Esophagus, Stomach and Duodenum

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Contents

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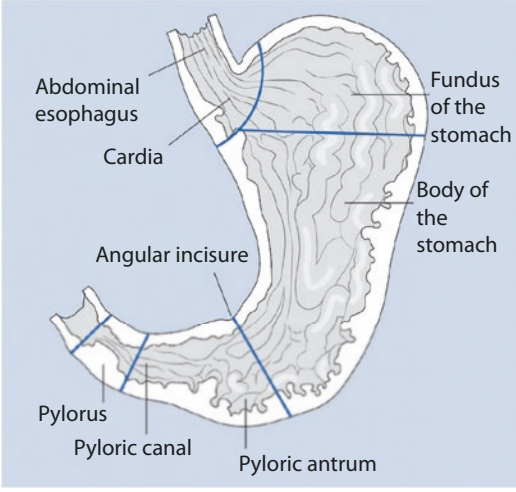
1.1 Anatomy and Physiology

1.1.1 Esophagus

- Cervical, thoracic and abdominal esophagus (total length approx. 25–28 cm)
- Physiological esophageal constrictions at the upper esophageal sphincter, tracheal bifurcation and lower esophageal sphincter
- Transition from the hypopharynx with striated, voluntarily innervated musculature to the esophagus with smooth musculature
- Histologically lined with squamous epithelium
- Transport function for food pulp and saliva

1.1.2 Stomach

- The cardia, fundus, corpus and antrum form the stomach (◘ Fig. 1.1)
- Sphincter muscle at the stomach outlet (pylorus)
- Histologically lined with cylindrical epithelium
- Blood supply via arcade from right and left gastric artery (small curvature), arcade from left and right gastroomental artery (large curvature) as well as short gastric vessels
- Reservoir function for food pulp
- Production of pepsin and hydrochloric acid for digestion
- Production of intrinsic factor (vitamin B_{12} absorption in the terminal ileum)



◘ **Fig. 1.1** Classification of the stomach according to external shape. (Mod. according to von Lanz and Wachsmuth 2004)

1.1.3 Duodenum

- Superior part, descending part, inferior part, ascending part of the duodenum (pars I–IV duodeni)
- Common orifice of bile duct and pancreatic duct at the major duodenal papilla (papilla Vateri) in the descending part, sometimes accessory pancreatic duct with separate orifice into the duodenum (minor duodenal papilla)
- Histologically lined with cylindrical epithelium
- Blood supply via gastroduodenal artery as well as branches of the superior mesenteric artery (sup. and inf. pancreatoduodenal artery, Rio-Branko arcade to gastroduodenal artery)
- Mixture of food pulp with bile and pancreatic secretions

1.2 Leading Symptoms and Diagnosis

1.2.1 Leading Symptoms

- Different for esophagus and stomach/duodenum

Esophagus

- Swallowing disorders/dysphagia
- Regurgitation of food
- Retrosternal pain
- Weight loss

Stomach

- Upper abdominal pain
- Regurgitation
- Weight loss

Upper GI Bleeding

- Vomiting blood (hematemesis)/tarry stools (hematochezia)/anemia

1.2.2 Diagnosis

Endoscopy

- Esophagogastroduodenoscopy (EGD)
- Endosonography for staging examination of neoplasms
- Manometry for the diagnosis of the movement disorders of the esophageal musculature
- 24 h-pH-metry, impedance measurement for the diagnosis of gastroesophageal reflux disease

Radiology/Nuclear Medicine

- Contrast esophagogram
- Computed tomography chest/abdomen
- MRI and PET-CT for special indications
- Gastric emptying scintigraphy

1.2.3 Therapeutic Principles

- Conservative
- Endoscopic
- Surgical

1.3 Benign Diseases of the Esophagus

Summary

- Diverticular disease:
  - Zenker's diverticulum
  - Midesophageal diverticulum
  - Epiphrenic diverticulum
- Achalasia:
  - Endoscopic and surgical therapy
- Gastroesophageal reflux disease (GERD)/hiatal hernias
  - Axial hiatal hernia
  - Paraesophageal hiatal hernia
  - Gastroesophageal reflux disease—Diagnosis
  - Gastroesophageal reflux disease—pharmacological and surgical therapy
- Esophageal perforations

1.3.1 Diverticular Diseases of the Esophagus

Etiology

Zenker's Diverticulum and Epiphrenic Diverticulum (Pulsion Diverticulum)

- Due to a mismatch between increased intraluminal pressure and anatomical muscle gap; preferentially at predilection sites above the esophageal sphincter
- Pseudodiverticulum (diverticular sac consists only of mucosa and submucosa)

Midesophageal Diverticulum (Traction Diverticulum)

- Diverticulum includes all wall layers of the esophagus
- Due to traction on the esophagus from outside, e.g. due to residual embryonic tissue connections between trachea and esophagus or also in the context of inflammatory processes in the mediastinum
- Located mostly in the middle esophagus
- Overall very rare

Forms

Zenker's Diverticulum (Hypopharynx)

- Most frequent esophageal diverticulum (incidence 2:100,000/year)
- Age of manifestation: 70–80 years
- Predominantly men affected
- Location: Killian's triangle (between cricopharyngeal muscle and inferior constrictor of the pharynx)
- Increased tone of the cricopharyngeal muscle and impaired relaxation of the upper esophageal sphincter

Killian-Jamison Diverticulum

- Pulsion diverticulum
- Origin immediately below the upper sphincter

- Localization ventrolaterally under the cricopharyngeal muscle or
- Localization dorsal through the Laimer triangle
- Mostly small and asymptomatic diverticula

Epiphrenic Diverticulum

- Pulsion diverticulum
- Localized up to 10 cm above the Z-line
- Much rarer than Zenker's diverticulum
- Development due to increased intraluminal pressure
- Usually associated with achalasia or diffuse esophageal spasm

Symptoms

- Dysphagia
- Regurgitation of undigested food
- Foetor ex ore
- Chronic cough and aspiration of food debris
- Recurrent pneumonias
- Lump feeling in the throat
- Cervical borborygmi (pathognomonic in Zenker diverticulum)
- Retrosternal pain and heartburn (epiphrenic diverticula)

Therapy

Zenker-Diverikel

- Cervical diverticular resection and myotomy of the cricopharyngeal muscle
- Cervical diverticulopexy and myotomy of the cricopharyngeal muscle
- Endoscopic interventional transoral splitting (stapler, laser, needle knife)

Epiphrenic Diverticulum

Caution

Treatment of the underlying esophageal motility disorder required.

- Laparoscopic diverticulotomy and myotomy of the lower esophageal sphincter (+/– fundoplication)
- Transthoracic diverticulotomy and myotomy of the lower esophageal sphincter

1.3.2 Achalasia

- Incidence = 1/100,000 population/year

Etiology

Pathogenesis

- Degeneration of the myenteric plexus
- Lack of relaxation of the lower esophageal sphincter

Forms

Primary Achalasia

- Etiology unknown

Secondary Achalasia

- Chagas disease
- Gastric Cancer
- Esophageal Cancer

Symptoms

- Dysphagia
- Regurgitation of food
- Retrosternal pain
- Aspiration

Complications

- Aspiration pneumonia
- Esophageal cancer (increased incidence in achalasia)

Staging

- Stage I: Hypermotile form
- Stage II: Hypomotile form
- Stage III: Amotile form

Diagnosis

Esophagogastroduodenoscopy (EGD)

- Exclusion of malignancy and benign stenosis

Manometry

- Pressure measurement in the esophagus via probe
- Slow retraction of the probe during the swallowing act and digital recording and evaluation of peristalsis

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- Highest sensitivity for the diagnosis of achalasia
- Findings: Combination of hypermotility/hypomotility/peristalsis and lack of relaxation of the lower esophageal sphincter

X-Ray (Esophagogram)/Computed Tomography

- Classic "champagne glass" shape of the esophagus with prestenotic dilatation

Therapy

Medical Therapy

Principle

- Drug-induced relaxation of the smooth muscle fibers of the lower esophageal sphincter

Preparations

- Calcium channel blockers (e.g. nifedipine)
- Long-acting nitrates
- Phosphodiesterase-5 inhibitor (sildenafil)

Drug Therapy

- Best results with nifedipine (10–30 mg) sublingually approx. 30 min before a meal (leads to a relaxation of the lower oesophageal sphincter lasting approx. 60 min).
- Overall, the effect of drug therapy is usually not satisfactory: persistence of symptoms and side effects of the drugs (headache, hypotension, etc.).

