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THYROID CANCER

Aggressive Surgery in Certain Patients with Familial Medullary Thyroid Cancer Is Safe and Results in High Cure Rates

WHAT IS THE STUDY ABOUT?

Medullary thyroid cancer can be inherited, either as part of a genetic syndrome known multiple endocrine neoplasia 2A (MEN 2A) or by itself (familial medullary thyroid cancer). This cancer is caused by genetic mutations and can be diagnosed at a young age. Patients with a specific genetic mutation (codon 634) start with an increase in the number of thyroid C-cells (C-cell hyperplasia) which then progresses to medullary cancer. Genetic screening for this mutation can identify patients that have either no changes in the thyroid, only C-cell hyperplasia or very early medullary cancer. In these patients, surgery to remove the entire thyroid can prevent the development of a significant medullary cancer. This surgery is usually done before the age of 5 years. While removing the thyroid is clearly helpful, it is not clear if removing all lymph nodes from the central neck (behind the thyroid) even before they appear to be involved with cancer is beneficial for patients with this gene mutation. The aim of the present study was to determine the effect of removing all lymph nodes from the central neck (central neck dissection) at the time of thyroid surgery.

THE FULL ARTICLE TITLE:

Schellhaas et al. Prophylactic central neck dissection with total thyroidectomy in familial medullary thyroid cancer with codon 634 mutations is safe and results in high cure rates. Surgery 2009; 146: 906-912.

WHAT WAS THE AIM OF THE STUDY?

The aim of the present study was to determine the effect of removing all lymph nodes from the central neck at the time of thyroid surgery.

WHO WAS STUDIED?

Seventeen patients with codon 634 mutations who had a total thyroidectomy and central neck dissection between 1992 and 1999 were evaluated. Fourteen of these patients had MEN 2A and 3 familial medullary thyroid cancer. Their median age was 13 years (range 4 to 36). Median follow up was 147 months (range 90 to 181 months).

HOW WAS THE STUDY DONE?

The seventeen patients included in this study had their initial surgery performed at a single institution in Germany. The surgery included a total thyroidectomy with central neck dissection. None of the patients had any symptoms of their cancer at the time of surgery. The patients and their primary doctors were contacted via telephone or mail and asked about the course of their cancer, their blood calcitonin levels (a hormone marker of medullary cancer) and any other treatment that they had received because of their MTC or other MEN 2A related problems.

WHAT WERE THE RESULTS OF THE STUDY?

Three patients had C-cell hyperplasia, 12 patients had small cancers < 1 cm and 2 patients had cancers between 1-2 cm. Although an average of 10 lymph nodes were removed per patient, only 2 patients had evidence of spread of the cancer to the lymph nodes. Of these 2 patients, 1 had repeat surgery and is currently free of the cancer. The other patient still has increased calcitonin levels but no clinical evidence of the cancer. All of the other patients remained free of cancer during the study period (potential 94% cure rate). The preoperative calcitonin levels correlated with size of the cancers but not with the presence of spread to the lymph nodes.
Fifteen patients had a preoperative ultrasound and in 9 no abnormalities, were found, even though 6 of them had cancer and one had spread to the lymph nodes. The other 3 patients had C-cell hyperplasia. Six patients were found to have nodules and all of them were medullary cancer. The incidence of complications related to the surgery was low. Nine patients had no complications at all. One patient (5.9%) had permanent hypoparathyroidism which required calcium pills and 5 patients had hypoparathyroidism that resolved with follow up. One patient had temporary injury to the laryngeal nerve, but this resolved on follow up examination.

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HOW DOES THIS COMPARE WITH OTHER STUDIES?

Many studies had shown that early thyroidectomy in patients with certain genetic mutations can prevent the development of medullary thyroid cancer. One study suggested that total thyroidectomy is sufficient in patients without invasive cancers. However, because many young and asymptomatic patients already have lymph node involvement at the time of the initial "preventive" operation, many physcians favor a preventive central neck dissection in almost all patients with medullary cancer.

WHAT ARE THE IMPLICATIONS OF THIS STUDY?

Medullary thyroid cancer can be prevented in patients with genetic mutations by performing early thyroidectomy. This study shows that in the hands of experienced surgeons, preventive central neck dissection at the time of initial thyroidectomy can be done safely in young children with minimal complications and results in high cure rates. However, because in this study there were few patients with spread to the lymph node , a general conclusion regarding the benefits of preventive central neck dissection could not be reached.

— M. Regina Castro, MD

ATA THYROID BROCHURE LINKS

Thyroid Cancer. http://thyroid.org/patients/patient_brochures/cancer_of_thyroid.html

Thyroid surgery. http://thyroid.org/patients/patient_brochures/surgery.html

ABBREVIATIONS & DEFINITIONS

Medullary thyroid cancer — a relatively rare type of thyroid cancer that also may be inherited.

Central neck compartment — the central portion of the neck between the hyoid bone above, and the sternum and collar bones below and laterally limited by the carotid arteries.

Prophylactic central neck dissection — Careful removal of all lymphoid tissue in the central compartment of the neck, even if no obvious tumor is apparent in these lymph nodes

Thyroidectomy — Surgery to remove the entire thyroid gland. When the entire thyroid is removed it is termed a total thyroidectomy. When less is removed, such as in removal of a lobe, it is termed a partial thyroidectomy.

MEN 2A — Multiple endocrine neoplasia, type 2A. A hereditary syndrome in which medullary thyroid cancer is often seen in association with other endocrine tumors such as pheochromocytoma (a tumor of the adrenal glands) and hyperparathyroidism (elevated parathyroid hormone levels usually caused by tumors of the parathyroid glands).

C-cell hyperplasia — An abnormal growth of parafollicular (C-cells) cells that usually occurs before the development of familial forms of medullary thyroid cancer and is considered a pre-cancerous condition

Lymph node — bean-shaped organ that plays a role in removing what the body considers harmful, such as infections and cancer cells.

Hypoparathyroidism — low calcium levels due to decreased secretion of parathyroid hormone (PTH) from the parathyroid glands next to the thyroid. This can occur as a result of damage to the glands during thyroid surgery and usually resolves. This may also occur as a result of autoimmune destruction of the glands, in which case it is usually permanent.