Hx Ex Ix Rx

SURGICAL CASES EXPLAINED

CLINICAL INTEGRATION

SURGERY

Chee, Raz & Arachchi

- Hx why are certain features key to making a diagnosis?
- Ex what do these features tell us?
- Ix what are the best tests to order?
- Rx what are the best management options?

CLINICAL INTEGRATION:

SURGERY

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Hx Ex Ix Rx

CLINICAL INTEGRATION:

SURGERY

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PREFACE

Following on from the success of *Clinical Integration: Medicine*, we, the editors, felt there was a gap in resources when it came to clinically applicable surgical cases for medical students. From my experiences going through medical school, studying for the clinical exams (or OSCEs) was certainly more difficult when the topic was surgical, especially in the more niche specialties such as ENT, Plastics and Paediatric Surgery. We aim for *Clinical Integration: Surgery* to be used as a tool to prepare for medical school clinical exams, especially in the final years where information synthesis and the management of certain conditions become important.

The book is a compilation of case studies that have been written by doctors who are either training or have an interest in that specialty. A history, examination, investigations and management section are provided, with certain key words being highlighted for further discussion. These points of discussion are intended to prompt readers to understand why the case was written in such a way and deepen their knowledge of the case diagnosis.

Clinical Integration: Surgery can be used in many ways. It can be read from front to back as a study tool to learn about different surgical topics and their work-up and management. It can also be used as a testing tool, where the reader could review the case history and examination to formulate a list of differential diagnoses, and then review the investigations to identify the true diagnosis and propose a management plan to compare to what is provided.

We hope that this book is a pleasure to read as much as it was to write.

Samuel Chee
Manda Raz
Asiri Arachchi

ABBREVIATIONS

AAA	abdominal aortic aneurysm
AAD	acute aortic dissection
ABCDE	airway, breathing, circulation, disability and exposure
ABG	arterial blood gas
ABI	ankle–brachial index
AC	alternating current
ACDF	anterior cervical discectomy and fusion
ACE	angiotensin-converting enzyme
ACL	anterior cruciate ligament
ACS	acute coronary syndrome
ACTH	adrenocorticotrophic hormone
ADLs	activities of daily living
ADPKD	autosomal dominant polycystic kidney disease
AFP	alpha-fetoprotein
AJCC	American Joint Committee on Cancer
AKI	acute kidney injury
ALP	alkaline phosphatase
ALT	alanine aminotransferase
AMP	adenosine monophosphate
AOM	acute otitis media
AP	anteroposterior
aPTT	activated partial thromboplastin time
ARDS	acute respiratory distress syndrome
ASIA	American Spinal Injury Association
ASOT	antistreptolysin O titre
AVM	arteriovenous malformation
AVPU	Alert; responds to Voice, responds to Pain, Unresponsive
BAL	blood alcohol level
BAV	bicuspid aortic valve
BCC	basal cell carcinoma
BGL	blood glucose level
BMD	bone mineral density
BMI	body mass index
BP	blood pressure
BPH	benign prostatic hyperplasia
bpm	beats per minute
BTM	biodegradable temporising matrix
CABG	coronary artery bypass graft
CAP	community-acquired pneumonia
CCB	calcium channel blocker
CCP	cyclic citrullinated peptide
CEA	carcinoembryonic antigen
CES	cauda equina syndrome
CK	creatine kinase
CKD	chronic kidney disease
CMP	calcium, magnesium and phosphate
CMV	cytomegalovirus
CN	cranial nerve
CNS	central nervous system
COPD	chronic obstructive pulmonary disease
CPA	cerebellopontine angle
CPB	cardiopulmonary bypass
CPR	cardiopulmonary resuscitation
CRP	C-reactive protein
CSF	cerebrospinal fluid
CSL	compound sodium lactate
CSM	cervical spondylotic myelopathy
CT	computed tomography
CTA	CT angiography
CTB	CT brain
CUS	compression ultrasonography
CVS	cerebral vasospasm
CXR	chest X-ray
dADLs	domestic activities of daily living
DC	direct current
DCM	degenerative cervical myelopathy
DHCA	deep hypothermic circulatory arrest
DIC	disseminated intravascular coagulation
DOAC	direct oral anticoagulant
DRE	digital rectal examination
DVT	deep vein thrombosis
EBUS	endobronchial ultrasound
EBV	Epstein–Barr virus
ECG	electrocardiography

ED	Emergency Department
EDH	extradural haemorrhage
eGFR	estimated glomerular filtration rate
EMG	electromyography
ENT	ear, nose and throat
ERCP	endoscopic retrograde cholangiopancreatography
ESR	erythrocyte sedimentation rate
ETT	endotracheal tube
EUA	examination under anaesthesia
EVAR	endovascular aneurysm repair
EVD	external ventricular drain
EVLT	endovenous laser therapy
FAST	focused assessment with sonography in trauma
FBC	full blood count
FBE	full blood examination
FDG	fluorodeoxyglucose
FDP	flexor digitorum profundus
FDS	flexor digitorum superficialis
FESS	functional endoscopic sinus surgery
FISH	fluorescence *in situ* hybridisation
FNA	fine needle aspirate/aspiration
GABHS	group A beta-haemolytic streptococcus
GBM	glioblastoma multiforme
GCS	Glasgow Coma Scale
GGT	gamma-glutamyl transpeptidase
GI	gastrointestinal
GORD	gastro-oesophageal reflux disease
GP	general practitioner
GTN	glyceryl trinitrate
HAP	hospital-acquired pneumonia
Hb	haemoglobin
hCG	human chorionic gonadotrophin
Hct	haematocrit
HDL	high density lipoprotein
HHT	hereditary haemorrhagic telangiectasia
HIT	heparin-induced thrombocytopenia
HIV	human immunodeficiency virus
HLM	heart-lung machine
HOCM	hypertrophic obstructive cardiomyopathy
HPOA	hypertrophic pulmonary osteoarthropathy
HPV	human papillomavirus
HR	heart rate
HTM	heart team meeting
HTN	hypertension
ICC	intercostal catheter
ICH	intracranial haemorrhage
ICP	intracranial pressure
ICU	intensive care unit
IDC	indwelling urinary catheter
IDH	isocitrate dehydrogenase
IE	infective endocarditis
Ig	immunoglobulin
IHD	ischaemic heart disease
IM	intramuscular
INR	international normalised ratio
IPJ	interphalangeal joint
IV	intravenous
IVC	inferior vena cava
IVDU	intravenous drug use(r)
IVP	intravenous pyelogram
JVP	jugular venous pressure
KUB	kidneys, ureter, bladder
LAD	left anterior descending
LCL	lateral collateral ligament
LDH	lactate dehydrogenase
LDL	low density lipoprotein
LFT	liver function test
LITA	left internal thoracic artery
LL	lower limb
LMCA	left main coronary artery
LMN	lower motor neurone
LMWH	low molecular weight heparin
LOC	loss of consciousness
LV	left ventricular
LVH	left ventricular hypertrophy
MCL	medial collateral ligament
MCPJ	metacarpophalangeal joint
MC&S	microscopy, culture and sensitivities
MDT	multidisciplinary team
MEN	multiple endocrine neoplasia

MGMT	methyl guanine methyl transferase
MI	myocardial infarction
mJOA	modified Japanese Orthopaedic Association
MMA	middle meningeal artery
MMSE	mini mental state examination
MOCA	Montreal Cognitive Assessment
MR	mitral regurgitation
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
MRSA	methicillin-resistant *Staphylococcus aureus*
MS	multiple sclerosis
MUA	manipulation under anaesthesia
MVA	motor vehicle accident
NAI	non-accidental injury
NEC	necrotising enterocolitis
NF	neurofibromatosis
NGT	nasogastric tube
NIHSS	National Institutes of Health Stroke Scale
NOF	neck of femur fracture
NSAID	non-steroidal anti-inflammatory drug
NSCLC	non-small cell lung cancer
OA	osteoarthritis
OME	otitis media with effusion
OP-CAB	off-pump CABG
OPG	orthopantomogram
ORIF	open reduction, internal fixation
PAD	peripheral arterial disease
pADLs	personal activities of daily living
PCA	patient-controlled analgesia
PCI	percutaneous coronary intervention
PCKD	polycystic kidney disease
PCL	posterior cruciate ligament
PCR	polymerase chain reaction
PDA	patent ductus arteriosus
PE	pulmonary embolism
PET	positron emission tomography
PFT	pulmonary function test
PICC	peripherally inserted central catheter
PIPJ	proximal interphalangeal joint
PND	paroxysmal nocturnal dyspnoea
PNET	primitive neuroectodermal tumour
PR	per rectum
PSA	prostate-specific antigen
PT	prothrombin time
PTH	parathyroid hormone
PTS	post-thrombotic syndrome
PTX	pneumothorax
PVD	peripheral vascular disease
RA	rheumatoid arthritis
RadA	radial arteries
RCA	right coronary artery
RCC	renal cell carcinoma
RHD	rheumatic heart disease
ROM	range of motion
RR	respiratory rate
RSI	rapid sequence induction
RTx	radiotherapy
RUQ	right upper quadrant
RVA	radiofrequency vein ablation
SABR	stereotactic ablative body radiotherapy
SAH	subarachnoid haemorrhage
SAM	systolic anterior motion
SBP	systolic blood pressure
SCC	squamous cell carcinoma
SCLC	small cell lung cancer
SCM	sternocleidomastoid
SGLT2	sodium-glucose co-transporter-2
SIADH	syndrome of inappropriate antidiuretic hormone secretion
SIRS	systemic inflammatory response syndrome
SLE	systemic lupus erythematosus
SLNB	sentinel lymph node biopsy
SLT	straight leg test
SOB	shortness of breath
SPECT	single-photon emission CT
SPF	sun protection factor
STI	sexually transmitted infection
SVC	superior vena cava
SVG	great saphenous vein
T2DM	type 2 diabetes mellitus
TAI	traumatic aortic injury

TAVR	transcatheter aortic valve replacement
TB	tuberculosis
TBI	traumatic brain injury
TBSA	total body surface area
TEVAR	thoracic endovascular aortic repair
TFT	thyroid function test
TIA	transient ischaemic attack
TNM	tumour, node, metastasis
TOE	transoesophageal echocardiogram
TSH	thyroid-stimulating hormone
TTE	transthoracic echocardiography
TTF	thyroid transcription factor
TURP	transurethral resection of the prostate
UEC	urea, electrolytes and creatinine
UFH	unfractionated heparin
UL	upper limbs
UMN	upper motor neurone
URTI	upper respiratory tract infection
UTI	urinary tract infection
UV	ultraviolet
VA	visual acuity
VATS	video-assisted thoracoscopic surgery
VBG	venous blood gas
VSD	ventricular septal defect
VTE	venous thromboembolism
WBC	white blood cell
WCC	white cell count

CASE 58: Neonatal bowel atresias

History

- A **36-hour-old** [1] **neonate** [2] is transferred from a rural hospital to a tertiary centre with **repeated non-bilious vomiting** [3].
- They were **born at term** [4] via a normal vaginal delivery.
- They have not **passed meconium** [5].
- They have not had any **fevers** [6].
- There is **no significant family history** [7].
- The patient's mother did not take any **medications or illicit drugs** [8] or **smoke** [9] during her pregnancy.
- The patient's mother reports her **antenatal testing** [10] was normal but she did have **polyhydramnios** [11] on her antenatal ultrasounds.
- The mother states she also underwent **maternal screening tests** [12] for congenital abnormalities.

