76 T_h e vermiform appendix

- unexpected tumour of the appendix Incidental neopl

ANATOMY

ANATOMY

The vermiform appendix is present only in humans, certain anthropoid apes and the wombat. It is a blind muscular tube with mucosal, submucosal, muscular and serosal layers. Morphologically , it is the undeveloped distal end of the large caecum found in many lower animals. At birth, the appendix is short and broad at its junction with the caecum, but di ff erential growth of the caecum produces the typical tubular structure by about the age of 2 years (Condon). During childhood, contin - ued growth of the caecum commonly rotates the appendix into a retrocaecal but intraperitoneal position (Figure 76.1). In approximately one-quarter of cases, rotation of the appendix does not occur, resulting in a pelvic, subcaecal or paracaecal position. Occasionally , the tip of the appendix becomes extraperitoneal, lying behind the caecum or ascending colon. Rarely , the caecum does not migrate during development to its normal position in the right lower quadrant of the abdomen. In these circumstances, the appendix can be found near the gallbladder or, in the case of intestinal malrotation, in the left iliac fossa, causing diagnostic di ffi culty if appendicitis develops (Figure 76.2).

Figure 76.2 Left-sided caecum and appendix due to intestinal mal rotation.

Aetiology

Aetiology

There is no unifying hypothesis regarding the aetiology of acute appendicitis. Decreased dietary fibre and increased consumption of refined carbohydrates may be important. As with colonic diverticulitis, the incidence of appendicitis is lowest in societies with a high dietary fibre intake. In resource- poor countries that are adopting a more refined Western-type diet, the incidence continues to rise. This is in contrast to the dramatic decrease in the incidence of appendicitis in Western countries observed in the past 30 years. No reason has been established for these paradoxical changes; however, improved hygiene and a change in the pattern of childhood gastrointes tinal infection related to the increased use of antibiotics may be responsible. While appendicitis is clearly associated with bacterial prolif eration within the appendix, no single organism is responsible. A mixed growth of aerobic and anaerobic organisms is usual. The initiating event causing bacterial proliferation is contro v ersial. Obstruction of the appendix lumen has been widely held to be important, and some form of luminal obstruction, either by a faecolith (Figure 76.5) or by a stricture, is found in the majority of cases. A faecolith (sometimes referred to as an appendicolith) is composed of inspissa ted faecal material, calcium phosphates, bacteria and epithelial debris. Rarely , a foreign body is incor porated into the mass. The incidental finding of a faecolith is a rela tive indication for prophylactic appendicectomy or an Reginald Heber Fitz , 1843–1913, Professor of Medicine, Harvard University , Boston, MA, USA. Charles McBurney , 1854–1913, Professor of Surgery , Columbia College of Physicians and Surgeons, New Y ork, NY , USA. In 1889 McBurney published a paper on appendicitis in which he stated, ‘I believe that in every case the seat of greatest pain “determined by the pressure of one finger” has been very exactly between an inch and a half and two inches from the anterior spinous process of the ilium on a straight line drawn from that process to the umbilicus.’ - - - - - - - ter interval appendicectomy in a patient treated conservatively . A fibrotic stricture of the appendix usually indicates previ - ous appendicitis that resolved without surgical intervention. Obstruction of the appendiceal orifice by tumour, particularly carcinoma of the caecum, is an occasional cause of acute appendicitis in middle-aged and elderly patients. Intestinal Enterobius vermicularis (pinworm), can parasites, particularly proliferate in the appendix and occlude the lumen. -

Figure 76.5 Coronal reformat of a computed tomography scan of the abdomen obtained with oral and intravenous contrast, demonstrating an in /f_l amed, enhancing and enlarged appendix that is curled in the midline extending towards the pelvis (arrow). It contains multiple radiopaque appendicoliths. There is extensive periappendiceal fat stranding (courtesy of Professor P MacMahon, FRCR, Dublin, Ireland).

Appendicitis in pregnancy

Appendicitis in pregnancy

Appendicitis appears to be less common in pregnant than in non-pregnant females; however, it is the most common extrauterine acute abdominal condition in pregnancy , with an incidence of 0.5–1 per 1000 pregnancies. Appendicitis is slightly more common in the second trimester and, when compared with the non-pregnant population, presentation during pregnancy is more likely to be complicated. The diag - nosis is often complicated by a delay in presentation as early non-specific symptoms are often attributed to the pregnancy . Obstetric teaching has been that the caecum and appendix are progressively pushed to the right upper quadrant of the abdomen as pregnancy develops during the second and third trimester s. However, pain in the right lower quadrant of the abdomen remains the cardinal feature of appendicitis in) or a pregnancy . Every attempt should be made to establish the diagnosis preoperatively as negative appendicectomy is associated with fetal loss in 4% and preterm labour in 10% of patients. Clinical diagnosis can be di ffi cult and may be facilitated by ultrasound scanning, w hich carries no risk to the fetus and is highly spe - cific; however, sensitivity in some series is low and its reliability varies according to the trimester. MRI scanning carries greater sensitivity but is more expensive and may not be widely avail - able. - Dela ys in diagnosis or in the initiation of definitive treatment pose the greatest risk to the mother and pregnancy . - Fetal loss occurs in 3–5% of cases of acute appendicitis in pregnancy but increases to 20% or more in the presence made. - of perforation. There is insu ffi cient evidence to support a non-operative approach and the pregnant patient with acute appendicitis should proceed to surgery . If the fetus is at a viable gestational age (23 weeks or more), appropriate obstetric and neonatal support should ideally be available. A laparoscopic approach is now considered to be safe in any trimester and, if used, should be initiated via the open Hasson technique. Data from a large series of pregnant women undergoing abdominal surgery (appendicectomy or cholecystectomy) reported a rate of obstetric complications of approximately 5%.

(c) Figure 76.13 Laparoscopic appendicectomy. (a) Hook diathermy dissection of the mesoappendix. is divided. (c) The appendix base is ligated with absorbable ties.

Differential diagnosis

Differential diagnosis

Although acute appendicitis is the most common abdominal surgical emergency , the diagnosis can be extremely di ffi cult at times. There are a number of common conditions that it is wise to consider carefully and, if possible, exclude. The di ff erential diagnosis di ff ers in patients of di ff erent ages; in women, addi tional di ff erential diagnoses are diseases of the female genital tract (Table 76.1). Children The diseases most commonly mistaken for acute appendicitis are acute gastroenteritis and mesenteric lymphad enitis . In mesenteric lymphadenitis, the pain is colicky in nature and cervical lymph nodes may be enlarged. It may be impossible to clinically distinguish Meckel's diverticulitis from acute appendicitis. The pain is similar; however, signs may be central or left sided. Occasionally , there is a history of antecedent abdominal pain or intermittent lower gastrointes tinal bleeding. It is important to distinguish between acute appendicitis and intussusception . Appendicitis is uncommon before the age of 2 years, whereas the median age for intussusception is 18 months. A mass may be palpable in the right lower quad rant, and the preferred treatment of intussusception is reduc tion by careful barium enema. Henoch–Schönlein purpura is often preceded by a sore throat or respiratory infection. Abdominal pain can be severe and can be confused with intussusception or appendici tis. There is nearly always an ecchymotic rash, typically a ff ect ing the extensor surfaces of the limbs and on the buttocks. The face is usually spared. The platelet count and bleeding time are within normal limits. Microscopic haema turia is common. Johann Friedrich Meckel (the younger), 1781–1883, Professor of Anatomy and Surgery , Halle, Germany , described the diverticulum in 1809. Eduard Heinrich Henoch , 1820–1910, Professor of Diseases of Children, Berlin, Germany , described this form of purpura in 1868. Johann Lucas Schönlein , 1793–1864, Professor of Medicine, Berlin, Germany , described this form of purpura in 1837. Burrill Bernard Crohn , 1884–1983, gastroenterologist, Mount Sinai Hospital, New Y ork, NY , USA. Alexandre Emile Y ersin , 1863–1943, bacteriologist, Paris, France. Lobar pneumonia and pleurisy , especially at the right base, may give rise to right-sided abdominal pain and mimic appendicitis. Abdominal tenderness is minimal, pyrexia is marked and chest examination may reveal a pleural friction rub or altered breath sounds on auscultation. A chest radio - graph is diagnostic. Adults Terminal ileitis in its acute form may be clinically indis - tinguishable from acute appendicitis unless a doughy mass of inflamed ileum can be felt. An antecedent history of abdominal cramping, weight loss and diarrhoea suggests regional ileitis rather than appendicitis. The ileitis may be non-specific, due to Crohn's disease (Figure 76.7) or Yersinia infection. Yersinia entero - - colitica causes inflammation of the terminal ileum, appendix and caecum with mesenteric adenopathy . If suspected, serum antibody titres are diagnostic, and treatment with intravenous tetracycline is appropriate. If Yersinia infection is suspected at operation, a mesenteric lymph node should be excised and divided, with half submitted for microbiological culture - (including tuberculosis) and half for histological examination. Ureteric colic does not commonly cause diagnostic dif - ficulty , as the character and radiation of pain di ff ers from that of appendicitis. Urinalysis should always be performed, and the presence of red cells should prompt a supine abdominal radiograph. A renal ultrasound or urogram will provide the -

