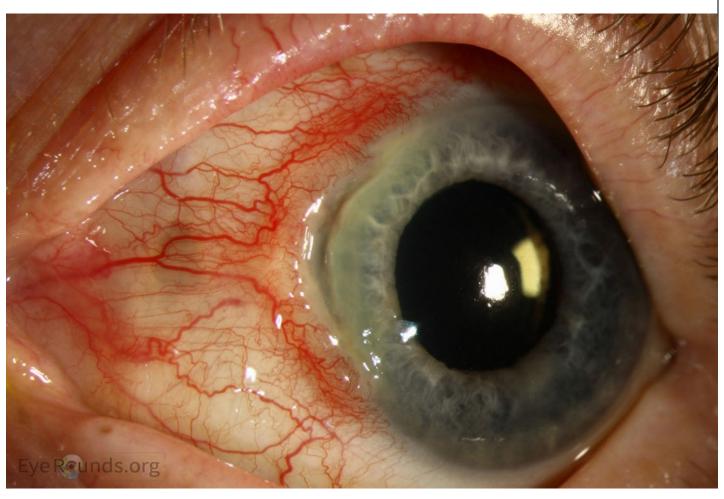


Autoimmune PUK is usually
 and circumferential extent
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Autoimmune PUK is usually unilateral and sectoral





Autoimmune PUK



- Autoimmune PUK is usually unilateral and sectoral
- It often heralds improvement vs worsening of systemic disease



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With what general category of autoimmune dz is PUK associated?

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granulomatosis with polyangiitis (GwP)

If you're having trouble remembering that granulomatosis with polyangiitis (GwP) is the entity formerly known as Wegener's...



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Sinusitis. Don't diagnose a pt with GwP without it!*



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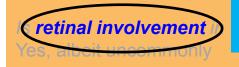
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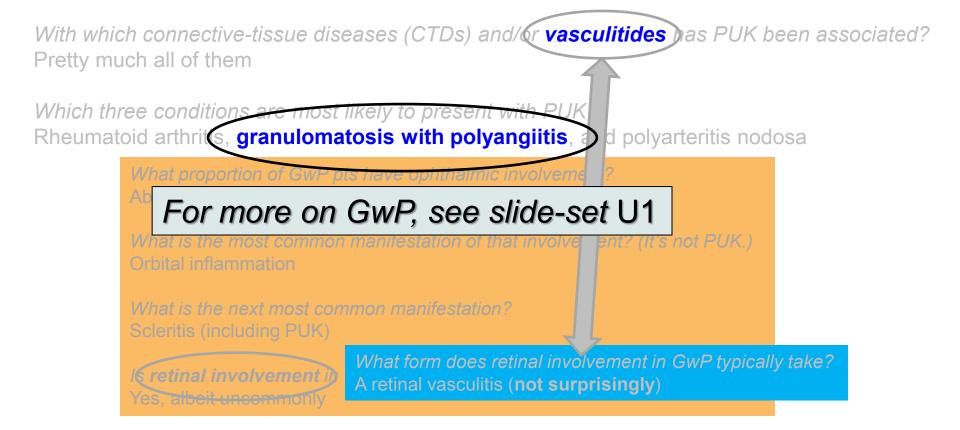
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PAN is strongly associated with seropositivity for what virus?

Hepatitis

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What proportion of PAN pts develop ophthalmic involvement? About 20%



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- Autoimmune Connective-tissue dz, especially vasculitides
- It often heralds exacerbation of systemic disease

With which connective-tissue diseases (CTDs) and/or vasculitides has PUK been associated? Pretty much all of them

Which three conditions are most likely to present with PUK?

Rheumatoid arthritis, granulomatosis with polyangiitis, and polyarteritis nodosa

If a PUK pt does not carry a CTD/autoimmune diagnosis, what should the ophthalmologist do?



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Autoimmune

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If the workup is negative, what non-autoimmune diagnosis should you consider?