1 An intestinal atresia is a congenital defect along the GI tract resulting in complete or partial obstruction of the lumen of the bowel. It can occur anywhere from oesophagus to rectum. Vomiting due to small bowel intestinal atresia will begin 24–48 hours after birth.

2 The term 'neonate' encompasses any newborn in their first 28 days of life.

3 In duodenal atresia the vomiting may or may not be bilious, depending on whether or not the defect is proximal or distal to the hepatopancreatic ampulla. For jejunal, ileal and colonic atresia, the vomiting is always bilious. Intestinal atresias range in incidence from approximately 1.3–3.5 per 10 000 live births. Of these, the most common is duodenal atresia, making up 60% of all atresias. This is followed by jejunal and ileal atresias, representing 20% of all atresias, while colonic atresia contributes <10% of all atresias.

4 The majority of neonates with intestinal atresias are born at term or near term. Term refers to a gestation of 37–41 weeks. A gestation <37 weeks is referred to as preterm and a gestation >41 weeks is referred to as post-term.

5 While it is more common for a neonate to not pass meconium after birth, it is still possible for infants with intestinal atresia to pass meconium despite their bowel obstruction. This is more commonly seen in more proximal obstructions, such as duodenal atresia, but can also occur with ileal or jejunal atresia if meconium is present in the bowel distal to the obstruction. Neonates with colonic atresia are unlikely to pass meconium.

6 Fevers may indicate sepsis as a possible alternative cause of the patient's symptoms. Sepsis could also develop from an aspiration pneumonia caused by the repeated vomiting.

7 Familial genetic factors do not play a large role in the development of intestinal atresias in the foetus. However, there have been some cases of familial atresia syndromes, as well as newborns being more likely to have atresias in families with known thrombophilia. A family history of cystic fibrosis may also point towards the same diagnosis in the neonate. During foetal development, thick inspissated meconium may cause a segmental volvulus affecting the mesenteric vascular supply of the developing bowel, causing failure of that portion of the bowel to develop. Most jejunal and ileal atresias are acquired from impaired vascular supply.

8 The use of vasoconstrictive medications such as pseudoephedrine found in over-the-counter decongestants or methylenedioxymethamphetamine (MDMA) triples the risk of intestinal atresia in the newborn. It is thought this could be due to interruption of mesenteric vessel blood flow during development. Significant vomiting is also a feature of neonatal abstinence syndrome in neonates whose mothers used opioids during the pregnancy.

9 Similar to vasoconstrictive medications, smoking in the first trimester of pregnancy also triples the risk of formation of intestinal atresias in the foetus, thought to be by a similar mechanism of interrupting blood flow.

10 Antenatal testing and screening are a routine part of antenatal care for an expectant mother to ensure a safe pregnancy and delivery for mother and baby, and to try to pre-empt and correct any issues early should they arise. Antenatal testing involves regular reviews with GPs, midwives and obstetrician gynaecologists. During these reviews, general physical check-ups including assessment of height, weight and BP for the mother, as well as fundal height measurements for an estimation of the development of the foetus, are performed. The mother will also be referred for antenatal ultrasound scans for monitoring of foetal morphology and growth as well as observation of amniotic fluid volume and blood flow in the foetus and placenta. Urine samples are taken to look for evidence of infection or

pre-eclampsia. Glucose tolerance testing is performed to identify early evidence of gestational diabetes mellitus. Blood tests are carried out, including for mother's blood group and antibodies, haemoglobin levels, vitamin D levels, and to identify the presence of or immunity to various infections that could be passed on to the foetus or neonate by vertical transmission. This includes rubella, varicella, CMV, parvovirus, herpes simplex virus, toxoplasmosis, hepatitis B and C, syphilis and HIV. The above collection of conditions may lead to infant morbidity and mortality through a variety of means including various neonatal infections leading to sepsis, congenital abnormalities, rashes, intrauterine growth restriction and developmental delays. A swab of the vagina late in pregnancy assesses for the presence of Group B streptococcus. Group B streptococcus can be a cause of early-onset sepsis in the newborn, and a positive finding of Group B streptococcus on swab or urine microscopy and culture means a woman should receive antibiotic coverage during labour to prevent infecting her newborn during delivery. For an unwell, vomiting neonate, sepsis as a cause of their symptoms must always be excluded.

11 Antenatal ultrasounds have a low sensitivity for detecting GI atresias and fewer than half of neonates with an atresia will be picked up on routine screening. During the third trimester, there may be a high suspicion for the neonate having duodenal atresia if a dilated, fluid-filled stomach and proximal duodenum are visualised on ultrasound. They appear like large 'bubbles', hence the term 'double bubble' sign. In up to 50% of cases of duodenal atresia polyhydramnios is present on antenatal ultrasounds, usually developing well into the second trimester. Jejunal and ileal atresias also may have findings of polyhydramnios on antenatal ultrasounds, as well as findings of ascites and dilated bowel loops.

12 Maternal serum screening tests are blood tests available to women during their pregnancy to help identify the likelihood of their unborn baby having Down syndrome (trisomy 21), Edwards syndrome (trisomy 18) or a neural tube defect. Small bowel intestinal atresias can be associated with chromosomal abnormalities or other congenital anomalies. Fewer than 5% of neonates with Down syndrome will have duodenal atresia, but they represent 30% of total cases. Duodenal atresia may also be seen as part of the VACTERL association of congenital anomalies. VACTERL association refers to a constellation of non-syndromic congenital abnormalities which frequently occur in conjunction with each other. It incorporates Vertebral anomalies, Anal atresia, Cardiac Anomalies, Tracheo-oEophageal fistula, Renal anomalies and Limb anomalies. Additionally, other congenital abnormalities not covered in the above list have also been seen with VACTERL association, including ear anomalies, laryngomalacia, laryngeal or tracheal stenosis, ambiguous genitalia, omphalocele, single umbilical artery and intestinal malrotation. As VACTERL is an association and there is no identified genetic cause, it cannot be tested for with any currently available maternal screening tests. Some features of VACTERL may be identified antenatally on ultrasounds when looking at foetal morphology. With this in mind, isolated duodenal atresia is present in up to half of all cases without any other anatomical or karyotypical abnormalities. In contrast, only 5% of cases of jejunal and ileal atresia are associated with chromosomal abnormalities. Jejunal atresia may be seen with cardiac or other GI malformations. Ileal atresia and colonic atresia are less commonly associated with other major anatomical malformations.

Examination

- The neonate appears **flat and unwell** [1].
- The **heart rate is 190bpm** [2].
- The **respiratory rate is 60 breaths per minute with mild work of breathing** [3] evident. **Oxygen saturations on air are 95%** [4].
- The **temperature is 37°C** [5].
- The neonate has **sunken fontanelles and a sluggish capillary refill time** [6].
- Their **skin is mildly tinged yellow** [7].
- Their **heart sounds are dual and there are no murmurs** [8].
- **Chest is clear** [9] to auscultation with no added sounds and good air entry throughout.
- Upon general inspection of the abdomen, there is **no evidence of an abdominal wall defect** [10].
- The **abdomen is mildly distended** [11].
- The abdomen is soft to palpation with **no guarding or rigidity and the neonate tolerates the examination** [12] of the abdomen well.
- The patient's **nappy is dry** [13].
- The **anus is patent and in a normal position** [14].
- The patient does not have any **facial or form features associated with genetic or congenital anomalies** [15].

1 Due to repeated vomiting the neonate is unable to sufficiently absorb any fluids such as breast milk or formula that they have been taking orally, and thus the neonate may present in hypovolaemic shock. Repeated vomiting may also be a sign of sepsis, which is a critical differential to consider in an unwell neonate. Bilious, but occasionally non-bilious vomiting is also part of the presentation of intestinal malrotation with volvulus.

2 Normal heart rate for a term neonate is 110–170bpm. A tachycardia may be present as a response to hypovolaemia, sepsis, pain or distress.

3 Normal respiratory rate for a term neonate is 30–60 breaths per minute. Respiratory compromise may be evident due to abdominal distension causing diaphragmatic elevation that is preventing adequate chest expansion. Respiratory changes may also be present in the setting of an aspiration pneumonia caused by the repeated bouts of vomiting. Increased respiratory rate may also be seen with dehydration and a subsequent metabolic acidosis.

4 In term neonates, oxygen saturations should be ≥95%. If saturations are <95% further investigation is warranted, for example looking for signs of sepsis, an airway or respiratory pathology or a cardiac pathology.

5 A fever is not usually seen in an isolated presentation of a small bowel intestinal atresia. However, a fever may be present in other diagnoses such as sepsis or aspiration pneumonia.

6 Neonates are particularly susceptible to rapid volume depletion as a consequence of vomiting, so a careful fluid balance assessment should be performed. Other signs to identify are evidence of oliguria or anuria, poor skin turgor, dry mucous membranes and altered conscious level.

7 Jaundice may be present in presentations of intestinal atresia, as intestinal obstruction increases the distribution of bilirubin in the enterohepatic circulation. Jaundice may also be intensified in the setting of associated volume depletion.

8 A full assessment of the neonate should be performed, particularly when the presentation is undifferentiated. Cardiac examination may identify added sounds or murmurs suggestive of a congenital heart defect which may be associated with a small bowel atresia. Atrioventricular septal defects are the most common cardiac defect seen in patients with Down syndrome, and may be found along with an intestinal atresia in these patients.

