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My46 Trait Profile

Hereditary Papillary Renal Cell Carcinoma

Renal cell carcinoma is a cancer of the kidney. There are different types of renal cell carcinoma (RCC). Hereditary Papillary Renal Cell Carcinoma is a genetic condition that increases the chance of developing papillary type 1 RCC. It is caused by mutations in a gene called MET that makes the hepatocyte growth factor receptor protein.

Characteristics of Hereditary Papillary Renal Cell Carcinoma

Most cancer occurs as a result of normal aging, lifestyle, and exposures. However, approximately 2-4% of renal cell carcinoma (RCC) is caused by inherited risk factors (genetic changes or mutations) that increase the chances of developing RCC. Papillary type 1 renal cell carcinoma is a specific type of kidney cancer. Individuals with Hereditary Papillary Renal Cell Carcinoma (HPRCC) have a higher chance to develop papillary type 1 RCC. An individual with HPRCC can develop one or more RCCs in one or both kidneys. The onset of RCC in individuals with HPRCC is typically in adulthood. However, not all individuals with HPRCC develop RCC. HPRCC is an uncommon condition, and few families have been reported in the literature. It is considered an adult-onset condition and RCC in children have not been reported in these families. Based on current knowledge there are no other types of cancer (i.e. cancers in other parts of the body) associated with HPRCC.

Diagnosis/Testing

HPRCC is suspected when an individual has multiple papillary type 1 RCCs in one or both kidneys, or when multiple members of the family have papillary RCC. Most individuals with HPRCC will have a change or mutation in a gene called MET. MET is an oncogene that promotes the growth of cells in the body. Mutations in this gene are thought to activate the growth of a cell. When a kidney cell loses the ability to maintain control over growth, a RCC can begin to develop. A RCC is a cluster of cells within the kidney that have an abnormal ability to grow and divide into new cells (thereby forming a mass), in comparison to normal cells within the same kidney.

Management/Surveillance

There are currently no standard guidelines for surveillance. However, adults with HPRCC should undergo regular monitoring through imaging such as CT or MRI of their kidneys to screen for kidney tumors. For lifelong monitoring, use of MRI may be preferred over CT in order to limit cumulative radiation exposure.

If a kidney tumor is detected on imaging, the individual should be referred to a urologic oncology surgeon for further evaluation.

Mode of inheritance

HPRCC is inherited in an autosomal dominant pattern. This means inheriting one MET mutation is enough for an individual to be at risk of developing papillary type 1 RCC. The mutation can be inherited from an affected parent or it can occur for the first time (de novo) in an individual.

Risk to family members

An individual with a MET gene mutation has a 50% chance of passing the mutation to each of his/her children, and a 50% chance of not passing the mutation.

Special considerations

None

Resources

Kidney Cancer Association
http://www.kidneycancer.org
Cancer.net: Hereditary Papillary Renal Cell Carcinoma
http://www.cancer.net/cancer-types/hereditary-papillary-renal-cell-carcinoma

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