### Table 30.5 Essential features of common bone tumours.

<table>
<thead>
<tr>
<th>Tumour</th>
<th>Age (Yrs)</th>
<th>Common sites</th>
<th>Location</th>
<th>Clinical features</th>
<th>X-ray picture</th>
<th>Differential diagnosis</th>
<th>Pathology</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteosarcoma</td>
<td>15-25</td>
<td>Lower end of femur, upper end of tibia</td>
<td>Metaphysis</td>
<td>Pain ++ Swelling ++ Duration wks - mths *</td>
<td>Sun-ray appearance, Codman's triangle, tumour new bone +</td>
<td>Ewing's tumour</td>
<td>Tumour cells with osteoid or bone formation, Alkaline phosphatase increased in 50% cases</td>
<td>Immunotherapy + chemotherapy</td>
</tr>
<tr>
<td>Ewing's tumour</td>
<td>5-15</td>
<td>Femur, tibia flat bones, multi-centric **</td>
<td>Diaphysis</td>
<td>Pain ++ Swelling + Duration wks-mths, often lever+</td>
<td>Onion-peel appearance</td>
<td>Osteosarcoma, Osteomyelitis Sheaths of round cells</td>
<td></td>
<td>Radiotherapy + chemotherapy</td>
</tr>
<tr>
<td>Osteoclastoma</td>
<td>20-40</td>
<td>Lower femur upper tibia, lower radius</td>
<td>Epiphysis region</td>
<td>Pain + Swelling + Duration - month</td>
<td>Soap-bubble appearance No tumour new bone</td>
<td>Aneurysmal bone cyst, Fibrous dysplasia Multi-nucleate giant cells in fibrous stroma</td>
<td>Excision of tumour + reconstruction</td>
<td>CA + Radiotherapy</td>
</tr>
<tr>
<td>Chondrosarcoma</td>
<td>30-60</td>
<td>Flat bones, upper end of femur</td>
<td>Anywhere in the bone</td>
<td>Pain + Swelling +++ Duration - mths-yrs</td>
<td>Mottled calcification within the tumour</td>
<td>Osteosarcoma Chondroblasts, and cartilaginous matrix</td>
<td></td>
<td>Local ablation + radiotherapy</td>
</tr>
</tbody>
</table>

---

1. Ewing's tumour is the commonest malignant tumour of flat bones.
2. Ewing's tumour is the commonest malignant bone tumour which has multi-centric origin.
* Management of Surgical Causes of
  - Gall bladder
  - Bile ducts
  - Cholecystitis
  - Ca Head of Pancreas

* Splenomegaly
  - Rectum
  - Anus
  - Anal canal

Complication of Ulcers
  - Fistula pancreatic
  - Leakage
  - Hemorrhage
  - Infection
  - Trouble & Stomach emptying
  - Diarrhoea
  - Weight loss
  - Insulin resistance
Gall Stone Disease 1994

Risk Factors

- Metabolic causes: obesity, high cholesterol, diet, diabetes
- Infection: e. coli etc. of duodenum, bowel
- Bile stasis & BA pool: cirrhosis, pregnancy, ileal resection, obesity, malabsorption
- Hemolytic anemia
- Schull's triad: G. Stone, Diverticulosis, H. Icterus
- Parasites: Ascaris
- Cystic fibrosis

Types
- Cholesterol: 10%
- Mixed: 80%
- Brown stones
- Pigment stones

Complications (40%)

- In GI:
  - Sickle stone
  - Peptic ulcer dyspepsia
  - Colic
  - Acute cholecystitis
  - Chronic cholecystitis
  - Diverticulitis
  - Emphysema
  - Perforation
  - Tamari Syndrome

- In Intestine:
  - Obstructive jaundice
  - Choledochitis
  - White bile
  - Acute pancreatitis

- Obstruction
- Gall Stone
Acute Cholecystitis

- Calculous
- Acalculous
- Acute Emphysematous

Clinical

Murphy's sign, Roadd's sign = Hyperesthesia between 9-11 ribs posterolaterally
- Quivering rigidity
- Nausea

Investigation

Blood = Total WBC, B.S.