9 Lung field auscultation may indicate signs of pneumonia, which may have developed due to aspiration following repeated vomiting.

10 A finding of an abdominal wall defect on abdominal inspection may indicate the presence of other GI abnormalities. Most abdominal wall defects will be picked up on antenatal ultrasounds from 10 weeks' gestation. There are two main types of abdominal wall defects – omphalocele and gastroschisis. Omphalocele, also known as exomphalos, is where internal abdominal organs – most frequently the stomach, liver and bowel – protrude through an opening where the umbilical cord joins the abdomen. This results in the organs being enveloped in a membranous sac of peritoneum. Omphalocele is seen in only 2.5 per 10 000 births and is largely associated with genetic and chromosomal abnormalities and congenital cardiac defects. The abdominal wall defect in gastroschisis is not associated with the umbilical cord and small and large bowel may protrude through the abdominal wall without a membranous covering, exposing them to the surrounding amniotic fluid. Gastroschisis is seen in 2–6 per 10 000 births. It is rarely associated with genetic and chromosomal abnormalities and less commonly with other congenital defects. 10% of cases of gastroschisis are associated with a small bowel atresia. During development, the extracorporeal intestine may volve or be compressed at the opening in the anterior abdominal wall. Due to the impaired blood supply, the affected bowel will undergo necrosis and subsequently be reabsorbed, causing an atresia.

11 Abdominal distension is a frequent finding in presentations of intestinal atresia. The more distal the atresia, often the greater the level of abdominal distension. Visible or palpable loops of bowel may also be present. In cases of colonic atresia, symptoms and examination findings may present later than in more proximal atresias.

12 The abdomen should be assessed for evidence of peritonism, which may be present in the setting of a bowel perforation or intra-abdominal cause of sepsis. Assessing for the presence of abdominal tenderness in a neonate and infants can be challenging, so paying close attention to their behaviour during palpation, palpating when they are settled and performing serial examinations may be required for an accurate assessment.

13 The neonate's nappy should be inspected to assess for any passed meconium, and as part of a fluid assessment to ensure urine is still being produced. Haematochezia may be visible in an infant's nappy in cases of intestinal malrotation with volvulus, indicating bowel ischaemia and necrosis.

14 Anal atresia can be associated with more proximal GI atresias as well as part of VACTERL associations. Therefore, assessment of anus position and patency should be performed.

15 A full top-to-toe assessment of the child should be completed, looking for physical indications of chromosomal anomalies or congenital malformations. In an acute presentation this assessment can be delayed until the neonate is stabilised. External features that may be present in neonates with Down syndrome include small low-set ears, brachycephaly, epicanthal folds, a low nasal bridge with a short nose and small nostrils, a small down-turned mouth, single palmar creases and a wide space between the first and second toes. Other structural malformations to identify, which may or may not be evident on physical examination, include cardiac, other GI, renal and skeletal abnormalities. Physical features that may be observed clinically in VACTERL association include anal atresia, cleft palate and limb anomalies such as radial aplasia or hypoplasia, polydactyly, syndactyly and hypoplastic thumb, club foot and tibial hypoplasia.

Investigations

- **Urinalysis** [1] is unremarkable.
- A **full blood examination** [2] is normal.
- A VBG demonstrates a **metabolic acidosis** [3] and **urea, electrolytes and creatinine** [4] show electrolyte disturbances.
- There is no growth seen on **blood microscopy and culture** [5].
- **Coagulation studies** [6] are normal.
- The neonate's **blood type** [7] is O+.
- Liver function testing shows a raised **bilirubin** [8] level.
- An **abdominal plain film** [9] is performed, which reveals dilatation of the stomach and proximal duodenum and an absence of distal gas.
- There is no **free air** [10].
- An **upper GI contrast study** [11] is performed which correlates the findings of the abdominal plain film.
- A **contrast enema** [12] is not required.
- A **CXR, renal ultrasound and echocardiogram** [13] are all normal.
- **Genetic screening** [14] is performed.

1 A urinalysis is a quick and simple test that can be done to screen for signs of a urinary tract infection. A formal microscopy and culture should also be sent, as part of a septic work-up for an unwell neonate presenting with vomiting.

2 An FBC may reveal an elevated WCC in the setting of infection and sepsis. Haemoglobin and platelet counts should also be on hand prior to proceeding to operative management.

3 A metabolic acidosis may develop in the setting of severe dehydration due to ongoing losses from vomiting.

4 Deranged electrolytes may develop due to ongoing losses from vomiting, as well as due to altered volume status.

5 If sepsis is suspected, a blood culture should be obtained as part of a septic work-up. Results of a blood culture will take >24 hours to return, so if sepsis is suspected antibiotics should be commenced immediately following collection of blood samples. Antibiotics should be rationalised after results of the culture and sensitivities are known.

6 Coagulation studies should be performed as part of a preoperative work-up to evaluate potential coagulopathies.

7 Blood type and available compatible blood on hand should be organised preoperatively, particularly if the neonate is proceeding to invasive abdominal surgery, as risk of blood loss and consequential need for blood transfusion are high.

8 Bilirubin levels will be elevated in intestinal atresia as bilirubin is unable to be faecally excreted and therefore remains in enterohepatic circulation. A bilirubin panel will show increased levels of unconjugated bilirubin. Raised bilirubin levels may also be seen in sepsis or in biliary atresia, another GI malformation that may be present with intestinal atresia, among many other pathologies causing raised bilirubin.

9 Abdominal X-rays should be ordered in all suspected cases of neonatal bowel obstruction. The findings on plain film can be diagnostic in some cases. A classical finding on plain film is the 'double bubble' sign. Combined with an absence of distal bowel gas, this appearance is strongly suggestive of duodenal atresia. The double bubble sign may also be seen with other pathologies causing high grade duodenal obstruction. Intestinal malrotation may also present with a similar-appearing double bubble sign. Rare causes of neonatal bowel obstruction, including duplication cysts and duodenal webs, can in some cases also present with similar abdominal radiograph findings. Dilated small bowel loops with air–fluid levels is suggestive of a more distal bowel obstruction, such as a jejunal or ileal atresia.

10 A finding of free air, or pneumoperitoneum, on plain abdominal radiograph indicates intestinal perforation, a surgical emergency requiring urgent treatment and management.

11 An upper GI contrast study is the next investigation to proceed with if plain film confirms suspicions of obstruction, providing there is no evidence of pneumoperitoneum. A duodenal atresia would demonstrate similar findings on contrast study to those seen on radiography, with a dilated stomach and proximal duodenum and pooling of contrast in those areas and lack of contrast movement into the more distal small bowel. An upper GI contrast study is important to differentiate between a small bowel atresia and an intestinal malrotation with volvulus. Signs of malrotation on contrast study include a displaced duodenojejunal junction and coiling of the bowel. Malrotation occurs during embryonic development due to interruption of the normal rotation of the embryonic gut. This leads to the eventual midgut being supported on a narrow vascular pedicle instead of the wide-based mesentery that is normally seen, increasing the risk of volvulus. The development of volvulus is a surgical emergency as it leads to small bowel ischaemia, necrosis and intestinal perforation causing peritonitis. Presentation is often within the first year of life, but it can be diagnosed at any age. More than half of cases are associated with other congenital anomalies.

12 If a diagnosis has not been reached from both the plain film and upper GI contrast study, a contrast enema study is the next modality used to identify the cause of the bowel obstruction. Often, a microcolon may be present in cases of small bowel atresia or meconium ileus, due to lack of use of this section of the GI tract. A distal small bowel atresia is confirmed by the inability for contrast to move into the dilated loops of more proximal small bowel. Meconium ileus, an obstruction of the distal ileum due to thick, impacted meconium, may be confirmed by intraluminal filling defects on the contrast enema. The contrast enema may also be a therapeutic study, with meconium being passed post the enema in approximately 60% of studies. Meconium ileus is commonly seen in neonates with cystic fibrosis. Colonic atresia may be identified on contrast enema with limited to no filling with contrast of more proximal sections of colon with a distal microcolon. A contrast enema is also included as part of the work-up for Hirschsprung disease. Colonic atresia and Hirschsprung disease may occur together.

13 If a finding of a small bowel atresia is confirmed, particularly if it is a duodenal atresia, the neonate should be evaluated for any evidence of other congenital anomalies. This includes anomalies associated with Down syndrome, particularly if a concurrent diagnosis of Down syndrome is suspected or confirmed, most importantly being any evidence of cardiac anomalies. An echocardiogram should be performed to identify any possible cardiac malformations. It is essential that this is done prior to proceeding to operative management for a small bowel atresia. Further imaging is required to identify any other anomalies that may not be immediately apparent or that have not revealed themselves so far.

14 To confirm a clinical suspicion of Down syndrome, FISH (fluorescence *in situ* hybridisation) and formal karyotype testing should be ordered. For neonates with a small bowel atresia and particularly if they also had meconium plugs, cystic fibrosis mutation testing should be performed. A microarray to look further into the neonate's genetic make-up will also be performed in certain cases to identify any underlying genetic disorders leading to congenital abnormalities. VACTERL association is not associated with an underlying genetic defect and its occurrence is thought to be sporadic.

Management

Immediate

Gain **IV access** [1]. **Stop feeding** [2]. **Insert an NGT** [3]. **Correct fluid and electrolyte balance** [4]. Monitor **vital signs** [5] and **urine output** [6]. Commence broad-spectrum **antibiotics** [7]. **Refer** [8] to the paediatric general surgical team.

Short-term

Once the neonate is **stable** [9], proceed to **operative management** [10] of the atresia. Postoperatively, the neonate should be **monitored closely** [11] in the neonatal ICU. Commence **feeding** [12] 3–5 days postoperatively.

Long-term

Once the neonate has been established on full feeds, is making good weight gains, and has been fully investigated for any other congenital malformations, they are suitable for **discharge home** [13]. They should continue to be **reviewed** [14] in the community by their GP and maternal and child health nurse and should be reviewed in the **paediatric surgical outpatient clinic** [15].

1 It is important to establish IV access early. It will be an essential route for fluids and medications since the neonate is not able to have anything orally.

2 The neonate must be made nil by mouth until definitive management of the atresia has been undertaken. This will also reduce the risk of further vomiting, in turn reducing the chance of the neonate developing an aspiration pneumonia.

