Approach to a child with red urine

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Causes of red urine

- Hemoglobinuria : G6PD deficiency
- Myoglobinuria :trauma,seizures,rhabdomyolysis
- Drugs (rifampicin),food
- Inborn errors of metabolism(porphyria)
- Urate crystals
- Hematuria :macroscopic

Analysis of hematurea

- Onset
- Color :Red if fresh(bladder), or brown color as Hb converted to acid haematin by urinary acids in renal causes
- Timing :Early hematuria:urethral cause ,Terminal hematuria:bladder cause
- Presense of clots : extrarenal causes
- Painful/painless,symptomatic,asymptomatic
- gross/ microscopic
- Transient /persistant
- With or without proteinurea

History and associated symptoms

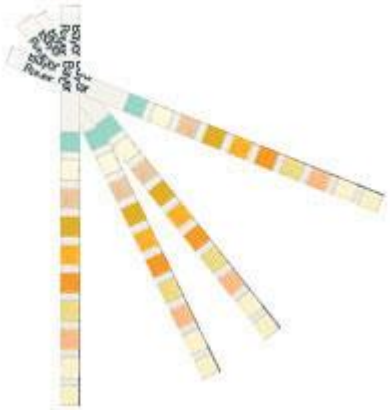
- Fever, urinary symptoms ,dysuria, frequency, loin pain/ suprapubic pain .(looking for cystitis,pyelonephritris/stones
- Age/gender
- Periorbital edema,lower limb edema, decreased urine output
- Preceding URTI…..PSGN,IgA nephropathy

- History of previous attacks of red urine
- Rash,arthritis …HSP,SLE
- Coagulopathy,bleeding tendency
- trauma
- FH of hematuria,deafness,renal failure…Alport
- FH of renal stones

Examination

- Vital signs: fever for UTI, hypertension for glomerulonephritis
- Looking for edema :lower limbs,eyes
- Abdomen exam : masses ,(PCKD) ,tenderness
- Genitalia exam:

Investigation



- Urine dipstick positive for hemoglobinuria,myoglobin (Positive heme,negative analysis), hematurea and neg if factitious
- Microscopy: look for RBC, wbc,bacteria (uti), high grade proteinurea (GN) ,crystals

- dysmprhic RBC by phase contrast microscopy ,RBC cast:glomerular bleeding

Distinguishing extraglomerular from glomerular hematuria

	Extraglomerular	Glomerular
Color (if macroscopic)	Red or pink	Red, smoky brown, or "Coca-Cola"
Clots	May be present	Absent
Proteinuria	Usually absent	May be present
RBC morphology	Normal	Dysmorphic
RBC casts	Absent	May be present

RBC: red blood cell.