Radiology

X-ray = 40% opaque, rule out perforation, Mercedes Benz sign, seagull sign

USG = Stones, Posterior Acoustic Shadow, Murphy sign by USG probe

CT scan = if USG not clear

HIDA scan = Importance in Acalculous Cholecystitis

Notes of Dr. Ravindra Goswami (IAS-2015, AIR-153)
Tilt
↓
set up & experience

Excellent

Correction of comorbid cond (DM, HTN)

Early cholecystectomy

Good

- Conservative
- Aspiration
- Antispasmodic
- Antibiotics

After 6 wks
elective cholecystectomy

Emergency Cholecystectomy

Pseudoappendicitis - du DM, Hemolytic anemia, sepsis, Acalculous cholecystitis

Bacteremia

For empyema - Urgent laparotomy, aggressive antibiotics, putting a drain to drain pus as cholecystectomy
Tilt of Gall Stone.

Lap chole

Indication
- In functions of GB
- Young, thin, female
- Tiny stones

Draw backs
- Recurrence
- Life long

Oral
- UDCA, CDCA
- HMG CoA

Direct contact
- Methyl tert butyl ether
- Other

Notes of Dr. Ravindra Goswami (2015, AIR-153)
Causes of Obstructive Jaundice

1. Causes in the lumen - Stones, Ova, Cysts (Ascariis)
2. Causes in the wall - Periampullary Ca, bile duct stricture, Stenosis of sphincter of Oddi, choledochal cyst, Klatskin's tumour (Ca)
3. Causes from outside - Ca head of pancreas, chronic pancreatitis, LIV at porta hepatis

C/F:
1. Age - 30-50
2. Sex - F = M
3. Duration - Long
4. Symptoms -
   - Pain - Colicky (Intermittent)
   - Fever - Rarely (Intermittent)
   - Jaundice - Intermittent
   - Stool - Milk/Clay coloured
   - Nausea - Rare
   - Loss of appetite
   - Weight loss
5. Signs - Jaundice - Deep yellow
   - Anemia -
- Percendemen - Lives can be cured, with some nodular than secondaries.

- CT = Contracted, palpable yes.

- Convolviers fallacy = Double: one stone in CBD, and one in cystic duct.

- Pancreatic = CA in previously cholezystitis patient.

- Pt. Oriental cholangihepatitis causing GB stone (GB remains normal).

If cholangitis had developed then Berynettes' A Me may be seen: Periesterict pain, cholelithiasis (T/T), periasm, persistent jaundice, Shock, acetazolamide status.

- Antibiotics T/T of CBD stones.
**Dr. Ravindra Goswami (IAS-2015, AIR-153)**

**Symptoms of Obstructive Jaundice**

1. **Blood** - HB, TLC, DLC, (BT, CT, PTINR)
   - Serum alkaline phosphatase
   - Gross elevation

- Obstructive Jaundice
- Biliary cirrhosis
- Chole disease

2. **Radiology**
   - **USG** - Stones, mass, secondary
   - **CECT** - To decide the operability of CA Head or periampullary
     - Obliteration of fat plane
     - b/w mass and sup mes vessels
     - But cannot differentiate b/w CA Head and chronic pancreatitis's
     - PET scan can come out
   - **MRCP** - IOC DUS obstructive jaundice

3. **Endoscopic**
   - **Endoscopy** - For direct visualisation
     - periampullary CA - Ulceration and Biopsy is taken
     - Stone is behind II part of duo - Bridge can be seen

   - **ERCP** - Ampulla of vater is cannulated and cholangi and pancreatic system is visualised

   - **Interpretation**
     - **Stone** - Filling defect
     - **Periampullary CA** - Irregular filling defect
     - **Pancreatitis** - Chain of lake appear
Uses of ERCP
- Stone extraction
- Dilation of stone by lithotriptor
- Sphincterotomy
- Stenting in cholangitis, Obs. Jaundice
- Malignancy
- Stricture
- Selected Chronic Pancreatitis