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If the PUK is associated with copious mucopurulent discharge, what infectious etiology should you consider?



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If the PUK is associated with copious mucopurulent discharge, what infectious etiology should you consider? Gonococcal disease



- Autoimmune PUK is usually unilateral and sectoral
- It often heralds exacerbation of systemic disease
- The treatment goal is to stop K melting through 3 maneuvers:
 - 1) Improve
 - 2) Promote via 4 treatment maneuvers

3) Suppress two words



- Autoimmune PUK is usually unilateral and sectoral
- It often heralds exacerbation of systemic disease
- The treatment goal is to stop K melting through 3 maneuvers:
 - 1) Improve wetting
 - 2) Promote re-epithelialization via lubes, BCL, patching, glue

(bandage contact lens)

3) Suppress systemic inflammation



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 - 2) Promote re-epithelialization

Of these three maneuvers, which is paramount?

3) Suppress systemic inflammation



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 - 2) Promote re-epithelialization via

Of these three maneuvers, which is paramount? Controlling the underlying disease process--without this, the other maneuvers are akin to rearranging the deck chairs on the *Titanic*

3 Suppress systemic inflammation



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```
How should one improve wetting?
```



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How should one improve wetting?
With frequent dosing of preservative-free artificial tears (PF ATs)
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In addition to improving wetting, what other benefit derives from frequent PF AT use?

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1) Improve wetting

How should one improve wetting?
With frequent dosing of preservative-free artificial tears (PF ATs)

In addition to improving wetting, what other benefit derives from frequent PF AT use? They will remove inflammatory cytokines from the ocular surface



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 - 2) Promote re-epithelialization via lubes, BCL, patching glue

What specific sort of glue is being referred to here?



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How does glue assist in PUK healing?

1)

2)



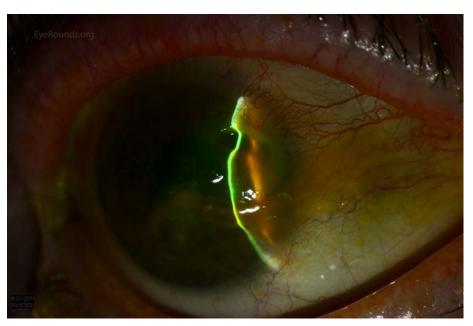
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How does glue assist in PUK healing?

- 1) It provides tectonic stability, thereby reducing the risk of perforation
- 2) It acts as a barrier preventing PMNs from reaching (and destroying) corneal stroma





Just prior to perfing



Same eye s/p gluing (and on IMT)



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Cyanoacrylate adhesive

Use of cyanoacrylate adhesive mandates that what other therapeutic maneuver be applied as well?



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Use of cyanoacrylate adhesive mandates that what other therapeutic maneuver be applied as well? A BCL must be placed over the glued cornea



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An antibiotic drop should be used to prophylax against the possibility of a BCL-induced bacterial superinfection



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What bacteria species must you be certain is adequately covered by the antibiotic?



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An antibiotic drop should be used to prophylax against the possibility of a BCL-induced **bacterial superinfection**

What bacteria species must you be certain is adequately covered by the antibiotic? Pseudomonas



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 - 1) Improve wetting
