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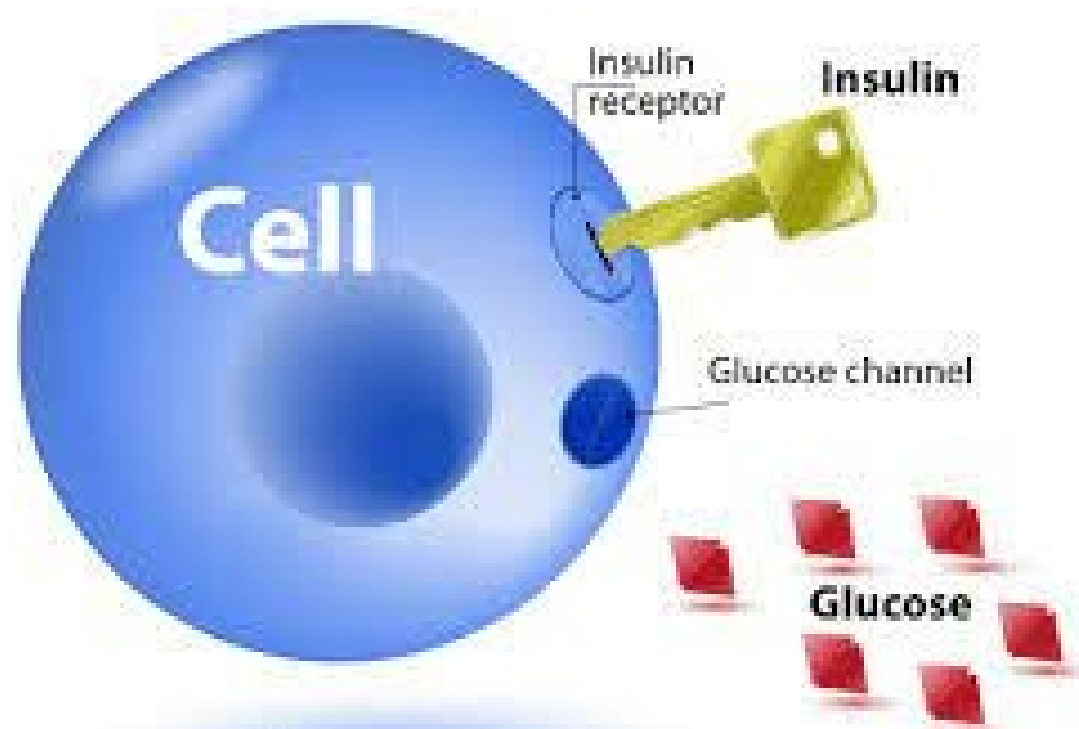
Prader-Willi syndrome: Nutrition Guidance and Recommendations

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The Endocrinologist's Perspective



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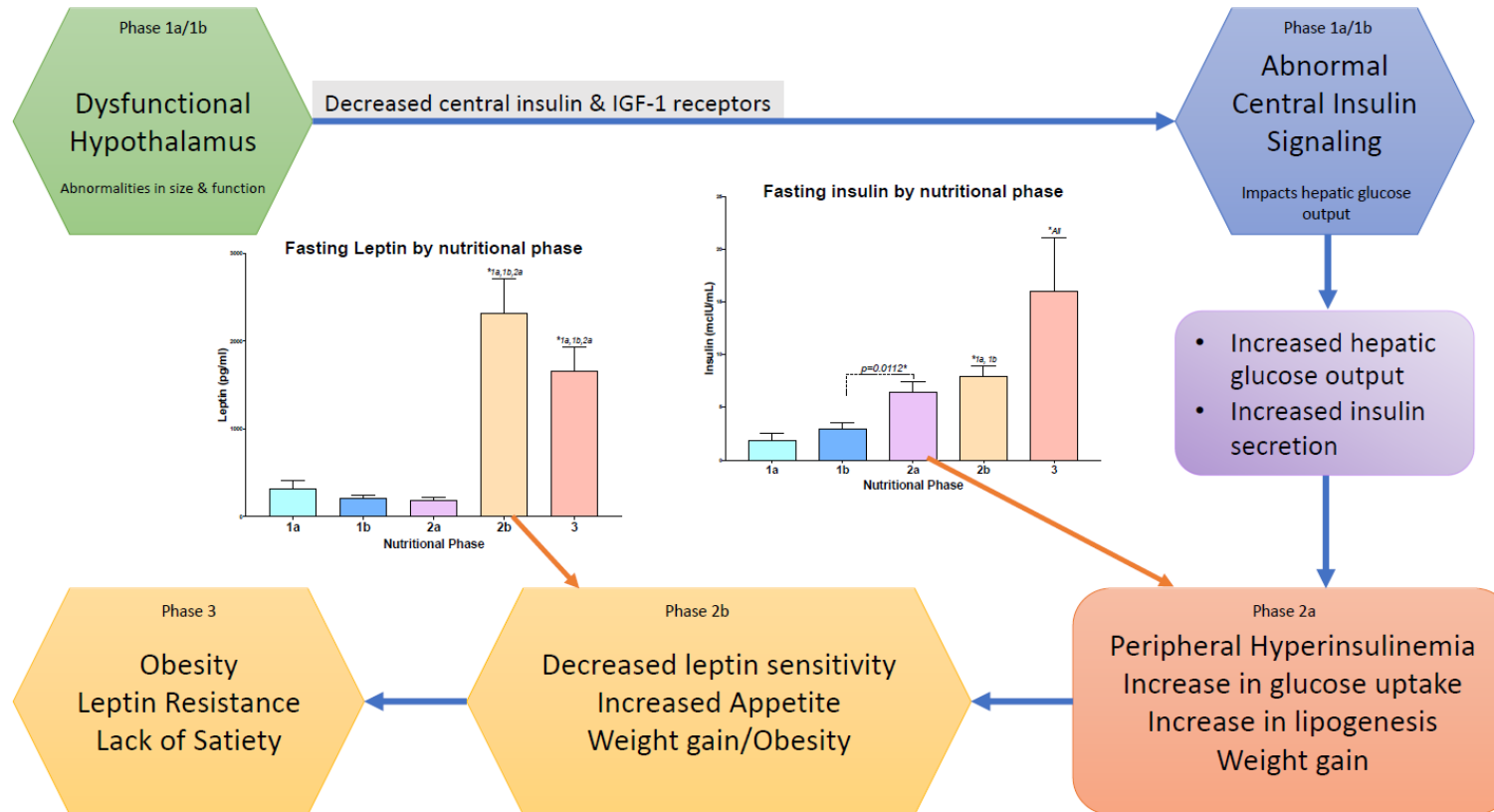
PWS Nutritional Phases