Complications: cholangitis

(2) Endoscopic: Stones, Mass, Guided FNAC

Percutaneous PTC: For dx of duodenum
and ERCP failure cases

In Klatkin's tumour

Dx by laparoscopy
T/T
Preoperative Preparation
- Fluid and electrolyte correction
- Inj. Vit K for 3 days or FFP
- Antibiotics
- Blood transfusion

T/T as per case etiology

CBD Stones
- Cholecystectomy
- O + C
  - Filling defect
    - On proven CBD stones
    - H/P Cholecystitis
  - Palpable stones in CBD

Supraduodenal
Choleccholithotomy
- Exploration for stone
- T-Tube insertion for 8-10 days
- Repeated cholangiogram
- Removal

Cholecdocholithotomy
- If CBD caliber > 1.5 cm
  - Recurrent stone
  - Multiple intrahepatic stones

Cholecdochoduodenostomy
2. Preoperative ERCP, sphincterotomy, extraction of stones followed by cap. choled. SC currently. 844

3. T/t of periampullary CA
   - Whipple's
   - Partial gastrectomy
   - Loop of duodenum up to DJ flexure + proximal Jejunum
   - Head of pancreas up to neck

Followed by EJ Anastomosis

4. pylorus preserving PDectomy

3. For palliation - Triple bypass
   - EJ
   - Entero-enterostomy
   - GJ
   (To prevent food particles to enter in duodenum)
Cystic Neoplasm of Pancreas

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>SCN</th>
<th>MCN</th>
<th>IPMN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>F &gt;&gt; M♂ (4:1)</td>
<td>F &gt;&gt; M♂ (10:1)</td>
<td>F = M♂</td>
</tr>
<tr>
<td>Age (years)</td>
<td>60-70</td>
<td>50-60</td>
<td>60-70</td>
</tr>
<tr>
<td>Location</td>
<td>Head♂</td>
<td>Body and tail♂</td>
<td>Head♂</td>
</tr>
<tr>
<td>Appearance</td>
<td>Multiple small cysts (microcyst) separated by internal septations with central sunburst calcifications♂</td>
<td>Thick-walled, septated macrocyst♀ with smooth contour; ± solid component, egg-shell calcifications♀</td>
<td>Poorly demarcated, lobulated, polycystic mass with dilation of main or branch ducts♀</td>
</tr>
<tr>
<td>Communication with ducts</td>
<td>No</td>
<td>No</td>
<td>Yes♀</td>
</tr>
<tr>
<td>Cytology</td>
<td>Scant glycogen-rich cells, with positive Periodic Acid Schiff stain♂</td>
<td>Sheets and clusters of columnar, mucin-containing cells</td>
<td>Tail, columnar mucin-containing cells</td>
</tr>
<tr>
<td>Mucin stain</td>
<td>Negative</td>
<td>Positive♂</td>
<td>Positive♀</td>
</tr>
<tr>
<td>Amylase</td>
<td>Low</td>
<td>Low</td>
<td>High♂</td>
</tr>
<tr>
<td>CEA</td>
<td>Low</td>
<td>High♂</td>
<td>High♂</td>
</tr>
</tbody>
</table>

CARCINOMA PANCREAS ETIOLOGY AND RISK FACTORS

108. Ans. a. Her-2-neu (Ref: Sabiston 19/e p1535-1536; Schwartz 9/e p1219-1220; Bailey 26/e p1137, 25/e p1148-1152; Blumgart 5/e p912-918; Shackelford 7/e p1187-1189)
K-ras mutations and HER2/neu over expression are the earliest changes to occur in pancreatic carcinoma.

**Risk Factors for Pancreatic Carcinoma**

- There is association between risk of pancreatic cancer, H. pylori colonization, and ABO blood groups.
- Older age, African American race, low socioeconomic status, Ashkenazi Jewish heritage are associated with increased risk of pancreatic cancer.
- Host etiologic factors associated with increased risk of pancreatic cancer include history of diabetes mellitus, chronic cirrhosis and pancreatitis, a high fat or cholesterol diet, and prior cholecystectomy.

