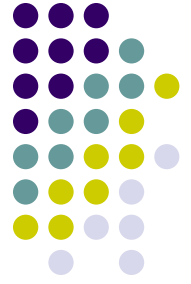


Q

Re ***Stevens-Johnson syndrome:***
True, False, or Controversial?

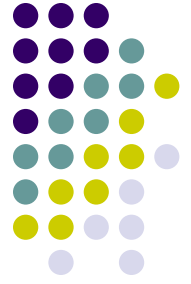
- SJS is also known as *erythema multiforme minor*



A

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

- SJS is also known as *erythema multiforme* ^{major} ~~minor~~ **False**



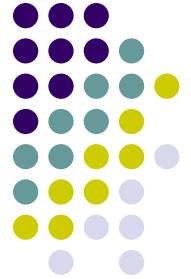
Q

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

- SJS is also known as *erythema multiforme*^{major}_^~~minor~~ **False**

Acute inflammatory vesiculobullous reaction =

two words



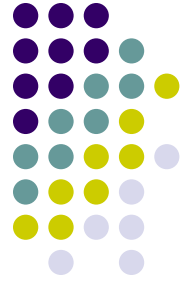
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- SJS is also known as *erythema multiforme*^{major}_{minor} **False**

Acute inflammatory vesiculobullous reaction =

Erythema Multiforme



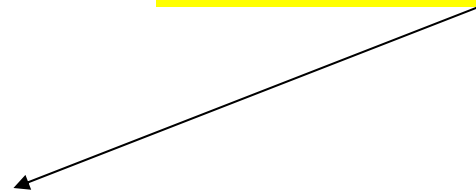
Q

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True, False, or Controversial?

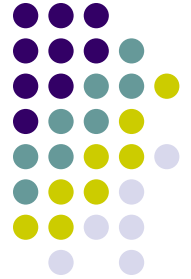
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Erythema Multiforme



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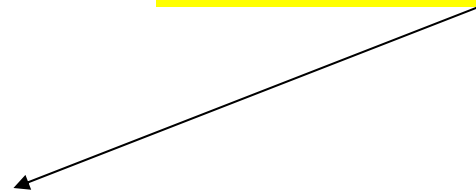
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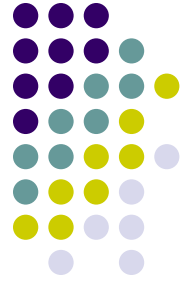
Re *Stevens-Johnson syndrome:*
True, False, or Controversial?



Erythema multiforme

Q

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?



- SJS is also known as *erythema multiforme* ^{major} ~~minor~~ **False**

Acute inflammatory vesiculobullous reaction =

Erythema Multiforme

Erythema multiforme limited to skin =
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Erythema multiforme involving skin
and mucous membranes =
Erythema Multiforme aka

three words and their (abb.)

A

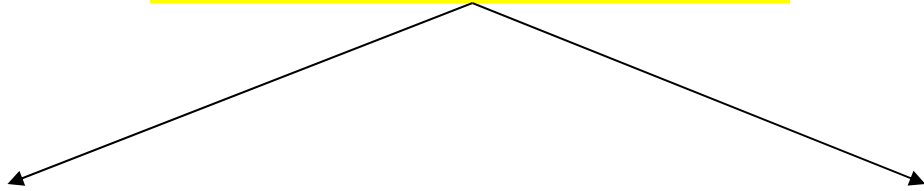
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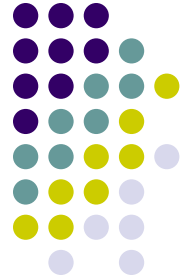
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Erythema multiforme limited to skin =
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Stevens-Johnson syndrome (SJS)

Re *Stevens-Johnson syndrome*:
True, False, or Controversial?



SJS

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?



- SJS is also known as *erythema multiforme*^{major}_{minor} False

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and mucous membranes =
Erythema Multiforme **Major** aka
Stevens-Johnson syndrome (SJS)

Note: The latest (in my possession) version of the BCSC *External Disease* book is the source I used in asserting the equivalency of these two terms. However, some well-respected sources (eg, *UpToDate*) insist that SJS is **not** a form of erythema multiforme at all—that it is a distinct and separate disease entity.

All that said, until/unless the description in the BCSC changes, I recommend going with the above for the OKAP/Boards.

Q

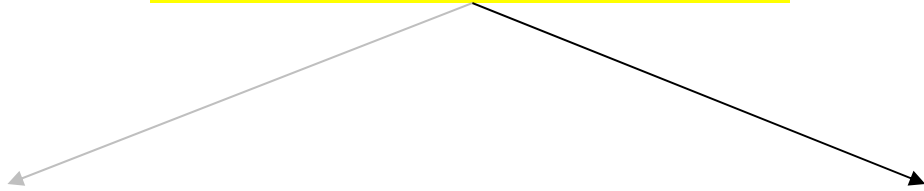
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Mucous membranes at what three sites are most frequently involved?

A

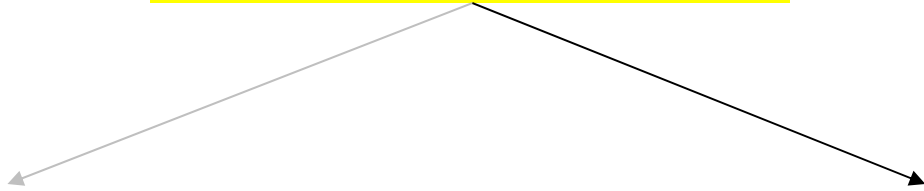
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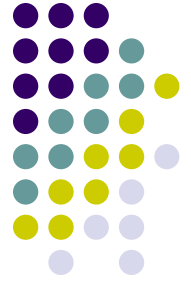
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Mucous membranes at what three sites are most frequently involved?
The eyes, the mouth, and the genitals

Q

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True, False, or Controversial?



