

## Board Review Practice Images



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# Disclosures

I have no financial disclosures or conflicts of interests with the presented materials in this presentation

# Question 1



**What is the diagnosis?**

- A. Central retinal artery occlusion**
- B. Diabetic retinopathy**
- C. Ocular toxoplasmosis**
- D. Optic neuritis**
- E. Malignant hypertension**

## B. Diabetic retinopathy



The fundus photograph shows findings consistent with a diagnosis of diabetic retinopathy.

### Features

- Microaneurysms
- Dot and blot hemorrhages
- Flame-shaped hemorrhages
- Retinal edema and hard exudates
- Cotton-wool spots
- Venous loops and venous beading
- Intraretinal microvascular abnormalities
- Macular edema

**Microaneurysms:** The earliest clinical sign of diabetic retinopathy; these occur secondary to capillary wall outpouching due to pericyte loss; they appear as small, red dots in the superficial retinal layers

**Dot and blot hemorrhages:** Appear similar to microaneurysms if they are small; they occur as microaneurysms rupture in the deeper layers of the retina, such as the inner nuclear and outer plexiform layers

**Flame-shaped hemorrhages:** Splinter hemorrhages that occur in the more superficial nerve fiber layer

**Retinal edema and hard exudates:** Caused by the breakdown of the blood-retina barrier, allowing leakage of serum proteins, lipids, and protein from the vessels

**Cotton-wool spots:** Nerve fiber layer infarctions from occlusion of precapillary arterioles; they are frequently bordered by microaneurysms and vascular hyperpermeability

**Venous loops and venous beading:** Frequently occur adjacent to areas of nonperfusion - reflect increasing retinal ischemia, and their occurrence is the most significant predictor of progression to proliferative diabetic retinopathy (PDR).

**Intraretinal microvascular abnormalities:** Remodeled capillary beds without proliferative changes; can usually be found on the borders of the nonperfused retina

**Macular edema:** Leading cause of visual impairment in patients with

Source: [Medscape.com/article/1225122-overview](https://www.medscape.com/article/1225122-overview)

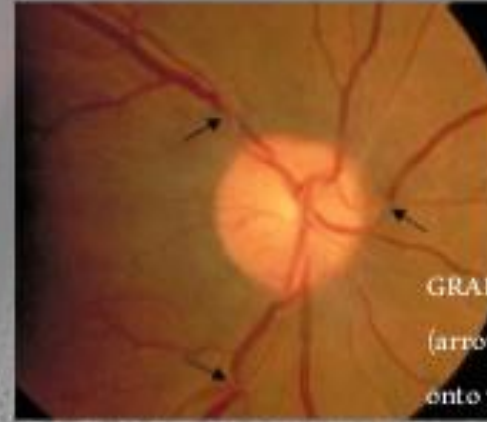


# Hypertensive retinopathy

GRADE 1: Tortuosity of retinal arteries and silver wiring



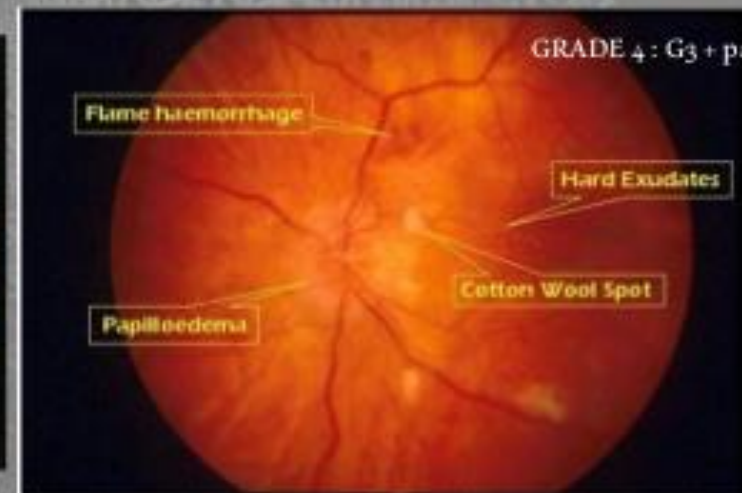
GRADE 2: G1 + AV nipping  
(arrow – artery cross over onto vein)



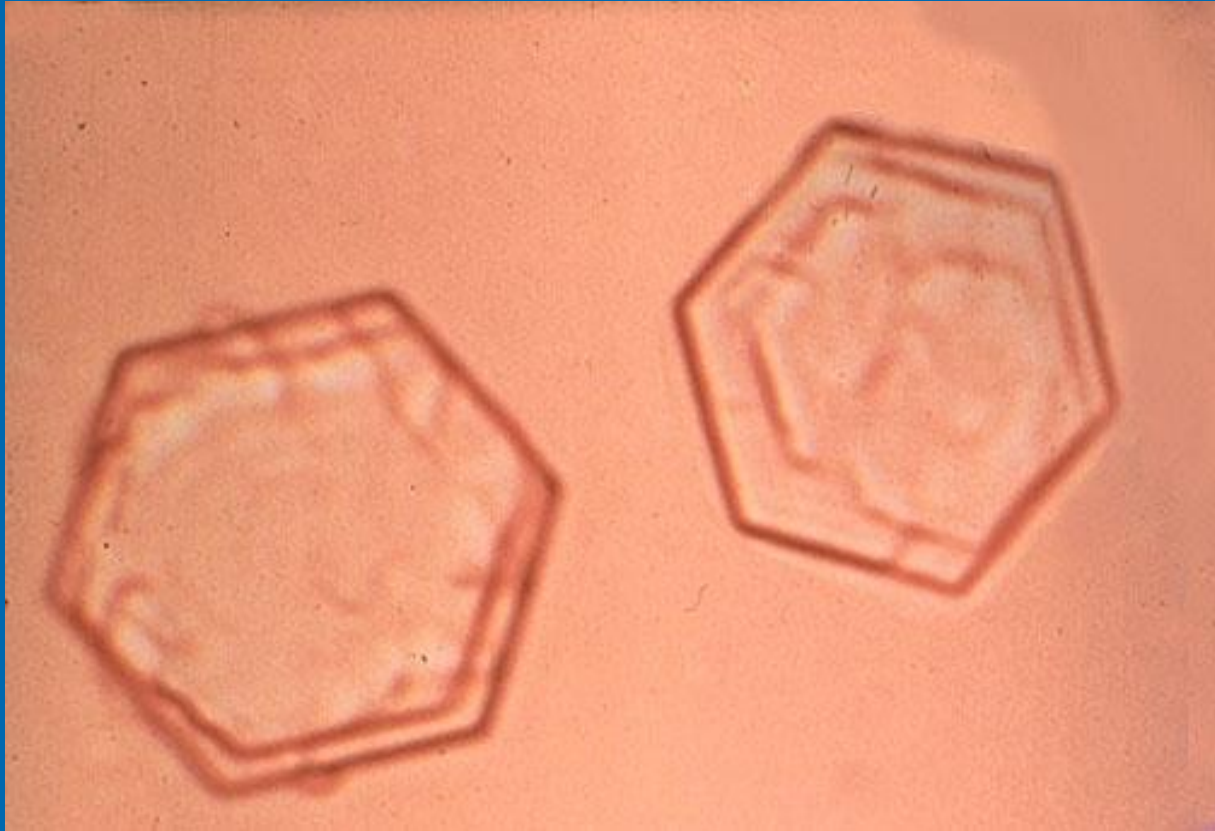
GRADE 3: G2 + flame-shaped haemorrhage and cotton wool exudate (whitish)



GRADE 4: G3 + papilloedema



# Question 2



The urine sediment shows what form of crystals?

