SJS is also known as *erythema multiforme minor*
Re *Stevens-Johnson syndrome*: True, False, or Controversial?

- SJS is also known as *erythema multiforme major* minor. False
Q

- SJS is also known as *erythema multiforme* minor. **False**

Acute inflammatory vesiculobullous reaction =  

**two words**
SJS is also known as *erythema multiforme minor* \(^\text{major}\) False

Acute inflammatory vesiculobullous reaction = *Erythema Multiforme*
- SJS is also known as \textit{erythema multiforme minor}. \textbf{False}

Acute inflammatory vesiculobullous reaction = \textbf{Erythema Multiforme}

Erythema multiforme limited to skin = \textbf{Erythema Multiforme}
- SJS is also known as *erythema multiforme minor* False

Acute inflammatory vesiculobullous reaction = **Erythema Multiforme**

Erythema mutliforme limited to skin = **Erythema Multiforme Minor**
Q

- SJS is also known as \textit{erythema multiforme minor}. \textbf{False}

Re \textit{Stevens-Johnson syndrome: True, False, or Controversial?} \textbf{False}

Acute inflammatory vesiculobullous reaction = \textit{Erythema Multiforme}

Erythema multiforme limited to skin = \textit{Erythema Multiforme Minor}

Erythema multiforme involving skin \textbf{and} mucous membranes = \textit{Erythema Multiforme Major}, aka three words and their (abb.)
SJS is also known as *erythema multiforme minor*. False

Acute inflammatory vesiculobullous reaction = **Erythema Multiforme**

- Erythema multiforme limited to skin = **Erythema Multiforme Minor**
- Erythema multiforme involving skin **and** mucous membranes = **Erythema Multiforme Major**, aka **Stevens-Johnson syndrome (SJS)**
SJS is also known as *erythema multiforme minor*. False

Acute inflammatory vesiculobullous reaction =

- Erythema Multiforme

Erythema mutliforme limited to skin =

- Erythema Multiforme Minor

Erythema mutliforme involving skin 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.
SJS is also known as erythema multiforme minor. False

Acute inflammatory vesiculobullous reaction = 

Erythema Multiforme

Erythema multiforme limited to skin = Erythema Multiforme Minor

Erythema multiforme involving skin and mucous membranes = Erythema Multiforme Major, aka Stevens-Johnson syndrome (SJS)

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%
- SJS is also known as *erythema multiforme minor* False

**Acute inflammatory vesiculobullous reaction =**

*Erythema Multiforme*

Erythema multiforme limited to skin =

Erythema Multiforme Minor

Erythema multiforme involving skin and mucous membranes =

Erythema Multiforme Major, aka

Mucous membranes at what three sites are most frequently involved?

The eyes, the mouth, and the genitals
SJS is also known as *erythema multiforme minor*. False

Acute inflammatory vesiculobullous reaction =

**Erythema Multiforme**

Erythema multiforme limited to skin =

*Erythema Multiforme Minor*

Erythema multiforme involving skin and mucous membranes =

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

*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?*
- SJS is also known as *erythema multiforme minor*. False

Acute inflammatory vesiculobullous reaction =

\[ \text{Erythema Multiforme} \]

Erythema multiforme limited to skin =

\[ \text{Erythema Multiforme Minor} \]

Erythema multiforme involving skin and mucous membranes =

\[ \text{Erythema Multiforme Major, aka Stevens-Johnson syndrome (SJS)} \]

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%)
Q

- SJS is also known as *erythema multiforme minor*. **False**

Acute inflammatory vesiculobullous reaction =

*Erythema Multiforme*

- Erythema multiforme limited to skin = *Erythema Multiforme Minor*
- Erythema multiforme involving skin and mucous membranes = *Erythema Multiforme Major*, aka *Stevens-Johnson syndrome (SJS)*

Severe *erythema multiforme major* =

*three words and their (abb.)*

Re *Stevens-Johnson syndrome*: **True, False, or Controversial?**
- SJS is also known as *erythema multiforme minor*. **False**

Acute inflammatory vesiculobullous reaction = **Erythema Multiforme**

- Erythema multiforme limited to skin = **Erythema Multiforme Minor**
- Erythema multiforme involving skin and mucous membranes = **Erythema Multiforme Major**, aka **Stevens-Johnson syndrome (SJS)**

Severe erythema multiforme major = **Toxic Epidermal Necrolysis (TEN)**
How is severe defined here?

- SJS is also known as *erythema multiforme minor* - False

Acute inflammatory vesiculobullous reaction = *Erythema Multiforme*

- Erythema multiforme limited to skin = *Erythema Multiforme Minor*
- Erythema multiforme involving skin and mucous membranes = *Erythema Multiforme Major*, aka *Stevens-Johnson syndrome (SJS)*

Severe *erythema multiforme major* = *Toxic Epidermal Necrolysis (TEN)*
**A/Q**

- SJS is also known as *erythema multiforme minor*. False

How is severe defined here? With respect to the % of four words involved.