 - 2) Promote re-epithelialization via lubes, BCL, patching, glue
 - You should also consider stopping common ocular drug which can delay reepithelialization. As a general rule: If the cornea is significantly thinned, avoid same drug.
 - 3) Suppress systemic inflammation



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 - 2) Promote re-epithelialization via lubes, BCL, patching, glue
 - You should also consider stopping topical steroids, which can delay reepithelialization. As a general rule: If the cornea is significantly thinned, avoid topical steroids.
 - 3) Suppress systemic inflammation



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 - 3) Suppress systemic inflammation
 - 4) Conj flap over the peripheral defect?

What about using a conj flap to cover the peripheral defect?



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 - You should also consider stopping topical steroids, which can delay re-

In what clinical scenario might a conj flap over a PUK defect be an appropriate treatment maneuver?

- 3) Suppress systemic inflammation
- 4) Conj flap over the peripheral defect? YES!



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In what clinical scenario might a conj flap over a PUK defect be an appropriate treatment maneuver? In **infectious** PUK, especially when the organism is type of bug

- 3) Suppress systemic inflammation
- 4) Conj flap over the peripheral defect? YES!



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 - You should also consider stopping topical steroids, which can delay re-

In what clinical scenario might a conj flap over a PUK defect be an appropriate treatment maneuver? In **infectious** PUK, especially when the organism is **fungal**

- 3) Suppress systemic inflammation
- 4) Conj flap over the peripheral defect? YES!



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 - 3) Suppress systemic inflammation

4) Conj surgery:

What about using a conj flap to cover the peripheral defect?
Conj flaps are contraindicated in autoimmune PUK because they bring the conj vasculature (and thus all those nasty blood-borne inflammatory mediators) even closer to the melt

What conj surgery is very helpful in autoimmune PUK?



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 - 3) Suppress systemic inflammation
 - 4) Conj surgery: Sectoral conj resection

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Conj flaps are contraindicated in autoimmune PUK because they bring the conj vasculature (and thus all those nasty blood-borne inflammatory mediators) even closer to the melt

What conj surgery is very helpful in autoimmune PUK?
Sectoral conj resection (ie, cutting the conj away from the PUK zone) can be very effective

Q

For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)

Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



Saddle-nose deformity (2):



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Saddle-nose deformity (2): RP; GwP



Saddle-nose deformity





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Saddle-nose deformity (2): RP; GwP

If a pt with a saddle nose had interstitial keratitis rather than PUK, what diagnosis should you consider?



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Churg-Strauss (CS)



Saddle-nose deformity (2): RP; GwP

If a pt with a saddle nose had interstitial keratitis rather than PUK, what diagnosis should you consider?

Congenital syphilis

Q

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Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia:



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Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



- Saddle-nose deformity (2): RP; GwP
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Q