- A. Oxalate crystals
- B. Cystine crystals
- C. Uric acid crystals
- D. Struvite crystals
- E. Acyclovir crystals

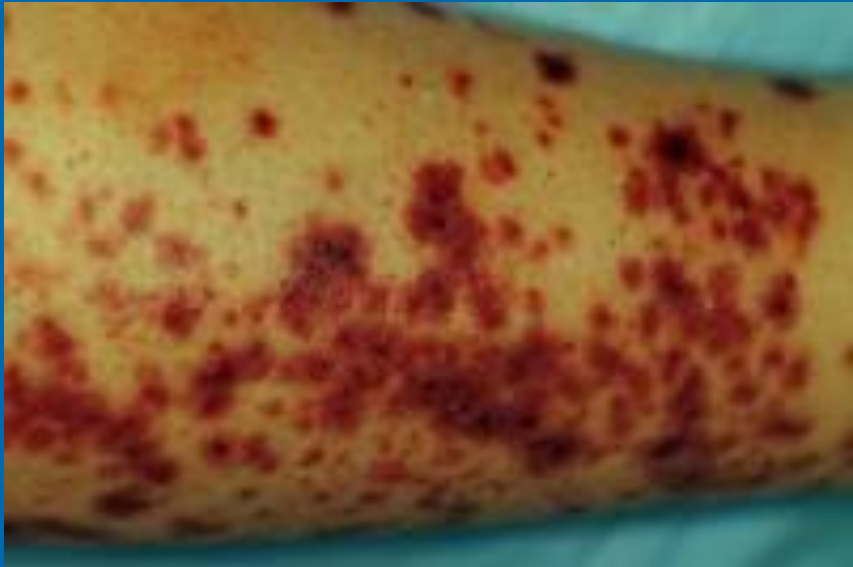


## B. Cystine crystals

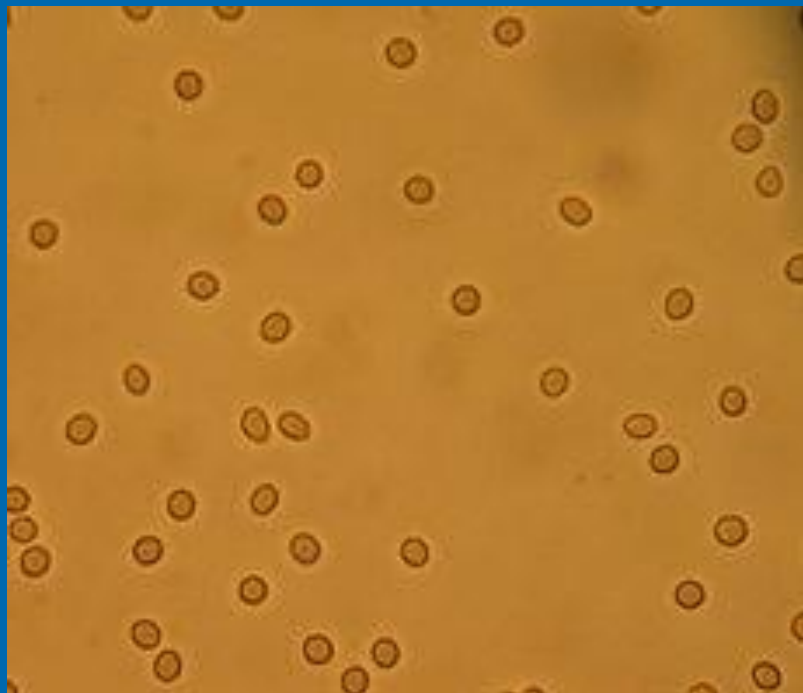


- The crystals are usually hexagonal, translucent, white
- Cystinuria is an autosomal recessive disorder caused by mutations in the [SLC3A1](#) and [SLC7A9](#) genes
- These genes encode two parts of a transporter protein in the kidneys
- Under normal circumstances, this protein allows certain amino acids, including cysteine, to be reabsorbed into the blood from the filtered fluid that will become urine
- As the levels of cystine in the urine increases, cystine crystals typical of form, leading to kidney stones

# Question 3



A 22-year old woman presents with joint and abdominal pain and a rash. Rectal exam is positive for occult blood. Urine shows hematuria. What is the most likely diagnosis?

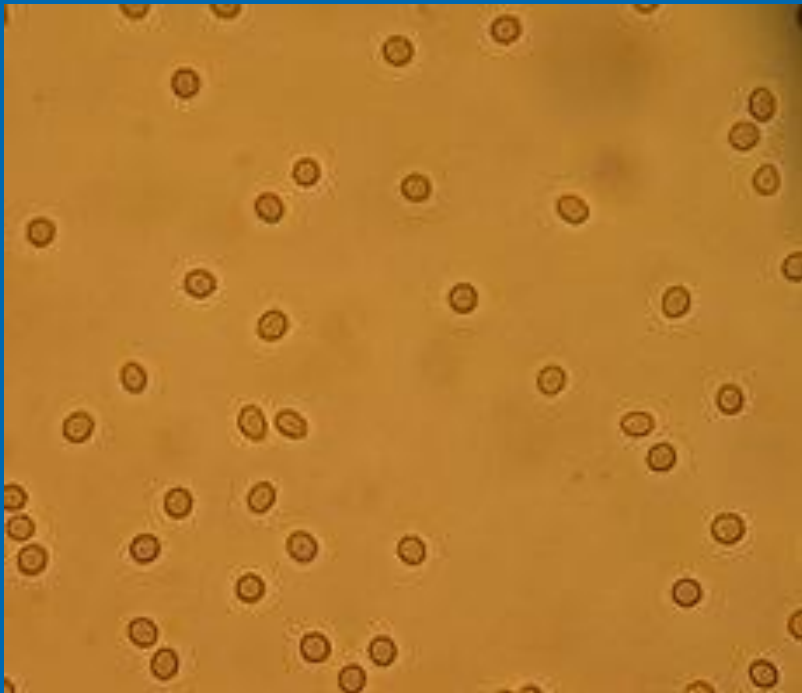
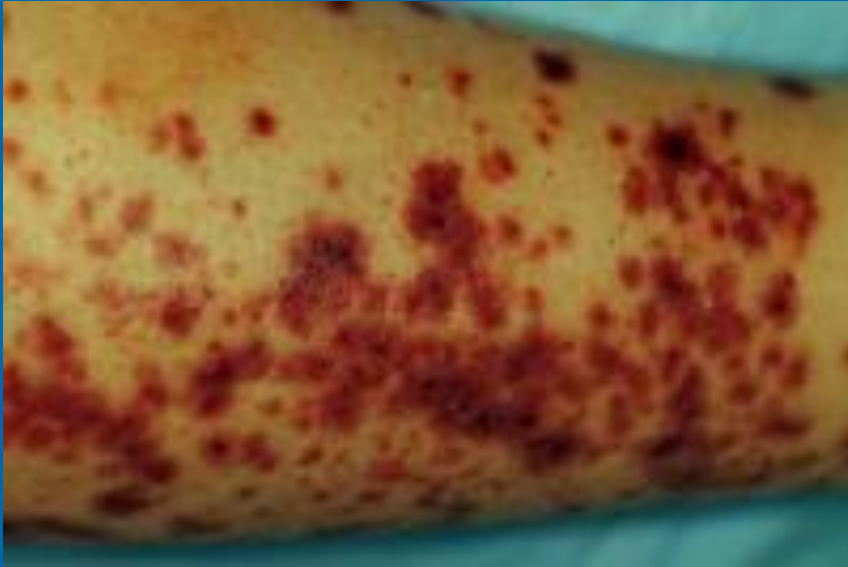


- A. Renal cell cancer
- B. Renal infarction
- C. Kidney stone
- D. Goodpasture's syndrome
- E. Henoch-Schonlein purpura

## E. Henoch-Schonlein purpura

The patient has palpable purpura and hematuria. The the red cells appear crenated - - dysmorphic red cells. The combination of the clinical presentation and the hematuria makes HSP nephritis the most likely etiology.

