An Information leaflet from Hereditary Fructose Intolerance (HFI) UK

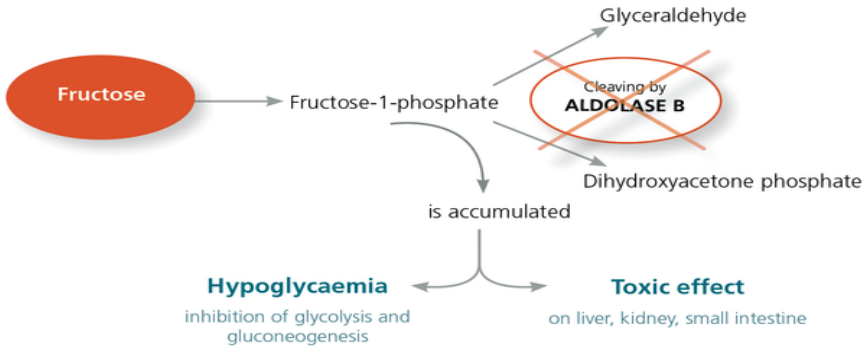
This information has been gathered from lived experience and has not yet been endorsed by a specialist paediatric team. Please consult your Metabolic Specialist.

What you need to know about the metabolic inherited disorder Hereditary Fructose Intolerance (HFI) and how to create a safe environment for a child with HFI in a school setting.

The disorder:

Hereditary fructose intolerance is an autosomal recessive disease caused by deficiency of the enzyme aldolase B. Aldolase B is found primarily in the liver, with some also in the kidneys and small intestine.

HFI manifests with abdominal pain and vomiting, symptomatic hypoglycaemia (which may induce seizures), low phosphate levels, acidosis, and other metabolic disturbances after consumption of foods and drinks containing fructose, sucrose or sorbitol.



It typically presents at weaning, when fatalities can occur. However, HFI may come to light at any age.

Diagnosis is by molecular analysis of the aldolase B gene by DNA test.

There is no cure as such for HFI, but with a strict, safe diet, avoiding fructose, sucrose and sorbitol, someone with the condition can live a normal, healthy lifespan.

A child with HFI might sometimes feel very tired or have tummy ache. These symptoms can all be usual for someone with the condition. It is also common to have diarrhoea after accidental ingestion of unsafe sugars. This is not the same as diarrhoea from pathogens.

Children and adults with HFI have a strong aversion to anything that tastes sweet. This is quite literally their life saver, stopping them from eating the foods that their bodies cannot metabolise. If a child with HFI refuses to eat something, under no circumstances make

them do so.

What is an HFI-safe diet like?

A diet for a child with HFI is not one of choice, but is a medical necessity.

It is imperative that a strict safe diet is followed for well-being and normal development. It is recommended that a child with HFI is supervised whenever food or drink is being consumed.

A safe diet for someone with HFI is completely counter intuitive to all the general “Healthy Diet” information that is recommended for most people.

All fruits, most vegetables (including onions, tomatoes and most potatoes), whole grains, honey, sucrose, sorbitol – all are **toxic** to someone with HFI and **MUST BE AVOIDED.**

The only safe and palatable drinks for a child with HFI are water or milk. **No flavoured drinks, juice or squash drink are to be given.**

The diet of a child with HFI will consist mainly of meat, fish, eggs, unsweetened dairy products, white rice, some white breads and pasta (needs to be sugar free), some lentils and pulses and **small** amounts of olives, mushrooms and dark green vegetables (e.g. spinach, kale – stalks removed).

These dietary restrictions ***MUST*** be followed and **ALL STAFF** need to be aware.

Practicalities in the classroom.

If a child with HFI is unwell, **NEVER** give Calpol. Always consult with the caregiver for advice.

In an emergency situation where a 999 call might have to be made, the attending paramedics must be made aware that a child has HFI, as many routinely given medications are unsafe.

Having reviewed the specific mealtime requirements for a child with HFI, there are other classroom scenarios which may pose problems.

A child with HFI will of course, want to be treated similarly to their peers. This needs careful consideration, and awareness raising is required for the other children, to understand that a child with HFI faces very specific challenges.
The child with HFI will probably be aware that their food preferences are different to their peers and it is all too common for them to try to ‘fit in’ by eating whatever their friends are eating, with no comprehension of the potential severity of their actions. This can be

particularly relevant at **'snack time'**.
Extra supervision and vigilance will be needed.

There are instances where foods that are unsafe for a child with HFI will be looked at and handled e.g. preparing and cooking foods in class. If a safe alternative can be found for the HFI child to be making, that is ideal e.g. making cheese biscuits instead of cookies, trying to ensure that there is no cross contamination with sugars during the process. Great care must be taken that the unsafe foods are ***not*** tasted or ingested by a child with HFI. If this does occur, please call the caregiver immediately for advice.

Food preparation in general will of course include careful handwashing beforehand. For a child with HFI, handwashing ***after*** handling foods containing fructose, sucrose or sorbitol needs to become routine, as does surface cleaning wherever toxic sugars have been used.

There are instances such as oral hygiene and tooth brushing that may be part of the class learning. Most toothpastes contain **sorbitol or other sugars,** and are **not safe** for a child with HFI.

If treats are brought in for birthdays, treats etc., (these are almost always sweet) please ask the caregiver what may be suitable to give to a child with HFI beforehand.

It is recommended that discussions take place with the child's caregiver prior to any activities that may prove unsafe. If unsure, please *always* speak to the child's parent/caregiver.

For further information please visit:

Hereditary Fructose Intolerance (HFI) UK

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