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Mucous membranes at what three sites are most frequently involved?

The **eyes**, the mouth, and the genitals

In what percent of cases are the mucous membranes of the eye involved?

A

Re **Stevens-Johnson syndrome:**
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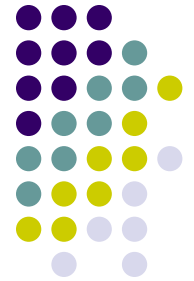
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Mucous membranes at what three sites are most frequently involved?
The **eyes**, the mouth, and the genitals

In what percent of cases are the mucous membranes of the eye involved?
About 80 (Note: The *Peds BCSC* book pegs the rate of ocular involvement in children at ~50%)

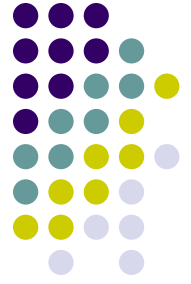
Re *Stevens-Johnson syndrome:*
True, False, or Controversial?



SJS with a corneal epithelial defect and diffuse conjunctival injection

Q

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True, False, or Controversial?



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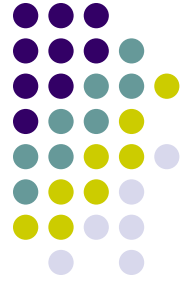
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Erythema Multiforme **Major**, aka
Stevens-Johnson syndrome (SJS)

Severe erythema multiforme major =

three words and their (abb.)

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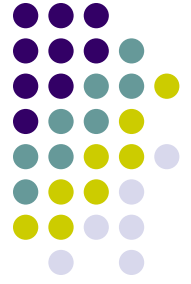
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Q

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How is severe defined here?

Severe erythema multiforme major =
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A/Q

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How is severe defined here?

With respect to the % of

four words

involved

Severe

erythema multiforme major =
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A

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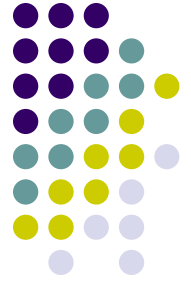
Erythema multiforme involving skin
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How is severe defined here?
With respect to the % of **total body surface area** involved

Severe erythema multiforme major =
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Q

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How is severe defined here?

With respect to the % of **total body surface area** involved

What %TBSA is involved in each?

--SJS:

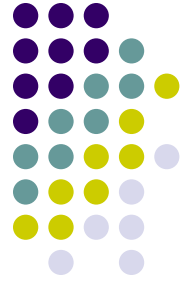
--TEN:

Severe

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A

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Erythema Multiforme **Major**, aka
Stevens-Johnson syndrome (SJS)

How is severe defined here?

With respect to the % of **total body surface area** involved

What %TBSA is involved in each?

--SJS: <10

--TEN: >30

Severe

erythema multiforme major =
Toxic Epidermal Necrolysis (TEN)

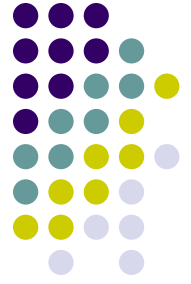
Re *Stevens-Johnson syndrome:*
True, False, or Controversial?



TEN

Q

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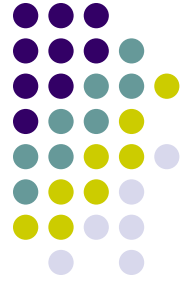
OK, but what if 10 to 30% TBSA is involved?

Severe

erythema multiforme major =
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A

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How is severe defined here?

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What %TBSA is involved in each?

--SJS: <10

--TEN: >30

OK, but what if 10 to 30% TBSA is involved?

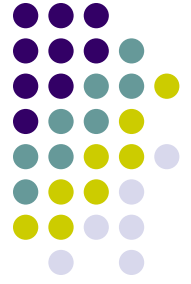
That's called SJS-TEN

Severe

erythema multiforme major =
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Q

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True, False, or Controversial?



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Erythema Multiforme **Major**, aka
Stevens-Johnson syndrome (SJS)

Where does ocular cicatricial pemphigoid
(OCP) fit in here?

Severe erythema multiforme major =
Toxic Epidermal Necrolysis (TEN)

A

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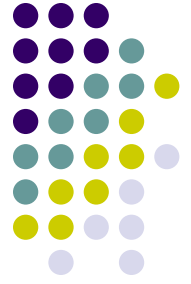
Where does ocular cicatricial pemphigoid (OCP) fit in here?
It doesn't. SJS is an immune dz (much more on this shortly), whereas OCP is an **autoimmune** dz. While these conditions inhabit one another's DDX, they are distinct entities.

Severe erythema multiforme major =
Toxic Epidermal Necrolysis (TEN)

Q

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

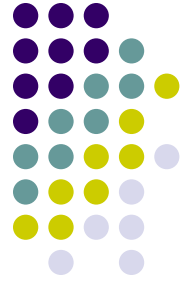
- SJS is also known as *erythema multiforme* ^{major} ~~minor~~ **False**
- Incidence is higher in young people



A

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True, False, or Controversial?

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Q

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

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What does 'young people' mean?