3 A nasogastric or orogastric tube should be inserted to decompress the stomach. It should be left on free drainage with regular aspirates.

4 It is important to ensure optimisation of fluid and electrolyte status before proceeding to operative management. It is likely that the neonate will be hypovolaemic from repeated vomiting and failure to absorb anything enterally and in turn will have altered electrolytes and acid–base status.

5 Continuous monitoring of vital signs helps guide response to treatment, ensures the patient remains stable while preparing for surgery and can identify any new issues that may arise and need to be addressed. These may include a new fever or change in oxygen saturations, which could indicate the development of an aspiration pneumonia from the repeated vomiting.

6 Monitoring of urine output is important as part of ongoing fluid status assessments, particularly when replacing losses and trying to ensure the patient remains euvolaemic.

7 Antibiotics may be commenced, particularly if surgery is imminent, to reduce the risk of postoperative infection. They are continued for several days postoperatively as well.

8 Neonates presenting with a small bowel obstruction should be managed in a tertiary centre in a neonatal intensive care unit under joint care of the neonatologists and the paediatric surgical team.

9 If an acute surgical emergency, such as a malrotation with volvulus, has been excluded on imaging, then surgery for correction of the atresia can be scheduled once the neonate has been optimised for surgical intervention. This includes adequate fluid and metabolic status and investigation for any other present congenital abnormalities. There are risks associated with waiting longer for surgical correction, however, including further vomiting leading to aspiration and potential sepsis; and complications of extended use of total parenteral nutrition, such as oral aversion, hyperlipidaemia, bone demineralisation and rebound hypoglycaemia.

10 A transverse right upper quadrant incision is made to access the duodenum in cases of duodenal atresia. Duodenal atresia is corrected by forming a duodeno-duodenostomy, either by a side-to-side or end-to-side anastomosis. Jejunal and ileal atresias are approached via a para- or transumbilical incision and are repaired primarily, usually with an end-to-end anastomosis. During the correction of atresias, the remaining bowel must be checked for the presence of other, previously unidentified, atresias. Primary repair is preferred when resecting multiple atresias, and care is taken to preserve adequate length of bowel to prevent short-gut syndrome – ideally >75cm, but some children may tolerate much less. While repairing colonic atresia, biopsies of the bowel wall should be taken intraoperatively to evaluate for evidence of Hirschsprung disease. Usually, the proximal end of the colon is externalised as a colostomy and anastomosis of the colon is delayed for several months. Babies tolerate colostomies well and this delayed anastomosis allows time for the dilated proximal colon to be decompressed and recover its tone, as well as allow time for biopsy results to demonstrate whether Hirschsprung disease is present.

11 Postoperatively, neonates should remain nil by mouth and continue with IV hydration and parenteral nutrition. The nasogastric or orogastric tube should also remain *in situ* for ongoing decompression. This allows for ongoing bowel rest in the initial days postoperatively. The neonate should be observed for any potential postoperative complications, e.g. peritonitis from an anastomotic leak or iatrogenic bowel injury.

12 Feeding can be reintroduced 3–5 days postoperatively and slowly increased until full oral feeds are established. Parenteral nutrition should be gradually weaned down as feeding is re-established.

13 Most neonates do well after repair of an intestinal atresia. Morbidity and mortality in these infants are more commonly attributable to concurrent issues, such as prematurity or other congenital anomalies, or the presence of multiple atresias or short-gut syndrome. If the neonate has required formation of a colostomy, they should be linked in with stomal therapy or hospital in the home to provide information and guidance for the care of colostomies in the community.

14 Neonates with small bowel atresias should have good follow-up in the community to ensure ongoing good feeding and weight gain is being maintained.

15 Regular review in the paediatric outpatient clinic is important, again to ensure good feeding and weight gain is being maintained. Surgical wounds should be reviewed to ensure adequate healing has occurred and there is no evidence of dehiscence or associated infection. The neonate should also be reviewed for the potential for late postoperative complications, such as anastomotic dysfunction or delayed emptying, or stricture or adhesion formation causing further full or partial obstructions. Reoperation should be considered if obstruction is confirmed on imaging, but return to theatre must be at least 3 weeks following the initial operation date.

CASE 59: Ovarian torsion

History

- A **15-year-old female** [1] with **no significant past medical history** [2] presents with a **4-hour history** [3] of **acute left-sided abdominal pain** [4].
- She has been having **waves of nausea associated with the pain and has vomited** [5] a few times.
- She had been **sitting** [6] in school when the pain came on. She has not had any **fevers** [7] at home.
- She has not had changes with **urination** [8].
- Her last **menstrual period finished a week ago and she is not sexually active** [9]. She has not had any abnormal **vaginal bleeding** [10].
- She has **no regular medications and no allergies** [11].

1 Ovarian torsion is an acute surgical emergency. It can affect women of any age from the neonatal period to postmenopausal but is most likely to affect women of reproductive age. Most paediatric general surgeons will manage ovarian torsion up until the age of 16, after which the adult obstetrics and gynaecology team will guide management. The ovary is suspended by the infundibulopelvic ligament (also known as the suspensory ligament of the ovary) which is a fold of the broad ligament. It is through the infundibulopelvic ligament that the ovarian vessels travel to reach the ovary. Due to this mobile nature, the ovary may sit laterally or posteriorly to the uterus depending on the position of the patient. It is on these ligamentous supports that the ovary can twist, obstructing its own blood supply. Often the Fallopian tube will twist with the ovary if this occurs.

2 Ovarian torsion is much more likely to occur due to the presence of an ovarian mass or cyst. The asymmetric size and weight of an affected ovary predisposes it to twist on its ligamentous supports. Masses or cysts >6cm in size are most likely to precipitate torsion. Approximately 40% of cases of ovarian torsion are due to ovarian neoplasms, the most common of which are benign, such as teratomas and adenomas. Up to 2% of ovarian torsions in adults are due to malignant neoplasms, but this percentage is much lower in children. Another 40% of cases of ovarian torsion are due to the presence of ovarian cysts. In the paediatric population, however, half the presentations of ovarian torsion are in females with normal ovaries, particularly if they are pre-menarchal. It is important to obtain a full past history from the patient to identify if there are any acute or chronic health problems that must be considered should the patient require surgery or to help refine the list of differentials for the presentation, such as if the patient has previously had any abdominal surgery as an infant or child, as this may make certain diagnoses such as bowel obstruction from adhesions or appendicitis more or less likely.

3 The time from symptom development to presentation is often short due to the severity of the symptoms.

4 Pain associated with ovarian torsion is acute, of a sudden onset, and may be constant or intermittent. The pain may be generalised abdominal pain or localised to the side of the affected ovary. The patient may report that the pain radiates to the groin, flank or back.

5 Nausea and vomiting are frequently seen with a presentation of ovarian torsion. However, these symptoms are non-specific and can also be present with many different pathologies. Diarrhoea is another non-specific symptom that may be present. Gastroenteritis can also present with abdominal pain, nausea, vomiting and diarrhoea, although usually the abdominal pain will begin after the other symptoms, unlike in ovarian torsion, and is usually much less acute. As the presentation of torsion can be very non-specific, it should always be considered when a female presents with abdominal pain. Ovarian torsion presents similarly from children to older adults although pre-menarchal children are more likely to present later after the onset of symptoms with diffuse pain and fever. Ovarian torsion can be challenging to identify in neonates whose only symptoms may be abdominal distension, irritability, vomiting and feed intolerance.

6 Some cases of ovarian torsion may be precipitated by sudden movements such as with strenuous exercise or brief, sudden increase in intra-abdominal pressure, such as when coughing, hiccuping or straining on the toilet.

7 Fever may be seen in cases of ovarian torsion but is uncommon. A fever associated with a presentation of ovarian torsion could indicate that the ovary is undergoing necrosis. The presence of a fever may also suggest an infective cause for the symptoms, such as appendicitis or a UTI.

8 Pyelonephritis can also present similarly to ovarian torsion with unilateral back, flank or abdominal pain. Fevers

are more likely to be a significant feature on history. Renal colic may also present with a similar picture of acute onset, unilateral back or flank pain with nausea and vomiting. Pain in renal colic is more likely to be intermittent and moving loin to groin, but will not always present with this classical history.

9 An ectopic pregnancy can present similarly to ovarian torsion so must be considered as part of the work-up for abdominal pain in females. Pregnancy itself is also a risk factor for ovarian torsion, with up to 20% of ovarian torsion cases occurring in pregnant women. Enlargement of the uterus as the foetus grows may displace the nearby ovary causing it to tort, particularly if it is already enlarged, such as due to a mass or a cyst. Most cases occur prior to 20 weeks' gestation. In the adolescent population it is important to obtain a gynaecological, menstrual and sexual history from the patient as part of the work-up for abdominal pain. Pelvic inflammatory disease can also present with non-specific diffuse abdominal pain and should be considered in females who are sexually active.

10 Abnormal vaginal bleeding is not commonly seen in ovarian torsion; however, it can be seen with an ectopic pregnancy, a differential that is important to exclude.

11 It is important to elicit a complete medication history to minimise any drug interactions or to avoid causing adverse reactions during management.

Examination

- The patient appears very **uncomfortable** [1] but **not unwell** [2].
- She is **mildly tachycardic** [3] and **hypertensive** [3].
- The respiratory rate and oxygen saturations are within normal limits for age. Her **temperature is 36.5°C** [4].
- The patient has **moist mucous membranes, peripheral and central capillary refill of <2 seconds and good skin turgor** [5].
- **Heart sounds are dual with no added sounds and the chest is clear** [6].
- Abdominal inspection is unremarkable, with **no previous surgical scars** [7] visible.
- There is **diffuse tenderness to palpation** [8] across the lower abdomen.
- There is **no guarding or rigidity** [9].
- An **exquisitely tender mass** [10] is palpable deep in the left iliac fossa.

1 Ovarian torsion is incredibly painful so patients will appear in pain. Younger patients may be restless or inconsolable.

2 Due to the acute onset and presentation of ovarian torsion, patients may still appear relatively well when they attend the ED.

3 Mild tachycardia and/or hypertension may be present as a response to the acute pain the patient is experiencing. In rare cases, haemorrhage may be seen with ovarian torsion, in which case evidence of hypovolaemia with tachycardia and hypotension may be present.