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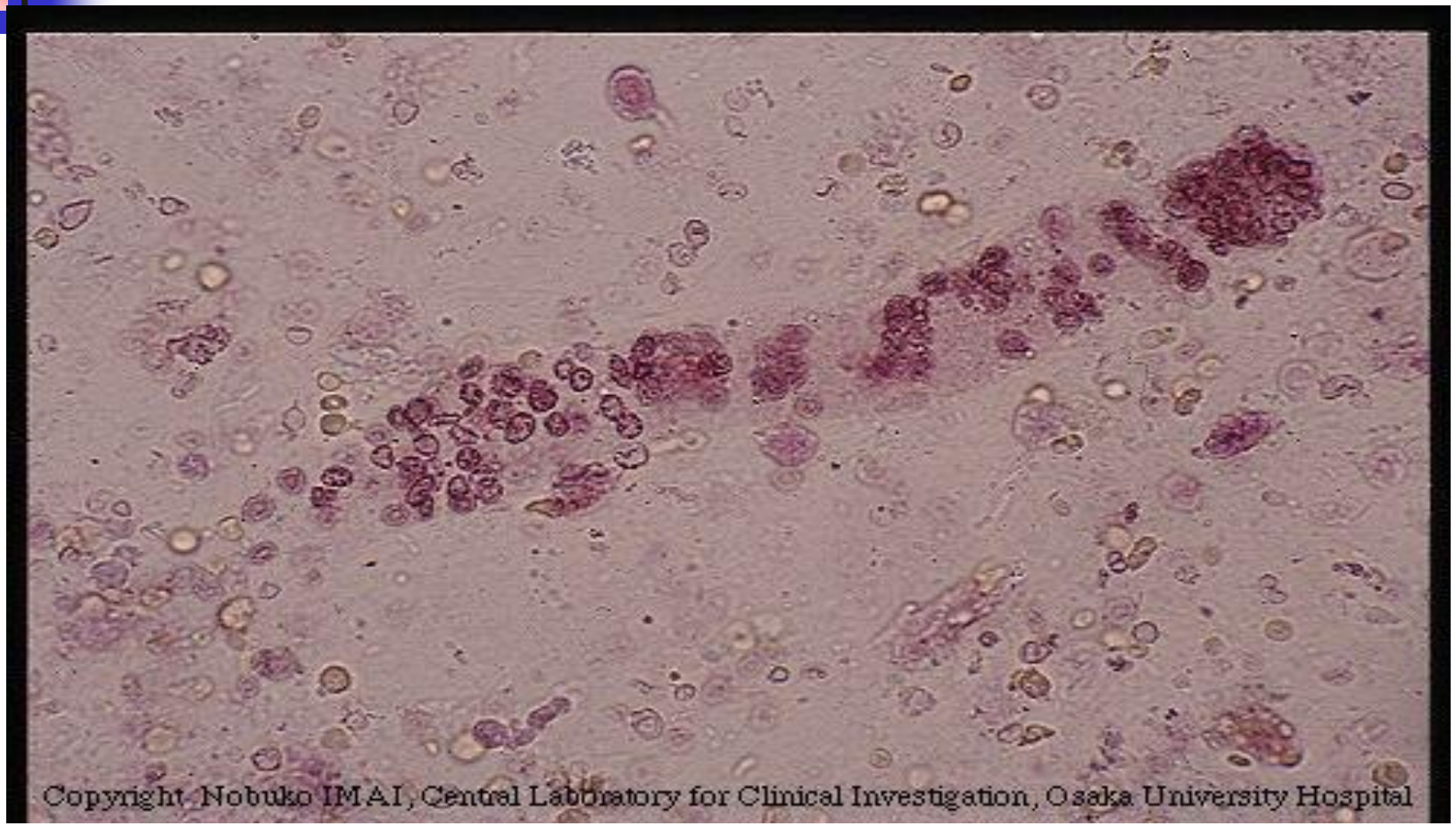
RBC casts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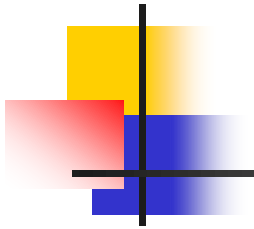
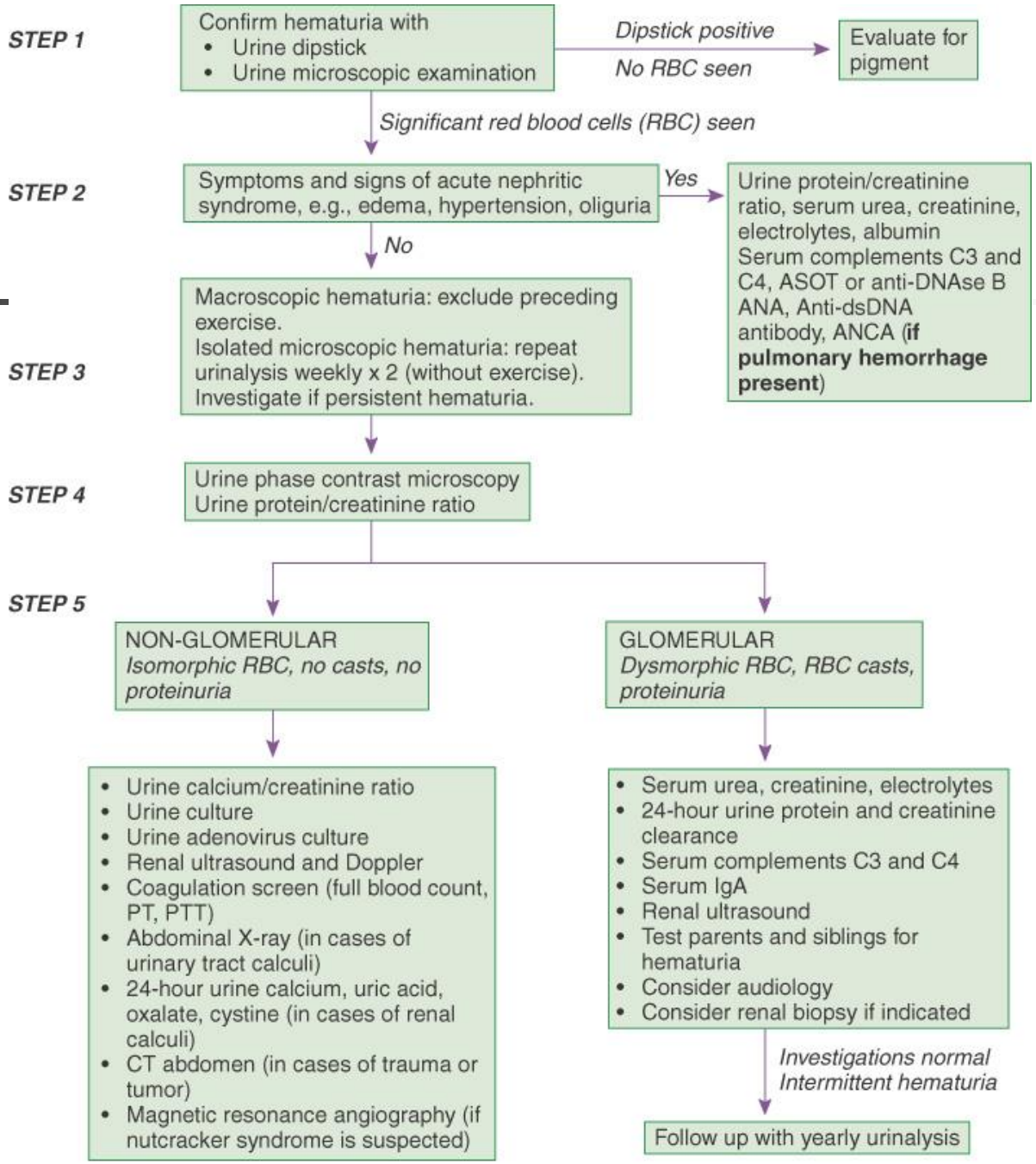