A

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

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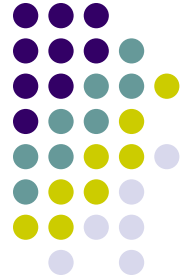


What does 'young people' mean?
Children and young adults

Q

Re **Stevens-Johnson syndrome:**
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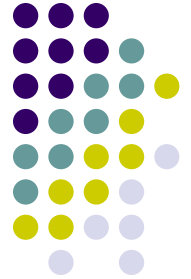
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Can it occur during other life-periods?

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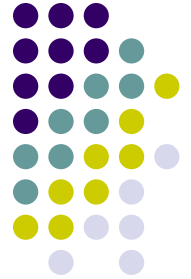
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Can it occur during other life-periods?
Yes, it can occur at any age

Q

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Children and young adults

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Is there a gender predilection?

A/Q

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Yes, it is more common in...

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Children and young adults

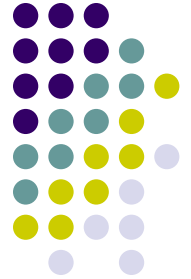
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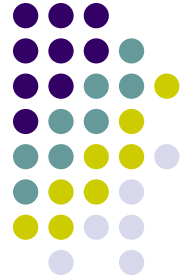
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How common is SJS?

Q/A

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True, False, or Controversial?



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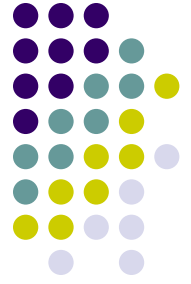
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How common is SJS?
Rare; the incidence is...

A

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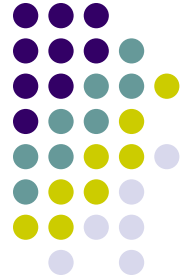
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How common is SJS?
Rare; the incidence is...~5 per million

Q

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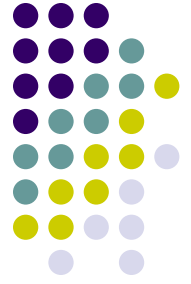
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Are HIV/AIDS pts at increased risk?

A

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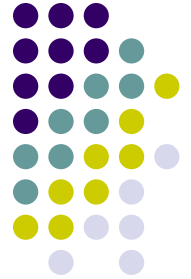
How common is SJS?
Rare; the incidence is...~5 per million

Are HIV/AIDS pts at increased risk?
Yes

Q

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?

- SJS is also known as *erythema multiforme*^{major}_{minor} False
- Incidence is higher in **young people** True



What does 'young people' mean?
Children and young adults

Can it occur during other life-periods?
Yes, it can occur at any age

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Yes, it is more common in...females

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Are pts with a malignancy at increased risk?

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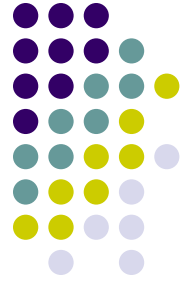
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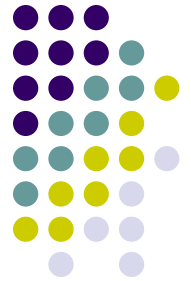


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What about for in TEN?



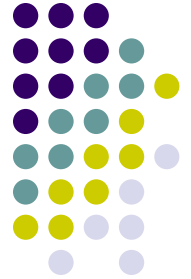
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What about for in TEN?

It's significantly higher--around 30%



Q

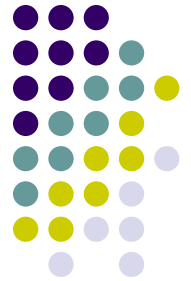
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Is there a way to remember this?



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Yup. Recall that SJS and TEN are differentiated based on the %TBSA involved. It so happens that the mortality rate for SJS (10%) and TEN (30%) are equal to their %TBSA criteria. (This is not a coincidence; the mortality rate in this disease spectrum correlates with the amount of TBSA involved.)

How is severe defined here?

With respect to the % of **total body surface area** involved

What %TBSA is involved in each?

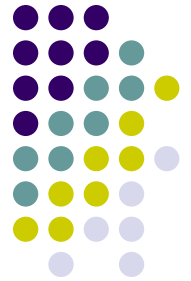
--SJS: <10

--TEN: >30

OK, but what if 10 to 30% TBSA is involved?

That's called SJS-TEN

← (Recall this box)



Q

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These numbers are for adults. Are the rates for children higher, or lower?

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Lower

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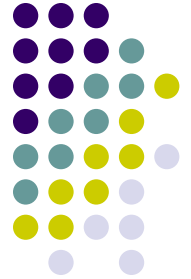
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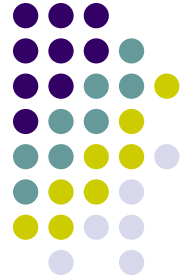
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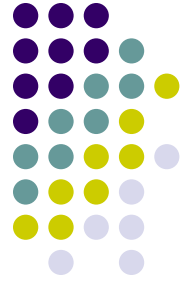


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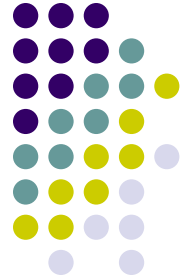


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Fever, HAs, fatigue, malaise, anorexia and GI upset

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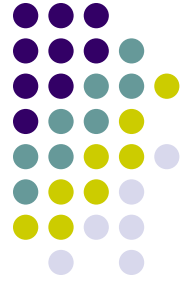
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Do these symptoms precede, or follow the skin findings?