Phase	Median ages	Clinical characteristics
0	Prenatal to birth	Decreased fetal movements and lower birth weight than sibs
1a	0–9 months	Hypotonia with difficulty feeding and decreased appetite
1b	9–25 months	Improved feeding and appetite and growing appropriately
2a	2.1–4.5 yr	Weight increasing without appetite increase or excess calories
2b	4.5–8 yr	Increased appetite and calories, but can feel full
3	8 yr to adulthood	Hyperphagic, rarely feels full



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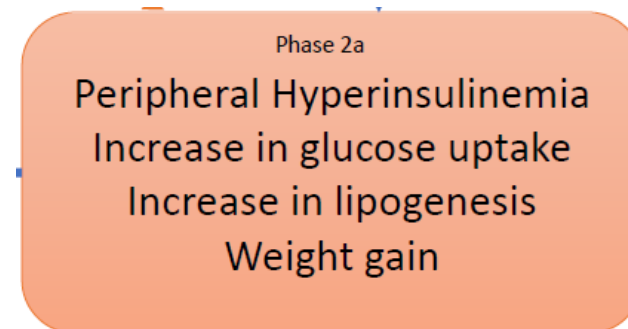
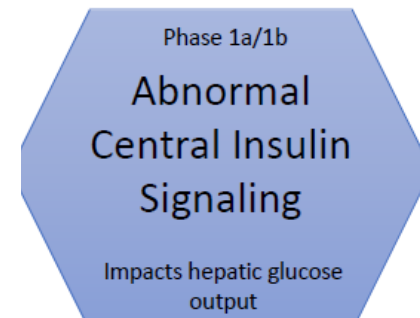
Progression to Hyperphagia



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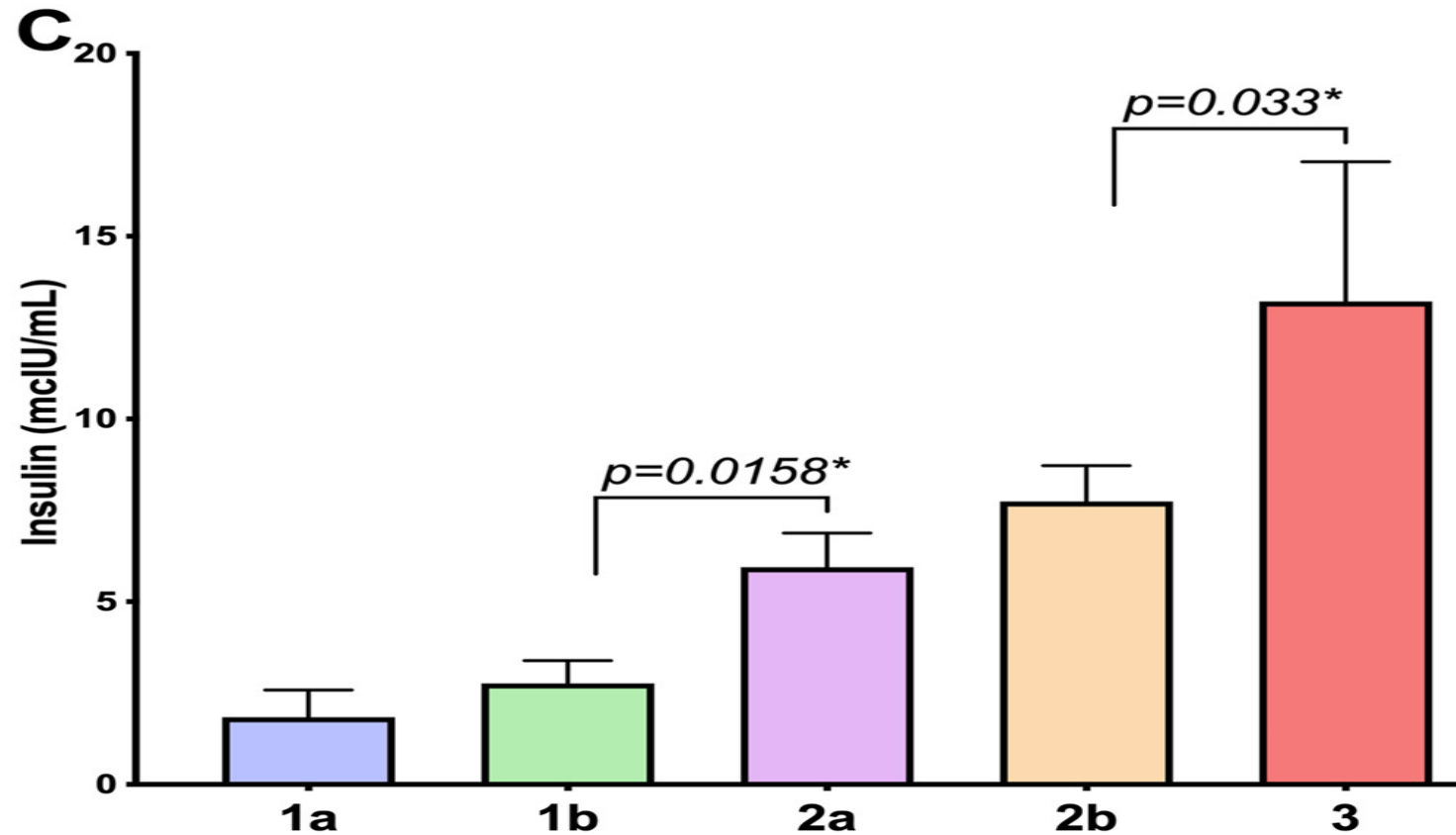
Glucose and Insulin in PWS

