Pallister-Killian Syndrome:
Medical Nutrition Therapy (MNT), Nutrition Interventions
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Best Practices in PKS

- Registered Dietitian (RD) should remain part of interdisciplinary team
  - continuous updates throughout lifecycle
  - continuous adjustment based on symptoms of PKS
    - medical nutrition therapy to improve outcomes
  - continuous adjustments based on unexpected modalities of PKS, changes in growth pattern over time
Nutrition Throughout the Lifecycle

- Infants - adjust formula/tube feed for kcal, protein, micronutrient needs
  - Calcium
  - Phosphorus
  - Iron
Nutrition Throughout the Lifecycle

- Toddlers: introduce new foods individually
  - increase range of textures
  - identify potential allergies
  - promote motor development
Nutrition Throughout the Lifecycle

- Adolescence- balanced diet with variety
  - promote individual choice in foods
  - promote variety for adequate vitamin/mineral intake
  - promote balance for appropriate calorie intake
Nutrition Throughout the Lifecycle

- Adulthood- sustain positive eating practices established in adolescence

- adopt increases in individual control of diet where possible

- promote simple cooking techniques where possible
Symptoms of PKS Potentially Affecting Nutrition

- Developmental Delay
- Walking Difficulties/ Limited Movement/ Brachymelia
- Hypotonia
- Osteopenia, Joint Contractures
- Gastroesophageal Reflux
- Cleft Palate, Delayed Dental Eruption
- Edema
MNT: Developmental Delay

- RD, parents to work with physical and/or occupational therapist
  - improve coordination of sucking, swallowing, chewing, breathing
  - sucking may be weakened, leading to fatigue

- Environmental changes-
  - regular eating schedule
  - 3 meals with set times
  - planned snacks
  - special meal order at school may be necessary
MNT: Walking Difficulties, Limited Movement, Brachymelia

- Decreased activity level decreases calorie needs
  - parent and RD must coordinate to identify appropriate calorie intake
  - may require planning more low fat meals and snacks, reducing sugar

- Smaller limbs
  - height not a good predictor of calorie needs
  - peripheral muscle mass likely less than average
  - adjust calorie needs accordingly
MNT: Hypotonia

- low muscle mass
- high fat mass
- may slow physical motor development
  - decrease activity level
- adjust calorie needs accordingly
MNT: Osteopenia, Joint Contractures

• To prevent or delay osteoporosis-
  • 175 mg/ 100 calories/ day Calcium
  • 91.5 mg/ 100 calories/ day Phosphorus
  • add human milk fortifier when breastfeeding
  • formula adequate

• consider supplementation in toddler, adolescent years
MNT: Gastroesophageal Reflux

- avoid feeding shortly before sleep
- avoid large, high fat meals
- thicken feedings for infant formula, enteral feedings
- low fat diet is often beneficial
- liquid diet may be beneficial
- avoiding heavily spiced foods may be beneficial
- decrease intake of acidic foods and carbonated beverages
MNT: Cleft Palate, Delayed Dental Eruption

- Surgical repair of cleft palate normally at 9 months
  - tube feed
  - concentrate formula
  - therapeutic bottle and nipple designs

- Delayed dental eruption does not indicate delaying solid foods
  - important for mandible muscle, motor skill development
  - softer textures, more chewing
MNT: Edema

- Swelling can be an issue
- Sodium and water restriction may be necessary
- adjust calorie needs to account for water weight in edematous tissues
MNT: Seizures, Epileptic Events

- Vitamin D supplementation recommended
- Avoid folic acid supplementation
- Separate enteral feedings from medication administration
- Consider ketogenic diet

- low carbohydrate
- high fat
- 4:1 (fat: carb/protein) calorie ratio
- high MCT - more nonketogenic foods OK
- MVI, Calcium supplement
MNT: Other Complications

- Deal with unique symptoms as they arise
- Endocrinologist may be important-
  - thyroid
- Routine evaluation/ reevaluation by interdisciplinary team
  - new or previously unknown symptoms arise routinely with PKS
Case Study in Temporary Overgrowth Syndrome

- 30 month old child with moderately severe to severe symptoms of PKS

- Fed enterally (G button) with monthly F/U with RD

- Very fast growth despite very low calorie intake
  - over 100 calories below estimated resting energy expenditure
  - sudden onset, temporary period of overgrowth
Weight-for-age percentiles: Boys, birth to 36 months

Length-for-age percentiles: Boys, birth to 36 months

SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).
Postnatal Overgrowth

- Be aware of this possibility
  - F/U regularly with RD
  - monitor growth closely in early childhood
- postnatal overgrowth recently noted in Croatian case*
  - 21 month old girl
- possible timeframe of 20 months to 36 months?
Summary

• Work with RD as part of interdisciplinary team

• Evaluate MNT for each symptom, combination of symptoms

• Continually reevaluate

• Promote new foods, more independence with age as appropriate
Thank You for Your Time

• Please feel free to
  • ask questions
  • comment
  • express your concerns

• contact me for my references
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Please feel free to contact me;
I will be available to speak in private after the presentation as well