- Prevalence of microscopic hematuria is 0.5-2 %
- Definition of hematuria is the presense of more than 5 cells per high power field of centrifuged urine
- Transient hematurea seen with fever and exercise
- Persistant asymptomatic hematuria weekly for three times needs to be investigated
- Urethrorrhagia:urethral bleeding associated with blood spots after voiding,prepubertal

Investigations

- Urine protein/creat ratio ,Electolytes,albumin,kft ,ASOT,C3,C4,ANA for GN causes
- Urine culture if UTI
- CBC if infection ,PT,PTT
- Urine calcium/creat ratio, 24 h urine collection
- U/S ,XRAY, spiral CT
- Later :Urine analysis on parents ,cystoscopy
- Renal biobsy



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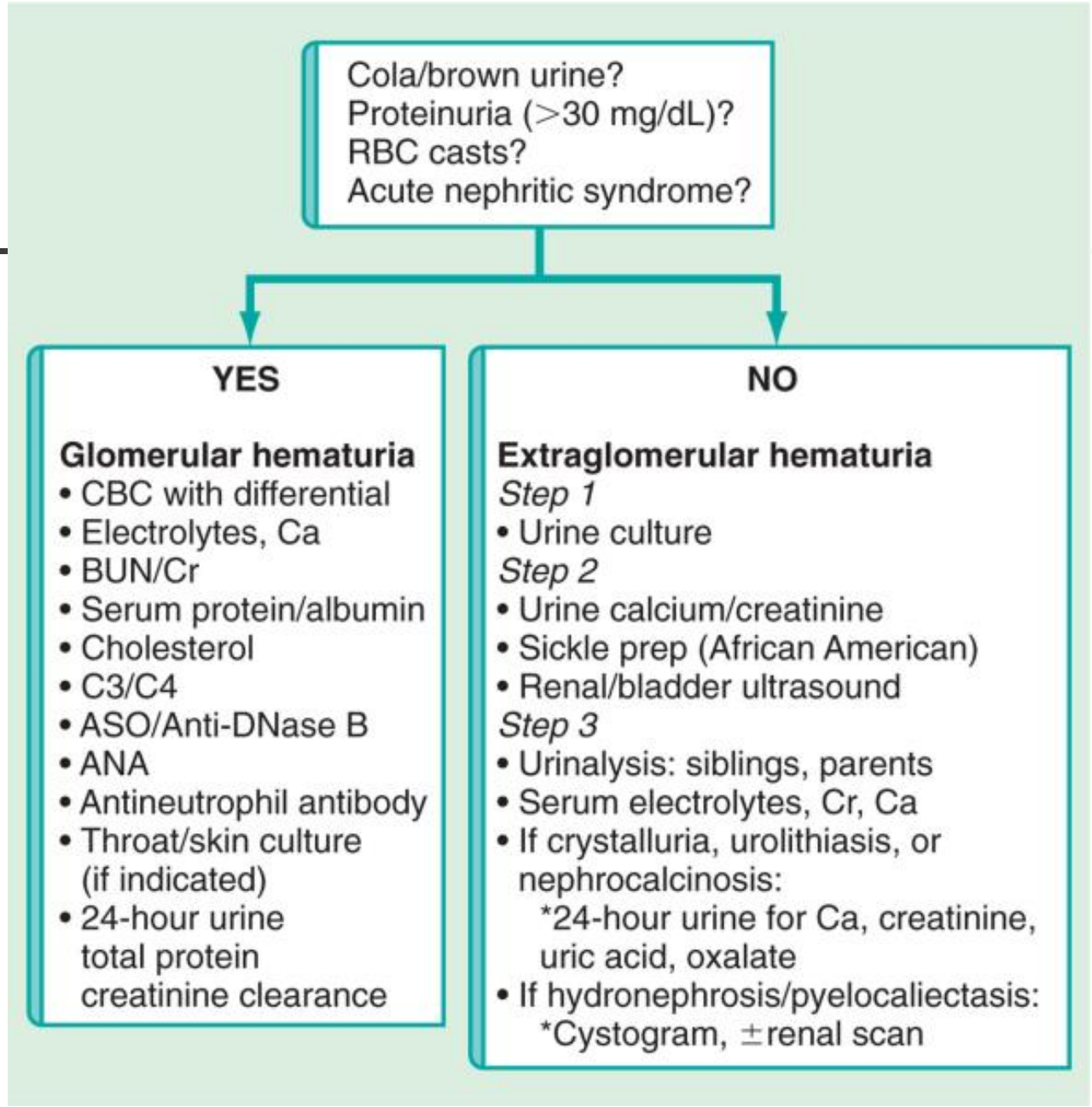