- Individuals with PWS can have hypoglycemia in infancy, which improves with growth hormone treatment
- Glucose levels tend to be within normal range in individuals with PWS
- There is increased insulin sensitivity in individuals with PWS
- Elevated insulin signaling in the face of increased insulin sensitivity in young children with PWS is a probable trigger for weight gain and onset of hyperphagia in this population.



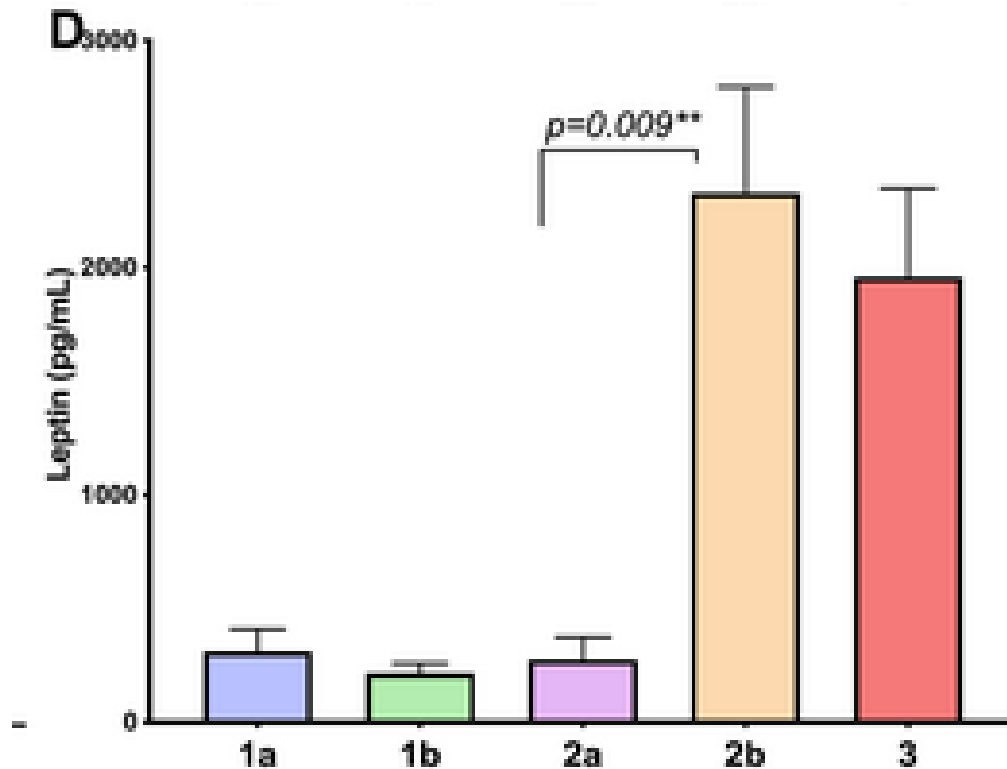
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Insulin Response to Sweet Taste in PWS



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Other Metabolic Changes in PWS



- As insulin release increases it begins to block leptin entry to brain, creating leptin resistance at blood-brain-barrier (this is not unique to PWS).
- Leptin levels rise because the brain starts to not see the leptin it was used to seeing.
- Leptin is a satiety hormone, so brain not seeing it = increased appetite