- Severe *erythema multiforme major = Toxic Epidermal Necrolysis (TEN)*

Acute inflammatory vesiculobullous reaction = *Erythema Multiforme*

- Erythema mutliforme limited to skin = *Erythema Multiforme Minor*

- Erythema mutliforme involving skin and mucous membranes = *Erythema Mutliforme Major*, aka *Stevens-Johnson syndrome (SJS)*

Re *Stevens-Johnson syndrome*: True, False, or Controversial?
How is severe defined here? With respect to the % of total body surface area involved

- SJS is also known as *erythema multiforme minor* - False

Acute inflammatory vesiculobullous reaction = *Erythema Multiforme*

- Erythema multiforme limited to skin = *Erythema Multiforme Minor*
- Erythema multiforme involving skin and mucous membranes = *Erythema Multiforme Major, aka Stevens-Johnson syndrome (SJS)*

Severe *erythema multiforme major* = *Toxic Epidermal Necrolysis (TEN)*

Re *Stevens-Johnson syndrome*: True, False, or Controversial?
How is severe defined here? With respect to the % of total body surface area involved

What %TBSA is involved in each?
--SJS: 
--TEN: 

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

SJS is also known as erythema multiforme minor False

Acute inflammatory vesiculobullous reaction = Erythema Multiforme

Erythema multiforme limited to skin = Erythema Multiforme Minor

Erythema multiforme involving skin and mucous membranes = Erythema Multiforme Major, aka Stevens-Johnson syndrome (SJS)

Severe erythema multiforme major = Toxic Epidermal Necrolysis (TEN)
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

---

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

- SJS is also known as *erythema multiforme minor*. False

Acute inflammatory vesiculobullous reaction = *Erythema Multiforme*

- Erythema multiforme limited to skin = *Erythema Multiforme Minor*
- Erythema multiforme involving skin and mucous membranes = *Erythema Multiforme Major*, aka *Stevens-Johnson syndrome (SJS)*

- *Severe erythema multiforme major = Toxic Epidermal Necrolysis (TEN)*
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?

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

- SJS is also known as erythema multiforme minor. False

Acute inflammatory vesiculobullous reaction = Erythema Multiforme

Erythema multiforme limited to skin = Erythema Multiforme Minor

Erythema multiforme involving skin and mucous membranes = Erythema Multiforme Major, aka Stevens-Johnson syndrome (SJS)

Severe erythema multiforme major = Toxic Epidermal Necrolysis (TEN)
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
- SJS is also known as *erythema multiforme minor* - False

Acute inflammatory vesiculobullous reaction = **Erythema Multiforme**

- Erythema multiforme limited to skin = **Erythema Multiforme Minor**
- Erythema multiforme involving skin and mucous membranes = **Erythema Multiforme Major**, aka **Stevens-Johnson syndrome (SJS)**

Severe erythema multiforme major = **Toxic Epidermal Necrolysis (TEN)**

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.
- SJS is also known as erythema multiforme minor. False

Acute inflammatory vesiculobullous reaction = **Erythema Multiforme**

Erythema mutliforme limited to skin = **Erythema Multiforme Minor**

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

Severe erythema mutliforme major = **Toxic Epidermal Necrolysis (TEN)**

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.
SJS is also known as *erythema multiforme minor* \(^{\text{major}}\) False

Incidence is higher in young people
- SJS is also known as *erythema multiforme major* \(^\text{major}\) **False**
- Incidence is higher in young people **True**
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as erythema multiforme major False
- Incidence is higher in young people True

What does ‘young people’ mean?
SJS is also known as *erythema multiforme minor* True

Incidence is higher in young people True

What does ‘young people’ mean?
Children and young adults
• SJS is also known as *erythema multiforme* **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?**
- SJS is also known as *erythema multiforme 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
SJS is also known as *erythema multiforme 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

Is there a gender predilection?
SJS is also known as erythema multiforme 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

Is there a gender predilection?
Yes, it is more common in...
- SJS is also known as *erythema multiforme 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

Is there a gender predilection?
Yes, it is more common in...females
Q

- SJS is also known as *erythema multiforme minor*  False
- Incidence is higher in **young people**  True

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

- Major

What does ‘young people’ mean?
Children and young adults

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

Is there a gender predilection?
Yes, it is more common in…females

How common is SJS?
Q/A

- SJS is also known as *erythema multiforme minor*  - False
- Incidence is higher in **young people**  - True

---

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

- What does ‘young people’ mean?  
  Children and young adults

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

- Is there a gender predilection?  
  Yes, it is more common in...females

- How common is SJS?  
  Rare; the incidence is...
- SJS is also known as *erythema multiforme 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

**Is there a gender predilection?**
Yes, it is more common in...females

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

Re *Stevens-Johnson syndrome: True, False, or Controversial?*
SJS is also known as *erythema multiforme minor*  
Incidence is higher in **young people**  

---

**What does ‘young people’ mean?**  
Children and young adults

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

**Is there a gender predilection?**  
Yes, it is more common in…females

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

**Are HIV/AIDS pts at increased risk?**
SJS is also known as *erythema multiforme 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

**Is there a gender predilection?**
Yes, it is more common in...females

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

**Are HIV/AIDS pts at increased risk?**
Yes
- SJS is also known as *erythema multiforme minor* \(^\text{major}\) 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