### Familial Pancreatic Cancer

- **Predisposing Conditions:** Hereditary pancreatitis, HNPCC, Hereditary Breast Cancer associated with the BRCA2 mutation, Ataxia Telangiectasia, FAMMM, and Peutz-Jegher's syndrome. (H3-AFP)
- BRCA-2 mutation is the most common mutation in patients with hereditary pancreatic cancer.
- K-ras is the most common mutation in patients with carcinoma pancreas.
- Peutz-Jegher's syndrome carries the highest relative risk of pancreatic cancer.
- Patients with pancreatic cancer with DNA mismatch repair mutations have a better prognosis.
- K-ras mutations and HER2/neu over expression are the earliest changes to occur.

### Risk Factors for Pancreatic Carcinoma

<table>
<thead>
<tr>
<th>Established</th>
<th>Associated</th>
<th>Possible</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tobacco</td>
<td>Chronic pancreatitis</td>
<td>Physical inactivity</td>
</tr>
<tr>
<td>Inherited susceptibility</td>
<td>Diabetes mellitus T2D</td>
<td>Certain pesticides</td>
</tr>
<tr>
<td></td>
<td>Obesity</td>
<td>High carbohydrate/sugar intake</td>
</tr>
</tbody>
</table>

### Genetic Mutation in Pancreatic Cancer

<table>
<thead>
<tr>
<th>Gene</th>
<th>Pancreatic Cancer %</th>
</tr>
</thead>
<tbody>
<tr>
<td>p16</td>
<td>82</td>
</tr>
<tr>
<td>K-ras</td>
<td>95-100 (MC)</td>
</tr>
<tr>
<td>p53</td>
<td>75</td>
</tr>
<tr>
<td>DPC4</td>
<td>55</td>
</tr>
<tr>
<td>BRCA2</td>
<td>7</td>
</tr>
</tbody>
</table>

### Predisposing Conditions for Familial Pancreatic Cancer (H-AFP)

- Hereditary pancreatitis
- HNPCC
- Hereditary Breast Cancer associated with the BRCA2 mutation
- Ataxia Telangiectasia
- FAMMM (Familial atypical multiple mole melanoma) syndrome
- Peutz-Jegher's syndrome

109. **Ans. a. Acute pancreatitis** (Ref: Sabiston 19/e p1535-1536; Schwartz 9/e p1219-1220; Bailey 26/e p1137, 25/e p1149; Blumgart 5/e p912-918; Shackelford 7/e p1188)

110. **Ans. a. K-ras** (Ref: Sabiston 19/e p1537; Schwartz 9/e p1220; Bailey 26/e p1137, 25/e p1149; Blumgart 5/e p884; Shackelford 7/e p1188)

111. **Ans. d. Cronkhite-Canada syndrome**

112. **Ans. d. FAP**

113. **Ans. b. K-ras**

114. **Ans. d. History of partial gastrectomy**

115. **Ans. a. Urgent weight reduction, b. Strict vegetarian diet, c. Stop alcohol, e. Stop cigarette smoking**

116. **Ans. c. BRCA activated**
### Pancreas Clinical Features and Diagnosis

19. **Anus. c. Jaundice** (Ref: Sabiston 19/c p1535-1544; Schwartz 9/e p1220-1225; Bailey 26/e p1137, 25/e p1149-1152; Blumgart 5/e p919-925; Shackelford 7/e p1190-1196)

#### Carcinoma Pancreas

- **MC type** is pancreatic ductal adenocarcinoma (PDAC).
- More common in Men, African Americans, mean age at diagnosis is 72 years.
- Overall, <5% of individuals will survive 5 years beyond their diagnosis.

  - **Association between risk of pancreatic cancer, H. pylori colonization, and ABO blood groups**
  - **Established risk factors:** Smoking (Tobacco) and Inherited susceptibility.