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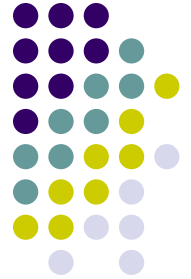
What non-skin/mucosal symptoms (and signs) are commonly associated with SJS?
Fever, HAs, fatigue, malaise, anorexia and GI upset

Do these symptoms precede, or follow the skin findings?
They precede them, usually by several days

Q

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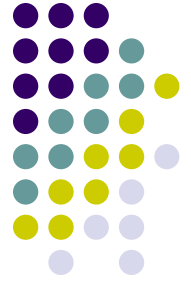


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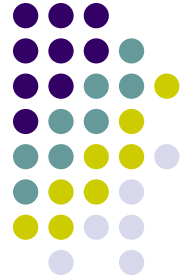
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At least 50 (most sources put the number closer to 80)

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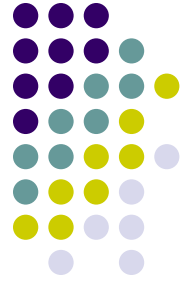
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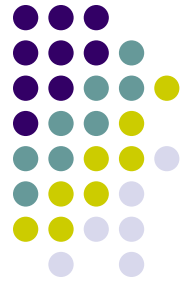
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Higher

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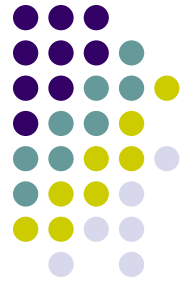
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(mnemonic forthcoming)

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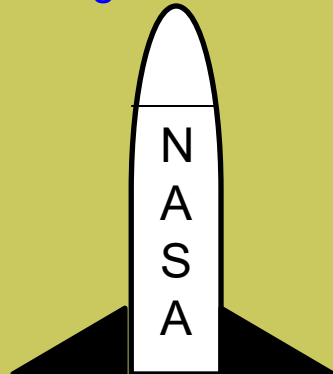
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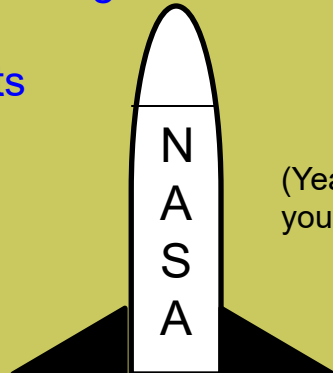
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--NSAIDs

--Anticonvulsants

--Sulfonamides

--Allopurinol



(Yeah, it's a lame mnemonic. But I bet you won't forget it!)

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If SJS occurs, it will commence within #, and unit of time of the start of drug use

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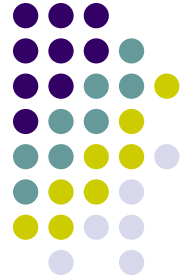
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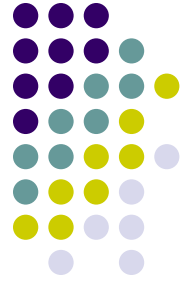
There is a possible exception to this--what is it?

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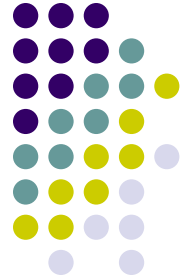
Some experts contend that a recent **dose increase in a longstanding drug** can also be the inciting event. So in addition to assessing for new drugs in the previous 8 weeks, be sure to inquire about dose increases in 'old' drugs!

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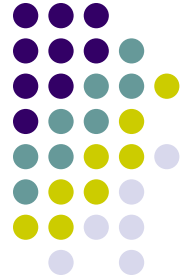
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Very much so. Prompt cessation of the offending drug can significantly impact the clinical course (including mortality). Thus, rapid identification of the offending agent can literally be a matter of life and death.

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Very much so. Prompt cessation of the offending drug can significantly impact the clinical course (including mortality). Thus, rapid identification of the offending agent can literally be a matter of life and death.

What if an offending agent can't be identified?

In this scenario, some clinicians contend that **all** home meds should be discontinued. In addition to discontinuing all meds, ... (the *in addition* will be addressed shortly)

If SJS occurs, it will commence within **8 weeks** of the start of drug use

Q

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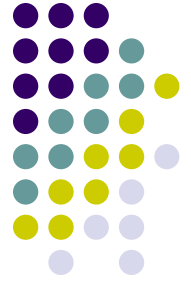


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What is the second-most common inciting factor in SJS?

A

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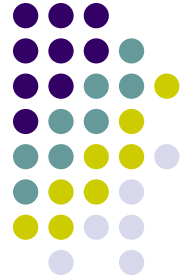
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Infectious agents

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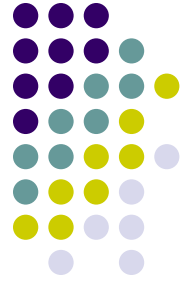
What is the second-most common inciting factor in SJS?

Infectious agents

Which infectious agent is most commonly implicated?

A

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What is the second-most common inciting factor in SJS?

Infectious agents

Which infectious agent is most commonly implicated?

Depends on who you ask. Per both *EyeWiki* and *UpToDate*, *Mycoplasma pneumoniae* is far-and-away the most common cause (*EyeWiki* attributes almost 90% of non-drug-induced cases to it). However, in the most recent version of the BCSC *Cornea/External Disease* book, *M pneumoniae* isn't even mentioned as a cause--HSV, strep and adeno are. On the other hand, the *Peds* book states that infection-related cases are usually secondary to "Mycoplasma species or herpes simplex virus." (Of course, this difference could be because *M pneumoniae* is a more common cause in *kids*.) At any rate, caveat emptor.

