

Prader Willi Syndrome

Dietetic Q&A Session

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Weaning



- *Is there a booklet anything that can guide us through weaning?*

Weaning booklet available on <https://www.safefood.net/getattachment/193da6f1-5830-4775-97ec-7ea19624701c/feeding-your-baby-hse.pdf?lang=en-IE>

- *What amounts should we be giving.*

Weaning for example 9- 12 months old aim for 4-6 tablespoons at each meal.
Depends on the age and stage

- *Can we give fruit?*

Yes

- *Do we need to give protein at every meal?*

Offer protein (HBV) at 2 meals

- *Is there a specific diet for weaning*

No specific diet

Age appropriate, healthy weaning diet

As you progress through weaning you are aiming for 3 family meals +/- 1 to 2 snacks depending on what fits in your family and your child's needs

- Don't give your baby:**
- honey or sugar
 - unpasteurised cheese
 - salt, gravies, packets or jars of sauce or soup, mealmakers
 - undercooked eggs
 - bran
 - tea or coffee
 - liver
 - processed or cured meats (such as sausages, ham, bacon)
 - high fat or high sugar foods (such as crisps, chocolate, cake, sweets), and
 - whole or chopped nuts (as there is a risk of choking).

| Stage 3 between 9 and 12 months | | | | |
|---|---|---|--|---|
| Foods to choose | Food textures | Aim for | Skills learned | Suitable drinks |
| Most homemade family foods (without added salt, gravies, sauces, packets or jars of sauce or sugar) are now suitable. | Lumpy foods. Chopped foods. Harder finger foods. Wide variety in foods and textures. | Three meals of about 4-6 tablespoons. Two to three snacks. Be able to manage more than two textures in one meal. All drinks (other than breastfeeds) taken from a cup or beaker. | Lumpier textures will help with speech, chewing and increase the variety of baby's diet in later childhood. Self feeding. | Breastmilk (on demand). Infant formula (approx 400mls or 14ozs a day). Cooled boiled water. |

| between 9 & 12 months | | | |
|--|-------|---|---|
| breakfast | | dinner | tea |
| Readybrek™ made with breast, formula or whole cow's milk | day 1 | Meatballs in tomato sauce with pasta | Wholemeal fingers of toast with baked beans |
| Porridge made with made with breast, formula or whole cow's milk | day 2 | Flaked cod, carrots and potatoes | Well cooked pasta with slices of cold chicken and cucumber |
| Weetabix™ made with breast, formula or whole cow's milk | day 3 | Minced lamb or beef, carrots and potatoes | Boiled or scrambled egg with fingers of toast |
| Readybrek™ made with breast, formula or whole cow's milk | day 4 | Flaked salmon, turnip, peas and potatoes | 1 egg omelette with cheese, slice of pitta bread and hummus |
| Porridge made with breast, formula or whole cow's milk | day 5 | Chopped chicken, broccoli and potatoes | Homemade soup and a sandwich |
| Weetabix™ made with breast, formula or whole cow's milk | day 6 | Chilli con carne with rice | Fingers of toast with cheese spread and slices of tomato |
| Readybrek™ made with breast, formula or whole cow's milk | day 7 | Flaked trout, parsnip and potatoes | Pasta shapes with tomato sauce and grated cheese |

| | Why | Good food sources |
|---------------------|---|--|
| Iron | Important for brain development between 6 months to 2 years of age. | Meat (this form of iron is easily absorbed). Red meat – beef, pork, lamb Poultry – chicken, turkey Non-meat sources (this form of iron is not so easily absorbed). Eggs. Baked beans, lentils, chickpeas, Dark green leafy vegetables (cabbage, broccoli, spinach) Cereals with added iron Dried fruits (raisins, sultanas, apricots). |
| Vitamin C | Helps the absorption of iron in the body when eaten at the same time as non meat sources. | Oranges or mandarins. Plums. Berries (strawberries, blueberries, raspberries). Kiwi fruit. Broccoli. Cauliflower. Peppers. |
| Omega-3 fats | Essential for brain and eye development. | Breastmilk. Oily fish (salmon, trout, mackerel, sardines) – tinned, fresh or frozen. Include two 1oz portions of oily fish a week from 7 months of age. |
| Calcium | Important for good bone development. | Oily fish (salmon, trout, mackerel, sardines) – tinned, fresh or frozen. Eggs. Fortified breakfast cereals. |

