

The Management of Thyroid Cancer

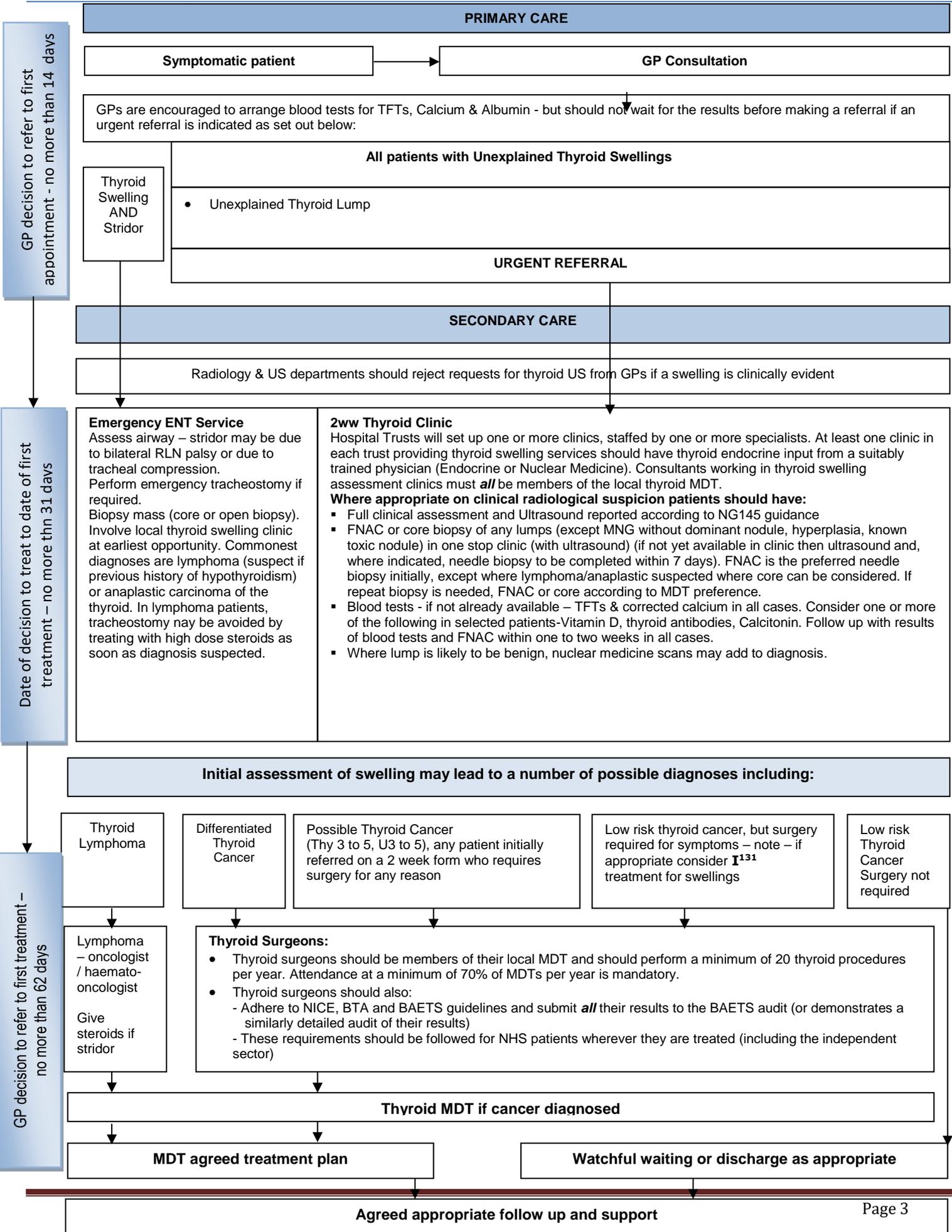
Pathway of Care

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1.0 Pathway Overview



2.0 Notes on Thyroid Swelling Assessment Clinics

- Each NHS Trust in Kent which wishes to treat patients with thyroid swellings of any form will set up one or more designated thyroid swelling assessment clinics and will apply to the Kent & Medway Cancer Collaborative (KMCC) for approval as a thyroid cancer unit. **All patients with thyroid swellings / lumps should be referred initially to one of the swelling assessment clinics** (except those with bilateral goitre *and* thyroid dysfunction, without 'urgent' symptoms, who will usually be seen in a routine thyroid endocrine clinic). Clinical Commissioning Groups (CCGs) will notify GP's that patients with thyroid swellings should not be referred elsewhere. Radiology and Ultrasound departments should reject requests for thyroid ultrasound from GP's if a swelling is clinically evident – these patients should be referred via the pathways above, to avoid diagnostic delay. Clinicians who do not work in a thyroid swelling assessment clinic should not accept referrals from GP's of patients with thyroid swelling but should redirect them, without delay, to a thyroid swelling clinic. The only exception to this is that Physicians with endocrine training may continue to accept referrals for bilateral thyroid swelling if hyper- or hypothyroidism is present, and there are no urgent 'triggers'.
- The clinicians in the thyroid swelling clinics will usually be from at least one of four specialties – Endocrinology, Nuclear Medicine with thyroid endocrine training, General Surgery with endocrine interest, ENT surgery with thyroid interest. The clinics may be run by one specialist, or several, but must a) be agreed between the local Trusts and the Tumour Site Specific Group (TSSG) and b) must comply with the pathways set out in this document. The clinicians involved should be members of the thyroid Multidisciplinary Team (MDT). There should be a trained individual who can perform needle biopsy in the clinic (or an adjoining area), or access to that within 7 days. Trusts should eventually aim to include a cytopathologist and ultrasound guidance in these clinics in the future. Small gauge trucut biopsy, either presented to the pathology laboratory in the form of a smear or as a core biopsy, is an acceptable alternative to Fine-needle aspiration cytology (FNAC), but the TSSG advises the FNAC as the initial needle biopsy in most cases. If lymphoma or anaplastic cancer suspected, a core should be considered initially. Arrangements should allow reporting of the cytopathology *on the same day* where appropriate.
- By referring directly to the clinics set out above, patients should have a complete diagnosis made at the earliest opportunity, and other necessary treatment will be arranged without delay.
- If more than one treatment option is available, patients must be fully informed so they may make a decision based on all available treatment options. For example, a patient with benign nodular hyperplasia and compression symptoms should be informed of the surgical and nuclear medicine options for treatment.