Re **Stevens-Johnson syndrome:**
True, False, or Controversial?



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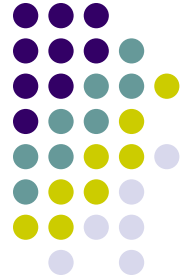
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Note: At least one reputable (but non-BCSC) source recommends that, if an inciting drug cannot be identified, the pt should undergo serologic testing to assess for acute *M pneumoniae* infection. (This is the *in addition* referred to earlier.)

other hand, the *Peds* book states that infection-related cases are usually secondary to "Mycoplasma species or herpes simplex virus." (Of course, this difference could be because *M pneumoniae* is a more common cause in *kids*.) At any rate, caveat emptor.

Q

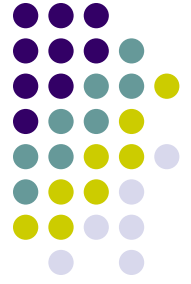
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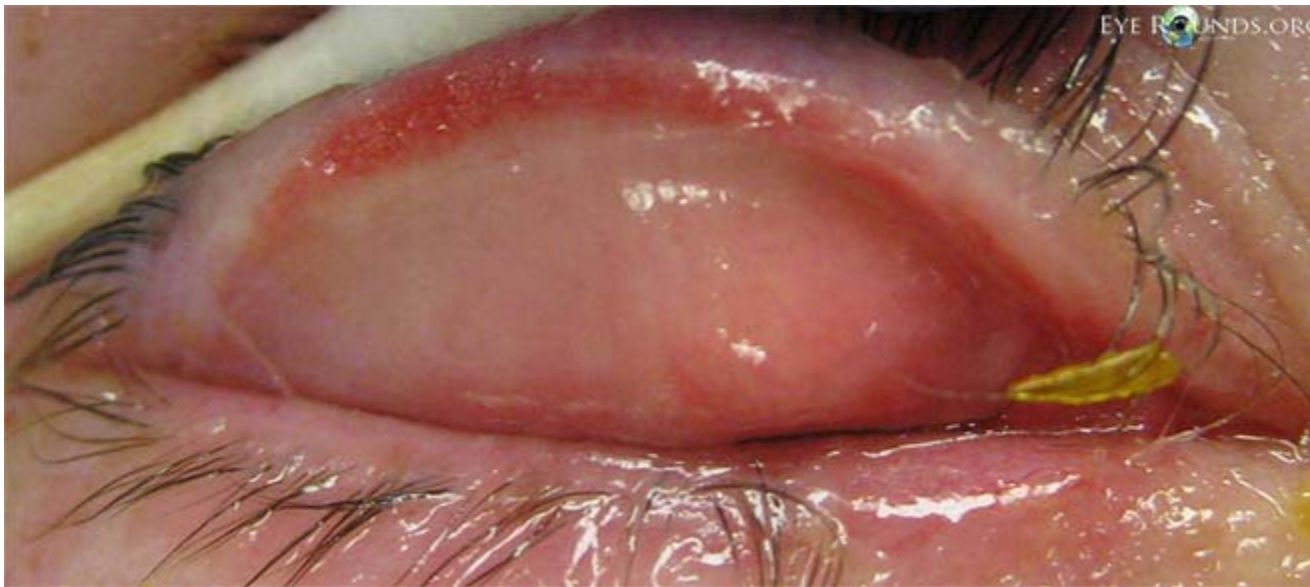
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Re *Stevens-Johnson syndrome:*
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SJS. Eversion of the right upper eyelid shows a conjunctival membrane

Q

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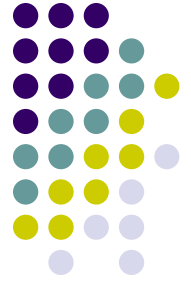


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What is the difference between a membrane and a pseudomembrane?