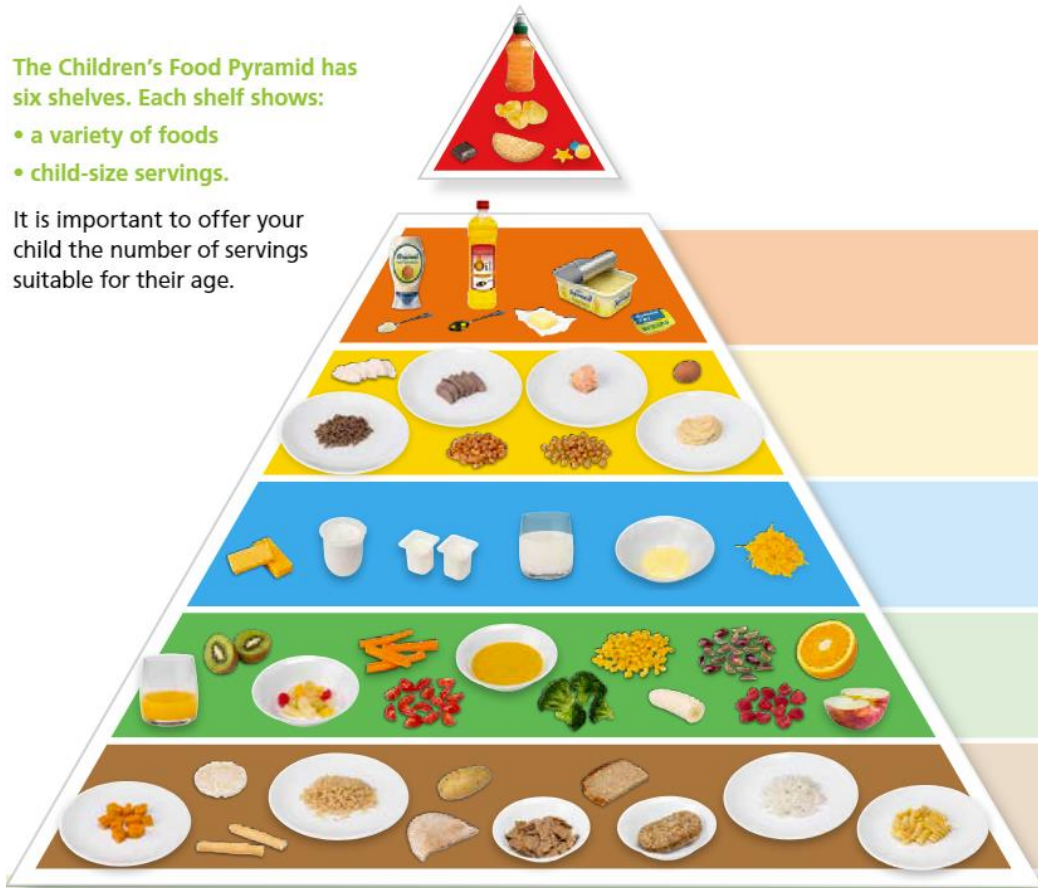
- Nutrients to focus on specifically during weaning
Iron, vitamin C, Calcium, vitamin D and Omega 3

Healthy eating guidelines

The Children's Food Pyramid has six shelves. Each shelf shows:

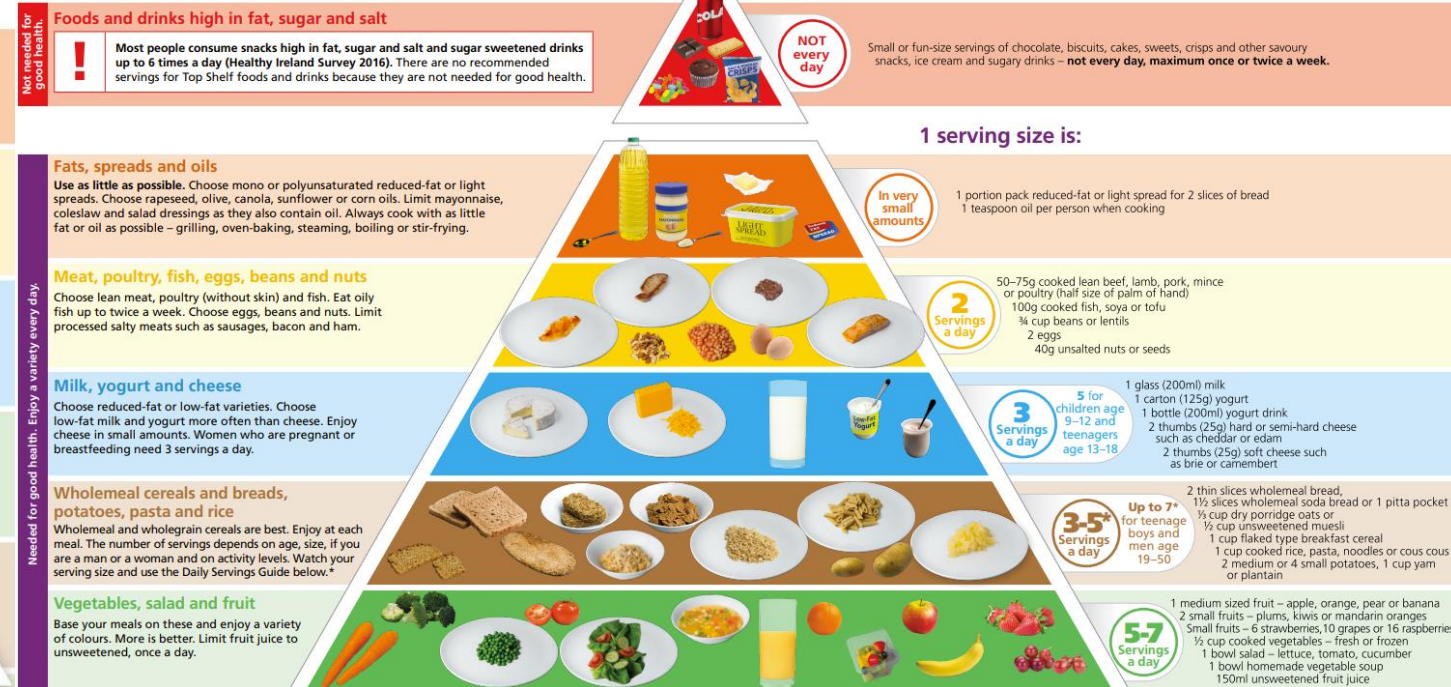
- a variety of foods
- child-size servings.

It is important to offer your child the number of servings suitable for their age.



The Food Pyramid

For adults, teenagers and children aged five and over



www.healthyireland.ie





| Ages one and two | Ages three and four |
|-----------------------|-----------------------|
| In very small amounts | In very small amounts |
| 2 Servings a day | 3-4 Servings a day |
| 3 Servings a day | 3 Servings a day |
| 2-3 Servings a day | 4-5 Servings a day |
| 3-4 Servings a day | 4-6 Servings a day |

Servings

1 and 2 year olds
3 servings a day

3 and 4 year olds
3 servings a day


Breastmilk counts towards these servings. Breastfeeding mothers can add cow's milk to their child's cereal or offer cow's milk as a drink. It is important to include yogurt and cheese also to help meet your child's nutritional needs.

