



# case summary

- A previously healthy 9-year-old boy reported **3 episodes of painless passage of red colored urine, occurring immediately after exercise and lasting for 2 or 3 urinations**
- No fever or other symptoms were reported. The family history was negative for kidney disease and hematological disorders. physical examination revealed no pathological signs
- The CBC, levels of serum electrolytes and creatinine, and liver function tests were all within the normal range.
- **C3 and C4 were in the lower normal range** (C3 84 mg/dl, normal range 83–177 mg/dl, and C4 15 mg/dl, normal range 15–45 mg/dl). The antistreptolysin O antibodies (**ASTO**) titer was **613 IU/ml (normal < 200 IU/ml)**, and throat swab culture was positive for group A  $\beta$ -hemolytic Streptococcus
- Urinalysis revealed density 1.015, pH 6, negative albumin, **hemoglobin +++**, and **erythrocytes 80–100 cells/hpf**.
- **24 hour urine collection revealed calcium excretion 5 mg/kg/d and total protein excretion 84 mg/m<sup>2</sup> /d**,

- urine culture was negative. Urinary tract ultrasound was normal.
- He was hospitalized for 4 days, during which time no further episode of macroscopic hematuria occurred
- Urinalysis and 24 h urine calcium and protein on discharge was normal.

#### Questions:

- 1. Is hematuria glomerular or non glomerular in origin?
- 2. Which diseases should be considered in the differential diagnosis of this patient?
- 3. What is the most likely diagnosis?
- 4. What investigations would you perform to reach a definitive diagnosis?
- 5. How should this patient be treated?



**Is hematuria glomerular or non glomerular in origin?**

# Glomerular hematuria

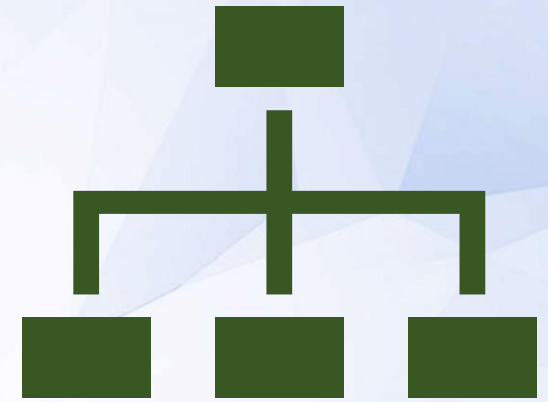
- Is often associated with brown, cola or tea-colored
- Proteinuria  $>100$  mg/dL via dipstick
- Urinary microscopic findings of RBC casts, and deformed urinary RBCs (particularly acanthocytes)
- Is usually associated with edema , oliguria, and hypertension



# Non glomerular hematuria

- May be associated with gross hematuria that is bright red or pink, terminal hematuria (gross hematuria occurring at the end of the urine stream), blood clots
- Normal urinary RBC morphology
- Minimal proteinuria on dipstick (<100 mg/dL).
- May be associated with LUT symptoms





**Which diseases should be considered in the differential diagnosis of this patient?**

# Causes of gross hematuria

- Acute glomerulonephritis if edema and hypertension are also present
- Urinary tract infection, hemorrhagic cystitis, urethritis, perineal irritation, urolithiasis, or hypercalciuria (conditions usually accompanied by voiding symptoms such as dysuria, frequency, and urgency)
- Trauma
- Coagulopathy
- Malignancy
- Recurrent gross hematuria suggestive of IgA nephropathy, familial benign hematuria, nutcracker syndrome, or Alport syndrome