4 Often patients are afebrile but a low grade fever may be seen in some patients with ovarian torsion. The presence of a fever can indicate that the ovary is undergoing necrosis. A fever may also suggest an infective cause for the symptoms, such as appendicitis or a UTI. Children are more likely to present with fever in cases of ovarian torsion.

5 A fluid balance assessment is important as part of the examination, particularly if the patient has been experiencing losses, such as through vomiting and diarrhoea.

6 A general physical assessment of the patient, including cardiovascular and respiratory assessment, should be performed, particularly as part of a pre-theatre assessment.

7 The presence of previous surgical scars may make a diagnosis of a bowel obstruction secondary to adhesions higher on the list of differentials.

8 In 66% of presentations of ovarian torsion, the patient will experience abdominal or pelvic tenderness during palpation. This may be localised to the side of the torsion or the pain may be diffuse.

9 Signs of peritonism may be present in a few patients and raise concern for the presence of ovarian necrosis.

10 A palpable, tender adnexal mass raises the suspicion for ovarian torsion. The adnexa may be palpable due to swelling from venous congestion or due to the presence of an associated mass or cyst. However, this finding is not present in all examinations where the patient has ovarian torsion. An adnexal mass may also suggest other pathologies such as an ectopic pregnancy or a tubo-ovarian abscess.

Investigations

- **Urinalysis** [1] is unremarkable.
- A **urinary and serum pregnancy test** [2] is negative.
- An **FBE is normal**. **Urea, creatinine and electrolytes are normal** [3].
- **Serum tumour markers** [4] are taken and pending.
- An **abdominal ultrasound reveals an enlarged left ovary with probe tenderness and decreased Doppler flow** [5] within the ovary.
- There is a small amount of **free fluid** [6] in the pelvis.
- A **normal appendix** [7] is visualised.

1 Urinalysis should be performed to look for evidence of white cells and nitrites and red blood cells, which may be present in UTI, or for isolated red blood cells which may be present in nephrolithiasis, causing renal colic.

2 Pregnancy testing is important as torsion is more common in pregnant women and an ectopic pregnancy needs to be excluded as a differential. hCG (human chorionic gonadotrophin) is also elevated with some ovarian tumours.

3 In most cases of ovarian torsion, laboratory studies will be normal. In rare cases, a drop in haemoglobin may be seen as a result of haemorrhage associated with the torsion or from a large, ruptured ovarian cyst. A white cell rise may be present with ovarian necrosis but is also non-specific and may be seen in many other alternative pathologies such as appendicitis, gastroenteritis, a UTI or a tubo-ovarian abscess. Thrombocytosis may also be seen in ovarian malignancies in children and adolescents.

4 If a mass is found on examination or further investigation, tumour markers should be ordered in consultation with the paediatric surgical or obstetrics and gynaecological team to investigate for ovarian tumours. In the paediatric population malignancy is unlikely, so senior advice should be sought before exploring further. Generally, tumour markers may only be ordered if a mass is confirmed on ultrasound and appears suspicious for malignancy. Examples of tumour markers include CA-125, a sensitive but not specific marker for epithelial ovarian cancer; AFP, an antigen produced by teratomas and mixed germ cell tumours; LDH, which is elevated with dysgerminomas; CEA, which is produced by germ or epithelial cell tumours; and inhibin and MIS (Müllerian-inhibiting substance), which are elevated in children with granulosa-theca cell tumours.

5 Ultrasound is the imaging modality of choice for patients with ovarian torsion. An abdominal and transvaginal approach is preferred, but in children only an abdominal ultrasound should be performed. A full bladder is needed for best visualisation, which can sometimes be difficult for children, particularly when they are already in pain and distressed. Findings on ultrasound can vary depending on duration of symptoms and degree of torsion, but can include an enlarged ovary relative to the other side due to lymphatic and vascular congestion causing oedema; visualisation of an ovarian mass; abnormal ovarian location; absent or reduced Doppler flow (although the presence of Doppler flow does not rule out torsion); heterogeneous appearance of ovarian stroma due to oedema and haemorrhage; multiple small peripheral follicles as they have been displaced by oedema (this finding is also seen in polycystic ovarian syndrome but is not associated with acute pain or oedema); and a 'whirlpool sign' caused by side-by-side arrangement of vessels with opposing directions of blood flow, representing twisting of the vascular pedicle. CT is not typically used to diagnose ovarian torsion, but may be performed when another cause of acute abdominal pain, such as appendicitis, is thought to be more likely. Its findings are similar to those seen on ultrasound, with an enlarged, abnormally placed ovary, and with contrast enhancement the whirlpool sign may also be visible. However, CTs are rarely used in the paediatric population unless essential, due to the associated radiation exposure. MRIs are another modality that can be used to diagnose ovarian torsion, especially if the findings on ultrasound are equivocal, although due to time, cost and centre availability it may not be frequently used. In the neonatal population, ovarian cysts are often diagnosed on antenatal ultrasounds and will then be followed up with serial ultrasounds in the prenatal and neonatal period and beyond to track progress/resolution. Most cysts are expected to resolve by 6 months of age. If cysts do not resolve, continue increasing in size or become symptomatic they may be excised surgically, or if the cyst is simple and >4mm in size the fluid from the cyst can be aspirated. Parents and carers should be informed of the signs of ovarian torsion in a neonate and instructed to present to the ED immediately if their child is displaying concerning symptoms for torsion.

6 Both ovarian torsion and a ruptured ovarian cyst can have findings of free fluid in the pelvis on ultrasound.

7 Appendicitis can present similarly to ovarian torsion with abdominal pain, nausea and fever. Differentiation between the two should be made on history, examination and investigations. In this case a normal appendix has been visualised on ultrasound, which lowers clinical suspicion for appendicitis being the cause of the pain, particularly with the other imaging findings present being suggestive of ovarian torsion.

Management

Immediate

Gain **IV access** [1]. Provide **analgesia and anti-emetics** [2]. Keep patient **nil by mouth** [3]. Monitor **vital signs** [4]. Commence **IV fluids** [5]. Refer to **paediatric surgical team** [6].

Short-term

Patients should proceed **as an emergency** [7] to the **operating theatre** [8] for exploration of suspected ovarian torsion. Postoperatively they should be **monitored** [9] and have **examination of the abdomen** [10] and **surgical sites** [11]. They should be started back on a **full diet** [12].

Long-term

Patients are suitable for discharge if their **observations remain stable, they are pain-free and tolerating a full diet** [13]. They should be **educated** [14] on signs of ovarian torsion. They should be followed up in **paediatric surgical outpatient clinic** [15].

1 IV access should be established quickly so as not to delay getting the patient to theatre, and it also provides an access for IV medication and fluid administration.

2 Effective analgesia should be provided to the patient to help ease the pain from a torted ovary. Anti-emetics can help settle nausea and vomiting.

3 Patients should be kept nil by mouth due to the urgent requirement for surgery to reduce the risk of aspiration while under anaesthetic.

4 Vital signs should be observed to ensure the patient remains haemodynamically stable.

5 While the patient is fasting, provide fluids in the form of 0.9% sodium chloride with 5% glucose at full maintenance hydration. Consider fluid boluses of 0.9% sodium chloride if the patient appears hypovolaemic.

6 Refer to the paediatric surgical team for all patients ≤16 years presenting with suspected ovarian torsion for urgent surgical intervention. If a paediatric service is not available, consider transfer to a tertiary centre or alternatively referral to the obstetrics and gynaecology team.

7 Surgical intervention should be performed swiftly in order to preserve ovarian function and reduce the chance of other adverse effects associated with ovarian necrosis, such as haemorrhage, peritonitis and subsequent formation of adhesions.

8 A diagnosis of ovarian torsion can only be made once visualised intraoperatively. A laparoscopic approach is usually used. The ovary should be detorted and viability assessed. In most cases the ovary is viable even with a blue–black appearance, and should be left in situ, particularly in young patients and those who are premenopausal, to preserve fertility. A necrotic ovary or Fallopian tube may appear gelatinous or have loss of its usual structure. If an ovary is clearly necrosed, an oophorectomy should be performed. For patients with a large ovarian cyst identified, cystectomy can be done at the same time as the detorsion procedure. For patients in whom malignancy is suspected, unless malignancy is confirmed at the time of procedure via frozen section, the mass should be excised and the ovary reconstructed. However, if malignancy is confirmed, a salpingo-oophorectomy should be performed on the affected side. It is better in a paediatric population, if malignancy is suspected but not definitively diagnosed, to manage only the torsion and excision of the lesion and then confirm malignancy with specimens and serum tumour markers. It is preferable to have the patient undergo a second procedure for salpingo-oophorectomy rather than remove them without absolute confirmation of the diagnosis.

9 Observations should be monitored postoperatively to ensure haemodynamic stability. Tachycardia or hypotension may indicate postoperative bleeding, while the concurrent presence of a fever could indicate sepsis from a retained necrotic ovary, something that warrants urgent re-exploration.

10 The abdomen should be examined for any evidence of guarding or rigidity, indicating peritonitis which may also be present with a retained necrotic ovary.

11 Surgical wounds should be observed to ensure good wound healing, and that there are no signs of dehiscence or wound infection.

12 Patients can be restarted on a normal diet as tolerated postoperatively and encourage children to resume feeding as normal as soon as able.

13 Once the patient appears well and is back to their usual baseline functioning, they are suitable for discharge from hospital.

14 Patients, parents and carers should be advised of the symptoms of ovarian torsion and instructed to re-present if the symptoms re-occur. Ovarian torsion may re-occur but the incidence and risk factors for recurrence are not known.

15 Patients should be followed up in the outpatient clinic to ensure they have been able to resume their usual activities, to review their surgical wounds to ensure good healing and to review any outstanding pathology from their admission.

CASE 60: Pyloric stenosis

History

- A **6-week-old** [1] **male** [2] infant with **no significant past medical history** [3] is sent in to the ED with his parents by his maternal and child health nurse, due to a 2-week history of recurrent vomiting and **irritability** [4].
- His parents describe a progressive history of **forceful milky vomiting not long after feeding** [5].
- There is no **blood** [6] noticed in the vomits.
- They have tried **multiple different formulas** [7] over the past couple of weeks but there has not been any improvement in symptoms.
- The maternal and child health nurse has documented that **weight gain** [8] has stagnated over the past few weeks.
- The infant has **not opened his bowels** [9] for two days and his **wet nappies** [10] have been lighter and less frequent.
- His mother had an unremarkable pregnancy and the infant was born at **term** [11] via a normal vaginal delivery.
- She is a **non-smoker** [12].
- The infant has had no **sick contacts** [13].
- This is the couple's **first child** [14].
- The infant's **mother had pyloric stenosis** [15] as a baby.

