#### ACP Rheumatology Pearls

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## Disclosures

□ I have no personal or professional disclosures

27 yo woman with a history of systemic lupus complicated by lupus nephritis contacts the clinic to let you know that she has unexpectedly become pregnant.

Her lupus was diagnosed two years ago when she presented with rashes, arthritis, pericarditis, and renal insufficiency associated with proteinuria.

Her serologies are notable for a high titer ANA, dsDNA, SSA, and SSB. At diagnosis she has very low levels of C3 and C4.

- A kidney biopsy demonstrated Class III (proliferative) Lupus Nephritis
- At diagnosis she was started on high dose steroids, hydroxychloroquine, and mycophenolate. Her steroids had been tapered to a dose of 5 mg daily when she contacts the clinic.

## Case #1: Question A

- Which of the following medications is <u>NOT</u> safe for use in lupus patients who are pregnant?
  - 1. Prednisone
  - 2. Mycophenolate
  - 3. Azathioprine
  - 4. Tacrolimus
  - 5. Hydroxychloroquine

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 She stops Mycophenolate and starts Azathioprine in its place. She continues her hydroxychloroquine

 2 months later she contacts you to let you know that she is short of breath and has new leg swelling.
 Labs done by her primary OB are notable for new renal insufficiency.

## Case #1: Question B

- Which of the following is the best parameter to monitor for lupus nephritis activity in a patient with well established lupus and a history of nephritis
  - 1. dsDNA
  - 2. C3
  - 3. Creatinine
  - 4. Urine Pr:Cr
  - 5. ESR

## Case #1: Question B

- Which of the following is the best parameter to monitor for lupus nephritis activity in a patient with well established lupus and a history of nephritis
  - 1. dsDNA
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  - 3. Creatinine
  - 4. Urine Pr:Cr
  - 5. ESR

 She has 3 gm of proteinuria and her lupus parameters are all suggestive of active disease.
 She resumes a moderate dose of prednisone and tacrolimus is added to her regimen

She develops refractory hypertension with worsening proteinuria. With concern for progressive renal disease her steroid dose is increased to 60 mg daily.

 Labs are notable worsening transaminases and falling platelets.

 She is diagnosed with HELLP and opts to terminate her pregnancy at 20 weeks

She has rapid normalization of her blood pressure, proteinuria, cell counts, and transaminases within two weeks.

- 71 yo female with acute onset of severe right sided occipital headache and right upper extremity pain associated with numbness, tingling, and pain following an iron infusion
- An ultrasound of the right upper extremity revealed a right brachial artery thrombus.
- During a CT scan of the head, she developed the sudden onset of severe right sided jaw pain and vision loss in the right eye.

- An urgent ophthalmology evaluation is notable for vitreous hemorrhage.
- □ ESR 80
- She is started on prednisone 60 mg daily for possible GCA with dramatic improvement in her symptoms of claudication and headache.
- Her vision does not improve

# Case #2 Question A

- You have 3 weeks after starting steroids to get a temporal artery biopsy
  - 1. True
  - 2. False

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## Diagnostic yield of TA biopsy

Patients Receiving Corticosteroids before Temporal Artery Biopsy	Patients with Positive Biopsy Results	
	n (%)	
Entire cohort $(n = 535)$	175 (33)	
1-7 days of treatment (n = 107)†	46 (43)‡	
8-14 days of treatment (n = 10)†	3 (30)	
>14 days of treatment (n = 32)†	9 (28)	

<sup>†</sup> Refers to patients who received more than 15 mg/d of prednisone or equivalent dose of a different corticosteroid for the indicated number of days before temporal artery biopsy.

 A temporal artery biopsy was obtained that showed a 3.5 mm segment of vessel wall inflammation without classic findings of giant cells.

A subsequent ophthalmologic evaluation raised concern for possible endophthalmitis for which she received intravitreal and systemic antibiotics.

 Vitreal cultures were collected but were not received in the microbiology lab

She continued prednisone 60 mg daily

 Thrombectomy was performed in the right upper extremity.

With broad spectrum antibiotics and antifungals along with prednisone, the patient reports gradual improvement in her symptoms.