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- Saddle-nose deformity (2): RP; GwP
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- Deformed auricular pinnae:



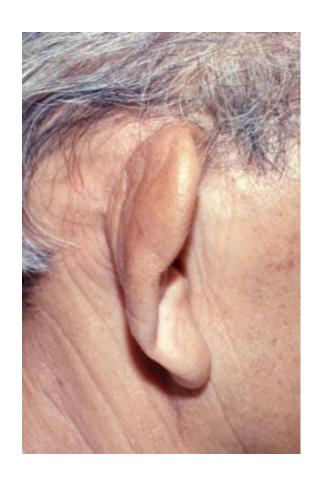
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- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP







Auricular damage in RP

Q

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Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2):



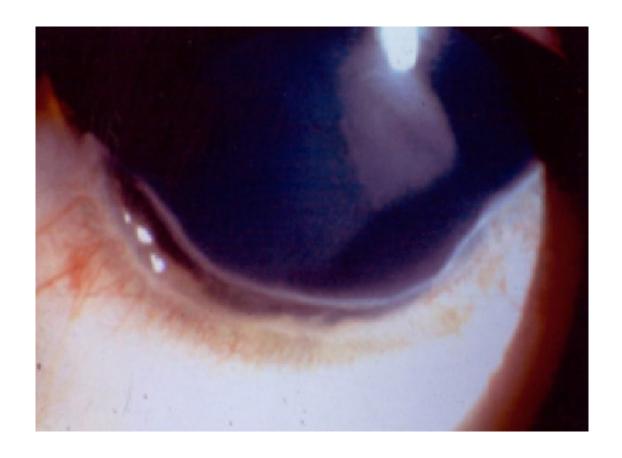
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- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN





Mooren's ulcer: Note the overhanging edge



Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)
Mooren's ulcer (MU)

Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN

What is the classic description regarding the pattern of progression for PUK in both Mooren's and PAN?

Starts...

Then extends...

And finally progresses...



For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN)

Relapsing polycho

Mooren's ulcer (MU)

Relapsing polychondritis (RP) Rheumatoid arthritis (RA) Granulomatosis with polyangiitis (GwP) **Churg-Strauss (CS)**



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN

What is the classic description regarding the pattern of progression for PUK in both Mooren's and PAN?

Starts...sectoral

Then extends...

And finally progresses...



For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)
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Mooren's ulcer (MU)

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- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN

What is the classic description regarding the pattern of progression for PUK in both Mooren's and PAN?

Starts...sectoral

Then extends...circumferentially

And finally progresses...



Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)
Mooren's ulcer (MU)

Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN

What is the classic description regarding the pattern of progression for PUK in both Mooren's and PAN?

Starts...sectoral

Then extends...circumferentially

And finally progresses...centrally

Q

For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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Churg-Strauss (CS)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved:



Polyarteritis nodosa (PAN)
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Mooren's ulcer (MU)

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Churg-Strauss (CS)



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- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved: MU

Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)

Relapsing polychondritis (RP)
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Churg-Strauss (CS)

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- Saddle-nose deformity (2): RP; GwP
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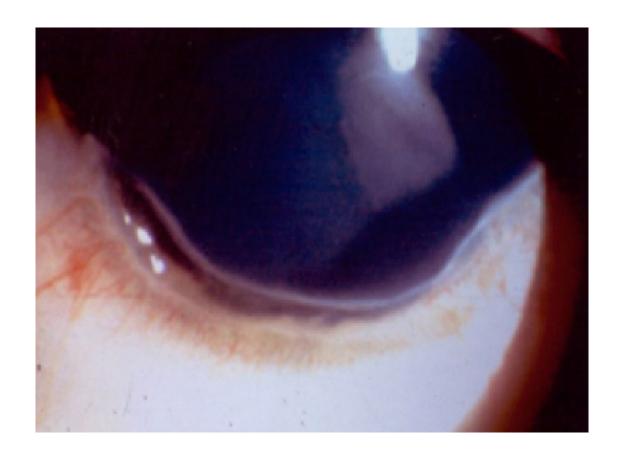
Mooren's ulcer (MU)

- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera <u>never</u> involved: MU

Take note! This is a key factor differentiating between Mooren's and other forms of PUK.

Concerning PUK





Mooren's ulcer. Note the adjacent sclera is totally quiet

Q

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- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved: MU
- ANCA positive (2):



Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU) Relapsing polychondritis (RP)
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- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved: MU
- ANCA positive (2): GWP; CS (Hey, what about PAN??!! Un momento, por favor)



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Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