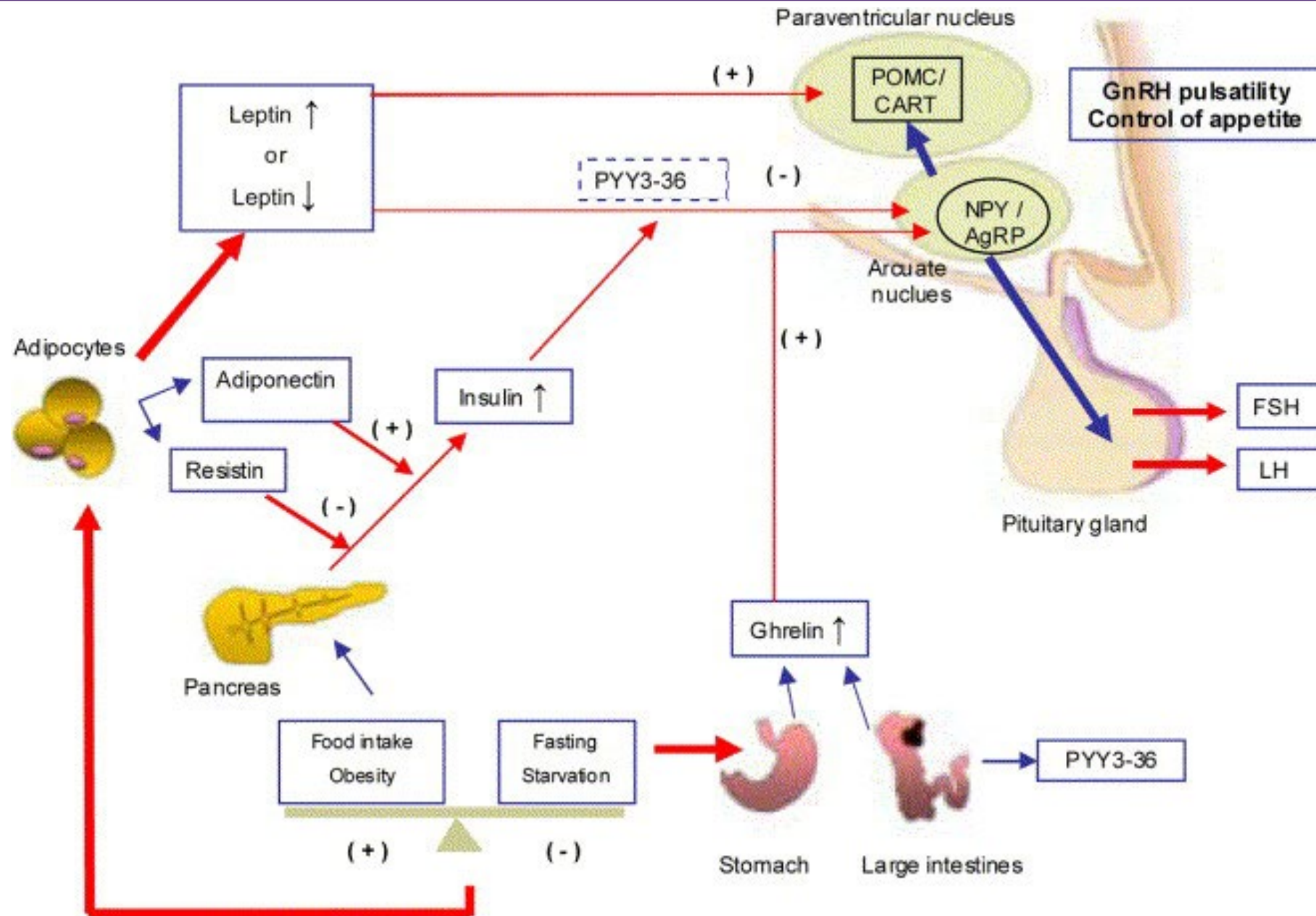
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Hormone Changes in PWS through the nutritional phases

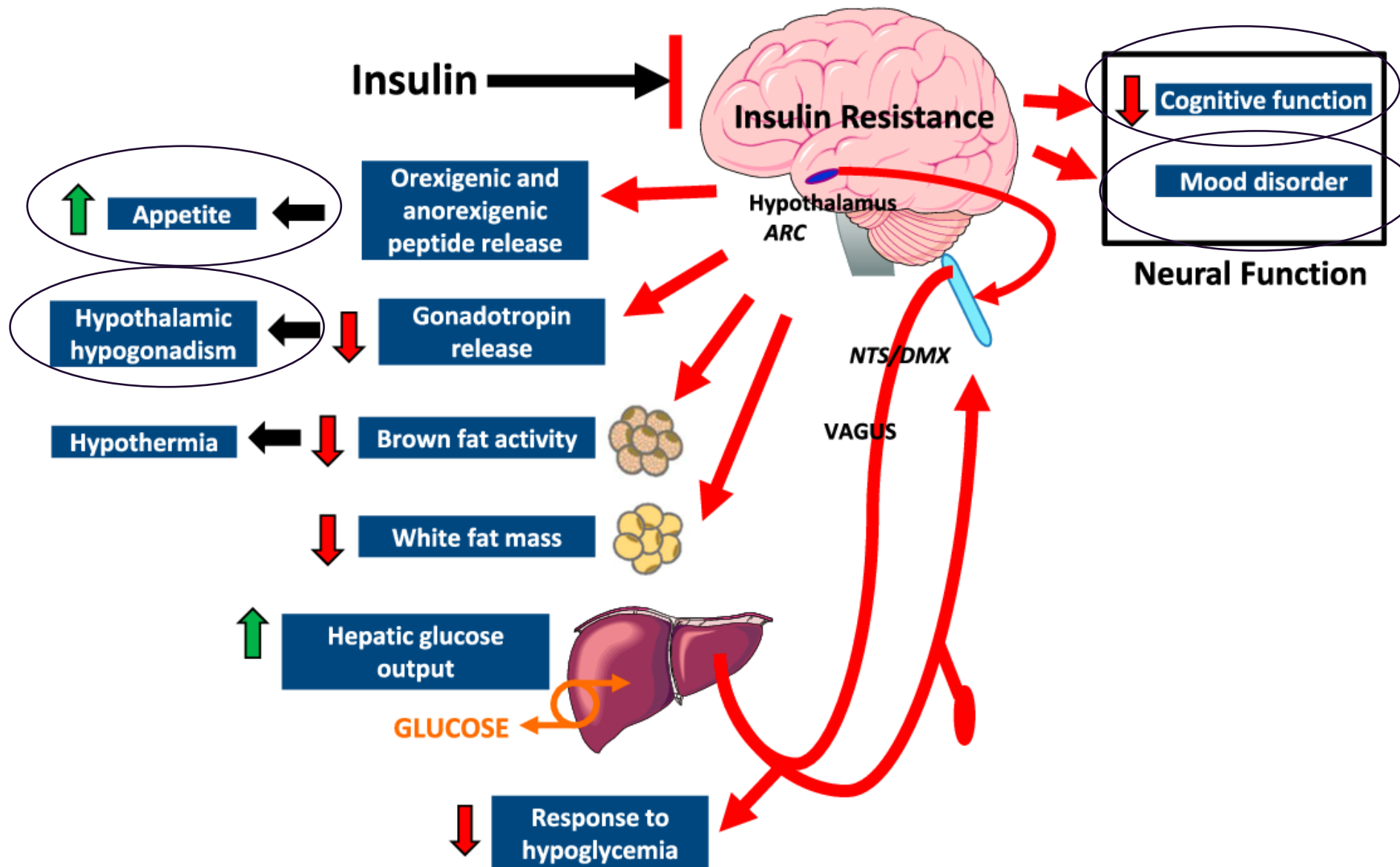
- As adipose tissue amount increases in PWS, insulin and leptin continue to increase.
- Ghrelin (hunger hormone) continues to go up as metabolic changes in nutritional phases cause increased appetite drive
- Adiponectin (cardioprotective hormone) decreases as adipose tissue amount increases



