All About Hürthle Cell Carcinoma

What is the thyroid?

The thyroid gland is located on the anterior (front) part of the neck. It can be felt just below the thyroid cartilage, or "Adam's apple." It is a butterfly-shaped organ that stretches across the middle of the neck just below the thyroid cartilage. Its "wings" spread towards the head, on either side of the thyroid cartilage. These "wings" are called the lobes of the thyroid, with the middle section being called the isthmus.

As an endocrine gland, the thyroid gland produces, stores, and secretes the thyroid hormones thyroxine (T4) and triiodothyronine (T3) into the bloodstream. The thyroid does this as a response to a hormone produced by the pituitary gland, called thyroid stimulating hormone, or TSH. When the thyroid gland is "turned on" by TSH, it increases its uptake of iodine, which is required to make thyroid hormone.

The principal cells of the thyroid are called follicular cells and are mainly responsible for the production and secretion of thyroid hormones. Thyroid hormone plays an important role in controlling our metabolism. Many functions such as body temperature and heart rate are regulated by thyroid hormone. Too much thyroid hormone in your system is called hyperthyroidism, while too little is called hypothyroidism. An imbalance of thyroid hormone may cause serious problems if not properly treated.

What is Hürthle Cell Carcinoma?

Hürthle Cell Carcinoma is a type of thyroid cancer. It is estimated there are about 52,890 new cases of thyroid cancer diagnosed each year in the United States. The four major types of thyroid cancer are papillary carcinoma, follicular carcinoma, medullary carcinoma, and undifferentiated. Hürthle cell carcinoma is a type of follicular carcinoma and accounts for about 3% of all thyroid cancers. Researchers believe that its behavior is similar to follicular cell carcinoma, but it tends to be more aggressive. These tumors are more common in women than men and affect Caucasians at a higher rate than other races.

Hürthle cell cancer has a high chance of metastases compared with other thyroid cancers. Hürthle cell cancer may metastasize to lymph nodes in the neck but follicular cancer does not. Both Hürthleccell and follicular cancers may spread by invasion into blood vessels, which may give rise to metastases outside of the neck. The likelihood of metastasis can be predicted by the pathology at the time of surgery. Hürthle cell cancers that are small and show no sign of invasion into blood vessels or lymph nodes are unlikely to spread.
What causes Hürthle cell carcinoma and am I at risk?

It is not clear what causes Hürthle cell carcinoma. Known risks of developing Hürthle cell carcinoma are a history of external beam radiation to the head and/or neck, iodide deficiency, genetic mutations, and a family history of thyroid cancer.

How can I prevent Hürthle cell carcinoma?

As most cases of Hürthle cell carcinoma are not linked with any risk factors, there is usually no way to prevent it. It is important that the thyroid is shielded in children who have diagnostic X-rays, such as dental X-rays. If you have a family history of thyroid cancer, be sure to notify your primary care provider.

What screening tests are used for Hürthle cell carcinoma?

Currently, there are no screening standards for thyroid cancer. However, thyroid cancer can be diagnosed early on. Part of your yearly physical exam should be visual and physical examination of the neck. Palpation (feeling) of the neck will detect many clinically significant thyroid cancers, which is part of a routine physical exam. In addition, the thyroid gland is included on many radiology studies performed to evaluate other concerns, such as CT scans of the lungs and cervical spine.

All patients suspected to have thyroid nodules (abnormal growths), either from physical examination or from another radiology study, should have a thyroid ultrasound done. Thyroid ultrasound uses sound waves to take a picture of the thyroid gland and surrounding structures. The ultrasound view of the nodule can help healthcare providers determine if a fine-needle aspiration biopsy is needed to further test the nodule.

What are the signs of Hürthle cell carcinoma?

