Autoimmune PUK is usually unilateral and sectoral.
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Concerning PUK

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

- Autoimmune PUK is usually **unilateral** and **sectoral**
- It often heralds **exacerbation** of systemic disease
Concerning PUK

- Autoimmune
- It often heralds exacerbation of systemic disease

With what general category of autoimmune dz is PUK associated?
Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

Concerning PUK:

- With what general category of autoimmune dz is PUK associated? Connective-tissue dz, especially vasculitides
- It often heralds exacerbation of systemic disease
Concerning PUK

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

With what general category of autoimmune dz is PUK associated?

With which connective-tissue diseases (CTDs) and/or vasculitides has PUK been associated?
Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

Concerning PUK:
- With what general category of autoimmune dz is PUK associated?
  - Connective-tissue dz, especially vasculitides
- With which connective-tissue diseases (CTDs) and/or vasculitides has PUK been associated?
  - Pretty much all of them

It often heralds exacerbation of systemic disease.
With what general category of autoimmune dz is PUK associated? Connective-tissue dz, especially vasculitides

Which three conditions are most likely to present with PUK?

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

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Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

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

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Which three conditions are most likely to present with PUK?
Rheumatoid arthritis, Wegener’s granulomatosis, and polyarteritis nodosa

Of these three, which is most likely to be associated with PUK?
Rheumatoid arthritis

The sclera
Concerning PUK

- Autoimmune PUK is usually unilateral and sectoral.
- It often heralds exacerbation of systemic disease.

With what general category of autoimmune dz is PUK associated?
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Pretty much all of them.

Which three conditions are most likely to present with PUK?
**Rheumatoid arthritis**, Wegener’s granulomatosis, and polyarteritis nodosa.

Of these three, which is most likely to be associated with PUK?
RA, by a substantial margin.
Concerning PUK

- Autoimmune
- It often heralds exacerbation of systemic disease

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RA, by a substantial margin

What percentage of PUK pts have RA as their underlying condition?
Concerning PUK

- Autoimmune
- It often heralds exacerbation of systemic disease

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RA, by a substantial margin

What percentage of PUK pts have RA as their underlying condition?
Up to 40
Concerning PUK

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**Of these three, which is most likely to be associated with PUK?**
RA, by a substantial margin

**What percentage of PUK pts have RA as their underlying condition?**
Up to 40

**In addition to the peripheral cornea, what other ocular structure is commonly affected in these pts?**
Concerning PUK

- Autoimmune PUK is usually unilateral and sectoral.
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Is PUK the most common ocular manifestation of RA?
No

In addition to the peripheral cornea, what other ocular structure is commonly affected in these pts?
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What is, then?

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Is PUK the most common ocular manifestation of RA?
No

What is, then?
Keratoconjunctivitis sicca

In addition to the peripheral cornea, what other ocular structure is commonly affected in these pts?
The sclera
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, Wegener’s granulomatosis, and polyarteritis nodosa

The term ‘Wegener’s granulomatosis’ has fallen out of favor.
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, Wegener’s granulomatosis, and polyarteritis nodosa

Concerning PUK

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The term ‘Wegener’s granulomatosis’ has fallen out of favor.
What term is preferred in its place?
Concerning PUK

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granulomatosi with polyangiitis

The term ‘Wegener’s granulomatosis’ has fallen out of favor.
What term is preferred in its place?
‘Granulomatosis with polyangiitis’
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, Wegener’s granulomatosis, and polyarteritis nodosa.

Autoimmune dz

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The term ‘Wegener’s granulomatosis’ has fallen out of favor. What term is preferred in its place? ‘Granulomatosis with polyangiitis’.
With what general category of autoimmune dz is PUK associated? Connective-tissue dz, especially vasculitides

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Which three conditions are most likely to present with PUK? Rheumatoid arthritis, Wegener’s granulomatosis, and polyarteritis nodosa

Granulomatosis with polyangiitis

The term ‘Wegener’s granulomatosis’ has fallen out of favor.

Why did the name ‘Wegener’s granulomatosis’ fall out of favor? Because Dr. Wegener was a Nazi
Concerning PUK

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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…
Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

With what general category of autoimmune disease is PUK associated? Connective-tissue disease, especially vasculitides.

