APPROACH TO HEALTHY NUTRITION IN PWS

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Kindercare Pediatrics, PWS clinics
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Sanjukta Basak and Karen Balko have no disclosures to report.
Objectives

- Provide an overview of the different approaches to healthy nutrition and best practices in the literature and clinical experience.
Outline

- Planning a PWS Diet
- Fluid Intake
- Different Diet Approaches
- Creation of PWS Friendly Food Environment
- Nutritional Supplements
- About us…KinderCare PWS Clinic
PLANNING A PWS DIET

NUTRIENTS TO CONSIDER, ENERGY, PROTEIN, FAT, CARBS
Nutrients to Consider when Planning your Child’s Diet

- Aims to meet adequate:
  - Energy/calories – metabolic needs
  - Protein – muscles, cellular growth and repair
  - Carbohydrates – energy for brain and muscles
  - Fat – nervous system, brain
  - Vitamins and Minerals – immune system, bones, heart,
Calorie/Energy requirements are individual

- Variations based on:
  - Age
  - Activity
  - Individual variations in calorie needs
  - Weight goals – growth hormone vs non growth hormone treated
What is a calorie?

- By definition a calorie is the energy it takes to raise the temperature of 1 gram of water to 1 degree celsius.

  \[ \text{CALORIES} = \text{ENERGY} \]

- It is the energy or fuel for our organs to function.

- A calorie does not speak to nutrient balance, and the quality of the food.
What is the right amount of calories?

Usually: 60-70% of calories but varies

<table>
<thead>
<tr>
<th>INFANTS, AND TODDLERS</th>
<th>CHILDREN, ADOLESCENTS AND ADULTS:</th>
</tr>
</thead>
<tbody>
<tr>
<td>6.5-8.0 calories/cm</td>
<td>8-11 calories/cm for normal growth</td>
</tr>
<tr>
<td>Eg. 2 year old boy 80 cm x 8 cal/cm = approx. 640-700 calories</td>
<td>7-8 calories/cm for weight loss</td>
</tr>
</tbody>
</table>

Berall et al. unpublished data.
Normal growth = adequate energy
Suboptimal Growth = Inadequate Energy Intake
Suboptimal Gain = Excess Energy Intake
Protein – muscles, cell, growth, satiety

**Ideal protein sources**
- Unprocessed poultry and meat
- Pink fish more often than white
- Plain greek yogurt
- Beans/legumes/nut butters/seeds/nuts
- Eggs

**Not so great protein sources**
- Hot dogs
- Boxed burgers, chicken and fish fingers or deep fried meats
- Processed meats/chicken/fish
Carbohydrates: Energy

**Ideal Sources:** Complex, high fibre, low simple sugars, lower glycemic index

- Oatmeal (not instant)
- Legumes (beans/lentils)
- Fresh fruit and vegetables
- Brown rice, whole wheat pasta, quinoa, buckwheat, millet, pot barley, teff
- Sweet potato, root vegetables
- Whole grain/sprouted bread/higher fibre (i.e. Ezekiel or Stonemill)
- High fibre, low sugar cereals >4 g fibre <8 g sugar (shreddies)

**Not so great:** Simple, low fibre, high simple sugars, higher glycemic index

- Low fibre, high sugar cereals (fruit loops/rice krispies)
- White bread, rice, pasta, crackers
- Cookie, cakes, candies, muffins, granola bars (most)
- Ice cream, frozen yogurt
- 100 calorie snack packs
- Sugar, High fructose corn syrup
- Juice
Fats: Brain, Nervous System, Satiety, Absorption
vit A,D,E,K, Essential Fatty Acids

CHOOSE...
I SAID YOU'RE THE GOOD KIND OF FAT!
Achieving adequate fluids
“Yucky water”

