Welcome to this NDRS training module on tumours of the small intestine which has been designed to help Cancer Administration staff gain a better understanding of the diseases and the terminology used by the clinical teams.

Agenda

- Small intestine
- Summary
- Acknowledgements

This module may be paused at any time

We're going to give you a brief introduction to tumours of the small intestine including some of the symptoms that patients might experience. We'll look at the anatomy & physiology will then go through diagnosis & treatment options. This module can be paused at any time.

Lower / Upper GI
Small intestine

In this section we will cover:

- Causes and Risk Factors
- Signs and Symptoms
- Anatomy & Physiology
- Regional Lymph Nodes
- Diagnosis
- Morphology
- ICD10 coding
- Grade
- Stage
- Treatment

We'll start off by looking at the causes and risk factors …

Small intestine – Causes & Risk Factors

Risk factors for a tumour of the small intestine include:

- Increasing age
- Familial adenomatous polyposis – a genetic condition leading to the development of polyps in the small intestine
- Lynch syndrome – a genetic condition that increases the risk of several different types of cancer at a younger age
- Crohn's disease
- Coeliac disease
- A diet rich in red meat, smoked foods or fat

The risk of a tumour in the small intestine increases as we age. There are also genetic conditions such as Lynch syndrome and certain pre-existing medical conditions including Crohn's disease that have been shown to be risk factors.

Small intestine – Signs & Symptoms

- Pain, swelling or a lump in the abdomen
- Weight loss
- Nausea or vomiting
- Diarrhoea
- Tiredness
- Dark, blackened faeces – due to bleeding in the small intestine
- An intestinal blockage
- Anaemia

Patients may present with a swollen abdomen, anaemia or blackened faeces due to intestinal bleeding.

Small intestine – Signs & Symptoms – Neuroendocrine tumours

Additional symptoms may be reported in the case of a possible neuroendocrine tumour:

- High temperature
- Flushed skin
- Wheezing
- Fast heartbeat

Additionally, neuroendocrine tumours may cause the patient to experience a raised temperature and flushed appearance.

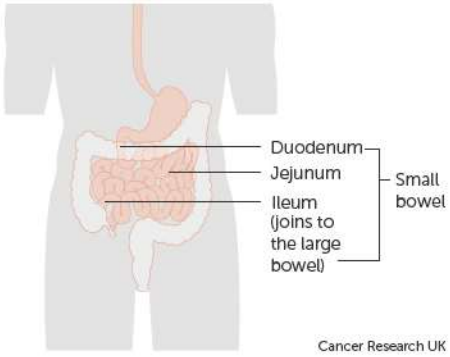
Small intestine – Anatomy & Physiology

The small intestine connects the stomach and the large intestine. The small intestine is approximately 6 metres in length and is divided into three sections:

- First is the duodenum which is connected to the stomach. The primary function of the duodenum is to secrete enzymes that digest the food
- Next is the jejunum which also secretes certain enzymes and absorbs many of the nutrients from the food
- The last and longest part of the small intestine is the ileum which carries out the bulk of nutrient absorption and connects to the large intestine

Most small intestine tumours are found in the duodenum

Please note that the Ampulla of Vater (where digestive juices enter the duodenum from the pancreas and bile ducts) is covered in the hepatobiliary module

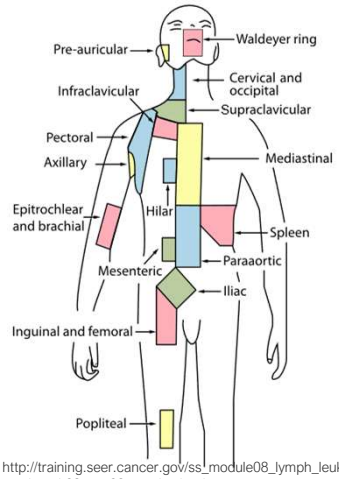


The small intestine lies between the stomach and the colon. Classified in three parts: duodenum, jejunum and ileum, its primary function is the digestion of food and the absorption of nutrients.

