**What you should know about Multiple Endocrine Neoplasia Type 2 (MEN2)**

MEN2 is a condition caused by mutations in the RET gene. Approximately 25% (1 in 4) individuals with medullary thyroid cancer have a mutation in the RET gene. Individuals with RET mutations may also develop tumors in their parathyroid and adrenal glands (pheochromocytoma). There are three types of MEN2, based on the family history and specific mutation found in the RET gene:

- **MEN2A** is the most common type of MEN2, with medullary thyroid cancer developing in young adulthood. MEN2A is also associated with adrenal and parathyroid tumors.
- **MEN2B** is the most aggressive form of MEN2, with medullary thyroid cancer developing in early childhood. MEN2B is associated with adrenal tumors, but parathyroid tumors are rare. Individuals with MEN2B can also develop benign nodules on their lips and tongue, abnormalities of the gastrointestinal tract, and are usually tall in comparison to their family members.
- **Familial Medullary Thyroid Cancer (FMTC)** is characterized by medullary thyroid cancer (usually in young adulthood) without adrenal or parathyroid tumors.

**The risk for cancer associated with MEN2**

- MEN2A is associated with a ~100% risk for medullary thyroid cancer; 50% risk of adrenal tumors; and 25% risk of parathyroid tumors
- MEN2B is associated with a 100% risk for medullary thyroid cancer; 50% risk of adrenal tumors; and rare risk of parathyroid tumors
- FMTC is associated with ~ 100% risk for medullary thyroid cancer; and no risk for adrenal or parathyroid tumors

Tumors that develop in the adrenal glands in individuals with MEN2 are typically not cancerous, but can produce excessive amounts of hormones called catecholamines, which can cause very high blood pressure.

Tumors that develop in the parathyroid glands in individuals with MEN2 are also typically not cancerous, but they can produce excessive amounts of parathyroid hormone. This causes calcium to be moved from the bone to the blood, which causes the bones to become weak, and can cause kidney stones to develop.

**The risks to family members**

MEN2 is inherited as an autosomal dominant condition. This means that the children, brothers, sisters, and parents of individuals with a RET mutation have a 50% risk to have the mutation. Approximately 50% of individuals with MEN2B have no family history of MEN2, which means that they have a new mutation.

**Managing the risk**

**Thyroid cancer:** Removal of the thyroid gland is the best way to prevent medullary thyroid cancer, and is recommended for individuals with a RET mutation; yearly calcitonin testing (blood test) following thyroid removal is recommended.

**Adrenal and Parathyroid tumors:** Yearly biochemical screening followed by imaging tests if the biochemical tests are abnormal; surgical removal of tumors if present.