diagnosis. Right-sided acute pyelonephritis is accompanied and often preceded by increased frequency of micturition. It may cause di ffi culty in diagnosis, especially in women. The leading features are tenderness confined to the loin, fever (temperature - 39°C) and possibly rigors and pyuria. - In perforated peptic ulcer , the duodenal contents pass along the paracolic gutter to the right iliac fossa. As a rule there is a history of dyspepsia and a very sudden onset of pain that starts in the epigastrium and passes down the right - paracolic gutter. In appendicitis, the pain starts classically in - the umbilical region. Rigidity and tenderness in the right iliac fossa are present in both conditions but, in perforated duodenal ulcer, the rigidity is usually greater in the right hypochondrium.

Adult female Elderly Mittelschmerz Diverticulitis Pelvic in /f_l ammatory disease Intestinal obstruction Pyelonephritis Colonic carcinoma Ectopic pregnancy Torsion appendix epiploicae Torsion/rupture of ovarian cyst Mesenteric infarction Endometriosis Leaking aortic aneurysm

An erect chest radiograph will show gas under the diaphragm in 70% of patients. An abdominal computed tomography (CT) examination is valuable when there is diagnostic di ffi culty . Testicular torsion in a teenage or young adult male is easily missed. Pain can be referred to the right iliac fossa, and shyness on the part of the patient may lead the unwary to sus pect appendicitis unless the scrotum is examined in all cases. Acute pancreatitis should be considered in the di ff er ential diagnosis of all adults suspected of having acute appen dicitis and, when appropriate, should be excluded by serum or urinary amylase measurement. Rectus sheath haematoma is a relatively rare but eas ily missed di ff erential diagnosis. It usually presents with acute pain and localised tenderness in the right iliac fossa, often after an episode of strenuous physical exercise. Localised pain with out gastrointestinal upset is the rule. Occasionally , in an elderly patient, particular ly one taking anticoagulant therapy , a rectus sheath haematoma may present as a mass and tenderness in the right iliac fossa after minor trauma. Pelvic inflammatory disease comprises a spectrum of /uni00A0 diseases that include salpingitis, endometritis and tubo ovarian sepsis. The incidence of these conditions is increasing, and the diagnosis should be considered in every young adult female. Typically , the pain is lower than in appendicitis and is bilateral. A history of vaginal discharge, dysmenor rhoea and burning pain on micturition is a helpful di ff erential diagnos tic point. The physical findings include adnexal and cervical tenderness on vaginal examination. When suspected, a high vaginal swab should be taken for Chlamydia trachomatis Neisseria gonorrhoeae culture, and the opinion of a gynaecologist should be obtained (see Chapter 87). lower abdominal and pelvic pain, typically midcycle, which is characteristic of mittelschmerz . Systemic upset is rare, a pregnancy test is negative and symptoms usually subside within hours. Occasionally , diagnostic laparoscopy is required. Retro - grade menstruation may cause similar symptoms. T orsion or haemorrhage of an ovarian cyst can prove a di ffi cult di ff erential diagnosis. When suspected, pelvic ultrasound and a gynaecological opinion should be sought. It is unlikely that a ruptured ectopic pregnancy , with its well-defined signs of haemoperitoneum, will be mistaken for acute appendicitis, but the same cannot be said for a right- sided tubal abortion or, still more, for a right-sided unruptured tubal pregnancy . In the latter, the signs are very similar to those of acute appendicitis except that the pain commences on the right side and stays there. The pain is severe and continues unabated until operation. Usually , there is a history of a missed menstrual period, and a urinary pregnancy test may be pos - itive. Severe pain is felt when the cervix is moved on vaginal examination. Signs of intraperitoneal bleeding usually become apparent and the patient

should be questioned specifically regarding referr ed pain in the shoulder. Pelvic ultrasonography should be carried out in all cases in which an ectopic pregnancy is a possible diagnosis. In some patients with a long sigmoid loop, the colon lies to the right of the midline and it may be impossible to di ff er - entiate between diverticulitis and appendicitis. Abdominal CT scanning is particularly useful in this setting and should be considered in the management of all patients over the age of 60 years. Right-sided diverticulitis is more common in Asia and may be clinically indistinguishable from appendicitis. Abdominal CT scanning is particularly useful in making the distinction. As with left-sided diverticulitis, treatment should - be conservative with intravenous antibiotics with recourse to laparoscopy or laparotomy in the face of clinical deterioration. - The diagnosis of intestinal obstruction is usually - clear; the subtlety lies in recognising acute appendicitis as the occasional cause in the elderly . An abdominal CT scan will clarify the diagnosis. - When obstructed or locally perforated, carcinoma of the caecum may mimic or cause obstructive appendicitis in adults. A history of antecedent discomfort, altered bowel habit - or unexplained anaemia should raise suspicion. A mass may be palpable and an abdominal CT scan diagnostic. Rare differential diagnoses Preherpetic pain of the right 10th and 11th dorsal nerves is localised over the same area as that of appendicitis. It does - not shift and is associated with marked hyperaesthesia. There is no intestinal upset or rigidity . The herpetic eruption may be delayed for 3–8 hours. Spinal conditions are sometimes associated with acute abdominal pain, especially in children and the elderly . These - may include tuberculosis of the spine, metastatic carcinoma, osteoporotic vertebral collapse and multiple myeloma. The pain is due to compression of nerve roots and may be aggra - and vated by movement. There is rigidity of the lumbar spine and intestinal symptoms are absent.

Figure 76.7 First presentation in a 19-year-old man with terminal ile itis, later con /f_i rmed to be Crohn’s disease. Short arrow demonstrates abnormally thickened and in /f_l amed terminal ileum. Long arrow indi cates wall enhancement and

enlargement of the appendix, indicating secondary acute appendicitis (courtesy of Professor P MacMahon, FRCR, Dublin, Ireland).

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need to be remembered. A urinalysis should be undertaken in every abdominal emergency . In cyclical vomiting of infants or young children, there is a history of previous similar attacks and abdominal rigidity is absent. Acetone is found in the urine but is not diagnostic as it may accompany starvation. Typhlitis or leukaemic ileocaecal syndrome is a rare but potentially fatal enterocolitis occurring in immunosuppressed patients. Gram-negative or clostridial (especially Clostridium septicum) septicaemia can be rapidly progressive. Treatment is with appropriate antibiotics and haematopoietic factors. Sur gical intervention is rarely indicated.

Epithelial tumours of the appendix

Epithelial tumours of the appendix

Epithelial neoplasms are found in 0.6% of appendicectomy specimens. Numerous classification systems have been proposed, leading to much confusion and di ffi culty when comparing treatment modalities and outcomes. Following a modified Delphi consultation process, a group of interna tional experts proposed an updated classification system for appendiceal epithelial neoplasms. Tumours may be classified as mucinous or non-mucinous (intestinal type) and according architectural features to the degree of cytological atypia and (infiltrative versus pushing invasion) (Table 76.3).

FURTHER READING

FURTHER READING

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Goblet cell carcinoma

Goblet cell carcinoma

Goblet cell carcinomas (GCCs) of the appendix are a rare vari ant, accounting for less than 5% of appendix tumours. They display both neuroendocrine and glandular di ff erentiation and are not considered true NETs. They ma y be classified as typical GCC or adenocarcinoma ex-GCC (Tang classification), with the latter group carrying a worse prognosis. GCCs display higher grade features than typical NETs and have a greater propensity for nodal and peritoneal dissemination; they should be treated in a similar fashion to appendix adenocarcinoma.

