



## ***Advice on Nutritional Problems for Patients with Fanconi Anaemia***

Many patients with Fanconi Anaemia [FA] experience nutritional problems such as poor appetite, poor weight gain, early satiety, nausea, diarrhoea and/or constipation and in younger children there be an element of feeding behaviour problems. There is great variation between patients with some patients able to eat an adequate diet and not require any nutritional intervention whilst others may require calorie supplements, nutritionally complete sip feeds or in some cases tube feeding either by a nasogastric (a small fine bore tube inserted via the nose and goes down the food pipe [oesophagus] into the stomach) or a gastrostomy (a tube inserted directly into the stomach).

There are separate fact sheets with this leaflet which give specific advice on poor appetite, poor weight gain, nausea and vomiting, constipation, diarrhoea and food refusal/feeding behaviour problems. You can just print off the specific ones you feel would be useful for you.

As well as ensuring an adequate diet to maximise growth in children with FA and weight gain, it is important to eat a well balanced diet which includes plenty of fruit and vegetables. This is for two main reasons. Firstly FA patients are known to have an increased tendency to certain types of cancers. Secondly FA is a disorder in which the mechanisms protecting the body against oxidative injury are impaired and there is evidence of increased oxidative stress in FA. Fruit and vegetables are naturally good source of antioxidants and therefore a good intake may help to reduce oxidative stress.

It may be difficult to increase your fruit and vegetable intake when your appetite is generally poor, you feel full after eating or experience nausea. One of the fact sheets included gives hints on how to include more fruit and vegetable in the diet. There is also an information sheet discussing dietary antioxidant supplements which you may have read or heard about in relation to FA.

Patients with FA are more prone to osteoporosis (brittle bones) and therefore there is a fact sheet giving information on good sources of vitamin D and calcium in the diet.

A good model to base a healthy diet on is the "The Balance of Good Health" (The Food Standards Agency) plate model given below. Eating a combination of foods from all the food groups will give you all the nutrients your body needs (energy, protein, vitamins and minerals).

# The eatwell plate

Use the eatwell plate to help you get the balance right. It shows how much of what you eat should come from each food group.



**Fruit and Vegetables** – Always have fruit and vegetables at every meal and aim for at least 5 servings a day

**Bread, rice, potatoes, pasta and cereals** – Have at least 1 food from this group with each meal

**Milk and dairy foods** – Have at least 3 servings of milk, yoghurt or cheese each day

**Foods high in fat and sugar** – Individuals who have a good appetite and a weight within the normal range for their height should eat these foods in small amounts. They are high in calories but have very few nutrients. However for those who have a poor appetite and weight gain they are a useful way of helping to increase your calorie intake without having to eat a lot of extra foods. When using fats and oils choose some with omega 3 fat such as olive oil, rapeseed oil or walnut oil.

**Meat, fish, eggs, beans, dahl and nuts** – Have foods from this group twice a day

## Is there anything which should be avoided?

FA patients should avoid the consumption of alcohol (this includes alcohol containing mouthwashes) as this has been linked to an increased risk of oral cancers in FA patients.

## Dietary Antioxidants and Fanconi Anaemia

Oxidative stress is caused by an imbalance between the production of substances called reactive oxygen species, which cause damage to the cells in the body, and the body's ability to detoxify them and repair the resulting damage. There is a relationship between oxidative stress and FA patients being more prone to certain types of cancers. Laboratory evidence also suggests that increased oxidative stress in FA may trigger certain stress pathways which may contribute to the progressive bone marrow failure in FA. Therefore the role of certain antioxidants providing protection from this increased oxidative stress has been suggested.

Antioxidants are important constituents of the diet and are involved in DNA and cell maintenance and repair. They reduce the production of free radical oxidants which are involved in oxidative stress by reacting with them and eliminating them. Antioxidants in the diet may be in the form of vitamins or minerals, such as vitamins C, vitamin E, beta-carotene and selenium or they may be found in flavonoids in vegetables.