Concerning PUK

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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… Think of the little ‘w’ as standing for ‘Wegener’s.’
Concerning PUK

- Autoimmune
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*What is the classic triad of granulomatosis with polyangiitis (GwP)?*
Concerning PUK

- Autoimmune
- It often heralds exacerbation of systemic disease

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What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
- ?
- ?
- ?
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With what general category of autoimmune dz is PUK associated? Connective-tissue dz, especially vasculitides.

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.

What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of: the upper and lower respiratory tract, the kidneys, small and medium-sized arteries and veins.
Concerning PUK

- Autoimmune
- It often heralds exacerbation of systemic disease

With what general category of autoimmune dz is PUK associated?
Connective-tissue dz, especially vasculitides

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What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract?
--the kidneys?
--small and medium-sized arteries and veins?

What is the classic manifestation of the classic triad? That is, with what specific condition do these pts always present?*

*On the OKAP and Boards, that is
Concerning PUK

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- the upper and lower respiratory tract
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What is the classic manifestation of the classic triad? That is, with what specific condition do these pts always present?*
Sinusitis. Don’t diagnose a pt with GwP without it!*
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What proportion of GwP pts have ophthalmic involvement?
Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

With what general category of autoimmune dz is PUK associated? Connective-tissue dz, especially vasculitides.

What proportion of GwP pts have ophthalmic involvement? About half.

What is the most common manifestation of that involvement? (It’s not PUK.) Orbital inflammation.

What is the next most common manifestation? Scleritis (including PUK).

Is retinal involvement in GwP a thing? Yes, albeit uncommonly.

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.
Concerning PUK

- Autoimmune
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Concerning PUK

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What form does retinal involvement in GwP typically take?
A retinal vasculitis (not surprisingly)

Which three conditions are most likely to present with PUK?
Rheumatoid arthritis, granulomatosis with polyangiitis, and polyarteritis nodosa
Concerning PUK

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Pretty much all of them.

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A retinal vasculitis (**not surprisingly**).
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For more on GwP, see slide-set U1.
Concerning PUK

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**Q**

- With what general category of autoimmune dz is PUK associated?
  - Connective-tissue dz, especially vasculitides

**In a nutshell, what is the pathophysiology of PAN?**

- Subacute episodes of focal necrotizing inflammation of arteries
- Uncommon
- A male between 40 and 60 years old
- No
- Strongly associated with seropositivity for hepatitis B
Concerning PUK

With what general category of autoimmune dz is PUK associated?
Connective-tissue dz, especially vasculitides

Autoimmune dz

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In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

What is the general category of autoimmune dz associated with PUK?
Connective-tissue dz, especially vasculitides

What is the most likely presentation of PUK?
Rheumatoid arthritis, granulomatosis with polyangiitis, and polyarteritis nodosa
With what 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.

Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic disease.

In a nutshell, what is the pathophysiology of PAN? Subacute episodes of focal necrotizing inflammation of arteries.

Is it a common, or uncommon condition? Uncommon.

Who is the typical PAN pt? A male between 40 and 60 years old.

Is there a racial predilection? No.

PAN is strongly associated with seropositivity for what virus? Hepatitis B.
With what general category of autoimmune dz is PUK associated?
Connective-tissue dz, especially vasculitides

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In a nutshell, what is the pathophysiology of PAN?
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Concerning PUK

Polyarteritis nodosa
Concerning PUK

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A male between 40 and 60 years old

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B
Concerning PUK

With what general category of autoimmune dz is PUK associated?
Connective-tissue dz, especially vasculitides

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?
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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

Autoimmune

It often heralds exacerbation of systemic disease

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If a PUK pt does not carry a CTD/autoimmune diagnosis, what should the ophthalmologist do?
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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 workup is negative, what non-autoimmune diagnosis should you consider? Infectious PUK.

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

Concerning PUK:

- Autoimmune
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Autoimmune PUK is usually unilateral and sectoral. It often heralds exacerbation of systemic 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 wetting
2) Promote re-epithelialization via
3) Suppress systemic inflammation

You should also consider stopping topical steroids, which can delay re-epithelialization. As a general rule: If the cornea is significantly thinned, avoid topical steroids.

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(bandage contact lens)
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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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*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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How does glue assist in PUK healing?
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What specific sort of **glue** is being referred to here?

