INTERESTING CASE

16/10/2023

PEDIATRICS PMK

4th floor ward

Patient Profile

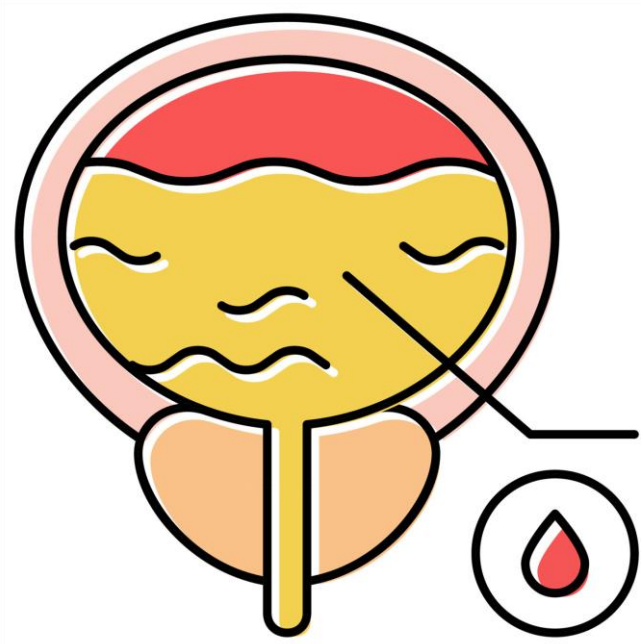
ผู้ป่วย : เด็กหญิงไทยอายุ 6 ปี 10 วัน

ภูมิลำเนา : จังหวัด นครปฐม

รับเข้ารักษาในโรงพยาบาล : วันที่ 10 ตุลาคม 2566

ประวัติได้จาก : ผู้ป่วย มารดา ใบส่งตัว
และเวชระเบียน มีความน่าเชื่อถือสูง

Chief complaint



ปัสสาวะสีโค้ก
1 วัน ก่อนมาโรงพยาบาล

History interview for approach

Present illness

1 สัปดาห์ ก่อนมารพ.(30/8/66) มีไข้ (ไม่ได้วัด) ไอมีเสมหะ เจ็บคอ ไม่ได้ไปพบแพทย์และไม่ได้ยาปฏิชีวนะ

5 วัน ก่อนมารพ.จังหวัด(3/9/66) มีไข้สูง เจ็บคอมาก ไม่อยากอาหาร จึงไปรักษาตัวแบบผู้ป่วยใน ที่ รพช. ได้ยาปฏิชีวนะไม่ทราบชนิด และสารน้ำทดแทนทางเส้นเลือดดำ

1 วัน ก่อนมารพ.จังหวัด(6/9/66) ขณะรักษาตัวที่ รพช พบว่ามีปัสสาวะสีโค้ก เป็นตลอดสาย ไม่มีลิ่มเลือด ไม่มีปัสสาวะแสบขัด ไม่ปวด ไม่มีตัวบวม ไม่มีหอบเหนื่อย นอนราบได้ ไม่มีปัสสาวะเป็นฟอง ถูกส่งตัวมารักษาต่อที่โรงพยาบาลจังหวัด

Past history and Past illness

- Late preterm female newborn GA 36+4 weeks C/S due to previous C/S with preterm labour , APGAR 8,9 , no complication after birth
- แจ้งว่าเคยตรวจการได้ยินตอนวัยเด็ก ผลเป็นปกติ
- ปฏิเสธประวัติโรคประจำตัว ปฏิเสธประวัติผื่นแพ้แสงปวดข้อหรือไข้เรื้อรังมาก่อน
- ปฏิเสธประวัติแพ้ยา แพ้อาหาร
- ปฏิเสธประวัติการใช้สมุนไพร ยาต้ม ยาลูกกลอน หรือ ยาต้านเกร็ดเลือด
- ปฏิเสธการใช้สารเสพติด
- ปฏิเสธประวัติปัสสาวะเป็นเลือดก่อนหน้านี้ ปฏิเสธประวัติปวดท้องเรื้อรังหรือคลำได้ก้อนที่ท้องน้อยมาก่อน
- ปฏิเสธประวัติเลือดออกง่ายหยุดยากมาก่อนหน้านี้
- ปฏิเสธประวัติออกกำลังกายหนัก หรือ อุบัติเหตุ ก่อนมานอนโรงพยาบาล

Personal history

- Development : พัฒนาการปกติ

- เรียนอยู่ชั้น ป.1 ผลการเรียนปกติ บวกลบเลขได้ , วิ่งหลบสิ่งกีดขวางได้ , วาดรูปทรงต่างๆได้ เข้ากับเพื่อนได้มีกลุ่มเพื่อนที่สนิท ไม่มีปัญหาด้านการสื่อสาร

- Netritional history

- รับประทานอาหาร 3 มื้อ ครบ 5 หมู่ , ดื่มนมกล่อง 2 กล่องต่อวัน (180 ml)

- Vaccination

- ครบตาม EPI : BCG*1 HBV*1 DTwP-HB-Hib*3 OPV*2 IPV*1 Rota*2 MMR*2 JE*2

Family history

- บิดาอายุ 35 ปี ปฏิเสธประวัติโรคประจำตัว ภูมิลำเนา จังหวัดนครปฐม ปฏิเสธประวัติปัสสาวะเป็นเลือด การได้ยินเป็นปกติ
- มารดาอายุ 35 ปี ปฏิเสธประวัติโรคประจำตัว ภูมิลำเนา จังหวัดนครปฐม ปฏิเสธประวัติปัสสาวะเป็นเลือด การได้ยินเป็นปกติ
- ปฏิเสธประวัติการได้ยินบกพร่องหรือปัสสาวะเป็นเลือดของญาติผู้ชายในครอบครัวฝ่ายมารดา
- พี่ชาย อายุ 8 ปี โรค G6PD deficiency ปัสสาวะปกติ
- ปู่ อายุ ปี 73 ปี เป็น ESRD จาก เบาหวานและความดันโลหิตสูง เป็นขณะอายุประมาณ 70 ปี
- ปฏิเสธประวัติแต่งงานในเครือญาติ บิดาและมารดาอยู่คนละอำเภอ
- ปฏิเสธประวัติคนในครอบครัวเป็นโรคแพ้ภูมิตัวเอง

Urinalysis (6/9/66) รพช.

Color	Sp.gr.	pH	Blood	Protein	Glu	WBC	RBC	Epi
Pale Yellow	1.010	6	3+	2+	Neg	3-5	30-50	0-1

Serum Cr 0.5 mg/dL

Interpretation of urine analysis >>

Color / Clarity	Yellow / Clear
Sp.Gr.	1.010
pH	6
Protein	2+
Sugar	Negative
Blood	3+
Leukocyte	Negative
Nitrite	Negative
Ketone	Negative
Bilirubin / Urobilinogen	Normal / Negative
RBC	30-50
WBC	3-5
Squa Epi	0-1

Urine analysis

Color : Red Brown Orange Blue green

1.002-1.035 if 1.010 = isoosteinuria = euvolumic status

4.5-8.0 : High protein > acidic Proteus/Fruits > base

Protein
Negative : < 10 mg/dL
Trace : 10-30 mg/dL
1+ : 30-100 mg/dL
2+ : 100-300 mg/dL
3+ : 300-1000 mg/dL
4+ : > 1 g/dL

Blood
Heme measurement
Hematuria
Myoglobinuria
Hemoglobinuria

Ketone : DKA , Starvation , IEM

Glucosuria : BS 180-200 mg% , Fanconi syndrome

Nitrite >> Nitrate ; Bacteriuria (except enterococcus)

Hematuria
RBC : > 5 cell/HPF
Dysmorphic : GN

Pyuria
WBC > 5 cell /HPF
: UTI , AIN

Urobillinogen
Bilirubin
= Hepatitis ,
Cirrhosis , Bile obs
Hemolysis

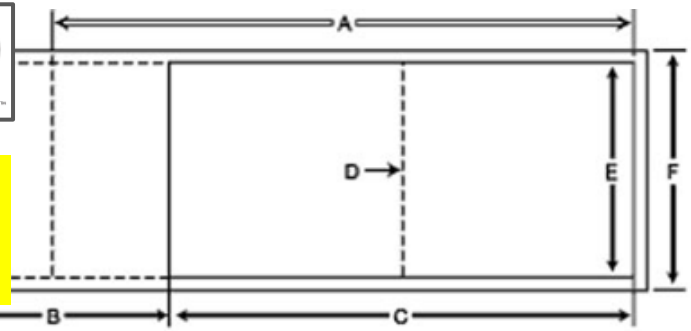
Physical examination at รพ.นครปฐม

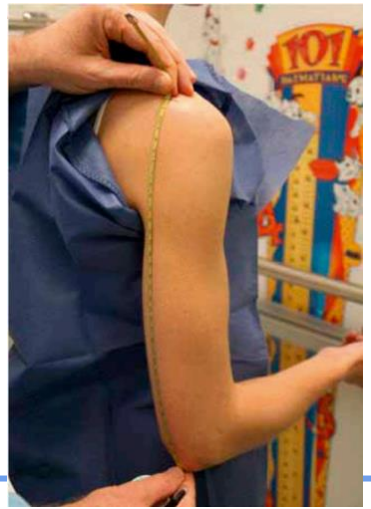
- Vital signs : Body temperature 37.6 C **Blood pressure 114/60 mmHg (P95)**
  Respirstory rate 20/min Heart rate 100 bpm
- Measurement : Weight 18.5 kg (P25) ,
  Height 113 cm (P10-P25)
- General appearance : A thin girl , well cooperate
- HEENT : not pale , anicteric sclerae ,
  no puffy eyelid , no discoid rash ,
  no oral ulcer , no malar rash , normal external ear canals , injected pharynge and tonsils , no exudative tonsils
- Neck : no cervical and supraclavicular lymphadenopathy