Cola/brown urine?
Proteinuria (>30 mg/dL)?
RBC casts?
Acute nephritic syndrome?
YES
Glomerular hematuria
• CBC with differential
• Electrolytes, Ca
• BUN/Cr
• Serum protein/albumin
• Cholesterol
• C3/C4
• ASO/Anti-DNase B
• ANA
• Antineutrophil antibody
• Throat/skin culture (if indicated)
• 24-hour urine total protein creatinine clearance
NO
Extraglomerular hematuria
Step 1
• Urine culture
Step 2
• Urine calcium/creatinine
• Sickle prep (African American)
• Renal/bladder ultrasound
Step 3
• Urinalysis: siblings, parents
• Serum electrolytes, Cr, Ca
• If crystalluria, urolithiasis, or nephrocalcinosis:
*24-hour urine for Ca, creatinine, uric acid, oxalate
• If hydronephrosis/pyelocaliectasis:
*Cystogram, ±renal scan

Macroscopic hematuria

- Most common cause is infection,then perineal irritation,trauma
- Viral infections,adenovirus 11,12 may cause hemorrhagic cystitis
- Exercise induced hematuria not associated with renal disease.
- Recurrent gross hematuria as IgA nephropathy,Alport,nut cracker (thin,loin pain,compression of renal vein bet aorta,SMA

Causes of hematuria

- 1.Glomerular causes
- Familial benign hematuria
- GN:primary as postinfectious,MPGN,IgA nephropathy,ALport
- secondary GN as SLE,HSP.
- HUS , Acute tubular necrosis,interstital nephritis,renal vein thrombosis,cystic renal disease
- Pyelonephritis,PCKD,Wilms tumor

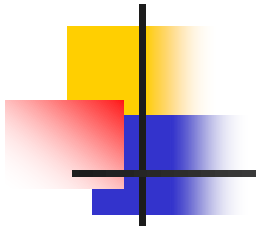
Non glomerular causes

- UTI
- Hypercalcuria,renal calculi,crystalluria
- Trauma,exercise
- Coagulpathy as sickle cell
- Vascular malformations
- Nut cracker syndrome
- Menarche
- Malignancy as nephroblastoma of the kidney or bladder tumors

- Persistent MA without proteinuria :
- 1.Beningn familial hematuria
- 2.idiopathic hypercalcuria
- 3.IgA nephropathy
- 4.Alport syndrome

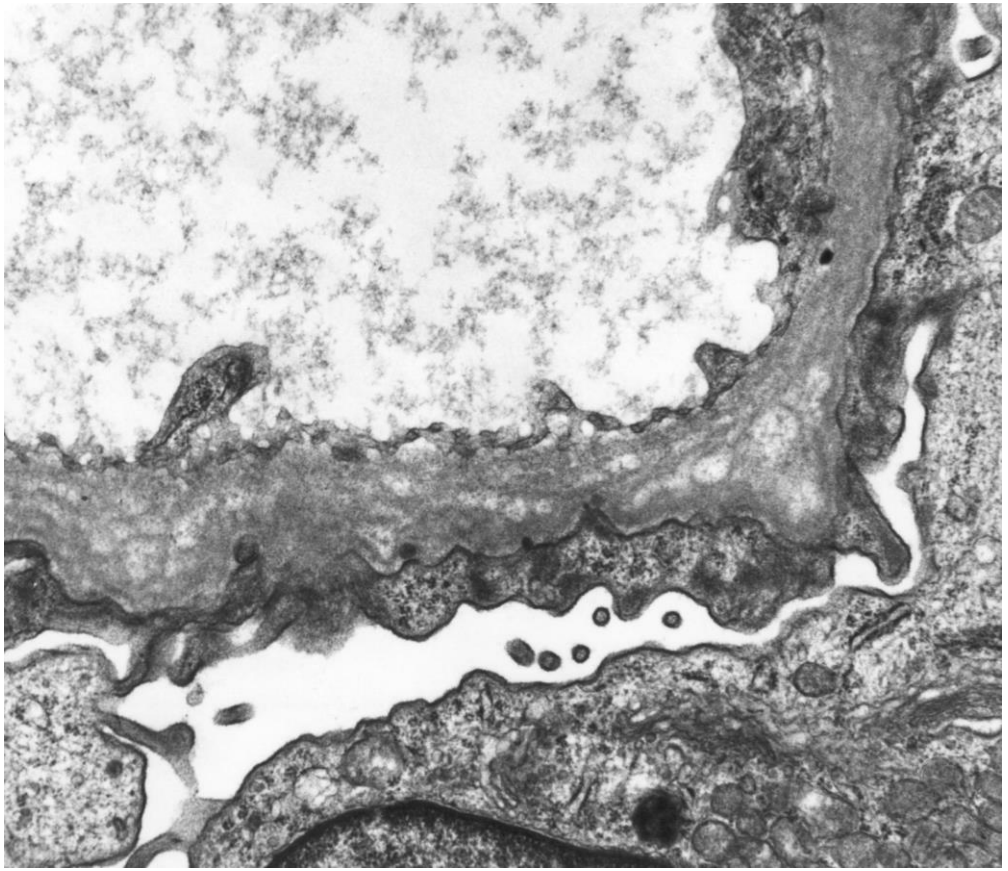
Alport Syndrome

- 80% XL,20% AR
- Renal failure,sensorineural deafness higher frequencies,ocular changes(anterior lenticonus,retinal changes
- Present as micro and rarely macroscopic hematauria with URTI
- Proteinura,HTN later age