The most common physical sign of Hürthle cell carcinoma is a quickly growing lump that you can feel below your Adam's apple. Other signs may include difficulty swallowing, pain, a hoarse voice if the vocal cords are involved, pressing down of the trachea, and enlarged lymph nodes. It can also cause superior mediastinal syndrome which causes facial swelling and dilated veins. Most patients with Hürthle cell carcinoma are euthyroid, meaning that the thyroid is functioning normally and does not cause any concerning symptoms.

How is Hürthle cell carcinoma diagnosed?

Once a thyroid nodule is found, the next steps are to determine if the nodule is a benign growth or malignant (cancerous) tumor. A physical exam should be done by a healthcare provider. Special attention should be given to examination of the neck to check for enlarged lymph nodes. The workup may also include having blood drawn to check the function of your thyroid.

Remember, you can have a properly functioning thyroid gland and still be diagnosed with Hürthle cell carcinoma. Your provider may order imaging of your thyroid including an ultrasound, a thyroid uptake and scan, a CT or MRI of the neck, and a PET scan. A thyroid uptake and scan is a nuclear medicine study using radioactive iodine. Functioning thyroid tissue takes up iodine to produce normal thyroid hormones. Therefore, the small amount of radioactive iodine used will be taken up by those areas of the thyroid that are producing thyroid hormone and then "seen" on the scan.

Unlike many cancers, Hürthle cell cannot be definitively diagnosed with a fine needle aspiration (biopsy). While the biopsy may be suggestive of a tumor that may or may not be cancer, called a Hürthle cell neoplasm, the large majority of these will be found to be benign (called a Hürthle cell adenoma). The only way to confirm the diagnosis of cancer is by identifying capsular or vascular invasion (that is, invasive growth that is seen with cancer). This can only be determined after the nodule is surgically removed and examined by a pathologist in the laboratory.

Once there is a diagnosis of a Hürthle cell neoplasm, surgery to remove the affected thyroid lobe is typically the next step. Only 15-30% of patients will receive a diagnosis of carcinoma after surgery, with the remainder finding the nodule was not cancerous.

How is Hürthle cell carcinoma staged?

The staging of a cancer describes how much the cancer has grown and invaded the area, explaining the extent of disease.
There is no specific staging system used for Hürthle cell carcinoma, but there is for thyroid cancer. The TNM (tumor, node, metastases) system is the most commonly used staging system. Most classification systems used in this type of cancer take into account factors such as tumor size, patient age, presence of metastases, and major capsular invasion. Below is the commonly used TNM staging for papillary and follicular thyroid cancer.

**NCCN TNM Staging System for Papillary and Follicular Thyroid Carcinoma (2.2017)**

**Papillary or Follicular(differentiated; under 45 years old):**

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<thead>
<tr>
<th>Stage</th>
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<th>N</th>
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<tr>
<td>Stage I</td>
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<td>M0</td>
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<tr>
<td>Stage II</td>
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<td>M1</td>
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**Papillary or Follicular(age 45 years and older):**

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<th>Stage</th>
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<tr>
<td>Stage I</td>
<td>T1</td>
<td>N0</td>
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<tr>
<td>Stage II</td>
<td>T2</td>
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<tr>
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<td>T3</td>
<td>N1a</td>
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<tr>
<td>Stage IVA</td>
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<td>T4a</td>
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<td>T1</td>
<td>N1b</td>
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<td>Stage IVC</td>
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Complete thyroid cancer staging available in the article "All About Thyroid Cancer."

**How is Hürthle cell carcinoma treated?**

Surgery remains the primary treatment for Hürthle cell carcinoma. However, other treatments such as radioactive iodine and radiation may be used alone or in combination with surgery.

**Surgery**

The main treatment of Hürthle cell carcinoma is surgery. However, there is some controversy as to how extensive a surgery needs to be, given that some tumors will behave aggressively, while others will not. The initial size of the nodule and the patient’s age and general health may help decide the extent of the first surgery. In some cases, the entire thyroid gland is removed (total thyroidectomy) along with any enlarged lymph nodes. Detection of cancer in any lymph nodes may lead to a more extensive lymph node dissection. Some experts feel total thyroidectomy is the best procedure because it allows follow up tests to be more effective, as residual thyroid tissue can affect these.

