ECTOPIA VESICA

1. Ans. d. Iliac bone (Ref: Smith 17/e p574-575; Campbell's 10/e p3221-3236; Bailey 26/e p1310, 25/e p1314)
   Posterior iliac osteotomy is done in ectopia vesicae.

   EXTRPHY OF BLADDER (ECTOPIA VESICA)

   - Exrophy of bladder is complete ventral defect of Urogenital sinus and the overlying skeletal system.
   - Defect in the infraumbilical part of the anterior abdominal wall, associated with incomplete development of the anterior wall of the bladder.

   Embryology
   - The basic defect is abnormal overdevelopment of the cloacal membrane and its rupture.
   - The timing of this rupture of this defective cloacal membrane determines the variant of the exophytic-epispadias complex that results.

   Clinical Features
   - The posterior wall of the bladder protrudes through the defect with mucosal edges fused with skin and urine spurs onto the abdominal wall from the ureteral orifices.
   - The rectus muscles which are inserted on the pubic rami are also widely separated.
   - An umbilical hernia though usually small is present along with exostic bladder.
     - In males, complete epispadias with a wide and shallow scrotum. Undescended testis and inguinal hernias are common.
     - Females also have epispadias with bifid clitoris and wide separation of the labia.
   - The anus is dislocated anteriorly in both sexes and there may be rectal prolapse.

   Complications
   - The consequences of untreated bladder exrophy are total urinary incontinence and an increased incidence of bladder cancer, usually adenocarcinoma.
   - Many untreated exropy of bladder reveal fibrosis, derangement of muscularis mucosa and chronic infection leading to hydronephrosis.

   Treatment
   - Enterocystoplasty is the method of choice to augment bladder capacity and aid in reservoir function.
   - Urinary diversion with cystectomy is treatment of choice for small, fibrotic or inelastic bladder.
   - Complete reconstruction is achieved by:
     - Bladder closure with sacral osteotomy and lengthening of penis (Posterior iliac osteotomy is done in ectopia vesicae)
     - Antiretreal reflux procedure with bladder neck reconstruction
     - Repair of epispadiac penis

2. Ans. a. Hypospadias
3. Ans. b. Cloacal membrane is present
4. Ans. c. Hypospadias
5. Ans. b. Ventral curvature of penis
6. Ans. b. Iliac bone

URINARY BLADDER STONES

7. Ans. b. Uric acid stones are dropped from above (Ref: Smith 17/e p272-273; Campbell 10/e p2521-2527; Bailey 26/e p1320-1322, 25/e p1323-1325)
Special
- Bil. Stones = kidney with better function first,
2 weeks later the other.

Stone & Pyonephrosis = Nephrostomy → Drainage

After clearance of pus
Assessment of kidney functions
Non-function → Nephrectomy
(EWL, PCNL, open)

Ureteric Stones
See in Book notes

- Upper: ESWL or flexible ureteroscopy & lasers
- Middle: ESWL or Basketing or Ureterolithotomy
- Lower: Ureteroscopic removal or lasers, ULTH
- Vesico-Ureteric: Ureteroscopic removal or endoscopic meatotomy
- Impacted = Open Ureterolithotomy.
Renal mass

- Moves with respiration
- Enlarged in upward and downward direction
- Bimanually palpable and ballottable
- Obtain caution of colonic resonance
- Upper border, not palpable.

1. PCKS
   - ESR: manifest at 40 yrs, not at child
     - Dull pain
     - Hematuria, HTN, B/L Renal mass
   - Im = IVU = Spider leg deformity.

   More

   Asymptomatic
   - Follow up
   - Drugs - Antihyp.
   - Nephrectomy
   - Transplant

   E HTN

   Injected cyst or
   Pyelonephritis
   - Antibiotics
   - Aspiration of cyst

   Renal failure
   - Dialysis
   - Transplant

2. Hydrenephrosis
   - Aseptic dilation of renal or
   - Part of the pelvi-calyceal system.

Causes

   Child: Phimosis, meatal stenosis, PUV, B/L VU, Reflux

   Young adult: Stenosis, reflux, B/L aberrant vessels

   Middle age: BPH, bladder neck contracture, Ormondial

   Physiological: Pregnancy
4. Therapeutic embolisation
   - This can be used as a palliative measure in advanced carcinoma to relieve symptoms. This can also be used preoperatively to regresses the size of the large tumour.
   - A catheter is placed in the renal artery and substances such as gel foam, blood clot, crushed muscle are injected.
   - They block the lumen of the vessel and reduce size of the tumour so that radical nephrectomy can be undertaken later.