- Unclear etiology of PWS’ dislike of water
- Can teach a child to learn to drink water
- Role Model
- Persevere
- Fun water bottle
- Straws
- Water first
- Preference to avoid aspartame/artificial sweeteners (last resort to ensure hydration)
Promoting water intake: Ideas for flavoring water

- Lime or lemon slices with basil or mint
- Cucumber and mint
- Boil cinnamon sticks, cloves, apples – cool
- Lemon slices and vanilla bean or vanilla extract
- Lemon and lime slices
- Boil chunks of Ginger and lemon slices – cool – add stevia
- Cucumber and lemongrass
- Lemon and cayenne pepper
- Chilled flavored herbal teas
- Add any herbs you like: tarragon, rosemary, basil, mint, sage
DIETARY APPROACHES
Um, Eve, that's not a salad, that's my dirty laundry...
What Diet Approach is best?

- Individualized
- The one that works best for your child for growth, development, energy needs
- One that is practical for you and your family
- One that does NO harm
- **BALANCED**, meets all nutrient needs
A reduced-energy intake, well-balanced diet improves weight control in children with Prader-Willi syndrome

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†Department of Health Outcomes and Policy, University of Florida, Gainesville, FL, USA
‡Center for Epigenetics, University of Florida, College of Medicine, Gainesville, FL, USA

- 63 children aged 2-20 years
- Approach: 30% fat, 45% carbohydrate, 25% protein and 20 grams of fibre with energy restricted diet
- Result: Balanced macronutrient content IMPROVES weight and body composition in children with PWS compared with simple energy restricted diet
Diet Approach: Balanced Macronutrients

<table>
<thead>
<tr>
<th>Nutrient</th>
<th>Average Canadian</th>
<th>Ideal for PWS</th>
</tr>
</thead>
<tbody>
<tr>
<td>PROTEIN (%)</td>
<td>15-20</td>
<td>25</td>
</tr>
<tr>
<td>FAT (%)</td>
<td>30</td>
<td>30</td>
</tr>
<tr>
<td>CARBOHYDRATES (%)</td>
<td>50-55</td>
<td>45</td>
</tr>
</tbody>
</table>
# Diet Approach: Balanced Macronutrients

<table>
<thead>
<tr>
<th></th>
<th>Breakfast</th>
<th>Snack</th>
<th>Lunch</th>
<th>Snack 1</th>
<th>Snack 2</th>
<th>Dinner</th>
</tr>
</thead>
<tbody>
<tr>
<td>Protein</td>
<td>½ cup cooked oatmeal</td>
<td>½ cup greek yogurt</td>
<td>1 ounce roast turkey, 2 tbsp. hummus</td>
<td>1 boiled egg or 2 tbsp seeds/nuts</td>
<td>¼ cup cottage cheese and ¼ cup berries</td>
<td>3 oz rainbow trout or chicken</td>
</tr>
<tr>
<td>Grains</td>
<td>½ cup blueberries</td>
<td>¼ cup chopped apple with cinnamon</td>
<td>1 small ww tortilla Spinach/grated carrot or beet</td>
<td>Or</td>
<td></td>
<td>1/3 cup sweet potato roasted</td>
</tr>
<tr>
<td>Vegetable</td>
<td>1 cup unsw almond milk</td>
<td></td>
<td>½ cup greek yogurt</td>
<td>Sliced cucumbers or celery</td>
<td></td>
<td>1 cup sautéed veggies</td>
</tr>
<tr>
<td>Fruit</td>
<td>5 almonds</td>
<td></td>
<td>1 clementine or kiwi</td>
<td></td>
<td></td>
<td>Green salad</td>
</tr>
<tr>
<td>Dairy</td>
<td></td>
<td></td>
<td>2 tbsp avocado</td>
<td></td>
<td></td>
<td>½ cup almond milk</td>
</tr>
<tr>
<td>Fat</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Olive oil and vinegar dressing</td>
</tr>
</tbody>
</table>
Diet Approach: PWS Plate Method

