Dietary factsheet for people living with HNF1B-associated disease

HNF1B-associated disease is caused by a change, like a spelling mistake (often referred to as a 'variant'), in one copy of the *HNF1B* gene or a missing copy of the *HNF1B* gene. Because this gene helps your kidneys, pancreas, and other organs develop and function, or work properly, changes in this gene can affect how these organs work. Each person with HNF1B-associated disease may be affected differently. Most people do not need a special diet. A healthy balanced diet helps support your kidneys, pancreas and overall health.

How HNF1B-associated disease can affect you

Not everyone with HNF1B-associated disease has the same symptoms; some people have only a few health issues while others have more. These can include:

- Kidney changes – such as cysts or only having one kidney
- Diabetes – this can happen because the pancreas, which normally helps control blood sugar, can be underdeveloped (i.e. small)
- Low magnesium – this happens because the kidneys lose too much magnesium in the urine
- Abnormal liver blood tests – this is common and usually nothing to worry about
- Gout – pain or swelling in the joints
- Pancreatic insufficiency – problems digesting food, which can cause bowel symptoms and make it harder to keep your weight stable
- Learning or attention differences – linked to autism or ADHD. This is usually only seen in people who are missing one copy of the *HNF1B* gene, but not everyone is affected.

Dietary advice for HNF1B-associated disease

The condition affects people in different ways. This means everyone's needs may be slightly different. However, most people with HNF1B-associated disease can follow the same healthy eating pattern recommended for the general population.

You might find lots of information online about eating for diabetes or kidney health. Sometimes this advice can sound confusing or even conflicting. For example, one source may suggest eating more of a certain food, while another says to avoid it. This is often because much of the online information is out of date, not specific to the condition or not specific to your individual needs.

This factsheet explains how to eat well if you have HNF1B-associated disease. It focuses on a healthy way of eating that supports diabetes, kidney health and overall health.

What to include?

Overall the advice is to eat a healthy eating pattern of regular meals that include the following foods:

- Vegetables and fruit – aim for at least 5 portions each day (for example, 3 vegetables and 2 fruit), and more if you can
- Wholegrains and starchy foods – such as potatoes, wholemeal bread, brown rice, oats, or wholegrain pasta
- Lean proteins and plant proteins – such as beans, lentils, chickpeas, eggs, fish, or lean meat
- Small amounts of healthy fats – olive oil, avocado, nuts, and seeds
- Drink mostly water.

What to reduce?

- Limit salt, salty foods, sugary foods and drinks, foods high in saturated fat, and ultra-processed foods*
- Eat less red and processed meat
- Limit alcohol, especially beer and spirits, as these can increase the risk of gout attacks. The UK recommendation is no more than **14 units per week**, spread across several days.

***Ultra-processed foods** are foods that have been heavily processed and often contain added ingredients such as sugar, salt, fats, flavourings, colours, preservatives and emulsifiers. *Examples include crisps, sweets, sugary drinks, instant noodles, packaged snacks, ready meals, processed meats (such as sausages or chicken nuggets), and shop-bought cakes and biscuits.*

Eating well for diabetes

Many people with HNF1B-associated disease have diabetes; this is because the pancreas, which helps control blood sugar, can be affected. The good news is **you do not need to follow a separate 'diabetic diet.**' The dietary advice for managing blood sugar overlaps with the healthy eating guidance above.

Eating for kidney health

Some people may have issues with their kidneys. Fortunately, a healthy diet also helps to look after your kidneys and reduce the risk of long-term kidney problems. **Unless your healthcare team advises otherwise, you do not need to follow a special 'renal diet' (such as low protein, low potassium or low phosphate).** The healthy eating advice above also helps keep your kidneys healthy. If you need more specific advice, your kidney team may refer you to a kidney dietitian.

Eating well for gout

Some people may develop gout, which causes pain or swelling in the joints. Gout happens when uric acid builds up in the blood and forms tiny crystals in the joints. In the past, people with gout were advised to eat a "low purine diet." However, current guidance (including NICE) no longer recommends focusing on purines as this approach is not very effective on its own and can be difficult to maintain.

Instead, eating a healthy, balanced diet as described above, along with staying well hydrated, can help reduce the risk of gout attacks.

It is also helpful to limit alcohol, particularly beer and spirits, as these can increase the risk of gout.

Eating well for magnesium

Some people have low magnesium levels because their kidneys lose too much magnesium in the urine. Magnesium helps your muscles, heart, and nerves work properly, and supports energy levels and bone health.

Eating a balanced diet that includes foods containing magnesium can help support your overall nutrition. However, magnesium supplements do not always raise magnesium levels because that extra magnesium is often lost in the urine.

Foods that contain magnesium include green vegetables, wholegrains (such as brown rice or wholemeal bread), beans and lentils, fish (such as salmon or mackerel), nuts, seeds and fruit.

Eating well for pancreatic insufficiency

Often people living with HNF1B-associated disease have pancreatic insufficiency, which means the pancreas does not make enough enzymes to digest food

properly. This can cause tummy pain, bloating, wind, or stools that are pale, oily, or difficult to flush away. It can also make it harder to maintain your weight.

If you have pancreatic insufficiency, your doctor or dietitian may prescribe enzyme tablets (such as Creon® or Nutrizym®). These should be taken with all meals and snacks. They help your body digest food and absorb nutrients. The dose you need to take with each meal is individual to you and what you eat, so seek support if you take this medication and are still having symptoms as your dose may need adjusting.

Eating well for liver health

Many people have mildly abnormal liver blood tests. This is common and usually nothing to worry about. It doesn't usually mean your liver is damaged or that you need a special diet.

You can help keep your liver healthy by eating a healthy, balanced diet as described above and keeping alcohol within the recommended limit (no more than 14 units per week).

Eating well with autism or ADHD

Some people with a missing copy of the *HNF1B* gene (called a gene deletion) also have autism or ADHD. This may affect how you eat. Examples may include being sensitive to certain tastes, textures, or smells, or preferring the same foods every day. These differences are common and nothing to be ashamed of.

You do not need a special diet for autism or ADHD, but these tips can help:
-Keep mealtimes calm and predictable
-Try new foods slowly, one at a time
-Make food fun and familiar
-Aim for balance over time, not perfection at every meal
-Listen to your body - if you forget to eat, set reminders
-Ask a dietitian for support if you find eating difficult.

In summary

Most people with HNF1B-associated disease stay well and live healthy lives. Most don't need a special diet - the same **healthy eating advice** that supports general wellbeing works for people with this condition too.

Useful resources

Kidney Kitchen – www.kidneykitchen.org
Kidney Care UK – www.kidneycareuk.org
Diabetes UK – www.diabetes.org.uk
Drink Aware – www.drinkaware.co.uk

About this leaflet

This leaflet was written by the Renal Dietetic Service at Royal Devon University Healthcare NHS Foundation Trust. It provides general information and should not replace advice from your own healthcare team.