- Diagnosis by EM:Thinning of GBM,split and duplicated lamina densa,basket weave
- Males progress to ESRD,deafness by 30y
- ACEI may delay progression to ESRD
- Deficiency of α5 of type 4 collagen

GBM in Alport syndrome



Benign Familial Hematuria(TBMN)

- AD inheritance
- Present as microscopic hematuria,no proteinuria or renal failure
- EM:thinning of GBM
- Follow up for proteinuria,HTN

IgA nephropathy

- Recurrent macroscopic hematuria,loin pain 1-2 days following URTI,last < 3 days.
- Persistent microscopic hematuria ±proteinuria
- Nephritic,nephrotic syndrome rare
- Present second decade,more in males

- Familial cases reported
- IgA high in 35-50%
- Diagnosis:LM:focal or diffuse mesangial cell proliferation,expansion of mesangial matrix
- IM:IgA,C3 deposits
- Prognosis for children better than adults
- Young children without macroscopic hematuria have the best long term outcome

- Heavy proteinuria is a risk factor for progression to ESKD.
- Progression to ESRD is slow(25% need dialysis in 20y)
- ACEI are used to delay progression,decrease proteinuria
- Fish oil

PSGN

- Follows GAS pharyngitis in winter,pyodrema in summer

- Certain nephritogenic M types,age 5-15 y,M:F 2:1
- Risk of PSGN following GABHS is 15%
- antibiotic treatment doesn’t prevent PSGN
- Pathogenesis

- Clinical features:latent period 10-14 days after pharyngitis, 3-6 wk pyoderma

Clinical manifestations

- Nephritic sydnrome :
- (edema,oliguria,HTN,hematuria,azotemia)
- Gross hematuria 30-70% resolves in 1-2 wk,cola urine,relapses appear after infection,exercise
- Microscopic hematuria in all,proteinuria
- Edema due salt &water retention,oliguria
- HTN 80% resolves in 4 wk
- Malaise ,lethargy ,flank pain

Clinical manifestations

- Complications:pulmonary edema,HTN encephalopathy,CHF,ARF
- Nephrotic syndrome,RPGN rare
- Subclinical disease in 90% of cases

Laboratory findings

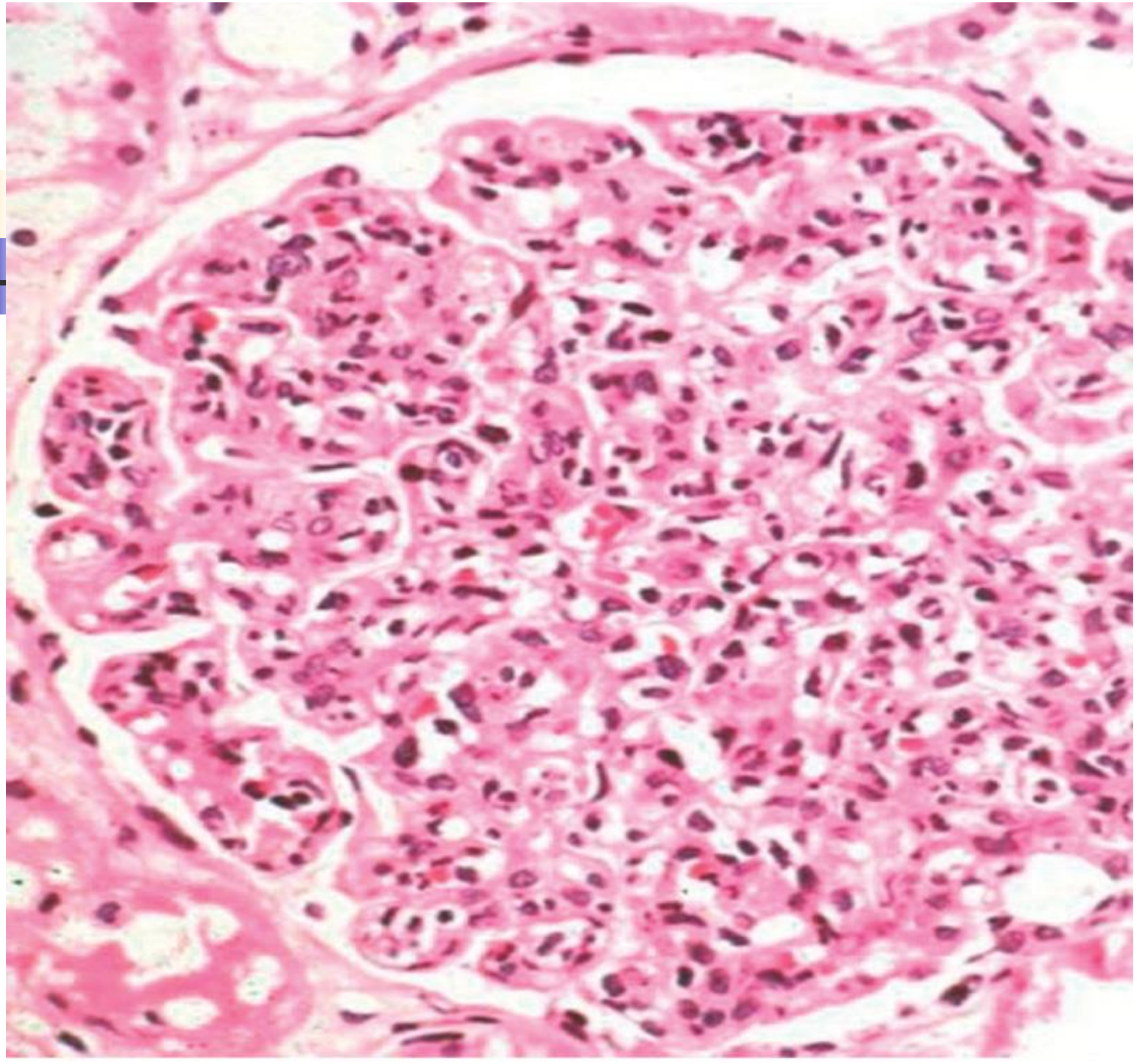
- Urine showes dysmorphic RBC,casts
- High K,acidosis,uremia,Throat culture
- High ASOT,antiDNAs after skin infections
- 90% have low C3, normalize 6-8 wk

