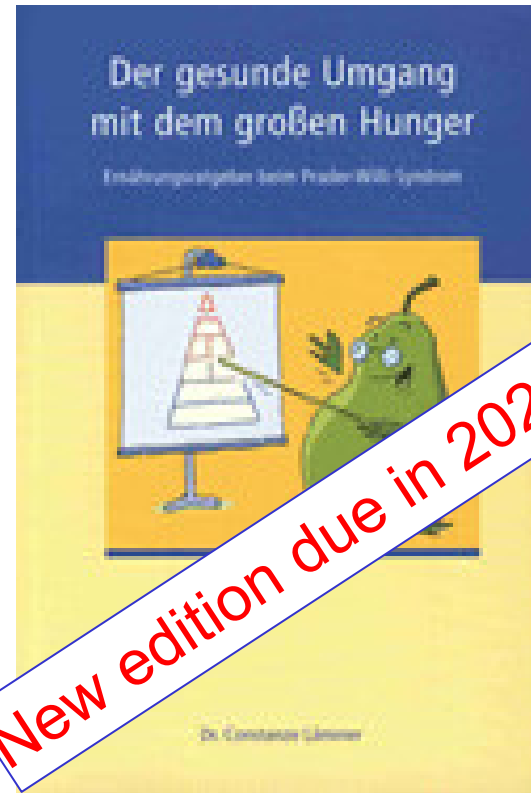
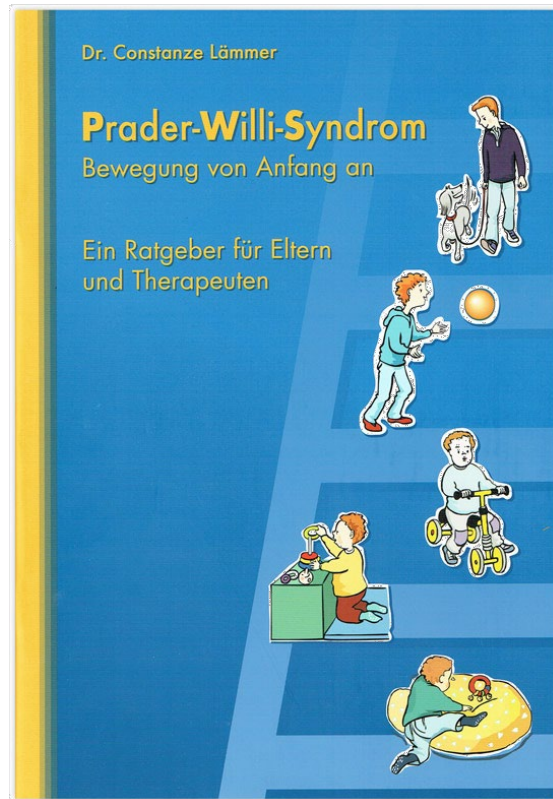
Solutions:

- Attention in case of accidents
- Need intense stimuli
- Caution for hot food but also bathing water and clothing selection
- Start dental care with the first tooth 2x per day, drink water, no sweets
- Watch infant carefully
- Structure the day, plan time for daytime naps

Booklets for PWS

The following publications are sponsored by Pfizer:

- PWS Sport book, 2016 (German, Dutch)
- PWS Nutrition book, 2006 (German)



Conclusion

- Early intervention supports infants with PWS and their families in daily life
- Muscular hypotonia is a central problem and has an impact on every aspect of life of an infant with PWS
 - Therefore, physiotherapy is necessary from the first day on to built up muscles
 - Additionally, growth hormone treatment from 6 month on helps to built up muscles
- Energy requirement is lower - up to 2/3 of a healthy infant
 - individual energy intake has to be adopted to the development according to the weight chart
- development and therapeutic measures shall be verified every three month
- Orthopedic problems are common