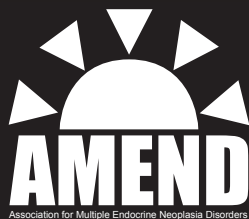


MTC

Patient Information

Medullary Thyroid Cancer



Registered Charitable Incorporated
Organisation no 1153890

MTC SUPPORT

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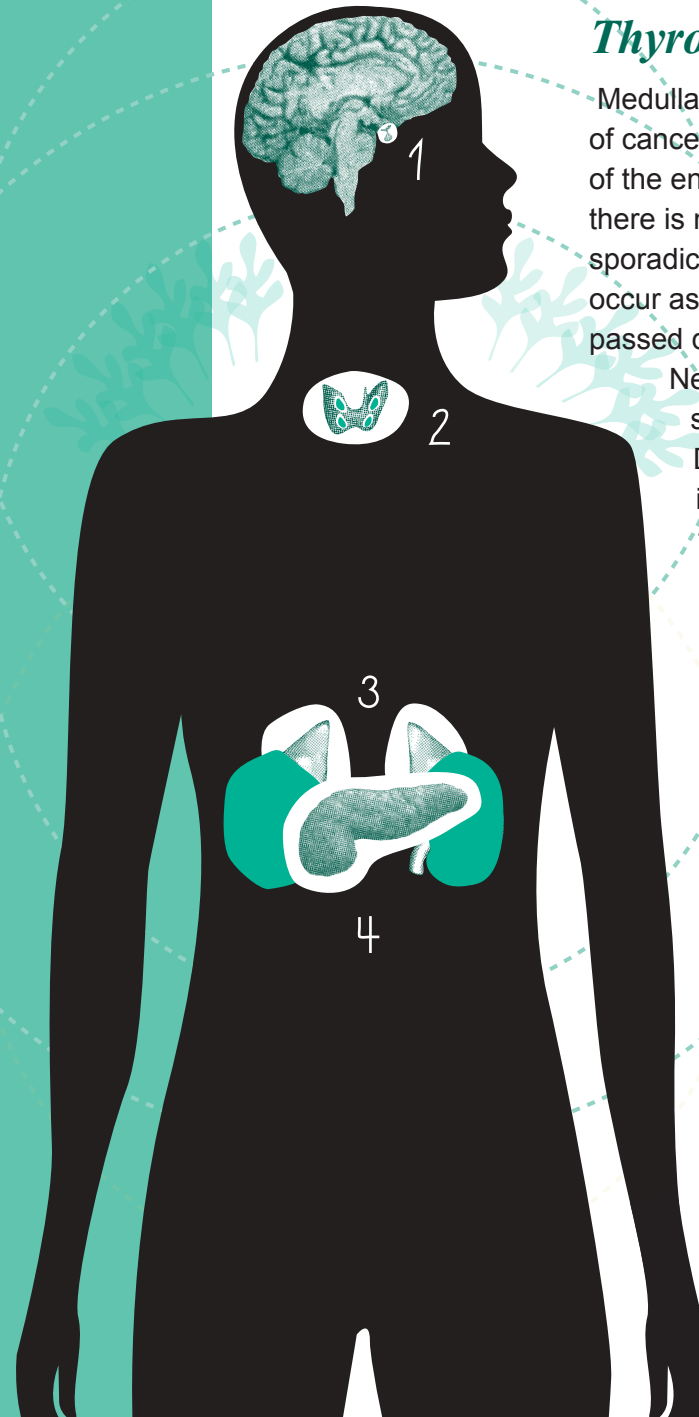
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What is Medullary Thyroid Cancer?