Small intestine – Regional Lymph Nodes

Duodenum:

- Pancreaticoduodenal – near the pancreas and the duodenum
- Pyloric – near the artery that supplies blood to part of the stomach and the duodenum
- Hepatic (including pericholedochal, cystic & hilar)
- Superior mesenteric



http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html

During an MDT, clinical teams will often make reference to particular groups of regional lymph nodes. This may indicate that the stage of the cancer has been determined. The lymph nodes considered to be regional for the duodenum are listed here…

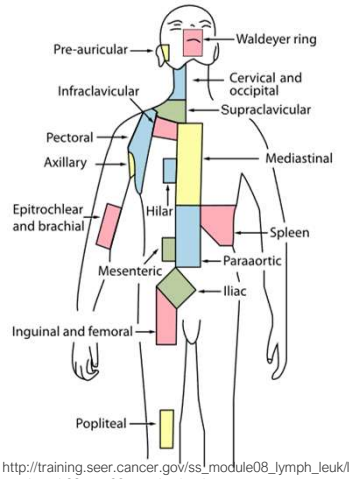
Small intestine – Regional Lymph Nodes

Jejunum & Ileum:

- Mesenteric (including the Superior mesenteric)

Terminal ileum:

- Mesenteric (including the Superior mesenteric)
- Ileocolic

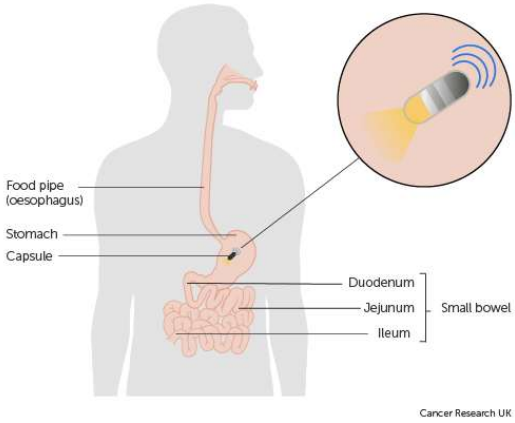


http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html

…while the lymph nodes considered to be regional for the jejunum, ileum and terminal ileum are listed here.

Small intestine – Diagnosis

- Blood tests – Liver Function Tests (LFTs) & tests for anaemia
- Urine tests - to check for unusual levels of certain hormones
- Barium x-ray
- Ultrasound scan
- Endoscopy
- Capsule endoscopy – illustrated
- Biopsy
- CT scan
- MRI scan
- PET scan
- Octreoscan – uses a radioactive tracer to detect neuroendocrine tumours



The diagnostic process generally involves blood tests, endoscopy and some form of imaging. Biopsies may also be taken

Small intestine – Morphology - Invasive

Invasive primary tumours of the small intestine may include:

- Adenocarcinoma – M8140/3
- Neuroendocrine (carcinoid) tumour, Grade 1 – M8240/3
- Neuroendocrine (carcinoid) tumour, Grade 2 – M8249/3

(please refer to the Neuroendocrine neoplasms – Key Points training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials)

- Gastrointestinal stromal tumour, malignant – M8936/3

(please note: **all** GISTs are to be reported as malignant M8936/3 in accordance with the WHO Classification of Tumour series, 5th edition and as such would be C coded in ICD10)

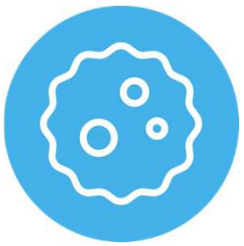
- Leiomyosarcoma – M8890/3
- Sarcoma – M8800/3
- Lymphoma, NOS – M9590/3
- Lymphoma, non-Hodgkin – M9591/3

The morphology codes for invasive primary tumours of the small intestine are shown here. Please be aware that ALL GISTs must be reported as malignant.

Small intestine – Morphology – Non-invasive

In situ tumours of the small intestine will have an ICD-O-3 morphology code ending with "/2"

Neoplasms of an uncertain or unknown behaviour will have an ICD-O-3 morphology code ending with "/1"



If the pathology report includes an ICD-O-3 morphology code ending in either /1 or /2, this indicates a non-invasive neoplasm.

Small intestine – ICD10 coding – Invasive

- Duodenum – C17.0
- Jejunum – C17.1
- Ileum – C17.2 (excludes ileocaecal valve – C18.0)
- Meckel diverticulum – C17.3
- Overlapping lesion of small intestine – C17.8
- Small intestine, unspecified – C17.9

The ICD10 codes for invasive tumours of the Small intestine are shown here.