You may have read that certain dietary antioxidants, such as quercetin, rutin, alpha- lipoic acid or resveratrol, may be beneficial to FA patients as they are known to reduce the production of one of the stress pathway triggers (TNF-alpha). However although in theory they would be beneficial to FA patients until controlled clinical studies are done or more is known about their safety it is difficult to make any recommendations. **It is important that you discuss with your consultant first if you are considering using any dietary antioxidants.**

Some points you may wish to consider first include:-

- Data on doses and safety, particularly in children is lacking and because of this generally the manufacturers do not recommend their use in children under 12 years or 18 years of age in some cases
- Generally data regarding the safety on long- term use of high doses or specific antioxidants is lacking, bearing in mind FA is a life long condition
- At certain doses or in certain clinical situations some antioxidants can be toxic or convert and exert the opposite (a pro-oxidant) effect instead of acting as an antioxidant. For example it is well documented that high doses of vitamin C can have a pro-oxidant effect in certain situations
- The quality of commercially available supplements can vary between manufacturers
- Generally most of the studies supporting supplementation of a single antioxidant supplement is based on lab or animal studies but such studies do not always show the same effects when studied in humans
- The effect of taking the antioxidant as a food may have added benefit due to the mixture between it and other antioxidants and compounds in the diet
- It is not known how high dose antioxidants react to other medications you may be taking

## **Food sources of the antioxidants quercetin, rutin, alpha-lipoic acid and resveratrol**

Fruit and vegetables are good sources of antioxidants and antioxidant vitamins and it is recommended to try to include 5 portions of fruit and vegetables daily. An advice sheet on giving hints on how to include more fruit and vegetables in the diet is included. However below is list of foods which are good sources of the above mentioned antioxidants which you may wish to try to include in your diet.

### **Quercetin**

Onions (especially red), apples, red grapes, citrus fruits, broccoli and other green leafy vegetables, raspberries and blueberries

### **Rutin**

Citrus fruits, tomato, rhubarb and buckwheat

### **Alpha-lipoic acid**

Dark green leafy vegetables e.g spinach, kale, okra, spring greens, broccoli

### **Resveratrol**

Red grapes (highest concentration is in the skin), peanuts, blueberries

## **What about a general multi-vitamin supplement?**

- FA patients who are eating well and include a good variety of foods in their diet should not need to take a vitamin supplement
- FA patients currently on tube feeds or nutritionally complete sip feeds should not need an additional vitamin supplement as their requirements will be met by the tube feed or sip feed
- FA patients not on any additional feeds or sip feeds and only eat a limited variety of foods may benefit from an additional multivitamin supplement. It is however preferable to try to get the vitamins and minerals you need from food. See the section on how to increase fruit vegetables in your diet for hints
- Vitamins in excess can be toxic therefore do not take a megadose of a single vitamin or combination of vitamins
- A general age appropriate supplement which does not give above the daily recommended amount [RDA] would be suitable to use. Ideally long term supplement use is not recommended
- Patients with FA who are receiving blood transfusions will receive iron from this and therefore do not need an additional source hence we would advise a supplement which does not contain any iron or only a small amount. [no more than 15% RDA]
- It may be that your doctor or dietitian has recommended a specific supplement due to the level of a particular vitamin or mineral being low in your blood e.g. Vitamin D
- If you are considering using a general multivitamin supplement please discuss this your doctor or dietitian first

## Fruit and Vegetable Ideas

The following gives some ideas on how to include more fruit and vegetables into the diet. Fruit and vegetables are naturally good sources of both antioxidants and antioxidant vitamins.