Gross anatomy

Gross anatomy

The position of the base of the appendix is constant, being found at the confluence of the three taeniae coli of the caecum, which fuse to form the outer longitudinal muscle coat of the appendix. At operation, use can be made of this to find an elusive appendix, as gentle traction on the taeniae coli, particularly the anterior taenia, will lead the operator to the base of the appendix. The mesentery of the appendix or mesoappendix arises from the lower surface of the mesentery Nikolai Kulchitsky , 1856–1925, Professor of Histology , Kharkov , Ukraine, who left Russia after the Revolution of 1917 and later worked at University College, London, UK. He described these cells in 1897. Sometimes, as much as the distal one-third of the appendix is bereft of mesoappendix. Especially in childhood, the mesoappendix is so transparent that the contained blood vessels can be seen (Figure 76.3). In many adults, it becomes laden with fat, which obscures these vessels. The appendicular artery , a branch of the lower division of the ileocolic artery , passes behind the terminal ileum to enter the mesoappendix a short distance from the base of the appendix. It then comes to lie in the free border of the mesoappendix. An accessory appendicular artery may be present but, in most people, the appendicular artery is an ‘end-artery’, thrombosis of which results in necrosis of the appendix (synonym: gangrenous appendicitis). Four, six or more lymphatic channels traverse the mesoappendix to empty into the ileocaecal lymph nodes.

Figure 76.3 Laparoscopic view of a normal appendix with meso- appendix displaying the appendicular artery.

High-grade and invasive neoplasms

High-grade and invasive neoplasms

Appendiceal mucinous tumours displaying high-grade dysplasia are classified as high-grade appendiceal mucinous neoplasm (HAMN). Mucinous tumours with infiltrative inva - sion are classified as mucinous adenocarcinoma and may be well, moderately or poorly di ff erentiated with or without the presence of signet ring cells. Non-mucinous adenocarcinoma is considered to be similar to typical colorectal cancer. Patients with high-grade tumour or invasive adenocarci noma are at risk of lymph node involvement. According to current paradigms, they should undergo right hemicolectomy . Because of the risk of peritoneal metastases, cytoreductive sur gery (CRS), to include right hemicolectomy with regional (right parietal) peritonectomy , omentectomy and HIPEC, may also be appropriate when the necessary expertise and experience are available. Considera tion may also be given to performing bilateral salpingo-oophorectomy because of the risk of tumour seeding to the ovaries, although in patients of childbearing age the decision making is complex.

L S TC C Figure 76.17 Pseudomyxoma peritonei with characteristic omental cake (C) encasing the transverse colon (TC) and extending to the greater curvature of the stomach (S). Tumour is seen to replace the lesser omentum (L).

Introduction

INTRODUCTION

The importance of the vermiform appendix in surgery arises primarily from its propensity for inflammation, which results in the clinical syndrome known as acute appendicitis. Acute appendicitis is the most common cause of an ‘acute abdomen’ in young adults and, as such, the associated symptoms and signs have become a paradigm for clinical teaching. Appen dicitis is su ffi ciently common that appendicectomy (termed appendectomy in North America) is the most frequently performed urgent abdominal operation and is often the first major procedure perfor med by a surgeon in training. Advances in modern radiographic imaging have improved diagnostic accuracy; however, the diagnosis of appendicitis remains essentially clinical, requiring a mixture of observation, clinical acumen and surgical science and as such it remains an enigmatic challenge and a reminder of the art of surgical A wombat is a nocturnal, burrowing Australian marsupial. Robert E Condon , 1929–2015, Emeritus Professor of Surgery , Medical College of Wisconsin, WI, USA. diagnosis. Although much more uncommon, the appendix also has a propensity to the formation of tumours, which, despite humble and innocuous beginnings, may disseminate widely with dramatic clinical consequences. Aside from its tendency to cause surgical pathology , the appendix, long thought to be a vestigial organ, may also have important roles in both immune function and maintaining - the gut microbiota. The putative role of the appendix in the pathogenesis of ulcerativ e colitis (appendicectomy seems to be protective), for example, may be explained by its interaction with the intestinal flora and gut immune function.

Preileal 1% Retrocaecal 74% Postileal 0.5% Pa racaecal 2% Pe lvic 21% Subcaecal 1.5% Figure 76.1 The various positions of the appendix (after Sir C Wakeley, London). Evolving concepts in the management of acute • appendicitis Basic surgical techniques, both open and laparoscopic • The management of postoperative problems • Tumours of the appendix and pseudomyxoma peritonei •

Investigation

Investigation

The diagnosis of acute appendicitis is essentially clinical; however, a decision to operate based on clinical suspicion alone can lead to the removal of a normal appendix in 15–30% of cases. The premise that it is better to remove a normal appen dix than to delay diagnosis does not stand up to close scrutiny , particularly in the elderly . A number of clinical and laboratory- based scoring systems have been devised to assist diagnosis. The most widely used is the Alvarado score (Table 76.2 scor e of 7 or more is strongly predictive of acute appendicitis. In patients with an equivocal score (5 or 6), abdominal ultrasonography or contrast-enhanced CT examination fur ther reduces the rate of negative appendicectomy . Abdominal ultrasonography is more useful in children and thin adults ticularly if gynaecological pathology is suspected, with a nostic accuracy in excess of 90% (Figure 76.8). Modern CT is both sensitive and specific (approximately 95%) in the diagno sis of acute appendicitis (Figure 76.9) and worldwide there has been a steady increase in its use for this purpose . CT has been shown to reduce the rate of negative appendicectomy without an associated increased perforation ra te (due to delay in diag nosis) and may be cost-e ff ective as a result of shorter hospital stay . While the diagnostic accuracy of modern CT scanning for appendicitis is well established, radiation exposure and the theoretical carcinogenic e ff ect ar e a concern. Low-dose proto cols, which reduce the radiation dose to the patient by up to 80%, can be as reliable as standard dose scanning and may be more appropriately applied when considering a diagnosis of acute appendicitis, particularly in the younger adult. Contrast-enhanced standar d dose CT is especially useful in patients in whom there is diagnostic uncertainty , particularly older patients, where acute diverticulitis, intestinal obstruction and neoplasm are likely di ff erential diagnoses.

Investigations

Investigations

The investigation of a patient with PMP should include a high-resolution CT scan of the abdomen and pelvis with oral and intravenous contrast. A full colonoscopy should be performed to exclude a primary colorectal cancer. Laparos copy may provide additional staging information by allowing direct visualisation of the small bowel where radiologically occult miliary disease may be found. Ideally laparoscopy should be performed at a centre wher e subsequent CRS would be undertaken. Laparoscopy also facilitates tissue diagnosis, although this is not always necessary . In experienced centres the clinical and radiological features are often su ffi cient to establish a diagnosis of PMP .

Learning objectives

Learning objectives

To understand: The aetiology and surgical anatomy of acute appendicitis • The clinical signs and differential diagnoses of • appendicitis The investigation of suspected appendicitis •

Low-grade neoplasms

Low-grade neoplasms

Most epithelial tumours of the appendix are classified as LAMNs. These lesions demonstrate minimal cytological Laura H Tang , contemporary , pathologist, Memorial Sloan Kettering Hospital, New Y ork, NY , USA. atypia and are characterised by pushing rather than infil - trative invasion without evidence of destruction. In the case of appendix perforation, mucin may be found outside the appendix, the significance of which is increased when accom - panied by the presence of epithelial cells. Low-grade tumours do not typically metastasise to regional lymph nodes and as - such right hemicolectomy is not required. The importance of these lesions lies in their propensity to disseminate thr oughout the peritoneal cavity , causing the syndrome known as PMP . Patients with low-grade epithelial neoplasms and no evidence of mucin or epithelial cells beyond the appendix are thought to be at low risk of future PMP development. A colonoscopy should be performed to exclude associated colonic epithelial lesions and patients entered into a surveillance protocol for at least 5 years. Surveillance may take the form of clinical review , annual low-dose abdominopelvic CT scan and monitoring of appendix-related tumour markers (carcinoembryonic anti - gen [CEA], CA-19-9, CA-125). In patients with perforated tumours, particularly when extraluminal mucin contains epithelial cells, the risk of PMP is higher. In some experienced ed for cytoreductive centres these patients may be consider surgery with hyperthermic intraperitoneal chemotherapy - (HIPEC), although a common approach is regular monitoring for the appearance of clinical or radiological features of PMP , which would then warrant surgery .