Endoscopic Therapy

Pneumatic Dilatation

- Dilatation with special balloon under fluoroscopy or under endoscopic guidance
- Disruption of the muscle fibers of the lower esophageal sphincter
- Multiple applications are often necessary, but long-term alleviation of symptoms is possible in up to 50–80% of patients
- Risk of esophageal perforation due to dilatation

Endoscopic Injection of Botulinum Toxin

- Low risk procedure
- High initial response rate (>75%)
- Frequent early recurrences (50%)

Peroral Endoscopic Myotomy (POEM)

- Endoscopic alternative to surgical myotomy
- Procedure:
  - Endoscopic incision of the mucosa, then submucosal tunnelling and long-stretch myotomy by diathermy
- In the short-term follow-up results comparable with the surgical procedure
- Centre-based expertise

Surgical Therapy

Surgical Therapy Options

- Myotomy of the lower esophageal sphincter possible via laparoscopy, laparotomy or thoracotomy (Vaezi et al. 2013) or robotically-assisted
- Gold standard: Laparoscopic Heller myotomy + fundoplication
- For stage III: discuss esophagectomy with gastric tube reconstruction

Results After Myotomy

- Partial or complete relief of symptoms in 90% of patients
- 30% of the patients develop reflux symptoms postoperatively, therefore an additional fundoplication is mandatory

Surgical Procedure

Laparoscopic Heller Myotomy

- Supine position with spread legs
- Insertion of the camera trocar, insertion of the pneumoperitoneum; operation may be robotically assisted
- Exposure of the esophageal hiatus by means of a liver retractor and positioning of the patient
- Ventral mobilization of the esophagus into the lower mediastinum
- Identification and protection of the vagus nerve
- Longitudinal severing of the muscle fibres of the lower oesophageal sphinc-

ter including cardia while sparing the mucosal tube
- Covering the defect with an anterior fundoplication (► Sect. 1.3.5), or Nissen fundoplication

1.3.3 Esophageal Perforation

Summary

- Different etiologies (malignant, spontaneous, iatrogenic)
- Life-threatening disease
- Therapy today mostly endoscopic (stent or endoluminal vacuum therapy EndoVAC)
- Surgical therapy (primary suture + fundoplication or muscle flap) as effective therapy option

Etiology

- Iatrogenic:
  - As a result of endoscopic procedures (EGD, ERCP, endosonography)
  - Complication of cardiothoracic surgery
  - Gastric tube
- Malignant:
  - Tumor perforation due to esophageal cancer
- Spontaneous:
  - Boerhaave Syndrome
  - Often as a result of sudden vomiting
  - Lower/middle third esophageal rupture

Symptoms

- Acute chest pain:
  - Differential Diagnosis:
    - Myocardial Infarction
    - Pulmonary Embolism
    - Aortic dissection
  - Frequently misdiagnosed
- Hematemesis
- Dyspnea
- Fever
- Complications:
  - Mediastinitis
  - Pleural effusion/empyema
  - Pneumothorax
  - Septic shock

Diagnosis

Endoscopy (EGD)

- Localization of the perforation
- Assessment of the size of the perforation

Computed Tomography of the chest

- Oral and intravenous contrast agent
- Confirmation of transmural perforation
- Detection of pleural empyema/mediastinal abscess

Therapy

- Closure of the perforation
- Endoluminal vacuum therapy
- Drainage of the contaminated cavities (mediastinum, pleura)

Surgical Therapy

- Primary suture of the esophageal perforation
- Additional coverage by means of fundoplication or muscle flap
- Insertion of mediastinal drains
- Insertion of chest drain(s)
- Video-assisted thoracoscopy (VATS) for pleural empyema
- Ultima Ratio: Esophageal resection and reconstruction via gastric tube or diversion

Endoscopic Therapy

- Actually Gold standard
- Less invasive and comparably effective
- Endoscopic insertion of a partially coated metal stent
- Endoscopic vacuum therapy-EndoVAC
- Over-the-scope clip (OTSC) only for small and recent perforations

Conservative Therapy

- Indicated only in palliative situation or very old perforation without sepsis
- Transcutaneous or endoluminal drainage of the contaminated cavity

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Prognostic Factors

- Delayed therapy (>48 h) unfavourable
- Septic shock at the time of therapy
- Spontaneous esophageal perforation (Boerhaave syndrome; Connelly et al. 2013)
- Size and localization of the perforation not prognostically relevant

1.3.4 Hiatal Hernias

Etiology

- Acquired pathology in the majority of patients
- Risk factors:
  - Overweight
  - Pregnancy
  - Connective tissue aging

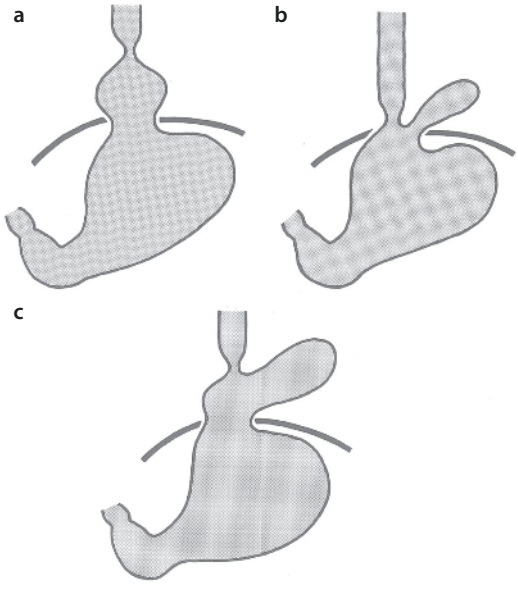
Types of Hiatal Hernias (◘ Fig. 1.2)

Cardiofundal Malposition

- Mildest type
- Often incidental finding

Axial Sliding Hernia

- 90% of all hernias
- Intrathoracic position of the gastric cardia



◘ **Fig. 1.2** Types of hiatal hernia. (**a**) Axial sliding hernia. (**b**) Paraesophageal hernia. (**c**) Mixed Hernia. (From von Lanz and Wachsmuth 2004)

Paraesophageal Hernia

- Lower esophageal sphincter intraabdominal
- Partial or complete intrathoracic stomach (upside-down stomach)

Mixed Forms

- Rarely, additional herniation of omentum, small or large intestine

Symptoms

Axial Hernias

- Often asymptomatic (90%)
- But predisposition for gastroesophageal reflux with insufficient function of the lower esophageal sphincter

Paraesophageal Hernias

- Initially often asymptomatic as well
- Possibly postprandial unspecific abdominal or thoracic complaints
- Complications:
  - Dysphagia
  - Incarceration
  - Ulceration
  - Chronic anemia
  - Dyspnea

Therapy

Symptomatic Therapy

- Proton-pump inhibitors
- See below: Therapy of reflux disease (► Sect. 1.3.5)

Surgical Therapy

- Strategy: Reduction of the hernia, anterior and/or posterior hiatoplasty, fundoplication or gastropexy
- Indication:
  - In axial hernia only in case of symptomatic reflux (GERD, 24 h pH-metry, volume reflux, metaplasia)
  - Indication for elective surgical intervention for paraesophageal hernia
  - Emergency indication for incarcerated hernias (usually paraesophageal)

Reinforcement of hiatoplasty by reinforcement with non-resorbable or bioresorbable mesh possible, benefit not proven. Risk of injury of esophagus or severe complications.