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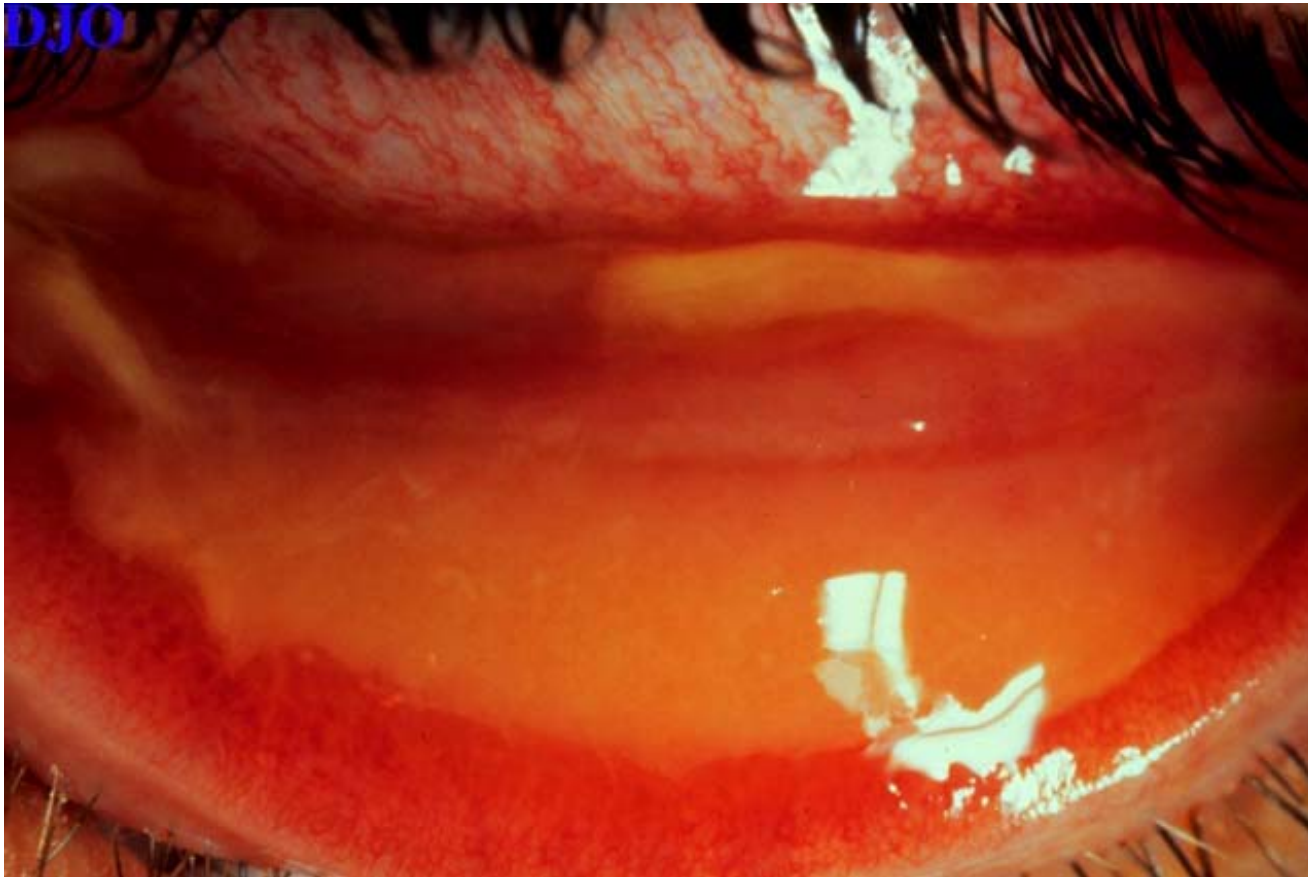
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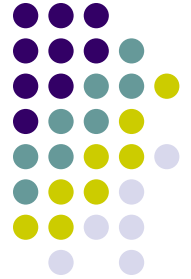
What is the difference between a membrane and a pseudomembrane?
Removal of a membrane leaves the epithelial bed bleeding; removing a pseudomembrane does not

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Membranous conjunctivitis is characterized by the formation of a layer of tissue consisting of fibroblasts, blood vessels, fibrin and inflammatory cells. Fibroblasts and blood vessels from the conj substantia propia provide a scaffold for a vascularized inflammatory membrane containing granulation tissue, **which bleeds markedly if stripped from the underlying conjunctiva.**

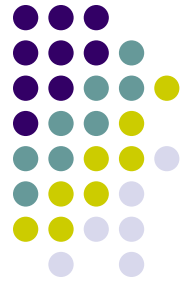
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Pseudomembranes consist of coagulated exudate that is loosely adherent to the inflamed conjunctiva. They are typically not integrated with the conjunctival epithelium and **can be removed by peeling, leaving the conjunctival epithelium intact**. Their removal produces little if any bleeding.

Q

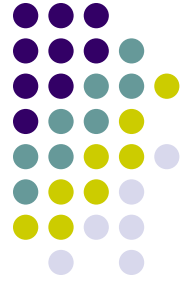
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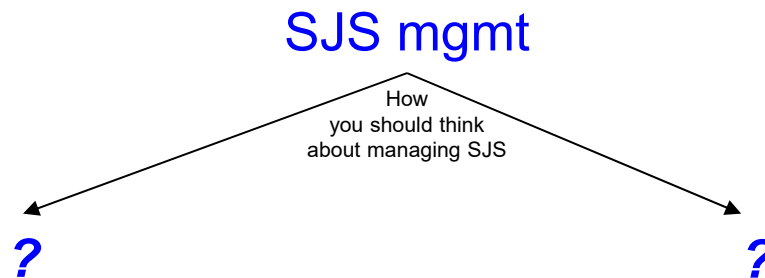
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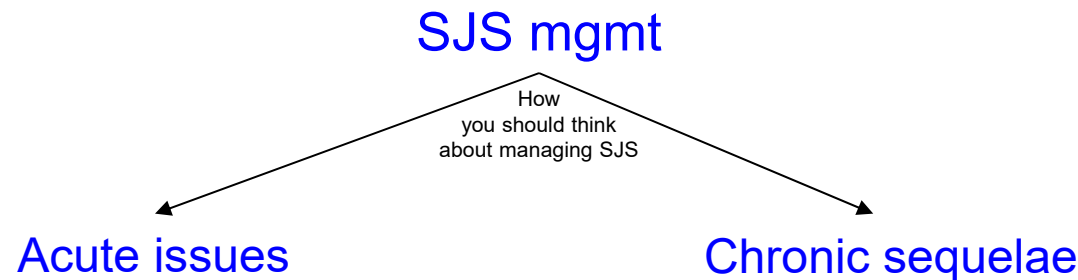


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In a nutshell, how should acute SJS be managed?