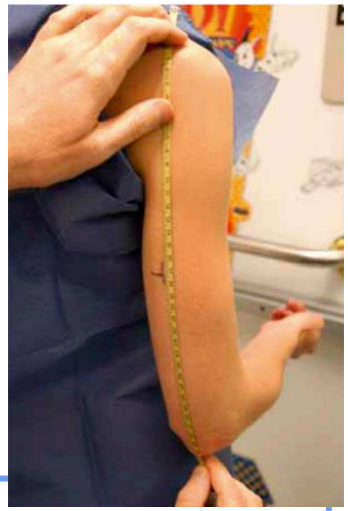
Interpretation	Children aged 1-13yr
Normal BP	< P90th P
Elevated BP	P90th - < P95th P
Stage 1 hypertension	P95th - < P95th+12
Stage 2 hypertension	P95th + 12 or > 140/90 mmHg

American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™

Best Blood Pressure Measurement







F : bladder width 40%MAC C : bladder length80%-100%MAC

TABLE 7 Best BP Measurement Practices

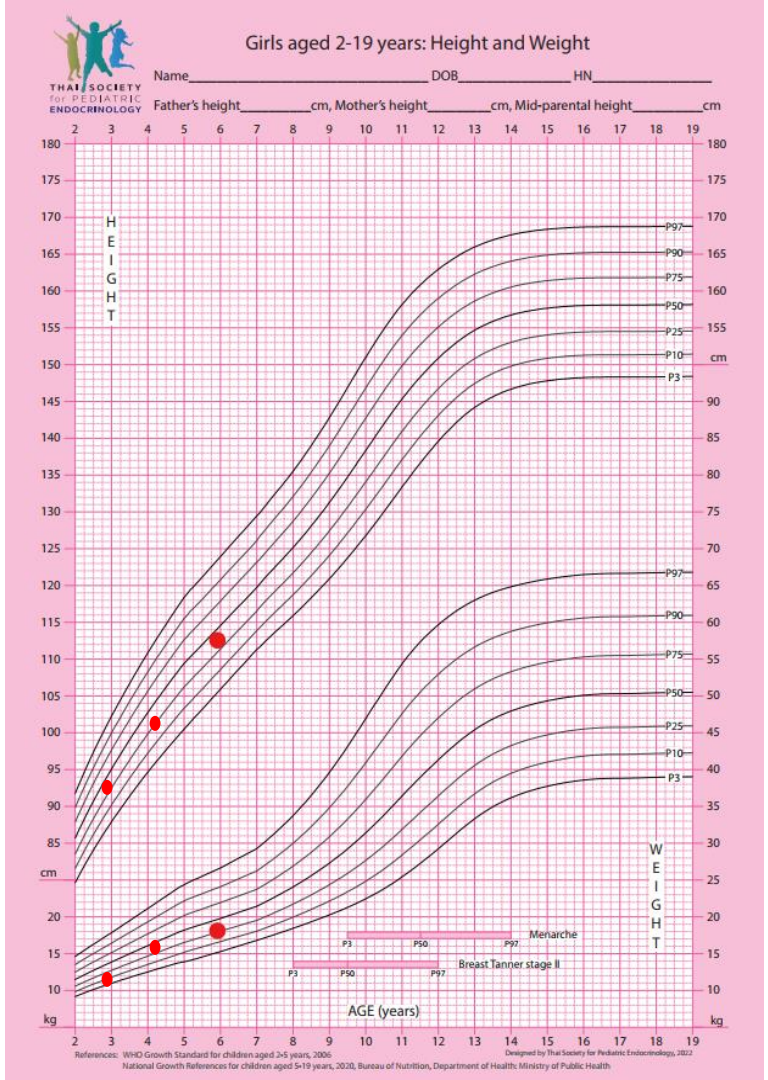
1. The child should be seated in a quiet room for 3–5 min before measurement, with the back supported and feet uncrossed on the floor.
2. BP should be measured in the right arm for consistency, for comparison with standard tables, and to avoid a falsely low reading from the left arm in the case of coarctation of the aorta. The arm should be at heart level,[90] supported, and uncovered above the cuff. The patient and observer should not speak while the measurement is being taken.
3. The correct cuff size should be used. The bladder length should be 80%–100% of the circumference of the arm, and the width should be at least 40%.
4. For an auscultatory BP, the bell of the stethoscope should be placed over the brachial artery in the antecubital fossa, and the lower end of the cuff should be 2–3 cm above the antecubital fossa. The cuff should be inflated to 20–30 mm Hg above the point at which the radial pulse disappears. Overinflation should be avoided. The cuff should be deflated at a rate of 2–3 mm Hg per second. The first (phase I Korotkoff) and last (phase V Korotkoff) audible sounds should be taken as SBP and DBP. If the Korotkoff sounds are heard to 0 mm Hg, the point at which the sound is muffled (phase IV Korotkoff) should be taken as the DBP, or the measurement repeated with less pressure applied over the brachial artery. The measurement should be read to the nearest 2 mm Hg.
5. To measure BP in the legs, the patient should be in the prone position, if possible. An appropriately sized cuff should be placed midthigh and the stethoscope placed over the popliteal artery. The SBP in the legs is usually 10%–20% higher than the brachial artery pressure.

Adapted from Pickering TG, Hall JE, Appel LJ, et al. Recommendations for blood pressure measurement in humans and experimental animals: part 1: blood pressure measurement in humans: a statement for professionals from the Subcommittee of Professional and Public Education of the American Heart Association Council on High Blood Pressure Research. *Circulation.* 2005;111(5):697–716.

Physical examination at รพ.นครปฐม

- Cardiovascular : full and regular pulse , capillary refill < 2 seconds , normal S1S2 , no murmur
- Lungs : clear and equal breath sound both lungs , no adventitous sound
- Abdomen : soft , no distension , no hepatosplenomegaly , no palpable mass
- Extremities : no deformity , no edema both legs and feet , no arthralgia , no arthritis , no limit ROM , no joint swelling
- Skin : no rash , no petechia , no ecchymosis , no palpable purpura
- Neuro : E4V5M6, alert , oriented to time place person , pupil 2 mm RTLBE , full EOM , Motor grade V all , sensory intact , deep tendon reflex 2+ all extremities

Growth curve



Pertinent Finding and Problem lists

Pertinent Finding

Positive pertinent findings

- Gross hematuria
- A 7 years old girl
- Stage I Hypertention
- Recent history of upper respiratorytract infection : synpharyngitic
- Urine analysis evidence of true hematuria and significant proteinuria

Negative pertinent findings

- No history of trauma
- No history of bleeding tendency
- No back pain or dysuria , foamy urine or hematuria
- No history of urinary tract infection
- No malar rash , palpable purpura
- No history of medicine use
- Denied family history of hematuria or hearing loss and early ESRD

