Sickle cell disease and Urology

Definition
- Sickle cell anemia is characterized by alterations stemming from the predominance of type S hemoglobin, which, when polymerized, causes the appearance of rigid, sickle-shaped erythrocytes that are unable to pass through small capillaries, leading to obstructive vascular problems.
- The main urological complications of the disease are renal problems, priapism, and sometimes testicular infarction.

Renal complications
- Glomerular disease and papillary necrosis: the most frequent complications.
- Hypostenuria: inability to produce urine with a specific density <1010.
- Nocturnal enuresis: multifactorial, affecting over 1/3 of younger patients.
- Hematuria: due to vascular infarction in the renal medulla/papilla. More common in the left kidney.
- Hypertension: affects only 5% of patients.
- Acute renal failure: affects 10% of patients and often occurs due to rhabdomyolysis.
- Chronic renal failure: in the 3rd-5th decades of life due to glomerulopathy.
- Urinary infections: mainly in women and caused by Gram-negative. Recurrent in 1/3.
- Renal medullary carcinoma: very aggressive renal tumor with poor prognosis.

Priapism
- Prevalence: affects 1/3 of patients; occurs during sleep in 75% of cases.
- Mechanism: hypoventilatory acidosis induced during sleep decreases oxygen tension and pH in the corpora cavernosa, which is compounded by the sinusoidal venous stasis resulting from the special morphology of the sickle-shaped erythrocytes.
- Systemic treatment (see chapter on Priapism):
  - Abundant iv hydration, Bicarbonate iv, and O2 supplementation to reverse hypoxia and secondary acidosis.
  - Blood transfusion to achieve hemoglobin values >10 g/dL or a hematocrit >30%.
  - Opioid analgesics for relief of pain and anxiety.
- Intracavernous treatment:
  - Aspiration-irrigation: one CC is punctured with a 19G-21G needle, compressing the penis to extract the blood from the CC. This is followed by irrigation with 30-40 mL of NSS. BP and pulse should be monitored (if SBP >200, administer ADALAT® sl).
  - Intracavernosal Phenylephrine: agent of choice. The initial recommended dose is 1 mL of a 0.2 mg/mL solution, which can be repeated every 5 min up to a max of 1 mg (5 mL).
- Prophylaxis: in recurrent cases, administer Phenylpropanolamine, Ethylephrine, Bicalutamid, Cyproterone acetate, Finasteride/Dutasteride, Diazepam, Baclofen, or Ketokonazole + Prednisone (see chapter on Priapism).
- Surgical treatment: if the aforementioned methods fail (see chapter on Priapism).
  - Distal glans-cavernosum shunt: the glans is connected to the CC at one or more points with the aid of a biopsy needle or scalpel.
  - Proximal caverno-spongiosum or caverno-saphenous vein shunt: last resort.