Small intestine – ICD10 coding – Non-invasive

- Carcinoma in situ of other and unspecified parts of intestine – D01.4 (excludes ampulla of Vater – D01.5)
- Carcinoma in situ of digestive organ, unspecified – D01.9
- Neoplasm of uncertain or unknown behaviour of small intestine – D37.2

- While your clinical team may request that these D coded tumours are recorded, these do not currently require a COSD submission from your cancer data management system. NDRS obtains data on these tumours direct from pathology laboratories

Non-invasive tumours would be coded differently in ICD10… and while your clinical team may request that non-invasive tumours are recorded, these do not currently require a COSD submission from your cancer data management system – NDRS obtains this data direct from the pathology labs

Small intestine – Grade – Carcinoma / Neuroendocrine tumours

- Grade 1 – Well differentiated: Tumours look very similar to the normal tissue and retain a degree of functionality. Grade 1 tumours have the best prognosis
- Grade 2 – Moderately differentiated: Tumours are formed of cells that somewhat resemble the normal tissue and retain limited functionality
- Grade 3 – Poorly differentiated: Tumours have very abnormal cells with little or no functionality

The grade of both carcinomas and neuroendocrine tumours is assessed under a microscope by comparing the appearance of tumour cells to normal healthy cells. The higher the grade, the less similar they appear and the lower the functionality.

Small Intestine – Grade – Soft tissue Sarcoma

The grading system used for soft tissue sarcomas in the UK is the French Federation of Cancer Centres Sarcoma Group (Fédération Nationale des Centres de Lutte Contre Le Cance - FNCLCC) which is derived following assessment of:

- Mitotic rate – the rate at which cancer cells reproduce, scored from 1 (low reproduction rate) to 3 (high reproduction rate)
- Presence of necrosis – the presence and proportion of dying tissue in the sarcoma, scored from 0 (no necrotic tissue) to 2 (50% or more necrotic tissue)
- Differentiation – how closely the sarcoma cells resemble normal tissue in appearance and functionality, scored from 1 (similar to normal cells) to 3 (very abnormal cells)

- Scores are added together to give the grade:
  - Grade 1 means a total score of 2 or 3 (sometimes called LOW grade)
  - Grade 2 means a total score of 4 or 5 (grade 2 or 3 are sometimes called HIGH grade)
  - Grade 3 means a total score of 6 or more

Grading of Soft Tissue Sarcomas relies not only on the resemblance to normal tissue but also on the rate of reproduction and the presence of any dead tissue within the tumour. Each of these factors is given a score which is then added to the other factors to assign the Grade.

Small intestine – Grade – Lymphoma

Grading of lymphomas can vary depending on the type of lymphoma:

Non-Hodgkin Lymphomas (NHL) are the more common type of intestinal lymphoma, and they include MALT lymphoma, DLBCL and follicular lymphoma (among others)

- Indolent (low grade, slow growing)
- Aggressive (high grade, fast growing

For other types of lymphoma, please refer to your clinical team

The grading system used for lymphomas of the small intestine will depend on the type of lymphoma. Non-Hodgkin Lymphoma grading is shown here. For other types of lymphoma, please refer to your clinical team.

Small intestine – Stage - TNM

- Neuroendocrine (carcinoid) tumours are staged using **TNM, ENETS version**.
- Other invasive tumours are staged as follows:
  - **For diagnosis dates up to 31st December 2025 use UICC TNM v8**
  - **For diagnosis dates from 1st January 2026 use UICC TNM v9**
- Please note that the TNM version must be accurately recorded – if you are unable to amend the version on your cancer data management system, please refer to your line manager
- If, after 1st January 2026, your cancer data management system has not been amended to include TNM v9 please record the TNM v9 stage and add the following statement to the Primary Diagnosis Subsidiary Comment field:
  - **Patient staged using TNM9 not TNM8 as per CR2070**

Neuroendocrine tumours are staged using TNM, ENETS version. All other invasive neoplasms are staged using the appropriate UICC TNM version as shown.