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Natural History of PWS

Metabolic Changes

Increased insulin
Increased leptin
Increased ghrelin
Low adiponectin

Body composition changes

Increased weight gain
Increasing body fat

Appetite Changes

Progression through nutritional phases
Insatiable appetite



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Nutritional Guidance for all ages

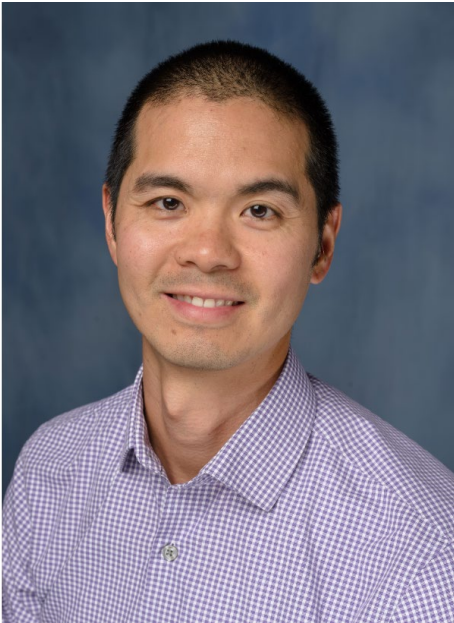
- Using complex carbs (whole grain) vs simple carbs (white flour, processed) helps mediate insulin response
- Pairing carbs with a protein or healthy fat helps mediate insulin response
- A very low carb diet is not recommended because too much of anything (eg dietary fat) will also cause increased insulin response
- Key to good nutrition is balance – a well-balanced diet results in appropriate weight gain, growth, muscle mass



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Prader-Willi Syndrome Nutrition Guidance and Current Recommendations

Michael Tan, MS, RD, LDN, CDCES



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Nutrition Guidance and Current Recommendations

Overview

- **Traditional** Guidelines
- **CURRENT** Recommendations
 - Quality
 - Quantity (portion sizes)
 - Balance (protein, carbs/fiber, fat)
 - Variety
 - Drinks
- Convenience
- Meal Scheduling
- Special Occasions
- Family Responsibilities
- Summary



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TRADITIONAL PWS Nutrition Recommendations

- Strict calorie-restriction targeting a specific weight
- Disadvantages:
 - Limits dietary fat intake due to high calorie content
 - Dietary fats are important for brain health, growth, development, satiety
 - Does NOT limit sugar substitutes/non-nutritive sweeteners (NNS)
 - NNS may increase appetite
 - Calorie counting can be tedious, labor intensive, and time-consuming
 - Inappropriately associates weight/lower calorie to health



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CURRENT PWS Nutrition Recommendations



- Unfortunately, there is a lack of clear, standardized medical nutrition therapy for PWS
- Current nutrition recommendations are not necessarily unique or highly specialized, but typically are stricter and more disciplined
 - Focuses on sustenance (physiologic nourishment) vs indulgence
 - De-emphasizes indulgence, celebratory/glorification of food
 - Minimizes/limits NON-hunger eating (emotional, behavioral, time-based, food as a reward, etc.)
 - Due to significantly lower calorie needs, there are less opportunities for “empty” or low-nutrient density foods
 - Limited flexibility (due to absolute/rigid thinking, “black and white” thinking)
- Recommendations are based on Mediterranean diet
- Recommendations focus on four key aspects
 - Quality
 - Quantity (i.e. portions)
 - Balance
 - Variety



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QUALITY

- Focus on
 - WHOLE grains, beans, legumes
 - Vegetables, fruits
 - Protein (emphasizes fish and plant-based proteins: nuts/seeds)
 - Other animal-based proteins and dairy are OK too
 - Extra virgin olive oil (unsaturated fat)
- Limits
 - Highly processed, “convenience” foods (“bars”, breakfast cereals, jelly/jams, etc.)
 - Refined grains
 - Excessive added sugar
 - Sweets and treats and other “sweet-tasting” foods
- **NO sugar substitutes/sweeteners**