- You can choose from fresh, frozen, tinned, dried or juiced
- Frozen vegetables and frozen berry fruits are good to use as they are easily incorporated into dishes and always there if you are in hurry. They are just as good as fresh, do not go off, which means you get more for your money
- Add vegetables, especially onions, to soups, stews, curries and pasta dishes. Cut into small pieces they are not so readily noticed
- Tinned tomatoes are easy to disguise and can be added to pasta sauces
- Tomato puree can be added to soup, pasta dishes and curries
- Mix some mashed carrot, swede or turnip into mashed potato
- As well as roast potatoes why not try roast parsnip and carrots
- Try serving some stir fried vegetables (onion, mushroom, baby corn, peppers, spinach, sugar snap peas) with rice
- Add extra vegetables and fruit to pizzas (tomato, sweetcorn, pineapple, peppers, mushrooms)
- Add crunch to your sandwich with lettuce, tomato, cucumber, radishes or grated carrot. Children often prefer cherry tomatoes as they are sweeter and are also easy to include in lunchboxes
- A glass of fresh fruit juice will count as one of your 5-a-day
- Try sticks of carrot, celery, cucumber or pepper dipped into a salsa or guacamole dip

- Include an apple or pear as a snack
- Do not throw away over ripe fruit – use them to make smoothies
- Make ice lollies by pouring fruit smoothies, 100% fruit juice or pureed fresh fruit into ice lolly moulds and freezing them
- Add tinned, fresh or stewed fruit to milk puddings, jelly, ice cream or yoghurts
- Most children like grapes so offer them as snacks, in lunch boxes and try adding to jelly
- Try fruit pies and crumbles
- Add fruit or dried fruit to breakfast cereals
- Buy fruit that is in season to save cost. Make up a bowl of brightly coloured fruit and put it in an area you or your child walk through a lot so it is appealing for them to pick up
- Let your child feel involved by choosing what vegetable topping to put on pizza or let them help you make a fruit salad
- Keep trying to encourage your child to eat more fruit and vegetables. It can take up to as many as 17 attempts to get a child to try something new
- Consider trying a reward or sticker chart to encourage your child to eat more fruit and vegetables
- At family meal times, preferably sitting around your dinner table, make sure you eat some fruit and vegetables yourself so your child sees that they are not horrible. If they see you eating and enjoying fruit and vegetables hopefully they will want to try some as well
- Try making fruit smoothies  
Chopped fruit e.g banana, strawberries, raspberries, blueberries (if not in season try defrosted frozen berries), pineapple, peaches (fresh or tinned) or mango  
100-150g natural yoghurt  
Add extra ice cream or cream if desired

Put all the ingredients in a liquidiser or blender and puree to a smooth consistency

Put in the fridge and serve chilled

## Osteoporosis and Fanconi Anaemia

Patients with FA are at increased risk of developing osteoporosis (brittle bones with a risk of fractures). This is due to several factors which will vary from patient to patient. Maximising peak bone mass (maximum bone strength) during skeletal growth in infancy, childhood and adolescence is considered to be important for the primary prevention of osteoporosis however this is not always achievable in children with FA due to problems with growth. Other risk factors include:

- Gut abnormalities resulting in a reduced absorption of calcium
- Poor dietary intake of calcium and vitamin D
- Being underweight (in adults a body mass index (BMI) <19)
- Reduced exposure to sun. Patients with FA are recommended not to have over exposure to sunlight and to use a high factor sun block. This recommendation is important to adhere to due to the increased risk of skin cancers. Vitamin D, which is needed to absorb calcium from our diet, is obtained from the action of sunlight on the skin. However, there are also foods which are good sources of vitamin D
- Some patients with kidney problems may be unable to convert vitamin D to its active form a process which takes place in the kidneys
- In woman low levels of the female hormone oestrogen
- In men low levels of the male hormone testosterone
- Patients who have undergone a bone marrow transplant who required a prolonged period of time on corticosteroids ( e.g. prednisolone) for management of graft versus host disease
- Patients who have undergone a bone marrow transplant who had a prolonged period of gut graft versus host disease resulting in problems with malabsorption

Calcium is vital for bone health and vitamin D is needed to absorb the calcium from our diets and therefore it is important to ensure an adequate intake of both nutrients.

FA patients who are on nutritionally complete sip feeds or tube feeds will obtain both vitamin D and calcium from the feed and your dietitian will be able to check if this is adequate.