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 it is called sporadic MTC. 1 in 4 or 25% of cases of MTC occur as part of a rare disorder which can be passed down in families called Multiple Endocrine

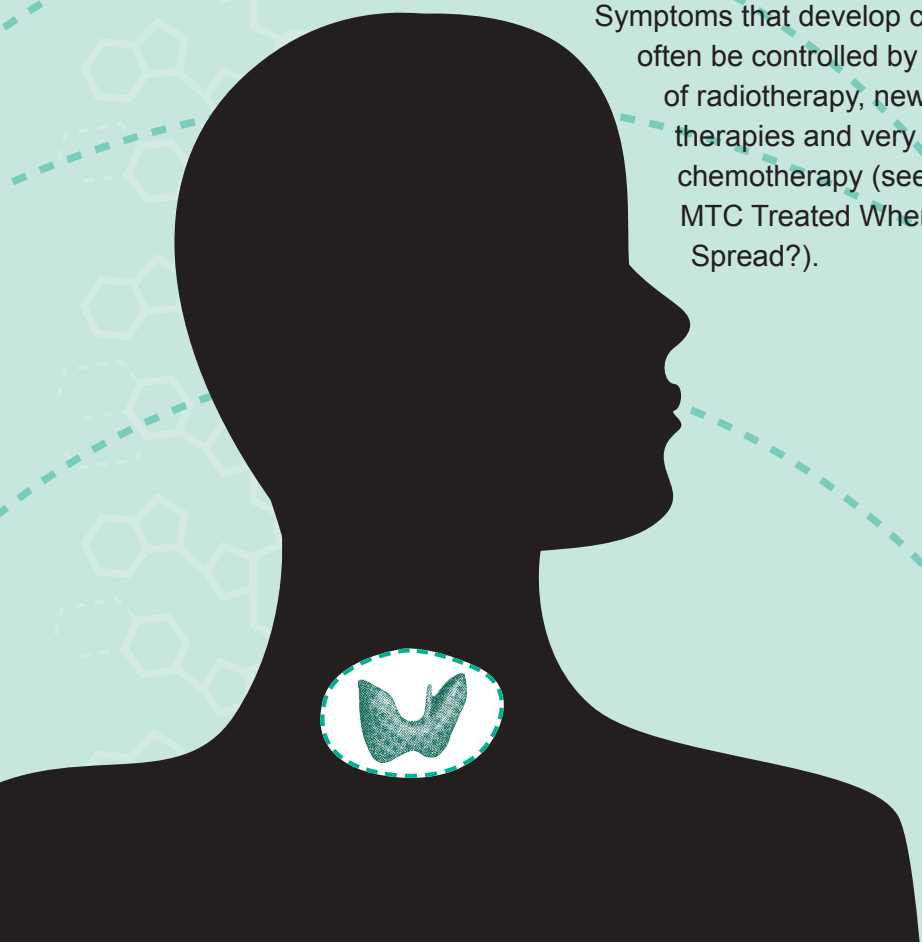
Neoplasia Type 2 (MEN2). All apparently sporadic MTC patients must be offered DNA screening (a gene test) in case theirs is the first case of a gene disorder in the family. AMEND has produced separate information booklets on the inherited forms of MTC (MEN2). The thyroid gland is found at the front of the neck. This gland makes 3 hormones; thyroxine and tri-iodothyronine (essential for maintaining the body's metabolism and mental and physical development), and calcitonin.

MTC starts in the C-cells of the thyroid which make the hormone calcitonin. An increase in the number of C-cells (C-cell hyperplasia) occurs before they become cancerous. MTC can spread to nearby lymph nodes. Even so, there may be no physical symptoms of this. As MTC grows, calcitonin levels increase.



If the thyroid and nearby lymph nodes are removed by surgery while the cancer is still contained within the thyroid (total thyroidectomy and central lymph node dissection), a patient is usually cured. If calcitonin levels are still raised after surgery, this shows that the cancer has spread (metastatic) or has not been completely removed. In this case further surgery and other therapies may be used to control it. As yet there is no complete cure for MTC that has spread; however, it is often slow-growing and can exist without symptoms for many years.

Symptoms that develop can often be controlled by the use of radiotherapy, new drug therapies and very rarely chemotherapy (see How Is MTC Treated When It Has Spread?).



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 MTC may cause diarrhoea, although it is not immediately clear that this may be related to a problem in the neck. In some families there may be a history of MTC which warrants gene testing of other family members. A diagnosis of medullary thyroid cancer may be confirmed by a fine needle biopsy of the neck lump, and ultimately by surgery (see Testing for MTC). A blood test to measure calcitonin is sometimes used to make the diagnosis of MTC, but levels may be raised for a variety of reasons other than MTC.

