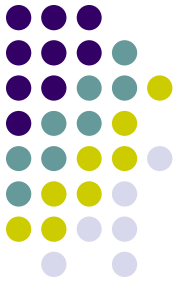


Q

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

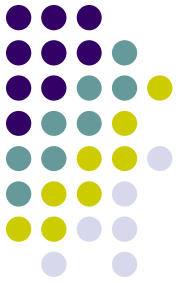
- SJS is also known as *erythema multiforme minor*



A

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

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



Q

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True, False, or Controversial?

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Acute inflammatory vesiculobullous reaction =

two words



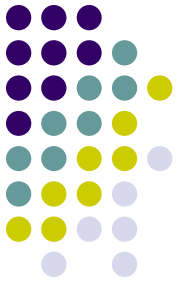
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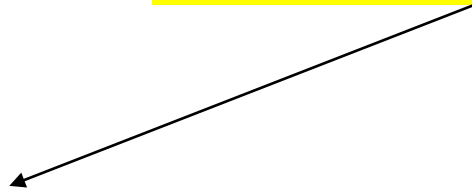
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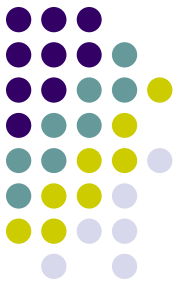
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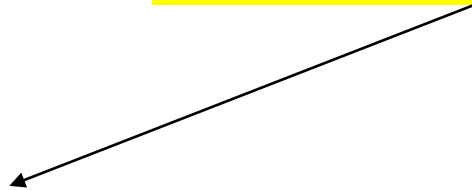
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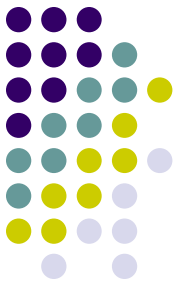
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Re *Stevens-Johnson syndrome*:
True, False, or Controversial?



Erythema multiforme

Q

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True, False, or Controversial?



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Erythema Multiforme

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Erythema multiforme involving skin
and mucous membranes =
Erythema Multiforme aka

three words and their (abb.)

A

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

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



SJS

Q

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Mucous membranes at what three sites are most frequently involved?

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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?

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

Severe erythema multiforme major =

three words and their (abb.)

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Severe erythema multiforme major =
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A/Q

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With respect to the % of involved

Severe erythema multiforme major =
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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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--TEN: >30

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



TEN

Q

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

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

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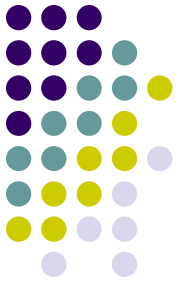
Where does ocular cicatricial pemphigoid (OCP) fit in here?

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.

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

Re **Stevens-Johnson syndrome:**
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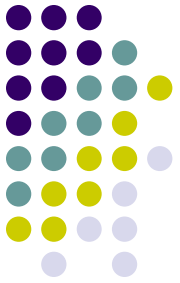
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What does 'young people' mean?

A

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- SJS is also known as *erythema multiforme*^{major}_{minor} False
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What does 'young people' mean?
Children and young adults

Q

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What does 'young people' mean?

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Can it occur during other life-periods?

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A/Q

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How common is SJS?

Q/A

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Rare; the incidence is...

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Are pts with a malignancy at increased risk?

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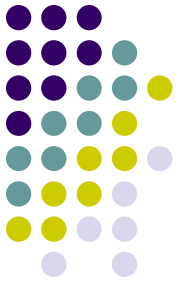
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Q

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**
- Mortality rate is about 10%



A

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True, False, or Controversial?

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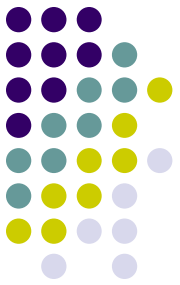


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What about for in TEN?



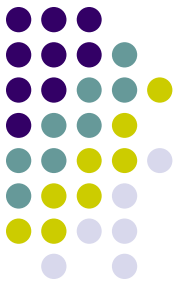
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- Incidence is higher in young people True
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What about for in TEN?

It's significantly higher--around 30%



Q

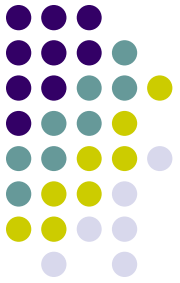
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Is there a way to remember this?



A

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It's significantly higher--around 30%

Is there a way to remember this?

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

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



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

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What about for in TEN?

It's significantly higher--**around 30%**

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

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True, False, or Controversial?

- SJS is also known as *erythema multiforme*^{major}_{minor} **False**
- Incidence is higher in young people **True**
- Mortality rate is about 10% **True**
- Systemic (non-skin/mucosal) symptoms are common



A

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What non-skin/mucosal symptoms (and signs) are commonly associated with SJS?

A

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- Mortality rate is about 10% True
- Systemic (non-skin/mucosal) symptoms are common True

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

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
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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?

