

# HNF1B and diabetes

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Why do HNF1B patients get diabetes?

Do all patients get diabetes?

When is diabetes likely?

How is the diabetes best treated?

Testing for diabetes complications

Pancreatic enzymes – do they matter?

# Why do HNF1B patients get diabetes?

HNF1B is important in the development of the pancreas

When reduced HNF1B due to genetic change you get a much smaller pancreas.

Reduced number of beta-cells that make insulin

Too few beta-cells results in too little insulin

Too little insulin means blood sugar goes up

(Also reduced pancreatic enzymes)

# Do all patients get diabetes?

About 50% of patients have diabetes now

Develop later in life so most children don't have diabetes

Likely to get when older; very rare to be over 45 without having diabetes

Later you get diabetes the less severe the diabetes is but still most people need insulin treatment at some point

# When is diabetes likely?

Rarely have diabetes in first few weeks of life (neonatal diabetes) but rapidly goes away.

Usually in adolescence as a young adult

Typically 12-30 years

Can be as old as 60

Should have regular (yearly) testing for diabetes (HbA1c) if don't have diabetes now (if >10 years at kidney clinic)

# How is diabetes best treated?

Best treatment depends on how much insulin you are making

Most patients make little insulin and need insulin treatment

Insulin usually with meals and overnight

In early stages tablets can be used but insulin often needed within 3-4 years

# How is diabetes best treated?

New drug SGLT2inhibitors:  
examples Empagliflozin, Canagliflozin,  
Dapagliflozin etc.

New treatment that works by increasing the  
sugar in the urine

Good because improves how kidneys work but  
not proven when not protein in the urine

Problems: might make very ill (ketoacidosis) if  
insulin low and not good at lowering glucose if  
kidneys not good

# Testing for diabetes complications

Like all types of diabetes need annual check to make sure problems not developing

Check

- Eyes
- Kidneys
- Feet
- Cholesterol
- Blood pressure



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## Is HNF1B diabetes Type 1 diabetes?

Cause different – failure of the insulin producing cells to be made (HNF1B monogenic) v disruption by the immune system (Type 1 diabetes)

Treatment is mainly the same: Insulin injections, pumps, closed loops.

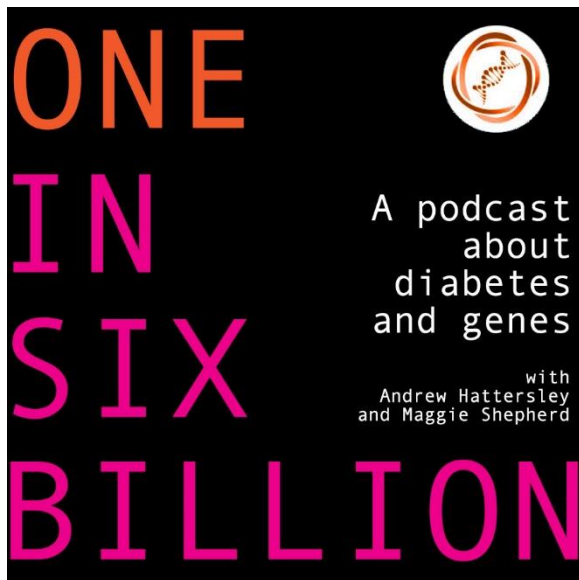
Compared to Type 1 will have a bit more help from the pancreas (in the early stages) but often need more insulin per kg than Type 1 as “insulin resistance” due to HNF1B.

Very few people have heard of HNF1B so best to call it “a type of Type 1 diabetes”!

## Further information

Diabetes information (search HNF1B diabetesgenes)

<https://www.diabetesgenes.org/what-is-mody/hnf1b-mody-rcad/>



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