Management of an incidental or

Management of an incidental or

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Microscopic anatomy

Microscopic anatomy

The appendix varies considerably in length and circumference. The average length is between 7.5 and 10 /uni00A0 cm. The lumen is irregular, being encroached on by multiple longitudinal folds of mucous membrane lined by columnar cell intestinal mucosa of colonic type (Figure 76.4). Crypts are present but are not numerous. In the base of the crypts lie argenta ffi n cells (Kulchitsky cells), which may give rise to neuroendocrine tumours (NETs) (see Neuroendocrine tumours of the appendix). The submucosa contains numerous lymphatic aggregations or follicles. While no discernible change in immune function results from appendicectomy , the prom - inence of lymphatic tissue in the appendix of young adults seems to be important in the aetiology of appendicitis (see Acute appendicitis).

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Figure 76.4 Normal vermiform appendix. The narrow lumen is bounded by mucosa, which may be arranged in folds. There is usu

ally abundant lymphoid tissue in the mucosa, especially in younger individuals. This may encroach on and further narrow the lumen. The mucosa is bounded by a relatively thin muscularis mucosa (courtesy of Dr P Kelly, FRCPath, Dublin, Ireland).

While there are isolated reports of perityphlitis (fatal inflam mation of the caecal region) from the late 1500s, recognition of acute appendicitis as a clinical entity is attributed to Reginald Fitz, who presented a paper to the first meeting of the Asso ciation of American Physicians in 1886 entitled ‘Perforating inflammation of the vermiform appendix’. Soon afterwards, Charles McBurney described the clinical manifestations of acute appendicitis, including the point of maximum tender ness in the right iliac fossa tha t now bears his name. The incidence of appendicitis seemed to rise greatly in the first half of the twentieth century , particularly in Europe, America and Australasia, with up to 16% of the population undergoing appendicectomy . In the past 30 years , the inci dence has fallen dramatically in these countries, such that the individual lifetime risk of appendicectomy is 8.6% and 6.7% among males and females, respectively . Acute appendicitis is relatively rare in infants and becomes increasingly common in childhood and early adult life, reach ing a peak

incidence in the teens and early tw enties. After mid dle age, the risk of developing appendicitis is quite small. The incidence of appendicitis is equal among males and females before puberty . In teenagers and young adults, the male-to female ratio increases to 3:2 at age 25; thereafter, the grea incidence in males declines.

NEOPLASMS OF THE APPENDIX AND PSEUDOMYXOMA PERITON

NEOPLASMS OF THE APPENDIX AND PSEUDOMYXOMA PERITONEI

Tumours of the appendix may occur in up to 0.97 per 100 /uni00A0 000 of the population. NETs account for approximately 30% of appendix neoplasms while epithelial tumours account for most other cases. Epithelial tumours may show mucinous or non-mucinous features and range in the aggressiveness of their behaviour from low grade to high-grade invasive with signet ring features (Table 76.3). A small percentage of appendix neoplasms fall into a third category known as mesenchymal tumours and include lymphoma, neuroma, GIST , Kaposi’s sarcoma and granular cell tumour of the appendix. Most patients with appendix neoplasms are asymptomatic at diagnosis and the appendix tumour is commonly an inci dental finding at appendicectomy . Perforation of a mucinous appendix tumour with dissemination of epithelial cells and mucin production leads to a condition known as pseudomyx oma peritonei (PMP) (Figure 76.15).

Neuroendocrine tumours of the appendix

Neuroendocrine tumours of the appendix

NETs of the appendix are slightly more common in females and have an overall incidence of 0.15–0.6 per 100 /uni00A0 000 per year. They arise in subepithelial neuroendocrine cells (Figure 76.16 and the majority (70%) are located in the appendix tip. The average age at presentation is 40–50 years and most patients are asymptomatic with early-stage disease typically found at appendicectomy for acute appendicitis. Uncommonly , patients may present with symptoms due to a mass or metastatic disease. Carcinoid syndrome is extremely rare. A diagnosis of NET is based on immunohistochemical staining for synaptophysin and chromogranin A and tumours are classified as grade 1–3 according to their proliferative capacity (determined by the Ki-67 index and mitotic rate). Treatment The treatment and prognosis of NETs of the appendix is governed by their grade, the tumour size and the extent of tumour invasion. Fully resected low-grade tumours less than 1 /uni00A0 cm in size with minimal serosal or mesoappendix invasion ustria. tologist, Vienna, A Moritz Kaposi , 1837–1902, der ma are considered fully treated by appendicectomy alone and no further treatment or follow-up is required. The optimum treatment of patients with tumours 1–2 /uni00A0 cm in size that have been fully resected is less clear as metastases may occur, albeit rarely . Current guidelines recommend a single CT or MRI of the abdomen to out rule regional or distant metastatic disease. Further surgery in the form of oncological resection of the right colon should be considered in patients with - larger tumours (>2 /uni00A0 cm), in the case of incomplete resection at appendicectomy or for higher tumour grade (2 or 3), T4 disease or vascular invasion. In these patients the risk of - regional lymph node involvement is increased; however, the potential benefit of further surgery must be weighed against the increased operative risk. No further follow-up is required when the completion hemicolectomy shows no evidence of residual disease. The presence of nodal disease or high-grade tumour mandates subsequent follow-up, typically with CT or)

Figure 76.15 A low-grade mucinous tumour of the appendix with mucinous ascites and low-volume pseudomyxoma peritonei. TABLE 76.3 Classi /f_i cation of epithelial neoplasia of the appendix. Adenoma (tubular, tubulovillous, villous) Serrated polyp Non-mucinous adenocarcinoma Mucinous neoplasm Low-grade appendiceal mucinous neoplasm High-grade appendiceal mucinous neoplasm Mucinous adenocarcinoma Adenocarcinoma with signet ring cells (<50%) Signet ring (>50%) carcinoma Adapted from Carr NJ, Cecil TD, Mohamed F et al . A consensus for classi /f_i cation and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia. The results of the Peritoneal Surface Oncology Group International (PSOGI) modi /f_i ed Delphi process. Am J Surg Pathol 2016; 40 : 14–26.

MRI at 6 and 12 months and then annually . For patients with early-stage NET the prognosis is excellent with 5-year survival of close to 100%. For those with advanced disease or distant metastases 5-year survival is typically less than 25%.

Figure 76.16 (a) Cross-sectional view of the appendix with outer, pink, muscularis propria (MP) and inner paler mucosa (Muc). The lumen of the /uni00A0 appendix (L) has been compressed by an adjacent well-differentiated neuroendocrine tumour (NET). Haematoxylin and eosin stain, × 20. (b) Higher power view of synaptophysin immunohistochemical stain showing characteristic positive staining (brown) within tumour cells. /uni00A0 Synaptophysin immunohistochemistry, × 100 (courtesy of Dr J Aird, FRCPath, Dublin, Ireland).

Pathology

Pathology

- Obstruction of the appendiceal lumen seems to be essential for appendiceal perforation. However, in many cases of early appendicitis, the appendix lumen is patent despite the presence - of mucosal inflammation and lymphoid hyperplasia. Occa - sional clustering of cases among children and young adults suggests an infective agent, possibly viral, which initiates an inflammatory response. Seasonal variation in the incidence is also observ ed, with more cases occurring between May and August in northern Europe than at other times of the year. Lymphoid hyperplasia narrows the lumen of the appen - - dix. Once obstruction occurs, continued mucus secretion and inflammatory exudation increase intraluminal pressure, obstructing lymphatic drainage. Oedema and mucosal cosa. Resolution may occur at this point either spontaneously or in response to antibiotic therapy . If the condition progresses, further distension of the appendix may cause venous obstruc tion and ischaemia of the appendix wall. With ischaemia, bacterial invasion occurs through the muscularis propria and submucosa, producing acute appendicitis (Figure 76.6 Finally , ischaemic necrosis of the appendix wall produces gangrenous appendicitis, with free bacterial contamination of the peritoneal cavity . Alternatively , the greater omentum and loops of small bowel become adherent to the inflamed appen dix, walling o ff the spread of peritoneal contamination and resulting in a phlegmonous mass or paracaecal abscess. Rarely , appendiceal inflammation resolves, leaving a distended mucus filled organ termed a mucocele of the appendix. Peritonitis occur s as a result of free migration of bacteria through an ischaemic appendicular wall, frank perforation of a gangrenous appendix or delayed perforation of an appendix abscess. Factors that promote this process include extremes of age, immunosuppression, diabetes mellitus, faecolith obstruc tion of the appendix lumen, a free-lying pelvic appendix and previous abdominal surgery that limits the ability of the greater omentum to wall o ff the spread of peritoneal contamination. In these situations, a rapidly deteriorating clinical course is accompanied by signs of di ff use peritonitis and systemic sepsis syndrome. Summary box 76.1 Risk factors for perforation of the appendix /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF John Benjamin Murphy , 1857–1916, Professor of Surgery , Northwestern University , Chicago, IL, USA. History - Appendicitis is relatively rare in infants under 36 months of age and, for obvious reasons, the patient is unable to give a history . Because of this, diagnosis is often delayed, and thus). the incidence of perforation and postoperative morbidity is considerably higher than in older children. In older age groups the classical features of acute appendi - citis begin with poorly localised colicky abdominal pain. This - is due to midgut visceral discomfort in response to appendi - ceal inflammation and obstruction. The pain is frequently first noticed in the periumbilical region and is similar to, but less - intense than, the colic of small bowel obstruction. Central abdominal pain is associated with anorexia, nausea and usually one or two e pisodes of vomiting that follow the onset of pain (Murphy). Anorexia is a useful