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What does ANCA stand for?



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What does ANCA stand for?
Antineutrophil cytoplasmic antibodies



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What does ANCA stand for?
Antineutrophil cytoplasmic antibodies

What are they?



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What does ANCA stand for?
Antineutrophil cytoplasmic antibodies

What are they?

Autoantibodies against antigens found within the







Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)
Mooren's ulcer (MU)

Relapsing polychondritis (RP)
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Churg-Strauss



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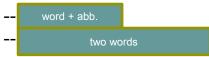
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For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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What's the difference between a 'small-sized' artery and a 'smaller' artery? Rule of thumb: Classic PAN only affects arteries large enough to be named, whereas microscopic angiitis only affects vessels smaller than that.

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Which hepatitis virus is definitely associated with PAN?



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Which hepatitis virus is definitely associated with PAN? Hepatitis B



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Which hepatitis virus is definitely associated with PAN?

Hepatitis B

What percent of PAN pts test positive for are Hep B surface Ag?



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Which hepatitis virus is definitely associated with PAN? Hepatitis B

Which form is probably associated, but the evidence is not as strong as for B?



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Which hepatitis virus is definitely associated with PAN? Hepatitis B

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- Associated with hepatitis seropositivity: PAN
- Associated with helminthic seropositivity:



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- Associated with hepatitis seropositivity: PAN
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Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved: MU
- ANCA positive (2): GwP; CS; PAN
- Is a diagnosis of exclusion: MU
- Chest X-ray likely abnormal (3): GwP; CS; RP
- Associated with hepatitis seropositivity: PAN
- Associated with helminthic seropositivity: MU
- Renal function may be impaired (4):



Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



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- Chronic, tx-resistant sinusitis common:



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- Extremely painful:



Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)
Mooren's ulcer (MU)

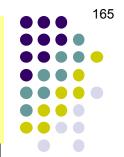
Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



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- Renal function may be impaired (4): GwP; PAN; CS; RP
- Chronic, tx-resistant sinusitis common: GwP
- Extremely painful: MU

All forms of inflammatory PUK are painful, but Mooren's is *exceptionally* so!

Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)



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- Extremely painful: MU
- Anti-CCP Ab positive:



Polyarteritis nodosa (PAN)
Rheumatoid arthritis (RA)
Mooren's ulcer (MU)



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Q

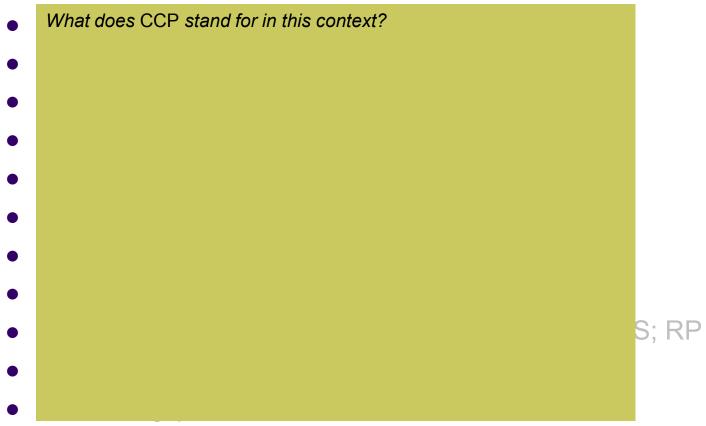
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)

Relapsing polychondritis (RP)
Granulomatosis with polyangiitis (GwP)
Churg-Strauss (CS)