Prader-Willi Plate

- **High Quality Protein**
- **Whole Grains**
- **Fats like oils, flax**
- **Fruit (but no fruit juice)**
- **Vegetables**
- **Dairy (like yogurt, cheese, milk)**
Traditional Plate vs PWS Plate

Prader-Willi Plate
- High Quality Protein
- Whole Grains
- Fats like oils, flax
- Fruit (but no fruit juice)
- Vegetables
- Dairy (like yogurt, cheese, milk)
PWS Plate Method

Benefits
- Easy guidelines
- Applies for all family members
- Ensures dietary balance

Drawbacks
- Plate sizes vary
- Portion sizes are unclear – heaping vs flatter portions
- Does not identify best choices within food groups
- Amount of added fat unclear
Diet Approach: Red Yellow Green (RYG) System
RYG: Average servings per day for healthy balanced diet for PWS

**Toddler/preschooler**
- 3 meals, 2 snacks per day
- Typical meal:
  - 1/4 cup high quality grain
  - 1-2 ounces protein
  - 1/2 cup vegetables (low starch)
  - 1-2 tsp healthy fat (MCT/avocado/olive/coconut)
  - Plus 2 servings per day dairy (yogurt, milk, milk alternative)

**Children/Adults**
- 3 meals, 2-3 snacks per day
- Typical meal:
  - 1/4-1/2 cup high quality grain or 1 slice of whole grain bread
  - 2-3 ounces of protein
  - 1-3 cups vegetables
  - 2-3 tsp of healthy fat
  - Plus 2 servings per day dairy/dairy alternative
RYG Method

Benefits
- Concrete, simple, easy to understand, no grey areas
- Can be planned to equal best macro nutrient ratios
- Clear portions
- Easy for meal planning
- Clear cut portion sizes to help with meeting the right balance for your child

Drawbacks
- A dietitian usually required to help plan to ensure appropriate ratio of macronutrients to meet your child’s individual needs
- Misunderstood as originally was lower in fat
Dietary Approach: Ketogenic Diet

- Mostly fat, little protein, very little carbohydrates
- Forces body to burn fat as fuel
- Ketosis leading to appetite reduction
- Used to treat seizures
Ketogenic Diet

Benefits
- Anecdotal positive reports – Diet trend initiative and parental reports:
  - Reduced appetite/hyperphagia
  - Improved behavior
  - Improved focus
  - Improved performance/energy

Drawbacks
- Difficult to follow
- Multiple follow-ups: needs constant monitoring of ketones and diet
- Time: users report burnout with meal preparation
- Side effects: high levels of ketones (acidosis), and severe constipation
- Strict weighing/measuring of foods to ensure accuracy
- No clinical trials yet to support, lack of understand of long term effects
<table>
<thead>
<tr>
<th>Ketogenic</th>
<th>Modified Ketogenic</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Fat to non fat (protein/carb) ratio 1 g:3-4 g</td>
<td>• Still lower in carb but more liberal – ratios 1:1 or 1:2</td>
</tr>
<tr>
<td>• All foods weighed, energy calculation.</td>
<td>• High in fat, liberalized protein, no counting calories</td>
</tr>
<tr>
<td>• Vegetables, fruit, fat, and protein</td>
<td>• Limit in carbs from grains</td>
</tr>
</tbody>
</table>
Modified Ketogenic AKA Lower carbohydrate, moderate protein/higher fat

**Benefits**
- Anecdotal positive reports similar to ketogenic diet

**Drawbacks**
- Evidence limited- no clinical trials completed to date
- Risk of acidosis still exists with too low carb
- Sustainability
- Nutrient deficiency if not planned properly
- Toddlers often need a little more carb for energy under age 2 years
• **Population:** age 5 years to 17 years
• **Intervention:** low carbohydrate diet (15%carb; 65%fat; 20% protein) vs low fat diet (65%carb; 15%fat; 20% protein)
• **Outcome:** the effects of low carbohydrate diet versus low fat diet on levels of ghrelin, appetite suppressing hormones and changes in subjective hunger
CREATION OF PWS FRIENDLY FOOD ENVIRONMENT
MY FACE