and constant clinical feature, particularly in children, who invariably also have vomiting. The patient often gives a history of similar discomfort that - settled spontaneously . A family history is also useful as up to one-third of children with appendicitis have a first-degree rela - tive with a similar history . In women of childbearing age pelvic disease can mimic acute appendicitis and a careful gynaecolog - ical history should be taken, concentrating on menstrual cycle, vaginal discharge and possible pregnancy . Summary box 76.2 Symptoms of appendicitis /uni25CF /uni25CF /uni25CF /uni25CF

Extremes of age Faecolith obstruction Immunosuppression Pelvic appendix Diabetes mellitus Previous abdominal surgery (a) Figure 76.6 Acutely in /f_l amed appendix with purulent exudate extending to the mesoappendix in a 28-year-old man as seen at laparoscopy and a photomicrograph (original magni /f_i cation × 20) (b) from the same patient showing the appendix with pus- /f_i lled lumen (L) and in /f_l ammation extending to in /f_l amed serosa (S) (courtesy of Professor C O’Keane, FFPath, FRCPI, Dublin, Ireland). Periumbilical colic Anorexia Pain shifting to the right iliac Nausea fossa (b) S L (a)

etal peritoneum in the right iliac fossa becomes irritated, pro ducing more intense, constant and localised somatic pain that begins to predominate. Patients often report this as an abdom inal pain that has shifted and changed in character. Typically , coughing or sudden movement exacerba tes the right iliac fossa pain. The classic visceral–somatic sequence of pain is present in only about half of those pa tients subsequently proven to have acute appendicitis. A typical presentation includes pain that is predominantly somatic or visceral and poorly localised. Atyp ical pain is more common in the elder ly , in whom localisation to the right iliac fossa is unusual. An inflamed appendix in the pelvis may not produce somatic pain involving the anterior abdominal wall, but instead cause suprapubic discomfort and tenesmus. In this circumstance, tenderness may be elicited only on rectal e xamination and is the basis for the recommendation that a rectal examination should be performed on every patient who presents with acute lower abdominal pain. During the first 6 hours, there is rarely any alteration in temperature or pulse rate. After that time, slight pyrexia (37.2–37.7°C) with a corresponding increase in the pulse rate to 80–90 beats per minute is usual. However, in 20% of patients there is no pyrexia or tachycardia in the early stages. In children, a temperature greater than 38.5°C suggests other causes (e.g. mesenteric adenitis; see Di ff erential diagnosis, Children). Typically , two clinical syndromes of acute appendicitis can be discerned: acute catarrhal (non-obstructive) appendicitis and acute obstructive appendicitis, the latter characterised by a more acute course. The onset of symptoms is abrupt and there may be generalised abdominal pain from the start. The temperature may be normal and vomiting is common, so the clinical picture may mimic acute intestinal obstruction. Signs The diagnosis of appendicitis rests more on thorough clinical examination of the abdomen than on any aspect of the history or laboratory investigation. The cardinal features are those of an unwell patient with low-grade pyrexia, localised abdom inal tenderness, muscle guarding and rebound tenderness. Inspection of the abdomen may show limitation of respiratory movement in the lower abdomen. The patient is then asked to point to where the pain began and w here it moved (pointing sign). Gentle superficial palpation of the abdomen, beginning in the left iliac fossa and moving anticlockwise to the right iliac Summary box 76.3 Clinical signs in appendicitis /uni25CF /uni25CF /uni25CF /uni25CF Neils Thorkild Rovsing , 1862–1937, Professor of Surgery , Copenhagen, Denmark. Sir Vincent Zachary Cope , 1881–1975, surgeon, St Mary’s Hospital, London, UK James Douglas , 1715–1742, anatomist and midwife who practised in London, UK, described this pouch in

1730. tenderness, classically McBurney’s point. Asking the patient to - cough or gentle percussion over the site of maximum tender - ness will elicit rebound tenderness (see Chapter 63). - Deep palpation of the left iliac fossa may cause pain in the right iliac fossa, Rovsing’s sign, which is helpful in supporting a clinical diagnosis of appendicitis. Occasionally , an inflamed appendix lies on the psoas muscle, and the patient, often a young adult, will lie with the right hip flexed for pain relief (the psoas sign). Spasm of the obturator internus is sometimes - demonstrable when the hip is flexed and internally rotated. If an inflamed appendix is in contact with the obturator internus, this manoeuvre will cause pain in the hypogastrium (the obtu - rator test; Zachary Cope). Cutaneous hyperaesthesia may be demonstrable in the right iliac fossa, but is rarely of diagnostic value. Summary box 76.4 Signs to elicit in appendicitis /uni25CF /uni25CF /uni25CF /uni25CF Special features, according to position of the appendix Retrocaecal Rigidity is often absent, and even application of deep pressure may fail to elicit tenderness (silent appendix), the reason being that the caecum, distended with gas, prevents the pressure exerted by the hand from reaching the inflamed structure. However, deep tenderness is often present in the loin, and rigidity of the quadratus lumborum may be in evidence. Psoas spasm, due to the inflamed appendix being in contact with that muscle, may be su ffi cient to cause flexion of the hip joint. Hyperextension of the hip joint may induce abdominal pain when the degree of psoas spasm is insu ffi cient to cause flexion - of the hip. Pelvic Occasionally , early diarrhoea results from an inflamed appen - dix being in contact with the rectum. When the appendix lies entirely within the pelvis, there is usually complete absence of abdominal rigidity , and often tenderness over McBurney’s point is also lacking. In some instances, deep tenderness can be made out just above and to the right of the symphysis pubis. In either event, a rectal examination reveals tenderness in the rectovesical pouch or the pouch of Douglas, especially on the right side. Spasm of the psoas and obturator internus muscles may be present when the appendix is in this position. An inflamed appendix in contact with the bladder may cause frequency of micturition. This is more common in children.

Pyrexia Localised tenderness in the right iliac fossa Muscle guarding Rebound tenderness Pointing sign Psoas sign Rovsing’s sign Obturator sign

Postileal In this case, the inflamed appendix lies behind the terminal ileum. It presents the greatest di ffi culty in diagnosis because the pain may not shift, diarrhoea is a feature and marked retching may occur. Tenderness, if any , is ill defined, although it may be present immediately to the right of the umbilicus.