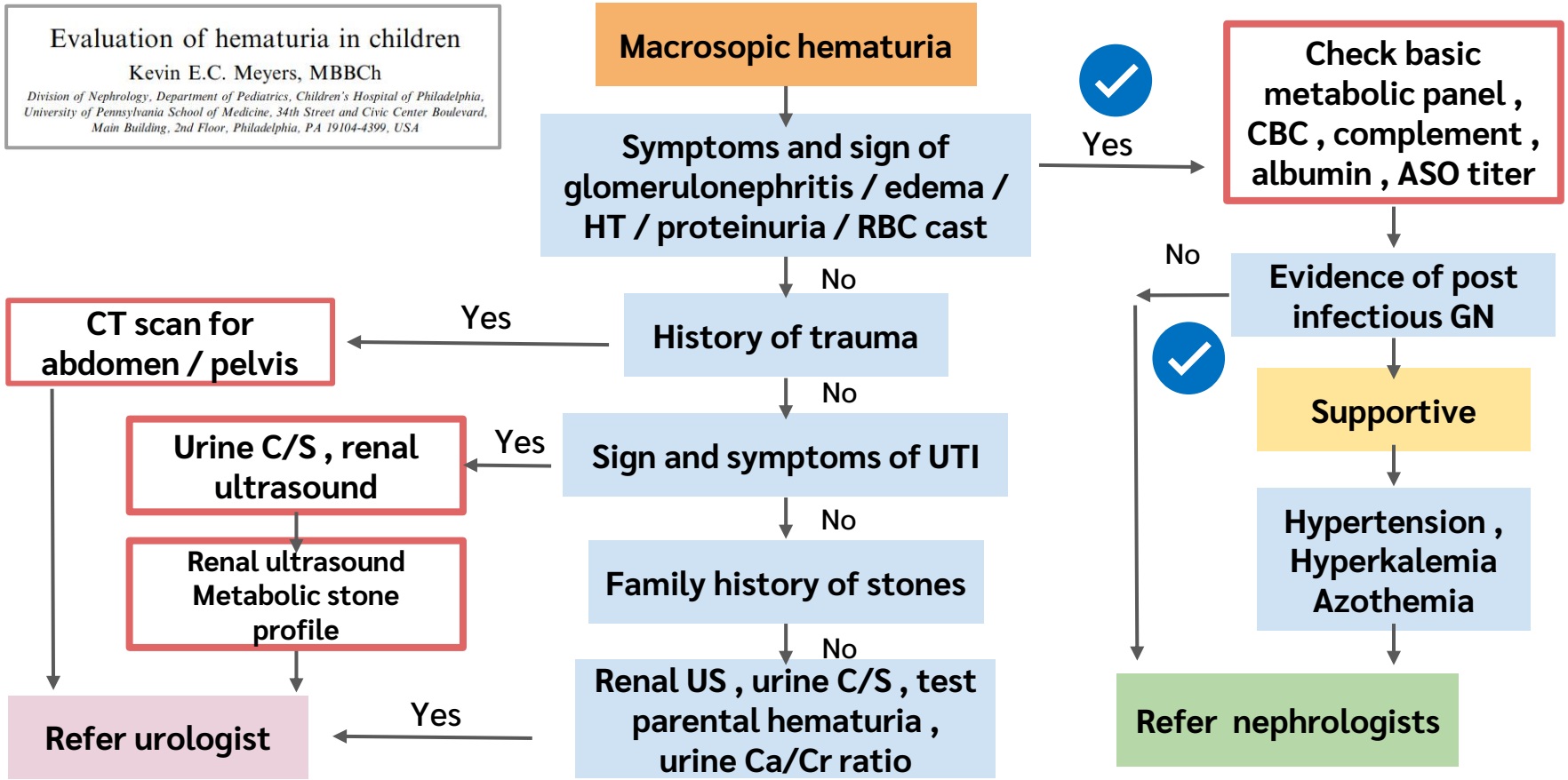
Problem list

A 6-years-old Thai girl presented with
gross hematuria with significant proteinuria
with stage I hypertention
with recent upper respiratory tract infection

Approach to this patient health problems

Approach to Hematuria by symptoms and signs

Evaluation of hematuria in children
Kevin E.C. Meyers, MBBCh
Division of Nephrology, Department of Pediatrics, Children's Hospital of Philadelphia, University of Pennsylvania School of Medicine, 34th Street and Civic Center Boulevard, Main Building, 2nd Floor, Philadelphia, PA 19104-4399, USA



History and Physical signs for approach hematuria

Box 4. Specific history and physical examination in a patient with hematuria

History
Trauma (recent bladder catheterization, blunt abdominal trauma)
Exercise
Menstruation
Recent sore throat, skin infection
Viral illness
Dysuria, frequency, urgency, enuresis
Urine color; stream discolored at initiation, throughout, or at termination of micturition
Abdominal pain, costovertebral angle pain, suprapubic pain
Medications (eg, cyclophosphamide), environmental toxins, or herbal compounds
Passage of a calculus
Joint or muscle pain

Family history
Hematuria
Deafness
Hypertension
Coagulopathy
Hemoglobinopathy
Calculi
Renal failure, dialysis, or transplant

Physical examination
Fever, arthritis, rash
Blood pressure
Edema
Nephromegaly
Costovertebral angle tenderness

(*Adapted from* Lieu TA, Grasmeder M, Kaplan BS. An approach to the evaluation and treatment of microscopic hematuria. Pediatr Clin North Am 1991;38:579–92.)

Box 5. Drugs and toxins associated with urine dipsticks positive for blood

Hemoglobinuria
Carbon monoxide
Mushrooms
Naphthalene
Sulfonamides
Tin compounds
Lead
Methicillin
Phenol
Sulfonamides
Turpentine
Ticlodipine [14]

Hematuria
Amitriptylene
Anticoagulants
Aspirin
Chlorpromazine
Cyclophosphamide
Toluene [13]
Ritonavir, indinavir [15]

(*Adapted from* Lieu TA, Grasmeder M, Kaplan BS. An approach to the evaluation and treatment of microscopic hematuria. Pediatr Clin North Am 1991;38:579–92.)

Box 3. Urine color

Dark yellow or orange
Normal concentrated urine
Rifampin pyridium

Dark brown or black
Methemoglobinemia
Bile pigments
Homogentesic acid, thymol, melanin, tyrosinosis, alkaptonuria
Alanine, cascara, resorcinol

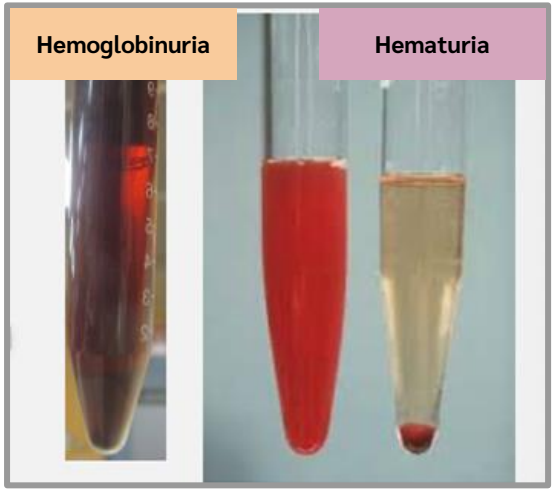
Red or pink urine
RBCs, free hemoglobin, myoglobin, porphyrins
Benzene, chloroquine, desferoxamine, phenazopyridine, phenolphthalein
Beets, blackberries, red dyes in food
Urates

Approach to Hematuria by urine analysis

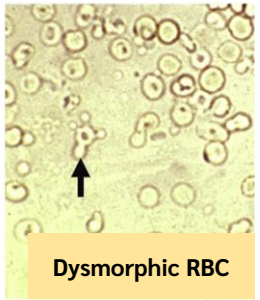
Hematuria
- Gross : Red , Tea-colored , Dark cola
- Microscopic hematuria

Confirm by urine dipstick & examination

- Dipstick Negative → No RBC → Pigmented uria
- Dipstick Positive
  - No RBC → Plasma color
    - Red/Pink → Hemoglobinuria
    - Clear → Myoglobinuria
  - RBC seen → True Hematuria
    - Glomerular hematuria
    - Hx , PE , UA
    - Non glomerular hematuria




RBC cast


Dysmorphic RBC

True Hematuria

Glomerular Hematuria

- UPCR
- BUN Cr Elyte
- Albumin

If indicated

- C3 , C4
- ASO titer , Anti-DNaseB
- P-ANCA , C-ANCA
- Anti-GBM Ab
- Hearing test
- UA in family members

Post infectious glomerulonephritis : APSGN
Primary glomerular disease : IgAN , MPGN , FSGS
Systemic : SLE , HSP , ANCA associated vasculitis
Hereditary GN : Alport , TBMN

Non glomerular Hematuria

- Urine C/S

If indicated

- Urine Ca / Cr ratio
- Renal US +- Doppler
- CBC
- Coagulogram
- Abdominal xray
- CT xray

UTI , Trauma
Vascular : RVT , Nutcracker
CAKUT : PKD , MCDK
Tumor : Wilms ,Rhabdomyosarcoma
Tubulointerstitial nephritis
Stone / Nephrocalcinosis
Excercise induce hematuria
Med : Cyclophosphamide

Features	Glomerular	Non glomerular
Dysuria	-	+/-
Abdominal Flank pain	-	+/-
Timing	Total	Initial / Terminal
Edema	+	-
Hypertension	+	+/-
Extrarenal	rash arthritis	abd mass
Dysmorphic RBC,RBC Cast	+	-
Clot	-	+/-
Proteinuria	> 1+	<=1+
Crystal	-	+/-