1 Most infants with hypertrophic pyloric stenosis present within 3–6 weeks of age and very rarely after 12 weeks of age. Hypertrophic pyloric stenosis, also known as infantile hypertrophic pyloric stenosis or simply pyloric stenosis, occurs at a rate of 2–3.5 per 1000 live births on average, but is often region-dependent, with pyloric stenosis more common in western populations as opposed to those in Africa and Asia.

2 Pyloric stenosis is more common in males, with a male to female ratio of 4:1.

3 Infants with a history of previous abdominal surgery who develop vomiting may have their symptoms initially attributed to adhesions causing bowel obstruction.

4 Infants with pyloric stenosis are often irritable, crying and difficult to console as they are hungry due to ineffective feeding. They will often want to feed again soon after vomiting.

5 Vomiting is the most common symptom seen in pyloric stenosis. It is projectile in nature, non-bilious as only food products are being regurgitated, and occurs following feeds. Sometimes this may be mistakenly assumed to be gastroenteritis, though other features of gastroenteritis such as loose stools and fever are not present. UTI can also present in a similar way, with repeated vomiting and irritable infants, and it is an important differential diagnosis to exclude as part of the work-up. The presence of bilious vomiting is indicative of a more distal intestinal obstruction, such as malrotation with volvulus or Hirschsprung disease.

6 In approximately 10% of cases, parents may notice blood stains in the vomit.

7 Early pyloric stenosis can be easily confused with intolerance to certain formulas or gastro-oesophageal reflux, and may be misdiagnosed before later stage symptoms present. Vomiting in gastro-oesophageal reflux tends not to be projectile and occurs 10 or more minutes after finishing feeding. Pyloric stenosis is also seen more often in bottle-fed infants as opposed to exclusively breastfed infants.

8 There is often inadequate weight gain in these infants due to insufficient ingestion and retention of food, and consequently poor nutritional absorption.

9 Constipation may develop in late stages of pyloric stenosis, secondary to dehydration due to poor oral intake, as most food is being vomited rather than digested.

10 Decreased wet nappies is another sign of dehydration secondary to poor oral intake.

11 Preterm delivery is a risk factor for pyloric stenosis, though diagnosis is usually at a later chronological age than term infants, and it can be more difficult to reach the diagnosis due to atypical symptoms or investigation findings. Term infants are more likely to present with classical features of pyloric stenosis with non-bilious, postprandial projectile vomiting.

12 The risk of an infant developing pyloric stenosis is increased by 1.5–2 times in children whose mothers smoked during their pregnancy.

13 Contact with other patients with GI symptoms such as diarrhoea and vomiting may make a diagnosis of gastroenteritis more likely.

14 Pyloric stenosis is most likely to affect the first-born in a family, as opposed to later children, at a rate of 2:1.

15 There is a strong familial risk involved in developing pyloric stenosis and it is seen more often when the mother had pyloric stenosis.

Examination

- The infant appears **flat** [1] and lethargic.
- His **heart rate** [2] is 190bpm.
- **Blood pressure** [3] is 75/50mmHg.
- **Respiratory rate** [4] is 40 breaths per minute. Oxygen saturation is 97% on air.
- The **temperature** [5] is 37°C.
- The infant has **sunken fontanelles and dry mucous membranes** [6].
- There are **no changes to the skin or sclera** [7].
- **Heart sounds are dual** [8] with no added sounds.
- **Chest is clear** [9] to auscultation.
- **Inspection of the abdomen** [10] is normal with **no surgical scars visible** [11].
- Careful palpation of the abdomen reveals a **small, firm mass in the right upper quadrant, lateral to the rectus abdominis** [12].
- **Bowel sounds** [13] are present.
- The **nappy is dry** [14].

1 A change in conscious state is a sign of shock from severe volume depletion.

2 Tachycardia is seen in severe dehydration associated with prolonged course of symptoms and delayed presentation.

3 The infant may be in hypovolaemic shock due to severe volume depletion. In some cases, infants with a delayed presentation of pyloric stenosis need to be stabilised in the ICU before they are suitable for surgery.

4 Respiratory rate may be increased in dehydration states.

5 Fever is not usually an examination finding in pyloric stenosis and if present, may suggest an alternative diagnosis.

6 A fluid balance assessment is an important part of the work-up for pyloric stenosis to assess hydration status, as infants can become severely dehydrated from repeated vomiting and inability to absorb feeds. Other features of the fluid balance assessment include assessing skin turgor, urine output and capillary refill time.

7 The presence of eczema raises suspicion for a food allergy as the cause of the patient's symptoms. Additionally, jaundice or icterus may be seen in infants with pyloric stenosis due to hyperbilirubinaemia. The presence of jaundice would also warrant an investigation into the possibility of underlying liver disease. Mottled skin is a sign of severe dehydration.

8 A full physical work-up is necessary as part of assessing suitability for surgery. A flow murmur may be appreciated during tachycardia which resolves after adequate volume repletion.

9 Repeated vomiting increases an infant's risk of aspiration during these episodes. Assessment of the lungs should be completed to rule out a concurrent pneumonia.

10 Visible peristalsis may be observed as waves travelling from left to right across the abdomen as the stomach attempts to force contents across the hypertrophied pylorus and narrowed pyloric outlet. It can be seen just prior to vomiting in some infants. Abdominal distension may indicate obstruction rather than pyloric stenosis.

11 Previous abdominal surgery may cause adhesions and bowel obstruction, which can also present with repeated vomiting.

12 A small, firm mass in the right upper quadrant lateral to the rectus abdominis described as an 'olive-shaped mass' is pathognomonic for pyloric stenosis and its presence confirms a clinical diagnosis without need for imaging. It is most easily palpable when the infant is settled and immediately post vomiting so that its presence is not obscured by tensed abdominal muscles. In earlier presentations of pyloric stenosis, the olive-shaped mass may be difficult to palpate.

13 High-pitched bowel sounds are suggestive of bowel obstruction, and if concurrently associated with bilious rather than non-bilious vomiting, should be investigated with a plain film to identify an obstruction.

14 A dry nappy may also be a sign of volume depletion as urine output is reduced. Stool should be inspected for evidence of bleeding. Occult blood may be present in otherwise healthy-appearing infants with a cow's milk protein allergy, while frank blood PR may suggest intussusception, particularly if the symptoms are of a shorter duration.

Investigations

- **Urinalysis** [1] is normal.
- An **FBE** [2] is normal.
- A VBG shows a **raised pH and bicarbonate levels and low serum chloride and potassium** [3].
- Formal UECs confirm low serum chloride and potassium and **mildly elevated creatinine and urea** [4].
- There is a finding of **unconjugated hyperbilirubinaemia** [5] on liver function testing.
- An **abdominal ultrasound** [6] confirms the diagnosis of pyloric stenosis.

1 A urine dipstick is a quick way to test for signs of a UTI that may be causing vomiting and irritability. A full urine microscopy and culture should be sent, as infants presenting in severe dehydration present similarly to those in sepsis, so investigating for all possible causes while the diagnosis is not confirmed is important.

2 FBE is usually normal in pyloric stenosis. There may be a mild WCC rise as a response to repeated vomiting. A large inflammatory marker rise may indicate symptoms are more likely due to an infection rather than pyloric stenosis. Patients with early presentation and diagnosis will often still have normal pathology results, as changes due to repeated vomiting and dehydration are seen in delayed presentations.

3 A hypochloraemic hypokalaemic metabolic alkalosis is a typical finding in late presentations of pyloric stenosis. It has a positive predictive value of 88% in diagnosing pyloric stenosis when the infant's main presenting symptom is vomiting. The degree of abnormality on the blood samples is proportional to the duration of symptoms prior to presentation.

4 Raised urea and creatinine are also a marker of dehydration status and give an assessment of effects of dehydration on renal function.

5 14% of cases of pyloric stenosis are associated with unconjugated hyperbilirubinaemia – the combination of which is termed icteropyloric syndrome. In many cases it is an early manifestation of Gilbert syndrome. Hyperbilirubinaemia resolves quickly after surgical correction of the pyloric stenosis.

6 If a diagnosis of pyloric stenosis cannot be made clinically then abdominal ultrasound is the gold standard to confirm the suspected diagnosis. The sensitivity and specificity for diagnosing pyloric stenosis on ultrasound is >95%. If the ultrasound is negative or equivocal for pyloric stenosis but based on history and examination pyloric stenosis still seems the most likely diagnosis, ultrasonography is repeated in a few days to see if classical imaging findings of pyloric stenosis develop with more time.

Management

Immediate

Gain **IV access** [1] and begin **fluid resuscitation** [2]. **Stop feeding** [3]. Consider **NGT insertion** [4]. Monitor **vital signs** [5] and **urine output** [6] and escalate to intensive care team if required. **Repeat blood gases every 6 hours** [7]. Refer to **paediatric surgical team** [8] or **transfer** [9] to nearest paediatric surgical service if in a rural area.

Short-term

Once the patient has been stabilised with hydration status, acid–base status and electrolyte disturbances all corrected, they can proceed to definitive management in the form of a **pyloromyotomy** [10]. Postoperatively patients should receive **apnoea monitoring** [11] for at least 24 hours. **Feeding** [12] can be resumed within a few hours after surgery. Examine the **abdomen** [13] and review surgical **wounds** [14].

Long-term

Patients are suitable for discharge once they are pain-free and **tolerating full oral feeds** [15]. They should be monitored in the community by their maternal and child health nurse or GP to ensure adequate **weight gain** [16] is made. They should be followed up in **surgical outpatient clinic** [17].

1 Ensure quick establishment of IV access to begin fluid replacement and electrolyte correction as quickly as possible.

2 Initial fluid resuscitation should be with 10–20ml/kg bolus of 0.9% sodium chloride. Not all children will require fluid resuscitation if they have presented early, appear clinically euvolaemic, have good urine output and normal vital signs. Replace any ongoing deficits and commence maintenance fluids using 0.9% sodium chloride with 5% glucose.