Children Adult Gastroenteritis Regional enteritis Mesenteric adenitis Ureteric colic Meckel’s diverticulitis Perforated peptic ulcer Intussusception Torsion of testis Henoch–Schönlein purpura Pancreatitis Lobar pneumonia Rectus sheath haematoma

Postoperative complications

Postoperative complications

Postoperative complications following appendicectomy are relatively uncommon and reflect the degree of peritonitis that was present at the time of operation and intercurrent diseases that may predispose to complications. Harrith Hasson , 1931–2012, Professor of Gynecology , Chicago, IL, USA. Summary box 76.7 Checklist for unwell patients following appendicectomy /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Wound infection Wound infection is the most common postoperative complica - tion, occurring in 5–10% of all patients. This usually presents with pain and erythema of the wound on the fourth or fifth

(d) (b) The appendicular artery, ligated with clips, (d) Appendicectomy complete. Examine the wound and abdomen for an abscess Consider a pelvic abscess and perform a rectal examination Examine the lungs – consider pneumonitis or collapse Examine the legs – consider venous thrombosis Examine the conjunctivae for an icteric tinge and the liver for enlargement, and enquire whether the patient has had rigors (pylephlebitis) Examine the urine for organisms (pyelonephritis) Suspect subphrenic abscess

postoperative day , often soon after hospital discharge. Treat ment is by wound drainage and antibiotics when required. The organisms responsible are usually a mixture of Gram-negative bacilli and anaerobic bacteria, predominantly Bacteroides species and anaerobic streptococci. Intra-abdominal abscess Approximately 8% of patients following appendicectomy will develop a postoperative intra-abdominal abscess. In an era of hospital discharge 24–48 hours following appendi cectomy , patients should be advised prior to discharge that a spiking fever, malaise and anorexia developing 5–7 days after operation is sug gestive of an intraperitoneal collection and that urgent medical advice should be obtained. Interloop, paracolic, pelvic and subphrenic sites should be considered. Abdominal ultrasonography and CT scanning greatly facilitate diagnosis and allow percutaneous drainage (Figure 76.14). Surgical exploration should be considered in patients suspected of having intra-abdominal sepsis but in whom imaging fails to show a collection, particularly those with continuing ileus. Ileus A period of adynamic ileus is to be expected after appendicec tomy , and this may last a number of days following removal of a gangrenous appendix. Ileus persisting for more than 4 or 5 days, particularly in the presence of a fever, is indicative of contin uing intra-abdominal sepsis and should prompt further investigation. Rarely , early during postoperative recovery , a Richter’s type of hernia may occur at the site of a laparoscopic port insertion and may be confused with a postoperative ileus. A CT scan is usually definitive. August Gottlieb Richter , 1742–1812, lecturer in surgery , Göttingen, Germany . - Respiratory In the absence of concurrent pulmonary disease, respiratory complications are rare following appendicectomy . Adequate postoperative analgesia and physiotherapy , when appropriate, reduce the incidence. Venous thrombosis and embolism These conditions are rare after appendicectomy . Patients should undergo preoperative assessment of risk factors for - venous thromboembolism and appropriate prophylactic measures should be taken. Portal pyaemia

(pylephlebitis) This is a rare but very serious complication of gangrenous appendicitis associated with high fever, rigors and jaundice. It is caused by septicaemia in the portal venous system and leads to the development of intrahepatic abscesses (often multiple). Treatment is with systemic antibiotics and percutaneous drain - age of hepatic abscesses as appropriate. Faecal fistula Leakage from the appendicular stump occurs rarely , but may follow if the encircling stitch has been put in too deeply or - if the caecal wall was involved by oedema or inflammation. Occasionally , a fistula may result following appendicectomy in Crohn’s disease. Adhesive intestinal obstruction This is the most common late complication of appendicec - tomy . At operation, a single band adhesion is often found to be responsible. Occasionally , chronic pain in the right iliac fossa is attributed to adhesion formation after appendicectomy . In such cases, laparoscop y is of value in confirming the presence of adhesions and allowing division.

Figure 76.14 (a) Rim-enhancing collection in the right iliac fossa, 1 week after open appendicectomy for perforated appendicitis. radiological drainage with resolution of the abscess (courtesy of Professor P MacMahon, FRCR, Dublin, Ireland). (b) Successful

Rarely , inflammation of the appendix may present as a chronic condition characterised by recurrent episodes of lower abdominal pain. Recurrent appendicitis is thought to arise as a consequence of incomplete self-limiting obstruction of the appendix lumen. The attacks vary in intensity and may occur every few months, and the majority of cases ultimately culminate in severe acute appendicitis. If a careful history is taken from patients with acute appendicitis, many remember having had milder but similar attacks of pain. The appendix in these cases is thickened and shows fibrosis indicative of previous inflammation.

Pseudomyxoma peritonei

Pseudomyxoma peritonei

PMP is a rare condition typified by progressive peritoneal tumour deposits, mucinous ascites, omental cake (76.17) and ovarian involvement in females. The vast majority of cases arise as a result of perforation of a mucinous appen diceal tumour. Patients typically present with progressive and massive abdominal distension, anor exia and symptoms of bowel dysfunction. The condition is invariably fatal without intervention. Traditionally , PMP was thought to have an incidence of 1 per 1 /uni00A0 000 /uni00A0 000 per year, but it is now thought to be at least double that with recent estimates of 3.2 cases per 1 /uni00A0 000 /uni00A0 000 per year. The overall risk of developing pseudomyx oma following removal of an appendix harbouring epithelial tumour is approximately 9%, with the risk varying according to the tumour subtype and the mode of presentation, while it may be as high as 30–50% in the case of a mucinous adeno carcinoma of the appendix (Figur e 76.18). PMP is classified according to the degree of cytological atypia within the peritoneal deposits (Table 76.4) and its grading may di ff er from that of the causative primary appendiceal tumour. Elevated tumour mar kers (CEA, CA-125, CA-19-9) - - Figure - - -

(b) Figure 76.18 (a) Contrast-enhanced axial computed tomography (CT) image demonstrates a tubular cystic structure with calci /f_i cation adjacent to the caecum compatible with an abnormally distended appendix (arrow). (b) Six-year follow-up postcontrast axial CT image demonstrated a 19 × 10 × 17 cm complex cystic mass in the right lower quadrant (arrow) highly suspicious for a mucinous tumour of the appendix with extensive peritoneal involvement and pseudomyxoma peritonei (courtesy of Professor Helen Fenlon, Dublin, Ireland). TABLE 76.4 Classi /f_i cation of pseudomyxoma peritonei. Acellular mucin Low-grade mucinous carcinoma peritonei High-grade mucinous carcinoma peritonei High-grade mucinous carcinoma peritonei with signet ring cells Adapted from Carr NJ, Cecil TD, Mohamed F et al . A consensus for classi /f_i cation and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia. The results of the Peritoneal Surface Oncology Group International (PSOGI) modi /f_i ed Delphi process. Am J Surg Pathol 2016; 40 : 14–26.

predict a more aggressive phenotype and are associated with a worse prognosis.

P T Figure 76.19 Hyperthermic intraperitoneal chemotherapy (HIPEC) delivery following cytoreductive surgery using a closed abdomen technique. The perfusion pump (P) heats and circulates chemother apy throughout the abdominal cavity via in /f_l ow and out /f_l ow tubing (T), typically for 60–90 minutes.

Treatment

Treatment

Non-operative management There are two scenarios in which patients with acute appendi citis may be considered for non-operative treatment. Uncomplicated appendicitis While surgery remains the standard teaching, there is evidence to support a trial of conservative management in patients Alfredo Alvarado , contemporary , surgeon, Plantation, FL, USA. Preoperative investigations in appendicitis /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF - /uni25CF /uni25CF /uni25CF -). A - , par - diag - - - - -

Routine Full blood count Urinalysis Selective Pregnancy test Urea and electrolytes C-reactive protein Supine abdominal radiograph Ultrasound of the abdomen/pelvis Contrast-enhanced abdomen and pelvic CT scan (consider low-dose protocol in young adults) TABLE 76.2 The Alvarado (MANTRELS) score. Score Symptoms M igratory

RIF pain 1 A norexia 1 N ausea and vomiting 1 Signs T enderness (RIF) 2 R ebound tenderness 1 E levated temperature 1 Laboratory L eukocytosis 2 S hift to left 1 Total 10 RIF , right iliac fossa. Figure 76.8 Ultrasound image of the right iliac fossa (RIF) demonstrat

ing a mildly enlarged appendix, measuring 8 mm in diameter, consis

tent with acute appendicitis in a 40-year-old man. Arrow indicates a small pocket of free /f_l uid more inferiorly in the RIF (courtesy of Dr D Byrne, Dublin, Ireland).