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Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)

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```
What does CCP stand for in this context?
Cyclic citrullinated peptide
                                                                      S: RP
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Polyarteritis nodosa (PAN) Mooren's ulcer (MU)

Relapsing polychondritis (RP) Rheumatoid arthritis (RA) Granulomatosis with polyangiitis (GwP) **Churg-Strauss (CS)**



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S: RP



Polyarteritis nodosa (PAN) Mooren's ulcer (MU)

Relapsing polychondritis (RP) Rheumatoid arthritis (RA) Granulomatosis with polyangiitis (GwP) **Churg-Strauss (CS)**



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S; RP

Q

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S: RP



- Saddle-nose deformity (2): RP; GwP
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 Cyclic citrullinated peptide
- What does it mean to say a peptide has been citrullinated?
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•

Anti-CCP Ab positive: RA

S; RP

Q

For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN)
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 - OK, so what does this have to do with RA?

S: RP



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 - OK, so what does this have to do with RA?
- The citrulline moieties alter the conformation of the proteins within
- which they occur, rendering the proteins novel to the immune system.
- The immune system of an RA pt will attack them as foreign antigens—
- hence the presence of anti-CCP antibodies in their serum.

S; RF



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- When evaluating a pt for RA, I usually check for the presence of serum blood test

S; RP



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S; RP



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- hence the presence of anti-CCP antibodies in their serum.
- When evaluating a pt for RA, I usually check for the presence of serum rheumatoid factor (RF). Is anti-CCP a better test?

S; RP



Polyarteritis nodosa (PAN) Rheumatoid arthritis (RA) Mooren's ulcer (MU)

Relapsing polychondritis (RP) **Granulomatosis with polyangiitis (GwP) Churg-Strauss (CS)**



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- hence the presence of anti-CCP antibodies in their serum.
- When evaluating a pt for RA, I usually check for the presence of serum rheumatoid factor (RF) . Is anti-CCP a better test?
- Yes, because it has the same

sensitivity v specificity

but higher sensitivity v specificity

(98%)

S: RP

for RA c/w the RF test



Polyarteritis nodosa (PAN)
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- When evaluating a pt for RA, I usually check for the presence of serum rheumatoid factor (RF) . Is anti-CCP a better test?
- Yes, because it has the same sensitivity but higher specificity (98%)
- for RA c/w the RF test
- Anti-CCP Ab positive: RA

S; RP



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

(IBD = Inflammatory bowel disease)



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

Why is Mooren's the oddball in this group?



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

Why is Mooren's the oddball in this group?
PUK in the others is due to a systemic condition, whereas Mooren's is, by definition, ocular only



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

- And in this group?
 - Mooren's, Terrien's marginal, Sarcoid, SLE



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Why is Terrien's the oddball in this group?





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Why is Terrien's the oddball in this group?

Two reasons:

--PUK in the others is an process; Terrien's is