Investigation for this patient

Lab investigation : Urine analysis

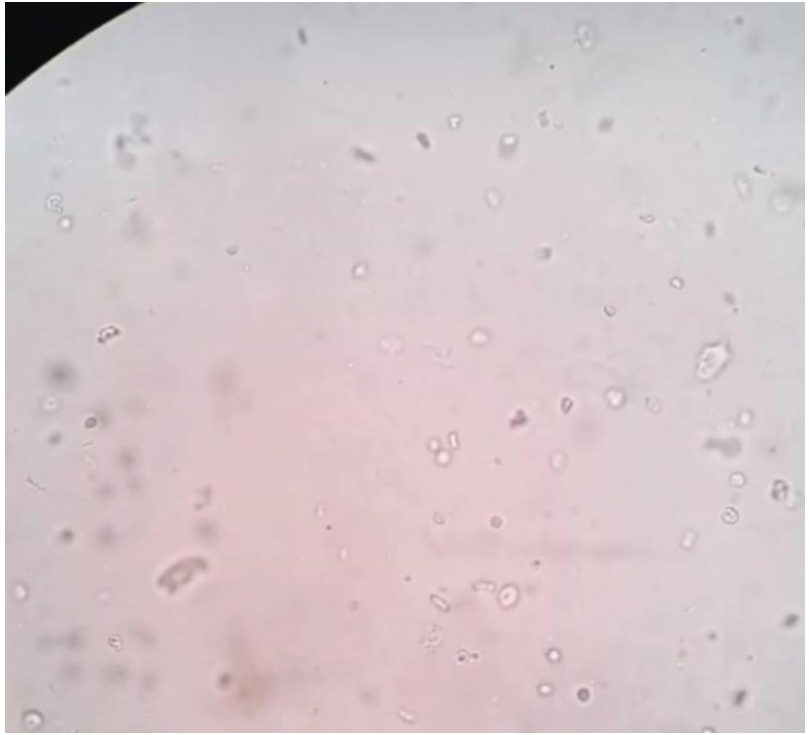
	color	Sp.gr.	pH	Blood	Protein	Glu	WBC	RBC	Epi
7/9/66	yellow /clear	1.006	7.5	3+	2+	Neg	1-2	30-50	0-1
3/10/66	yellow/ turbid	1.012	6.5	3+	1+	Neg	3-5	30-50	0-1
11/10/66	yellow /clear	1.010	7	2+	1+	Neg	5-10	30-50	0-1

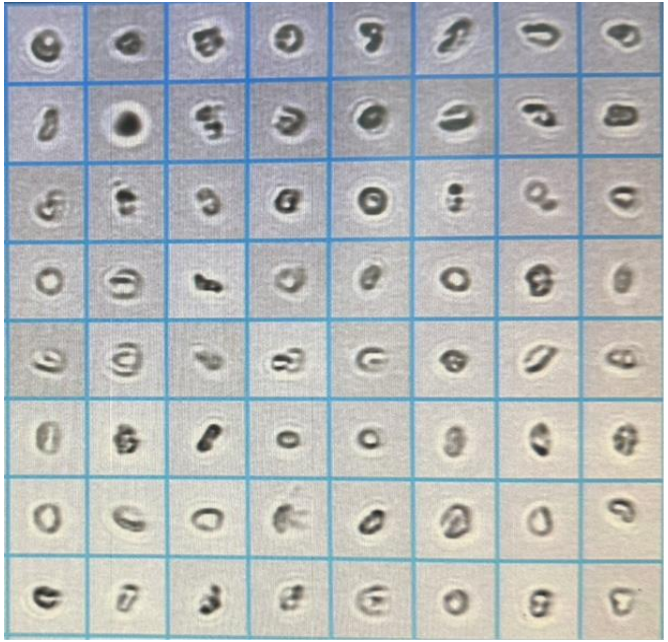
UPCR date	7/9/66	3/10/66	11/10/66
UPCR result	1.1 mg/mg	1.4 mg/mg	2.0 mg/mg

UPCR >__ significant proteinuria
UPCR > __ nephrotic range proteinuria

ผล UA บิดามารดา no hematuria / proteinuria

Urine microscopic examination





Dysmorphic RBC : acanthocytes , irregular erythrocytes with disrupted basement memnraneand vesicles on outer surface

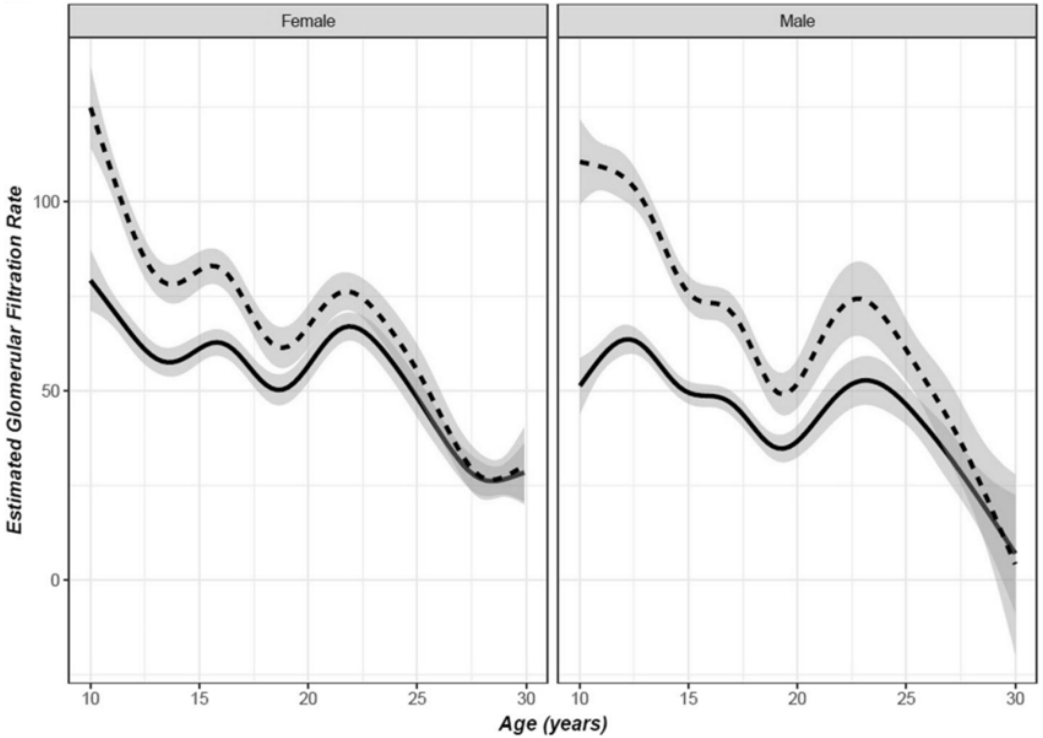
CBC	7/9/66	unit
Hb	12.5	g/dL
Hct	39	%
WBC	5,850	cells
Neutrophil	35	%
Lymphocyte	54	%
Monocyte	9	%
Basophil	1	%
MCV	74.6	fL
RDW	13.4	%
Platelet	475,000	cells

eGFR = 97.3 ml/min/1.73m^2

Electrolyte	7/9/66	unit
Na	136	mmol/L
K	4.1	mmol/L
Cl	102	mmol/L
HCO3	23	mmol/L
Ca	9.6	mmol/L
Mg	2.8	mmol/L
P	4.2	mmol/L
BUN	9	mg/dL
Cr	0.5	mg/dL
Albumin	3.6	mg/dL

Nasal swab for influenza A : positive

Tips : specific issues of kidney fuction measurement



Webster-Clark et al. BMC Nephrology (2018) 19:194

Transition-Age Period 16-18 years

Adolescents eGFR calculation changes as soon as they turn 18 years of age

Bedside Schwartz formula	only for height

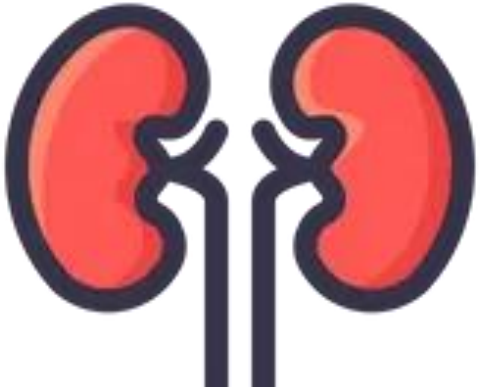
eGFR = 0.413 x Ht(cm) / Scr (mg/dL)

CKD-EPI formula	sex race height weight

Factors of person associated with GFR calculation : Growth , Muscle mass , Bodies habbitus , Health status , Illness

Lab : Complement , ASO titer , ANA , Anti DNaseB are pending form outsource lab

Approach and differential diagnosis of glomerulonephritis from those clinical and basic lab



Glomerulonephritis

1. Glomerular disease due to immune mediated damage	Postinfectious glomerulonephritis	APSGN
	IgA nephropathy	IgAN
	Primary glomerular diseae	MPGN , C3GN
	Systemic diseases	SLE with LN , IGAVN HSP
2. Glomerular disease due to an inherited abnormality of basement membrane collagens	Isolated microscopic hematuria	Thin basement membrane nephropathy
	Hematuria with extrarenal additional criteria	Alport syndrome