HSP is an IgA-mediated, small-vessel vasculitis that predominantly affects children but also is seen in adults. HSP is a subset of necrotizing vasculitis characterized by fibrinoid destruction of blood vessels

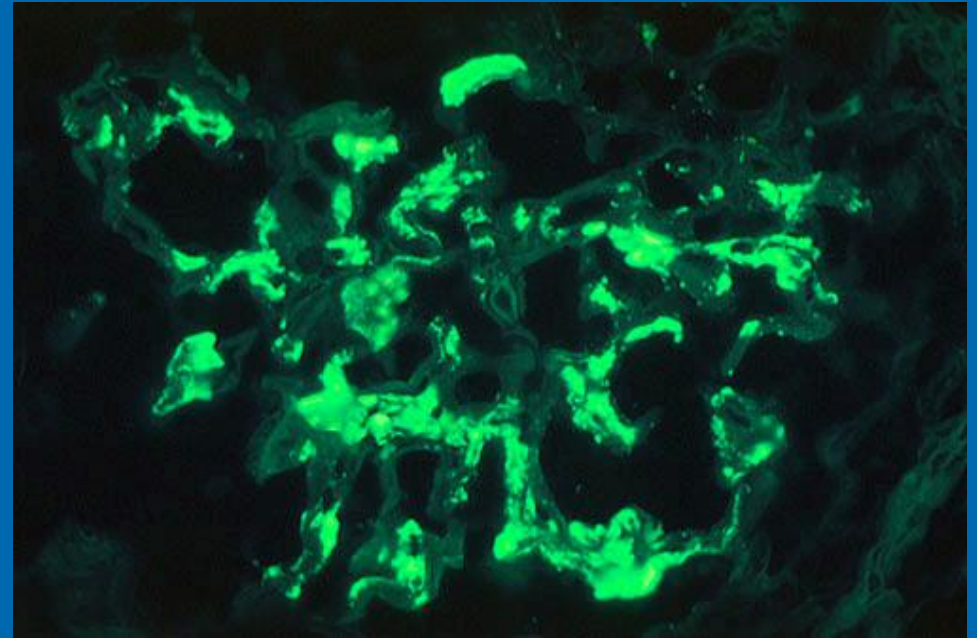
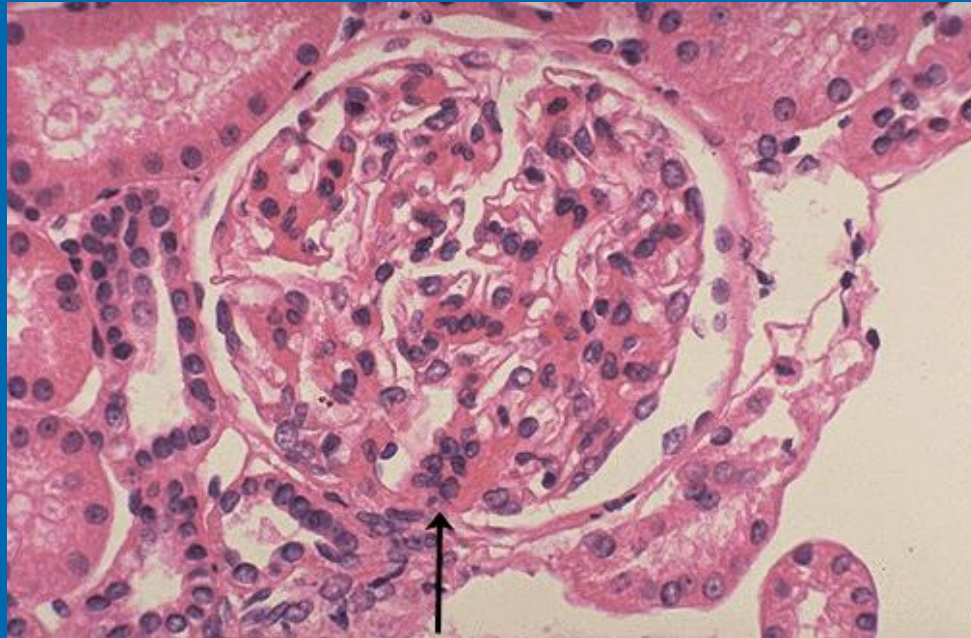


# Glomerular Syndromes

- Nephritis: Hypertension, Azotemia, proteinuria, hematuria, RBC casts / dysmorphic RBCs
- Nephrosis: edema, proteinuria, hypoalbuminemia, lipid abnormalities
- RPGN: rapid renal failure, crescents on renal biopsy + nephritis
- Isolated urinary abnormalities: hematuria / proteinuria



# Pathology of IgAN/HSP nephritis





# HSP nephritis

## *Clinical Features*

### **Dermal**

Purple, nonblanching,  
urticarial, purpuric papules  
may become confluent  
Bx: leukocytoclastic vasculitis

### **Renal**

33% children, 63% adults  
Hematuria, macroscopic / microscopic  
Proteinuria  
Azotemia

### **GI**

Abd pain (2/3rds of cases)  
may precede rash  
Vomiting  
Diarrhea  
Periumbilical pain  
Major complications (5%)  
    intussusception  
    bowel ischemia  
necrosis

### **Joints**

Arthralgias and periarticular edema (2/3)  
knees, ankles, elbows, wrists

# Question 4



**What is the most likely diagnosis?**

- A. Renal tubular acidosis**
- B. Primary hypoparathyroidism**
- C. Familial hypocalciuric hypercalcemia**
- D. Salicylate overdose**
- E. Paget's disease**



## A. Renal tubular acidosis

The film reveals bilateral symmetric calcification of the renal parenchyma, sparing only the renal pelvis. This patient had been diagnosed with renal tubular acidosis at 9 years of age, but did not undergo medical follow-up for 20 years. The other listed choices are not common causes of nephrocalcinosis.

Type	Type 1	Type 2	Type 4
Location	Collecting Tubules	Proximal tubules	Adrenal
Acidemia	Yes (severe)	Yes	Mild when present
Potassium	Hypokalemia	Hypokalemia	Hyperkalemia
Pathophysiology	Failure of $\alpha$ intercalated cells to secrete $H^+$ and reclaim $K^+$	Failure of proximal tubular cells to reabsorb $HCO_3^-$	Deficiency of aldosterone, or a resistance to its effects, (hypoaldosteronism or pseudohypoaldosteronism)

## Distal RTA

1. Classical form of RTA (described first)
2. Failure of  $\alpha$  intercalated cells to secrete  $H^+$  and  $K^+$
3. Hypokalemia, hypocalcemia, hyperchloremia
4. Urinary stone formation (related to alkaline urine, hypercalciuria and low urine citrate)
5. Nephrocalcinosis
6. Bone demineralization (rickets in children, osteomalacia in adults)

**Table 1: Differences between the two common types of renal tubular acidosis**

Feature	Proximal RTA	Distal RTA
Plasma potassium	Normal/Low	Normal/Low
Urine pH	<5.5	>5.5
Urine anion gap	Positive	Positive
Fractional bicarbonate excretion	>10-15%	<5%
Urine calcium excretion	Normal	High
Nephrocalcinosis and bone disease	Absent	Present
Other tubular defects	Often present	Absent
Treatment	5-20 mEq/kg/day of $\text{NaHCO}_3$	$\text{NaHCO}_3$ 1-2 mEq/kg/day in divided dosage