Children and MTC

It is rare for children to develop MTC. However, those that do should be seen by a genetics consultant to be tested for MEN2 (a

disorder that can be passed down in families), as they are likely to have this condition and will need testing for other potential MEN-related problems. A referral to a genetics consultant at a Regional Genetics Service Centre should be made by your specialist or GP if appropriate.

Testing for MTC

You may have the following tests to confirm a diagnosis of MTC:

Baseline Calcitonin Blood Test
Calcitonin levels are usually raised when MTC is present (note that once drawn, the blood must be taken immediately and on ice to a chilled centrifuge in the lab).

Ultrasound Scan and Fine Needle Aspiration (FNA)
A painless scan of the neck provides pictures of the thyroid and any lumps or cysts. A tissue biopsy (sample) is obtained from the insertion of a very fine needle into the thyroid lump. The sample is looked at under a microscope and is a very reliable way of diagnosing MTC.

Treating MTC

MTC is different from other types of thyroid cancer. It is best treated in a hospital that is a centre of expertise for MTC, and by an experienced endocrine or head-and-neck surgeon who regularly operates on such patients.

Once a diagnosis of MTC has been made, an ultrasound scan of the neck should be done to help map the extent of the disease (staging ultrasound). If there is no suspicion of enlarged lymph nodes, removal of the thyroid together with central lymph nodes is done (thyroidectomy with central node dissection). If enlarged nodes are detected, removal of other neck lymph nodes (lateral neck) should be undertaken at the same time as the thyroidectomy. If MTC is diagnosed, special blood and urine tests must be done (even if there is no family history of MEN2) to rule out a growth of the adrenal gland which could cause problems during or after surgery.

SURGERY

Total thyroidectomy + central node dissection A cut is made at

the base of the front of the neck from which the thyroid and nearby lymph nodes can be removed. A larger cut is needed if the cervical neck lymph nodes need to be removed as well. Eating and drinking is possible almost straight away after waking up from the operation.

Hospital Stay

You will probably stay in the hospital for around 2-3 days in total.

Risks

Thyroid surgery is generally safe but there are some possible risks that you need to be aware of:

- Injury to the nerves that control the vocal cords which may affect the voice (1-2%).
- Unavoidable removal of or injury to the parathyroid glands may occur which might result in a temporary drop in calcium levels in the blood (hypocalcaemia). Sometimes this may be permanent. Symptoms of low blood calcium include tingling lips, fingers and toes, and eventually cramping. All these symptoms can be corrected with medication.

Often there are no obvious symptoms of very mild low calcium

although some subtle signs are shown in the table below:

Symptoms of HYPOcalcaemia		
- calcium	- - calcium	- - - calcium
<ul style="list-style-type: none">• Tingling of the face and extremities (fingers and toes)• Pins and needles in the face and extremities (fingers and toes) <p>These symptoms may be worse when crossing one's legs or sitting on the toilet, for example.</p>	<p>all of the previous plus:</p> <ul style="list-style-type: none">• Muscle cramps• Clawing of hands or feet (tetany)• twitching of the face• spasm of the hand and fingers	<p>all of the previous plus:</p> <ul style="list-style-type: none">• Convulsions• irregular heartbeat• Difficulty breathing <p>Over a long period of time, low calcium can lead to:</p> <ul style="list-style-type: none">• osteoporosis• cataracts

What Medication will I need to take?

Levothyroxine

Levothyroxine (Thyroxine) must be taken life-long after a total thyroidectomy. Tablets are taken once a day and doses are based on the body weight of the individual (typically between 100-150mcg for adults, lower for children). You will need regular blood tests to make sure that you are on the right dose. A dose that is too big may cause symptoms such as rapid

heartbeat, sweating, anxiety, shaking and weight loss. A dose that is too small may cause symptoms such as tiredness, a slow heartbeat, sensitivity to cold, and weight gain. Although the above symptoms may suggest a need to change the dose, the same symptoms can occur in other conditions. Only a blood test measuring the thyroid stimulating hormone (TSH) level can help doctors be sure when a change of dose is needed. Once your ideal dose is found, as judged by blood

tests, repeat tests only need to be done once a year.