Cyanoacrylate adhesive

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.
Concerning PUK

Just prior to perfing

Same eye s/p gluing (and on IMT)

PUK in RA
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Use of cyanoacrylate adhesive mandates that what other therapeutic maneuver be applied as well?
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Use of a BCL mandates that what other therapeutic maneuver be applied as well? 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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_What about using a conj flap to cover the peripheral defect?_
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*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.
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*In what clinical scenario might a conj flap over a PUK defect be an appropriate treatment maneuver?*

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**Concerning PUK**

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**

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*In infectious PUK, especially when the organism is **fungal***

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  4) Conj surgery:

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*What conj surgery is very helpful in autoimmune PUK?*
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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.
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN)  Relapsing polychondritis (RP)
Rheumatoid arthritis (RA)  Granulomatosis with polyangiitis (GwP)
Mooren’s ulcer (MU)  Churg-Strauss (CS)

- Saddle-nose deformity (2):
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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- Saddle-nose deformity (2): RP; GwP
Concerning PUK

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

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

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If a pt with a saddle nose had interstitial keratitis rather than PUK, what diagnosis should you consider?
Congenital syphilis
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- Saddle-nose deformity (2): RP; GwP
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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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- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
Auricular damage in RP

Concerning PUK
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- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2):
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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

Mooren’s ulcer: Note the overhanging edge
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer):

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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)

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

Polyarteritis nodosa (PAN)  Relapsing polychondritis (RP)
Rheumatoid arthritis (RA)  Granulomatosis with polyangiitis (GwP)
Mooren’s ulcer (MU)         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…
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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

- Polyarteritis nodosa (PAN)
- Relapsing polychondritis (RP)
- Rheumatoid arthritis (RA)
- Granulomatosis with polyangiitis (GwP)
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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
- Sclera never involved:
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

Polyarteritis nodosa (PAN)  Relapsing polychondritis (RP)
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- Sclera never involved: MU
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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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
- Sclera *never* 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
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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- Asthma and eosinophilia: CS
- Deformed auricular pinnae: RP
- Ulcer has overhanging edge (2): MU; PAN
- Sclera never involved: MU
- ANCA positive (2):
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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*(Hey, what about PAN??!! Un momento, por favor)*
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

- 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**

What does ANCA stand for?
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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*What does ANCA stand for?*
Antineutrophil cytoplasmic antibodies
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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*What does ANCA stand for?*
Antineutrophil cytoplasmic antibodies

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

- Saddle-nose deformity (2): RP; GwP
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- Sclera never involved: MU
- **ANCA positive (2): GwP; CS**

*What does ANCA stand for?*
Antineutrophil cytoplasmic antibodies

*What are they?*
Autoantibodies against antigens found within the **cytoplasm** of **neutrophils**
Saddle-nose deformity (2): RP; GwP
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**What does ANCA stand for?**
Antineutrophil cytoplasmic antibodies

**What are they?**
Autoantibodies against antigens found within the cytoplasm of neutrophils

**With which specific ANCA pattern is each condition associated?**
Granulomatosis with polyangiitis: ?
Churg-Strauss

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

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What does ANCA stand for?
Antineutrophil cytoplasmic antibodies

What are they?
Autoantibodies against antigens found within the cytoplasm of neutrophils

With which specific ANCA pattern is each condition associated?
Granulomatosis with polyangiitis: Cytoplasmic (c-ANCA)
Churg-Strauss
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

- Saddle-nose deformity (2): RP; GwP
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Autoantibodies against antigens found within the cytoplasm of neutrophils

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ANCA positive (2): GwP; CS

What does ANCA stand for?
Antineutrophil cytoplasmic antibodies

What are they?
Autoantibodies against antigens found within the cytoplasm of neutrophils

With which specific ANCA pattern is each condition associated?
Granulomatosis with polyangiitis: Cytoplasmic (c-ANCA)
Churg-Strauss: Perinuclear (p-ANCA)
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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What about PAN? I thought it was ANCA-positive as well.
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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- Ulcer has overhanging edge (2): MU; PAN
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- ANCA positive (2): GwP; CS; PAN?

What about PAN? I thought it was ANCA-positive as well.

This is a sticky widget. In the 1990s, rheumatologists determined that the label PAN was being applied to conditions that were actually separate disease entities. Thus, PAN was subdivided into several conditions:

-- Classic PAN
-- Microscopic polyangiitis
What about PAN? I thought it was ANCA-positive as well.