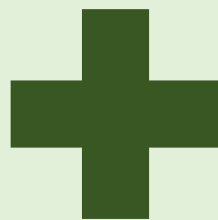




**What is the most likely diagnosis?**



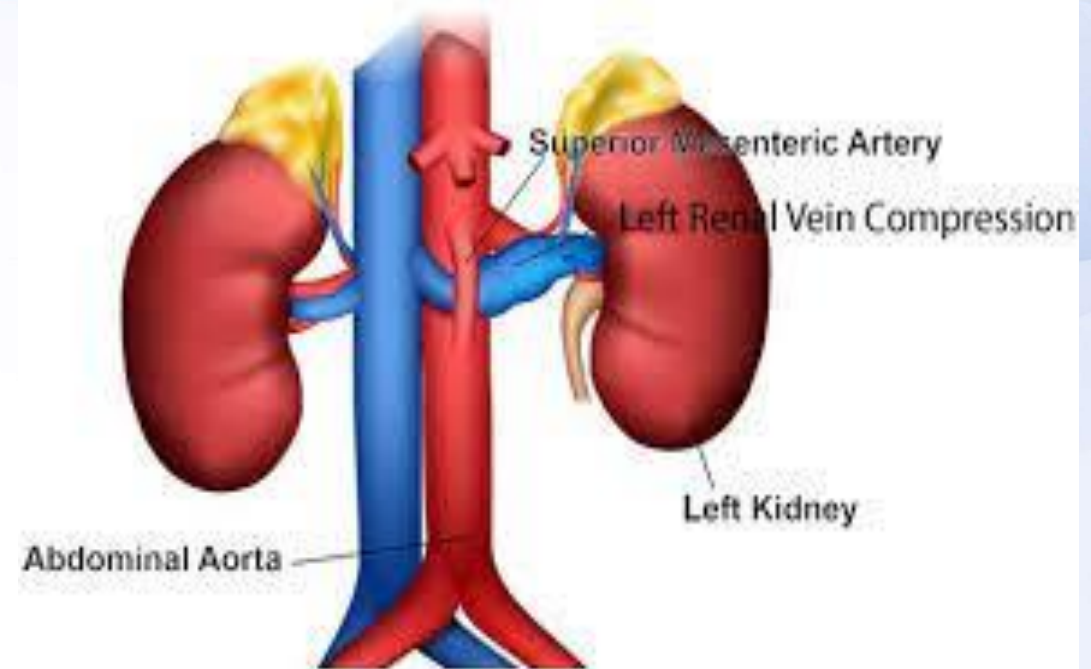
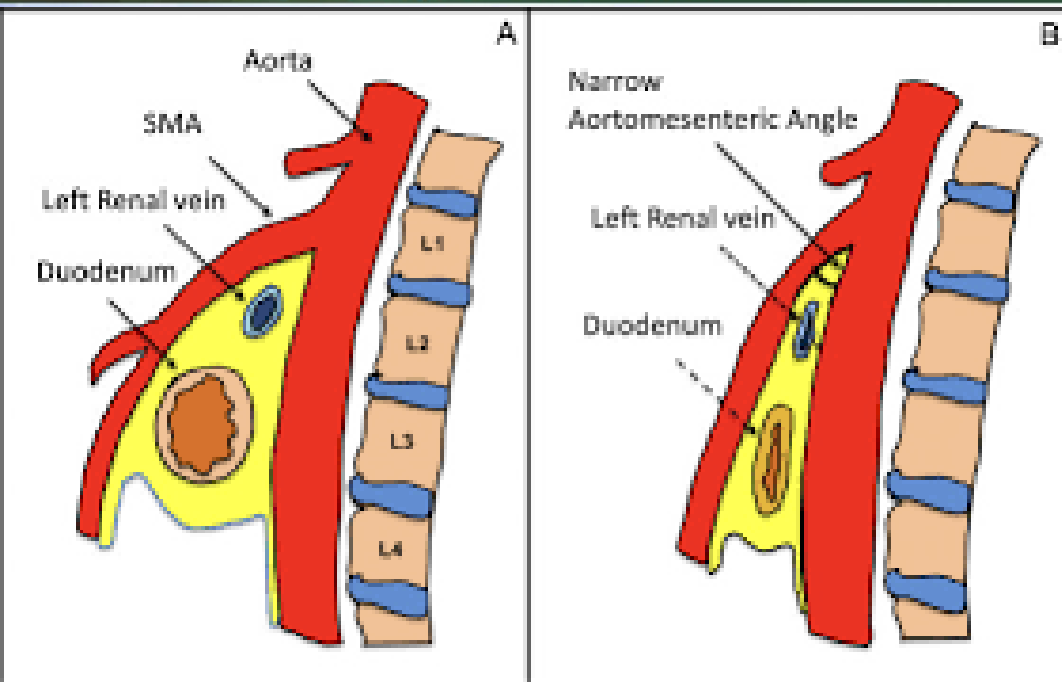
Macroscopic hematuria after exercise with resolution at rest



Exclusion of other common causes of gross hematuria



**NUTCRACKER SYNDROME (NCS)**



- NCS, also known as left renal vein entrapment syndrome, was first described in 1950
- Entrapment of the left renal vein between the superior mesenteric artery and the abdominal aorta