3.0 Referral

Patients will be referred according to the scheme set out in the pathway overview in section 1.

3.1 Patients with thyroid swelling and stridor

Patients with thyroid swelling and stridor should be referred, urgently, to the ENT emergency service.

3.2 Urgent referral to the thyroid swelling assessment clinic

Patients with the following symptoms should be referred, urgently, to the thyroid 2ww swelling assessment clinic:
All patients with unexplained thyroid swelling

4.0 Thyroid multidisciplinary team meeting

- Core team members:
 - At least two surgeons who specialise in thyroid/endocrine oncology (each of whom should be performing at least 20 thyroid operations a year)
 - An endocrinologist
 - A clinical oncologist
 - Specialist Pathologists (both histopathology and cytopathology)
 - Radiologist
 - A nuclear medicine specialist
 - Clinical Nurse Specialist (who may be a H&N CNS)
 - MDT-Co-ordinator
- One of the core members will be responsible for patient and carer issues
- One of the above will be the named chair of the MDT and will have a job description outlining the key responsibilities of the chair agreed with the Trust Lead Cancer Clinician (or Trust Medical Director)
- One of the core members will be responsible for service improvement
- To be a core member of the MDT, participation in a minimum of 70% of MDT discussions per year is mandatory. Participation may be by attendance, or by video-link. In the case of video link participation, the individual should participate in, and thus contribute to, the whole meeting – not just that part of the meeting which deals with his/her patients
- At least one core member of the MDT should attend at least 70% of Thyroid TSSG meetings
- The MDT leads will provide an annual summary of arrangements, including defined membership, to the chair of the TSSG. This is a governance requirement to ensure ongoing compliance with national standards
- There will be two thyroid MDT's – one based at the Kent Oncology Centre, Maidstone and the other at the East Kent Hospitals Trust. Both MDT's leads will ensure the pathway, as set out here, is followed and that the organisation of the MDT's also meets the nationally agreed standards
- MDTs should meet at least monthly

4.1 Thyroid Surgeons

- Thyroid surgeons should be members of their local MDT and should perform a minimum of 20 thyroid procedures per year. Attendance at a minimum of 70% of MDT's per year is mandatory.
- Thyroid surgeons should also:
 - Adhere to National Institute for Health & Care Excellence (NICE), British Thyroid Association (BTA) or British Association of Endocrine & Thyroid Surgeons (BAETS) guidelines and submit *all* their results to the BAETS audit (or demonstrate a similarly detailed ongoing audit of their results).
 - Thyroid surgery should not be undertaken by surgeons who are not members of their local MDT. These requirements should be followed for NHS patients wherever they are treated (including the independent sector)
- The majority of operations for thyroid cancer consist of hemi – or total thyroidectomy, combined with a central compartment neck dissection where indicated, and all surgeons who are MDT members should have the skills necessary to undertake this surgery. However each MDT should agree which of the surgeons within the MDT are able to undertake 'complex' thyroid surgery. Complex cases are generally those requiring lateral neck dissections or those requiring tracheal resection. Because of the geographically spread out nature of the

population in Kent, it is proposed that all trusts who currently provide thyroid surgical services continue to do so, provided that all the surgeons involved meet the criteria laid down above.

- The TSSG recognises that each contributing NHS Trust is likely to have a number of surgeons who currently undertake thyroid surgery. Each Trust should provide to the TSSG the names of surgeons who will continue to work within the framework set out above. The reporting Trust should confirm that the nominated surgeons meet (or will meet) the criteria set out above. Trusts will be asked to confirm this information on an annual basis. It is hoped that Trusts will continue to support all those surgeons who undertake regular thyroid surgery, wherever possible. However, where the minimum criteria of 20 thyroid operations per year cannot be met by individuals (i.e. too many surgeons in the Trust wanting to continue to undertake thyroid surgery), then the Trust will need to decide which surgeons may continue to undertake thyroid surgery and be members of the MDT. Trusts will need to decide whether they want a number of surgeons to attend the MDT, provided the minimum 20 procedures can be met, or whether rationalisation is required in the interest of the Trust – i.e. not having an unnecessarily large number of surgeons attending the MDT. Trusts should ensure that there are adequate cross-cover arrangements for leave etc.
- These arrangements have the support of the members of the thyroid TSSG, but it is recognised that further rationalisation may be required if central guidance makes this imperative. Treatment of proven thyroid cancer usually consists of total thyroidectomy (with parathyroid preservation where possible), and post operative radioiodine therapy. If a simultaneous neck dissection is required, an appropriate surgeon should be involved. Where a cancer is diagnosed following partial thyroid surgery, and a completion thyroidectomy is required, this should be performed by a surgical member of the MDT within four weeks of the date on the diagnostic histopathology report.