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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

Re ***Stevens-Johnson syndrome:***
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At least half, and perhaps as high as 80%

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The *Cornea* book pegs it right at 80%

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Which four drugs/drug classes are most commonly implicated?

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(mnemonic forthcoming)

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Q

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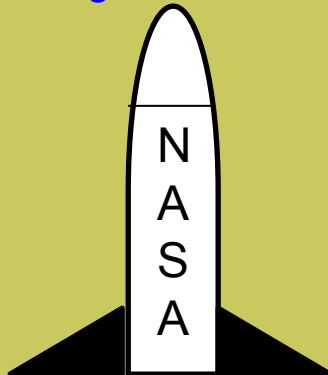
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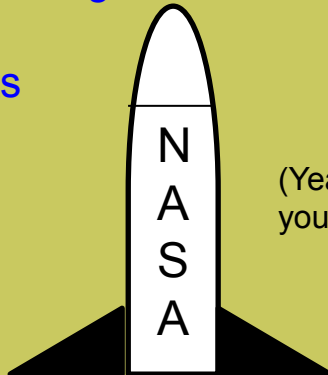
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--NSAIDs

--Anticonvulsants

--Sulfonamides

--Allopurinol



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

Q

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Q/A

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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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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)

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--M

--A

--S

--H

The mnemonic is...MASH

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Which infectious agent is considered the most common?

Mycoplasma, by a mile

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.)

Q

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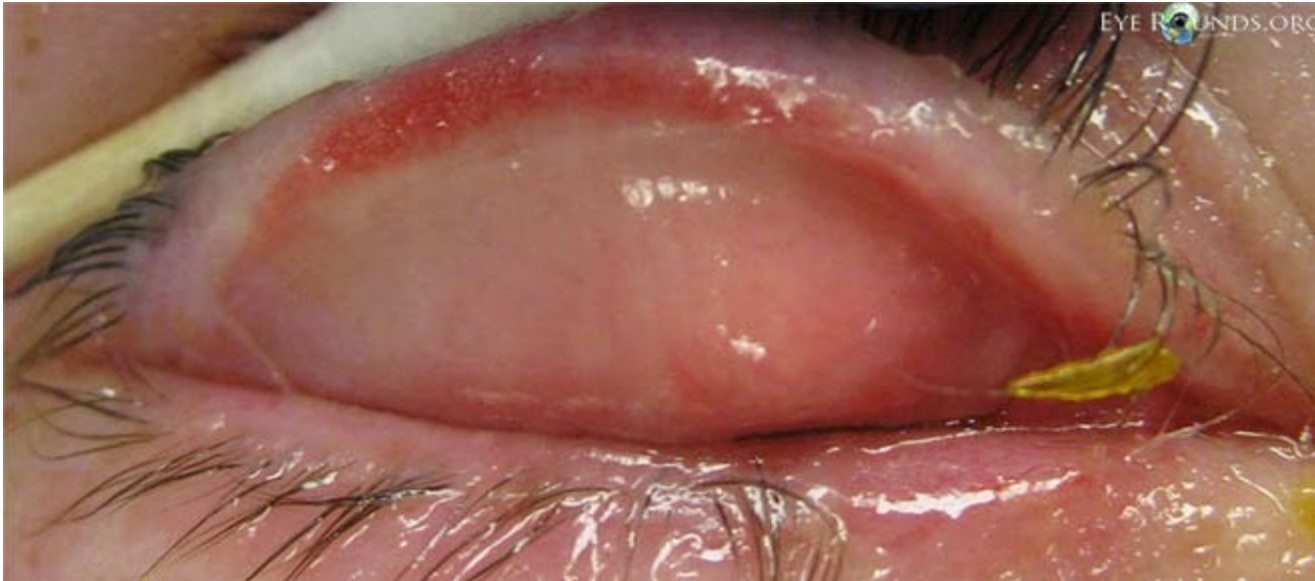
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SJS. Eversion of the right upper eyelid shows a conjunctival membrane

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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

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



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.**

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



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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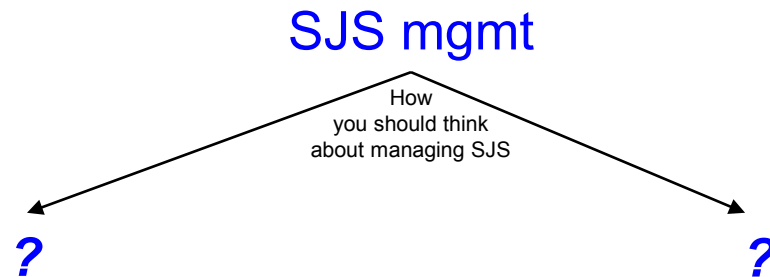
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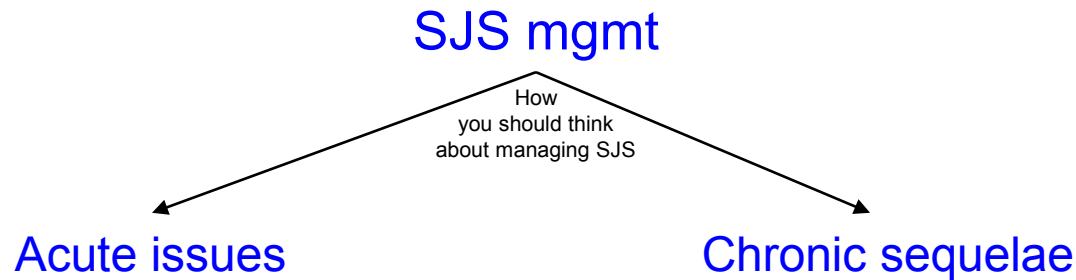


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In a nutshell, how should acute SJS be managed?