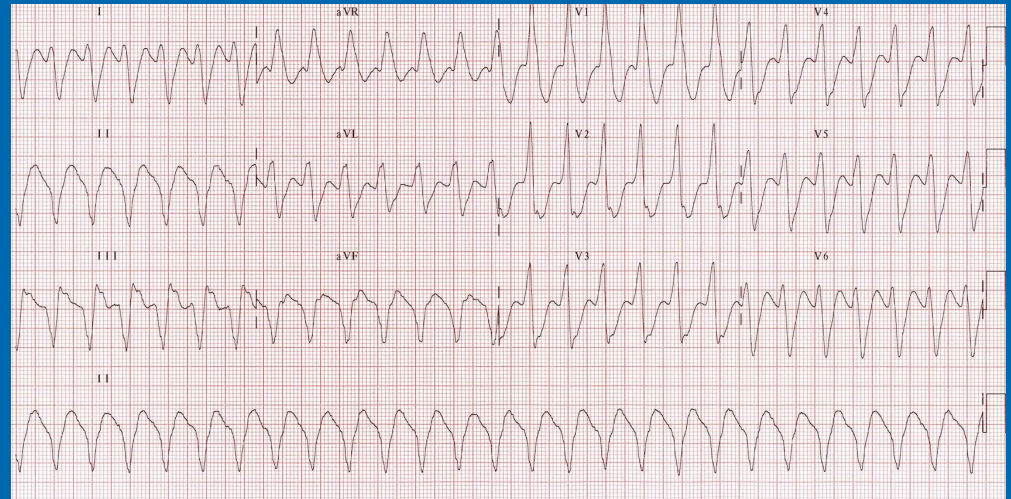


# Question 5

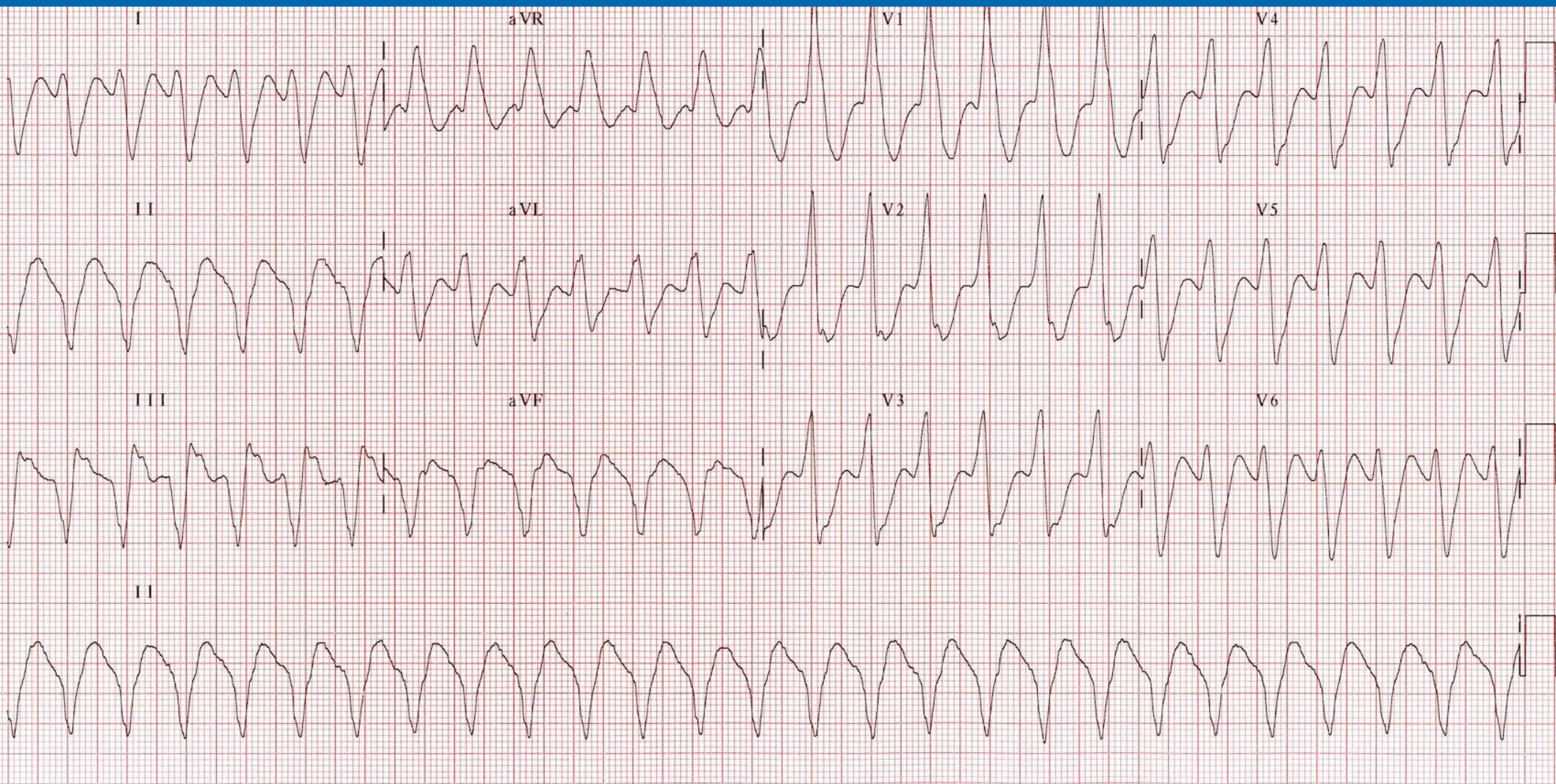
A middle-age white female is brought into the ED after suffering what appears to be a fall while jogging in Larz Anderson Park. She is unresponsive, has a faint pulse and labored breathing. SBP 55 mm/Hg, HR 278 bpm. She has scratches and lacerations on her face and trunk.

The ECG is shown:

- A. Atrial fibrillation
- B. Ventricular fibrillation
- C. Ventricular tachycardia
- D. Electrical artefact
- E. Atrial flutter with 2:1 block