Treatment

- 1-salt and water restriction
- 2-loop diuretics
- 3_antiHTN drugs,calcium channel blockers
- Pulse steriods in cresentic RPGN
- Dialysis in ARF

- Histopathology:exudative proliferative GN

- EM:deposits,humps on subepithelial side of GBM



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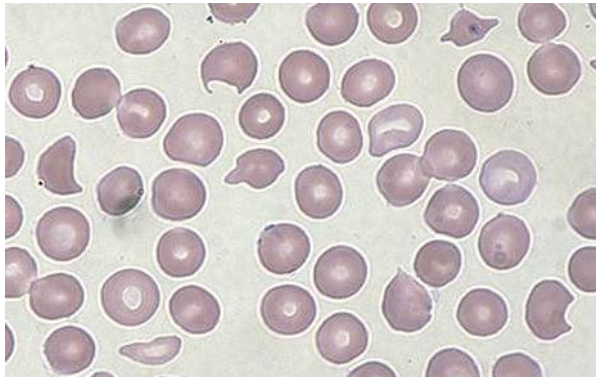
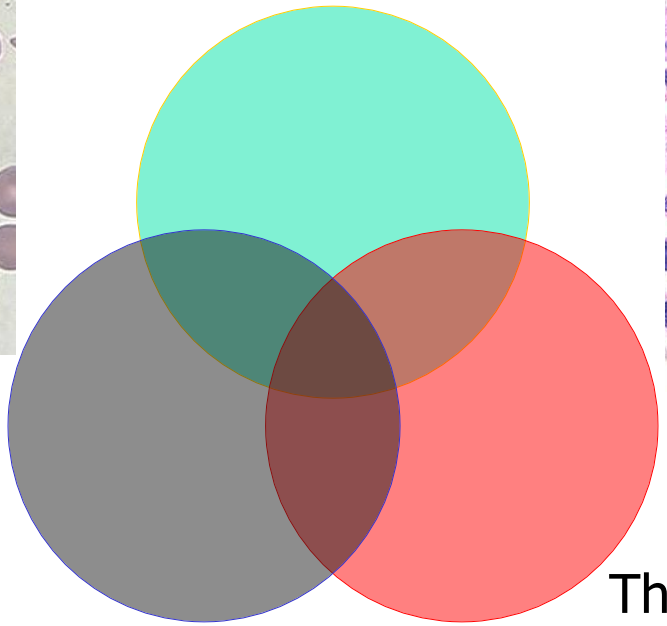
Prognosis

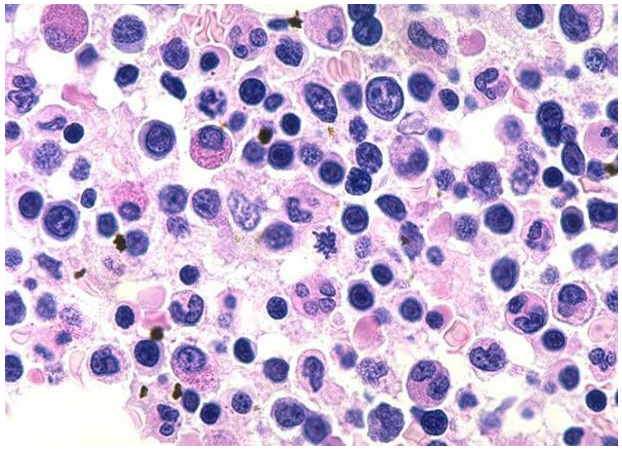
- Acute phase resolve in 6-8 weeks
- Persistent proteinuria for 6 m
- Persistent hematuria for 1-2 y
- Excellent long term outcome
- Rare recurrence
- Mortality <1%