4.2 Clinical Oncologists

Each MDT should have a Clinical Oncologist as a core member. The role of the clinical oncologist is to advise specifically about the role of radiotherapy, radioactive iodine (unless this is undertaken by a Nuclear Medicine Specialist for that patient) and, rarely, systemic therapy, and more generally about the general support of the patient with cancer. Patients considered at low risk of recurrence need not see an oncologist (unless he/she is administering the radioactive iodine) but those with macroscopic residual disease, local recurrence or distant metastasis should do so.

4.3 Nurse Specialists and other support disciplines

All patients to be referred to Clinical Nurse Specialist (CNS) at same time as the patient is told the diagnosis. The Head and Neck Cancer CNS's will cover the thyroid cancer service. Wherever possible a CNS should be present the 'breaking of bad news' meeting. The CNS's will liaise with other health professionals as required including Macmillan Radiographers, Speech and Language therapists and dieticians.

4.4 Thyroid Physicians

Thyroid physicians may be endocrinologists or nuclear medicine physicians with thyroid endocrine training *and* specific experience of dealing with patients who have thyroid cancer. To participate in thyroid swelling assessment clinics, and to treat patients with thyroid cancer (including post-treatment follow-up), these physicians must be core members of the local MDT. In setting up thyroid swelling clinics, the Trusts involved will need to ensure that the clinicians involved meet the criteria for core membership of the MDT.

4.5 Thyroid Clinicians in general

Wherever possible, the TSSG supports all clinicians currently involved in the treatment of thyroid cancer in Kent who wish to continue to treat these patients. Clinicians will need to adapt their practices according to those elements of the pathway which are compulsory in the Improving Outcomes Guidance (IOG). When clinicians retire, or otherwise leave the local service, Trusts should liaise with the Chairman of the TSSG (acting in an advisory role) *before* appointing replacement thyroid cancer clinicians (includes surgeon, physician, pathologist, and radiologist) – this will allow areas of need to be identified, and also avoid duplication of resources between Trusts. Trusts should not assume new appointees will be able to join the local MDT unless there has been discussion prior to the appointment. This requirement is in line with the Manual for Cancer Services 2004, 2.2.

5.0 Data Collection

Collection of data at each stage of the pathway is the responsibility of the team looking after the patient at that time. The minimum dataset agreed by the TSSG will be a combination of those data items that meet national requirements, and additional items as agreed by the TSSG.

National data requirements will include:

- Cancer Waiting Times monitoring, including Going Further on Cancer Waits. The data items required will be as defined in ISB0147 at the time of referral and/or treatment.

Cancer Waiting Times data will be submitted according to the timetable set out in the National Contract for Acute Services.

- The Cancer Outcomes and Services Dataset. The data items will be as defined in ISB1521, and any subsequent versions, at the time of diagnosis and/or treatment. The requirement will include those fields listed in the “Core” section of the dataset, and any additional tumour site specific sections, as applicable.

Details of the COSD are available from:

http://www.ncin.org.uk/collecting_and_using_data/data_collection/cosd.aspx

Cancer Registration and Cancer Outcomes and Services (COSD) data will be submitted according to the timetable set out by National Cancer Registration Service (NCRS).

- Where applicable, teams will also collect additional data items as defined in any corresponding National Clinical Audit Support Programme (NCASP) and /or HQIP Quality Accounts audit dataset(s).

Details of these datasets are available from:

<https://www.hqip.org.uk/wp-content/uploads/2020/01/nhs-england-quality-accounts-list-2020-21-vjan2020.pdf>

Data for NCASP audits will be submitted, where applicable, according to timetables as agreed by the TSSG, and within the overall submission deadlines for each audit.

Submission of data to meet these national requirements will be the responsibility of each individual Trust.

Note that these standards are subject to variation from time to time, and where these requirements change, the data items required to be collected by the team will also change in line with national requirements.

Local data requirements will include any additional data items as agreed by the TSSG. These must be selected to avoid overlap with any existing data items, and where possible must use standard coding as defined in the NHS Data Dictionary.

Where possible and applicable, InfoFlex will be used for the collection and storage of data.

Additional areas of the COSD, relating to pathology, radiotherapy, Systemic Anti-Cancer Therapy (SACT), diagnostic imaging and basic procedure details will feed into the dataset from other nationally mandated sources. It is the responsibility of each team to ensure that the whole of the relevant dataset is collected, and it is acknowledged that this may come from a variety of sources.

It is a mandatory requirement that all surgeons recognised for thyroid cancer surgery collect data on outcomes and upload this data to an appropriate national database for access by patients. All thyroid surgeons who are members of MDTs covered by this Pathway of Care are strongly encouraged to upload their thyroid outcome data onto the British Association of Endocrine and Thyroid Surgeons (BAETS) national audit.