with uncomplicated (absence of appendicolith, perforation or abscess) appendicitis. Treatment is bowel rest and intra venous antibiotics, often metronidazole and a third-generation cephalosporin. The available data indicate initial successful outcomes in approximately 85% of pa tients; however, between one-quarter and one-third of patients initially treated conservatively will require surgery within 1 year for recurrent symptoms. Subsequent surgery , if needed, tends to be uncomplicated and the overall postoperative complication rate is similar when patients treated conservatively and later needing surgery are compared with those undergoing surgery at the outset. Overall hospital length of stay is also similar when comparing patients in each group. Thus, antibiotic treatment of acute uncomplicated appendicitis appears to be safe and it allows a large number of patients to

avoid invasive treatment; however, this information must be balanced by the high treatment failure rate and need for interval intervention. Conservative treatment may be considered in the well patient with limited signs or those with high operative risk (multiple comorbidities); the patients must be aware of the high failure rate. As with conservative treatment of an appendix Albert John Ochsner , 1858–1925, Professor of Clinical Surgery , The University of Illinois College of Medicine, Chicago, IL, USA. James Sherren , 1876–1945, surgeon, The London Hospital, London, UK. ensure that there is no underlying malignancy (see Neoplasms of the appendix and pseudomyxoma peritonei). In children, conservative treatment of uncomplicated appendicitis also appears to be safe in the short term with r es - olution of acute symptoms in approximately 90% of patients. Re-presentation with complicated appendicitis appears to be ver y rare; however, recurrent symptoms requiring sur gery have been reported in up to 46% of patients. Notably , histological features of acute appendicitis may be present in only one-fifth of patients needing interval surgery . Appendix mass If an appendix mass is present and the condition of the patient is satisfactory , the standard treatment is the conserva - tive Ochsner–Sherren regime. This strategy is based on the premise that the inflammatory process is already localised and that inadvertent surgery is di ffi cult and may be dangerous. It may be impossible to find the appendix and, occasionally , a faecal fistula may form. For these reasons, it is wise to observe a non-operative programme but to be prepared to operate should clinical deterioration occur. Summary box 76.6 Criteria for stopping conservative treatment of an appen - dix mass /uni25CF /uni25CF /uni25CF Careful recording of the patient’s condition and the extent of the mass should be made and the abdomen regularly re - examined. It is helpful to mark the limits of the mass on the abdominal wall using a skin pencil. A contrast-enhanced CT - examination of the abdomen should be performed and anti - biotic therapy instigated. An abscess, if pr esent, should be drained radiologically . Temperature and pulse rate should be recorded 4-hourly and a fluid balance record maintained. Clinical deterioration or evidence of peritonitis is an indication for early laparotom y . Clinical improvement is usually evident within 24–48 hours. Failure of the mass to resolve should raise suspicion of a carcinoma or Crohn’s disease. Using this regime, approximately 90% of cases resolve without incident. The need for interval appendicectomy in this cohort is much debated. The majority of patients will not develop recur - rent appendicitis; however, recently published studies have identified higher than expected rates of underlying appen - diceal neoplasm in those patients who do go on to interval appendicectomy , particularly those patients over the age of 40. A recent randomised clinical trial was termina ted early owing to the unexpected finding of appendiceal tumour during follow-up in 29% (12/41) of pa tients aged more than 40 /uni00A0 years who initially presented with periappendicular abscess. Low-grade appendiceal mucinous neoplasms (LAMNs)

Figure 76.9 Sagittal reformat of a computed tomography scan of the abdomen obtained with oral and

intravenous contrast, demonstrating an enlarged (10 mm), enhancing retrocaecal appendix with periappen diceal fat stranding. There is no evidence of necrosis, perforation or collection. No radiopaque appendicolith can be seen. Figure 76.6 refers to the same patient (courtesy of Professor P MacMahon, FRCR, Dublin, Ireland). A rising pulse rate Increasing or spreading abdominal pain

Increasing size of the mass

incidental appendix tumour in patients presenting with abscess appears to be much higher than in the general population undergoing appendicectomy , suggesting a di ff erent patho genesis in the former group. Careful consideration should be given to interval appendicectomy in this cohort of patients. At the very least, follow-up CT or magnetic r esonance imaging (MRI) should be performed to ensure complete resolution of findings and patients should undergo colonoscopy . Operative management General principles The traditional treatment for acute appendicitis is appendicec

tomy . While there should be no unnecessary delay , all patients, particularly those most at risk of serious morbidity , benefit by a short period of intensive preoperative preparation. Intravenous fluids, su ffi cient to establish adequa te urine output (catheterisa tion is needed only in the very ill), and appropriate antibiotics should be given. Risk factors for venous thromboembolism should be considered and appropriate prophylaxis (mechanical and/or phar macological) initiated. There is evidence that, in the absence of purulent peritonitis, a single perioperative dose of antibiotics reduces the incidence of postoperative wound infection. When peritonitis is suspected, therapeutic intravenous antibiotics to cover Gram-negative bacilli as well as anaerobic cocci should be given. Hyperpyrexia in children should be treated with salicylates in addition to antibiotics and intravenous fluids. With appropriate use of intravenous fluids and parenteral antibiotics, a policy of deferring appendicec tomy after midnight to the first case on the following morning does not increase morbidity . However, when acute obstructive appendicitis is recognised, operation should not be deferred longer than it takes to optimise the patient’s condition. Gridiron , a frame of crossbeams to support a ship during repairs. Lewis Linn McArthur , 1858–1934, surgeon, St. Luke’s Hospital, Chicago, IL, USA. James Rutherford Morison , 1853–1939, Professor of Surgery , Durham University , Durham, UK. Otto Lanz , 1865–1935, surgeon, Amsterdam, The Netherlands. anaesthetic with the patient supine on the operating table and may be undertaken using either an open or laparoscopic - approach. When the appropriate equipment and expertise are available and cost allows, the laparoscopic appr oach is advantageous. The initial laparoscopy allows the diagnosis to be established and may reduce the negative appendicectomy rate. Furthermore, the patient may benefit from the quicker recovery a ff orded by a minimally invasive approach, the rate of wound infection is lower (when compared with open sur - gery) and, contrary to initial concerns, the incidence of post - operative pelvic collection does not appear to be increased. When a laparoscopic technique is used, the bladder must - be empty (ensure that the patient has voided before leaving the ward). Prior to preparing the entire abdomen with an appropri - ate antise ptic solution, the right iliac fossa should be palpated for a mass. If a mass is felt, it may , on occasion, be preferable - to adopt a conservative approach. Draping of the abdomen is in accordance with the planned operative technique , taking account of any requirement to extend the incision or convert a laparoscopic technique to an open operation. Surgical technique: conventional appendicectomy When the preoperative diagnosis is considered reasonably certain, the incision that is widely used for appendicectomy is the so-called gridiron incision. The gridiron incision (described first by McArthur) is made at right angles to a line joining the anterior superior iliac spine to the umbilicus, its centre being along the line at McBurney’s point (Figure 76.10). If better - access is required, it is possible to convert the gridiron to a Rutherford Morison incision (see below) by cutting the internal oblique and transversus muscles in the line of the incision. In recent years, a transverse skin crease (Lanz) incision has become more popular, as the exposure is better and extension,

2 / 3 1 / 3 Figure 76.10 Gridiron incision for appendicitis, at right angles to a line joining the anterior superior iliac spine and umbilicus, centred on McBurney’s point (courtesy of Professor M Earley, FRCSI, Dublin, Ireland). Figure 76.11 Transverse or skin crease (Lanz) incision for appendicitis, 2 cm below the umbilicus, centred on the midclavicular–midinguinal line (courtesy of Professor M Earley, FRSCI, Dublin, Ireland).

the size and obesity of the patient, is made approximately 2 /uni00A0 cm below the umbilicus centred on the midclavicular–midinguinal line (Figure 76.11). When necessary , the incision may

be extended medially , with retraction or suitable division of the rectus abdominis muscle. When the diagnosis is in doubt, particularly in the presence of intestinal obstruction, a lower midline abdominal incision is to be preferred over a right lower paramedian incision. The lat ter, although widely practised in the past, is di ffi cult to extend, more di ffi cult to close and provides poorer access to the pelvis and peritoneal cavity . Rutherf ord Morison's incision is useful if the appendix is para- or retrocaecal and fixed. It is essentially an oblique muscle-cutting incision with its lower end over McBurney's point and extending obliquely upwards and laterally as neces sary . All layers are divided in the line of the incision. The caecum is identified by the presence of taeniae coli and, using a finger or a swab, the caecum is withdrawn. A turgid appendix may be felt at the base of the caecum. Inflammatory adhesions must be gently broken with a finger, which is then hooked around the appendix to deliver it into the wound. T William Wayne Babcock , 1876–1963, surgeon, Philadelphia, PA, USA. Sir William Arbuthnot Lane , 1856–1943, surgeon, Guy's Hospital, London, UK forceps applied in such a way as to encircle the appendix and yet not damage it. The base of the mesoappendix is clamped in artery forceps, divided and liga ted (Figure 76.12a). When the mesoappendix is broad, the procedure must be repeated with a second or, rarely , a third artery forceps. The appendix, now completely freed, is clamped with an artery forceps near its base and then ligated close to the junction with the caecum using an - absorbable 2/0 ligature. The appendix is amputated between the artery forceps and the ligature (Figure 76.12b). When the appendix is retrocaecal and adherent, it may be an advantage to divide the base first. The appendiceal vessels are then ligated and gentle traction on the caecum will enable the surgeon to deliver the body of the appendix, which is then removed from base to tip. Occasionally , this manoeuvre requires division of - the lateral peritoneal attachments of the caecum. An absorbable 2/0 or 3/0 purse-string or 'Z' suture may then be inserted into the caecum about 1.25 /uni00A0 cm from the base (Figure 76.12c). The stitch should pass thr ough the muscle coat, picking up the taeniae coli. The stump of the appendix is invaginated (Figure 76.12d) while the purse-string or 'Z' he suture is tied, thus burying the appendix stump.