HUS

Acute haemolytic anaemia







Reduced GFR

Thrombocytopaenia

- Most commom cause of ARF

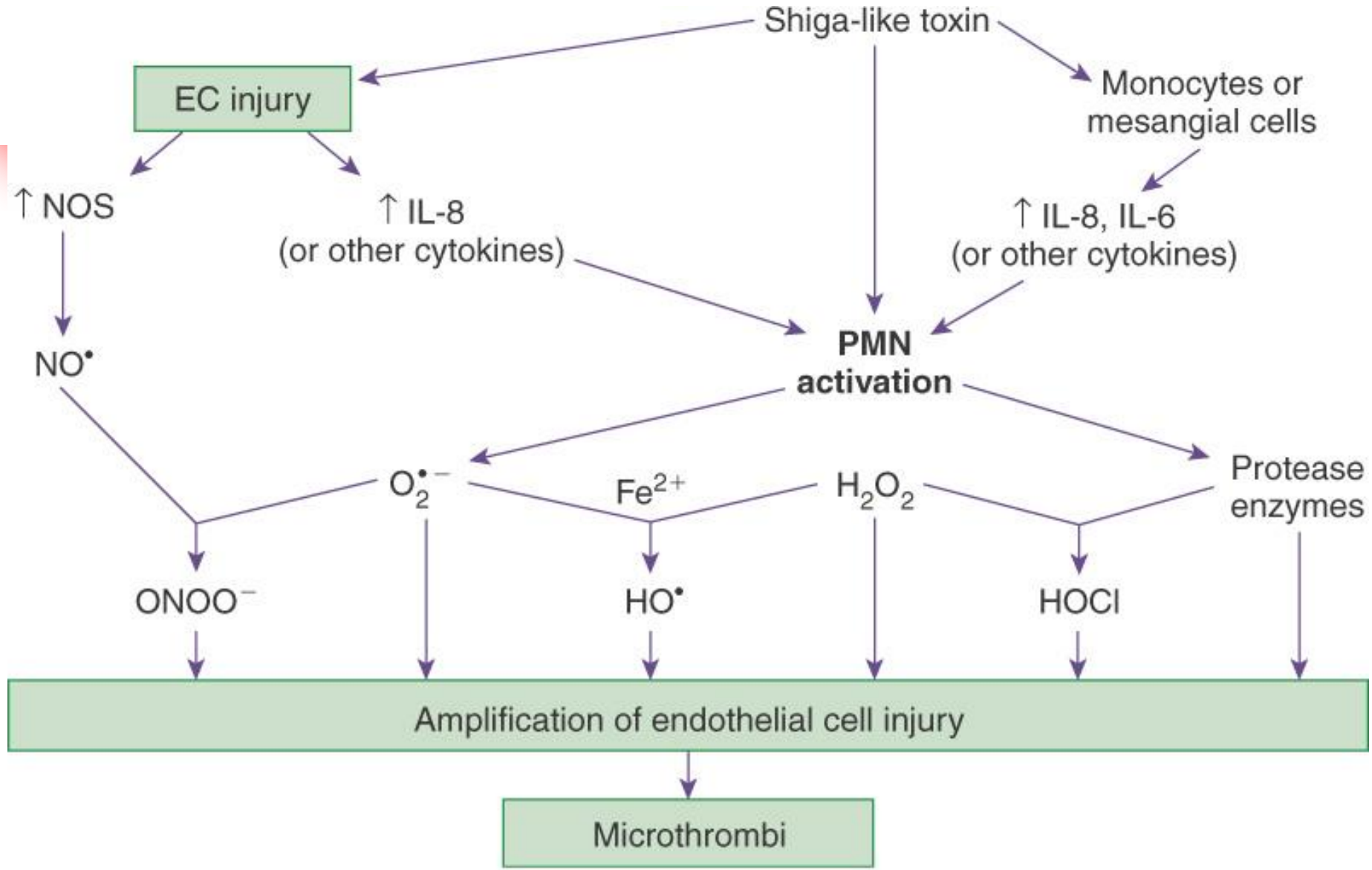
- Triad:MAH,ARF,thrombocytopenia

- Pathology:endothelial cell injury microthrombi ,ischemic injury to multiple organs

- Kidney :glomerular,arterial thrombotic microangiopathy ,cortical necrosis

Classification of HUS

- Infectious (Stx)
  - E coli 0157:H7
  - Shigella dysenteriae type I (D+ HUS)
- Hereditary
  - Factor H deficiency, VWF proteinase def,ADAMTS-13
- Secondary
  - Pregnancy
  - Malignancy
- Medication
  - CNIs



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Diarrhae + HUS

- D+HUS:follows STEC, shigella
- Transmitted undercooked hamburgers,milk,person to person
- O157:H7 E. coli most common serotype
- 5-15% of kids infected STEC develop HUS

- Risk of HUS increase with age <5y, WBC >13,000/mm^3,antimotiliy drugs (retention of toxin
- Antibiotic can increase risk?? Release toxin