- **Hereditary risk factors:** Hereditary pancreatitis, HNFCC, Hereditary Breast Cancer associated with the BRCA2 mutation, Ataxia Telangiectasia, FAMMM, and Peutz-Jegher's syndrome. (H3-AFP)

  - K-RAS2 oncogene is activated (by point mutation) in >95% of pancreatic cancers (MC gene mutation).

#### Pathology

- Macroscopically, ductal adenocarcinoma is a scirrhous (scar forming) type of carcinoma.
- It is associated with abundant desmoplastic stroma, in which the neoplastic glands are widely scattered.

#### Clinical Features

- **MC symptom** for patients with PDACs in the peripancreatic region is jaundice.
- Pain typically arising in the epigastrium and radiating to the back.
- **Weight loss** affecting more than 50% of individuals.
- For tumors of the body and tail of the pancreas, pain and weight loss become more common at presentation.
- A palpable distended gallbladder in 1/3 of patients with peripancreatic PDAC (Courvoiser Law).
- With widespread disease, a left supraclavicular node (Virchow's node) may be palpable. Periumbilical lymphadenopathy may be palpable (Sister Mary Joseph's node).
- In cases of peritoneal dissemination, perirectal tumor involvement may be palpable via digital rectal examination, referred to as Blumer's shelf.

#### Table: Autosomal Dominant, Autosomal Recessive, X-Linked Disorders

<table>
<thead>
<tr>
<th>Autosomal Dominant</th>
<th>Autosomal Recessive</th>
<th>X-Linked Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Familial hypercholesterolemia</td>
<td>Deafness</td>
<td>Hemophilia A&lt;sup&gt;a&lt;/sup&gt; (recessive)</td>
</tr>
<tr>
<td>HNPCC</td>
<td>Albinism&lt;sup&gt;a&lt;/sup&gt;</td>
<td>G6PD deficiency&lt;sup&gt;a&lt;/sup&gt; (recessive)</td>
</tr>
<tr>
<td>FAP&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Wilson's disease&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Duchenne/Becker muscular dystrophy&lt;sup&gt;a&lt;/sup&gt; (recessive)</td>
</tr>
<tr>
<td>BRCA1 and BRCA2 breast cancer</td>
<td>Hemochromatosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Fabry's disease</td>
</tr>
<tr>
<td>Hereditary hemorrhagic telangiectasia</td>
<td>Sickle cell anemia&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Ocular albinism</td>
</tr>
<tr>
<td>Marfan's syndrome&lt;sup&gt;a&lt;/sup&gt;</td>
<td>beta thalassemia&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Testicular feminization</td>
</tr>
<tr>
<td>Hereditary spherocytosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Cystic fibrosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Chronic granulomatous disease</td>
</tr>
<tr>
<td>Adult polycystic kidney disease</td>
<td>Hereditary emphysema (α, antitrypsin deficiency)</td>
<td>Hypophosphatemic rickets&lt;sup&gt;a&lt;/sup&gt; (dominant)</td>
</tr>
<tr>
<td>Huntington's chorea&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Homocystinuria&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Fragile-X syndrome&lt;sup&gt;a&lt;/sup&gt; (recessive)</td>
</tr>
<tr>
<td>Acute intermittent porphyria&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Friedreich's ataxia&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Color blindness&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
<tr>
<td>Ostogeneisis imperfecta&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Phenylketonuria&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
</tr>
<tr>
<td>von Willebrand's disease&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Fanconi's Syndrome</td>
<td></td>
</tr>
<tr>
<td>Myotonic dystrophy&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Gaucher's Disease</td>
<td></td>
</tr>
<tr>
<td>Familial hypertrophic cardiomyopathy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurofibromatosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tuberous sclerosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Osteopetrosis&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Achondroplasia&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
**Presenting Symptoms of Periampullary Tumors**

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaundice (75%)§</td>
<td>Pruritus (11%)</td>
</tr>
<tr>
<td>Weight loss (51%)§</td>
<td>Fever (3%)</td>
</tr>
<tr>
<td>Abdominal pain (39%)</td>
<td>Gastrointestinal bleeding (1%)</td>
</tr>
<tr>
<td>Nausea/vomiting (13%)</td>
<td></td>
</tr>
</tbody>
</table>

**Diagnosis**

- Tumor markers: CA19-9 (most sensitive)§ and CEA.
- Individuals with blood Lewis antigen-negative status (10-15%) do not develop elevation of the CA19-9.
- MDCT is investigation of choice for the evaluation of lesions arising in the pancreas.