Glomerulonephritis

3. Glomerular disease due to thrombotic microangiopathy	Hemolytic uremic syndrome	EHEC induced , Strep. pneumo related HUS , Atypical HUS
	Thrombotic - thrombocytopenic purpura	TTP
4. Pauci - immune vasculitis	C-ANCA vasculitis	Granulomatosis with polyangiitis
	P-ANCA vasculitis	Eosinophilic granulomatosis with polyangiitis
5. Anti-GBM antibodies disease	Pulmonary renal syndrome	Goodpasture syndrome

Differential diagnosis from clinical

Suspected disease	Age group	Finding from Hx and PE	Pathogenesis	Invest.
1 IgA Nephropathy	6-18 years M : F 1 : 1	Synpharyngitic (URTI 2-3 days) Recurrent microscopic hematuria with or without proteinuria Hypertension Mixed nephritic-neprotic / RPGN	Hyperactivity of mucosal immune systems against antigens (virus , bacterial)	
2 APSGN	5-12 years M : F 2 : 1	Hematuria Edema HT proteinuria Nephritis after period of infection - URTI 1-2 wk - Skin infection 3-5 wk Rarely with nephrotic syndrome	M protien (nephrogenic strains) streptococcus pyogenes Immune complex mediated GN	
3 Alport syndrome	Child-Adult M > F 30%-50% of asymp. hemat	XLAS(80%) , ADAS(15%) ESRD and Bilat sensorineural hearing loss at 15 - 40 yrs (90%) Anterior lenticonus (M > F) (40y)	Mutation of the gene encoding for type IV collagen cause thin GBM	

Differential diagnosis from clinical

Suspected disease	Age group	Finding from Hx and PE	Pathogenesis	Invest.
4 Lupus nephritis	Adolescent F : M 4.5 : 1 (adolescent) > 1 : 100,000	Clinical by EULAR/ACR criteria Extrarenal menifestration Moderate proteinuria with microscopic RBC / RPGN	Loss of immune intolerance Genetics promoting organ specificity	
5 C3 glomerulopathy	5-17 years rare 1-2 per million	Micro/Macroscopic hematuria Protienuria (Neprotic>non nephro.) Hypertension Renal insufficiency	Terminal complement complex > influx of leukocyte > damage capillary wales from cytokines release	
Thin basement membrane nephritis 6	Child 7 yr Adult 37 yr F > M	Isolated hematuria Syntharyngitic GN AD , ⅓ de novo (familial hematuria) no ESRD or hearing loss	Mutation of the gene encoding for type IV collagen cause thin GBM	

From those differential diagnosis

Further investigation for this patient

Lab investigation

7/9/66	
ASO	200 IU/mL (<200)
Anti-DNase B	< 75 U/mL
C3	161 mg/dL (90-180)
C4	35 mg/dL (10-40)

10/10/66	
ASO	170 IU/mL (<200)
Anti-DNase B	pending
C3	114 mg/dL (90-180)
C4	17 mg/dL (10-40)

Discussion laboratory features of APSGN

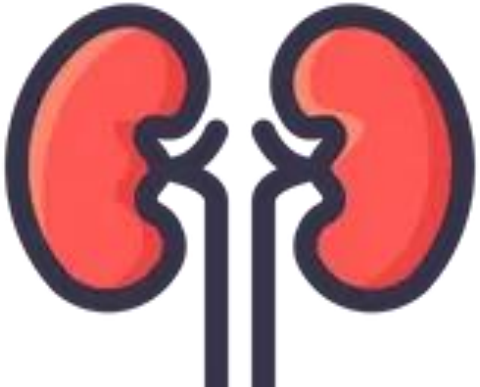
This patient lab result : ASO 200 IU/ml C3 161 mg/dL C4 35 mg/dL : Normal range

Serology	Sensitivity	Specificity	Onset	Peaks	Decline	Response
ASO titer	72.7%	93.2%	7-14 days	3-4 weeks	1-6 mo	URTI
Anti-DNAseB	70.5%	93.2%	2 weeks	6-8 weeks	3 mo	URTI/Skin

- Streptozyme test : positive 95% of APSGN form pharyngitis , 80% from from pyoderma
- Normal C3 is up to 10% and C4 may be slightly decrease
- Throat and skin C/S are negative in majority (75%)

Braun MC. Clinician's Manual of Pediatric Nephrology 2011 , Adapted from Srisuwan K. APSGN handout2021

Differential diagnosis of Glomerulonephritis



Glomerulonephritis complement approach

Normal C3 C4	Anti - GBM disease
	ANCA associated GN
	Ig A nephropathy / HSP
	Hereditary glomerulonephritis
Low C3 C4	Lupus nephritis in SLE
	MPGN with immunoglobulin deposit
	MPGN without immunoglobulin or complement deposit
Low C3 / Normal C4	APSGN MPGN with complement deposit
	PIGN ; bacterial endocarditis , shunt nephritis

Differential diagnosis

Suspected disease	Age group	Finding from Hx and PE	Complement	Investigation
1 IgA Nephropathy	6-18 years M : F 1 : 1	Synpharyngitic (URTI 2-3 days) Recurrent microscopic hematuria with or without proteinuria Hypertension Mixed nephritic-neprotic / RPGN	Normal C3 C4	Serum IgA : increase Galactose def. IgA1 LM IM EM : Biopsy
2 Alport syndrome	Child-Adult M > F	XLAS(80%) , ADAS(15%) ESRD and Bilat sensorineural hearing loss at 15 - 40 yrs (90%) Anterior lenticonus (M > F)	Normal C3 C4	Parental hematuria protienuria Hearing&Eye exam COL4A5 ,Renal Biopsy
Thin basement membrane nephritis	Child 7 yr Adult 37 yr F > M	Isolated hematuria Synpharyngitic AD , ⅓ de novo (familial hematuria) no ESRD or hearing loss	Normal C3 C4	Parental UA IM EM : Biopsy Gene study COL4A3 COL4A4

Differential diagnosis

Suspected disease	Age group	Finding from Hx and PE	Complement	Investigation
APSGN	5-12 years M : F 2 : 1	Hematuria Edema HT proteinuria Nephritis after period of infection - URTI 1-2 wk - Skin infection 3-5 wk Rarely with nephrotic syndrome	Low only C3	ASO titer Anti-DNaseB
Lupus nephritis	Adolescent F : M 4.5 : 1 (adolescent) > 1 : 100,000	Clinical by EULAR/ACR criteria Extrarenal menifestration Moderate proteinuria with microscopic RBC / RPGN	Low C3 C4	ANA , Anti-dsDNA Coombs test Anticardiolipin IgG Lupus anticoagulant
C3 glomerulopathy	5-17 years rare 1-2 per million	Micro/Macroscopic hematuria Protienuria (Neprotic>non nephro.) Hypertension Renal insufficiency	Low only C3	IgG IgA IgM ANA Anti-dsDNA Hepatitis serology

You’re a general pediatrics in this hospital and don’t have nephrologist

Further management for this patient

Management at รพ. นครปฐม

- Enalapril (5) 1.5 tab x 1 PO PC (0.13mg/kg/day)

For proteinuria reduction and blood pressure control

- Refer to PMK hospital for kidney biopsy

Refer to nephrologist for kidney biopsy

Decision tree for the consideration of a kidney biopsy in patients with proteinuria and/or glomerular hematuria

Patients classically treated without kidney biopsy diagnosis

Children:
- Steroid-sensitive nephrotic syndrome <12 yrs old
- Poststreptococcal GN

The kidney biopsy is the gold standard for diagnostic evaluation of glomerular diseases

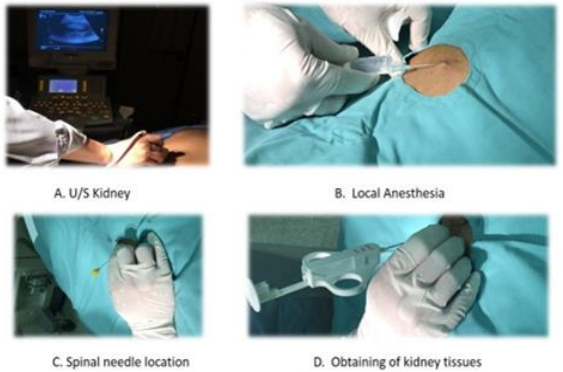
Biopsy should be performed if the biopsy result is expected to modify treatment and/or if additional prognostic information is needed

Examples of clinical conditions for which treatment may be considered without diagnostic confirmation by kidney biopsy:
- PLA2Rab+ membranous (especially with normal eGFR)
- MPO+ or PR3+ ANCA vasculitis
- Anti-glomerular basement membrane disease
- Alport disease
- Fabry disease
- Familial focal and segmental glomerulosclerosis in families with well-characterized mutations
- Biopsy contraindicated as judged by a synthesis of history, physical exam, and laboratory testing
- Systemic lupus erythematosus

KDIGO2021 figure2
Considerations for a kidney biopsy in patients with proteinuria and/or glomerular hematuria.