1.3.5 Gastroesophageal Reflux Disease (GERD)

Summary

- GERD subsumes different disease entities
- Prevalence: 15% of the western population
- Multifactorial pathophysiology
- Therapy initially conservative with proton pump inhibitors (PPI)
- Laparoscopic fundoplication as an effective therapeutic option in case of failure of drug therapy or patient preference

Definition

- "GERD" = "gastroesophageal reflux disease"
- The term subsumes the following entities:
  - Erosive reflux esophagitis of various degrees of severity (ERD)
  - Non-erosive reflux disease (NERD)
  - Hypersensitive esophagus
  - Extraesophageal manifestations
  - Complications of GERD
  - Functional reflux complaints

Etiology

Demographics

- Prevalence: about 15% with increasing incidence
- Approximately 50% of patients with GERD do not have endoscopically definable lesions (NERD)
- Approx. 5% of GERD patients develop Barrett's esophagus = intestinal metaplasia of the epithelium in the (distal) esophagus

Pathophysiology and Risk Factors

- Pathophysiology:
  - Primary GERD: Unclear dysfunction of the lower esophageal sphincter
  - Secondary GERD: in the context of other diseases or as a consequence of surgical treatment (esophageal cancer, post-heller myotomy)
- Predisposing factors (due to dysfunction of the lower esophageal sphincter):
  - Pregnancy
  - Obesity
  - Hiatal Hernia
  - Nutritional factors
- Predisposing factors (due to irritating reflux):
  - Overproduction of gastric acid, e.g. due to Helicobacter pylori
  - Alkaline reflux (e.g. bile reflux after gastrectomy)
  - Alcohol, coffee, nicotine, various drugs affect both the lower esophageal sphincter and gastric content
- Frequently increased symptoms postprandially and due to bending or pressing

Symptoms

- Chief complaint = heartburn
- Other symptoms:
  - Diffuse epigastric pain
  - Retrosternal pain
  - Belching
  - Volume reflux with regurgitation of food residues
  - Dysphagia
  - Irritative cough, hoarseness

Diagnosis

Esophagogastroduodenoscopy (EGD)

- Detection of erosive lesions
- Classification according to the Los-Angeles classification (◘ Table 1.1)
- Often no correlation between intensity of complaints and endoscopic findings

◘ **Table 1.1** Los-Angeles classification of gastroesophageal reflux disease

Stage	Findings
A	Erosions <5 mm
B	Erosions >5 mm
C	Confluent erosions <75% of circumference
D	Confluent erosions >75% of circumference

- Exclusion of complication:
  - Stenosis
  - Barrett's Esophagus
  - Dysplasia
  - Ulceration
  - Esophageal Cancer

24 h pH-Metry/Impedance Measurement

- Quantification of gastroesophageal reflux via probe
- Prior discontinuation of PPI
- Highest sensitivity and specificity for the detection of GERD

With pH-metry only acid reflux is detected, with impedance measurement: detection of any type of reflux.

Manometry

- Relevant for the exclusion of motility disorders, especially before surgical therapy (fundoplication, magnetic sphincter augmentation)
- Hypomotility of the distal esophagus often associated with long-lasting GERD
- Details ► Sect. 1.3.2

Therapy

Conservative Therapy

- Changes in "life style": weight loss, avoidance of noxious substances, sleeping with the upper body elevated, discontinuation of triggering medications
- Medical: Proton pump inhibitors (PPI)
- Objective: Symptom control and healing of existing erosions
- Therapy failure of PPI: always detailed diagnosis with endoscopy and pH-metry/impedance measurement
- Asymptomatic erosive reflux esophagitis: therapy indicated

Surgical Therapy

- Indication:
  - Long-term need for therapy
  - Inadequate symptom control with PPI
  - Volume reflux
  - Side effects of drug therapy
  - Patient preference
- Operation of choice = laparoscopic fundoplication:
  - 360°—Nissen, 270° posterior—Toupet
  - Always with hiatoplasty (if necessary reduction of a hiatal hernia)
- Results:
  - Success rate approx. 85% with thorough patient selection
  - Lower success rate for re-operation (re-fundoplication)

Even after surgical treatment, endoscopic controls are recommended for pre-existing Barrett's esophagus.

Surgical Procedure

Laparoscopic Fundoplication 360° According to Nissen

- Supine split-legs position (french position)
- Insertion of the camera trocar, insertion of the pneumoperitoneum; robot-assisted operation possible
- Exposure of the esophageal hiatus by means of a liver retractor and positioning of the patient
- Visualisation of the left and right diaphragmatic crus
- Mobilization of the esophagus into the lower mediastinum, with creation retroesophageal window
- Identification of the vagal nerve
- Partial mobilization of the gastric fundus (division short gastric vessels)
- Calibration of the esophagus with large lumen gastric tube (bougie)
- Posterior hiatoplasty with non-absorbable suture material
- Retroesophageal pulling of the fundus
- Creation of the wrap: suture of the retroesophageal fundus anterior to additional part of fundus with 2–3 non-absorbable sutures
- Fixation of the wrap to the esophagus with distal suture

1.3.6 Guidelines

AWMF guideline registry: gastroesophageal reflux disease (German Society of Gastroenterology, Digestive and Metabolic Diseases, AWMF), 2014, AWMF registration number: 021/013—valid until May 31, 2019 currently under revision (7/2020).

1.4 Malignant Diseases of the Esophagus

1.4.1 Overview

Esophageal Cancer

- Squamous cell carcinoma
- Adenocarcinoma
- Adenosquamous carcinomas, undifferentiated carcinomas

Adenocarcinoma of the Gastroesophageal Junction (AEG)

- AEG 1, AEG 2 and AEG 3 with esophageal infiltration are classified as esophageal carcinoma according to UICC TNM 8th version (2017). Differentiation AEG 1–3: according to the epicenter of the tumor, not the upper margin.

1.4.2 Esophageal Carcinoma (Including AEG)

Summary

- Squamous cell carcinoma vs. adenocarcinoma: different etiology and tumor biology
- Leading symptoms: Dysphagia, weight loss, hematemesis
- Pretherapeutic staging: EGD/endosonography/CT (cTNM + histology)
- Neoadjuvant therapy for T3 or any T with N+: Neoadjuvant radiochemotherapy (adenocarcinoma and squamous cell carcinoma) or perioperative chemotherapy for adenocarcinoma
- Operative standard: Thoracoabdominal esophagectomy with 2-field lymphadenectomy and advancement of stomach into right chest and esophagogastric anastomosis
- High rate of postoperative complications after esophagectomy
- Minimally invasive laparoscopic and robotic-assisted procedures with potential benefits in terms of postoperative morbidity
- Minimally invasive abdominal gastric mobilisation/advancement (hybrid OP) with evidence-based (Ib) advantages
- In case of functional or oncological inoperability definitive radiochemotherapy or systemic chemotherapy, if necessary palliative insertion of esophageal stents

Definition

- All epithelial malignancies between upper and lower esophageal sphincter
- Adenocarcinomas of the gastroesophageal junction with infiltration of the esophagus are defined as esophageal carcinoma (UICC TNM 8)

Types

Adenocarcinoma

- 95% in the distal esophagus

Adenocarcinoma of the Gastroesophageal Junction (AEG)

- Definition: All adenocarcinomas with tumor center 5 cm proximal to 5 cm distal to the gastroesophageal junction. Definition by tumour epicentre, not upper margin (UICC 8th version)
- Classification according to Siewert:
  - AEG 1: Tumour centre 1–5 cm proximal to the gastro-esophageal junction
  - AEG 2: Tumour centre from 1 cm proximal to 2 cm distal to the gastro-esophageal junction
  - AEG 3: Tumour centre 2–5 cm distal to the gastro-esophageal junction

Squamous Cell Carcinoma
- May occur throughout the esophagus

Adenosquamous Carcinomas, Undifferentiated Carcinomas
- Rare entities

Epidemiology and Etiology

Occurrence
- Approx. 6000 new cases in Germany/year
- Significant increase in the incidence of adenocarcinoma in Europe and the USA
- 80% men, 20% women

Risk Factors
Squamous Cell Carcinoma
- Nicotine abuse
- Alcohol abuse
- Achalasia
- History of radiation therapy in the head and neck region

Adenocarcinoma
- Gastroesophageal reflux disease
- Barrett's Esophagus
- Nicotine abuse
- Achalasia

Tumor Spread

Continuous Spread
- Intramural
- Direct organ infiltration (pericardium, pleura, aorta)

Lymphogenous Spread
- Lymph node levels: cervical, mediastinal and abdominal

Hematogenous Spread
- Hepatic: via portal vein
- Pulmonary, osseous or cerebral: via vena cava or liver

Classification
UICC/AJCC TNM 8 Classification (2017)
- T (Tumor)
  - TX Tumor cannot be assessed
  - T0 No primary tumor detectable
  - Tis High-grade dysplasia (malignant cells above the basement membrane)
  - T1a Infiltration of the lamina propria and muscularis mucosa
  - T1b Infiltration of the tela submucosa (further subcategories)
  - T2 Infiltration of the tunica muscularis
  - T3 Infiltration of the adventitia
  - T4a Infiltration of pleura, pericardium, peritoneum, azygos vein or diaphragm
  - T4b Infiltration of other organs, such as aorta, vertebral body or trachea
- N (Lymph nodes)
  - NX Regional lymph nodes cannot be assessed
  - N0 No metastases in the lymph nodes
  - N1 Metastases in 1–2 regional lymph nodes
  - N2 Metastases in 3–6 regional lymph nodes
  - N3 Metastases in 7 or more regional lymph nodes
- M (Metastases)
  - M0 No Distant Metastasis
  - M1 Distant Metastasis(es)