(a) (c) Figure 76.12 Appendicectomy. (a) The mesoappendix divided between artery forceps and ligated. about to be divided. (c) 'Z' suture inserted prior to inversion of the appendiceal stump. been tied. (b) (d) (b) The appendix is ligated at its base and (d) The appendiceal stump inverted, the 'Z' suture having

suture is in danger of cutting out. If the oedema is of lim ited extent, this can be overcome by inserting the purse-string suture into more healthy caecal wall at a greater distance from the base of the appendix. Occasions may arise when, because of the extensive oedema of the caecal wall, it is better not to attempt invagination. Many surgeons believe invagination of the appendiceal stump is unnecessary . Should the base of the appendix be gangrenous, ligation should not be attempted. Two stitches are placed through the caecal wall close to the base of the gangrenous appendix, which is amputated flush with the caecal wall, after which these stitches are tied. Further closure is e ff ected by means of a sec ond layer of interrupted seromuscular sutures. An alternative but more costly option when the appendix base is compro mised is to resect the appendix with a cu ff of healthy caecum using a sing le firing of a linear stapling device. Surgical technique: laparoscopic appendicectomy The most valuable aspect of laparoscopy in the management of suspected appendicitis is as a diagnostic tool, particularly in women of childbearing age. The placement of operating ports may vary according to operator preference and previous abdominal scars. Typically , a pneumoperitoneum is established using an open infraumbilical approach. This umbilical port

serves as the camera port with two working ports inserted under direct vision, the first suprapubically and the second in the left lower quadrant. A moderate Trendelenburg tilt with elevation of the right side of the operating table improves exposure and assists delivery of loops of small bowel from the pelvis. The appendix is found in the conventional manner by identification of the caecal taeniae and is controlled using a laparoscopic tissue-holding forceps. Occasionally , it is necessary to divide the peritoneal attachments and mobilise the caecum in order to adequately expose the appendix. By elevating the appendix, the mesoappendix is then displayed. A dissecting forceps, hook or scissors diathermy is used to dissect the mesoappendix (Figure 76.13a) and expose the appendicular vessels, which may be coagulated or ligated using a clip applicator (Figure 76.13b). The appendix, free of its mesentery , can be ligated at its base with an absorbable loop ligature (Figure 76.13c linear stapling device, divided (Figure 76.13d) and removed in a specimen bag through one of the operating ports. It is not usual to invert the stump of the appendix. Absorbable sutures are used to close the fascia at the umbilicus and at any port sites greater than 5 /uni00A0 mm, and the small skin incisions may be closed with subcuticular sutures. Problems encountered during appendicectomy The finding of a normal appendix demands careful exclusion of other possible diagnoses, particularly terminal ileitis, Meck el’s diverticulitis and tubal or ovarian causes in women. It is usual to remove the appendix to avoid future diagnostic di ffi culties, even though the appendix is macroscopically normal, particularly if a skin crease or gridiron incision has been /uni00A0 Friedrich Trendelenburg , 1844–1924, Professor of Surgery successively at Rostock (1875–1882), Bonn (1882–1895), Leipzig (1895–1911), Germany . The Tren delenburg position was first described in 1885. - appendix seen at diagnostic laparoscopy , although approxi - mately one-quarter of seemingly normal appendices show microscopic evidence of inflammation. If the appendix cannot be found, the caecum should be mobilised and the taeniae coli should be traced to their conflu - ence on the caecum before the diagnosis of ‘absent appendix’ is made. If an appendix mass is found at operation, particularly at laparoscopy , it may be safer to abandon the procedure rather than risk bowel injury during attempted mobilisation. Any abscess should be drained, intra venous antibiotics administered - and the patient carefully monitored during the postoperative period. V ery rarely in the face of a frankly necrotic appendix, - a caecectomy or partial right hemicolectomy is required. Occasionally , a patient undergoing surgery for acute appendicitis is found to have concomitant Crohn’s disease of the ileocaecal region. Providing that the caecal wall is healthy at the base of the appendix, appendicectomy can be per - formed without increasing the risk of an enterocutaneous fis - tula. Rarely , the appendix is involved with the Crohn’s disease. In this situation, a conservative approach may be warranted; a trial of intravenous corticosteroids and systemic antibiotics can be used to resolve the acute inflammatory process . Treatment

Patients with PMP should be referred to a specialist centre with multidisciplinary expertise in the assessment and management of patients with peritoneal malignancy . The accepted treatment is CRS combined with HIPEC (Sugarbaker). This approach combines multiple peritonectomy procedures with multivisceral resections as required to achieve a complete surgical clearance of the tumour (complete cytoreduction), which is augmented by HIPEC (typically mitomycin C or oxaliplatin) to eradicate presumed residual microscopic disease (Figure 76.19). The combined operation can take in excess of 10 hours and may require total abdominopelvic peritonectomy , greater and lesser Paul H Sugarbaker , contemporary , surgeon, Washington DC, USA. Brendan J Moran , contemporary , surgeon, Basingstoke, UK. cholecystectomy , splenectomy , partial gastrectomy , colectomy and anterior resection of the rectum. The largest re ported CRS/HIPEC for

PMP comes from Basingstoke, UK series of (Moran). In their experience involving more than 1000 patients, a complete cytoreduction was achieved in approximately 75% of patients, with the remainder undergoing maximal tumour debulking. Although a potentially morbid procedure, in experienced centres the operative mortality rate following CRS/HIPEC is less than 2% with major postoperative morbidity in 15% of patients. Appropriate patient selection is critical and patients must have a su ffi cient performance status and be without major comorbidity in order to withstand the surgery . Preoperative evaluation, including nutritional assessment and optimisation, is paramount. Following a complete cytoreduction 5- and 10-year survival rates of 87% and 70%, respectively , can be achieved. Poorer outcomes are seen in males, patients with elevated tumour marker s and following resection of tumour showing high- grade or invasive features. Follow-up typically comprises at least annual clinical evaluation, monitoring of tumour markers and CT scan. Surveillance should be continued for at least 10 years as late recurrence is documented. Systemic chemotherapy may be considered as first-line treatment in patients with high-grade or invasive unresectable disease or in the adjuvant setting following CRS/HIPEC, ag ain in patients with high-grade tumour. Chemotherapy is not typically considered in patients with low-grade PMP .

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Unexpected appendix tumour encountered at surgery

Unexpected appendix tumour encountered at surgery

An inspection of the abdominal cavity should be performed to establish the presence of metastases or PMP . The appendix should be removed, with care taken to avoid spillage of its contents, to allow a pathological diagnosis, which will deter - mine the need for subsequent intervention. In most cases the tumour can be removed by appendicectomy alone, with or without a cu ff of caecum. encountered at surgery The surgeon should perform a careful inspection of the abdominal cavity and record the extent and distribution of disease. A peritoneal or omental biopsy can be performed; however, care should be taken to minimise disruption of the anatomical planes. If the appendix is abnormal, an appendi cectomy should be performed to allow a histological diagnosis. Ideally the planned procedure should then be aborted with a view to referring the patient to a centre experienced in the management of peritoneal malignancy .

unexpected tumour of the appendix Incidental neopl

unexpected tumour of the appendix Incidental neoplasm in appendicectomy specimen

In a patient with an incidental finding of an appendix neoplasm and no current evidence of metastatic disease or PMP , subsequent treatment is dependent on the degree of cytological atypia within the primary tumour, the estimated risk of lymph node metastases and the future risk of developing PMP . In general, small low-grade tumours require surveillance only , whereas patients with larger tumour or adverse features may need further surgery .