Each of these examples shows one serving

- 200ml or 2x100ml plain milk or milk with added vitamins
- 200ml or 2x100ml unsweetened soya 'milk' fortified with calcium
- 2 adult thumbs of cheese
- 1 pot (125g) plain yogurt
- 2 small pots (47g) plain or natural fromage frai
- Homemade custard or rice pudding made with 200ml of milk

Servings can be split during the day between a meal and a snack. 1 serving can combine two foods or drinks. For example, 1 serving could be 100ml of milk with 1 small pot fromage frai or 1 thumb of cheese.

Your child can start drinking from a cup or a beaker when they are between 1 and 2 years old.



Servings


1 and 2 year olds
3 or 4 servings a day

3 and 4 year olds
4 to 6 servings a day

Each of these examples shows one serving

- 1/2 cup (30g) flaked cereal fortified with iron
- 1-1 1/2 wheat biscuits
- 1 plain rice cake
- 1-2 crackers
- 1/2-1 slice bread
- 1/2-1 small roll
- 1/2 pitta pocket
- 1/2 small wrap
- 1/2 chapatti
- 1 unsalted breadstick
- 1/2-1 small cooked potato
- 1/2 cup (30-40g) cooked sweet potato or yam
- 1/2 cup (30-40g) cooked pasta
- 1/2 cup (30-40g) cooked rice
- 1/2 cup (30-40g) cooked noodles
- 1/2 cup (30-40g) cooked couscous

Serving guide
Adjust these servings to suit your child. Smaller, younger children will eat less. Taller, older and more active children will eat more.



Servings


1 and 2 year olds
2 servings a day

3 and 4 year olds
3 or 4 servings a day

Each of these examples shows one serving

- 30g cooked beef
- 30g cooked lamb
- 30g cooked chicken or turkey
- 30g cooked salmon
- 1/4 cup (40g) baked beans
- 1/4 cup (40g) lentils
- 1/4 cup (40g) peas
- 1/4 cup (40g) chickpeas
- 35g hummus
- 35g cooked portion tofu
- 2 falafels
- 1 medium egg
- 1 heaped teaspoon smooth peanut or nut butter

Serving Guide
An average serving size of cooked beef, lamb, pork, chicken, turkey or fish is about 30g. A serving size that fits into one third the palm of your hand is about right for children aged 1 to 4.



Servings

1 and 2 year olds
2 or 3 servings a day

3 and 4 year olds
4 or 5 servings a day

Each of these examples shows one serving

- 1 plum
- 1 kiwi
- 1 mandarin
- 100ml unsweetened orange juice
- 1/4 orange
- 1/4 apple
- 1/4 pear
- 1/4 banana
- 3-4 cooked carrot sticks
- 3-4 cucumber sticks
- 5-6 berries cut in halves or quarters
- 5-6 grapes cut in quarters
- 3-4 cherry tomatoes cut in quarters
- 1/2 cup (30-40g) tinned fruit in natural juice
- 1/2 cup (30-40g) cooked, fresh or frozen vegetables
- 100ml homemade or readymade vegetable soup

Serving guide
An average serving size is about 40g. A serving size that fits into half the palm of your hand is about right for children aged 1 to 4.



Adjust these servings to suit your child – some children will take a little more and others a little less.

Fats, spreads and oils



Fats, spreads and oils provide essential fats in your child's diet. Only use very small amounts of these foods.

What is a small amount?

- 1 teaspoon of spread on bread
- 1/2 teaspoon oil in cooking.

Healthy eating habits

- Frying food is not recommended for young children.
- Choose mono or poly-unsaturated spreads. Choose rapeseed, olive, canola, sunflower or corn oils.
- Limit mayonnaise, coleslaw and salad dressings as they also contain oil.
- Always cook with as little fat or oil as possible.
- Try to:
 - grill
 - oven-bake
 - steam
 - boil
 - stir-fry.