Q/A

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Like a

two words

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In a nutshell, how should acute SJS be managed?
Like a **thermal burn**

Q

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SJS mgmt

How
you should think
about managing SJS

Acute issues

Chronic sequelae

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

What are the two overarching goals in thermal-burn management?

--
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Q/A

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

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- Maintain adequate...**hydration**
- Prevent...**superinfection**

Q

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How likely are SJS pts to develop chronic complications?

Q/A

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- Chronic ocular complications are usually ^{severe} ~~mild~~ False



How likely are SJS pts to develop chronic complications?
Quite. The rate for children is % ; for adults, % .

A

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
- Mortality rate is about 10% True
- Systemic (non-skin/mucosal) symptoms are common True
- Adverse drug reaction is the most common cause True
- Membranous/pseudomembranous conjunctivitis may develop True
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How likely are SJS pts to develop chronic complications?
Quite. The rate for children is ~25% ; for adults, ~50%.

Q

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SJS mgmt

How
you should think

What are some of the chronic ocular complications of SJS?

Chronic sequelae

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SJS mgmt

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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

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



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

Q

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True, False, or Cont



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A 'frozen' globe

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two words

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Amniotic membrane grafting of the ocular surface—**specifically,**
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Adhesions between conjunctival surfaces

Adhesions between which two conjunctival surfaces are involved?

non True

What about early treatment of them (with a glaucoma filter)?

While long a controversial issue, it may even worsen the condition. Its hoped-for effect is not achieved.

If symblepharolysis and rings are no longer favored, what is favored vis a vis preventing serious ocular-surface sequelae in SJS?

Amniotic membrane grafting of the ocular surface—specifically, early amniotic membrane grafting

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What about early amniotic membrane grafting? If symblepharolysis and rings are no longer favored, what is favored in breaking them (with a glass rod)? vis a vis preventing serious ocular-surface sequelae in SJS? While long a controversial issue, early amniotic membrane grafting may prolong the healing process or even worsen the disease. Early amniotic membrane grafting may prolong the healing process or even worsen the disease. The opposite of its hoped-for effect.

Amniotic membrane grafting of the **ocular surface**—specifically, early amniotic membrane grafting.

How much of the ocular surface are we talking about?

OK then, what about amniotic membrane grafting of the ocular surface?

This too has fallen out of favor

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What about early breaking them (with a glass rod) vis a vis preventing serious ocular-surface sequelae in SJS? While long a controversial issue, it may prolong or even worsen the disease, the opposite of its hoped-for effect.

If symblepharolysis and rings are no longer favored, what is favored?

Amniotic membrane grafting of the **ocular surface**—specifically, early amniotic membrane grafting.

How much of the ocular surface are we talking about?

The whole kit and kaboodle, up to and including the lid margins

OK then, what about amniotic membrane grafting? This too has fallen out of favor.

What are some of the chronic ocular complications of SJS?

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The first 3-7 days

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While lo...
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Acute management of SJS tl;dr:

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- If/when an epi defect appears anywhere (cornea, conj, or lid margin):

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 - If/when an epi defect appears anywhere (cornea, conj, or lid margin):
- Perform two words (and keep the steroids/abx going)

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te of

OK then

This too has fallen out of favor

What are some of the chronic ocular complications of SJS?

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

Chronic sequelae

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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 conjunctival surfaces are involved?

non True

What about them (w...
While lo...
or even...
its hope...

Acute management of SJS tl;dr:

- Start PFATs and ointment immediately; follow closely
- If/when the eye becomes injected: Add topical steroids and antibiotics
- If/when an epi defect appears anywhere (cornea, conj, or lid margin):
- Perform amniotic graft (and keep the steroids/abx going)

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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

Extensive symblephara can lead to what dreaded complication?

A 'frozen' globe

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What non-SJS conditions are associated with symblepharon formation?

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may develop True

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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.)

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SJS mgmt

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

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SJS mgmt

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All three result in damage to the corneal surface, and thus promote corneal decompensation and neovascularization

Chronic sequelae

What are some

--Symblephara

--Entropion

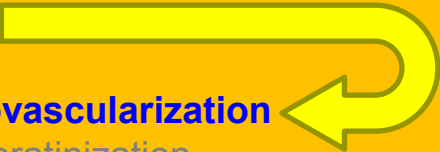
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(Recall this box...)

What are the two overarching goals in thermal-burn management?

--Maintain adequate...**hydration**

--Prevent...**superinfection**

Q

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That too

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