What is Medullary Thyroid Cancer (MTC)?

Medullary Thyroid Cancer, or MTC, is a rare form of cancer of the thyroid gland. This gland is part of the endocrine system as shown below. When there is no family history of the disease and MTC occurs as an isolated case, it is called sporadic MTC. 1 in 4 or 25% of cases of MTC occur as part of a rare inherited (genetic) disorder called Multiple Endocrine Neoplasia Type 2 (MEN2). All sporadic MTC patients must be offered DNA screening for MEN2. AMEND has produced separate information booklets on the inherited forms of MTC.

How is MTC Diagnosed?

In most cases, patients may notice a lump in the neck which was not there before, or this may be noticed by a partner or colleague. Sometimes the growth secretes a hormone or hormones, which may cause diarrhoea, and it is not immediately apparent that this may be associated with a problem in the neck. In some families there may be a history of medullary thyroid cancer which warrants testing other family members. A diagnosis of medullary thyroid cancer may be confirmed by blood test, a fine needle biopsy, and ultimately by surgery (see Testing for MTC).
Sporadic Medullary Thyroid Cancer (MTC)

The thyroid is situated at the front of the neck. This gland produces 3 hormones; thyroxine and triiodothyronine (essential for maintaining the body’s metabolism and mental and physical development) and calcitonin (which has no known action in healthy people). MTC occurs in the calcitonin-producing cells (parafollicular or C-cells) of the thyroid. MTC usually develops over a number of years but can spread early on to nearby lymph nodes. If the thyroid and nearby lymph nodes are surgically removed while the cancer is still contained within the thyroid (total thyroidectomy and central lymph node dissection), a patient is usually cured. If, after surgery, calcitonin levels are still raised, this indicates that the cancer has spread (become metastatic) or has not been completely removed, and so further surgery and therapies are used to control it. As yet there is no definitive cure for metastatic MTC; however, it is usually slow growing and may often be managed effectively and without symptoms for many years. Symptoms that may develop can sometimes be controlled by the use of radiotherapy and sometimes chemotherapy.

Children and MTC

It is rare for children to present with MTC. However, those that do present with apparently sporadic MTC (with no family history of the disease) should be seen by a genetics clinic for investigation of MEN2 (an inheritable genetic disorder), as they are likely to have this condition and will require screening for other potential MEN2-related problems. A referral to a genetic counsellor at your Regional Genetics Service Centre should be made by your specialist or GP if appropriate.

Testing for MTC

Tests you may have to confirm a diagnosis of MTC:

Blood Tests
Baseline Calcitonin A simple blood test to detect calcitonin levels [NB: once drawn, the blood must be taken immediately and on ice to a chilled centrifuge in the lab]

Scans/Other
Ultrasound and Fine Needle Aspiration (FNA) A painless scan of the neck using a probe provides pictures of the thyroid lump. A tissue biopsy (sample) is obtained from the insertion of a small needle into the thyroid lump and this often provides the definitive diagnosis.
**Treating MTC**

Once a diagnosis of MTC has been made, a staging ultrasound scan of the neck should be undertaken. If there is no suspicion of enlarged lymph nodes, a total thyroidectomy and central node dissection (removal) is undertaken. If enlarged or involved lymph nodes are detected, the removal of other lymph nodes should be undertaken at the same time. If MTC is diagnosed before surgery, 24-hour urine collections to test for metanephrines or a plasma metanephrine test should be performed (even if there is no family history of MEN2) to rule out the presence of an adrenal growth (phaeochromocytoma).

**SURGERY**

Total thyroidectomy + central node dissection  A small incision is made at the base of the front of the neck from which the thyroid and nearby lymph nodes can be removed. A larger incision is required if the removal of other neck lymph nodes is necessary. Eating and drinking is possible almost immediately after waking from the operation.

**Hospital Stay**

Approximately 3-5 days

**Risks**

Injury to the nerves that control the vocal cords (less than 1-2%); unavoidable removal of or injury to the parathyroid glands resulting in a temporary drop in calcium levels in the blood, although occasionally this may be permanent. Symptoms of low blood calcium include tingling lips, fingers and toes, and eventually cramping, all of which can be corrected with medication.

**MEDICATION**

**Levothyroxine**

This is a very well established and effective replacement for the thyroid hormones. It must be taken life-long after thyroidectomy. Tablets are taken once a day and doses are typically between 100-150mcg for adults, lower for children. Regular blood tests are required to ensure that the right dose is being prescribed. Too large a dose may cause symptoms such as rapid heartbeat, sweating, anxiety, tremor and loss of weight. Too small a dose may cause symptoms such as lethargy, slow heartbeat, sensitivity to cold, and weight gain. Although the above symptoms may suggest a need for a change of dosage, the same symptoms can occur in other conditions. Only a blood test (measuring the thyroid stimulating hormone or TSH level) can determine accurately whether a change in levothyroxine dose is required. Once a stable dose is achieved, as judged by blood tests, repeat tests need only be done annually in adults, or more frequently in children and teenagers as they grow.

**Calcium replacement medication** *(required if parathyroid glands are injured or unavoidably removed along with the thyroid)*

**Vitamin D** *(calcitriol, alfacalcidol or ergocalciferol)*

Vitamin D supplements in capsule form that aid absorption of calcium from the patient’s diet. Taken once a day, this is often the only life-long medication required after parathyroid loss.