6.0 Histopathology and Cytopathology

- Pathology from the thyroid swelling clinics should be reported by a pathologist specialising in thyroid cytology / core biopsies
- FNAC / Core biopsy should be performed at initial assessment. Where core biopsy is used, material may be submitted as a core, or in the form of smear on slide
- Cytologists to report using Thy score 1 to 5: 1 - Not diagnostic, 2 – benign, 3a – atypical, 3f – follicular lesion, 4 – suspicious of malignancy, 5 –diagnostic of malignancy
- If decision is made to put patient into watchful waiting pathway, FNAC / core biopsy to be repeated after four weeks – changes diagnosis in 1:50 patients
- Pathology samples will be processed and reported in accordance with the guidelines set out in the Kent and Medway Cancer Collaborative Pathology Guidance.

All Kent & Medway Cancer Collaborative reporting pathologists follow The Royal College of Pathologists Histopathology Reporting on Cancers guidelines.

7.0 Imaging

KMCC imaging guidelines are located on the KMCC website on the following link:

<http://www.kmcc.nhs.uk/tumour-sites/sub-groups-or-cross-cutting-groups/diagnostics-group>

Radiologists and ultrasonographers who are undertaking thyroid ultrasound should report according to NG145 standards. Ultrasounds should be reported on the day they are performed and should include the following information:

- Size and characteristics of left and right lobes of thyroid
- Position, size and U grade of thyroid nodules seen on ultrasound
- A comment on the presence or absence of pathological cervical lymph nodes, with full details of any pathological nodes
- Where FNAC or Core biopsy is indicated, this should be usually be performed in the same imaging session.

8.0 Surgical Provision

- All surgical patients with high risk of (or proven) cancer, any patient with voice change, and all patients with previous thyroid surgery, to have preoperative laryngoscopy to assess vocal cord function. This procedure is advisory in *all* surgical patients.
- All large MNG with pressure symptoms to have preoperative CT neck and Chest. There is usually no need for preoperative CT in other patients unless stridor or hoarse voice. If CT requested, consider whether it is appropriate to use contrast as this blocks iodine isotope uptake for up to 8 weeks.
- Surgery should consist of total thyroidectomy, hemithyroidectomy or isthmusectomy. Partial thyroidectomy should not usually be performed.
- All patients undergoing total/completion thyroidectomy for malignancy should usually be started on T4(levothyroxine) post operatively. Nearly all patients requiring RAI will have that administered with recombinant TSH – and therefore it is unusual to need to send a patient home on T₃ (liothyronine). If there is a case where thyroid hormone withdrawal is considered for RAI, this should be subject to MDT discussion and any patient in this category may be considered for T3 on discharge. Patients undergoing total thyroidectomy for benign disease to be started on thyroxine before discharge.
- All patients undergoing hemithyroidectomy to have TFT's performed approximately 6 weeks to three months after surgery.

8.1 Complex Surgery

The KMCC has agreed which surgeons may perform lymph node surgery (except level 6 which may be performed by all thyroid surgeons across K&M). The TSSG has agreed the surgeons / sites able to perform this work - they are, Mr Alistair Balfour, Mr Vikram Dhar, Mr Chris Theokli, Mr Rob Hone and Mr Ali Al-lami (East Kent), Mr John Shotton (Tunbridge Wells), Mr Jeremy Davis, Mr Praveen and Mr Nav Upile (Medway).

8.2 Post operative hypocalcaemia

Temporary early postoperative hypocalcaemia is common after surgery. This must be identified but requires treatment only if severe. Serum calcium should be measured within the first 24 hours after surgery and regularly thereafter until normal levels are achieved by spontaneous recovery or institution of calcium and/or vitamin D therapy. A suggested pathway is shown below. (Trusts should confirm to the TSSG that they are either using this pathway, or have a similar formal pathway of their own).