5. Radiotherapy
   - Not of much use. However, it is a good form of palliation for secondaries in the lung, bone and brain.

6. Immunotherapy
   - Administration of interferon or interleukin-2 has been found to improve the survival rate.

RENAL MASS IN SURGICAL WARD (Table 39.2)

Clinical features of kidney mass
   - Moves with respiration because the fascia of Gerota encloses the kidney and fuses above with the diaphragm.
   - Kidneys enlarge in the upward and downward direction.
   - Bi-laterally palpable and ballotable. It is ballotable because of renal pedicle and peri-ren al pad of fat.
   - Colonic band of resonance is obliterated when the kidney enlarges, as the colon is pushed laterally.
   - Upper border is not palpable because it is under the 12th rib.

<table>
<thead>
<tr>
<th>Table 39.2: Renal mass in surgical ward</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hypernephroma</strong></td>
</tr>
<tr>
<td>Chief symptoms: Hb, pain in loin, renal mass</td>
</tr>
<tr>
<td>Age of the patient: Over 50 years</td>
</tr>
<tr>
<td>Sex incidence: Common in males</td>
</tr>
<tr>
<td>Anaemia: Present</td>
</tr>
<tr>
<td>Features of renal failure: Absent</td>
</tr>
<tr>
<td>Renal mass: Unilateral, nodular, hard, may be fixed, nontender</td>
</tr>
<tr>
<td>Features of kidney mass: May not have free mobility due to fixation</td>
</tr>
<tr>
<td>IVU: Irregular calyces</td>
</tr>
<tr>
<td>CT Scan: Enhancing mass</td>
</tr>
<tr>
<td>Treatment: Radical nephrectomy</td>
</tr>
</tbody>
</table>

| **Hydronephrosis**                      |
| Asymptomatic, distension, abdomen pain |
| 20–30 years |
| Common in females |
| Absent |
| Can be present in bilateral cases (rare) |
| Can be bilateral, smooth, cystic, feels firm |
| Not fixed, nontender |
| Irregular calyces |
| CT Scan: Enhancing mass |
| Treatment: Percutaneous nephrostomy, nephrostomy-renal transplantation |

| **Polycystic kidney**                   |
| Mass abdomen, hypertension, haematuria |
| 30–40 years |
| Common in females |
| May be present |
| Bilateral, bossed, nodular, not fixed, nontender |
| Present |

PYONEPHROSIS

In this condition, the entire kidney is converted into a sac containing pus or purulent urine—almost always the renal parenchyma is destroyed totally.

Causes

1. Renal calculous disease is the most common cause of pyonephrosis.
2. Acute pyelonephritis is more common in children and in females. Inadequately treated cases may develop into pyonephrosis, especially when pyelonephritis is associated with urinary tract obstruction.
3. Infection of a hydronephrosis.

Clinical features
   - Anaemia and fever
   - Renal swelling
   - Large swelling with high grade fever with chills and rigors suggest an imminent danger of sepsis and calls for immediate drainage of the pus.

Investigations
   - Urine examination may be positive for coliforms and other gram-negative organisms.
   - Plain X-ray KUB may reveal a stone or an enlarged renal outline.
   - Ultrasound can confirm hydronephrosis.
Intravenous urogram demonstrates poor function of the kidney on the diseased side. As a rule the opposite kidney is normal.

**Treatment**

- Broad-spectrum antibiotics (parenteral) should be started immediately once the urine and blood is sent for culture and sensitivity.
- Ultrasound-guided aspiration of pus or a percutaneous nephrostomy (better), and drainage of pus greatly improves the general condition of the patient.
- If any obstruction or causative agent such as a stone is found, it should be removed.
- Nephrectomy should be considered if the kidney is non-functioning with significant damage.

**PERINEPHRIC ABSCESS**

It refers to the collection of pus in the perirenal area.

**Causes**

- Infection in a perirenal haematoma
- Pyonephrosis when it ruptures
- Tubercular perinephric abscess
- Pus from retrocecal appendix can extend into loin, perinephric area and may present as abscess.

**Clinical features**

- High swinging temperature
- Rigidity, tenderness, fullness in the loin
- Oedema in the loin

**Investigations**

- Total count: Raised above 20,000 cells/mm³
- Urine analysis: No organisms are usually found

- X-ray spine: Scoliosis with concavity towards abscess
- Screening chest: Diaphragm is immobile and elevated on the diseased side.

**Treatment**

- Pus is drained by an incision in the loin, breaking all the loculi.
- Dialysis and renal transplantation are discussed in page 1111.