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Examples of Sweeteners

- Acesulfame Potassium - Sunnett, Sweet One
- Aspartame - Nutrasweet, Equal
- Neotame
- Soluble Corn Fiber
- Saccharin - Sweet 'N Low, Sweet Twin, Sugar Twin
- Sucralose - Splenda
- Stevia/Rebaudioside - A Sweet Leaf, Sun Crystals, Steviva, Truvia, PureVia
- Allulose
- Monk fruit
- Sugar alcohols – xylitol, erythritol, mannitol, sorbitol, maltitol
- Tagatose
- Advantame
- Inulin (chicory root)
- Tapioca Fiber (prebiotic fiber)



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QUALITY

- **Guidance on “sweet tasting” foods**
 - **Fruit**
 - Fresh fruit only
 - Limited frequency (twice per day)
 - Limited portion sizes
 - Always balance with protein/fat
 - **Common “sweet tasting” foods generally NOT recommended**
 - Sweetened DRINKS
 - Breakfast cereals
 - “Bars” (granola, protein, fruit bars, etc.)
 - Dried/processed fruit
 - Jelly, jam, fruit spread, marmalade
 - Applesauce
 - “Fruit snacks”
 - Canned or highly processed fruit, “fruit cups”
 - Flavored yogurts, flavored oatmeal
 - Granola
 - Muffins
 - Jello/pudding
 - Sweet condiments (ketchup, teriyaki, BBQ, fruit-flavored vinaigrettes, honey mustard, etc.)
 - Syrup, honey



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QUALITY

- **Examples of sugar-containing foods that are generally OK to give (because they are not sweet)**
 - Plain Cheerios
 - Tomato Sauce
 - 100% whole grain bread/products
 - Sausage
 - Refried beans

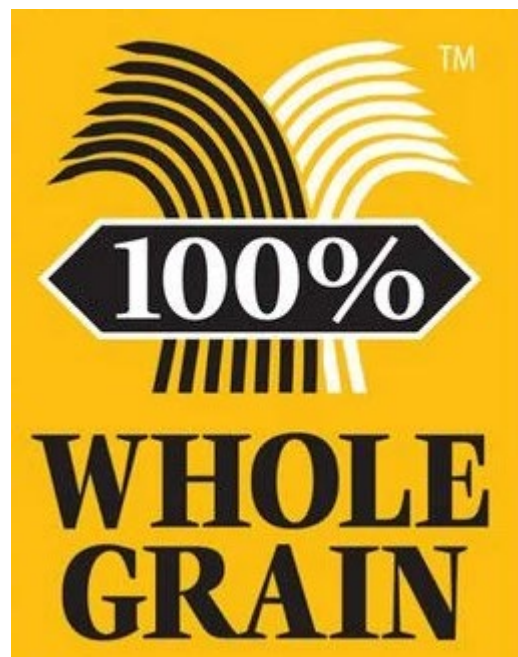


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QUALITY

- Low nutrient-density foods are not recommended because they (1) tend not to be satisfying or satiating and (2) offer minimal nutritional benefit
- **Examples:**
 - White flour
 - Rice cakes
 - Cassava flour
 - Tapioca flour
 - Rice flour
 - Potato starch
 - Veggie straws
 - Pirate's booty
 - Pretzels
- Look for **100% WHOLE** grain products



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QUANTITY

- Portion sizes are *highly* variable/individualized to specific needs
 - Based on growth, weight trends, activity level, body composition, genetics, etc.
 - General recommendations/handouts are only a starting point
 - **Most important: appropriate growth and weight gain**
- Generally, portions are significantly smaller when compared to general population
 - Due to low tone/lean mass, decreased energy needs
- Fruit is limited to twice per day and recommended to be served with a meal or with protein/fat

- Carbohydrates (whole grains, starchy vegetables, legumes, beans) – 1 fist (at each meal)
- Protein – 1 fist (at each meal)
- Non-starchy vegetables – 1 fist (at each meal)
- Nuts & Seeds – flat (*not* piled up) in the palm of hand (2x per day maximum)
- Fruit – flat (*not* piled up) in the palm of hand (2x per day maximum)



FIST



PALM



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QUANTITY

- Guidance on snacks
 - Do NOT assume a snack(s) is needed
 - Do not offer food unless requested/showing signs of hunger
 - If your child is easily distracted, they may not be hungry
 - Lighter snacks are typically preferred
 - i.e. Non-starchy veggies (carrots, cucumbers, tomatoes, etc.)
 - No carb-*only* snacks
 - If serving fruit or whole grain/starch, serve with protein/fat (nuts, seeds, cheese, meat, eggs, olives, avocado, etc.)
 - Do NOT schedule snacks (or meals)
 - Keep snacks “as needed”
 - Allow flexibility in mealtimes; intentionally build “noise” into the schedule



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BALANCE

1. Protein

2. Carbohydrates

- Starchy source of fiber (“complex carbs”)
 - WHOLE grains, starchy veggies, beans/peas/legumes
- Non-starchy source of fiber
 - Non-starchy veggies
- Do not recommend carb-free (or low carb) meals
- Do not recommend carb-heavy meals

3. Fats

- Unsaturated/plant-based fats are preferred
- Typically, fats are already included with the protein or added to the cooking process