Calcium replacement medication

(required if parathyroid glands are injured or unavoidably removed along with the thyroid)

Calcium Carbonate (*Calcichew, Adcal*)

This is a chalk-like tablet that has to be chewed or sucked. This is often used as a short term “top-up” after surgery, but is not always needed life-long. If the patient begins to suffer from headaches, nausea and vomiting, this may show that the dose is too high or that these tablets are no longer needed.

Vitamin D3 (*cholecalciferol*)

This more common form of Vitamin D is given in capsule form but needs PTH to work to keep your calcium level normal. If this does not work, you may need to move to taking a Vitamin D Analogue.

Vitamin D Analogue (*calcitriol*)

This ‘activated’ form of Vitamin D may be given in a capsule form to help the body absorb calcium from the diet. It is taken once a day and is often the only life-long medicine needed after parathyroid surgery. This is used if your body cannot make parathyroid hormone (PTH) and needs more regular tests to make sure that calcium levels do not rise to high.

Magnesium supplement

This may be in the form of an injection or a tablet but is rarely needed long-term.

Recombinant PTH

If hypocalcaemia cannot be controlled using Vitamin D, then some centres may be able to offer Recombinant PTH which replaces the hormone made by the parathyroid glands. These drugs are not yet approved for use by the NHS, but may be accessed on a named patient basis or through clinical trials.

How is MTC treated when it has spread?

Patients with MTC may have high levels of calcitonin in the blood even after surgery. However, although this shows that there are MTC cells left in the body, patients with calcitonin levels that are higher than normal, staying the same over a period of time, or slowly increasing, often do not need further tests or treatment. This is because scans are not always able to find a site of disease outside of the neck unless calcitonin levels are very high: calcitonin alone is not proof of a growing tumour. Even so, in some patients, the search for metastatic disease may involve various scans (including radioactive isotope scans), followed by treatment with more surgery or radiotherapy needed.

C-cells also produce a substance called carcinoembryonic antigen (CEA) which is measured

alongside calcitonin to help gather more information about disease that has spread outside of the neck.

MIBG / OCTREOTIDE THERAPY

Where surgery is no longer an option due to the extent or site of the disease, some expert medical centres may use radioactive treatment (octreotide or MIBG). These have very few side effects and can help to reduce or control the spread of MTC. However, they are only used if tests show that they will be taken up by the tumour. The agent is given through a vein by slow injection. The patient remains radioactive for a few days and therefore must be nursed alone in a special room. The treatment may need to be repeated several times at 3 or 6 month intervals. Possible side effects of MIBG / Octreotide therapy include feeling sick, and sometimes being sick too.

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 A change to the patient's diet may be required, together with medicine such as *Imodium* which can help to control it. In some cases diarrhoea can also be relieved by treatment with drugs called octreotide or lanreotide, although this is not the case for everyone. Some believe that in such cases it may also help slow down the growth of the tumour.

Flushing Medicines used to control stomach ulcers called H2 blockers (*cimetidine or ranitidine*) may sometimes be used to help ease flushing.

Painful bone metastases

External radiation therapy can sometimes provide rapid relief from pain when MTC has spread to the bone. In all cases, pain medications may be prescribed.

OTHER THERAPIES FOR MTC THAT HAS SPREAD

A class of drugs called Tyrosine Kinase Inhibitors (TKIs) and Multi-Kinase Inhibitors (MKIs)

are becoming available for use in metastatic MTC where other treatments do not work. These drugs are not cures, but may in some people help slow or stop the spread of MTC as well as relieve symptoms. At the moment, the drugs *vandetanib* and *selpercatinib* are approved for use in the UK. You should discuss this option, the possible serious side effects and other issues about these therapies with your specialist. TKIs and MKIs are not always suitable for everyone.

Vandetanib: This drug is taken in the form of a tablet once a day. It helps to block chemical messages that MTC needs to spread. Side effects may include:

- loss of appetite
- diarrhoea
- high blood pressure
- headaches
- tiredness

Your doctor will use tests to make sure that you find the right dose of the drug to help maintain a good quality of life.