This is a sticky widget. In the 1990s, rheumatologists determined that the label PAN was being applied to conditions that were actually separate disease entities. Thus, PAN was subdivided into several conditions:

--Classic PAN, which affects...[description of involved vessels]

--Microscopic polyangiitis
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): RP; GwP
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--- Classic PAN, which affects…medium- and small-sized ‘muscular’ arteries

--- Microscopic polyangiitis
What about PAN? I thought it was ANCA-positive as well.
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--Classic PAN, which affects…medium- and small-sized ‘muscular’ arteries; and
--Microscopic polyangiitis, which affects…[ditto]
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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--**Classic PAN**, which affects...medium- and small-sized ‘muscular’ arteries; and

--**Microscopic polyangiitis**, which affects...smaller arteries, arterioles, capillaries and venules.
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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-- **Classic PAN**, which affects…medium- and small-sized ‘muscular’ arteries; and

-- **Microscopic polyangiitis**, which affects…smaller arteries, arterioles, capillaries and venules.

It turns out microscopic polyangiitis is strongly ANCA-positive, but classic PAN is not.
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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--Classical PAN, which affects medium- and small-sized ‘muscular’ arteries; and
--Microscopic polyangiitis, which affects smaller arteries, arterioles, capillaries and venules.

It turns out microscopic polyangiitis is strongly ANCA-positive, but classic PAN is not. Because of its ANCA-positivity, microscopic angiitis is now considered to be more closely related to Churg-Strauss, and especially granulomatosis with polyangiitis, than it is to PAN.
What about PAN? I thought it was ANCA-positive as well.

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It turns out microscopic polyangiitis is strongly ANCA-positive, whereas classic PAN is not. (Because of its ANCA-positivity, microscopic polyangiitis is now considered to be more closely related to Churg-Strauss, and especially granulomatosis with polyangiitis with polyangiitis.

What's the difference between a 'small-sized' artery and a 'smaller' artery?

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

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<tr>
<td>Rheumatoid arthritis (RA)</td>
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<tr>
<td>Mooren’s ulcer (MU)</td>
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<tr>
<td>Relapsing polychondritis (RP)</td>
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<tr>
<td>Granulomatosis with polyangiitis (GwP)</td>
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</tr>
<tr>
<td>Churg-Strauss (CS)</td>
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</tr>
</tbody>
</table>

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

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<thead>
<tr>
<th>Cause</th>
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*What about PAN? I thought it was ANCA-positive as well.*

This is a sticky widget. In the 1990s, rheumatologists determined that the label PAN was being applied to conditions that were actually separate disease entities. Thus, PAN was subdivided into several conditions:

- **Classic PAN**, which affects…medium- and small-sized ‘muscular’ arteries; and
- **Microscopic polyangiitis**, which affects…smaller arteries, arterioles, capillaries and venules.

It turns out microscopic polyangiitis is strongly ANCA-positive, whereas classic PAN is not. (Because of its ANCA-positivity, microscopic polyangiitis is now considered to be more closely related to Churg-Strauss, and especially granulomatosis with polyangiitis.)

*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.
What about PAN? I thought it was ANCA-positive as well. This is a sticky widget. In the 1990s, rheumatologists determined that the label PAN was being applied to conditions that were actually separate disease entities. Thus, PAN was subdivided into several conditions:–*Classic PAN*, which affects only medium- and small-sized ‘muscular’ arteries; and–*Microscopic polyangiitis*, which affects smaller arteries, arterioles, capillaries and venules. It turns out microscopic polyangiitis is strongly ANCA-positive, but classic PAN is not. Because of its ANCA-positivity, microscopic angiitis is now considered to be more closely related to Churg-Strauss, and especially granulomatosis with polyangiitis, than it is to PAN.

Got it. So if it’s ANCA+ it’s not PAN, right?
What about PAN? I thought it was ANCA-positive as well.
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--- Classic PAN, which affects only medium- and small-sized ‘muscular’ arteries; and

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---

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

<table>
<thead>
<tr>
<th>Effect</th>
<th>Causes</th>
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<tbody>
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<tr>
<td>Asthma and eosinophilia</td>
<td>CS</td>
</tr>
<tr>
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<td>RP</td>
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<tr>
<td>Ulcer has overhanging edge (2)</td>
<td>MU; PAN</td>
</tr>
<tr>
<td>Sclera never involved</td>
<td>MU</td>
</tr>
<tr>
<td>ANCA positive (2)</td>
<td>GwP; CS; PAN (10% of cases)</td>
</tr>
</tbody>
</table>

---

OK, tell me this much at least--are both classic PAN and microscopic angiitis associated with PUK?
What about PAN? I thought it was ANCA-positive as well.