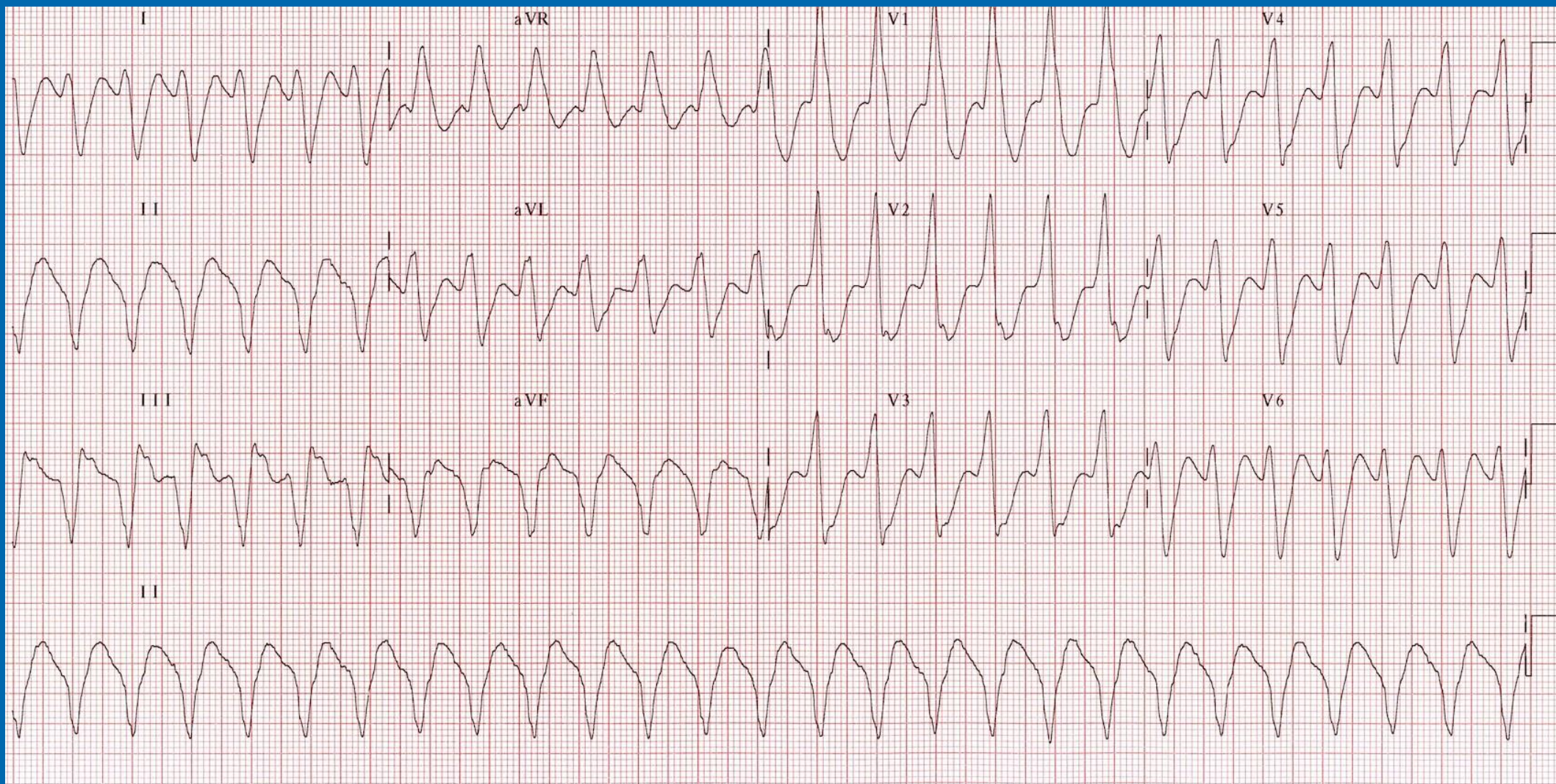






- A. Atrial fibrillation
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## C. Ventricular Tachycardia

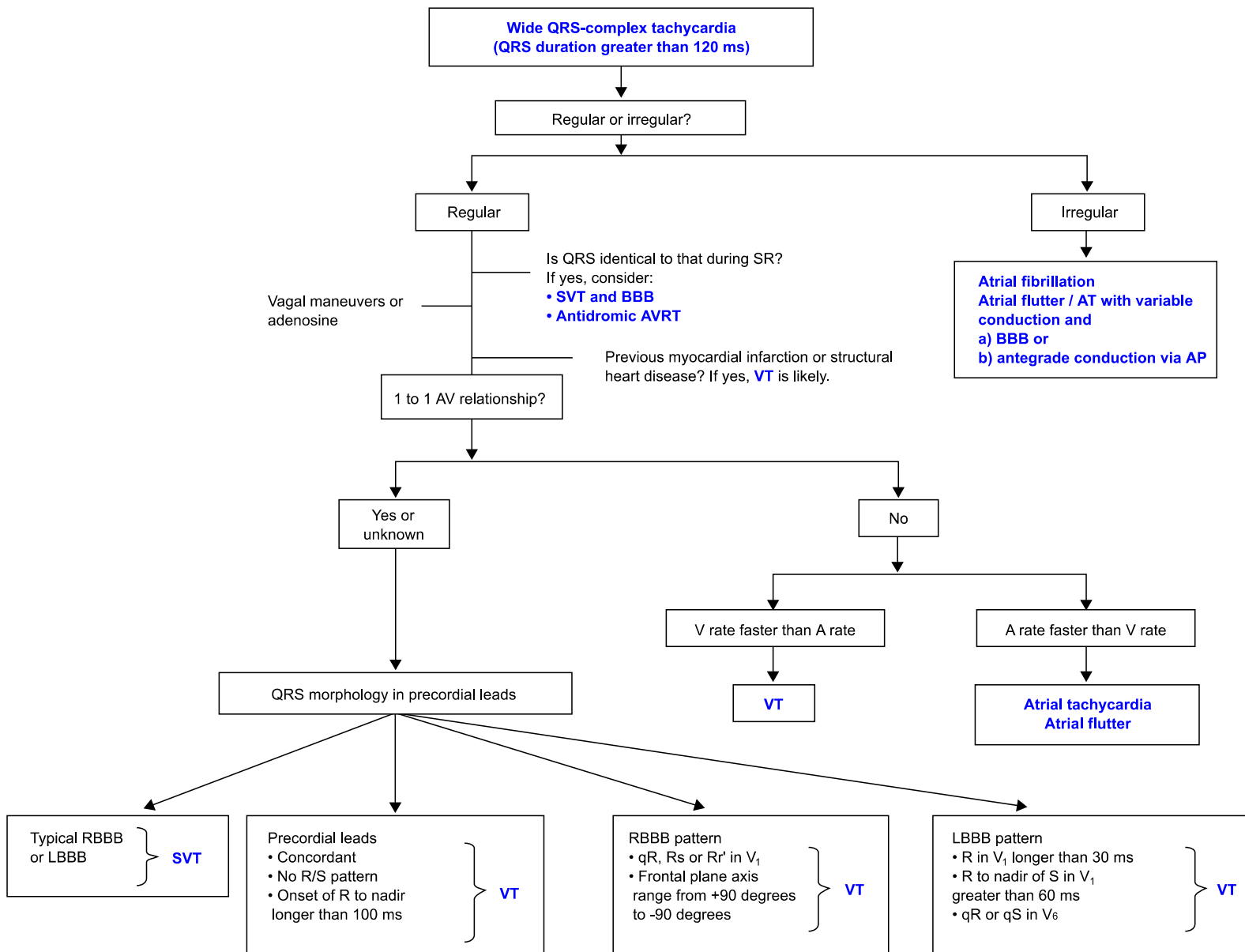
Rapid heart rate ( $> 100$  bpm).

Broad QRS complexes ( $> 120$  ms).



# VT

- Impairs cardiac output with consequent hypotension, collapse, and acute cardiac failure.
  - extreme heart rates and lack of coordinated atrial contraction (loss of “atrial kick”)
- Prompt recognition and initiation of treatment (e.g. electrical cardioversion) is required in all cases of VT
- **VT can be Sustained** = Duration > 30 seconds or requiring intervention due to hemodynamic compromise.  
**Or, Non-sustained** = Three or more consecutive ventricular complexes terminating spontaneously in < 30 seconds.
- **VT can be Hemodynamically stable or Hemodynamically unstable** — e.g hypotension, chest pain, cardiac failure, decreased conscious level.





# Question 6



A 78-year-old male presented with headache of 4 weeks' duration. Pain was excruciating, constant and predominantly over the right hemicranium, with the maximum being over the right temple. There was jaw and tongue claudication. The patient felt weak and there was low grade fever. He also **complained of proximal and axial joint arthralgias.** He had tenderness over his superficial temporal artery and over his scalp.

SOURCE:

[https://www.google.com/search?q=temporal+arteritis&biw=1327&bih=661&source=lnms&tbm=isch&sa=X&sqi=2&ved=0ahUK EwjdyPjXmufNAhUCDT4KHWfWD9kQ\\_AUIBigB#imgsrc=1Boe GCHqqtZAPM%3A](https://www.google.com/search?q=temporal+arteritis&biw=1327&bih=661&source=lnms&tbm=isch&sa=X&sqi=2&ved=0ahUKEwjdyPjXmufNAhUCDT4KHWfWD9kQ_AUIBigB#imgsrc=1BoeGCHqqtZAPM%3A)

SOURCE: <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2771971/>

A diagnosis of giant cell arteritis (GCA) was confirmed by a superficial temporal artery (STA) biopsy. Which one of the following is atypical of GCA?