Some patients considered more at risk, those who have a low level of vitamin D in their blood or those who are unable to obtain a good intake of vitamin D and calcium from food should be prescribed a vitamin D and calcium supplement. Your doctor will be able to advise you.

## Foods High in Vitamin D and Calcium

### Vitamin D

To absorb calcium from our diet we need vitamin D. Some vitamin D comes from our diet. Sources include:

- Margarine and fat spreads (fortified by law with vitamin D)
- Many breakfast cereals or fortified with vitamin D
- Oily fish like mackerel, fresh tuna, sardines, herring, pilchards
- Eggs
- Full fat dairy foods (if using reduced fat dairy products check that they have vitamin D added)





- Try to make meals interesting e.g. colourful, well arranged on the plate
- Avoid having drinks 1 hour before a meal so as you or your child is not too full to eat. Have drinks after meals and snacks
- Avoid rich fatty food. It may make you feel nauseous if your appetite is poor
- Never “force feed” a child. Give lots of encouragement and praise even if the amount eaten is small
- Encourage toddlers and young children to feed themselves if they are not doing so already
- Eat with your child or let them eat with their peers or siblings and try to make meal times enjoyable social occasions

### **Advice on how to increase the protein and calorie intake**

It is important to include well, balanced meals everyday for growth, repair and energy. Refer to page 2 for advice on what a balance meal should include.

Here are some ideas that will help you increase the protein and calorie content of meals and snacks.

- Add butter/margarine (not reduced fat) to bread, toast, crackers, mashed potato, vegetables, teacakes, crumpets, scones or chapatti. From a general health point it is preferable to use a fat spread high in polyunsaturated fats (e.g. sunflower oil based) or monounsaturated fats (e.g. olive oil based). They contain the same amount of calories as butter and other margarines but are a healthier type of fat
- Add cheese to sandwiches, toast and crackers. Add grated cheese to potatoes, soups, pasta dishes, pizza, tinned spaghetti and baked beans
- Full fat dairy products. Use full fat milk for puddings, cereals, porridge and added to drinks. Use full fat yoghurts, fromage frais, mousses and blancmanges
- Serve cream, ice cream or custard with cake, desserts and fruit
- Add cream, sour cream, plain yoghurt or crème fraiche to sauces, soups and meat dishes
- Add jam, marmalade, lemon curd, honey (not suitable for children under 1 year of age) or peanut butter (do not give foods containing peanuts to children under 3 years if their parents or siblings have allergy, eczema, asthma or hay fever) to bread, toast, teacakes and scones
- Try full fat milk flavoured with milk shake mix, ready made yoghurt style drinks or smoothies

These suggestions are intended as short term measures to help you or your child gain weight. If find your child needs to continue this advice longer term please request further information from your dietitian or ask to be referred to a dietitian if you are currently not seeing one.

Some patients may already have been advised to use proprietary calorie supplements, sip feeds or may be receiving tube feeds to help with their nutrition and weight gain. It is important to continue with your current supplements or feeds and to receive regular review from your dietitian.

## What If My Child Won't Eat?

### Food refusal and feeding behaviour advice

Many children go through phases of refusing to eat certain foods or at times refusing to eat anything at all. This is particularly common in children up to the age of 5 years, but may occur at any time and is a normal part of growing up. Feeding behaviour problems are common in children with FA due to other underlying problems of poor appetite, gastrointestinal problems and naturally parents of children with FA are anxious to ensure their child has a good food intake to promote weight gain and growth. Below are a few tips to try to help your child to eat.

- Establish regular meal and snack times, as this is better than letting your child “pick” throughout the day.