MDCT for suspected periampullary pathology, a three-phase (noncontrast, arterial, and portal venous) CT scan with 3-mm slices and coronal and three-dimensional reconstruction should be routine.

- ERCP: Reserved for cases requiring therapeutic or palliative intervention.
- Double duct sign on ERCP is highly suggestive of pancreatic head cancer.
- EUS: For identifying lesions <2 cm that do not appear on CT scans.
- Tissue diagnosis is not necessary prior to routine resection.
- A suspicious lesion by imaging should be treated with resection.

**Treatment**

- Surgical resection remains the only potentially curative treatment of pancreas cancer.

**Tumors of head of the pancreas**
- pylorus preserving pancreaticoduodenectomy or Langner-Traverso procedure is preferred.

**Tumors of body and tail of the pancreas**
- Distal pancreatectomy and en-bloc splenectomy.

- MC complication of pancreaticoduodenectomy is delayed gastric emptying.
- MC cause of death following pancreaticoduodenectomy is cardiopulmonary complications.
- Most important predictor of post-operative survival is R0 resection.
- Most important margin in pancreaticoduodenectomy is retroperitoneal or uncinate margin.

**Palliative Therapy for Pancreatic Cancer**

- **Biliary obstruction**
  - ERCP with metal stent placement (Best)§
  - Roux-en-Y hepaticojejunostomy

- **Gastric outlet obstruction**
  - Endoscopic stenting (Preferred)§
  - Double bypass (Roux-en-Y hepaticojejunostomy + gastrojejunostomy)
  - NSAIDs or opiates§
  - Celliac nerve block

**Chemotherapy**

- Gemcitabine§ is currently the standard of care for patients with metastatic pancreatic cancer.

**Prognosis**

- Five year survival after curative resection (pancreaticoduodenectomy) approaches 15-20%§.
- Overall, 5 year survival rate with pancreatic cancer is 5%§.

**Median survival in Carcinoma Pancreas**

<table>
<thead>
<tr>
<th>Disease Type</th>
<th>Median Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Resectable disease (stage I and II)</td>
<td>15-20 months§</td>
</tr>
<tr>
<td>Locally advanced disease (stage III)</td>
<td>6-10 months§</td>
</tr>
<tr>
<td>Metastatic disease (stage IV)</td>
<td>3-6 months§</td>
</tr>
</tbody>
</table>

- But in specific patients, a tissue diagnosis may be needed such as in patients entering a clinical trial, prior to neoadjuvant chemotherapy, and prior to chemotherapy in advanced tumors. In these patients, an EUS is highly accurate.
139. Cause of hemobilia are all except: (AIIMS June 2000)
   a. Trauma to abdomen
   b. Malignancy
   c. Rupture of hepatic artery aneurysm
   d. Hepatitis

140. All are seen in hemobilia except: (PGI Dec 99)
   a. Shock
   b. Colicky pain
   c. Melena
   d. Jaundice

141. Triad of hemobilia includes all, except: (NEET Pattern, AIIMS June 93)
   a. Pain
   b. Fever
   c. G.I. bleeding
   d. Jaundice

142. True regarding hemobilia: (DPG 2007)
   a. Triad of jaundice, pain, melena
   b. MC cause- rupture of portal vein into biliary system
   c. MR angiography is the IOC
   d. None of the above

143. All are true about hemobilia except: (PGI SS June 2006)
   a. Pain abdomen
   b. Melena
   c. Jaundice
   d. Hypoglycemia