Selpercatinib: This drug is also taken in the form of a tablet once a day and may be used either if *vandetanib* does not work or if it stops working after time. It blocks very specific chemical messages to stop MTC from spreading. Side effects are less common but may also include those listed above. This drug currently has a temporary approval for use in England and Scotland while more data is collected on its effects.

Research into MTC is ongoing. There are other drugs in the pipeline that are currently being tested on animals and humans. To view the various human clinical trials, please visit the Research section of the AMEND website.

Emotional Well-being

Living with a rare cancer like MTC is not always easy. Some people cope better than others, but most people will have periods of low mood at some point along the way. It is now recognised that overall health depends upon the health of both the body and the mind. For this reason, AMEND offers a free telephone counselling service to registered members. AMEND's Counsellor is also sometimes available for face-to-face sessions at our free events. See our website for more details.

AMEND has developed some specific resources about the psychological impact of living with rare disorders that we are sure our MTC patients will find useful. A range of leaflets on mental health, including 'Dealing with Diagnosis' and 'Living with Uncertainty' are free to download from the Resources section of our website. In addition, the project has produced a series of free podcasts and a video on Mindfulness. These are also available via our website.

Useful Information

Free Prescriptions In the UK, if you are to take lifelong thyroxine, you are entitled to free prescriptions for all medicines. You should obtain a FP92A application form from your doctor and complete parts 1 and 2. Your doctor will then sign it and send it on. You will then receive a Medical Exemption Certificate, which you must show to your pharmacist when collecting medicines. You can find more information on Medical Exemption Certificates on the following website: www.nhsbsa.nhs.uk/HealthCosts/2095.aspx

MedicAlert Emblem® 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. Other medical identification products are available.

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.

Useful Organisations

The British Thyroid Foundation

Tel: 0870 770 7933
www.btf-thyroid.org

Butterfly Thyroid Cancer Trust

Tel: 01207 545469
www.butterfly.org.uk

Macmillan Cancer Relief

Tel: 0800 808 0000
www.macmillan.org.uk

Parathyroid UK

Tel: 01342 316315
<https://parathyroiduk.org>

AMEND Medical Advisory Team

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Glossary

Adrenal Glands a pair of walnut-sized organs found above the kidneys that make stress hormones

Calcitonin a hormone made by the C-cells of the thyroid gland that has no known action in healthy people

CEA short for carcinoembryonic antigen; a substance that can be measured as a marker for MTC

Chemotherapy cancer treatment using chemicals

DNA short for deoxyribonucleic acid; the carrier of genetic information, stored in every cell in the body

Endocrine Glands organs in the body that make and release hormones which affect the activity of other organs

Hormones chemical messengers in the body which drive different processes by controlling the function of many different organs

Hypercalcaemia a state of having too much calcium in the blood

Hypocalcaemia a state of having too little calcium in the blood

Multiple endocrine neoplasia (MEN) a condition that can be passed down in families where more than one endocrine gland may develop a tumour

Osteoporosis a condition caused by having hypocalcaemia over a long time period making bones break more easily than normal

Parathyroid Glands four small organs found in the neck that make parathyroid hormone (PTH)

Radiotherapy a form of cancer treatment that uses X-ray radiation to destroy cancer cells

Thyroid Gland a butterfly-shaped organ found in the neck that makes and releases hormones called thyroxine and triiodothyronine

Ulcer a painful sore on or inside of the body

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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Written by Jo Grey (AMEND CEO) with the help of the AMEND Medical Advisory Team

Gunning Fog Index: 12:70

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About AMEND

AMEND is a Charitable Incorporated Organisation registered in England and Wales (number 1153890). It provides support and information services to families affected by multiple endocrine neoplasia and related endocrine tumours including MTC. AMEND encourages research into the conditions by awarding annual medical prizes and research awards. It hosts regular free patient information events every year and runs social media forums connecting patients from around the world.

Please visit our website for more information on AMEND or to make a donation: www.amend.org.uk

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