Clinical Manifestations

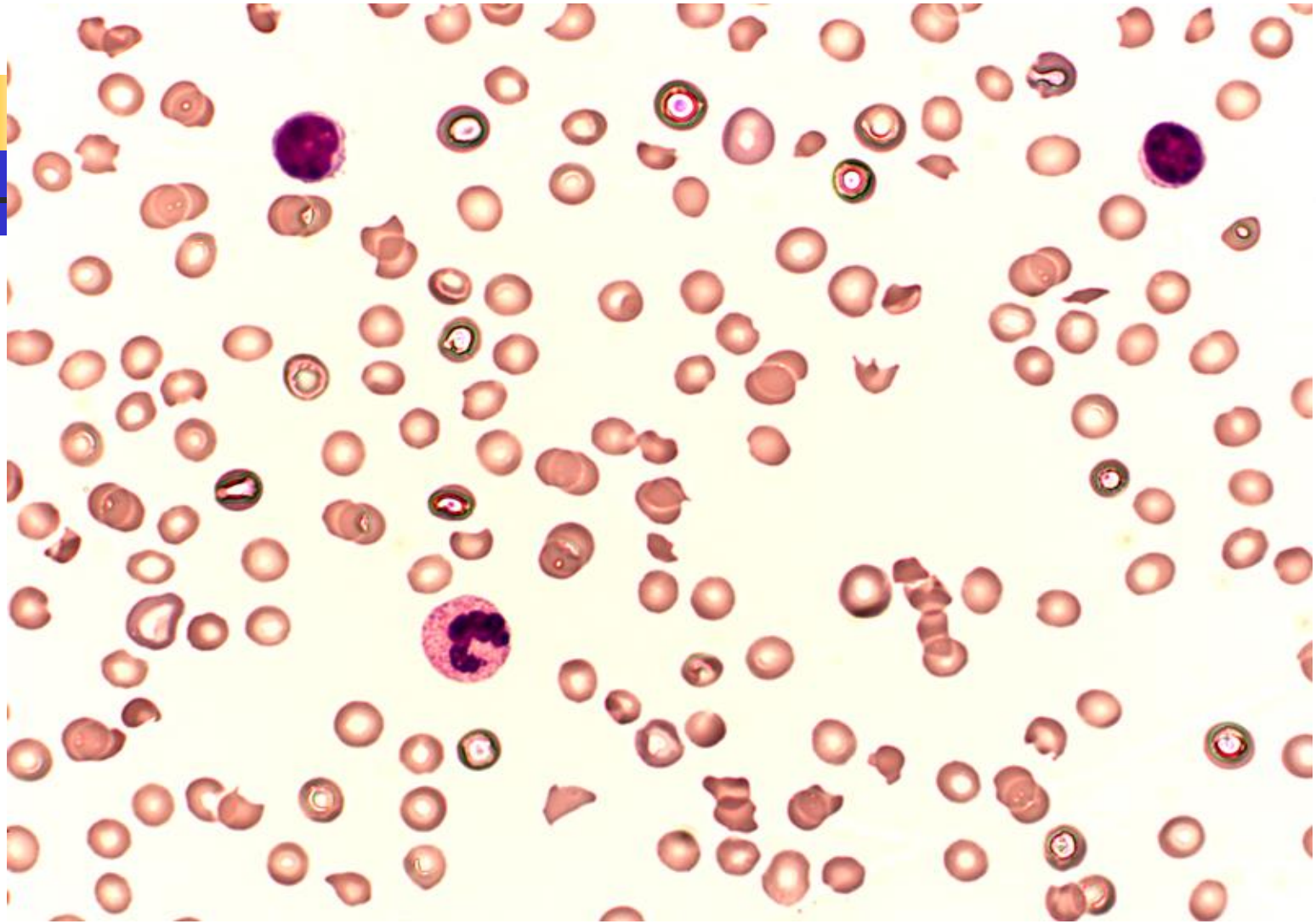
- Diarrhea 3-7 d after exposure to STEC,mostly bloody
- Pallor,oliguria 4-7 d post diarrhoea
- GIT:severe colitis,transmural
- necrosis,perforation,stricture,rectal prolapse
- Hepatitis,jaundice 35%

Pancreatitis

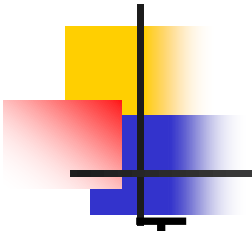
- Glucose intolerance,IDDM
- CNS: 20% seizures,irritability,confusion
- Mycardium ischemia rare,rhabdomyolysis
- HTN,renal cortical necrosis, 50% are anuric,75% needs dialysis

Investigations

- FBC,showes anemia and low platlets.
- LDH high,blood film shows schizocytes,fragmented RBC
- High urea and creatinine
- Elevated liver enzymes 40 %
- hematuria,proteinuria



Management



- Transfusion if severe hemolysis,slowly 4h
- Monitor fluid and electrolyte status
- Platlet if bleeding,can accelerate microthrombi formation
- 5-10% develop ESRD
- RRT if ARF


THANK YOU