Her prednisone dose is reduced to 20 mg

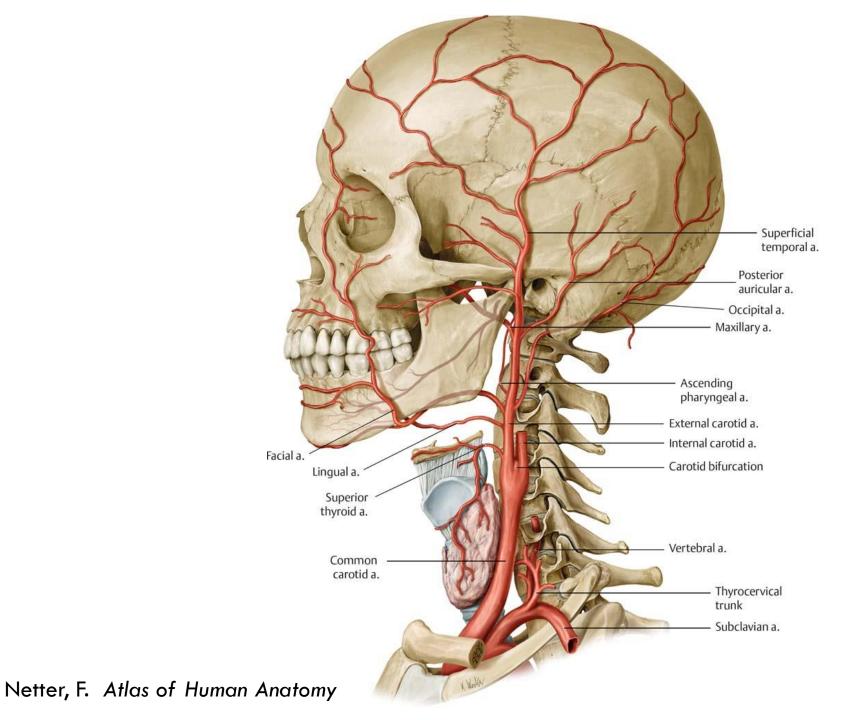
- Once prednisone is tapered below 10 mg daily, she experiences return of right sided jaw pain and severe occipital headaches.
- Prednisone is restarted with improvement in her symptoms
- Ophthalmology re-evaluates patient and feels that that her vision loss was due to right optic nerve damage.

## Case #3 Question B

- Other than the temporal arteries, what other branches of the external carotid are targets for potential biopsy
  - 1. Lingual
  - 2. Posterior Auricular
  - 3. Occipital
  - 4. Facial
  - 5. Maxillary

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## Case #2 Question B

- If you receive a pathology report on a temporal artery biopsy that is "negative" or "atypical" for changes consistent with GCA, what it is next best step?
  - 1. Ask Pathologist to take another look
  - 2. Biopsy the other side
  - 3. Biopsy another vessel
  - 4. Perform a vascular ultrasound of other branches of the external carotid
  - 5. Perform an MRA of the Great Vessels

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36 yo man with chronic tophaceous gout that began in his late teenage years. He has a heavy burden of tophi. His hyperuricemia is managed initially with Allopurinol and later transitioned to Febuxostat 80 mg. His current uric acid level is 8. His estimated GFR is 35 ml/min

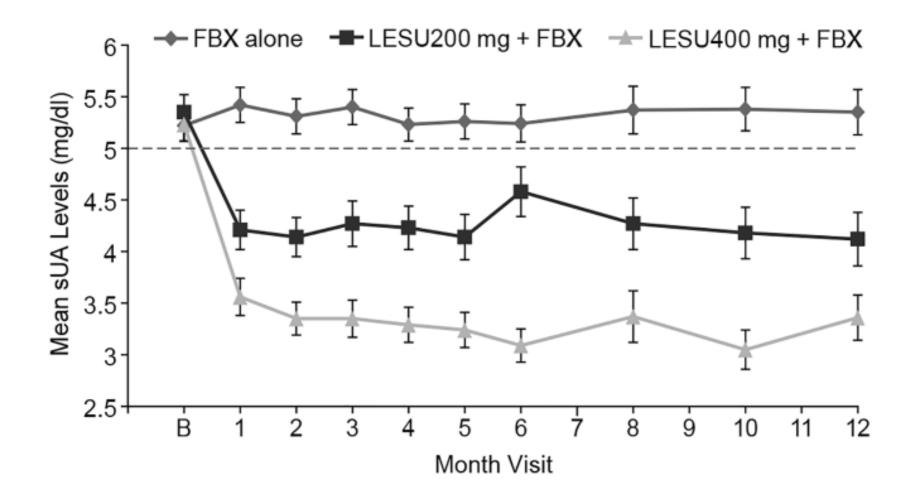


### Case #3: Question

- In a patient with Stage III CKD and tophaceous gout currently managed with febuxostat 80 mg daily but with a persistently elevated uric acid level of 8, what is the next most appropriate step:
  - 1. Increase dose of Febuxostat
  - 2. Add Probenecid
  - 3. Add Pegloticase
  - 4. Add Colchicine
  - 5. Add Lensinurad

### Case #3: Question

- In a patient with Stage III CKD and tophaceous gout currently managed with febuxostat (Uloric) 80 mg daily but with a persistently elevated uric acid level of 8, what is the next most appropriate step:
  - 1. Increase dose of Febuxostat (Ulolric)
  - 2. Add Probenecid
  - 3. Add Pegloticase (Krystexxa)
  - 4. Add Colchicine
  - 5. Add Lensinurad (Zurampic)



CRYSTAL Trial Arthritis Rheumatol. 2017 Aug;69:1903-1913

## Questions?