3 Feeding must be stopped as preparation preoperatively and to ensure emesis episodes subside.

4 If copious vomiting persists despite stopping feeds, an NG should be inserted to decompress the stomach.

5 Monitor vital signs to ensure tachycardia resolves, hypotension improves and respiratory rate is stable as fluid and electrolyte correction is provided.

6 Once urine output reaches 1–2ml/kg/hour, 20mmol of potassium can be added to maintenance fluids.

7 Blood gases should be repeated regularly so that fluids can be adjusted accordingly. Fluid and electrolyte deficits should be fully corrected within 48 hours from presentation. Serum bicarbonate levels should be fully corrected prior to surgery as ongoing metabolic alkalosis puts the patient at risk for hypoventilation and apnoea postoperatively.

8 A paediatric general surgical team should review and admit the patient for definitive management of their pyloric stenosis.

9 All patients with pyloric stenosis should be managed in a tertiary centre with paediatric surgical and paediatric intensive care capabilities. Once management has been initiated a patient should be transferred to a tertiary centre urgently.

10 To correct the pyloric narrowing due to the hypertrophic pylorus and therefore allow stomach contents to pass into the duodenum, a longitudinal incision is made in the pylorus and then blunt dissected down to the level of the submucosa. When an open approach is used this is referred to as a Ramstedt procedure, although a laparoscopic approach is typically favoured when possible.

11 Apnoea monitoring should be a routine part of postoperative monitoring, as infants are at higher risk for apnoea due to their young age, effects of anaesthetic and if the patient's alkalosis was not fully corrected preoperatively.

12 Infants may still demonstrate some regurgitation postoperatively, but the projectile-type vomiting should now have resolved. The rate of incomplete pyloromyotomy is 1%.

13 Routine abdominal examination should be a part of postoperative reviews. Ensure there is no evidence of abdominal distension, rigidity or abdominal tenderness out of proportion with surgical wounds, as this may be concerning for an intraoperative perforation and consequent peritonitis. However, the rate of mucosal perforations intraoperatively is <1%, and easily identified and corrected at the time.

14 Surgical wounds should be reviewed to ensure adequate wound healing, no signs of dehiscence or developing infection.

15 Feeds should be back at expected amount for age before discharge, to ensure the infant will receive adequate nutrition and hydration in the community.

16 Infants with delayed presentation may have had weight loss due to inadequate feeding at initial presentation. It is important to ensure the infant is having steady weight gain after correction of the pyloric stenosis, and length and weight should be plotted on child growth charts.

17 Patients should be reviewed in surgical outpatient clinic for review of wound healing and to ensure the child is gaining weight and reaching expected milestones. If infants are still having issues with vomiting or inadequate weight gain despite correction of the pyloric stenosis, a referral to a paediatrician or inpatient admission for further investigation is warranted in order to identify other causes to explain their persisting symptoms. Gastro-oesophageal reflux is common but no more so than in infants who have not had pyloric stenosis, and should only be investigated if the reflux is severe or causing other symptoms.

CASE 61: Breast lump and lymphoedema

History

- A **70-year-old** [1] **female** [2] has been referred to the outpatient plastic and reconstructive surgery clinic of a tertiary hospital with a **right-sided breast lump** [3].
- She has not yet had any investigations and tells you that she found the lump **about 6 months ago whilst showering** [4].
- She is up to date with her **mammography** [5] and has always had **'dense breasts'** [6].
- Throughout her teens and early 20s, she had **breast mice** [7] but nothing more serious.
- Her **sister and mother both died of breast cancer** [8], her sister aged 49 and her mother aged 74.
- She has a **BMI of 30** [9], **T2DM** [10] and **hypercholesterolaemia** [11].
- She is a retired **nurse** [12], and spends her spare time making small handicrafts for the local market.
- She has been married for 50 years and her husband has recently pointed out that she has **lost a lot of weight** [13] and on prompting, she says perhaps her **appetite has been low** [14] for the last year.
- She has not had **drenching night sweats or fevers** [15].
- When she first found the lump, it was about the size of a pea, but **grew rapidly** [16] over the last 6 months to now be the size of a golf ball.
- The patient has also noticed her **nipple has turned in** [17].
- She denies **any pain or tenderness** [18] in the lump and has not experienced any **nipple discharge** [19].

1 Breast cancer occurs most commonly in women over 50 years old and in men over 60 years old. Risk increases with age.

2 Both males and females can develop breast cancer.

3 A breast lump is a localised bulge or swelling in the breast that feels different from the surrounding breast tissue or compared with the other breast.

4 Knowing the time period that the breast lesion developed can be helpful in diagnosis and determining how aggressive a cancer is; the faster it has been growing, the more likely it is to be metastatic.

5 Mammography is a radiographic technique where the breasts are sandwiched between X-ray plates and imaged to view the ducts, the breast tissue and the nipple. Regular screening mammography is recommended for women after age 45.

6 Dense breast tissue comprises milk glands, supportive tissue and milk ducts as seen on a mammogram. Non-dense breast tissue is fat. Having 'dense breasts' means that there is less fat than breast tissue and breast cancer may not be able to be seen easily on a mammogram; it is more common in women with a lower BMI, taking hormone therapy for menopause or who are of a younger age. Levels of breast density are described using a reporting system called Breast Imaging Reporting and Data System (BI-RADS):

- A: almost entirely fatty; 1 in 10 women have this result
- B: scattered areas of fibroglandular density, most of the tissue is non-dense (fat); 4 in 10 women have this result
- C: heterogeneously dense (majority of breast tissue is dense tissue); 4 in 10 women have this result
- D: extremely dense (nearly all the breast tissue is dense); 1 in 10 women have this result.

7 'Breast mice' are breast fibroadenomas. Simple fibroadenomas are the most common and do not increase the risk of breast cancer; however, the complex subtype does. The latter has calcification, enlarged lobules (glands) and cysts. There can be one or several and can occur in one or both breasts. Both types are often tender and enlarge during the period of menstruation and are most common in women aged 20–30 years.

8 The majority of inherited cases of breast cancer are associated with two genes, *BRCA1* and *BRCA2* (which stands for BReast CAncer gene 1 and 2). These normally repair cell damage. Female carriers of a germline mutation in BRCA1 have a lifetime risk of breast cancer >80% and ovarian cancer risk of 60%. Female carriers with a germline mutation in *BRCA2* have a similar lifetime risk of breast cancer to this but a higher ovarian cancer risk in their lifetime.

9 Obesity increases incidence and mortality from multiple cancer types, including breast and endometrial. The current thought is that through a multitude of various endocrine

factors, adipose tissue contributes to a high inflammatory response and resultant tumorigenesis.

10 People with T2DM have much higher rates of all types of cancer compared to non-diabetics.

11 Hypercholesterolaemia is not a risk factor for breast cancer, but an overall reflection of general health.

12 Some studies suggest people who work night shifts long-term, such as nurses, have a higher risk of breast, GI and lung cancer than people who do not.

13 Loss of weight can be a sign of cancer or chronic inflammatory disease.

14 Loss of appetite can be a symptom of cancer but can also be a reflection of stress, anxiety, depression or other chronic disease (e.g. lung disease, heart failure, chronic kidney failure).

15 Drenching night sweats and fevers are symptoms of cancer (typical of Hodgkin's disease but can be any neoplastic process) or infectious disease (e.g. TB or HIV).

16 Rapid growth of a mass in any region is a concern for a neoplastic process.

17 Inverted nipples can be a symptom of breast cancer but also can be due to increased age, mammary duct ectasia (a non-cancerous occurrence during perimenopause), or Paget's disease of the nipple.

18 Breast cancers are not typically tender.

19 Nipple discharge (clear or bloody) is the most common presentation for intraductal papilloma. It is a wart-like tumour that develops within the breast ducts which are close to the nipple.

Examination

- The patient has a right breast mass about **3cm in diameter at 3 o'clock** [1].
- It is **firm** [2] to the touch and **adherent to the overlying skin** [3].
- It feels **irregular** [4], but is **non-tender** [5], and there is **no erythema or change in temperature** [6] when compared with the surrounding skin.
- There is no evidence of **tethering** [7] or **peau d'orange** [8] but the **nipple is partially inverted** [9].
- There are no dilated veins or ulceration of the breast and the **nipple looks otherwise normal** [10]. It is not possible to express anything from the nipple.
- The left breast does not have any masses. On a lymphatic examination, there is **one enlarged lymph node** [11] at **axillary node level I** [12].

1 When describing a breast lesion or skin change, it is reported using a clock face where the nipple is in the centre.

2 Texture of a breast mass can aid diagnosis. For example, breast mice are round, smooth, and move underneath the examiner's hand easily, which is why they are colloquially called mice. Concerning for malignancy are masses that are firm, irregular in shape or fixed to the skin.

3 This is a concerning feature for malignancy as the mass underneath has tethered to the skin overlying it.

4 Irregular breast masses are suspicious of cancer.

5 Non-tender breast lumps can be lipomas, a milk cyst (galactocele), a malignant breast cancer (e.g. lobular or ductal carcinoma) or lymphoma. Tender breast masses can be an abscess, mastitis, fibroadenoma (more common in those aged 20–30 years), or phylloides tumour (these can also be non-tender, but grow rapidly as a firm, round mass that is rarely malignant but treated with wide local excision).

6 This is looking for inflammatory lesions (such as a breast abscess or cellulitis) and inflammatory breast cancer. The latter is more common in younger women and those of African heritage, becoming clinically apparent when the cancer cells block the lymph vessels in the skin of the breast and cause it to become erythematous and swollen. It is highly aggressive and is often metastatic when diagnosed.

7 Tethering is highly suggestive of breast cancer.

8 Peau d'orange is an orange peel appearance of the skin caused by lymphatic blockage.

9 As mentioned above, nipple inversion can be due to age, mammary duct ectasia (a non-cancerous occurrence during perimenopause), Paget's disease of the nipple and breast cancer.

10 Clinical findings in Paget disease of the nipple include:
- crusting, scaling or flaking of the nipple
- erythema of the nipple and areola
- burning or itching in the nipple and breast
- bleeding or discharge from the nipple
- nipple inversion or flattening
- a lump felt underneath or around the nipple.