- *Examples of suitable finger foods for infant learning to feed themselves?*
- Finger Foods (6-9 months)
 - Food should be soft, manageable sized pieces
 - No pips, stones, tough skins, stringy bits or bones.
 - Peel fruit / vegetables with skins for example cucumber, apple, pear
 - Offer a wide variety of tastes and textures
 - Fruit: Berries, avocado, banana, pear, grapes (cut in quarters, lengthwise), apples (cut in thin slices) apricot
 - Vegetables: cooked vegetables (broccoli, carrots, green beans, peppers, peas, courgettes, cucumber)
 - Protein: grated cheese, hardboiled egg, butter beans, meat, flaked fish, chickpeas, slices of omelette
 - Carbohydrates: pasta, breadsticks, fingers of toast

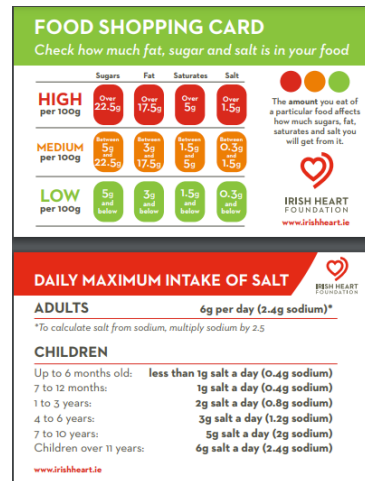
- *What commercially available products to avoid?*

Commercial products don't necessarily need to be avoided, they need to be healthy – read the food label and the ingredients list.

Food label:

Use the 3-6-5 rule to interpret the food label - a healthy product should meet or be as close as possible to the 3-6 -5 rule

| Nutrition Information | |
|-----------------------|---|
| | per 100g |
| 3 | < 3g Total Fat |
| 6 | >6g Fibre |
| 5 | <5g Carbohydrates of Which Sugars |
| | |



Ingredients list:

Ingredients are listed in order of descending weight.

The ingredients the food contains more of are at the top of the list and less of are listed at the end of the list.

If sugar or fat is close to the top of the ingredients list, this food is not a very healthy choice.

Some types of foods may be given different names in the ingredients list:

Sugar may be labelled as sucrose, glucose, glucose syrup, fructose, glucosefructose syrup, maltose, malto-dextrin, invert sugar, golden syrup, maple syrup, molasses or honey.

Fat may be labelled as butter, milk fat, cream, vegetable oil, vegetable fat, nut oil, monoglycerides, trans fats or hydrogenated fats. Try to avoid trans and hydrogenated fats and choose foods higher in monounsaturated or polyunsaturated fats instead.

- *What is the best way to reduce my child's portions?*



- How is your child growing, does the portion size need to be reduced?
- Use a smaller plate
- Keep the portion size the same but change the food
- Ensure the plate of food looks the same - overall volume of food on the plate is the same. For example if you want to reduce the portion of pasta then increase the portion of salad/veg/fruit on the plate

- *What is the portion of porridge for a 5 year old child?*

- 1/3 of a cup of raw porridge oats



- *If child doesn't eat what is provided should an alternative be offered?*
 - Division of responsibility: Parents / caregivers provide leadership on “the what, when and where” and the child or young person will decide "how much and whether” to eat.
 - Don't offer an alternative meal but always try to include 1 food in the meal that your child will eat
- *If a child doesn't like texture of food e.g steamed broccoli should it be offered up again and again or if how it is cooked be changed e.g in an omelette?*

If a food is refused, continue to offer the food as it may take 15-20 exposures for your child to accept the new food. You can change how the food is presented but don't hide the food.