UICC Stages According to the TNM Classification 8th Version (2017)

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage IIa	T1	N1	M0
Stage IIb	T2	N0	M0
Stage III	T2	N1	M0
	T3–4a	N0–1	M0
Stage IVa	T1–4a	N2	M0
	T4b	N0–2	M0
	T1–4	N3	M0
	Any T	N3	M0
Stage IVb	Any T	Any N	M1

Symptoms
- Weight loss
- Retrosternal pain
- Hematemesis/melena/anemia

Diagnosis

Esophagogastroduodenoscopy (EGD)

- Biopsy
- Confirmation of tumor diagnosis
- Localization of the tumor

Endosonography

- Infiltration depth
- Assessment of T- and N-stage

Thoracic CT, Abdominal CT

- Assessment of primary tumor and lymph node involvement
- Distant metastases

Bronchoscopy

- For squamous cell carcinoma to exclude second carcinoma
- If the tumor is closely located to the central airways (tracheal infiltration, bronchial infiltration)

Panendoscopy

- In the case of squamous cell carcinoma to exclude second carcinoma in the ENT area

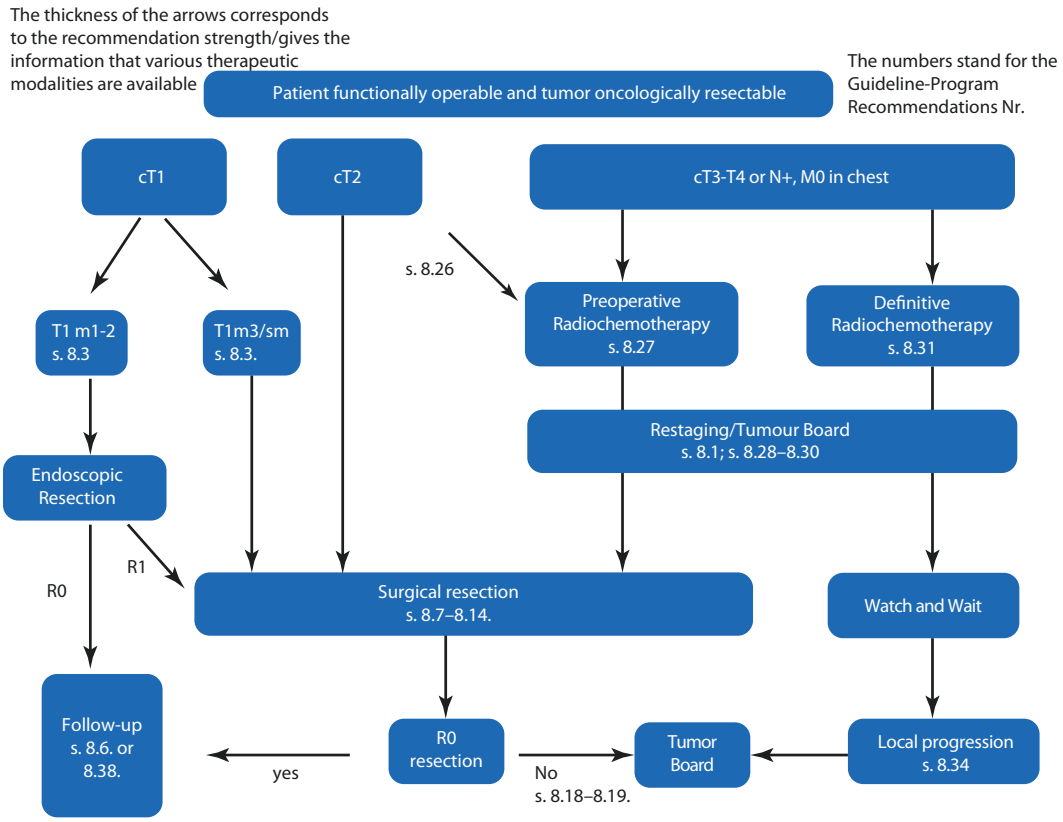
PET-CT, MRI Abdomen

- Not required for primary diagnosis
- Only to exclude metastases in rare and special indications
- Helpful in recurrence diagnosis

Therapy

Indication

- Therapeutic approach according to guideline (AWMF 021/023OL: 12/2018) depending on preoperative staging (◘ Figs. 1.3 and 1.4)
- Surgical therapy in specialized centers with high case load
- In early stages (T1a) consider endoscopic local therapy



◘ **Fig. 1.3** Treatment algorithm for functionally operable and oncologically resectable squamous cell carcinoma of the esophagus. (From Guideline Program in Oncology 2018; courtesy)

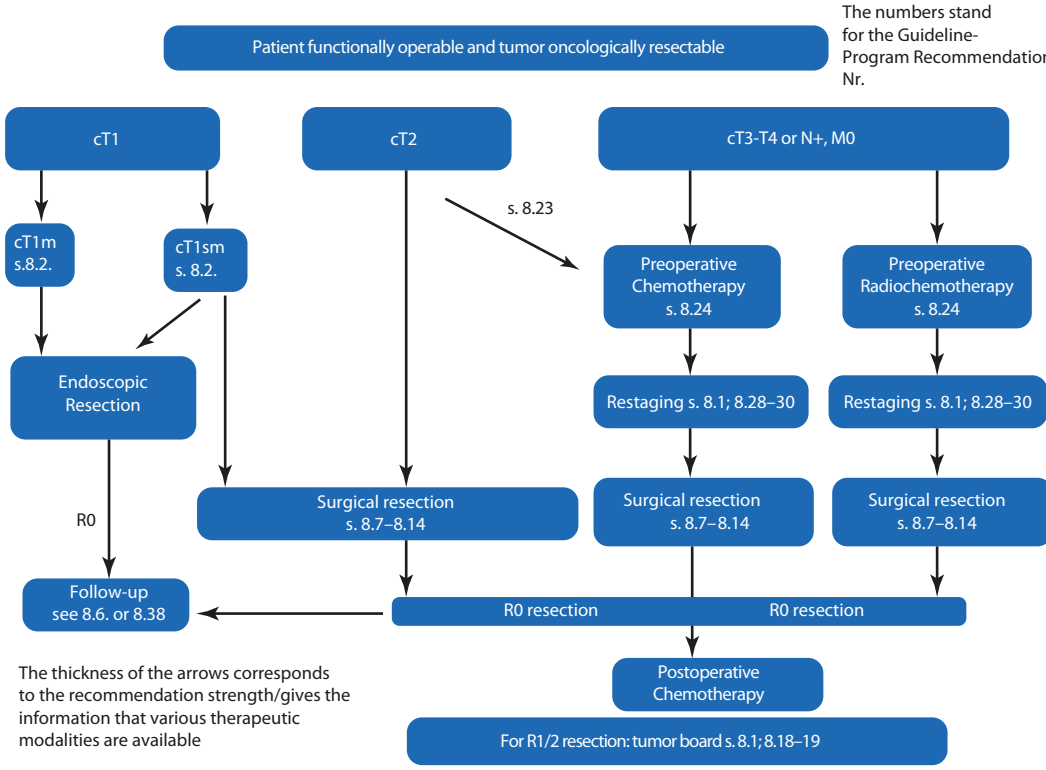


Fig. 1.4 Treatment algorithm for functionally operable and oncologically resectable adenocarcinomas of the esophagus and gastroesophageal junction. (From Guideline Program in Oncology 2018; courtesy)

- In case of planned two-cavity procedure, consider general operability, especially cardiac and pulmonary comorbidity
- In case of inoperability, severe comorbidity or patient preference: definitive radiochemotherapy

Multimodal Therapy

Principles

- Improved local and systemic tumor control
- Indication usually for tumor stage T3/4 and/or positive lymph node status
- Down staging of primary inoperable tumors
- Increased R0 resection rates
- Reduced recurrence rate
- Prolonged overall survival
- After neoadjuvant therapy: re-staging (endoscopy and CT)
- Surgery: usually at time interval of 4–6 weeks after neoadjuvant therapy

Neoadjuvant Radiochemotherapy

- For squamous cell carcinoma and adenocarcinoma
  - 36–54 Gy radiation dose with simultaneous chemotherapy
  - Currently different protocols (e.g. CROSS protocol: 41.4 Gy divided into 23 single doses of 1.8 Gy each plus chemotherapy with carboplatin and paclitacel)