Small intestine – Stage - TNM

- For details on recording stage, please see the NDRS training module – KPI-TNM Staging 101, available on this link: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials

- Staging data sheets detailing the specific staging requirements can also be downloaded from the NDRS website for clinical use: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

For more details on recording stage, please see the NDRS training module KPI-TNM Staging 101 and the relevant staging data sheets

Small intestine - Treatment

The treatment of intestinal cancers depends on the location/type of the tumour, the stage of disease at presentation and how fit the patient is to tolerate radical treatment

- Early stage - likely to receive surgery as their first treatment with radiotherapy and possibly chemotherapy as adjuvant treatment
- Advanced stage - neo-adjuvant treatment, usually with a combination of chemotherapy and radiotherapy may be offered prior to surgery

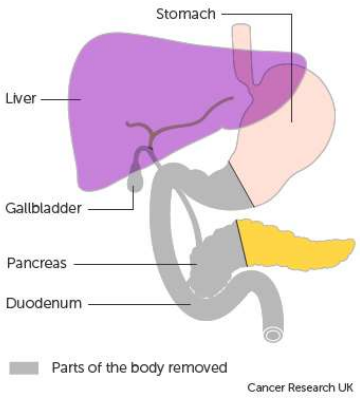
Treatment options and sequences vary according to… the location and type of the tumour, how far the tumour has spread and whether or not the patient is fit for surgery. Early stage cancers are more likely to be offered surgery as a first treatment, while more advanced cases may need a different approach

Small intestine – Treatment – Surgery

Surgery may be offered where the tumour is deemed to be resectable. Surgery is often the main treatment option for all morphologies of tumour in the small intestine, depending on stage

If the tumour is at the top of the duodenum, patients may be offered a pancreatoduodenectomy (illustrated) which, as well as removing the duodenum, may involve removal of part of the stomach, part of the pancreas and the gallbladder

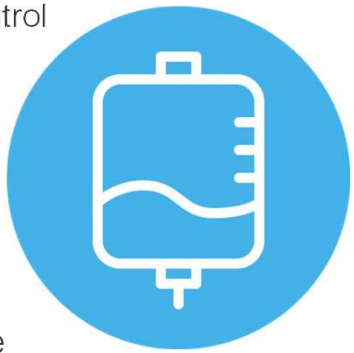
For tumours on the distal ileum, the surgeon may also remove part of the colon



Patients with a resectable tumour may be offered a surgical excision. Some types of surgical excision may require the removal of all or parts of associated organs.

Small intestine – Treatment - Chemotherapy

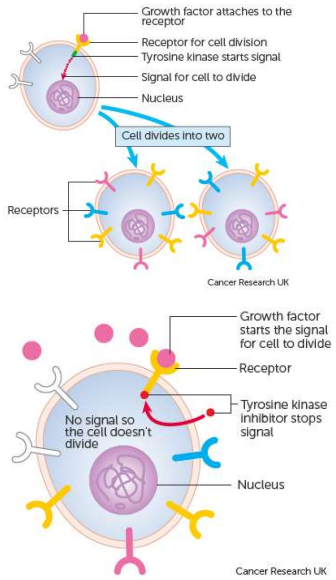
Chemotherapy can be a primary treatment for advanced tumours or for a recurrence. It is unlikely to be curative but chemotherapy may slow the growth of the tumour and control symptoms



- Neo-adjuvant Chemotherapy
- This may be used to shrink tumours to allow surgical resection with sufficient margins
- Adjuvant Chemotherapy
- At the surgery of the primary cancer, if it is found that the cancer has spread to the lymph nodes, or the tumour is of poor prognosis then chemotherapy may be offered

Chemotherapy can be used as a primary treatment and although unlikely to be curative it may slow the growth of a tumour. It can also be used to shrink the tumour prior to a surgical resection or to destroy any remaining cancer cells after surgery.

Small intestine – Treatment – Targeted treatments



Where surgery is not possible and the patient is fit enough, targeted treatments (sometimes called biological treatments) may be offered for some types of malignant GIST or for neuroendocrine tumours. The targeted treatments used for these tumours are a type of small-molecule inhibitor. The treatments act to prevent tumour growth

- When certain growth factors within the body attach to a receptor on the surface of a cancer cell, the base of the receptor releases a chemical that instructs the cell to divide into two new cells
- The targeted treatments aim to block these chemical signals from the base of the receptor
- Many of the targeted treatments also act to prevent the tumour developing its own blood supply

Small-molecule inhibitors will have a drug name ending in -nib

Targeted treatments may be an alternative for some types of malignant GIST or for neuroendocrine tumours. The targeted treatments used will block chemical signals within the cancer cells that would otherwise instruct them to divide. This prevents tumour growth

Small intestine – Treatment - Radiotherapy

Radiotherapy is not generally offered as a curative treatment for tumours of the small intestine

- Palliative Radiotherapy
- For more advanced tumours and where appropriate, radiotherapy may be used to target distant metastases to alleviate symptoms

Radiotherapy is not generally offered as a curative treatment for small intestine tumours but may be used palliatively to ease the symptoms of metastatic deposits.