1. Vegetables (NON-starchy)

Carrots	Cucumbers	Green Beans	Broccoli	Cauliflower	Spinach	Lettuce
Tomatoes	Celery	Zucchini	Yellow Squash	Asparagus	Bell Peppers	Onions
Brussel Sprouts	Artichoke	Cabbage	Kale	Snap Peas	Okra	Collards
Bok Choy	Eggplant	Spaghetti Squash	Swiss Chard	Turnips		

2. Protein

Beef	Pork	Chicken	Turkey	Salmon	Tuna	White Fish	Shrimp
Crab	Lobster	Scallops	Clams	Eggs	Greek Yogurt	Nuts	Seeds
Cottage Cheese	Duck	Soy/Tofu/Tempeh	Lamb/Goat				

3. Carbohydrate

WHOLE grain Bread	WHOLE Grain Pasta	Quinoa	Brown Rice	Wild Rice		
Barley	Oatmeal	Black Beans	Kidney Beans	Pinto Beans	White Beans	
Lima Beans	Chickpeas (<u>Garbanzo</u>)	Green Peas	Hummus	Black-Eyed Peas		
Edamame	Lentils	Sweet Potato	<u>Potato</u>	Whole Grain Couscous	Farro	Millet
Pumpkin	Butternut/Acorn Squash	Popcorn	Buckwheat	Amaranth		

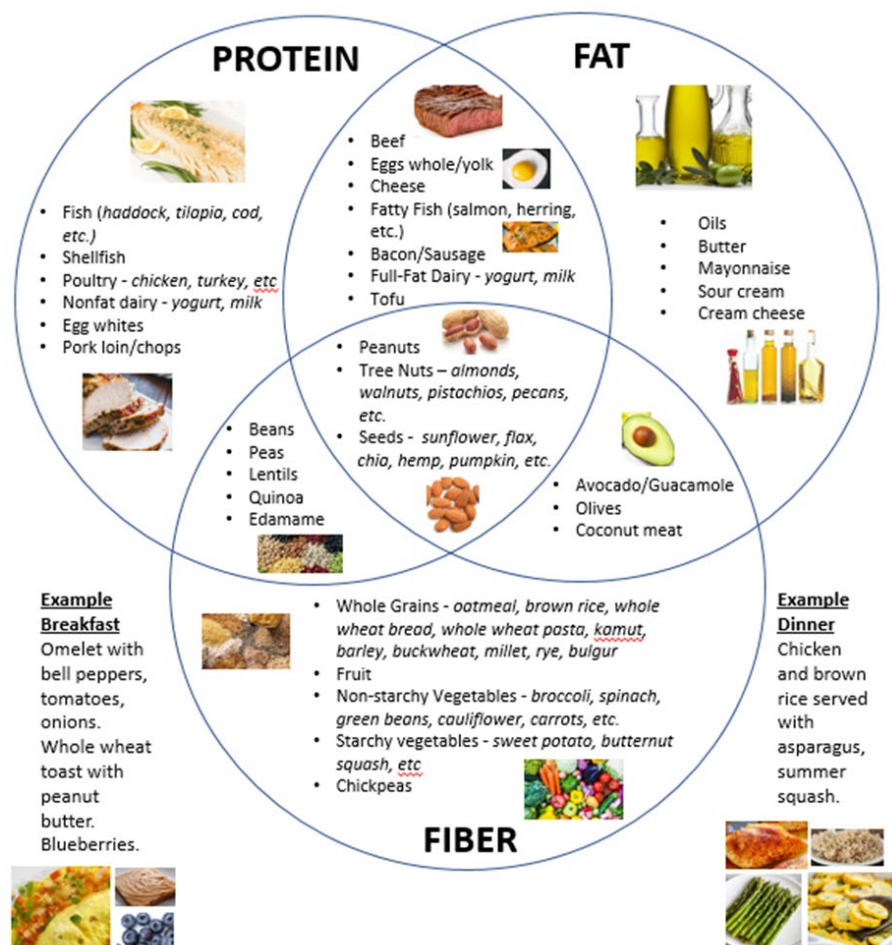


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BALANCE

Building a Balanced Meal

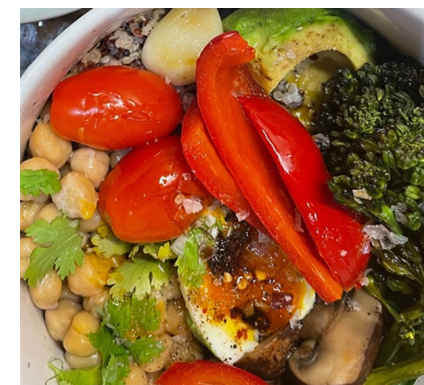


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VARIETY

- Encourage a large variety of foods, textures, and flavors from a *young* age
- Do *not* stop offering disliked foods
- Do not rely too much on favorite foods
 - Do not let your child's food preferences determine what is being served
- Continue to encourage new and different foods/flavors/textures etc.
- Nutritional variety:
 - Ensures the body is properly nourished
 - Reduces risk of nutritional deficiencies
 - Encourages diverse gut microbiome
 - Can reduce any selective ("picky") eating



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DRINKS!