Perioperative Chemotherapy

- Alternative treatment protocol for adenocarcinoma of the gastroesophageal junction
- Protocol analogous to multimodal therapy for gastric carcinoma (► Sect. 1.6)
- Advantages over neoadjuvant radiochemotherapy possibly due to improved systemic tumor control and reduced perioperative morbidity

Additive/Palliative Therapy

Principles

- Endoscopic stent insertion
- Installation of percutaneous endoscopic gastrostomy or catheter jejunostomy for enteral feeding
- Palliative radiochemotherapy
- Palliative chemotherapy

Strategy

- In case of locally inoperable tumor or functionally inoperable patient: definitive radiochemotherapy (long-term survival >10–35% in stage II/III)
- In case of R1/R2 resection and lack of possibility for surgical resection: postoperative radiochemotherapy
- In case of recurrence or tumor persistence after definitive radiochemotherapy: salvage esophagectomy may be necessary (**caution**: increased postoperative morbidity)
- In metastatic adenocarcinoma: palliative chemotherapy
- In metastatic squamous cell carcinoma: palliative chemotherapy
- In case of pronounced dysphagia and weight loss: endoscopic implantation of a self-expanding metal stent recommended, also possible prior to neoadjuvant therapy

Operative Therapy Principles

Local Endoscopic Interventional Procedures

- Indication:
  - If there is evidence of high-grade intraepithelial neoplasia or mucosal carcinoma (<2 cm, no lymphatic invasion L0, no venous invasion V0, no ulceration, grading G1/G2) in Barrett's esophagus
  - In case of lymphatic or blood vessel infiltration, poor degree of differentiation (≥G3), submucosal infiltration or tumor remnant at the basal resection margin: indication for esophageal resection
- Disadvantages:
  - No reliable assessment of the lymph node status
  - No certainty of R-status for extended resections in piece-meal technique
  - High risk of stenosis after (circular) resection of extensive findings
- Principle and Endoscopic Procedure:
  - Endoscopic resection depending on the extent and localization of the tumor
  - In case of Barrett's mucosa additional thermoablation of the complete area
  - Endoscopic mucosal resection (EMR)
  - Endoscopic submucosal dissection (ESD)

Esophagectomy

- Principles of resection:
  - For AEG 3: transhiatal extended gastrectomy (► Sect. 1.6)
  - For AEG 2 alternatively:
    - Transhiatal extended gastrectomy or
    - Esophagectomy
    - For tumors with massive infiltration on the stomach: esophagogastrectomy
  - For AEG 1: always abdominothoracic esophagectomy
  - Standard procedure = Ivor-Lewis esophagectomy (see "Operative Procedure"):
    - Mobilization and resection of the esophagus via right thoracotomy or thoracoscopy with mediastinal en bloc lymphadenectomy
    - Dissection of the esophagus at the level of the azygos arch or in the upper thoracic aperture
    - Lymphadenectomy in the abdominal compartment and gastric mobilization and advancement via upper abdominal laparotomy or laparoscopy
  - Alternatively, thoracic and abdominal parts can each be minimally invasive
    - Hybrid laparoscopic/thoracotomic or laparotomic/thoracoscopic procedure widely used
    - Complete laparoscopic and thoracoscopic (if necessary, with robotic-assisted technique)
    - Potential reduction of pulmonary complications
    - Less blood loss, faster recovery

  - For squamous cell carcinoma: resection of the esophagus into the upper thoracic aperture with cervical lymphadenectomy if necessary
  - One-stage resection-reconstruction as a standard procedure
  - Two-stage resection-reconstruction: with temporary cervical diversion of the esophagus and gastric blind closure, interval reconstruction in septic patients after (tumor) perforation
- Principles of Reconstruction:
  - Preferred Reconstruction via gastric mobilization, advancement and intrathoracic anastomosis
  - Alternatively cervical anastomosis via separate left cervical incision
  - Reconstruction with colonic interposition for tumor infiltration of the stomach
  - Reconstruction in the posterior mediastinum, alternatively retrosternal
- Postoperative complications:
  - High rate of perioperative complications (morbidity up to 70%)
  - Pulmonary complications (pneumonia, pleural effusion, pneumothorax)
  - Anastomotic leak (therapy by stent insertion or ENDOVAC, surgical revision)
  - Chyle Leak due to injury of the thoracic duct
  - Delayed gastric emptying
  - Wound infections, post-operative bleeding, etc.
  - Cardiac complications (high rate of arrhythmias, pericardial effusion)
- Perioperative management:
  - Preoperative respiratory therapy, exercise and nutrition program
  - Treatment of patients in specialized centers
  - Optimized anaesthetic and intensive care management
  - Peridural catheter for postoperative analgesia
  - Postoperative nutrition via catheter jejunostomy, alternatively parenteral nutrition
  - Aspiration prophylaxis, if necessary scheduled postoperative bronchoscopies

Surgical Procedure

Ivor Lewis Esophagectomy

- Preoperative colonoscopy: if colon interposition necessary!
- Anaesthetic preparation: Peridural catheter, central venous catheter, continuous arterial blood pressure measurement, double lumen endotracheal tube
- Positioning of the patient in a semilateral position, right thoracic elevation
- Alternative: Intraoperative repositioning Left lateral position → Supine position
- Right thoracotomy
- Division of the azygos vein
- Mobilization of the esophagus including the periesophageal lymphatic and fat tissue and the peribronchial lymph nodes. **Caution**: thoracic duct!
- Upper abdominal median laparotomy
- Mobilization of the stomach while sparing the gastroepiploic arcade
- Division of the left gastric artery and short gastric vessels
- Abdominal lymphadenectomy (hepatic artery, coeliac trunk, splenic artery)
- Resection of the esophagus and the proximal stomach
- Mobilization of the stomach (stapler-resection)
- Transhiatal advancement of the stomach into the posterior mediastinum
- End-to-side anastomosis using a circular stapler
- Insertion of chest tubes
- Alternatively, minimally invasive procedure

Prognostic Factors

- Postoperative staging (UICC)
- Lymph node ratio (quotient of affected and removed lymph nodes)
- Lymphatic/venous invasion
- Response to neoadjuvant therapy (clinical and histopathological regression)
- R status (residual status)

Follow-Up

Purpose

- Symptom-oriented follow-up
- Diagnosis and treatment of functional disorders (recurrence or benign complications of treatment)
- Nutritional medical follow-up, additional nutrition if necessary
- Early detection of potentially curable local recurrences

Implementation

- After successful endoscopic therapy of a high-grade intraepithelial neoplasia or an early carcinoma, regular control endoscopies (after 3 months, then every 6 months for 2 years and thereafter annually)
- After oesophagectomy, no predefined scheme, e.g. history, physical examination and computed tomography of abdomen/thorax every 6 months

1.5 Benign Diseases of the Stomach

1.5.1 Gastroduodenal Ulcer Disease

Summary

- Mostly due to Helicobacter pylori positive gastritis
- Non-specific symptoms or upper abdominal pain
- Complications: Hemorrhage, penetration, perforation, stenosis, neoplasia
- Differential diagnosis: gastric cancer
- Conservative therapy of the underlying pathology + proton-pump inhibitors
- Endoscopic therapeutic options for complications: Injection or clipping of bleedings, dilation of stenoses
- Surgical interventions for complicated ulcer disease:
  - Surgical hemostasis in case of endoscopically uncontrollable bleeding
  - Excision and suturing for perforation
  - Distal gastric resection for perforation or stenosis

Etiology

Appearance

- Incidence approx. 200/100,000
- Ratio men:women = 3:1
- Localization mostly at the small curvature, antrum and in first part of duodenum

Risk Factors

- Chronic gastritis due to Helicobacter pylori
- Genetic predisposition (blood group 0, HLA B-5)
- Use of NSAIDs (ASS, diclofenac, ibuprofen): increased risk especially in combination with glucocorticosteroids
- Smoking
- Zollinger-Ellison syndrome (gastrinoma)
- Hypercalcemia, usually with parathyroid adenoma
- Acute stress ulcer: risk factor independent etiology (after polytrauma, long intensive care stay, major operations, etc.)

Symptoms

- Epigastric pain
- Fasting pain: for duodenal ulcer
- Postprandial pain: for gastric ulcer
- Upper gastrointestinal bleeding
- Perforation with rapid onset (chemical) peritonitis. **Caution**: masked symptoms with occult perforation

Diagnosis

Endoscopy (EGD)

- Confirmation of diagnosis and biopsy to exclude malignancy
- With conservative therapy always re-endoscopy to record complete healing of the ulcer (DD cancer!)