Breakfast	Mid-afternoon snack
Mid-morning snack	Evening meal
Lunch	Supper (optional)
- Always offer 2 courses at a meal, one savoury and one sweet
- Eat at the same time as your child if possible, as they will learn from you. Try to make meals enjoyable social occasions. At nursery and school ensure that they are eating with their peers and not on their own
- Sit together at the table. Use brightly coloured plates and cups. Present food in fun and attractive ways
- Try to eat in a calm, relaxed area, without the television or toys. Children are easily distracted
- Try not to rush a meal, as your child may be slow to eat, yet try not to let meals drag on too long. A maximum of 30 minutes is about right
- Avoid giving drinks 1 hour before a meal so as your child is not too full to eat. Offer food first and drinks at the end of the meal

- Encourage toddlers and young children to feed themselves if they are not already doing so. Offer finger foods e.g. fish fingers, sandwiches, crackers, vegetable or fruit slices, slices of chicken or ham, chips. Cut food into small pieces
- Encourage your child to use their fingers to play with food. Don't worry about the mess
- Milk is a nutritious drink but should not replace meals. Be careful not to let your child drink lots of milk in the day or at night
- Never force a child to eat. Although a very difficult thing to do, try not to show that you are anxious or annoyed by your child not eating. Try once to encourage them to eat a little more. If they don't want it take the plate away without comment
- Do not offer lots of alternatives if they won't eat what you have offered
- Try offering new foods along with known favourites. It can take up to 17 attempts to get a child to try a new food
- If they do eat a small amount or try a small amount of a new food give them lots of encouragement and praise
- If your child eats well offer a reward such as going to the park or spending some special time playing together. Do not use food though to reward or punish your child
- Let them watch or help you prepare food e.g. help to decorate buns, biscuits, put topping on pizza etc
- Picnics can be fun either outdoors in the garden or on the floor in your home!
- Invite special friends for a meal together

### Advice on Problems with Nausea and Vomiting

Patients with FA may at times experience problems with nausea and vomiting. The following gives advice on what foods to try and the best way to eat if nausea and vomiting are a problem.

- Try to eat small amounts throughout the day, little and often, rather than large meals
- Avoid drinking until at least 30 minutes after eating, Let the food go down first. A large gulp of juice with food may make you vomit
- Cold foods or foods served at room temperature may be better tolerated as they usually smell less than hot foods. Strong smells may make the nausea worse
- Dry foods can sometimes stay down better e.g. crackers, toast, plain biscuits or ginger biscuits
- Avoid fatty greasy foods. They can sometimes make nausea worse
- Sips of cold juices can sometimes help
- Older children and adults may find sucking boiled sweets, fruit sweets, mints or ice-lollies may help to take the edge of an nausea

## Advice on Problems with Diarrhoea and Constipation

Patients with FA may experience intermittent problems with constipation or diarrhoea or both. Both of which can affect appetite and food intake. Below is some advice which may help.

### Diarrhoea

- Try to drink plenty but avoid really cold drinks straight from the fridge which may make the diarrhoea worse
- Try to eat small amounts of foods and snacks instead of big meals
- Avoid foods high in fibre (roughage) e.g. dried fruit, nuts, beans and lentils
- Avoid any specific foods which seem to make the diarrhoea worse
- Look out for symptoms of dehydration. These include passing urine less often and passing small amounts of dark urine
- Let your doctor know if diarrhoea persists

### Constipation

- Try to eat foods high in fibre (roughage) e.g.
  - Wholemeal bread
  - Wholegrain cereals – Weetabix, Shreddies, Cheerios, Branflakes, Porridge
  - Fruit and vegetables
  - Dried fruit
  - Lentils and beans
- Make sure you drink plenty e.g. 6-8 cupfuls daily. Fibre absorbs water as it travels through the gut so if you increase your fibre intake it is important to make sure you drink plenty
- Let your doctor know if the constipation persists

## Contact Details

Please use this page to list details of your local hospital, consultant, nurse and dietitian

**Hospital**.....

Address.....

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Telephone .....

**Consultant**.....

Contact details.....

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**Specialist nurse**.....

Contact details .....

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**Community Nurse**.....

Contact details .....

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**Dietitian**.....

Contact details.....

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Leeds. 2009