**Calcium Carbonate** *(Calcichew or Adcal)*

This is a chalk-like tablet that should be chewed or sucked. This is often used as a temporary “top-up” after surgery, but is not necessarily required life-long. Too large a dose or an indication that this supplement is no longer needed may become apparent if the patient begins to suffer from headaches, nausea and vomiting.

**Magnesium Supplement**

This may be in the form of an injection or tablet (e.g. magnesium glycerol-phosphate) but is rarely needed long-term.
The Future for Metastatic MTC Treatment

Research into treatments for MTC is currently focused on human trials which are testing the effectiveness of a group of drugs called tyrosine kinase inhibitors. These have been shown to work in the laboratory by stopping the uncontrolled cell division of MTC, or even making cells self-destruct. There are also other drugs under trial which may directly interfere with the growth factors which may be involved in maintaining tumour growth, but such agents are also only currently available in clinical trials.

Treating Metastatic MTC

Patients with MTC may have high blood calcitonin levels even after complete surgical treatment. However, although this indicates that there are MTC cells left in the body, patients with calcitonin levels that are higher than normal, remaining the same over a period of time, or slowly increasing do not necessarily need further investigation or treatment. This is because scans are unlikely to identify a site of disease outside of the neck unless calcitonin levels are significantly high: calcitonin alone is not an indication of a growing tumour. Nevertheless, in some patients, the search for metastatic disease may involve various scans (including radioactive isotope scans) and even a laparoscopy (telescopic inspection of the abdomen), followed by treatment with more surgery or radiotherapy if required.

MIBG / OCTREOTIDE THERAPY

Where surgery is no longer an option due to the extent of the disease, some specialised medical centres may use radiolabelled octreotide or MIBG radioactive therapies, which have very few side effects, to help reduce or control the spread. However, it is only appropriate if tests suggest that the radioisotope will be taken up by the tumour. The agent is attached to a radioactive substance, and is given through a vein by slow injection. The patient remains radioactive for a few days and therefore must be nursed in a lead-lined room. The treatment may need to be repeated several times at 3 or 6 month intervals. Possible side effects of MIBG Therapy include nausea, and occasionally vomiting. Until a complete cure is found, much of the current focus of treatment for extensive metastatic MTC is on the relief of the symptoms it causes:

Diarrhoea Adjustments to the patient’s diet may be required, together with an anti-diarrhoea medication such as Imodium, which contains loperamide. Some of the tumours contain somatostatin receptors, and in these instances treatment with a long-acting form of somatostatin (octreotide or lanreotide) may sometimes be helpful. Some believe that in such cases it may also help slow down the growth of the tumour.

Flushes Anti-ulcer medications called histamine receptor blockers (H2 blockers such as cimetidine or ranitidine) may occasionally be prescribed to help ease flushing.

Painful bone metastases may be suitable for external radiation therapy, which can provide rapid relief. In all cases, pain medications may be prescribed.

The Future for Metastatic MTC Treatment

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Useful Information

FREE PRESCRIPTIONS: In the UK, if you are to take lifelong levothyroxine, you are entitled to free prescriptions for all medicines. You should obtain a P11 leaflet from your doctor and fill in form B. Your doctor will then sign it and send it on. You will then receive an exemption certificate, which you must show to your pharmacist when collecting medicines.

MEDICALERT®: AMEND recommends that anyone taking lifelong medications obtain and wear a MedicAlert® identification emblem. The emblem contains summarised information of your medical condition and a 24-hour Helpline number for emergency medical staff to call in order to obtain detailed information on your medical condition from the MedicAlert database. This enables emergency medical staff to give appropriate treatment in full knowledge of your underlying condition and current medications. Emblems come in a range of styles so that there is something for everyone, even children.

Telephone AMEND for an order form and brochure or join online at www.medicalert.org.uk.

BENEFITS: Cancer patients may be entitled to state benefits, or even help with the costs of hospital parking. For UK information or support, contact Macmillan Cancer Relief.

Telephone 0808 808 0000, or visit their website at www.macmillan.org.uk/abetterdeal.

Useful Organisations

The British Thyroid Foundation
Tel: 0870 770 7933
www.btf-thyroid.org.uk

Butterfly Thyroid Cancer Trust
Tel: 01207 545469
www.butterfly.org.uk

Macmillan Cancer Relief
Tel: 0800 808 0000
www.macmillan.org.uk

Hypoparathyroidism UK
Tel: 01342 316315
www.hpth.org.uk

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- Dr Fiona Lalloo, Consultant Clinical Geneticist, St Mary’s Hospital, Manchester;
- Mr Tom Lennard, Professor of Surgery, University of Newcastle upon Tyne;
- Mr David Scott-Coombes, Department of Surgery, University Hospital of Wales, Cardiff;
- Professor Tony Weetman, University of Sheffield Clinical Sciences Centre, Sheffield.

Afterword

This book has been written for patients by patients with the help of a medical advisory panel. The aim of this book is to answer those questions, sometimes in great detail, that one may come across during a lifetime of living with MTC. It is not for use in self-diagnosis. It contains detailed information on tests, surgery and potential symptoms associated with MTC. However, it is possible that not all of this information will be relevant to you. This book is not intended to replace clinical care decisions and you should always discuss any concerns you may have carefully with your specialist. Every care has been taken to ensure that the information contained in this book is accurate, nevertheless, AMEND cannot accept responsibility for any clinical decisions.

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