**MISCELLANEOUS**

**INTERESTING MOST COMMON FOR RENAL CELL CARCINOMA**

- Most common renal cancer in adults are adenocarcinomas.
- Most common site is upper pole of the kidney.
- Most common presentation of renal cell carcinoma is with mass abdomen.
- Most common investigation of choice is contrast enhanced CT scan.
- Most common method of spread is by haematogenous route.
- Most common intra-abdominal malignancy which spreads within vena cava and into atrium is renal cell carcinoma.
- Most common cell of origin is proximal renal tubular epithelium.

**WHAT IS NEW IN THIS CHAPTER? RECENT ADVANCES**

- All the topics have been updated.
- New photographs and key boxes have been added.

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**MULTIPLE CHOICE QUESTIONS**

1. Following is not the feature of adult polycystic kidney:
   A. It can give rise to renal failure
   B. Hypertension is seen in about 75% patients
   C. It is autosomal recessive
   D. It is always bilateral

2. Relationships of right kidney include following except:
   A. Posteriorly are muscles
   B. Anteriorly pyloric antrum
   C. Lateral ascending colon
   D. Medial adrenals

3. Which of the following is the feature of horseshoe kidney?
   A. Classically it is the upper polar fusion of both kidney
   B. It does not cause angulation of the ureter causing hydronephrosis
   C. It can be associated with Down's syndrome
   D. Hyperextension of the spine results in pain, nausea and vomiting

4. Gout results in:
   A. Calcium stones
   B. Cystine calculi
   C. Phosphate stones
   D. Uric acid stones
GFR = Patchless Renal mass
- Slight pain in the loin
- Rarely HTN, hematuria
- Dietl's crisis - in Calculous hydronephrosis

Investigations:
- X-ray, USG, CT Scan
- Pyelogram
- DTPA scan - to know the anatomy
- Percussion
- Retrograde pyelography

T/T:
- Treat the cause
- For treatment of hydronephrosis: particularly
  - Non-functioning kidney with thin cortex - Nephrectomy
  - If cortex > 0.5 cm -> Nephrectomy
    - Decompress the system
    - Reassessment
      - Improved
      - Not improved
        - Definitive sx - Nephrectomy

For B12 -> Better kidney first

For Congenital hydronephrosis:
- Mild = 11-20 mm
- Moderate = 21-35 mm
- Severe = >35 mm

= Conservative & Medical monitoring
= Anderson Hynes pyeloplasty
renal neoplasms

Benign
- Adenoma
- Papilloma
- Hemangioma

Malignant
- Nephroblastoma
- RCC
- TCC
- SCC

1. Nephroblastoma (WT) ⇒ Epithelial and mesothelial components

- Y/F = Female > Male, 2-4 years
- Abdominal mass (51% = Neuroblastoma)
- Hematuria (if present is bad sign)
- Low grade fever

51% = Neuroblastoma
Adrenal tumours, retroperitoneal tumour

Investigations:
- USG, CT, IVP, PNAC

WT vs. Neuroblastoma

Calcification
- Intraspinal extension
- Aorta & IVC involvement

Location
- Intrarenal
- Above kidney

Midline Cross
- 4V4/VMA

Less

More

Dumbbell tumour
WILM’S TUMOR

124. Ans. b. Abdominal lump (Ref: Decita 9/e p1766-1769; Nelson 18/e p2140; Smith 17/e p339-343; Campbell 10/e p3714-3722; Bailey 26/e p1304.

WILM’S TUMOR

- Wilm’s tumor: MC primary renal tumor of childhood (2-5 years);
- Wilm’s tumor: 2nd MC malignant abdominal tumor in children (MC is neuroblastoma);
- Arise from kidney, composed of three elements- blastema, epithelium and stroma;
- MC presenting feature is asymptomatic abdominal mass or swelling;
- Mostly unilateral;
- Characterized by triad of abdominal mass, fever and microscopic hematuria;
- Fever typically resolve after tumor resection.

Associated peseformations
- WAGR Syndrome: It consists of aniridia, genital anomalies and mental retardation. The risk of Wilm’s tumor is increased by 5x in this syndrome.
- Denys-Drash Syndrome: It consists of gonadal dysgenesis (Male pseudohermaphroditism), nephropathy leading to renal failure. Majority of patients with this syndrome have renal failure.
- Beckwith-Wiedemann Syndrome: It consists of enlargement of body organs, hemi-hypertrophy, renal medullary cysts and abnormal large cells in adrenal cortex, macroglossia, omphalocele, hepatoblastoma.

Diagnosis
- USG (first investigation) or CT abdomen for staging.
- MRI is superior to other imaging modalities in delineating nephroblastomatosis elements.
- Calcification tends to be more crescent shaped, discrete and peripheral in comparison of finely stippled calcification of neuroblastoma.