| | | | |
|--|--|--|--|
| | | EATING | |
| | | <ul style="list-style-type: none"> ☐ Chews and swallows whole foods independently ☐ Chews, swallows whole foods with drink ☐ Chews, swallows some and spits some ☐ Takes, chews "a" times & spits out ☐ Takes pieces, holds in mouth for "a" seconds & spits out ☐ Takes off piece & spits out immediately ☐ Full tongue lick ☐ Licks lip or teeth | |
| | | TASTE | |
| | | <ul style="list-style-type: none"> ☐ Tip of tongue, tip of tongue ☐ Teeth ☐ Lip ☐ Nose, underneath nose ☐ Chin, cheek ☐ Top of head ☐ Chest, neck ☐ Arm, shoulder ☐ Outside hand ☐ Fingertips, fingertips ☐ Side finger tip | |
| | | TOUCH | |
| | | <ul style="list-style-type: none"> ☐ Reaches down or sticks up to smell ☐ Color in child's forward space ☐ Color at table ☐ Color in room | |
| | | SMELL | |
| | | <ul style="list-style-type: none"> ☐ Uses utensils or a container to stir or pour food/drink outside of own space ☐ Uses utensils or a container to stir or pour food/drink for others ☐ Assists in preparation/set up with food | |
| | | INTERACT WITH | |
| | | <ul style="list-style-type: none"> ☐ Looks at food when directly in child's space ☐ Being at the table with the food just outside of child's space ☐ Being at the table with the food in way across the table ☐ Being at the table with the food on the other side of the table ☐ Being in the same room | |
| | | TOLERATES | |

- *My child likes trying new food but won't give up what she already gets. Please advise on how to change meal plan?*

- How is your child growing, how are the energy levels, is there room to introduce a new food?
- Remember, if something new is introduced it can cause stress & anxiety if it needs to be removed.
- Depends on the age of your child
- Younger children, if there is a new food that you would like them to include in their diet then include it and reduce the portion of their existing food.
- Plan for introducing the new food, work with your child, agree the day and time and amount of the new food and agree that on that day the portion of their existing meal /snack will be reduced

Snacks

- *What are your top 5 snack options for sustenance and slow release of energy?*

Fresh fruit

Raw veg

Hummous/ low fat cheese / nut butters / sliced meat / hardboiled egg + wholemeal pitta bread fingers / oat bread/ breadsticks/ oatcakes / wholewheat cracker

Roasted chickpeas

Homemade smoothie

- *What commercial cereal do you like (if any) for our kids?*

Porridge / overnight oats, readybrek, Weetabix

- *Should I switch to low fat milk at 2 years of age?*

Suitable milk for children

Full fat up to 2 years

Low fat from 2 – 5 years

Fat free / skimmed from 5 years

Limit of 500mls of milk per day



- *How much milk should a 6year old with PWS have daily?*
 - No more than 500mls/day

Recommended daily vitamin D supplements

| | Daily requirement | When to take |
|--|---------------------------|---|
|  Infants 0–12 months | 5 micrograms (µg)/200 IU | All year round if breastfed exclusively <u>or</u> if taking less than 300ml of infant formula per day Exception: Infant formula has added vitamin D so if a baby drinks more than 300ml of formula, they do not need a vitamin D supplement. |
|  Children 1–4 years | 5 micrograms (µg)/200 IU | During the winter months (Halloween to St Patrick's day) |
|  Children 5–12 years | 10 micrograms (µg)/400 IU | During the winter months (Halloween to St Patrick's day) <u>or</u> all year round for those with reduced sun exposure or darker skin tone |
|  Teenagers and adults | 15 micrograms (µg)/600 IU | During the winter months (Halloween to St Patrick's day) <u>or</u> all year round for those with reduced sun exposure or darker skin tone and every day during pregnancy |
|  Older adults Over 65 years | 15 micrograms (µg)/600 IU | All year round |