1

Radiology

- Computed tomography with oral contrast medium: only indicated to exclude penetration or perforation

Further Diagnosis

- Diagnosis of Helicobacter pylori
- Determination of gastrin, calcium and parathormone to exclude a hormonal cause

Complications

- Bleeding
- Perforation/penetration
- Stenosis
- Neoplasia

Therapy

Conservative Therapy

- For Helicobacter-positive gastritis: eradication with proton-pump inhibitor and clarithromycin + amoxicillin (French triple therapy) or metronidazole (Italian triple therapy) for 7 days
- Avoidance of noxious substances (NSAR, smoking, coffee, alcohol, stress)
- Proton pump inhibitors

Interventional Therapy

- In case of ulcer bleeding: bleeding control by injection with adrenalin/histoacryl or clipping
- In case of endoscopically uncontrollable ulcer bleeding or high risk of recurrent bleeding after primary successful endoscopic hemostasis: Interventional angiography with endovascular hemostasis (coiling of gastroduodenal artery)
- In case of stenosis: endoscopic dilation (bougie) possible

Surgical Therapy

- In gastrinoma or parathyroid adenoma: surgical therapy of the underlying pathology (► Chaps. 6 and 9).
- Surgical therapy of ulcer complications:
  - Ulcer perforation: if possible excision and primary suture of the ulcer, otherwise distal gastric resection
  - Gastric stenosis: distal gastric resection or gastroenterostomy, if malignant cause of stenosis can be ruled out
- 2/3 gastric resection (Billroth I with gastroduodenostomy or Bilroth II with gastrojejunostomy) and the vagotomy procedures for the treatment of ulcer disease: obsolete today due to effective conservative treatment options—PPI, HP (Helicobacter pylori) eradication.
- In case of ulcer bleeding in the first part of duodenum: extra- and intraluminal (duodenum) closure of the gastroduodenal artery.

Surgical Procedure

Distal Gastric Resection Analogous to Billroth II

- Anesthesiology preparation: peridural catheter, central venous catheter, continuous arterial blood pressure measurement
- Supine position
- Upper abdominal median laparotomy, alternatively upper abdominal transverse laparotomy
- Distal gastric mobilization of the stomach
- Resection of approx. 2/3 of the distal stomach by transection of the postpyloric duodenum
- Closure of the duodenal stump with stapler, if necessary additional sutures
- Reconstruction using a small bowel loop (Y-Roux technique or classical omega reconstruction)

1.5.2 Guidelines

AWMF Guideline Register: Helicobacter pylori and gastroduodenal ulcer disease (German Society of Gastroenterology, Digestive and Metabolic Diseases, AWMF), 2014, AWMF registration number: 021/001, ► http://www.awmf.org/leitlinien.html

1.6 Malignant Diseases of the Stomach

1.6.1 Gastric Adenocarcinoma

Summary

- Frequent tumor worldwide, decreasing incidence in the western population
- Intestinal or diffuse histological differentiation according to Lauren
- Early lymphogenic, hematogenic and peritoneal spreading
- Tumour stage is often advanced at the time of diagnosis
- For T3/4 or N+: Multimodal therapy concept consisting of perioperative chemotherapy and surgical resection
- Therapeutic standard: Total gastrectomy with D2 lymphadenectomy
- For distal tumors: Subtotal gastrectomy possible

Definition

- All tumors of the gastric antrum, corpus and cardia with distance >5 cm from the Z-line
- AEG 3 tumors without esophageal infiltration (UICC TNM 7, 2010)
- Tumors of the gastroesophageal junction with esophageal infiltration are classified as esophageal cancers (UICC TNM 7, 2010)

Forms

- According to histology (Lauren classification)
  - Intestinal type
  - Diffuse type (signet ring cells)
- According to localization
  - Antrum
  - Corpus/Fundus
  - Subcardial (AEG 3)

Epidemiology and Etiology

Occurrence

- Approx. 15,000 new cases in Germany per year
- Predominantly men (ratio men:women = 3:2)
- Older patients
- Incidence varies considerably from region to region (increased incidence mainly in Asia, but also in South America, Southern and Eastern Europe)
- In the western population, decreasing incidence of gastric cancer, but increasing incidence of gastroesophageal junction tumors

Risk Factors

- Helicobacter pylori
- Age
- Low socioeconomic status
- Nicotine and alcohol abuse
- Family history
- Previous gastric surgery
- Pernicious anaemia
- Nutritional and environmental factors

Tumor Spread

- Continuous
  - Intramural
  - Direct organ infiltration (spleen, pancreas, colon, duodenum)
- Lymphogenic (◘ Fig. 1.5)
  - Early lymphogenic metastasis
  - Lymph node compartments D1–D4
- Hematogenic
  - Hepatic via portal vein
  - Pulmonary, osseous, cerebral
- Peritoneal carcinomatosis
  - Often early peritoneal seeding
  - Ovarian metastasis: "Krukenberg tumor"

Classification

TNM 7 Classification (2010)

- T (Tumor)
  - T0 No primary tumor detectable
  - Tis carcinoma in situ
  - T1a Infiltration of the lamina propria or muscularis mucosae
  - T1b Infiltration of the tunica submucosa
  - T2 Infiltration of the muscularis propria
  - T3 Infiltration of the subserosa

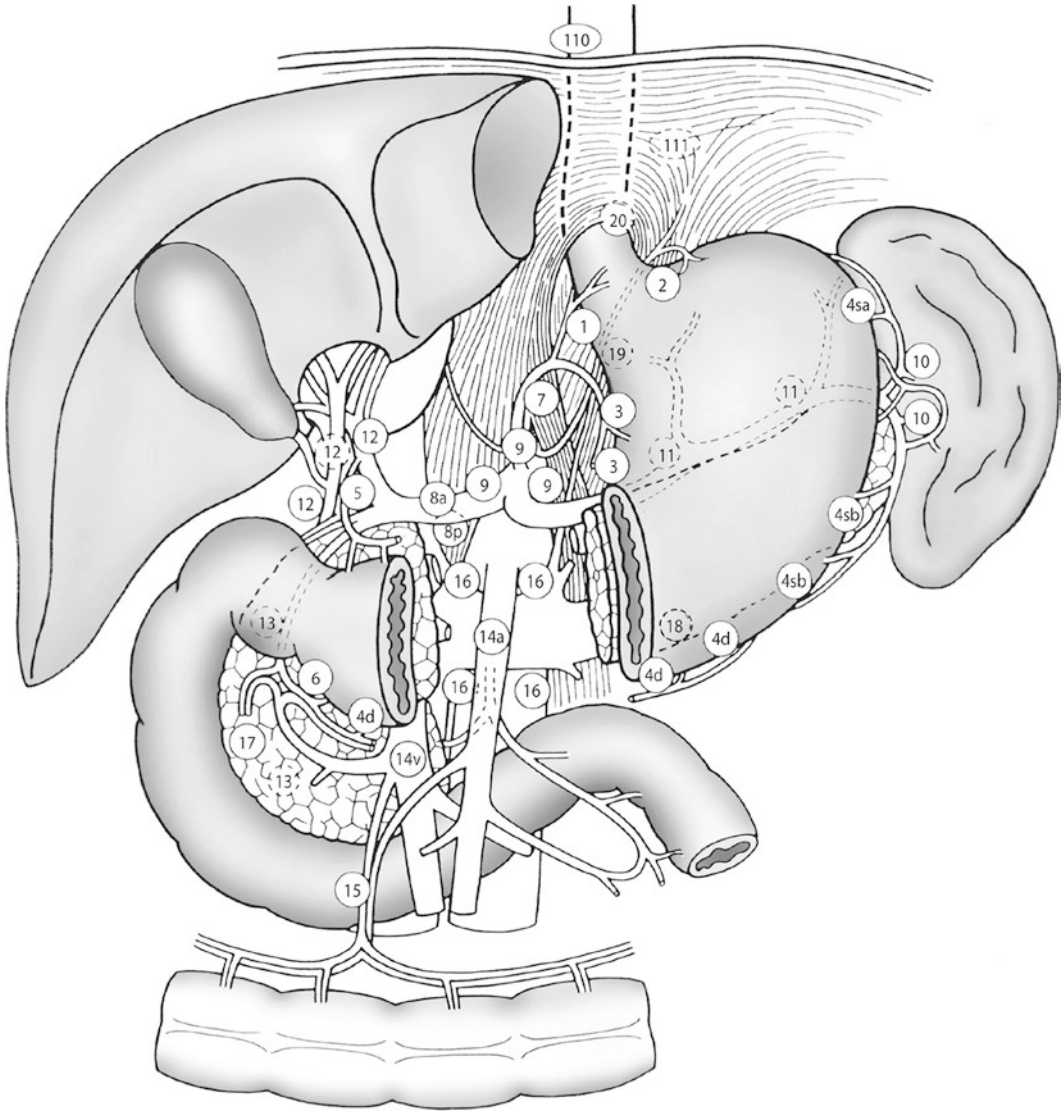


Fig. 1.5 Regional and distant lymph nodes of the stomach. Lymph node compartment D1: *1–6*, lymph node compartment D2: *7–11*, lymph node compartment D3: *12–14*, lymph node compartment D4: *15–16*. (According to Siewert et al. 2010)