WHEN I'M EATING MY SALAD
AND SOMEONE BRINGS DONUTS
PWS Friendly Family Home

- Groceries are mainly whole foods: high fibre, unprocessed, low sugar
- Healthy eating principles apply to all, regardless of weight/picky eating
- Plan meals ahead – even if it’s one day – less eating out/take out
- No edibles left in sight – put away food even fruit – visual triggers may reduce anxiety
- Locking food is not always necessary depending on person – but it’s ok to do it if reduces anxiety and food seeking behavior
- Siblings can be taken out for more treats/packed in lunch if necessary to create more normalcy
I am still hungry – what to do?

- Volumetrics – small plate, more veg, small containers, bento
- Adequate fat, protein, fibre on plate to help satiety
- Slow speed eaters down, satisfy the sensory seeker – flavour up!
- Consistency – in meal times and approach – whatever works stick with it!
Ways to add bulk to dishes

- Invest in a spiralizer or grate zucchini to make zucchini noodles
- Add veggies to dishes
- Small plates, cut up small pieces, thinner cuts of meat
- Cauliflower: makes rice, pizza crust, mash
- Spaghetti squash add to pasta or alone
NUTRITIONAL SUPPLEMENTS
Supplements

- Buying supplements: ensure NPH number
- Some lack rigorous data to support effectiveness.
- Important to weigh risk versus benefit and decide what is right for you and your family.
- Many can be obtained from your diet.
- Important to discuss with your health care team.
- You don’t “have to” start anything if you don’t want to...
Recommended Supplements

- Some important ones to consider with evidence in pediatric literature:
  - Vitamin D for bone health
  - Calcium for bone health (supplement if deficient in diet)
  - Omega 3/Fish oil: important for brain myelination, hard to obtain Health Canada RDI in PWS diet
  - Probiotics: helps regulate bowel movements
Vitamin D

- Important for bone health
- >90% of Canadians are deficient
- Supplementation based on CPS and PES guidelines

### Daily Recommendation for Vitamin D

<table>
<thead>
<tr>
<th>Age</th>
<th>International Units (IU) of vitamin D suggested per day (normal bone health)</th>
<th>International Units (IU) of vitamin D suggested per day (poor bone health)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-6 months</td>
<td>400 IU</td>
<td>400 IU</td>
</tr>
<tr>
<td>7-12 months</td>
<td>400 IU</td>
<td>400 IU</td>
</tr>
<tr>
<td>1-3 years</td>
<td>600 IU</td>
<td>600-1000 IU</td>
</tr>
<tr>
<td>4-8 years</td>
<td>600 IU</td>
<td>1000-2000 IU</td>
</tr>
<tr>
<td>9-18 years</td>
<td>600-1000 IU</td>
<td>1000-2000 IU</td>
</tr>
<tr>
<td>19+ years</td>
<td>600-1000 IU</td>
<td>1000-2000 IU</td>
</tr>
</tbody>
</table>
Calcium

- Supplementation determined based on evaluation of diet.
- Ideal: obtain calcium from dietary sources
- Supplement if necessary

<table>
<thead>
<tr>
<th>Age</th>
<th>Amount of calcium every day (as elemental calcium)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-6 months</td>
<td>210 mg</td>
</tr>
<tr>
<td>7-12 months</td>
<td>270 mg</td>
</tr>
<tr>
<td>1-3 years</td>
<td>700 mg</td>
</tr>
<tr>
<td>4-8 years</td>
<td>1000 mg</td>
</tr>
<tr>
<td>9-18 years</td>
<td>1300 mg</td>
</tr>
<tr>
<td>19+ years</td>
<td>1000 - 1200 mg</td>
</tr>
</tbody>
</table>
Omega 3 help lower your risk of heart disease. In infants, omega-3 fats help with brain, nerve and eye development.