Indication for kidney biopsy

-**Glomerular causes**
-Steroid resistant nephrotic syndrome
-Congenital nephrotic syndrome
-Atypical nephrotic syndrome
-Rapidly progressive glomerulonephritis
-Non resolving PIGN
-Recurrent gross hematuria
-*Tubulo-interstitial nephritis*
-Acute kidney injury >4 wks w/o cause



Complication post percutaneous kidney biopsy

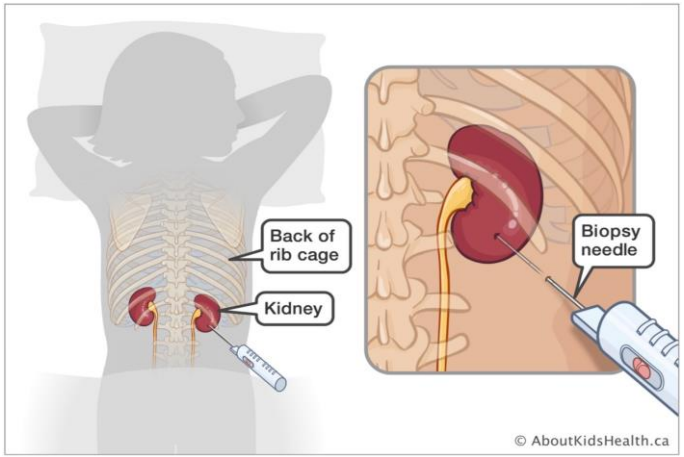
Native Kidney Biopsy: An Update and Best Practice Evidence

Ehab Mohammed[1], *Issa Al Salmi*[1*], *Shilpa Ramaiah*[1] and *Suad Hannawi*[2]

Nephrologist, The Renal Medicine Department, The Royal Hospital, Muscat, Oman

Major complications	Minor complications
PKBC transfusion 0.4-1.5%	Gross hematuria 0.3-14.5%
Nephrectomy 0.01%	Hematoma on CT 57-91%
Bladder obstruction 0.3%	
Death 0.02%	

Major complications (need for transfusion or invasive procedure, acute renal obstruction or failure, septicemia, or death occurred during the first 8 hours of observation, with 91% detected by 24 hours and 9% detected after 24 hours.



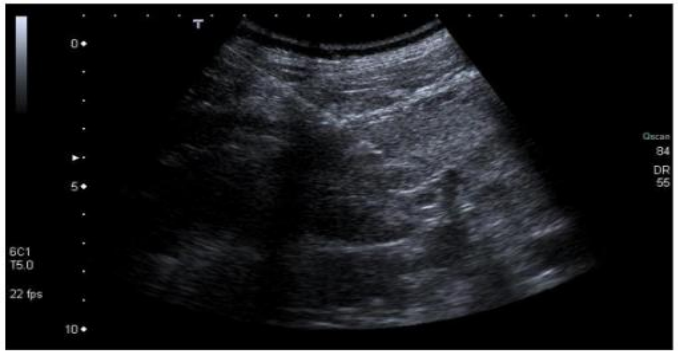
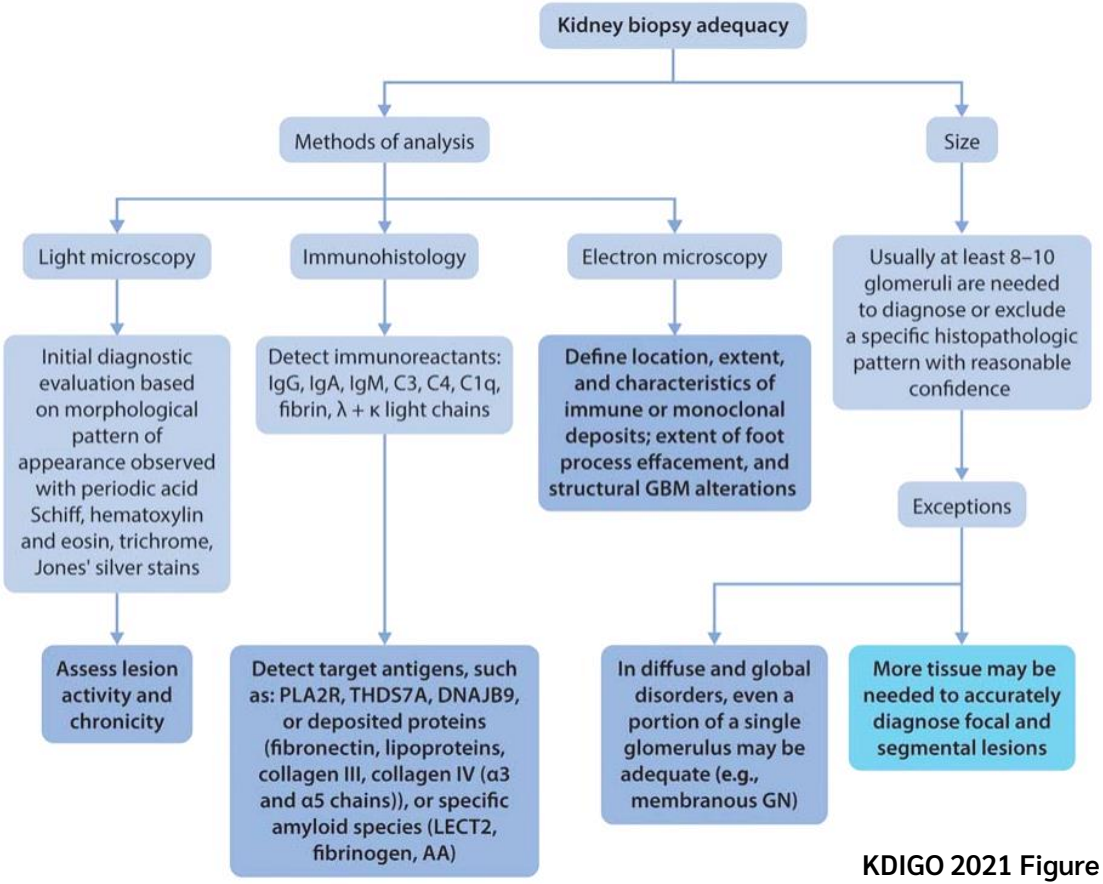
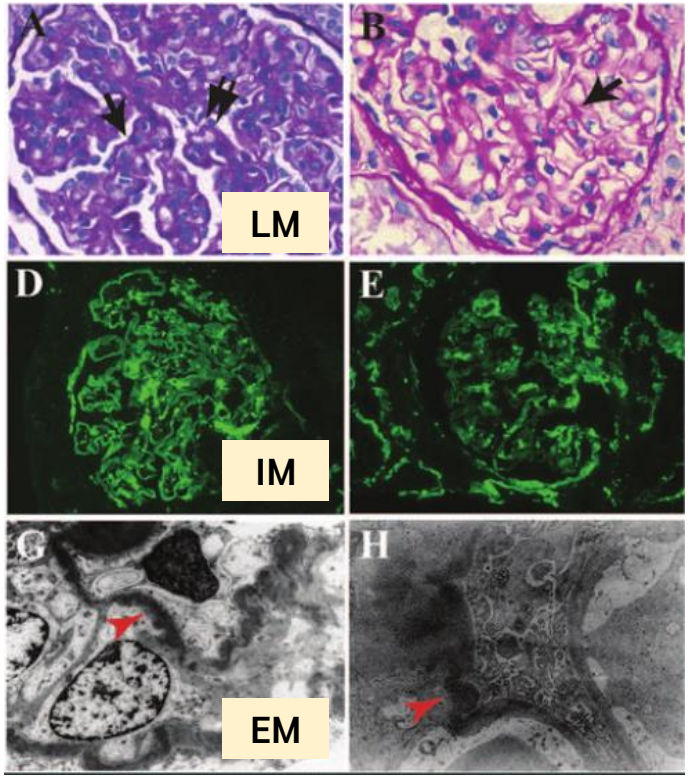


Figure 2 Needle pathway shown in ultrasonography.