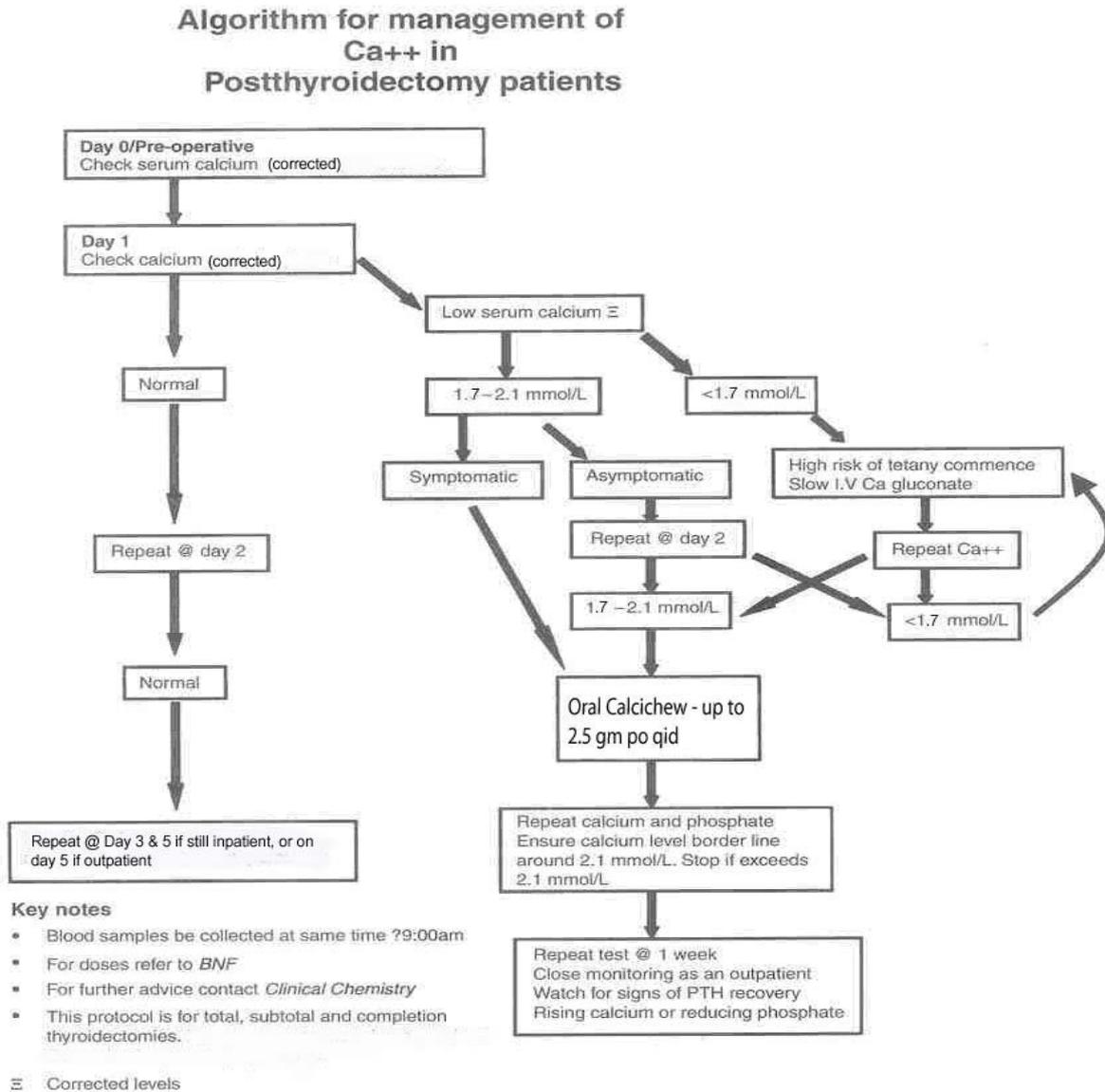
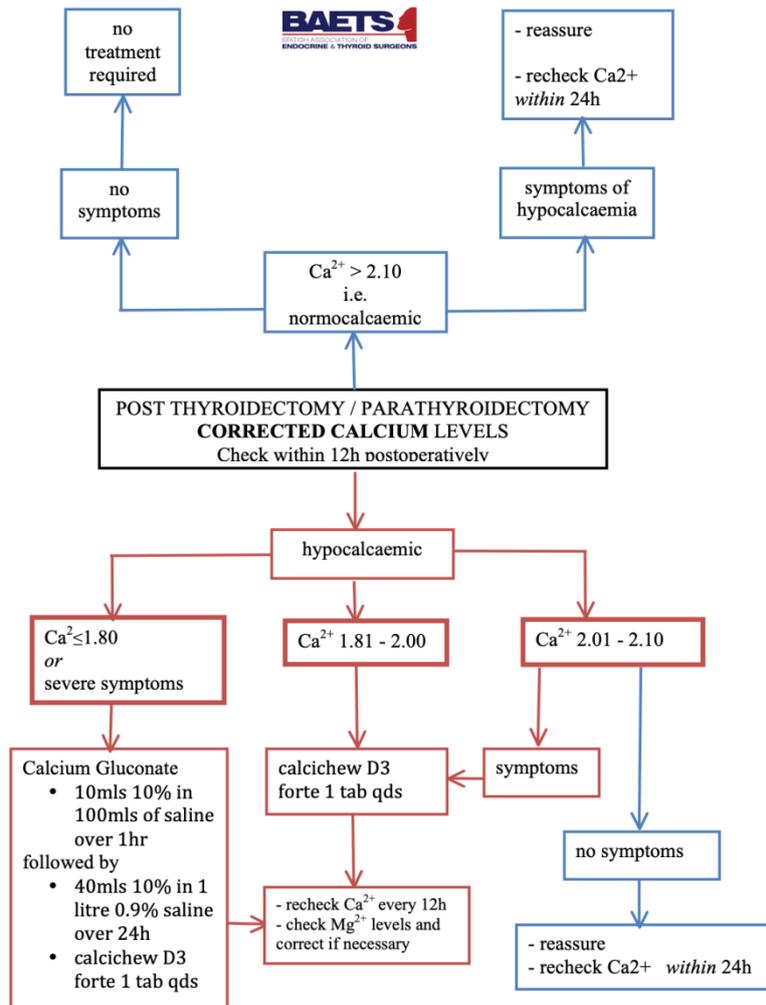


Figure 1. Tunbridge Wells protocol.



Postoperative calcium supplementation in patients undergoing thyroidectomy.
Wang TS, Roman SA, Sosa JA. Curr Opin Oncology 2012; 24(1): 22-28

All Patients having total or completion thyroidectomy to have serum calcium checked on day 1, 2 and (where defined by the local protocol) on day 5 postoperatively – and arrangements made to contact patients with results and take clinical action as appropriate within 24 hours of blood being sent to laboratory.

Note: Pathway deliberately does not utilise vitamin D analogues. These should only be used in the severely hypocalcaemic patient (Corrected Ca less than 1.6mmol/l) – and then should be monitored closely with a view to stopping this treatment as soon as possible.

