

# **Master class - HNF1B and diabetes**

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# Renal cysts and diabetes

Mild Kidney cysts  
Diabetes

Kidney disease  
No diabetes



Mild Kidney cysts  
No diabetes

No Kidney cysts  
Diabetes

**Why do HNF1B patients get diabetes?**

**Do all patients get diabetes?**

**When is diabetes likely?**

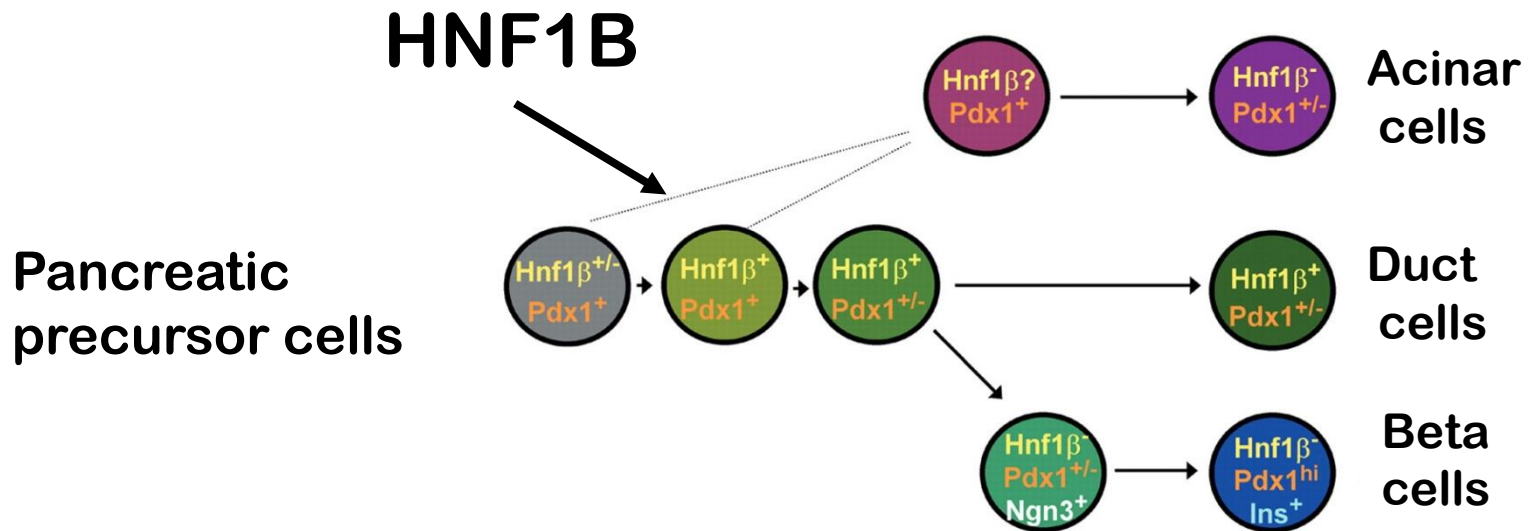
**How is the diabetes best treated?**

**Testing for diabetes complications**

**Pancreatic enzymes – do they matter?**

# Why do HNF1B patients get diabetes?

HNF1B is important in the development of the pancreas

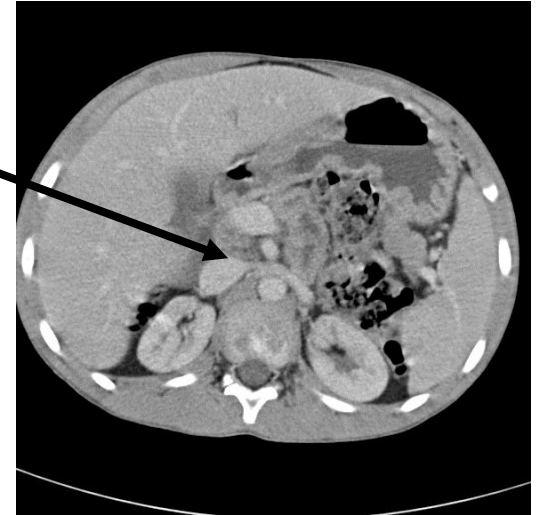


Maestro M *et al*, HMG 2004

Reduced HNF1B results in less of all the different cells in the pancreas

# Why do HNF1B patients get diabetes?

Reduced HNF1B results in a small pancreas with a reduced number of all cell types



When reduced HNF1B due to genetic change you get small pancreas.

Reduced number of beta-cells that make insulin

To little insulin means blood sugar goes up

(Also reduced pancreatic enzymes as less pancreatic cells that make these)

# Do all patients get diabetes?

Mild Kidney cysts  
Diabetes

Kidney disease  
No diabetes



Mild Kidney cysts  
No diabetes

No Kidney cysts  
Diabetes

# **Do all patients get diabetes?**

**About 50% of patients have diabetes**

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

**Likely to get diabetes when older:  
very rare to be over 50 without having diabetes**

# When is diabetes likely?

Very rarely seen in first few weeks of life (neonatal diabetes) but gets better

Usually in adolescence or young adult: typically diagnosed 12-30 years

Can be as old as 60

Should have regular (yearly) testing for diabetes (HbA1c or fasting glucose)



# 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 usually needed with a year**

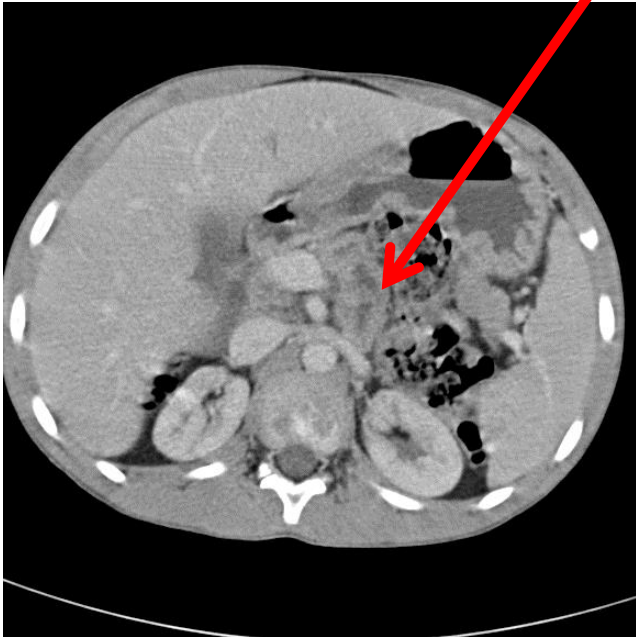
# Testing for diabetes complications

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

Check

- Eyes
- Kidneys
- Feet
- Cholesterol
- Blood pressure

# Pancreatic Enzymes – do they matter?



**Small pancreas – less enzyme secreting cells – so less enzymes (faecal elastase reduced)**

**Enzymes are needed for fat digestion**

**Most patients have made enough enzymes so don't need treatment**

**If problems with fatty meats – floating pale, fatty stools consider taking enzyme treatment with meals.**

# Conclusions

Almost all patients with HNF1B gene changes will get diabetes because the pancreas is small with a reduced number of insulin producing cells

Diagnosis usually after kidney disease typically 10 – 30 years but can be any age

Treatment is usually with insulin

Check for diabetes annually if not diagnosed and have an annual check up if have diabetes.