Q/A

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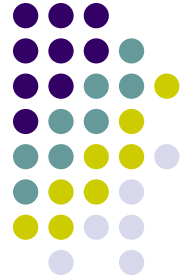
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Like a

two words

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Like a **thermal burn**

Q

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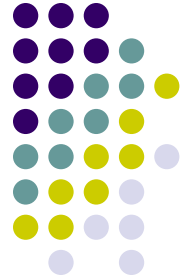
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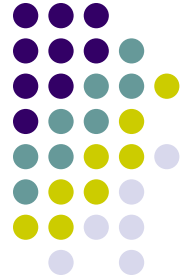
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--Maintain adequate...

--Prevent...

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In a nutshell, how should
Like a **thermal burn**

What are the two overarching goals in thermal-burn management?
--Maintain adequate...**hydration**
--Prevent...**superinfection**

Q

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How likely are SJS pts to develop chronic complications?

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Quite. The rate for children is % ; for adults, % .



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SJS mgmt

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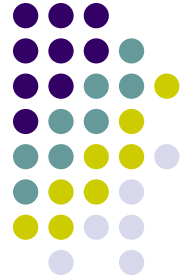
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Chronic sequelae

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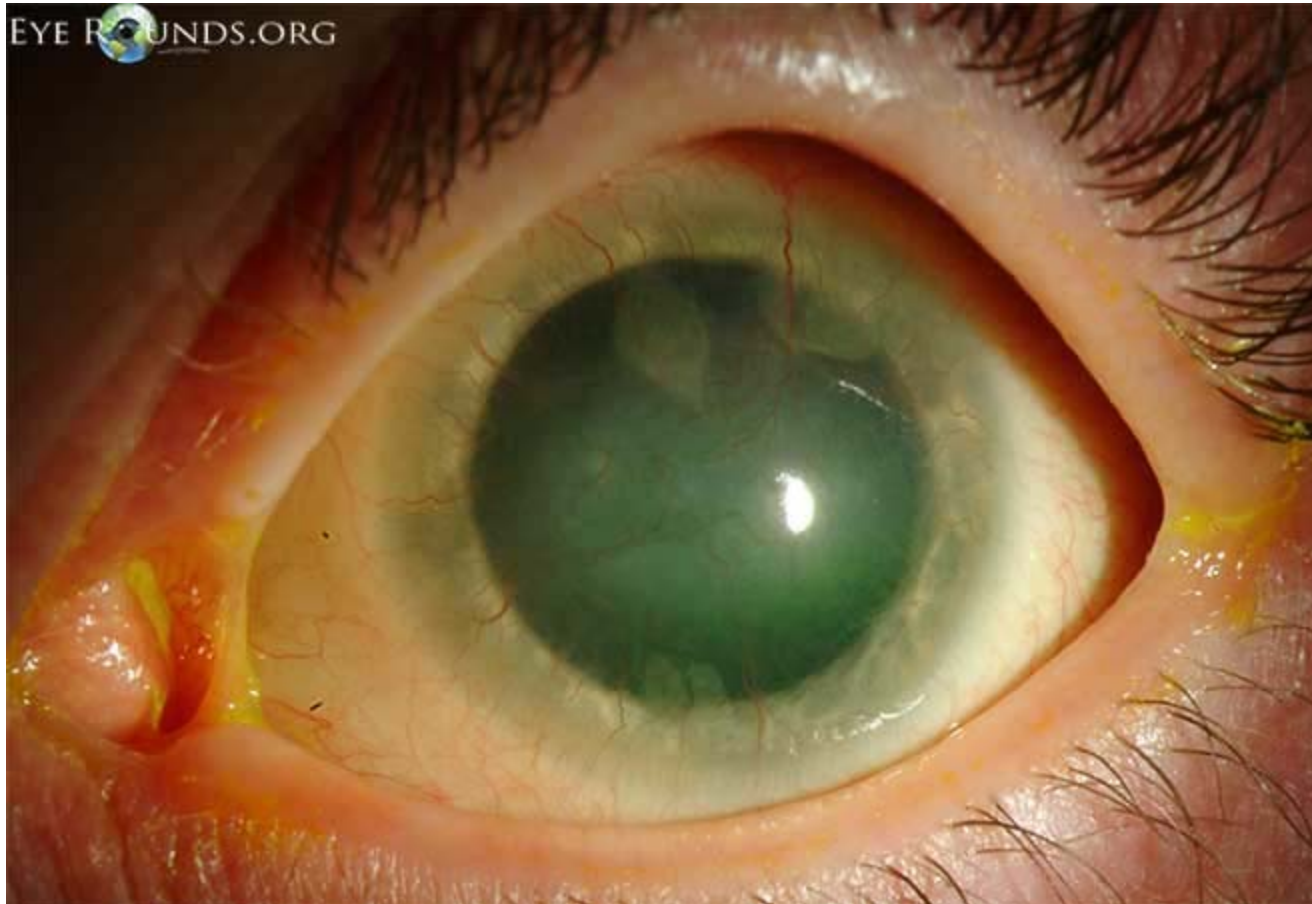
How
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What are some of the chronic ocular complications of SJS?

- Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization
- Limbal stem-cell deficiency

Chronic sequelae

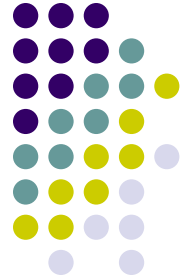
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Slit-lamp photograph showing the long-term consequences of SJS/TEN, which can include limbal stem cell deficiency, corneal vascularization and symblepharon formation, all pictured here

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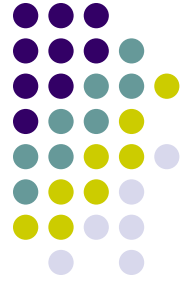
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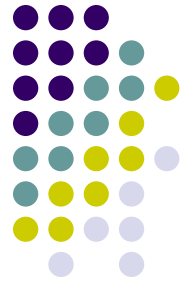
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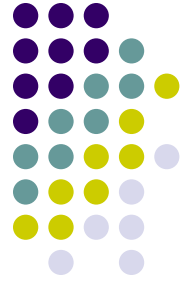
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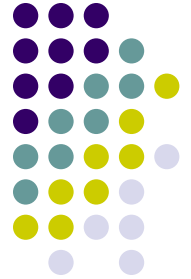
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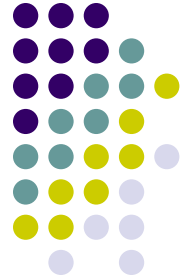
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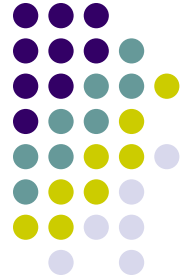
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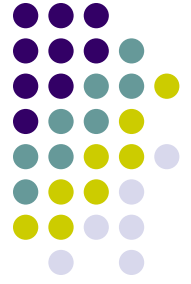
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While common, this practice is controversial. Some feel it reduces the development of significant symblephara, whereas others argue that repeated lysing prolongs (or even worsens) surface inflammation, thereby (and paradoxically) **increasing** the risk of symblephara formation.

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--Symblephara formation

- Entropion
- Ectropion
- Trichiasis
- Corneal neovascu
- Lid margin keratinization
- Limb stem-cell deficiency

Chronic sequelae

Under what circumstances would symblephara necessitate surgical redress?

A

Re *Stevens-Johnson syndrome*: *True, False, or Controversial?*



- SJS is also known as *erythema multiforme*^{major}_{minor} ~~False~~
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What are symblephara?

Adhesions between conjunctival surfaces

Adhesions between which two conj surfaces are involved?

Between the bulbar and tarsal/palpebral conj

What process produces the 'raw' conj surface needed to create symblephara?

The conj develops bullae, which leave a raw surface when they rupture

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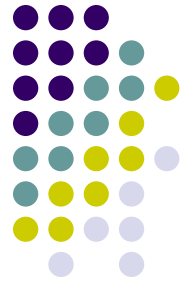
Chronic sequelae

Under what circumstances would symblephara necessitate surgical redress?

If they are impairing movement of the globe, or movement/function of the eyelid

Q

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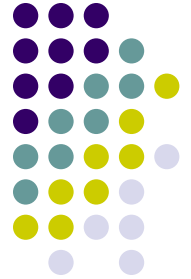
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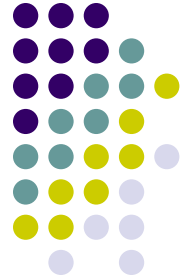
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Others include *atopic keratoconjunctivitis* (AKC), adenoviral conjunctivitis (especially *epidemic keratoconjunctivitis*, EKC), and trachoma. (There are a number of other causes to boot.)



Q

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SJS mgmt

What role do entropion, ectropion and trichiasis play in the disease process?

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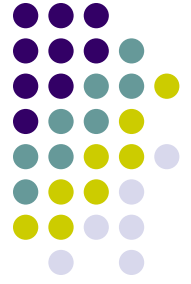
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SJS mgmt

What role do entropion, ectropion and trichiasis play in the disease process?
All three result in damage to the corneal surface, and thus promote corneal decompensation and neovascularization

Chronic sequelae

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--Symblephara

--Entropion

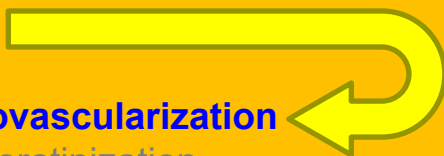
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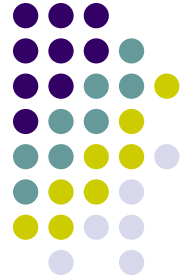


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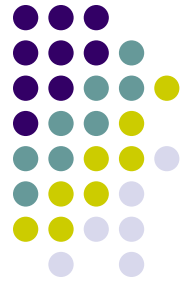
There have been reports linking systemic steroids in SJS to GI hemorrhage and sudden death. Further, as an immunosuppressant, systemic steroids increase the risk of an infection in these pts with severely compromised barrier function.

(Recall this box...)

What are the two overarching goals in thermal-burn management?
- Maintain adequate...hydration
--Prevent...**superinfection**

Q

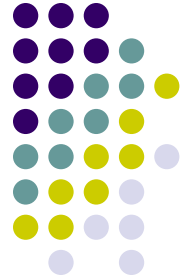
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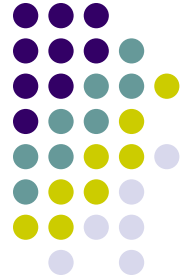
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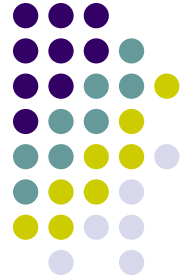
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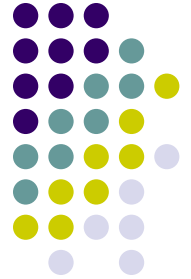
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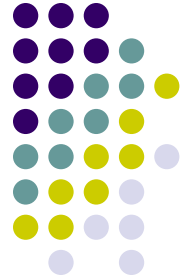


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The entire ocular surface (note that this includes the lid margins as well)