9.0 Post Surgical Treatment

9.1 Radioiodine (where appropriate)

- In general, radioactive iodine (where indicated) is given to all differentiated cancer patients following completion thyroidectomy, within 3 months of surgery. Only patients who have tumours above PT1b should be considered for RI or entry into trials.

- Nuclear Medicine Specialists or Clinical Oncologists prescribing radioiodine therapy must be ARSAC certificate holders and core members of the Thyroid MDM (Dr Acosta – Medway and EKHUFT cover), Dr Rowell - Maidstone).
- Radionuclide Radiologists and Nuclear Medicine Specialists responsible for the conduct of imaging studies must be core members of the Thyroid MDM.
- Radionuclide imaging (tracer) studies are not performed prior to the first radioactive iodine treatment.
- Post treatment imaging (with neck and whole body images) is carried out on all patients. Images must be viewed by a radionuclide radiologist or nuclear medicine specialist before the patient leaves the department so that the need for extended imaging or additional views can be determined.
- Serum TSH, thyroglobulin and antithyroglobulin antibodies are measured immediately prior to each radioactive iodine treatment.
- Further radioactive iodine treatment is given as clinically indicated following MDM discussion.
- Arrangements must be in place to ensure proper facilities for treatment, Medical Physics support, and radionuclide radiologist or nuclear medicine specialist supervision and support.

9.2 External beam radiotherapy

External beam radiotherapy should be considered for patients with:

- Macroscopic residual disease
- Unresectable locoregional disease
- Selected cases of recurrent nodal disease (postoperatively)
- Sites of distant metastasis (principally bone)

9.3 Anti-Cancer Drugs

Systemic Therapy may be considered for patients of good performance status and disease which is unresectable and resistant to radioactive iodine.

A copy of the KMCC Head & Neck & Thyroid Oncological Guidance can be located on the KMCC website:-

<http://www.kmcc.nhs.uk/medicines-and-prescribing-incorporating-sact-pathways/oncological-treatment-guidelines/>

10.0 Follow Up

Where Differentiated Thyroid cancer has been treated by hemi-thyroidectomy only due to the low risk of recurrence, TSH suppression is not indicated and long-term follow-up in secondary care is not indicated.

Other patients with DTC should have a post-operative risk stratification to determine if they are Low, Intermediate or High risk for recurrence. At 9-12 months post-treatment patients will require Dynamic risk Stratification to reassess response to treatment and re-classify as low, intermediate or high risk. The follow up of thyroid cancer should be lifelong, to ensure adequate TSH suppression for differentiated cancer where indicated.

- Where the patient is classified as low risk, and TSH suppression is not indicated and should be maintained in the low-normal range, follow up for 5 years, usually annually, is suggested. Individual decision making is important and more frequent follow up, or a longer period of follow up may be needed in some patients.
- Where the patient is classified as intermediate risk TSH suppression should be undertaken for 5 years. Follow-up should continue until this period is finished and thyroid hormone dose had been adjusted to bring the TSH into the lower end of the normal range.
- Where the patient is classified as high risk (including incomplete response) TSH suppression may be considered for up to ten years, and exceptionally lifelong, and follow-up is needed whilst TSH is suppressed (10) years to lifelong, with a member of the Thyroid MDT.

- During follow-up:
- Thyroid hormones, including TSH, and (in selected cases only) serum calcium should be monitored regularly.
- Thyroglobulin should be monitored in patients with differentiated thyroid cancer, and calcitonin in those with medullary cancer. MDT's should agree which assay that MDT, and its clinician members, will use for thyroglobulin measurements. Patients having thyroglobulin monitoring should also have anti thyroglobulin antibodies checked at the same time as the thyroglobulin measurement.

A small proportion of patients who have been treated for thyroid cancer may develop recurrence, sometimes many years after initial treatment. Many of these patients can be treated, and often cured, with further surgery and / or radioiodine. External beam radiotherapy may be used in addition to other forms of treatment. All such patients should be assessed and restaged, and their further management discussed by the thyroid cancer MDT.

Head & Neck and Thyroid TSSG agreed follow-up policy for intermediate and high risk patients:

years since diagnosis or relapse	frequency of appointments
First year	3-6 months
Second year	6-12 months
Third year onwards	12 months or annually. Some patients may be discharged after 5 years
After 10 years	most patients may be discharged

10.1 PET Scans

PET scans may be used if rising thyroglobulin post treatment, but no uptake of radioiodine in isotope scan.

11.0 Supportive and Palliative Care

Patients who have inoperable/recurrent/relapsed thyroid should be referred to the specialist supportive & palliative team if indicated. The prime aim of the treatment is to alleviate symptoms.

Open and frank discussions with patients should take place with patients at all stages of their journey so that patients are not confused about their prognosis or have unrealistic expectations of any of the forms of treatment offered to them.

Relatives and carers will need to be appropriately supported and given appropriate information. However, in accord with the recommendations set out in various Improving Outcomes Guidance, relatives and carers should not be given information different to that given to the patient.

A key worker will be assigned to each patient. The clinical nurse specialists will be responsible for ensuring an appropriate individual is assigned – this may vary according to the patients treatment plan – for instance for the small number of patients undergoing external beam radiotherapy, a Macmillan Radiotherapy specialist colleague may be an appropriate key worker.