BILHEMIA

144. Best investigation for bilhemia is: (JIPMER GIS 2011)
   a. ERCP
   b. MRCP
   c. CT
   d. EUS

145. Investigation of choice for bilhemia: (ILBS 2011)
   a. CT angiography
   b. MR angiography
   c. EUS
   d. ERCP

146. All are true about bilhemia except: (AIIMS GIS May 2008)
   a. Biliary pressure/portal pressure
   b. Diagnosed by ERCP
   c. Death due to embolism of bile in lungs
   d. Patient has hyperbilirubinemia with raised enzymes

MISCELLANEOUS

147. False statement about common bile duct: (PGI May 2011)
   a. Lies in free margin of lesser omentum
   b. Anterior to first part of duodenum
   c. Right to hepatic artery
   d. Anterior to portal vein
   e. Open in second part of duodenum

148. True regarding common bile duct is all except: (All India 2000)
   a. Opens 10 cm distal to pylorus
   b. Lies anterior to I.V.C
   c. Portal vein lies posterior to it
   d. Usually opens into duodenum separate from the main pancreatic duct

149. Which of the following statement is true regarding the relation of bile duct? (PGI Dec 2005)
   a. Posteriorly related to 1st part of duodenum
   b. Related posteriorly to the tunnel of pancreatic head
   c. Anteriorly related to 1st part of duodenum
   d. Related to IVC posteriorly
   e. It lies left to hepatic artery in the free border of lesser omentum

150. Predominant blood supply to the supraduodenal bile duct is derived from: (All India 2012)
   a. Vessels that run upward along the bile duct from the duodenal end of the duct such as the retro-duodenal and gastroduodenal arteries
   b. Vessels that run downward along the bile duct from the hepatic end of the duct such as the right hepatic artery
   c. Vessels that arise from the hepatic artery proper run up along the CBD and supplies it with twigs in non-axial distribution
   d. Vessels that arise from the cystic artery

151. All of the following conditions are associated with pneumobilia except:
   a. Sphincterotomy
   b. Rupture of hydatid cyst
   c. Mirizzi's syndrome
   d. Gall stone ileus

152. Pneumobilia is seen in:
   a. Gall stone ileus
   b. Mirizzi's syndrome
   c. TPN
   d. Volvulus

153. Normal length of CBD is:
   a. 7 cm
   b. 5 cm
   c. 3 cm
   d. 2 cm

154. Which of the following is true regarding the principle of use of MRCP? (AIIMS Nov 2012)
   a. Intraluminal dye is used to create the three dimension view of the structures
   b. Dye is instilled percutaneously first then MRI is used
   c. Use of heavily T2-weighted image without contrast to create the three dimensional image of the biliary tree using MIP algorithm
   d. Use of systemic Gadolinium as a contrast agent to create the three dimensional image of the biliary tree
**CHOLEDODHAL CYST**

1. Ans. c. IVa (Ref: Sabiston 19/e p1503-1505; Schwartz 9/e p1155; Bailey 26/e p1106, 25/e p1119; Blumgart 5/e p707-719; Shackelford 7/e p1397)

**Etiology**

- Most widely accepted hypothesis: Abnormal pancreaticobiliary ductal junction (APBD)\(^\text{Q}\)
- APBD results in reflux of pancreatic fluid into the distal common hepatic duct and results in mucosal injury, chronic inflammation, and weakening of the bile duct wall\(^\text{Q}\).
### Classification

- Bile duct cysts are classified on the basis of site, extent, and shape of the cystic anomaly of the ductal system.
- MC choledochal cyst: Type I > Type IV > Type III (44%)^q.

<table>
<thead>
<tr>
<th>Type</th>
<th>Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Dilatation of the extrahepatic biliary tree</td>
</tr>
<tr>
<td>II</td>
<td>Diverticular dilatation^q of the extrahepatic biliary tree</td>
</tr>
<tr>
<td>III</td>
<td>Cystic dilatation of the intraduodenal portion of the common bile duct (choledochocoele)^q</td>
</tr>
<tr>
<td>IVA</td>
<td>Dilatation of the extrahepatic and intrhepatic biliary tree^q</td>
</tr>
<tr>
<td>IVB</td>
<td>Dilatation of multiple sections of the extrahepatic bile ducts^q</td>
</tr>
<tr>
<td>V</td>
<td>Dilatation confined to the intrahepatic bile ducts (Carol's disease)^q</td>
</tr>
</tbody>
</table>