Three types: ALA (alpha-linolenic acid), EPA (eicosapentaenoic acid), DHA (docosahexaenoic acid).

Health Canada Daily Recommended intake = 1.2 - 1.6 g/day.
  - Canada’s Food Guide recommends = 2 servings of fish per week.

Side effect of supplementation: fishy odor in breath, better with Krill based supplements.
Probiotics: are live micro-organisms which can confer a health effect on the host when consumed in adequate amounts.

Prebiotics: are nonviable food components which can confer a health benefit on the host by modulating intestinal microflora.
# Indications for Pediatric Health

<table>
<thead>
<tr>
<th>Brand Name</th>
<th>Probiotic Strain(s)</th>
<th>Dosage Form</th>
<th>CFU/dose</th>
<th>No. doses/day</th>
<th>Report/Gr Meq</th>
<th>MEC (grown)</th>
<th>Colic</th>
<th>ID</th>
<th>AAD</th>
<th>CDAD</th>
<th>BDD-IEC</th>
<th>HP</th>
<th>C</th>
<th>FRS/RA</th>
<th>CID</th>
<th>NI</th>
<th>CE/AD</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>BioGaia® drops</strong>&lt;sup&gt;®&lt;/sup&gt; <strong>BioGaia® chew tabs</strong></td>
<td><em>L. reuteri</em> protectus DSM 17938</td>
<td>Drops Chew. tabs</td>
<td>100M/5 drops 100M/tab</td>
<td>5 drops 1 tab</td>
<td>1S25&lt;sup&gt;3&lt;/sup&gt;</td>
<td>25-50</td>
<td>16</td>
<td>14</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td><strong>Culturelle®</strong></td>
<td><em>L. rhamnosus</em> GG</td>
<td>Powder</td>
<td>1B/packet</td>
<td>6-10 packets</td>
<td>12-17</td>
<td>18-20</td>
<td></td>
<td></td>
<td></td>
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</table>
| **FloraBABY®** | *B. breve* HA-129 1.2B  
*L. rhamnosus* HA-131 1.8B  
*B. infantis* HA-137 0.8B  
*B. longum* HA-135 0.4B | Powder | 4B/scoop | 1 scoop | 0 |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| **Florastor®** | *Saccharomyces boulardii* lyo | Capsule | 5B/capsule 5B/sachet | 1-7 capsules 1-2 sachets | 50-90 | 164 |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| **Junior Daily Probiotic (formerly DDS® Junior)** | *B. lactis* LA-12 4.2B  
*L. acidophilus* DDS®-1 0.8B | Powder | 5B/gram | 2 grams |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| **Proxifor® (formerly Lacidofil)** | *L. rhamnosus* RO011 3.8B  
*L. helveticus* RO052 0.28 | Capsule | 4B/capsule | 1 capsule |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| **UltraFlora® Children's** | *L. acidophilus* NCFM®  
*B. animalis subsp. lactis* Bi-07 | Chewable tablet | 2.5B each/ chewable tablet | 1-2 chew tablets |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| **VSL®#3®**<sup>®</sup> | *L. acidophilus* SD5212  
*L. casei* SD5218  
*L. bulgaricus* SD520 | Sachet | 450B/sachet | 1-2 sachets |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
Probiotics

- Growing body of evidence in pediatric literature about impact of probiotics in improving
  - Constipation
  - Gastric motility
  - Regurgitation
Other Supplements in PWS

- **Coenzyme Q10**: To improve alertness, energy,
- **Carnitine**: To improve alertness, reduce muscle fatigue, improve tone and strength
- **B vitamins**: Reported effects on cognition, mood, energy
- **N-acetyl cysteine**: To improve skin picking
- **Lactoferrin**: To improve iron levels
- **MCT oil**: Increases energy expenditure spares lean body mass
- **Prebiotics**: Alter gut flora to prevent weight gain
About Us…
PEDIATRIC PWS CLINIC
Dr. Sanjukta Basak, Pediatric Endocrinology
Karen Balko, Registered Dietitian
2 clinics/month Thursdays