Palliative care provision should be made for all patients:

- Hospital teams, including the Clinical Nurse Specialists for thyroid cancer patients
- Primary Health Care Team would provide for palliative care at home
- General Practitioner should be informed within 24 hours of the diagnosis, treatment plan and medication changes.

The management of symptoms, psychological, social and spiritual issues, and the communication of the diagnosis, and any associated problems, should be within the domain of all health care professionals.

Referral to specialist palliative care services should be considered when these issues have not been resolved and in particular for patients with:

- Complex and difficult symptom management issues
- Difficulties in adjusting to the diagnosis or disease progression
- Psychological and family issues – such as communication problems within the family
- Spiritual issues – such as the challenging of belief system/faith/cultural values as a result of the cancer

Consideration of specialist palliative care or support should be given throughout the patient pathway, particularly:

- At the Multidisciplinary Team Meeting
- When no active treatment is considered
- After active treatment
- At relapse
- In the terminal stages

12.0 Clinical Trials

Wherever appropriate patients should be considered for entry into clinical trials.

13.0 TNM Staging

UICC TNM 8TH EDITION STAGING SUMMARY			
Under 55 Years			
ANY T	ANY N	M0	Stage 1
ANY T	ANY N	M1	Stage 2
Papillary or Follicular - 55 Years and Older			
T1a, T1b, T2	N0	M0	Stage 1
T1, T2, T3	N1	M0	Stage 2
T3	N0	M0	Stage 2
T4a	N0, N1	M0	Stage 3
T4b	ANY N	M0	Stage 4a
ANY T	ANY N	M1	Stage 4b
MEDULLARY			
T1a, T1b	N0	M0	Stage 1
T2, T3	N0	M0	Stage 2
T1, T2, T3	N1a	M0	Stage 3
T1, T2, T3	N1b	M0	Stage 4A
T4a	ANY N	M0	Stage 4A
T4b	ANY N	M0	Stage 4B
ANY T	ANY N	M1	Stage 4C
ANAPLASTIC CARCINOMAS - (ALL STAGE 4)			
T4a	ANY N	M0	Stage 4A
T4b	ANY N	M0	Stage 4B
ANY T	ANY N	M1	Stage 4C

- pTX Primary tumour cannot be assess
- pTO No evidence of primary tumour
- pT1a <10mm, limited to thyroid
- pT1b <20mm but>10mm, limited to thyroid
- pT2 >20mm, <40mm, limited to thyroid
- pT3a >40mm, limited to thyroid
- pT3b Tumour of any size with gross extrathyroidal extension invading strap muscles (sternohyoid, sternothyroid or omohyoid muscles)
- pT4a Tumour invades beyond thyroid capsule and invades any of: subcutaneous soft tissues, larynx, trachea, oesophagus or recurrent laryngeal nerve
- pT4b Tumour invades prevertebral fascia, mediastinal vessels or encases carotid artery

UICC TNM 8 staging applies to carcinomas and includes papillary, follicular, poorly differentiated. Hurthle cell and anaplastic carcinomas.

Multifocal tumours (>2 foci) of all histological types should be designated (m), the largest focus determining the classification, e.g. pT2(m).

Regional lymph nodes (pN)

- pNX Cannot assess regional lymph nodes
- pNO No regional nodes involved
- pN1a Metastasis in level V1 (pretracheal, paratracheal and prelaryngeal/Delphian) lymph nodes
- pN1b Metastasis in other unilateral, bilateral or contralateral cervical (levels I,II,III,IV or V) or retropharyngeal or superior mediastinal lymph nodes

Distant metastasis (M)

- MO No distant metastases
- M1 Distant metastases

14.0 Medullary Carcinoma

Types:

Sporadic

Familial

(a) as part of multiple endocrine neoplasia type IIa

(b) as part of multiple endocrine neoplasia type IIb

(c) as familial medullary thyroid cancer without other endocrine disease (FMTC)

Although appearing microscopically identical, the behaviour of the MTC varies according to the setting. The tumour is most indolent in FMTC and most aggressive in MEN IIb. The spectrum from indolent to aggressive is: FMTC * MEN IIa * Sporadic * MEN IIb.

The familial forms are inherited autosomal dominant conditions – there is a 50/50 chance of any child of an affected parent inheriting the condition.

Suspect in:

- Upper pole tumour
- Painful / tender thyroid lump
- Family history
- Diarrhoea

FNAC may be interpreted as follicular lesion, so diagnosis may only be made after hemithyroidectomy.

As soon as diagnosis made:

Bloods

- Calcium
- Calcitonin
- PTH
- Plasma Metanephrines

Urine

- Mets / Catecholamines

Imaging

- MR Neck
- CT Chest / Liver

Treatment

- Surgery – Total thyroidectomy and ipsilateral neck dissection (MRND) *with thymusectomy* as minimum. If tumour > 2cm, known neck nodes, or measurable calcitonin 12 weeks after initial treatment, contralateral MRND also. Plasma or Urinary metanephrines and catecholamines should be checked before surgery because of risk of coexisting pheochromocytoma.
- Radioiodine – no role in medullary thyroid cancer
- Systemic therapy – has role in palliation
- Post op raised calcitonin – up to 100, prob neck, 100 to 1000, prob mediastinal, > 1000 – prob liver mets. Note – liver mets not always seen on U/S or CT – if calcitonin >1000, consider liver biopsy or laparoscopy
- All patients – post op baseline MRI neck / mediastinum at three months.
- Radiotherapy – Radiotherapy should be considered for those with extensive nodal involvement and in those with extrathyroidal extension and microscopic or macroscopic residual disease. Palliative radiotherapy may also be of benefit to those with metastatic disease.
- Genetic Counselling – in the presence of a family history suggestive of MEN 2 or FMTC family genetic screening is required. It must be remembered that absence of family history does not preclude this patient being the index case for a new kindred.
- All patients must have RET gene testing – family screening if positive.