# Presentation

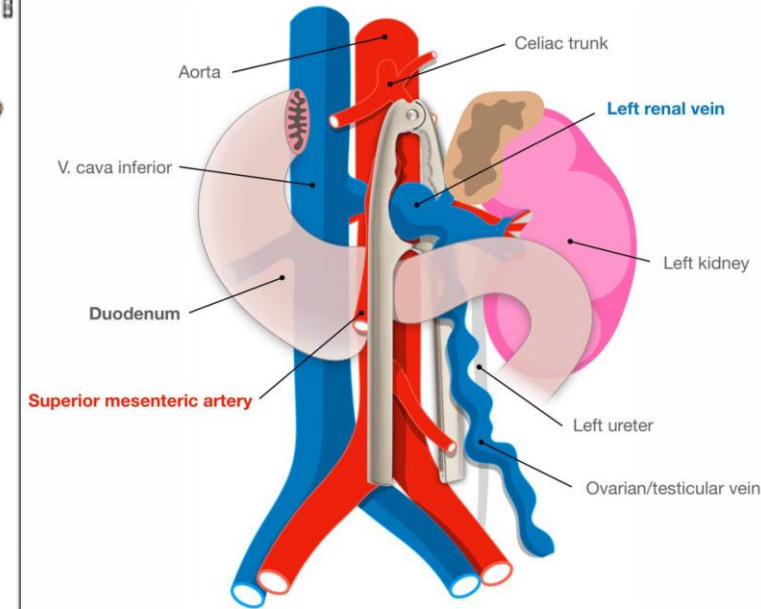
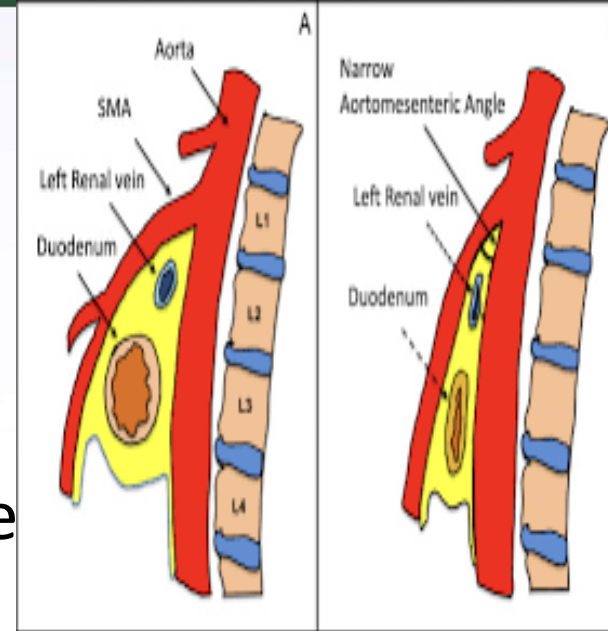
- Microscopic and macroscopic hematuria
- The pediatric patients present more frequently with microscopic hematuria
- 15% of the boys have a left-sided varicocele
- Hematuria may worsen after physical activity or in the upright position
- NCS can also cause postural proteinuria
- left-sided abdominal pain was reported in less than 40% of patients

- the hematuria is due to rupture of the septum separating the veins from the collecting system, related to venous hypertension
- There are no specific diagnostic criteria for NCS, and the diagnosis is usually made after exclusion of other common causes of hematuria
- diagnosis of NCS is often delayed
- The diagnosis should be considered when children present with orthostatic proteinuria and hematuria



**What investigations would you perform to reach a definitive diagnosis?**

- Doppler ultrasonography
- MRA or CT angio
- Venography is the most accurate diagnostic test for NCS



a reduced angle between the superior mesenteric artery and the abdominal aorta and a sharp change in the width of the left renal vein



# How should this patient be treated?





- Conservative monitoring is the most appropriate approach for patients aged under 18 years as automatic resolution of the stenosis may be observed with growth
- Reduce excessive exercise
- Monitoring clinically every 6 months and once a year with ultrasound and Doppler ultrasonography of the kidney and scrotum
- To gain weight in order to increase retroperitoneal fat, which leads to decompression of the left renal vein due to a change in the position of the left kidney
- Surgery may be necessary in patients with recurrent episodes of severe hematuria and pain