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- And in this group?
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Why is Terrien's the oddball in this group? Two reasons:

--PUK in the others is an inflammatory process; Terrien's is noninflammatory

__





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Why is Terrien's the oddball in this group? Two reasons:

- --PUK in the others is an inflammatory process; Terrien's is noninflammatory
- --As implied by the word 'ulcerative' in the name, the corneal epithelium is in PUK. In contrast, the epithelium is in Terrien's.



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

- And in this group?
 - Mooren's, Terrien's marginal, Sarcoid, SLE

Why is Terrien's the oddball in this group? Two reasons:

- --PUK in the others is an inflammatory process; Terrien's is noninflammatory
- --As implied by the word 'ulcerative' in the name, the corneal epithelium is disrupted in PUK. In contrast, the epithelium is **intact** in Terrien's.



- With respect to manifesting PUK, which of the following doesn't belong, and why?
 - RA, Mooren's, Behçet, IBD

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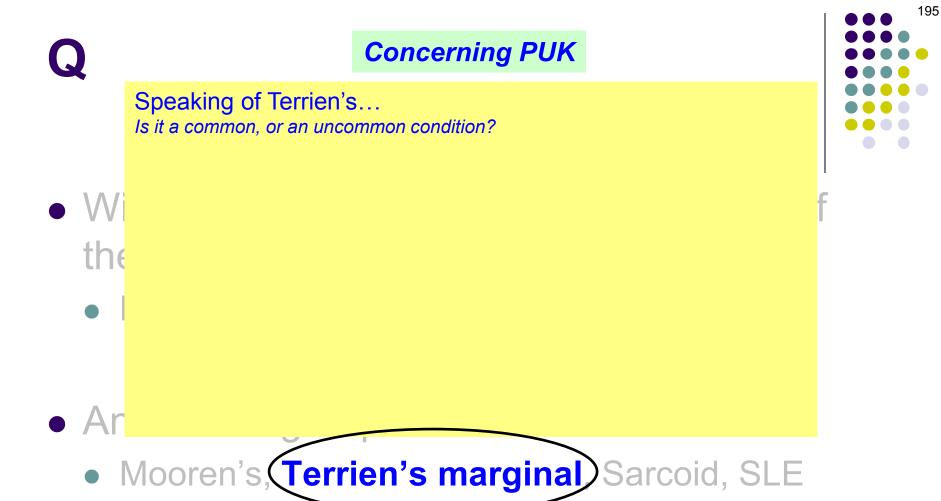
- And in this group?
 - Mooren's, Terrien's marginal, Sarcoid, SLE
 - If the epithelium is intact, what is going on that puts Terrien's on the DDx for PUK?

 Progressive peripheral thinning makes the limbal region in Terrien's resemble PUK
 - --PUK in the others is an inflammatory process; Terrien's is noninflammatory
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Mooren's, Terrien's marginal Sarcoid, SLE

Why is Terrien's the oddball in this group?

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Speaking of Terrien's...

*Is it a common, or an uncommon condition?*Uncommon

Does it have a gender predilection?
Yes, it is more common in

the

Ar

Mooren's, Terrien's marginal Sarcoid, SLE

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A

Concerning PUK

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Q

Concerning PUK

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During what life-stage does Terrien's typically first appear?

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Mooren's, Terrien's marginal Sarcoid, SLE

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Speaking of Terrien's...

Is it a common, or an uncommon condition?
Uncommon

Does it have a gender predilection?
Yes, it is more common in males

During what life-stage does Terrien's typically first appear? Young adulthood (late teens - early 30s)

Ar

Mooren's, Terrien's marginal Sarcoid, SLE

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Speaking of Terrien's...

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the Is it unilateral, or bilateral?

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Is it unilateral, or bilateral?

Bilateral (although involven

Bilateral (although involvement can be strikingly asymmetric)

Ar

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Which sector of the cornea is involved first, and how does it progress from there?

Ar

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Which sector of the cornea is involved first, and how does it progress from there? It starts superonasally, then spreads circumferentially

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Does it affect vision? If so, how?

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Does it have a gender predilection?
Yes, it is more common in males

Is it unilateral, or bilateral?
Bilateral (although involvement can be strikingly asymmetric)

Which sector of the cornea is involved first, and how does it progress from there? It starts superonasally, then spreads circumferentially

