AT THE FOREFRONT UChicago Medicine | Kovler Diabetes Center

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HNF1B-MODY (MODY 5)

Legend: A transcription factor molecule binds to the DNA at its binding site, and thereby regulates the production of a protein from a gene.

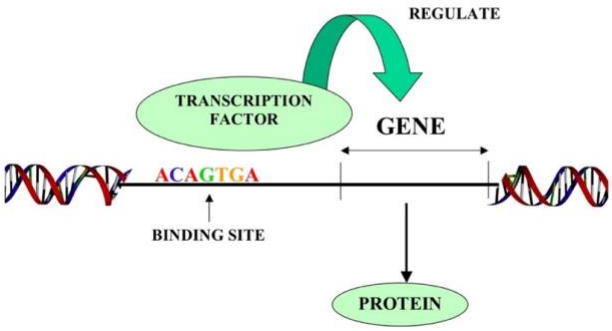

Image: http://www.bio.miami.edu/dana/pix/transcription_factor.jpg

Overview:
Diabetes caused by genetic changes (sometimes called 'mutations') in the *HNF1-beta* gene is a less common type of MODY. We call this HNF1B-MODY. This type of MODY used to be called MODY 5. The HNF1B gene contains instructions for a transcription factor called hepatocyte nuclear factor-1 beta that is important for the normal development of beta cells, kidneys, and genitourinary tracts. Beta cells are cells in the pancreas that make insulin and help to keep blood sugars in a normal range.

Presentation:
People with HNF1B-MODY often have diabetes in combination with kidney or genitourinary tract problems, such as kidney cysts, abnormally shaped kidneys, or abnormally shaped uterus.

These genetic changes in HNF1B can occur spontaneously, such as when a person does not inherit the change from their parent, but usually the genetic changes are passed on from a parent to a child. If a parent has HNF1B-MODY, there is a 50% chance that their child will inherit the genetic change and be at risk of developing diabetes at a young age.

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Features that make us suspect HNF1B-MODY include:

- Negative antibody testing. These tests are typically done around the time of diabetes diagnosis and, if positive, indicate an autoimmune type of diabetes such as Type 1 diabetes.
- Renal (kidney) cysts or structural problems of the genitourinary tract or kidneys
- No history of diabetic ketoacidosis
- A strong family history of diabetes - 2 or 3 generations of diabetes diagnosed at a young age (typically under 35 years old) - or kidney cysts/structural problems of the genitourinary tract or kidneys

Treatment:
Unlike other forms of MODY, patients with HNF1B-MODY do not necessarily respond well to pills like sulfonylureas. Insulin may be required.

Inheritance:
Finding out someone has HNF1B-MODYmay have implications for other family members as those family members may also have HNF1B-MODY. A child of a person with HNF1B-MODY will have a 50% chance of inheriting the genetic change and developing diabetes and/or kidney or genitourinary problems.

Research:
If you or a family member have HNF1B-MODY, or think you may have it, then please contact us at monogenicdiabetes@uchicago.edu to learn more about our studies.