  - T4a Penetration of the visceral peritoneum **without** infiltration of adjacent organs
  - T4b Penetration of the visceral peritoneum **with** infiltration of adjacent organs (pancreas, spleen, liver)
- N (Node)
  - N0 No metastases in the lymph nodes
  - N1 Metastases in 1–2 regional lymph nodes
  - N2 Metastases in 3–6 regional lymph nodes
  - N3a Metastases in 7–15 regional lymph nodes
  - N3b Metastases in 16 or more regional lymph nodes
- M (Metastasis)
  - M0 No distant metastases
  - M1 distant metastasis(s)

UICC Stages According to the TNM Classification

Stage 0	Tis	N0	M0
Stage Ia	T1	N0	M0
Stage Ib	T1	N0	M0
	T2	N0	M0
Stage IIa	T1	N2	M0
	T2	N1	M0
	T3	N0	M0
Stage IIb	T1	N3	M0
	T2	N2	M0
	T3	N1	M0
	T4a	N0	M0
Stage IIIa	T2	N3	M0
	T3	N2	M0
	T4a	N1	M0
Stage IIIb	T3	N3	M0
	T4a	N2	M0
	T4b	N0/1	M0
Stage IIIc	T4a	N3	M0
	T4b	N2/3	M0
Stage IV	Each T	Each N	M1

Symptoms

- Often unspecific
- Inappetence/weight loss
- Nonspecific upper abdominal pain
- Recurrent vomiting
- Hematemesis/melena/anaemia

Diagnosis

Esophagogastroduodenoscopy/ Endosonography

- Biopsy
- Confirmation of tumor diagnosis
- Localization of the tumor
- Infiltration depth
- Assessment of T- and N-stage

Thoracic CT, Abdominal CT

- Assessment of the primary tumor and lymph node involvement
- Distant metastases

Diagnostic Laparoscopy

- Staging of peritoneal carcinomatosis
- Biopsy if necessary
- Laparoscopy is an important diagnostic step prior to preoperative therapy

PET-CT, MRI Abdomen, Bone Scintigraphy

- Not required for primary diagnosis
- Only to exclude metastases in rare and special indications
- Helpful in recurrence diagnosis

Therapy

Indication

- Therapeutic procedure according to the guideline depending on the preoperative staging (◘ Table 1.2 and ◘ Fig. 1.6)
- Consider endoscopic mucosal resection at T1a stage

◘ **Table 1.2** Therapeutic strategy for gastric cancer depending on preoperative staging; T1 and T2 see Fig. 1.6

Tumor stage	Therapy
T2 N0 M0	Primary resection
T3/4 N0 M0 Tx N+ M0	Perioperative chemotherapy + resection
Tx Nx M1	Palliative therapy: palliative gastrectomy, gastroenterostomy, palliative chemotherapy Consider resection + peritonectomy + HIPEC for limited peritoneal carcinomatosis and good response to perioperative chemotherapy

HIPEC hyperthermic intraperitoneal chemoperfusion

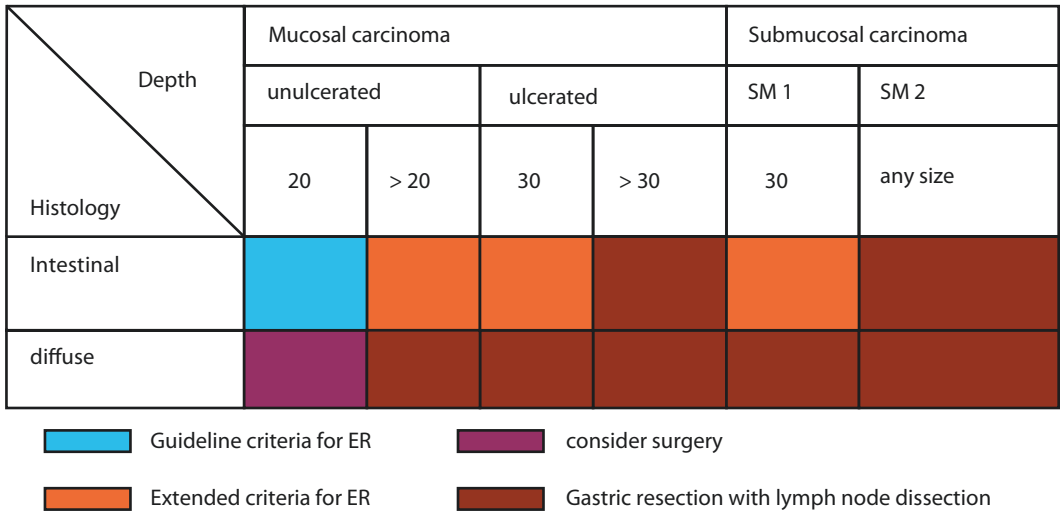


Fig. 1.6 Guideline criteria and expanded criteria for early gastric cancer. (From Guideline Program in Oncology 2019; used with permission)

- Curative therapy for localized and locally advanced tumors
- In case of inoperability, severe comorbidity or patient request palliative chemotherapy
- Palliative gastrectomy for limited metastasis in the young and/or low comorbid patient
- Palliative gastrectomy for tumor hemorrhage and tumor perforation

Multimodal Therapy

Principles

- Improved local and systemic tumor control
- Prognostic improvement in locally advanced tumors
- Indication usually for tumor stage T3/4 and/or positive lymph node status
- Down staging of inoperable tumors
- Increased R0 resection rates
- Reduced recurrence rate
- Prolonged overall survival
- After neoadjuvant therapy: re-staging (endoscopy, computed tomography)
- Surgery at intervals of 4–6 weeks after neoadjuvant therapy

Perioperative Chemotherapy

- Combination of pre- and postoperative chemotherapy
- Various platinum-based chemotherapy protocols:
  - ECF (epirubicin, cisplatin and 5-fluorouracil)
  - ECX (epirubicin, cisplatin, capecitabine)
  - EOX (epirubicin, oxaliplatin, capecitabine)
  - FLOT (docetaxel, folic acid, 5-fluorouracil, oxaliplatin)
  - DCF (docetaxel, cisplatin, 5-fluorouracil)
  - Cisplatin + 5-fluorouracil

Adjuvant Radiochemotherapy

- Widely used in the USA, in Europe only for limited lymphadenectomy (≤D1) or R1/2 resection according to interdisciplinary board recommendation.
- Consider in lymph node positive patients without preoperative chemotherapy even after D2 lymphadenectomy.