- A.) Jaw claudication
- B.) Lower extremity claudication
- C.) Tongue claudication
- D.) Proximal and axial joint arthralgias suggesting polymyalgia rheumatica (PMR)
- E.) Scalp tenderness

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# GCA

- Most common vasculitis
- Elderly women
- **Small, medium size arteries**
- Cranial Arteritis: temporal, facial, ophthalmic arteries. Also aortic arch (giant cell aortitis, uncommon)
- Throbbing headache that doesn't respond to NSAIDs, tender temporal area, firm & nodular temporal artery
- Facial pain, loss of taste, **tongue pain/ Claudication, scalp tenderness**
- **Jaw claudication**
- Visual abnormalities: amaurosis fugax, blurred vision, double vision, blindness
- **50% cases => Polymyalgia Rheumatica**
- Necrotizing vasculitis with granulomas: fragmentation of the internal elastic lamina due to the presence of multinucleated Giant Cells & intimal fibrosis.

# GCA Treatment

## ORIGINAL ARTICLE

### Trial of Tocilizumab in Giant-Cell Arteritis

John H. Stone, M.D., M.P.H., Katie Tuckwell, Ph.D., Sophie Dimonaco, M.Sc., Micki Klearman, M.D., Martin Aringer, M.D., Daniel Blockmans, M.D., Ph.D., Elisabeth Brouwer, M.D., Ph.D., Maria C. Cid, M.D., Bhaskar Dasgupta, M.B., B.S., M.D., Juergen Rech, M.D., Carlo Salvarani, M.D., Georg Schett, M.D., Hendrik Schulze-Koops, M.D., Ph.D., Robert Spiera, M.D., Sebastian H. Unizony, M.D., and Neil Collinson, Ph.D.

N Engl J Med 2017; 377:317-328 | July 27, 2017 | DOI: 10.1056/NEJMoa1613849

Stone JH et al N Engl J Med 2017; 377:317-328 July 27, 2017

N=251

Pred (26 w course) + tocilizumab (162 mg SC weekly/QOW)  
vs.

Pred (26 w or 52 w course) +placebo

Outcome: Rate of remission

At 52 weeks: ≈56% remission in combination therapy group  
versus 18% in the prednisone alone

More side-effects in the prednisone alone group



# Question 7



This 26-year old patient presented with a facial rash and diffuse arthralgias. She is not on any drug treatment. Which one of the following tests would confirm the diagnosis?

- A. ESR
- B. CRP
- C. Anti-ds DNA ab
- D. Anti-microsomal antibody measurement
- E. Anti-RNP ab measurement

## C. Anti-ds DNA ab

### DDx

- SLE
- Pellagra
- Bloom syndrome (BLM)

**Bloom = autosomal recessive** characterized by short stature and a butterfly facial rash that develops shortly after first exposure to sun. Other clinical features include a high-pitched voice; distinct facial features, such as a long, narrow face, pigmentation (hypo/hyper), moderate immune deficiency, sub-fertility in females.

Pellagra most commonly caused by a chronic lack of niacin (vitamin B3). Characterized by 4 D's: diarrhea, dermatitis, dementia, and death



# Anti-ds DNA Antibodies

- Group of anti-nuclear Abs that target double stranded DNA
- Low sensitivity for SLE, high specificity (absence of the antibodies does not rule out the disease)
- Anti-dsDNA antibodies are highly associated with lupus nephritis
- Some patients with high titers of anti-dsDNA antibodies do not develop renal disease -most likely because anti-dsDNA are a heterogeneous population and some non-pathogenic.
- Patients with rheumatoid arthritis can develop anti-dsDNA antibodies, however they are usually treatment related. Anti-TNF $\alpha$  biological therapies, such as adalimumab, infliximab, etanercept.
- Infection with viral pathogens can induce anti-dsDNA antibodies transiently e.g., HIV, human parvovirus B19, BK virus.

# Question 8



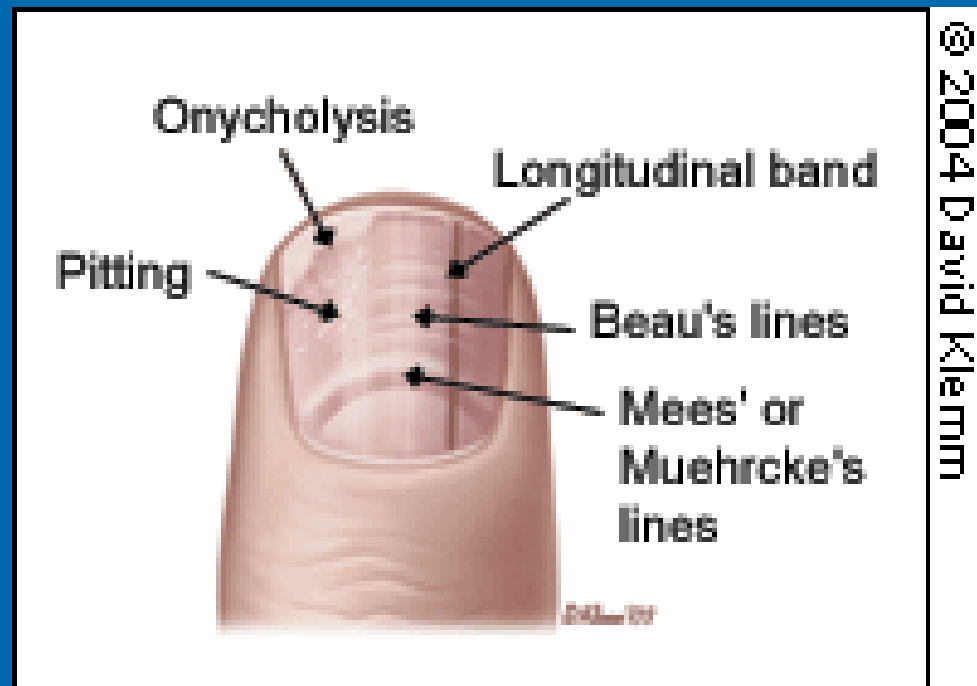
A 46-year-old woman with metastatic sarcoma who had been treated with five cycles of doxorubicin, ifosfamide, and mesna chemotherapy presented with two symmetrical, horizontal white lines on all of her fingernails but not on her toenails. **Which one of the following is the most likely diagnosis?**

SOURCE: Morrison-Bryant M, Gradon JD.  
N Engl J Med. 2007 Aug 30;357(9):917

- A. Chronic renal failure**
- B. Iron deficiency**
- C. Graves' disease**
- D. Chemotherapy treatment**
- E. Psoriasis**