Treatment
- Surgical excision (transperitoneal radical nephrectomy) is treatment of choice.
- Routine exploration of contralateral kidney is not necessary if imaging is satisfactory and doesn’t suggest bilateral process.
- In favorable histology, Radiation therapy should be started within 10 days after nephrectomy. Chemotherapy should be started 5 days after surgery.
- Chemotherapy: VACD (Vincristine + Cyclophosphamide + Doxorubicin or daunorubicin).
- Whole lung irradiation is recommended for pulmonary metastasis.

Preoperative treatment should be considered
- Solitary kidney
- Bilateral renal tumors
- Tumor in horseshoe kidney
- Tumor thrombus in IVC above the level of hepatic veins
- Respiratory distress due to metastatic disease

Prognosis
- The histology of Wilm’s tumor and tumor stage is identified as most important determinant of prognosis (Histology > Stage).

Pediatric Tumors

| MC malignant tumor of infancy | Neuroblastoma |
| MC extracranial solid tumor in children | Wilm’s tumor |
| MC abdominal malignancy in children | Congenital mesoblastic nephroma |
| MC primary malignant renal tumor of childhood | Rhabdomyosarcoma |
| MC renal tumor of infancy | Brain tumor |
| MC soft tissue tumor in infants and children | Leukemia (30%) > Brain tumors (22%) |
| MC solid tumor of childhood | Leukemia |
| MC cancer of childhood | Leukemia |
125. Ans. a. Bone metastasis
126. Ans. c. Histology
127. Ans. a. Within 10 days
128. Ans. c. Same location
129. Ans. c. Abdominal mass
131. Ans. c. Hematuria almost always present
132. Ans. b. Less than 1 year
133. Ans. a. Pre-operative use of actinomycin D
134. Ans. b. Lungs
135. Ans. b. International society of Pediatric Oncology (SIOP) (Ref: Devita’s 8/e p2051, Schwartz 8/e p1509)

The post-chemotherapy based staging system is the ‘SIOP’ staging system developed by the International Society of Pediatric Oncology. Two Staging Systems are currently being used for the staging of Wilm’s Tumor.

<table>
<thead>
<tr>
<th>Pre-chemotherapy Staging System</th>
<th>Post-chemotherapy Staging System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Developed by the National Wilm’s Tumor staging Group (NWTS - Staging system)</td>
<td>Developed by the International Society of Pediatric Oncology (SIOP - Staging system)</td>
</tr>
<tr>
<td>This staging system is widely used in North America and Canada</td>
<td>This staging system is widely used in Europe</td>
</tr>
<tr>
<td>NWTS approach involves employment of ‘primary surgery’.</td>
<td>SIOP approach involves employment of preoperative chemotherapy without histological confirmation of Wilm’s tumor.</td>
</tr>
<tr>
<td>Chemotherapy with or without Radiation therapy is given after surgery</td>
<td>Primary chemotherapy for all patients regardless of extent</td>
</tr>
<tr>
<td>Staging is done at time of surgery (Pre-chemotherapy)</td>
<td>Staging is done at time of surgery (Post-chemotherapy)</td>
</tr>
</tbody>
</table>

136. Ans. c. Arthogryposis multiplex congenita
137. Ans. b. Open nephroureterectomy (Ref: Sabiston 18/e p2082)

- The treatment of choice for stage I Wilm’s tumor is transperitoneal radical nephrectomy (radical nephroureterectomy) followed by chemotherapy with or without radiotherapy depending upon tumor histology.

TUMORS OF RENAL PELVIS


Carcinoma Renal Pelvis

- Transitional cell carcinoma accounts for 90% of upper urinary tract cancers.
- Urothelial cancer often presents as a widespread urothelial abnormality: Patients with a single upper-tract carcinoma are at risk for developing bladder carcinoma (30-50%) and contralateral upper urinary tract carcinoma (2-4%).
- More common in males

Etiology
- Smoking
- Industrial dyes or solvents
- Excessive analgesic (Phenacetin) intake
- Balkan’s nephropathy

Notes: Eckovation App Users (2015, AIR-153)
Tumour:

1. Renal Cancer

2. Tumour confined to capsule: Radical nephrectomy + chemo - Daunomycin + vincaistine for 6 months.

3. Tumour beyond capsule: Nephrectomy + chemo - (D+V) - 15 months + Radio.


5. Nephrectomy chemo - D + Doxorubicine

6. Bladder WT = Radical neph. on larger side, partial nephrectomy on smaller side.

Adenocarcinoma:

- on upper pole - WT - on lower pole
- Reniform side of kidney maintained (WT - it losses easily)