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-- **Classic PAN**, which affects only medium- and small-sized 'muscular' arteries;
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<th>Condition</th>
<th>Causes Associating</th>
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<td>Saddle-nose deformity (2)</td>
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<tr>
<td>Is a diagnosis of exclusion</td>
<td></td>
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</tbody>
</table>
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

<table>
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<tr>
<th>Cause</th>
<th>Associated Causes</th>
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<td>Polyarteritis nodosa (PAN)</td>
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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
- Sclera never involved: MU
- ANCA positive (2): GwP; CS; PAN
- Is a diagnosis of exclusion: MU
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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<tr>
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<th>Causes Associated</th>
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- Sclera never involved: MU
- ANCA positive (2): GwP; CS; PAN
- Is a diagnosis of exclusion: MU
- Chest X-ray likely abnormal (3):
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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- Is a diagnosis of exclusion: MU
- Chest X-ray likely abnormal (3): GwP; CS; RP
Q

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

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<th>Cause</th>
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- 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:
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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<tr>
<th>Cause of PUK</th>
<th>Associations</th>
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<td>Is a diagnosis of exclusion</td>
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<tr>
<td>Chest X-ray likely abnormal (3)</td>
<td>GwP; CS; RP</td>
</tr>
<tr>
<td>Associated with hepatitis seropositivity</td>
<td>PAN</td>
</tr>
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</table>
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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<th>GwP</th>
<th>CS</th>
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<th>PAN</th>
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Which hepatitis virus is definitely associated with PAN?
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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<tr>
<td>Associated with hepatitis seropositivity</td>
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Which hepatitis virus is definitely associated with PAN?  
Hepatitis B
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

Which hepatitis virus is definitely associated with PAN?
Hepatitis B

What percent of PAN pts test positive for are Hep B surface Ag?
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

<table>
<thead>
<tr>
<th>Causes</th>
<th>Polymyositis nodosa (PAN)</th>
<th>Relapsing polychondritis (RP)</th>
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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?

About 10
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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?**
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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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
- 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

*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?*
Hepatitis C
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:
- 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
Q

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

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

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- PAN: Polyarteritis nodosa
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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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All forms of inflammatory PUK are painful, but Mooren’s is exceptionally so!
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<tbody>
<tr>
<td>Saddle-nose deformity (2)</td>
<td>RP; GwP</td>
</tr>
<tr>
<td>Asthma and eosinophilia</td>
<td>CS</td>
</tr>
<tr>
<td>Deformed auricular pinnae</td>
<td>RP</td>
</tr>
<tr>
<td>Ulcer has overhanging edge (2)</td>
<td>MU; PAN</td>
</tr>
<tr>
<td>Sclera never involved</td>
<td>MU</td>
</tr>
<tr>
<td>ANCA positive (2)</td>
<td>GwP; CS; RP</td>
</tr>
<tr>
<td>Is a diagnosis of exclusion</td>
<td>MU</td>
</tr>
<tr>
<td>Chest X-ray likely abnormal (3)</td>
<td>GwP; CS; RP</td>
</tr>
<tr>
<td>Associated with hepatitis seropositivity</td>
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</tr>
<tr>
<td>Associated with helminthic seropositivity</td>
<td>MU</td>
</tr>
<tr>
<td>Renal function may be impaired (4)</td>
<td>GwP; PAN; CS; RP</td>
</tr>
<tr>
<td>Chronic, tx-resistant sinusitis common</td>
<td>GwP</td>
</tr>
<tr>
<td>Extremely painful</td>
<td>MU</td>
</tr>
<tr>
<td>Anti-CCP Ab positive</td>
<td>RA</td>
</tr>
</tbody>
</table>

What does CCP stand for in this context?
Cyclic citrullinated peptide

What does it mean to say a peptide has been citrullinated?
It means that some of the arginine moieties within proteins have been enzymatically converted to the amino acid citrulline (which isn’t one of the 20 standard AAs coded for in our genome)

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.