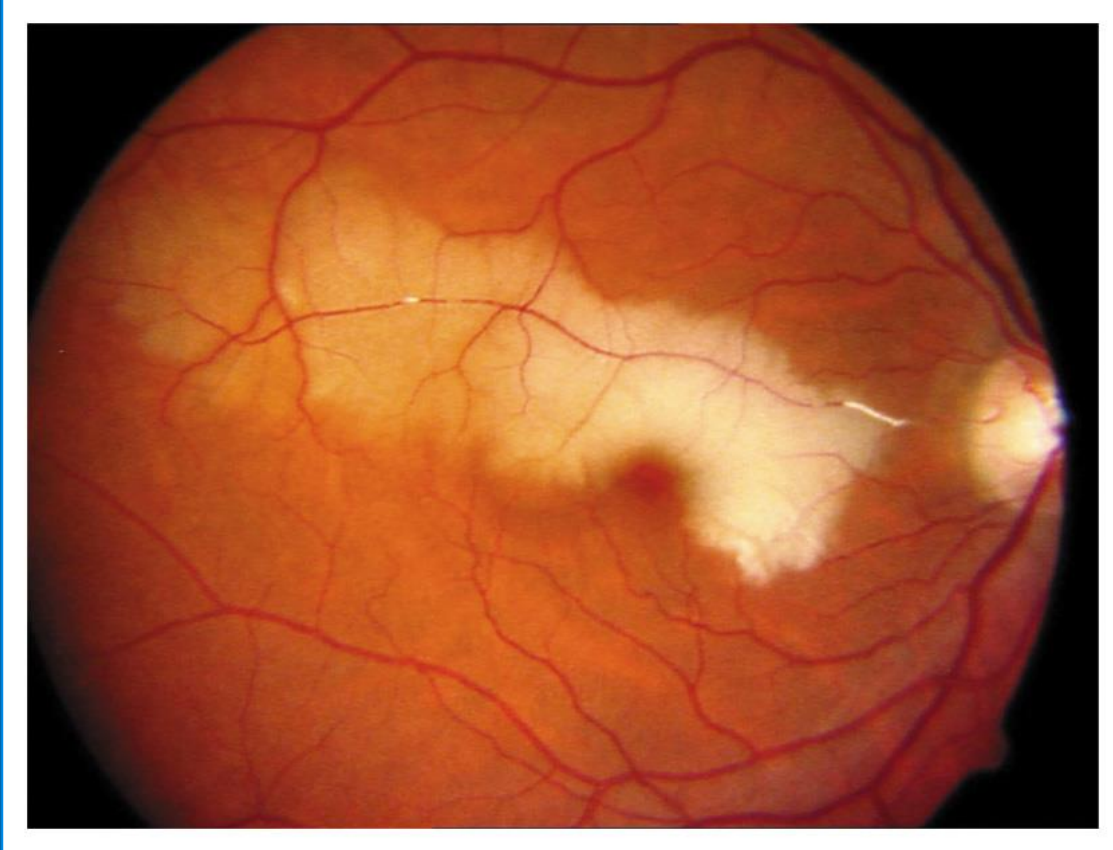
## D. Chemotherapy treatment

- A diagnosis of Muehrcke's lines related to chemotherapy treatment.
- Muehrcke's lines are the two smooth white bands that run parallel to the lunula across the width of the nail.
- The lines are nonpalpable and, unlike Beau's lines, do not indent the nail itself.
- Muehrcke's lines are a nonspecific finding that may be associated with periods of metabolic stress, which transiently impairs protein synthesis. Muehrcke's lines are also caused by infections and trauma.





# Question 9

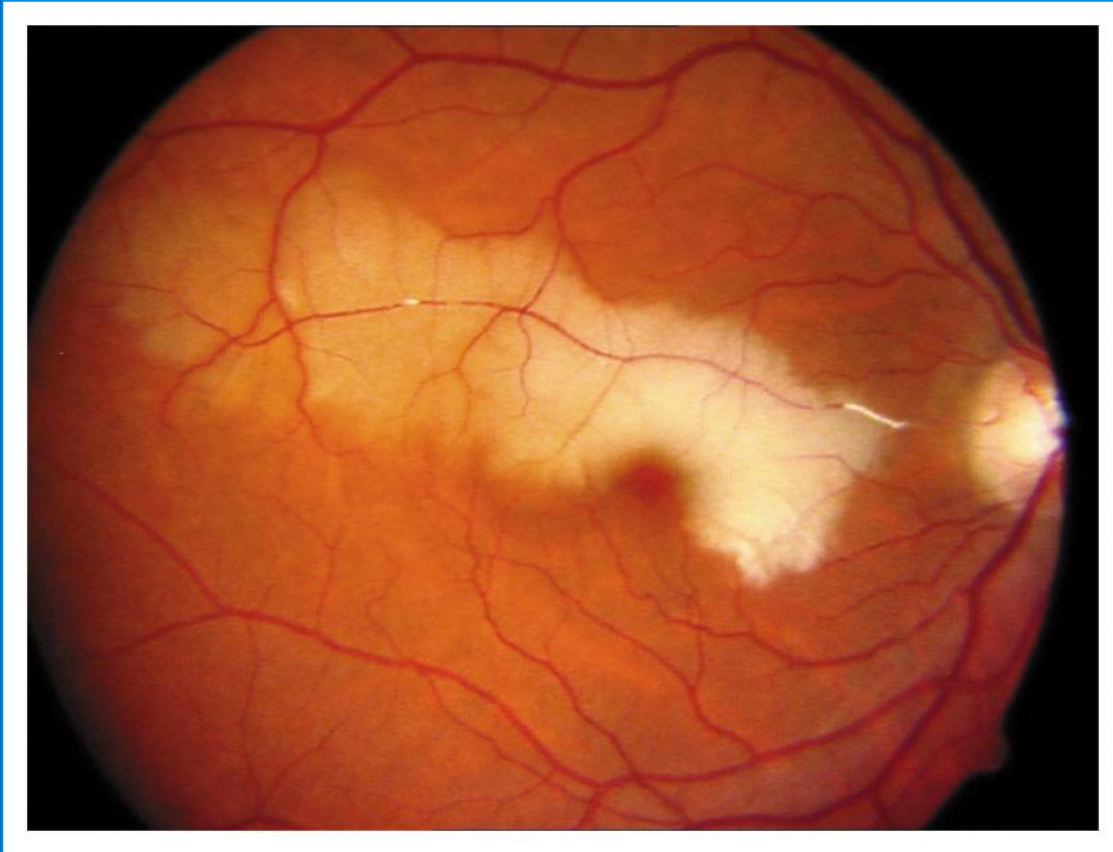


A 56-year-old woman with hypertension presented with a sudden onset painless impaired vision in the right eye. On the previous day, she had undergone cardiac catheterization for evaluation of hypertensive emergency. On physical examination, the visual acuity in the right eye was 20/100 with significant visual-field defect in the inferior temporal quadrant.

SOURCE: Meyer CH, Holz FG. Images in clinical medicine. Blurred vision after cardiac catheterization. N Engl J Med. 2009 Dec 10;361(24):2366.



# Question 9

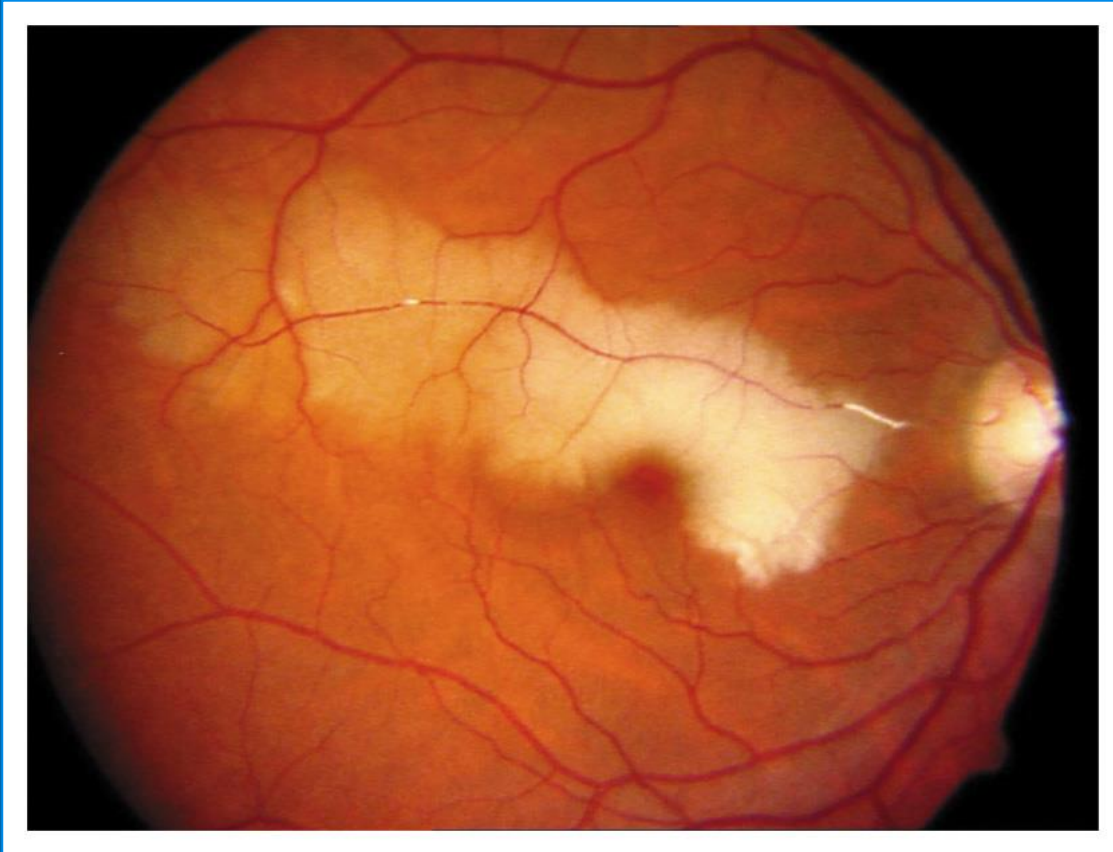


**Choose the most likely cause:**

- A.) Diabetic retinopathy
- B.) Hypertensive retinopathy
- C.) Cholesterol embolism
- D.) Toxoplasmosis
- E.) Retinal Detachment

SOURCE: Meyer CH, Holz FG. Images in clinical medicine. Blurred vision after cardiac catheterization. N Engl J Med. 2009 Dec 10;361(24):2366.