15.0 Personnel and Contact Information

A comprehensive, up to date list of MDM contact details can be found on the KMCC website via the following link: <http://www.kmcc.nhs.uk/tumour-sites/terms-of-reference>

16.0 Glossary

Acronyms in common usage throughout KMCC documentation

CNB	Cancer Network Board
CYP	Children & Young People (in relation to the IOG)
DCCAG	Diagnostic Cross Cutting Advisory Group
DOG	Disease Orientated Group (NSSG/TSSG/TWG)
DVH	Darent Valley Hospital
EK	East Kent
EKHUFT	East Kent Hospitals University Foundation Trust
HoP	High Level Operational Policy
IOG	Improving Outcomes Guidance
IOSC	Improving Outcomes: A Strategy for Cancer
K&C	Kent & Canterbury Hospital, Canterbury, (EKHUFT)
K&M	Kent & Medway
KMCA	Kent & Medway Cancer Alliance
KMCC	Kent & Medway Cancer Collaborative
KMCN	Kent & Medway Cancer Network
KMCRN	Kent & Medway Cancer Research Network
LSESN	London & South East Sarcoma Network
MFT	Medway Foundation Trust
MTW	Maidstone & Tunbridge Wells NHS Trust
NOG	Non Surgical Oncology Group <i>(Permanent oncologist sub group of the TSSGs with a specific responsibility for chemo/rad pathways and advice to the TSSG, KMCC and geographical locations on new drugs)</i>
PoC	Pathway of Care <i>(KMCC agreed disease site specific clinical guidelines)</i>
QEQM	Queen Elizabeth the Queen Mother Hospital, Margate (EKHUFT)
QoL	Quality of life
QVH	Queen Victoria Foundation Trust Hospital East Grinstead
RAT	Research and Trial Group <i>(Permanent sub-group of the TSSGs with a specific responsibility for taking forward the clinical trials agenda)</i>
RMH	Royal Marsden Hospital
RNOH	Royal National Orthopaedic Hospital
TSSG	Tumour Site Specific Group
UCLH	University College Hospital London
WHH	William Harvey Hospital, Ashford (EKHUFT)
WK	West Kent
MDT	Multidisciplinary Team
NICE	National Institute for Health & Care Excellence
BAETS	British Association of Endocrine & Thyroid Surgeons

BTA	British Thyroid Association
SACT	Systemic Anti-Cancer Therapy

17.0 Document Administration

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Revision History			
Date of revision	New Version Number	Nature of Revision	Confirmation of Accuracy by
June 2005 to June 2006	1 to 5	Initial draft – development of robust working document	J.Davis
July 2006	6	Draft – formatting	A.Jackson
July to November 2006	7	Final – final amendments from DOG members	J.Davis
November 2006	8	Final – final amendments from DOG members	C.Evans
December 2006	9	Final – final amendments from DOG members	C.Evans
February 2007	10	Published – final review for corrections	J.Davis/A.Jackson
September 2010	11	Update	J.Davis/A.Jackson
September 2010	12	Update – final updates (follow up and contacts)	C.Tsatsaklas/J.Davis
March 2012	12.1	Draft – updated all weblinks inc. imaging, pathology & contacts; general formatting & content checking	S.Stanley/C.Tsatsaklas
September 2012	12.2	Draft – data collection section updated / general formatting	A.Brittle/C.Tsatsaklas
November 2012	13.0	Final/Published – agreed by the Thyroid DOG 20/11/12	Thyroid DOG /I.Vousden/C.Tsatsaklas
March 2014	13.1	Draft – general admin updates made (i.e. removal of Cancer Network/DOG etc)	C.Tsatsaklas
March 2014	13.2	Draft - corrected the details for: surgeons for East Kent; Radioactive iodine personnel for East Kent. Rewritten the TMN staging as it has been updated	M.Harron
March 2014	13.3	Draft – final changes made as agreed in the TSSG	C.Tsatsaklas/M.Harron
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June 2014	14.0	Final/Published – ratified by the Operational & Quality Group (25/6/14)	Operational & Quality Group
May 2015	14.1	Pathway changes to 5.0, 6.0 and 8.1 by M.Harron and D.Hannant	M.Harron/D.Hannant
December 2015	14.2	Amendments as agreed in the November TSSG meeting. Sent to P.Ryan for review before ratification.	N.Aluwalia/P.Ryan/TSSG
February 2016	14.3	Revised documents received from P.Ryan, TSSG ratification now due.	P.Ryan/N.Aluwalia
March 2016	14.4	Amendments to abbreviations	N.Aluwalia
May 2016	14.5	Further Acronyms added/Amendments and addition of timeline	N.Aluwalia/ L.Caine
June 2016	15.0	Ratified final document to be published following grammatical corrections and O&Q Group meeting	N.Aluwalia
January 2021	15.1	Draft - Revision of full document & follow up	J Davis/A Wiltshire
June 2021	16.0	Final – full revision of document & follow up section	J Davis/A Wiltshire