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

Anti-CCP Ab positive: RA
Saddle-nose deformity (2): RP; GwP

Asthma and eosinophilia: CS

Ulcer has overhanging edge (2): MU; PAN

Sclera never involved: MU

ANCA positive (2): GwP; CS

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): GwP; PAN; CS; RP

Chronic, tx-resistant sinusitis common: GwP

Extremely painful: MU

Anti-CCP Ab positive: RA

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When evaluating a pt for RA, I usually check for the presence of serum rheumatoid factor (RF).

Anti-CCP Ab positive: RA
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

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

What does CCP stand for in this context?
Cyclic citrullinated peptide

What does it mean to say a peptide has been citrullinated?
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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?

Anti-CCP Ab positive: RA
For each statement, identify which of these causes of PUK is/are associated (some will more than one answer)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Associated with</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyarteritis nodosa (PAN)</td>
<td>PAN; RP; GwP</td>
</tr>
<tr>
<td>Rheumatoid arthritis (RA)</td>
<td>GwP; CS; RP</td>
</tr>
<tr>
<td>Mooren’s ulcer (MU)</td>
<td>PAN; CS; RP; GwP; RP</td>
</tr>
<tr>
<td>Ulcer has overhanging edge (2)</td>
<td>MU; PAN; GwP; CS; RP; RP</td>
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**Q/A**

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

**What does CCP stand for in this context?**
Cyclic citrullinated peptide

**What does it mean to say a peptide has been citrullinated?**
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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.

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

<table>
<thead>
<tr>
<th>Polyarteritis nodosa (PAN)</th>
<th>Relapsing polychondritis (RP)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatoid arthritis (RA)</td>
<td>Granulomatosis with polyangiitis (GwP)</td>
</tr>
<tr>
<td>Mooren’s ulcer (MU)</td>
<td>Churg-Strauss (CS)</td>
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- Saddle-nose deformity (2): RP; GwP
- Asthma and eosinophilia: CS

**What does CCP stand for in this context?**
Cyclic citrullinated peptide

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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
Q

Concerning PUK

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

(IBM = 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
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
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?
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 --
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
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.
Concerning PUK

- 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?
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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

If the epithelium is intact, what is going on that puts Terrien’s on the DDx for PUK?

- 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

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

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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
--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

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

**Concerning PUK**

If the epithelium is intact, what is going on that puts Terrien’s on the DDx for PUK?
- Progressive peripheral stromal thinning makes the limbal region in Terrien’s resemble PUK
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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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Concerning PUK

Speaking of Terrien’s…

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

Uncommon

---

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Concerning PUK

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Is it a common, or an uncommon condition?
Uncommon

Does it have a gender predilection?

Why is Terrien’s the oddball in this group?

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Concerning PUK

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 [male]

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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.
Concerning PUK

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

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Two reasons:
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--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.
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And 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 males

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

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

*Is it unilateral, or bilateral?*

Bilateral (although involvement can be strikingly asymmetric)

Does it affect vision? If so, how?

Yes, by inducing high astigmatism
Concerning PUK

Speaking of Terrien’s…
Is it a common, or an uncommon condition?
Uncommon

Does it have a gender predilection?
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Terrien’s marginal

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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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Two reasons:

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Concerning PUK

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Is it a common, or an uncommon condition?

Uncommon

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?

Terrien’s marginal
With respect to manifesting PUK, which of the following doesn’t belong, and why?

- RA, Mooren’s, Behçet, IBD
- Mooren’s, Terrien’s marginal, Sarcoid, SLE

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And in this group?

- Mooren’s, Terrien’s marginal, Sarcoid, SLE

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Concerning PUK

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

*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

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

Mooren’s, Terrien’s marginal, Sarcoid, SLE

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Concerning PUK

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 **intact**, as noted earlier
-- The **leading edge** of the thinned area is characterized by the presence of…

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, Sarcoid, SLE

Terrien’s marginal

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.
Concerning PUK

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 intact, as noted earlier
-- The leading edge of the thinned area is characterized by the presence of… Lipid

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

Why is Terrien’s the oddball in this group?
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Concerning PUK

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 **intact**, as noted earlier
--The **leading edge** of the thinned area is characterized by the presence of… **Lipid**
--The **trailing portion** is characterized by the presence of a…

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**

**Terrien’s marginal**

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.
Concerning PUK

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 **intact**, as noted earlier
--The **leading edge** of the thinned area is characterized by the presence of... **Lipid**
--The **trailing portion** is characterized by the presence of a... **Vascular pannus**