### Clinical Features

- Classic triad: Pain, jaundice (intermittent) and abdominal mass (10%)^q.
  - MC symptom in Infants: Jaundice (in 80%)^q
  - MC symptom in patients >2 years of age: Abdominal pain^q

- In children, the major clinical symptoms are recurrent abdominal pain (61.6%), nausea and vomiting (65.5%), mild jaundice (43.6%), an abdominal mass (29.0%), and fever (29.0%).
- In adults, abdominal pain (89%) and jaundice (42%) are present frequently. Less common clinical findings include nausea (29%), cholangitis (26%), pancreatitis (25%), and an abdominal mass (13%).

### Diagnosis

- ERCP: Most useful in defining the distal ductal anatomy and the presence of APBD/
- PTC: Useful in defining the proximal ductal anatomy and the presence of intrahepatic disease.

### Treatment of Choledochal Cyst

<table>
<thead>
<tr>
<th>Type</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Roux-en-Y hepaticojejunostomy^q</td>
</tr>
<tr>
<td>II</td>
<td>Excision with T-tube repair^q</td>
</tr>
<tr>
<td>III</td>
<td>Endoscopic sphincterotomy and cyst unroofing^q</td>
</tr>
<tr>
<td>IVA</td>
<td>Hepatic resection for localized disease^q</td>
</tr>
<tr>
<td>IVB</td>
<td>Liver transplantation for diffuse disease^q</td>
</tr>
<tr>
<td>V</td>
<td>Hepatic resection for localized disease^q</td>
</tr>
<tr>
<td></td>
<td>Liver transplantation for diffuse disease^q</td>
</tr>
</tbody>
</table>
Lilly technique: Serosal surface of the duct is left adhering to the portal vein, while the mucosa of the cyst wall is obliterated by curettage or cautery, when cyst is densely adhered to the portal vein secondary to long-standing inflammatory reaction.

In this situation, a complete, full-thickness excision of the cyst may not be possible.

2. Ans. d. Type V choledochal cyst (Ref: Sabiston 19/e p1504-1505; Schwartz 9/e p1119; Bailey 26/e p1106, 25/e p1119; Blumgart 5/e p713; Shackelford 7/e p1402-1403)

**CAROLI'S DISEASE (TYPE V CHOLEDOCHAL CYST)**

- Congenital malformation, consists of multiple sacular dilatations limited to the intrahepatic bile ducts (segmental bile ducts).
- About half the cases are associated with congenital hepatic fibrosis (affect interlobular bile ducts).
- Cyst with congenital hepatic fibrosis is known as Grumbach's disease.
- Portal hypertension is present in Caroli’s disease associated with congenital hepatic fibrosis.

**Clinical Features**

- Symptoms include cholangitis (64%), portal hypertension (22%), and abdominal pain (18%)
- More common in males
- Septa containing portal veins protrude into the lumen of the ectatic bile ducts (central dot sign).
- The main and often the only symptom of bacterial cholangitis secondary to Caroli’s disease is fever without abdominal pain and jaundice.
- Frequent episodes of cholangitis indicates poor prognosis.
- Most stones are pigmented in Caroli’s disease.

**Diagnosis**

- CT findings: Portal vein radicals can be seen after enhancement within dilated intra-hepatic bile ducts (central dot sign).

**Treatment**

- Hepatic resection for localized disease
- Liver transplantation for diffuse disease.

3. Ans. b. Extra hepatic duct dilatation
4. Ans. a. Caroli’s disease
5. Ans. c. Type IVa = choledochocele
6. Ans. a. Type I
7. Ans. b. Type II
8. Ans. a. Excision of cyst
9. Ans. c. Type III
10. Ans. a. Resection decreases the incidence of malignancy but risk persists (Ref: Sabiston 19/e p1505; Bailey 26/e p1106, 25/e p1119; Blumgart 5/e p713; Shackelford 7/e p1339)