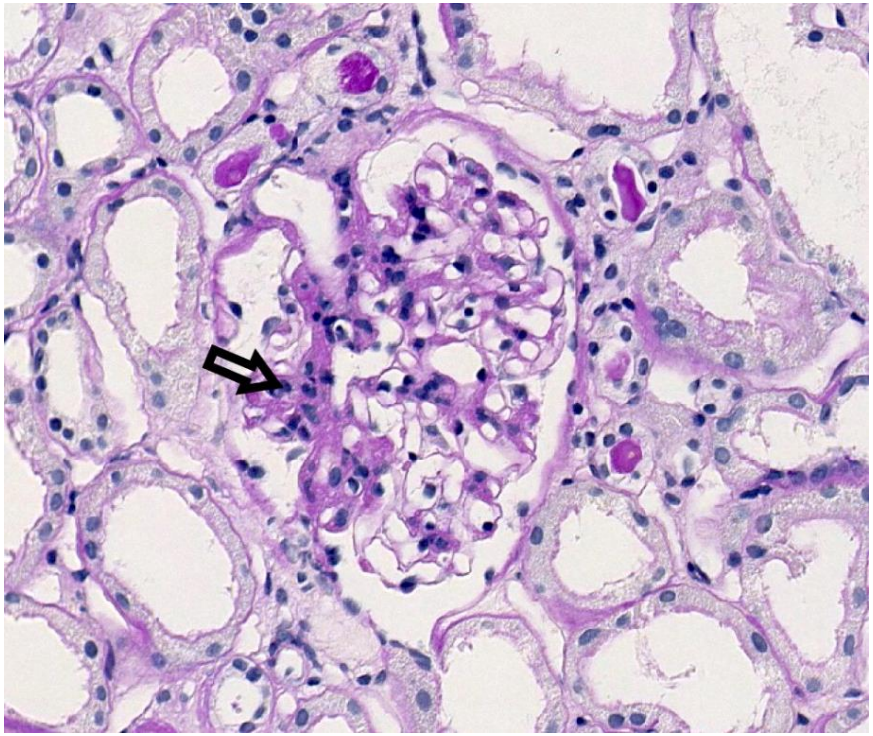
Kidney biopsy adequacy



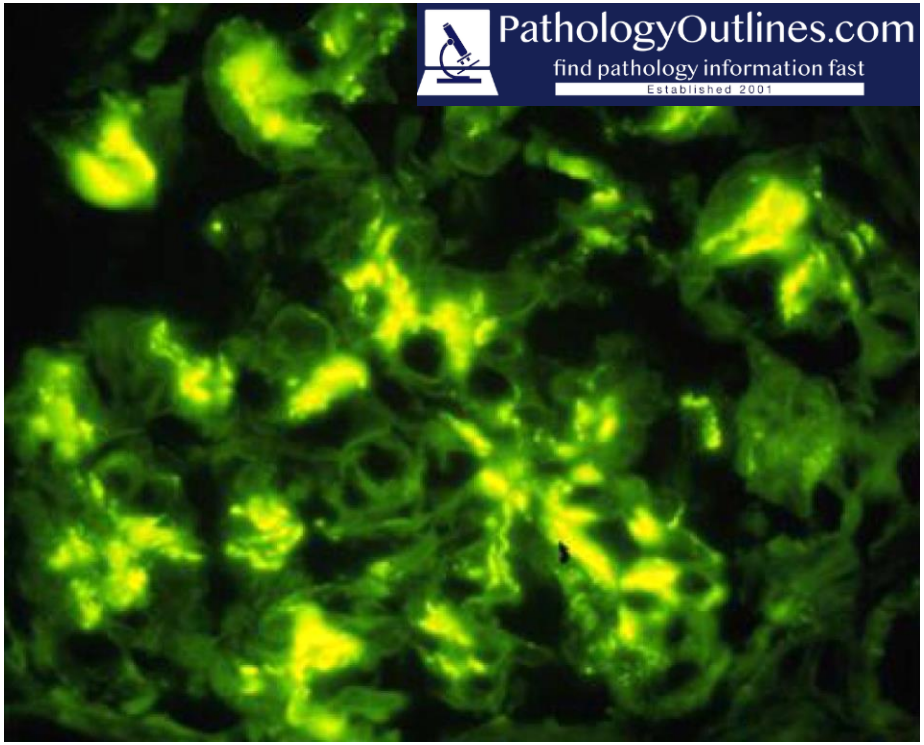


KDIGO 2021 Figure 3 | Evaluation of kidney tissue

Renal pathology in IgA nephropathy

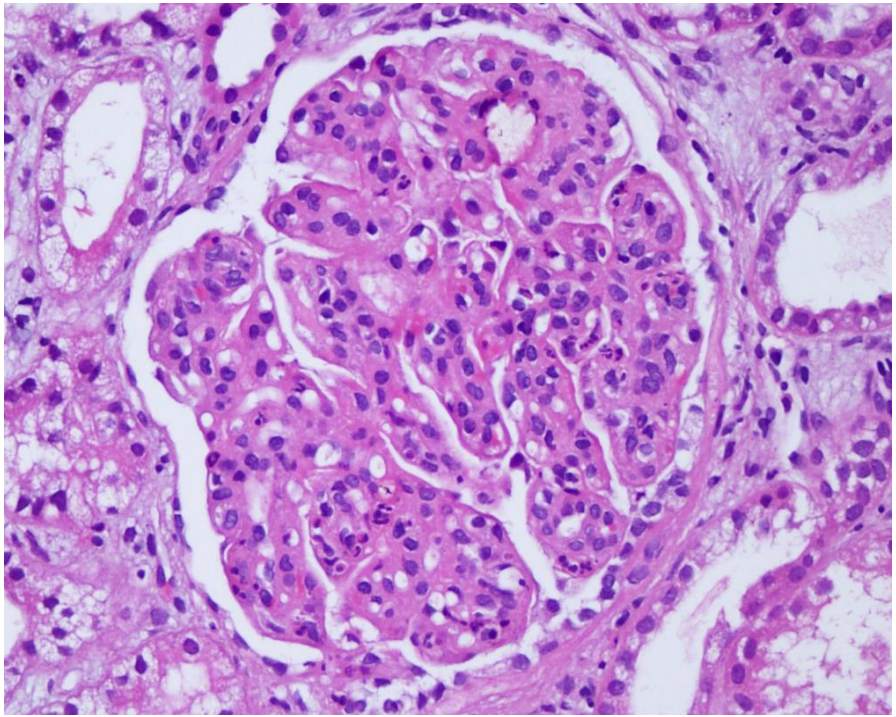


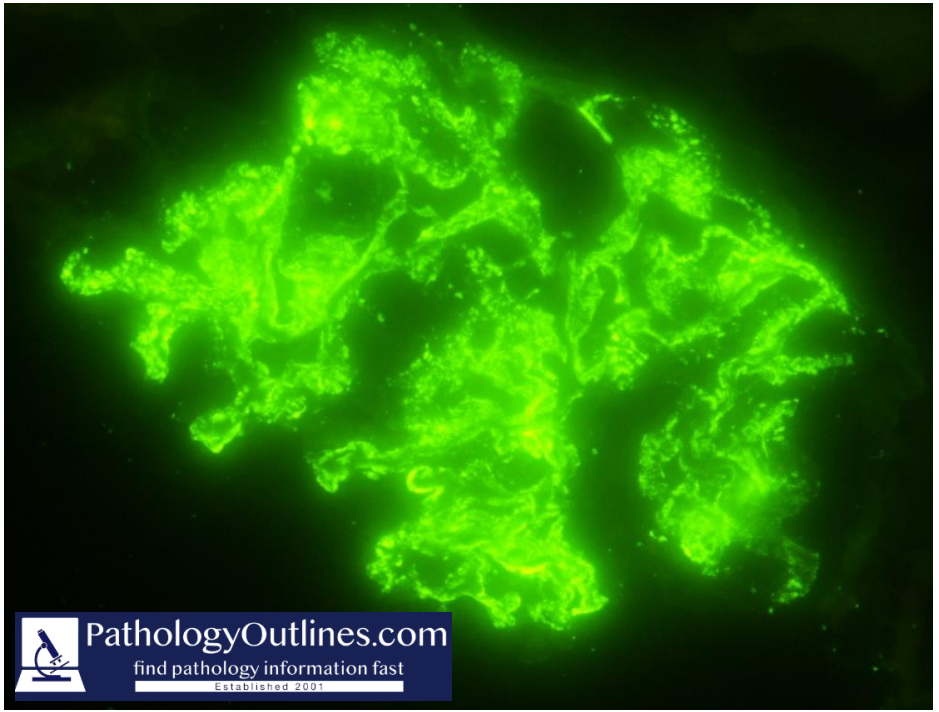
Light microscopic findings : Mesangial hypercellularity (≥ 4 cells/mesangial space) should be assessed in the mesangial spaces in PAS stained sections.