- *Should we be changing the diet for start of teen years?*

From 11 years – increased nutritional requirements for certain nutrients

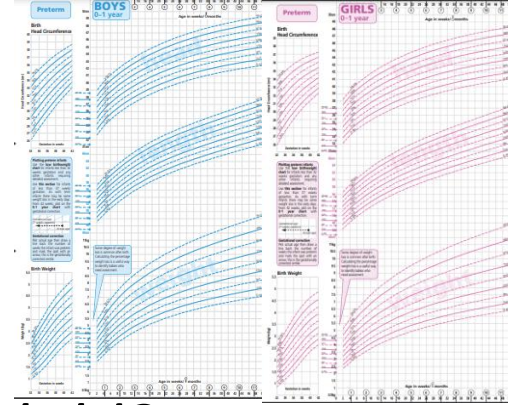
Multivitamin containing iron

Separate Calcium + vitamin D supplement (depending on vitamin D status)

Ensure nutrient dense diet (40-60% energy requirements of age matched peers)

| | Girl 7-10 years | Girl 11-14years |
|-----------|---------------------|----------------------|
| Calcium | 800mg (3 portions) | 1150mg (5 portions) |
| Vitamin D | 600IU | 600IU |
| Iron | 11mg | 13mg |
| Vitamin C | 45mg | 70mg |
| Fibre | 16g | 19g |

Energy Balance & Growth



- *How to work out the calorie count for my child?*

6-8 kJals/cm for weight loss

10-12KJals/cm for weight maintenance

Typically for children and young people with PWS the daily energy requirement depends on :

How your child is growing (growth trajectory)

How your child is growing with their current dietary intake

How your child is presenting (tiredness, energy levels, daytime sleepiness, developmental milestones).

Changes in energy demands at different stages (eg transitioning from tube feeding to oral feeding, starting to walk, increased movement or physical activity levels eg intensive physio etc)

- *Child who was on the same amount of calories as he was this time last year, he wasn't walking then but is now. His energy levels vary, should we increase his intake to try maintain energy levels?*

Check how your son is growing (growth centile charts)

Check his recent blood test results (FBC, ferritin and thyroid function)

? Increase his intake appropriately (transparently so if you need to reduce the intake at another stage this can be done without causing anxiety or stress for your child eg if necessary change from low fat to full fat products, add extra calories when cooking a meal so you are not adding extra food on the plate but you are making the meal more energy dense – add olive oil, cheese, ground almonds etc)

- *Would a lack of calories potentially have an effect on ability to stave off infections?*

Potentially if the diet is not nutritionally adequate – (energy, protein, fibre, calcium, vitamin D, iron, vitamin C)

Aim to get a balance of healthy nutritional foods



- *Child has bloating but no constipation. Have been advised to cut back on beans in diet. He has 4 servings of a variety of beans a week. Any suggestions on alternatives or other foods to watch? ? What are your thoughts on baked beans for our kiddos?*

Baked beans: fine to include as part of a balanced diet (good source of protein and fibre)

Alternatives for protein: meat, fish, eggs

Alternatives for fibre: wholegrain bread or cereals, fruit and veg with the skins on

Eat a well balanced diet – ?excessive amount of fibre

Regular meal pattern – no long gaps between meals

Take time to sit down and relax while eating

Adequate fluid intake

Regular movement / exercise

Use a 7 day food and symptom diary – identify the triggers

Trial a probiotic - 4-6 wks / probiotic yogurt/ natural yogurt/probiotic drink / linseeds

Make time to relax – try kids yoga, massage



| Day | Breakfast | Mid Morning | Lunch | Afternoon | Dinner | Supper | Symptoms |
|-----|-----------|-------------|-------|-----------|--------|--------|----------|
| | | | | | | | |
| | | | | | | | |
| | | | | | | | |
| | | | | | | | |

- *Should we be strictly focusing on daily calorie count?*

This depends on the child, their growth and how they are presenting. Overall as a parent you should focus on trying to achieve a healthy balance of food in the diet that is appropriate for your child's age and stage.