- **Unsweetened** drinks only
 - Plain water
 - Ice-cold may work better
 - Milk or milk alternatives
 - Unsweet tea
 - Squeeze of *sour* fruit (lemon, lime) into water
- Beware of clever marketing: “no sugar added”, “no artificial sweeteners”, “sugar free”, “zero calorie”, etc.
 - These products are likely sweetened with alternative sweeteners (i.e. stevia, monk fruit, erythritol, sucralose, etc.)
- Build PLAIN water into *daily* routine
- Start EARLY, be consistent, do NOT offer alternative

UNSWEETENED Drinks

Flavored Sparkling Waters (Polar, La Croix, Dasani, Bubly, Waterloo, AHA)



Flavored Waters (Hint, JUST water, Rethink, WonderWell, True Lemon FRUIT INFUSIONS)



Fruit Teas (Celestial, Twinings, Lipton)



Milk Alternatives (Unsweetened)



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MEAL AND SNACK IDEAS

	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	Sunday
Breakfast	Egg, Sausage, Raspberries, Whole grain toast	PLAIN Greek Yogurt Blueberries Cinnamon Oats	Omelet w/ Tomatoes Peppers Whole grain toast Strawberries	PLAIN Greek yogurt Whole grain pancake with peanut butter Cherries	Egg Oatmeal Cinnamon Strawberries	PLAIN Greek Yogurt Blueberries Whole grain toast	Peanut Butter on Whole grain pancake Sliced almonds Blackberries
Lunch	Steak Cauliflower Carrots Sweet Potato	Chicken and Bean Chili Onions Tomatoes Yellow Squash	Salmon Broccoli Wild rice	Ground Turkey Tomatoes Cauliflower Black Beans Avocado	Salmon Spinach Quinoa	Steak Asparagus Butternut Squash	Chicken Carrots Lentils
Snack	Apple Sunflower seeds	Pear Pecans	Cherries Walnuts	Apple Peanut butter	Pear Sunflower seeds	Orange Peanuts	Apple Peanut butter
Dinner	Turkey Meatballs Broccoli Whole grain or Chickpea Pasta	Salmon Brussel sprouts Brown Rice	Turkey Spinach Tomatoes Onions Black Beans Avocado	Shrimp Green beans Quinoa	Fish Roasted carrots Whole grain couscous	Baked chicken Green beans Sweet potato	Pork Asparagus Peas



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CONVENIENCE - meals

- Quick, “no cook” meal examples



Breakfast	Lunch	Dinner
Peanut butter, whole grain toast	Deli meat, Triscuits, carrots	Tuna, whole grain bread, cucumber
Cashews, Josephs Pita	Pre-boiled egg, popcorn, bell peppers	Sunflower seeds, canned black beans, carrots
Almonds, PLAIN/Original Cherrios	Plain Greek yogurt (+ fruit, cinnamon), edamame, tomatoes	Deli turkey, chickpeas/hummus, steam-in-bag green beans



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CONVENIENCE – snacks

- Harvest Snap Pea Crisps
- Whole grain crackers/bread (+ protein/fat)
- Hippeas
- Crunchsters
- Bada Bean Bada Boom
- Cheese
- Meat
- Eggs
- Popcorn (+ protein/fat)
- Nuts/seeds
- Beef jerky/meat stick



- Yogurt (PLAIN)
- Dried chickpeas
- Peanut butter
- Peanut butter puffs
- Chicken/tuna salad
- PLAIN Cherrios (+ protein/fat)
- Fresh fruit (+ protein/fat)
- Veggies (carrots, tomatoes, cucumbers, peppers, celery, etc.)
- Dried veggies
- Hummus
- Dried seaweed



- Edamame or dried edamame



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MEAL “Schedules”

- A strict schedule can exacerbate behaviors when schedule is not/cannot be followed
- Maintaining flexibility in schedules is preferable and recommended
 - May need to intentionally build “noise” into daily routine
 - Base meals around *activities*, not the clock/time
 - Ensure your child knows that food *will* be provided, but is *not* dependent on a specific time
 - Avoid “feeding the clock”



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SPECIAL OCCASIONS

- Sweet treats (cookies, cakes, cupcakes, brownies, ice cream, etc.) are OK on **extremely special** occasions (~5 times per *year*)
 - Birthdays
 - Special holidays
 - Big vacations
 - Examples of NOT special occasions:
 - “on weekends”
 - Friday nights
 - “Wednesdays”
 - After dinner (i.e., dessert)
 - “when we go out to eat”
 - Uncle’s birthday



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FAMILY ROLES

- TEAM effort
 - Whole-family approach
 - Proper nutrition is beneficial for everyone in the family
- Parents
 - Do *not* cater to children's likes and dislikes; your children are not "customers", and you are not a "waiter/waitress"
 - Should treat all children similarly
 - Need to set aside own food issues
 - Do not let own food preferences determine what is being served
 - Understand your food anxiety will spill over to your children
 - Acknowledge potential risk of disordered eating in other family members
- Grandparents
 - Food is *not* a reward or treat (or punishment)
 - Do *not* show love through food (use other forms of rewards: toys, books, game, quality time, money, adventures/trips, etc.)
- Siblings
 - No sharing of food
 - Awareness of access to food
 - Maintain a healthy relationship with food



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SUMMARY

- Quality – no sweet-tasting foods! Focus on Mediterranean Diet
- Quantity – small portions, especially fruit
- Balance – protein, complex carbs, non-starchy veggies (no carb-free meals & no carb-heavy meals)
- Variety – do not serve the same foods over and over. Continually offer new and non-preferred foods.
- Drinks – **UNSWEETENED** only
- Convenience – meals/snacks do not have to be fancy or complicated!
- Meal schedules – maintain flexibility; snacks not necessarily needed
- Special occasions foods ~ 5 times per year
- Family Roles – parents are in charge! Do not cater to too much to your child



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Questions?



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