Does it affect vision? If so, how? Yes, by inducing high astigmatism

Mooren's, Terrien's marginal Sarcoid, SLE

Why is Terrien's the oddball in this group?

- --PUK in the others is an inflammatory process; Terrien's is noninflammatory
- --As implied by the word 'ulcerative' in the name, the corneal epithelium is disrupted in PUK. In contrast, the epithelium is **intact** in Terrien's.





Speaking of Terrien's...

Is it a common, or an uncommon condition? Uncommon



The key to diagnosing Terrien's is its classic appearance:

--The epithelium is _____, as noted earlier

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The key to diagnosing Terrien's is its classic appearance:

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- --The leading edge of the thinned area is characterized by the presence of...

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- --The leading edge of the thinned area is characterized by the presence of...Lipid

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two words

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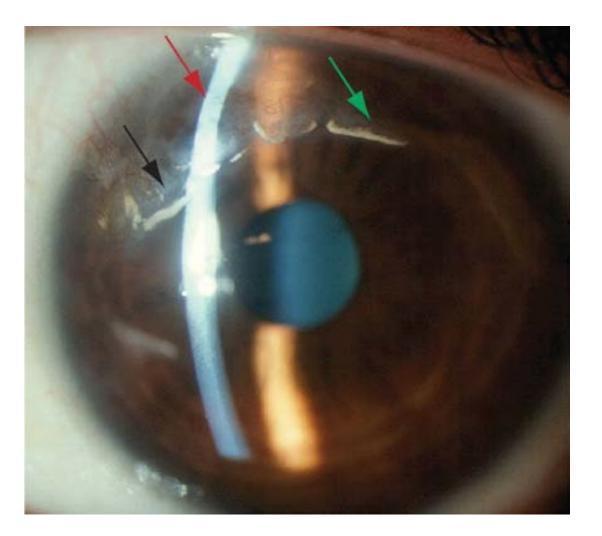
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Terrien marginal degeneration with a fine vascular pannus (black arrow), superior thinning (red arrow), and lipid deposits (green arrow) at the leading edge of the pannus







Terrien marginal degeneration. Note the leading lipids and the trailing pannus

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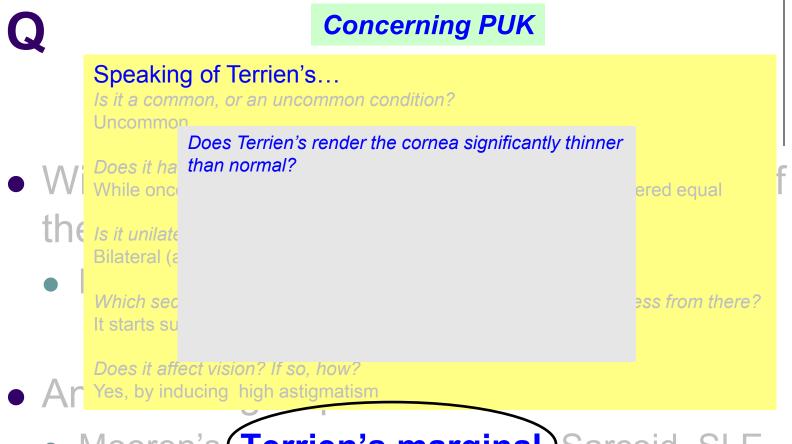
Remember this!!! Consider it your tl;dr for Terrien's

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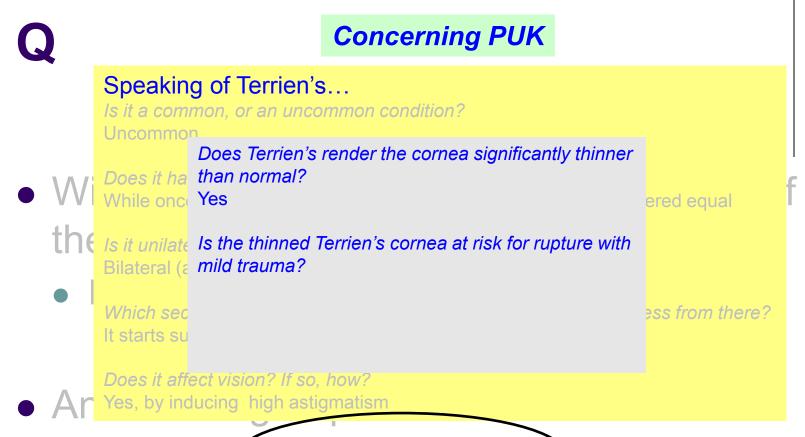


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Speaking of Terrien's...

Is it a common, or an uncommon condition?

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Does Terrien's render the cornea significantly thinner

Does it ha than normal?

While once Yes

Bilateral (a mild trauma?

Is it unilate Is the thinned Terrien's cornea at risk for rupture with

Yes

Which sec It starts su ess from there?

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Yes

Does it affect vision? If so, how?
Yes, by inducing high astigmatism

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but rather a manifestation of the same dz process as Terrien. What is it? Fuchs' superficial marginal keratitis

Yes

Does it affect vision? If so, how?

Yes, by inducing high astigmatism

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Fuchs' superficial marginal keratitis



- All of the following are true concerning Mooren's ulcer except (could be more than one):
 - Cause is unknown
 - One clinical type presents as a unilateral PUK in the elderly
 - The other type presents as bilateral disease in young African women
 - Patients with the 'African' variety often have a history of systemic helminth infection
 - Mooren's responds readily to aggressive local therapy



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Mooren's ulcer is a chronic, progressive PUK. By definition, the cause is unknown. It starts sectorally, progresses circumferentially, then finally centrally. The leading edge is undermined and de-epithelialized. Two clinical varieties are recognized: Unilateral disease in the elderly, and rapidly progressive, severe bilateral disease that strikes young African men. These men usually are seropositive for helminthic disease.

The plethora of treatments stands as gloomy testimony to the relative ineffectiveness of each. Ocular modalities include topical steroids, BCL, *n-acetylcysteine* drops, topical cyclosporine and conjunctival resection. Quite often, systemic immunosuppressives are needed: steroids, MTX, and/or cyclophosphamide.