11 Enlarged lymph nodes are >1cm and can be smooth, rubbery or hard in consistency.

12 Lymphatic drainage of the breast is to internal mammary nodes and via axillary lymph nodes. The latter is surgically divided into:

- level I – inferior and lateral to pectoralis minor muscle
- level II – posterior to the pectoralis minor muscle and inferior to the axillary vein
- level III – infraclavicular, medial to the pectoralis minor and against the chest wall; involvement of these lymph nodes carries a poor prognosis.

Investigations

- Breast lesion work-up is often called the **'triple assessment'** [1].
- **FBE and UECs** [2] are unremarkable and a **group and hold** [3] is ordered in preparation for surgery.
- **Coagulation studies** [4] are considered for ordering.
- **Mammography** [5] is ordered for this patient and the results show a **spiculated, irregular mass** [6] at 3 o'clock in the right breast **just next to the nipple** [7].
- It has **microcalcifications and areas of hyperdensity** [8].
- The left breast does not have any suspicious features. The mammography findings that are suspicious for breast cancer are conveyed to the patient at the next outpatient visit.
- **Core biopsy** [9] of the lesion is organised under ultrasound guidance and the results return as **invasive ductal carcinoma** [10].
- After the core biopsy has been taken to aid diagnosis, a **PET scan or CT chest** [11] will be further organised.

1 The 'triple assessment' consists of the clinical examination, imaging of the breast and a biopsy of the lesion.

2 In this patient, an FBE could be taken in order to have a baseline haemoglobin level in the event she needs blood transfusion after surgery. UECs are useful in this patient for a baseline, as many medications and anaesthetics are dosed based on renal excretion.

3 A blood group and hold tests for blood type and a few units are put aside in preparation for surgery. It is performed preoperatively in major surgeries; for example, a mastectomy or reconstruction.

4 Coagulation studies should be ordered if the patient has a history of liver disease, chronic alcoholism (with undiagnosed liver disease), has a clotting disorder or is on anticoagulation. Peri-operatively, these tests are used to guide whether the patient will require medications or blood products to reverse their anticoagulation in order to minimise blood loss during and after surgery.

5 Mammography is used in fatty breasts, but ultrasound and MRI are more sensitive for detecting breast cancer in non-fatty breasts.

6 These features suggest cancer. Spiculated lesions mean that the lesion's edges have spikes, like the sun, but can be found in benign processes such as a post-surgical scar, a Desmond tumour, an abscess or fat necrosis, but can be suspicious of malignancy (where the lesion is invading surrounding tissue).

7 Lesions next to the nipple can be papillomas or ductal carcinomas.

8 Microcalcifications, asymmetry and architectural distortion are suspicious features for malignancy but can also be seen in fat necrosis (from trauma to the breast) and scarring. Benign causes of hyperdense breasts are after breast irradiation, in pregnancy, and in breasts with minimal fat. Malignant and infective causes of hyperdense breasts include diffuse involvement with lymphoma, inflammatory carcinoma and mastitis (the latter two secondary to lymphatic or venous drainage obstruction in that breast).

9 A core biopsy is performed under ultrasound guidance and is the least invasive way to diagnose a breast lesion and therefore whether the patient requires surgery.

10 Invasive ductal carcinoma is the most common type of invasive breast cancer and metastasises via the lymphatics. Invasive carcinomas of the breast are reviewed on histopathology using the Nottingham Criteria that assess gland formation, nuclear atypical and mitosis counts in order to give the lesion a 'grade' – higher grades correspond to a lesion that is more invasive, more aggressive and has a higher risk of metastasis.

11 A PET scan uses a radioactive tracer administered through the vein which is taken up by tissues that have a high metabolic rate (e.g. cancerous lesions, the thyroid) and excreted through the kidneys. In breast cancer, it has been recently shown that PET and PET/CT imaging have low sensitivity and specificity in staging axillary lymph nodes or metastases and also have high false-positive results.

Management

Immediate

The patient should be offered **psychological support** [1]. Breast cancer resection, whilst important, is not an emergency so the patient should be **consented for surgery** [2] and placed on the **waiting list** [3].

Short-term

Treatment is by surgical excision +/– **sentinel node biopsy** [4] or **axillary node dissection** [5]. A **suction drain** [6] may be left in place after a mastectomy with axillary dissection to reduce the dead space and allow a path for **fluid to drain** [7]. Postoperatively, the patient may be fitted with a **compression garment** [8] to wear around their chest for 1–2 weeks to reduce formation of **seroma or haematoma** [9]. She should be presented at a multidisciplinary meeting where the **full course of treatment** [10] can be determined. Adjuvant treatment options include **chemotherapy** [11], **radiation therapy** [12] and **targeted hormone treatment** [13]. Reconstructive options after mastectomy include a **TRAM** [14] or **DIEP flap** [15] or **tissue expanders and breast implants** [16].

Long-term

If the patient develops lymphoedema post axillary lymph node dissection then **compression garments** [17] can be worn and venepuncture or vascular access **should be avoided** [18] in this region. They should be followed up **every 3–6 months** [19] initially to monitor response to treatment and recurrence. Often, follow-up imaging is in the form of mammography. If *BRCA1* or *BRCA2* gene positive, first-degree relatives should be offered **genetic screening and counselling** [20].

1 Psychological support should be long-term, as the diagnosis and treatment of breast cancer can understandably cause grief, stress, depression and anxiety. Additional long-term effects of the diagnosis and treatment of any cancer can be pain, fatigue, loss of appetite, weight loss, hair loss and other medication side-effects.

2 Surgical excision of breast cancer can be in the form of a wide local excision, lumpectomy or segmentectomy (i.e. breast-conserving surgery) or total mastectomy, depending on the type of breast cancer, the tumour stage, patient preference and breast size, and geographic region (i.e. surgeons that are available in that city or country, and if reconstructive options are offered in that region).

3 Time on the wait list for surgery depends on the geographical location of the hospital, but generally cancer surgery is urgent, warranting <30 days of waiting before the surgery date.

4 Sentinel node biopsy is performed when there is one lymph node that is involved by metastasis – the 'sentinel node'. Lymphatic mapping with technetium-99 or blue dye can be injected into the region that the breast cancer is occupying and the lymph drainage to this lymph node can be detected with a hand-held gamma probe during the operation. Risks involved in sentinel lymph node biopsy (SLNB) are reactions to the dye (e.g. anaphylaxis), lymphoedema and false-negative results.

5 Axillary lymph node clearance has higher morbidity than an SLNB as more lymph nodes are being removed. It is performed when there is evidence of metastasis on imaging or if the lesion is high risk for metastasis, such as invasive ductal carcinoma. During the procedure, the patient should be under general anaesthesia but not paralysed, so that the large motor nerves can be tested during the dissection.

6 Examples of surgical drains include:
- Jackson–Pratt – a perforated round or flat tube connected to negative pressure collection bottle
- Blake – round silicone radio-opaque drain with four channels along the sides connected to a negative pressure bottle
- Penrose – a flat, thin malleable tube that is put in a wound to hold it open; it cannot be attached to a drain bottle.

7 Fluid from any wound after an operation can be serous (yellow watery proteinaceous fluid), bloody, or haemoserous (blood-tinged serous fluid). Placing a drain in a wound that is on suction allows dead space to collapse down and promote internal wound healing, whilst also allowing any slow bleeding or build-up of serous fluid to drain.

8 Compression garments can be tailor-made or by using a Tubigrip sock in a size as large as the patient's chest and cut to size.

9 A seroma is a collection of a clear (serous) fluid collection under a skin flap and frequently occurs after mastectomy and axillary dissections. A haematoma is a collection of blood.

10 Whilst resection is the primary treatment for invasive ductal carcinoma, after histology has returned, an MDT

should discuss whether adjuvant chemo- or radiotherapy is required and what reconstructive options can be employed for this patient.

11 Chemotherapy for breast cancer includes doxorubicin, carboplatin and cyclophosphamide, among others, and can be used in combinations.

12 Radiation therapy can be delivered via external beam radiation or partial breast irradiation, being aimed at the chest and the lymph nodes in the chest, shoulder and axilla. This aims to eradicate residual disease that is not macroscopically seen during surgical resection of the tumour or where margins are considered close after resection. Some risks associated with radiotherapy include a rash, pain and change in colour or texture of the skin.

13 Targeted hormone therapy depends on what receptors the cancer expresses and are used only in postmenopausal women. These are:

- Herceptin (trastuzumab, a monoclonal antibody against HER2 receptors) for breast cancers that express HER2 (human epidermal growth factor-2).
- Anastrozole (Arimidex) and letrozole (Femara) for breast cancers that express oestrogen receptors (called ER-positive) or progesterone receptors (called PR-positive). Around 80% of breast cancers are ER-positive.
- Tamoxifen (a selective oestrogen receptor modulator) used in ER-positive breast cancers (largely replaced by anastrozole and letrozole).
- Palbociclib (Ibrance) – therapy in combination with letrozole for HER2-negative, hormone-receptor positive (ER- or PR-positive) breast cancer.

14 A TRAM (transverse rectus abdominis myocutaneous) flap is the surgical method of removing skin, fat and part of the rectus abdominis muscle from the abdomen to the chest with the superior epigastric artery.

15 A DIEP (deep inferior epigastric perforator) flap is the surgical method of removing skin and fat in the infraumbilical part of the abdomen to reconstruct a chest wall defect from a mastectomy. The benefits of a DIEP flap are that the rectus abdominis muscle is preserved, therefore reducing abdominal hernias in future.

16 In some patients who have small breasts or who have a mastectomy, tissue expanders are inserted under the skin and pectoralis major to slowly stretch the skin overlying it in preparation for a prosthetic breast implant.

17 Compression garments for upper limb lymphoedema are Tubigrip stockings which fit to the arm and aim at milking the lymph up the arm.

18 In patients without lymphoedema of the upper limb, there is minimal evidence to suggest venepuncture precipitates lymphoedema. Current suggestions state that if a venepuncture cannot be achieved on the contralateral arm, then venepuncture in the ipsilateral arm (the side of the axillary lymph node dissection) should be attempted rather than in sites with higher infection rates, such as the foot. The concern in arms that have had an axillary lymph node clearance is that it is at higher risk of infection and venous thrombosis due to slow or absent lymphatic clearance.

19 Frequency of follow-up depends on the tumour grade and staging and the treatment the patient undergoes.

20 Genetic counselling involves a healthcare practitioner trained in cancer genetics guiding patients and their families through genetic testing and helping them understand their risk and options for prevention in various hereditary conditions.