Summary

In summary …

Summary

- Risk factors for a tumour of the small intestine include increasing age, genetic conditions, Coeliac or Crohn's disease or a diet rich in fatty foods

The risk of a tumour in the small intestine may increase with age, certain pre-existing conditions and a high-fat diet.

Summary

- Risk factors for a tumour of the small intestine include increasing age, genetic conditions, Coeliac or Crohn's disease or a diet rich in fatty foods
- Signs of a possible small intestine tumour include weight loss, anaemia and abdominal pain. Additionally, neuroendocrine tumour patients may present with a fast heartbeat and flushed appearance

Symptoms of a possible tumour may include weight loss or abdominal pain. Neuroendocrine tumours may also cause a faster heartbeat and flushed appearance.

Summary

- Risk factors for a tumour of the small intestine include increasing age, genetic conditions, Coeliac or Crohn's disease or a diet rich in fatty foods
- Signs of a possible small intestine tumour include weight loss, anaemia and abdominal pain. Additionally, neuroendocrine tumour patients may present with a fast heartbeat and flushed appearance
- The diagnostic process may include blood & urine tests, imaging and biopsies

Diagnostic tests can include blood tests and biopsies as well as imaging

Summary

- Risk factors for a tumour of the small intestine include increasing age, genetic conditions, Coeliac or Crohn's disease or a diet rich in fatty foods
- Signs of a possible small intestine tumour include weight loss, anaemia and abdominal pain. Additionally, neuroendocrine tumour patients may present with a fast heartbeat and flushed appearance
- The diagnostic process may include blood & urine tests, imaging and biopsies
- All invasive tumours must be recorded. Stage must be recorded for all stageable cancers. Stage edition for neuroendocrine (carcinoid) tumours must be recorded as ENETS

All invasive cancers must be added to your cancer data management system and if stageable, also have a stage recorded. Stage version for neuroendocrine tumours should be recorded as ENETS, while all other tumour types are staged using the relevant UICC TNM version

Summary

- Risk factors for a tumour of the small intestine include increasing age, genetic conditions, Coeliac or Crohn's disease or a diet rich in fatty foods
- Signs of a possible small intestine tumour include weight loss, anaemia and abdominal pain. Additionally, neuroendocrine tumour patients may present with a fast heartbeat and flushed appearance
- The diagnostic process may include blood & urine tests, imaging and biopsies
- All invasive tumours must be recorded. Stage must be recorded for all stageable cancers. Stage edition for neuroendocrine (carcinoid) tumours must be recorded as ENETS
- If a tumour is diagnosed it may or may not be invasive. While all invasive tumours must be recorded, non-invasive tumours do not need to be recorded on a cancer data management system for the purposes of COSD - NDRS obtains these records directly from pathology laboratories

And non-invasive tumours do not need to be recorded for the purposes of COSD

Summary

- Additional guidance on recording COSD data including morphology, topography, staging and recording a diagnosis can be found at: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Staging data sheets can also be downloaded from the NDRS website for clinical use: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

Additional training modules as well as Staging sheets for clinical use may be downloaded from the NDRS website.

Summary

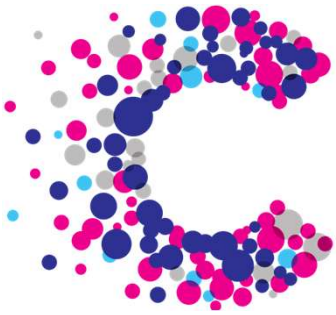
- If in any doubt as to whether you should be recording a diagnosis, please refer to the latest COSD User Guide, Appendices A & B
- For guidance on the required staging system, please refer to the latest COSD User Guide, Appendix E

- https://digital.nhs.uk/ndrs/data/data-sets/cosd#downloads

Do please remember, guidance **is** available on our website. You can download the COSD User Guide by clicking on this link and selecting the COSD version appropriate to your trust.

Acknowledgements

Many thanks to Cancer Research UK for the use of their images within this training module.



CANCER RESEARCH UK

We'd like to thank Cancer Research UK for the use of their images within this training module.

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If you have any questions on the information contained within this module or about COSD in general, do please feel free to email your regional Data Liaison Manager