**Radioactive Iodine**

Radioactive iodine is a useful nonsurgical treatment for many types of thyroid cancers, but Hürthle cell cancer is treatable only by radioactive iodine in a small number of cases. A dose of radioactive iodine may be given to destroy the remaining normal thyroid tissue after surgery. The iodine is taken up into the cell and the radiation within the radioactive iodine itself is released locally, delivering a lethal dose of radiation to these cells.
Radiation Therapy
In Hürthle cell carcinoma, radiation therapy is used to relieve symptoms of metastases, control recurrent tumors, and to prevent recurrence. It is only indicated when complete surgical removal is not possible. Radiation comes in the form of high energy x-rays, similar to those used for diagnostic x-rays, but they are of a much higher energy. The high energy of x-rays in radiation therapy results in damage to the DNA of cells. Cancer cells divide faster than healthy cells, so their DNA is more likely to be damaged than normal cells. Additionally, cancer cells are generally less able to repair damaged DNA than normal cells are, so cancer cells are killed more easily by radiation than normal cells.

Clinical Trials
There are clinical research trials for most types of cancer, and every stage of the disease. Clinical trials are designed to determine the value of specific treatments. Trials are often designed to treat a certain stage of cancer, either as the first form of treatment offered, or as an option for treatment after other treatments have failed to work. They can be used to evaluate medications or treatments to prevent cancer, detect it earlier, or help manage side effects. Clinical trials are extremely important in furthering our knowledge of disease. It is through clinical trials that we know what we do today, and many exciting new therapies are currently being tested. Talk to your provider about participating in clinical trials in your area. You can also explore currently open clinical trials using the OncoLink Clinical Trials Matching Service.

Follow-up Care and Survivorship
People facing a diagnosis of Hürthle cell carcinoma require a multidisciplinary team of healthcare providers, including an endocrinologist, head and neck surgeon or endocrine surgeon, nuclear medicine physician, pathologist and, in a few cases, a radiation oncologist. Finding an experienced team is important in treating this rare cancer. After surgery, patients will require thyroid hormone supplementation and monitoring for life. How often you need to be evaluated and which tests you will need will be determined by the providers caring for you. Monitoring for tumor recurrence is done with ultrasound, measurement of thyroglobulin levels (blood test), and in some cases iodine 131 total body scanning. Other radiology exams, including PET scan, CT or MRI imaging, may be used in cases where iodine-131 uptake is not present. It is important to follow your provider’s follow-up recommendations and to speak with your team regarding any recurrent or new symptoms you are experiencing as they could be related to your treatment or recurrence.

Fear of recurrence, the financial impact of cancer treatment, employment issues and coping strategies are common emotional and practical issues experienced by thyroid cancer survivors. Your healthcare team can identify resources for support and management of these practical and emotional challenges faced during and after cancer.

Cancer survivorship is a relatively new focus of oncology care. With nearly 17 million cancer survivors in the US alone, there is a need to help patients transition from active treatment to survivorship. What happens next, how do you get back to normal, what should you know and do to live healthy going forward? A survivorship care plan can be a first step in educating yourself about navigating life after cancer and helping you communicate knowledgeably with your healthcare providers. Create a survivorship care plan today on OncoLink.

Resources for More Information
ThyCa: Thyroid Cancer Survivors’ Association
Provides education and support for people with thyroid cancer as well as funds research into the disease.
www.thyca.org

Light of Life Foundation
A non-profit organization which provides education and support to people with thyroid cancer and the public.
www.lightoflifefoundation.org

Endocrine Web
Provides education about various endocrine conditions, including thyroid cancers.
www.endocrineweb.com/conditions/thyroid-cancer

REACT Thyroid Foundation
An advocacy organization dedicated to supporting research for thyroid cancer treatments.

www.reactthyroidfoundation.org/

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