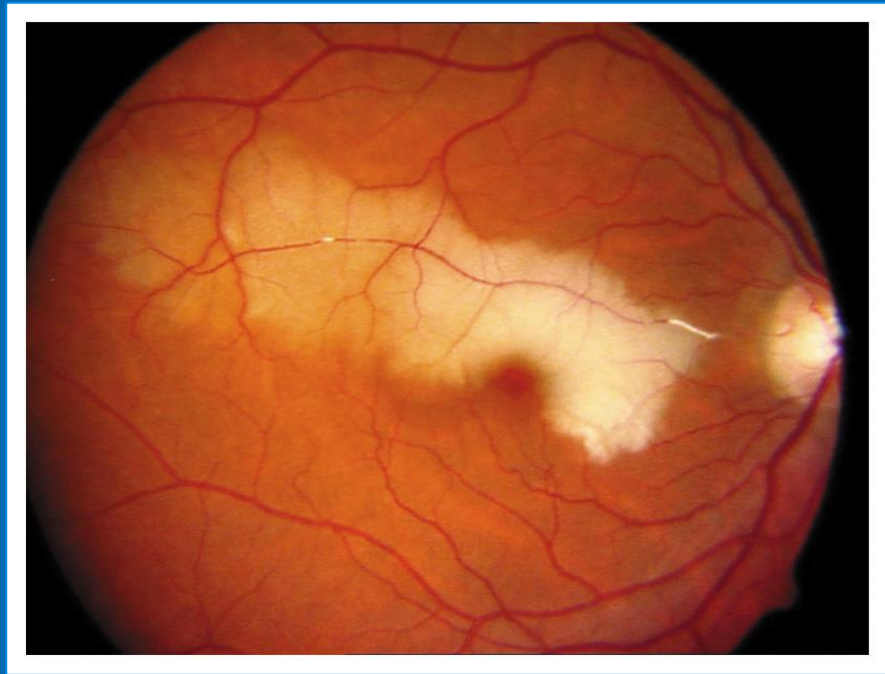
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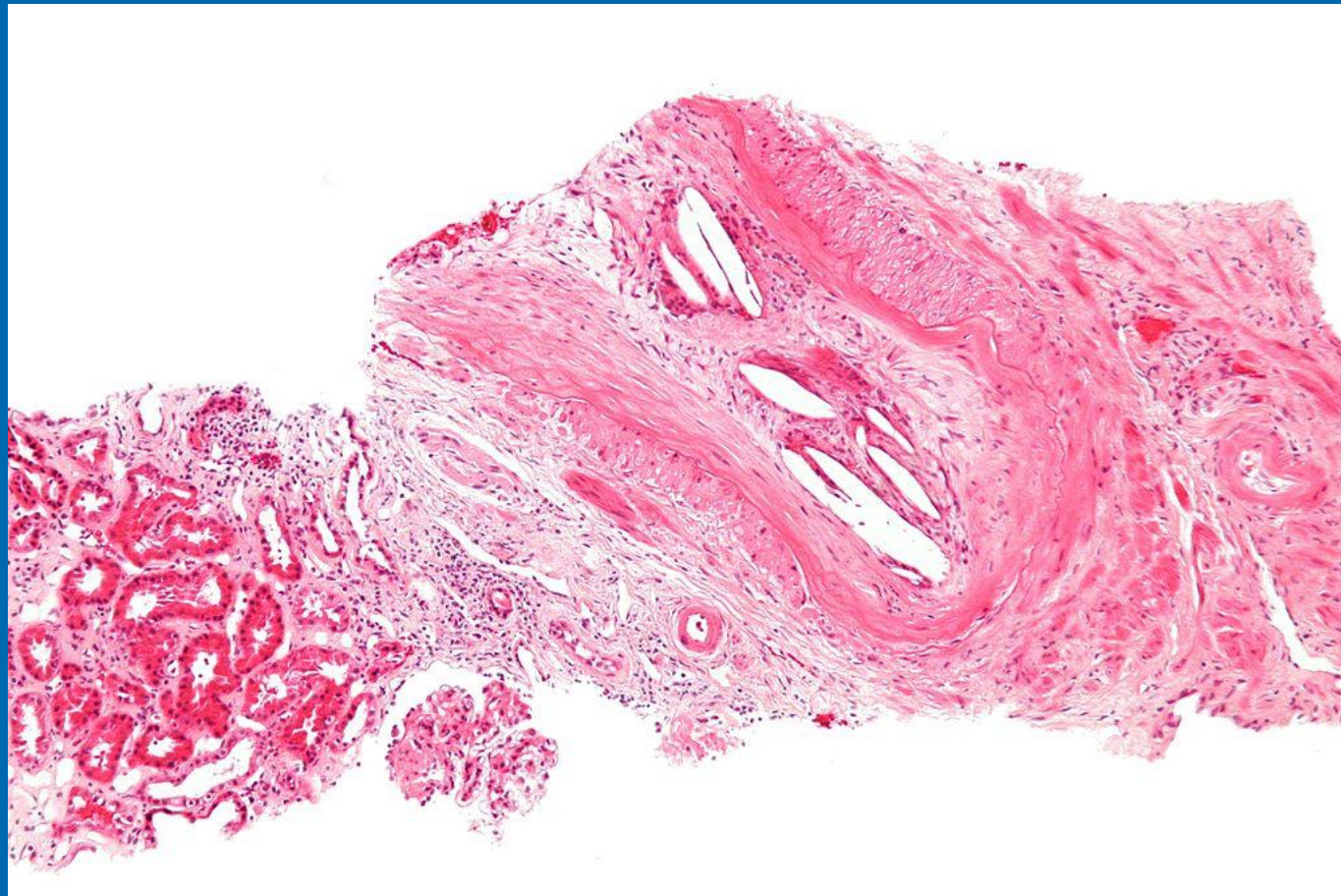
## C. Cholesterol embolism

The image demonstrates cholesterol emboli with surrounding white lucency representing retinal edema. Fluorescein angiography confirmed occlusion of the cilioretinal artery with nonperfusion of the tissue bed in the hypofluorescent areas. Cholesterol emboli are crystals that are released in the arterial bloodstream from ulcerated or disrupted atherosclerotic plaques and can be an initial sign of vascular disease.



- Cholesterol embolism syndrome should be suspected in a patient who develops worsening renal function, hypertension, distal ischemia or acute multisystem dysfunction after an invasive procedure.
- Middle-aged to elderly
- Men > women
- Step wise rise in Scr
- Bland urine classic

Cholesterol embolus in a medium sized artery of the kidney



SOURCE: [https://en.wikipedia.org/wiki/Cholesterol\\_embolism](https://en.wikipedia.org/wiki/Cholesterol_embolism)