Additive/Palliative Therapy

Principles

- Palliative gastrectomy for bleeding tumors or limited metastatic tumors with obstructive symptoms
- Gastroenterostomy for stenosis and inoperable tumor
- Palliative chemotherapy
- Definitive radiochemotherapy

Strategy

- In case of R1/R2 resection and no possibility of further surgical resection: adjuvant radiochemotherapy
- In case of locally inoperable tumor or distant metastasis: palliative chemotherapy
- Palliative gastrectomy only for bleeding tumors after exhaustion of endoscopic and angiographic methods
- Bypass procedure: for clinically manifest gastric outlet stenosis

Operative Therapy Principles

- Local endoscopic interventional procedures:
  - Indication:
    - Intraepithelial neoplasms of any size as well as early gastric cancers that meet all four of the following criteria **should be** resected endoscopically en-bloc: <2 cm in diameter, not ulcerated, mucosal carcinoma, intestinal type/grade of differentiation good to moderate "(G1/G2)"
    - Early gastric cancers with a maximum of one "extended criterion" can be endoscopically resected curatively. Extended criteria are; differentiated mucosal carcinoma (G1/2) without ulceration and size >2 cm; differentiated mucosal carcinoma with ulceration <3 cm; well-differentiated carcinoma with submucosal invasion <500 µm and size <3 cm; undifferentiated mucosal carcinoma <2 cm in diameter (provided there is no biopsy evidence of tumor cells <1 m)
    - Indication for gastric (partial) resection plus lymphadenectomy in the presence of risk criteria or residual tumor at the basal resection margin
  - Disadvantages:
    - No reliable assessment of the lymph node status
    - No assessment of R-status for extended resections in piece-meal technique
  - Principle and endoscopic procedure:
    - Endoscopic resection depending on extension and localization
    - Endoscopic mucosal resection (EMR)
    - Endoscopic submucosal dissection (ESD)
    - Goal: En-bloc resection
- Gastrectomy/Gastric Resection:
  - Principles of resection:
    - A safe resection distance of 5 cm for the intestinal type and 8 cm for the diffuse type should be achieved
    - Gastrectomy with lymphadenectomy of the compartments D1 and D2 is standard
    - For distal tumors, subtotal (4/5) gastrectomy is sufficient, leaving a small proximal gastric remnant
    - In case of subcardial gastric cancer (AEG 3) a transhiatal extended gastrectomy including the distal esophagus is necessary
    - In case of limited peritoneal carcinomatosis, gastrectomy with local peritonectomy and, if necessary, hyperthermic intraoperative chemotherapy can be performed with curative intention
    - Laparoscopic and robotic-assisted surgery can be performed in a specialized center
  - Principles of Reconstruction:
    - No clear recommendation, procedure depending on the experience of the surgeon
    - Classical Roux-en-Y reconstruction
    - Different techniques of jejunum pouch reconstruction, e.g. J-pouch (possible quality of life advantage)
  - Postoperative complications:
    - Duodenal Stump Insufficiency
    - Anastomotic leak of the esophagojejunostomy
    - Pulmonary and cardiac complications
    - Pancreatic fistula after D2 lymphadenectomy
    - Cachexia

Surgical Procedure
Gastrectomy

- Anaesthesiological preparation: Peridural catheter, central venous catheter, continuous arterial blood pressure measurement
- Supine position
- Upper abdominal median laparotomy, alternatively upper abdominal transverse laparotomy
- Incision of gastrocolic ligament and opening of lesser sac (great omentum resected en-bloc with gastric specimen)
- Transection of the gastro-splenic ligament and short gastric vessels near the spleen
- Division of the gastroomental artery close to the pancreatic head
- Transection of the right gastric artery
- Division of the postpyloric duodenum and closure of the duodenal stump
- D2 lymphadenectomy including the lymph nodes around the hepatic, celiac and splenic arteries, and division of left gastric artery at its origin near the coeliac trunk
- Division of the vagal nerve at the abdominal esophagus
- Dissection/Mobilisation of the abdominal esophagus
- Roux-en-Y Reconstruction using a small bowel loop (Jejunum)

Prognosis

Prognostic Factors

- Postoperative stage (UICC TNM)
- Lymph node ratio (quotient of affected and removed lymph nodes)
- Lymphatic/venous invasion
- Response to neoadjuvant therapy (clinical and histopathological regression according to Becker et al. 2011)
- R status (residual status)

Follow-Up

Goals

- Symptom-oriented follow-up
- Rule out functional disorders as a result of recurrence or as benign complications of treatment
- Nutritional medical follow-up, additional nutrition if necessary
- Early detection of potentially curable local recurrences
- Early detection of distant metastases

Implementation

- After successful endoscopic therapy of a high-grade intraepithelial neoplasia or an early cancer, regular control endoscopies (every 3 months in the first year, every 6 months in the second year, thereafter annually)
- After gastrectomy, so-called symptom-oriented follow-up without a predefined scheme, e.g. anamnesis, physical examination and computer tomography of abdomen/thorax every 6 months

1.6.2 Gastrointestinal Stromal Tumours (GIST)

► Chapter 14

1.6.3 Guidelines

Guideline program oncology (German Cancer Society, German Cancer Aid, AWMF): "Gastric carcinoma"—Diagnostics and therapy of adenocarcinomas of the stomach and esophagogastric junction, long version 2.0, 8/2019, AWMF registration number: 032-009OL

1.7 Diseases of the Duodenum

Summary

- Duodenal diverticulum: Common finding usually not requiring any treatment
- Duodenal ulcer: ► Sect. 1.5
- Duodenal cancer

- Rare tumor disease
- FAP (familial adenomatous polyposis)/duodenal polyps as risk factors

1.7.1 Diverticular Disease of the Duodenum

Incidence

- Approximately 10–20%

Types

- Duodenal diverticula are mostly found near pancreas head
- Rarely intraluminal or intramural duodenal diverticula, as congenital malformations, originating from a mucosal duplication

Symptoms

- Mostly incidental finding during ERCP
- Mostly asymptomatic and without need for therapy

Therapy

- Very rarely duodenal diverticula require surgery

Complications

- Rarely upper GI bleeding and perforation.
- Very rarely obstruction of bile duct (jaundice) and pancreatic duct (pancreatitis) due to compression.

1.7.2 Duodenal Cancer

Etiology and Tumor Manifestation

Appearance

- Rare tumor disease
- Duodenal adenoma as a precancerous condition
- Frequently in the context of hereditary tumor syndromes: FAP, HNPCC ("hereditary non-polyposis colorectal cancer"), Peutz-Jeghers syndrome, Gardner syndrome
- Other risk factors: Crohn's disease, celiac disease
- Up to 90% of patients with FAP develop duodenal polyps; lifetime risk of duodenal cancer is 3–4%

Symptoms

- Often asymptomatic
- Hematemesis/anaemia
- Stenosis/inappetence/weight loss/vomiting
- Obstructive jaundice or pancreatitis if infiltration of the duodenal papilla

Diagnosis and Therapy

Diagnosis

- Endoscopic diagnosis with biopsy: always + immediately in case of suspected tumor
- In case of tumor detection: CT abdomen/thorax and if necessary endosonography for reliable staging

Endoscopic Therapy

- Duodenal polyps are removed endoscopically analogous to colon polyps
- In the case of larger, flat polyps, consider ablation using the piece-meal technique with additional thermal ablation of the affected area, if necessary
- For duodenal polyposis, determine Spigelman score (polyp number, polyp size, histologic type, grading of intraepithelial neoplasia): Stage I–IV
  - In stage I–III regular endoscopic controls
  - In stage IV, consider surgical therapy

Surgical Therapy

- Surgical excision with transverse closure of the excision site: for adenomas that cannot be removed by endoscopy
- Transduodenal papillary excision with re-insertion of the main pancreatic duct and bile duct: in case of papillary adenomas
- Pancreas sparing duodenectomy (caution: morbidity): for benign tumors (e.g. duodenal polyposis)
- Radical oncological resection (pylorus preserving pancreatoduodenectomy or Whipple operation): in the case of duodenal cancer

Multimodal Therapy

- No recommendations due to conflicting data
- No neoadjuvant therapy
- Adjuvant therapy analogous to the recommendations for colon cancer (► Chap. 3)
- For ampullary cancer survival benefit with adjuvant chemotherapy with 5 FU/leukovorin or gemcitabine (ESPAC-3) after R0 resection

Palliative Therapy

- Surgical gastroenterostomy: as a bypass procedure in symptomatic patients with inoperable tumors
- Endoscopic biliary stent/prosthesis insertion or surgical palliative biliodigestive anastomosis: in the case of obstructive jaundice
- Palliative chemotherapy: analogous to colon cancer (► Chap. 3), consider palliative radiochemotherapy if necessary

Prognosis

- 5-year survival rate: approx. 30%
- For N0, M0, R0 = 50–70% 5-year survival

1.7.3 Guidelines

Guideline program oncology (German Cancer Society, German Cancer Aid, AWMF): S3 guideline colorectal carcinoma, long version 2.1, 2019, AWMF registration number: 021-007OL, ► http://leitlinienprogrammonkologie.de/Leitlinien.7.0.html

Oncology guideline program (German Cancer Society, German Cancer Aid, AWMF): Diagnostics and therapy of squamous cell carcinomas and adenocarcinomas of the esophagus, long version 2.0, 2018, AWMF registration number: 021/023OL, ► http://leitlinienprogrammonkologie.de/Leitlinien.7.0.html

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