ADULT PWS CLINIC
Dr. Reena Kilian, Family Physician
Karen Balko, Registered Dietitian
2 clinics/month Mondays

Dr. Daniel Flanders, Pediatrician
Owner Kindercare
A Short Walk Through our Practice

Growth & Development
- Review growth
- Growth Hormone

Nutrition & Behavior
- Nutrition counselling
- Adequacy of nutrients, caloric intake and fluids
- Food security and healthy food environment

Health Supervision
- Management and prevention of medical issues
- Physical activity
Our PWS clinics: Our 3 C’s

Caregiver Support
- Supportive counseling to reduce stress and direction for implementation of supports when necessary

Collaboration
- Encouraging streamlined, consistent and complete care
- Other RD’s, OT’s, specialists
- School advocacy

Caring
- We strive for compassionate family centered care
- Individualized flexible approach with a No harm philosophy
There are many dietary approaches to healthy nutrition in PWS.

There is no ideal “one” strategy but a healthy should include a balanced intake of nutrients to promote optimal growth.

Supplements can included in a healthy diet with consideration.
Any questions...
We can decide to have them in the ppt if people ask or distribute
Carnitor (L-carnitine)

- **Claims:**
  - Seems to work more at muscle level, reducing muscle fatigue
  - Critical role in transport of long chain fatty acids into the mitochondria of the cell for “burning” – produces energy for multiple systems in body- i.e. muscle contraction
  - Helps fatty acid metabolism for optimal available energy

- **Evidence:**
  - Anecdotal evidence only so far shown to improve mental and physical lethargy, muscle weakness
  - Unsure explanation of why carnitine may benefit some individuals with PWS – poorly understood

- **Safety:** helpful in setting of deficiency, best to be carried out with your health care provider

- **Side effects:** unknown, “fishy body odor”, diarrhea

- **Dose:** Infants and children: 50 -100 mg per kg/day divided 2-3 doses maximum 3 g/day – begin dose at 50 mg/kg/day
CoEnzyme Q10

- **Claims:**
  - naturally occurring vitamin like substance in body – present in all cells especially in muscle
  - Helps to provide energy to the cells

- **Evidence:**
  - Anectodal: Some parents see benefit, others see no results

- **Side Effects:** none known

- **Dose:**
  - Infants: 1-30 mg/kg
  - Children/Adults: 180-200 mg/day, usually 60-100 mg/day enough
Vitamin B12

- **Claims:**
  - Formation of red blood cells, helps with neurological functions, and productions of DNA
  - Can obtain this easily from food: (mainly animal) Meat, chicken, fish, eggs, dairy, nutritional yeast, non-dairy milks
Claim:
- Essential fatty acids – body can’t produce

Evidence:
- Best to get from our diets: Often difficult to get enough in calorie restricted PWS diets
  - Found in fatty fish (pink fleshed), fortified foods (eggs), walnuts, flax (ALA)

Dose:
# Variations of Volumes to Accommodate Carb/Fat/Pro Ratios

<table>
<thead>
<tr>
<th>Food Group</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dairy</td>
<td>2</td>
</tr>
<tr>
<td>Fruit</td>
<td>2-3</td>
</tr>
<tr>
<td>Grains</td>
<td>2-5</td>
</tr>
<tr>
<td>Meat/Alternatives</td>
<td>3-8 ounces</td>
</tr>
<tr>
<td>Fats and Oils</td>
<td>1-3 tbsp</td>
</tr>
<tr>
<td>Vegetables non starchy</td>
<td>Up to 7 cups/day (upper level for teens/adults)</td>
</tr>
</tbody>
</table>