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

Another point to make:

Mooren’s, Steroid-induced, 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.
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.
Concerning PUK

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

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 intact, as noted earlier
--The leading edge of the thinned area is characterized by the presence of…Lipid
--The trailing portion is characterized by the presence of a…Vascular pannus

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

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

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

Why is Terrien’s the oddball in this group?
Two reasons:
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Concerning PUK

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Concerning PUK

Speaking of Terrien’s…

Is it a common, or an uncommon condition?
Uncommon

Does Terrien’s render the cornea significantly thinner than normal?
Yes

Does it have gender predilection?
While once thought to be more common in males, it is now considered equal

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

Does Terrien’s render the cornea significantly thinner than normal?
Yes

Is the thinned Terrien’s cornea at risk for rupture with mild trauma?
Yes

Do Terrien pts need to wear protective eyewear?
Yes

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

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 \textbf{intact} in Terrien’s.
Concerning PUK

**Speaking of Terrien’s…**

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

Uncommon

*Does it have a gender predilection?*

While once thought to be more common in males, it is now considered equal

*Is it unilateral, or bilateral?*

Bilateral (although involvement can be strikingly asymmetric)

*Does Terrien’s render the cornea significantly thinner than normal?*

Yes

*Is the thinned Terrien’s cornea at risk for rupture with mild trauma?*

Yes

*Does it affect vision? If so, how?*

Yes, by inducing high astigmatism

*Is the thinned Terrien’s cornea at risk for rupture with mild trauma?*

Yes

*Does Terrien’s marginal render the cornea significantly thinner than normal?*

Yes

*Is the thinned Terrien’s marginal cornea at risk for rupture with mild trauma?*

Yes

*Does Terrien’s marginal affect vision? If so, how?*

Yes, by inducing high astigmatism

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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 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

And in this group?

Mooren’s, Terrien’s marginal, Sarcoid, SLE

Why is Terrien’s the oddball in this group?

Two reasons:

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--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.

Concerning PUK

Speaking of Terrien’s...

Is it a common, or an uncommon condition?

Uncommon

Does it have a gender predilection?

While once thought to be more common in males, it is now considered equal

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

Does Terrien’s render the cornea significantly thinner than normal?

Yes

Is the thinned Terrien’s cornea at risk for rupture with mild trauma?

Yes

Do Terrien pts need to wear protective eyewear?

Yes

Does Terrien’s marginal

Mooren’s, Terrien’s marginal, Sarcoid, SLE
With respect to manifesting PUK, which of the following doesn't belong, and why?

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And in this group?

Mooren’s, Terrien’s marginal, Sarcoid, SLE

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Concerning PUK

Speaking of Terrien’s?
- Is it a common, or an uncommon condition?  
  Uncommon
- Does Terrien’s render the cornea significantly thinner than normal?  
  Yes
- Does it have a gender predilection?  
  No
- While once thought to be more common in males, it is now considered equal
- 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.

Concerning Terrien’s?
- Concerning Terrien’s?
  - Is it a common, or an uncommon condition?  
    Uncommon
  - Does it have a gender predilection?  
    No
  - While once thought to be more common in males, it is now considered equal

There is a lookalike condition--rarer than Terrien’s—which differs in that 1) it is more likely to occur in children, and 2) it is inflammatory in nature. The Cornea book speculates that it might not be a separate condition, but rather a manifestation of the same dz process as Terrien. What is it?

- Fuchs’ superficial marginal keratitis

Are there any lookalikes?
- Yes

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

...
Concerning PUK

Speaking of Terrien’s?

Is it a common, or an uncommon condition?

Uncommon

Does Terrien’s render the cornea significantly thinner than normal?

Yes

While once thought to be more common in males, it is now considered equal

There is a lookalike condition--rarer than Terrien’s—which differs in that 1) it is more likely to occur in children, and 2) it is inflammatory in nature. The Cornea book speculates that it might not be a separate condition, but rather a manifestation of the same dz process as Terrien. What is it?

Fuchs’ superficial marginal keratitis

Does it affect vision? If so, how?

Yes, by inducing high astigmatism

Does Terrien’s marginal

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Two reasons:

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--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.
Concerning PUK

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
All of the following are true concerning Mooren’s ulcer *except* (could be more than one):

- Cause is unknown "T"
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- Mooren’s responds readily to aggressive local therapy

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.