Immunohistologic findings : Presence of IgA in glomerular mesengium

Renal pathology in APSGN

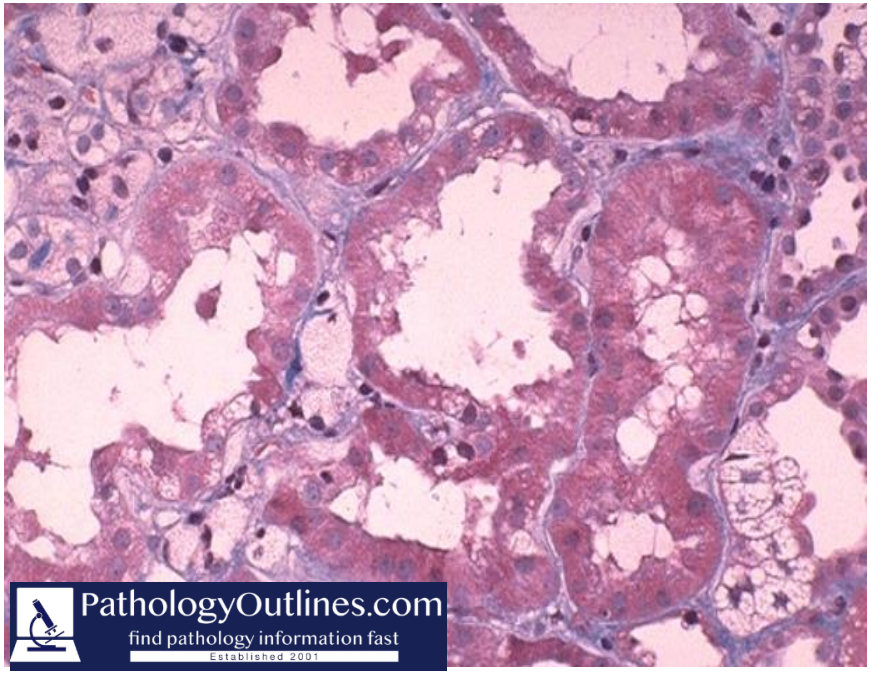




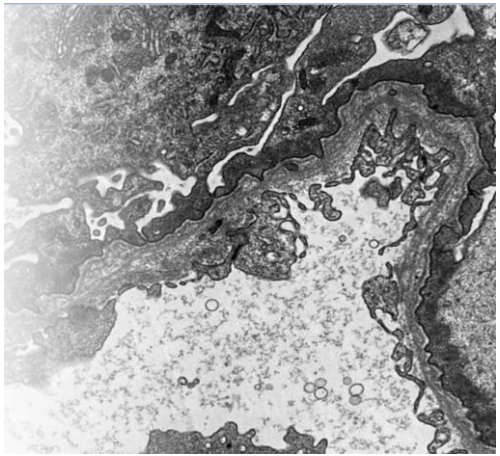
Light microscopic finding : Prominent endocapillary hypercellularity with numerous neutrophils both within capillary lumina and within mesangial area.

Immunohistologic findings : Coarse granular lumpy bumpy and garland pattern of immunofluorescence along glomerular basement membrane and in mesangium (IgG+ , C3+)

Renal pathology in Alport and Thin basement membrane

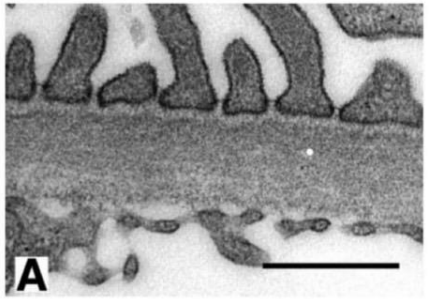


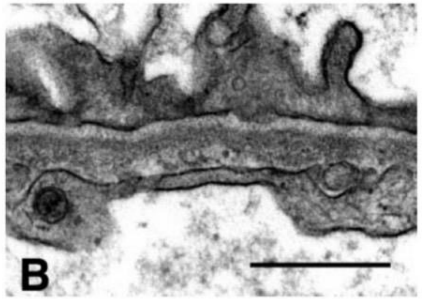
The glomeruli show irregular thickening and thinning and splitting of basement membranes due to an inherited abnormality in collagen



EM of AS
GBM becomes irregular with thin and thick segments an multi-laminated appearance

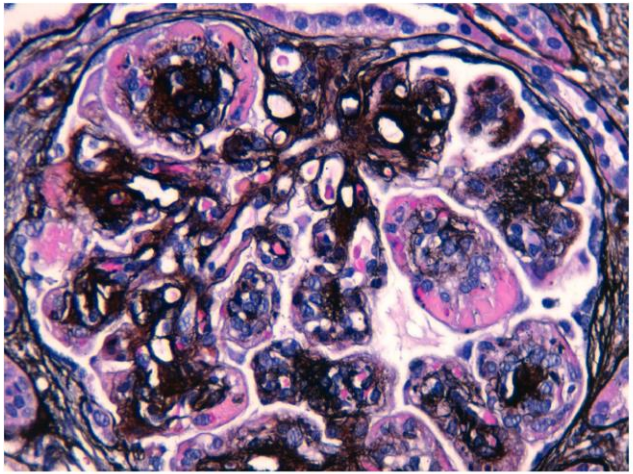
EM of TBM
Diffuse thinning of the GBM and uniform

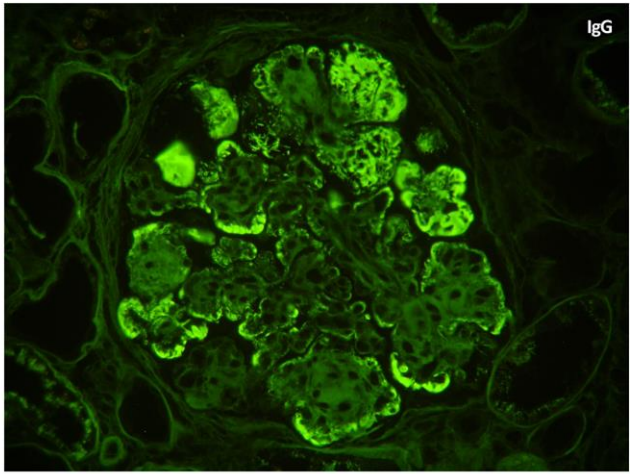




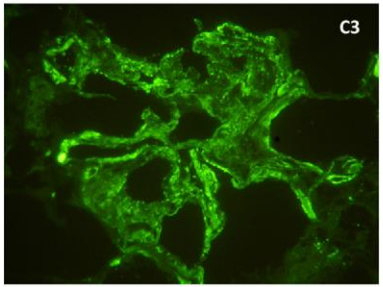
Renal pathology in C3 glomerulopathy

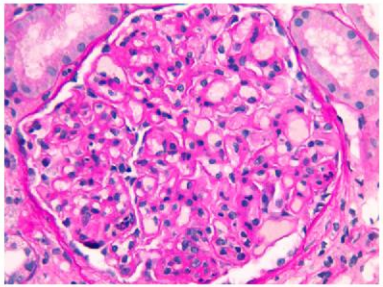
Illustrative case: MPGN I with immune complex Ig dominant (IgG), negative for C3. (Monoclonal gammopathy of renal significance).

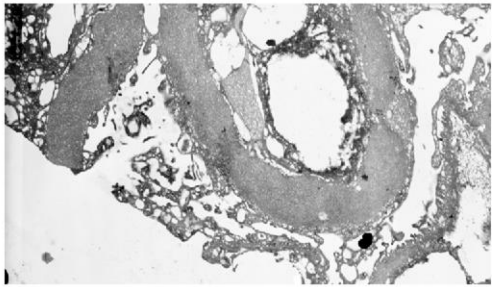




Illustrative case: DDD with exclusive C3 deposit, negative for immunoglobulin and with dense deposit (] by EM (C3 glomerulopathy).







Light microscope

- **mesengial proliferative pattern**
- **cresentic pattern**

IM

- **C3 intensely positive and dominant in both in GBM and mesangial regions**

Carvalho, Félix and Fernando Nolasco. "C3 Glomerulopathies: A New Category Encompassing Rare Complement Mediated Glomerulonephritis." (2016).

Pathological finding of kidney are pending

Management for this patient

Management in this patient

Hypertension control and proteinuria reduction	Use an ACEI or ARB to maximally tolerated - All child with IgAN and proteinuria > 200 mg/d or UPCR > 0.2 g/g should receive ACEI/ARB	• Do not stop if stable increase in serum creatinine • Stop ACEI/ARB if kidney function worsen and hyperkaleamia
	Target systolic blood pressure is < 90 th percentile for age,sex,Ht.	• KDIGO BP guidline
	Proteinuria goal is variable depending on primary disease process; < 1g/day	• Tritration of ACEI or ARB may cause AKI hyperkaleamia

KDIGO 2021 Figure 8,16 | Management of hypertension proteinuria and dietary in glomerular disease

Management in this patient

Dietary management	Restrict dietary sodium to control blood pressure and control proteinuria	• Dietary sodium < 2 g/d
	The safety of protein restriction in GN has not been established in children	• Nephrotic-range proteinuria : 0.8-1 g/kg/d protein intake in adult
Life style modifications	All GN patients as synergistic means for improving control of hypertension and proteinuria	• Normalize weight • Excercise regularly

KDIGO 2021 Figure 8,16 | Management of hypertension proteinuria and dietary in glomerular disease

Take home messages

- Macroscopic hematuria should prompt investigation
- Asymptomatic microscopic hematuria repeat UA and follow up clinical presentation first
- Patient with persistent or symptomatic proteinuria/hematuria : further investigation should be selected according to basic (Hx , PE , UA) findings
- Age group , extrarenal manifestation and renal manifestation are the key concept to approach GN
- Many of glomerular cause except APSGN need kidney biopsy for diagnosis