- *When it comes to medication and supplements, should we be wary of ingredients when our children are on such strict diets?*

Medication: adhere to the guidance of the medical team

Supplements: on a restricted diet it will be necessary to use multivitamins to meet your child's full micronutrient requirements

A general multivitamin should be used except in cases where particular micronutrient deficiencies have been identified then single supplementation is indicated for example: iron, vitamin D and calcium

- *Child recently started Eltroxin. Was advised to avoid calcium for at least 2 hours after taking eltroxin in the morning (some advice is 4 hours). Tolerating weetabix and readybrek mixed with water. Any other ideas for adjusting breakfast or how to add in proteins other than adding nut butters/seeds.*

Non Dairy Calcium Sources

| Food group | Food | Calcium (mg) |
|-----------------------------|--|--------------|
| Fruit, vegetables & legumes | Black berries (70g) | 22.5 |
| | 1 large orange (200g) | 50 |
| | Dried apricots (x4/ 32g) | 23 |
| | Dried Figs (25 g) | 60 |
| | Broccoli (85g) | 34 |
| | Cooked spinach (50g) | 49.5 |
| | Okra (70g) | 60 |
| | Boiled lentils (3tbs/ 120g) | 26 |
| | Chickpeas (2tbs/ 70g) | 30 |
| | Peas (50g) | 12.5 |
| Protein sources | Baked Beans (50g) | 30.5 |
| | Kidney beans (50g) | 71.5 |
| | Nuts & seeds - <small>Nuts must be ground or as a butter for children under 5 due to choking risk</small> | |
| | Smooth peanut butter (25g) | 10.75 |
| | Tahini (25g) | 106.5 |
| | Sesame seeds (1 tbsp / 12g) | 80 |
| | Fish | |
| | ½ tin sardines (60g) | 258 |
| | ½ tin salmon (with bones (52g) | 147 |
| | 1 tin mackerel (85g) | 250 |
| Eggs Tofu | Fish fingers (112g) | 103 |
| | Fish cakes (81g) | 89 |
| | Prawns (100g) | 110 |
| | 1 large egg | 35 |
| | Tofu (50g) | 175 |
| | Bread & cereal foods | |
| | Ready Brek (30g) | 402 |
| | 30g fortified mixed whole grain cereal with 125ml of fortified dairy free 'milk' | 303 |
| | 1 slice brown bread (40g) | 74 |
| | 1 slice granary bread (40g) | 84 |
| | 1 slice white bread (40g) | 71 |
| | 1 seeded burger bun (90g) | 117 |

- Weetabix / Porridge / Oatibix ? Readybrek + water/dairyfree unfortified drinks eg Flahavans organic oat drink
- Pancakes made using oats + dairyfree unfortified drinks/ Readybrek
- Eggs poached/scrambled/in a cup + wholegrain bread + fruit
- Beans + wholegrain bread + fruit

To conclude

- Work with your medical team to monitor your child's growth
- Dietary intake may need to be altered to support appropriate growth for your child (↑/↓ daily energy intake)
- Consider a general multivitamin supplement if on a very restricted diet. (key micronutrients calcium + vitamin D, iron and vitamin C)
- Food security- keep your child safe across all environments
No Doubt, No Hope, No Disappointment
- You are the expert in your child!

Resources

- Healthy Eating Guidelines for 1 to 4 year olds:

[https://www.safefood.net/Healthy-Eating/Childrens-Food-Pyramid-\(1\)](https://www.safefood.net/Healthy-Eating/Childrens-Food-Pyramid-(1))

<https://www.gov.ie/pdf/?file=https://assets.gov.ie/93351/bc5bec36-1748-4b0e-b356-300a25f018d5.pdf#page=null>

- Healthy Eating Guidelines for Adults, teenagers and children over 5 years old

<https://www.safefood.net/healthy-eating/guidelines/food-pyramid>

- Safe Food Portion Cup:

<https://orders.safefood.net/products/sfc033?variant=40821217951827>

- Food Shopping Card:

<https://irishheart.ie/publications/food-shopping-card/>