**Is there a gender predilection?**
Yes, it is more common in…females

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

**Are HIV/AIDS pts at increased risk?**
Yes

**Are pts with a malignancy at increased risk?**
SJS is also known as *erythema multiforme 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

**Is there a gender predilection?**
Yes, it is more common in…females

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

**Are HIV/AIDS pts at increased risk?**
Yes

**Are pts with a malignancy at increased risk?**
Yes
Q

- SJS is also known as *erythema multiforme major* ^ minor False
- Incidence is higher in young people True
- Mortality rate is about 10%
SJS is also known as *erythema multiforme minor*  \(^\text{False}\)
Incidence is higher in young people  \(^\text{True}\)
Mortality rate is about 10%  \(^\text{True}\)

Re *Stevens-Johnson syndrome*:  \(^\text{True, False, or Controversial?}\)
Q

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

- SJS is also known as erythema multiforme minor: False
- Incidence is higher in young people: True
- Mortality rate is about 10%: True

What about for in TEN?

It's significantly higher—around 30%.
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as erythema multiforme minor  **False**
- Incidence is higher in young people  **True**
- Mortality rate is about 10%  **True**

What about for in TEN?
It’s significantly higher--around 30%
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as *erythema multiforme minor* False
- Incidence is higher in young people True
- Mortality rate is about 10% True

What about for in TEN?
It’s significantly higher--around 30%

Is there a way to remember this?
**Re Stevens-Johnson syndrome: True, False, or Controversial?**

- SJS is also known as *erythema multiforme minor* **False**
- Incidence is higher in young people **True**
- **Mortality rate is about 10%** **True**

**What about for in TEN?**
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
Q

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

- SJS is also known as erythema multiforme minor False
- Incidence is higher in young people True
- Mortality rate is about 10% True

What about for in TEN?
It’s significantly higher—around 30%

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

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.)
SJS is also known as erythema multiforme minor: False
Incidence is higher in young people: True
Mortality rate is about 10%: True

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

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

Incidence is higher in young people True

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?**
SJS is also known as erythema multiforme minor. False
Incidence is higher in young people True
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
SJS is also known as *erythema multiforme minor*  False
Incidence is higher in young people True
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

Do these symptoms precede, or follow the skin findings?
SJS is also known as *erythema multiforme minor* False
Incidence is higher in young people True
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

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

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

- SJS is also known as *erythema multiforme 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
Re *Stevens-Johnson syndrome*: True, False, or Controversial?

- SJS is also known as *erythema multiforme 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
SJS is also known as *erythema multiforme* _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

*What percent of SJS cases are believed to be drug-induced?*
SJS is also known as *erythema multiforme minor*  \( ^{major} \) 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

*What percent of SJS cases are believed to be drug-induced?*

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

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

- SJS is also known as *erythema multiforme 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

---

*What percent of SJS cases are believed to be drug-induced?*

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

*How about TEN--is the percent of cases secondary to drug reaction higher, or lower?*
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as erythema multiforme major 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

What percent of SJS cases are believed to be drug-induced? At least 50 (most sources put the number closer to 80)

How about TEN—is the percent of cases 2ndry to drug reaction higher, or lower? Higher
Re *Stevens-Johnson syndrome*: True, False, or Controversial?

- SJS is also known as *erythema multiforme 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

**What percent of SJS cases are believed to be drug-induced?**
**At least** 50 (most sources put the number closer to 80)

**Which four drugs/drug classes are most commonly implicated?**
--
--
-- *(mnemonic forthcoming)*
--
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as *erythema multiforme 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

*What percent of SJS cases are believed to be drug-induced?*
*At least** 50 (most sources put the number closer to 80)*

*Which four drugs/drug classes are most commonly implicated?*
--N
--A
--S
--A
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as erythema multiforme 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

What percent of SJS cases are believed to be drug-induced? At least 50 (most sources put the number closer to 80)

Which four drugs/drug classes are most commonly implicated?
-- NSAIDs
-- Anticonvulsants
-- Sulfonamides
-- Allopurinol

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

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

- SJS is also known as erythema multiforme minor. True
- 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

What percent of SJS cases are believed to be drug-induced?
At least 50 (most sources put the number closer to 80)

Which four drugs/drug classes are most commonly implicated?
--NSAIDs
--Anticonvulsants
--Sulfonamides
--Allopurinol

What is the temporal relationship between drug use and the onset of SJS?
Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as *erythema multiforme 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

What percent of SJS cases are believed to be drug-induced?
**At least** 50 (most sources put the number closer to 80)

Which four drugs/drug classes are most commonly implicated?
- NSAIDs
- Anticonvulsants
- Sulfonamides
- Allopurinol

What is the temporal relationship between drug use and the onset of SJS?
If SJS occurs, it will commence within #, and unit of time of the start of drug use
SJS is also known as *erythema multiforme 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

---

**What percent of SJS cases are believed to be drug-induced?**

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

**Which four drugs/drug classes are most commonly implicated?**

-- *NSAIDs*

-- *Anticonvulsants*

-- *Sulfonamides*

-- *Allopurinol*

---

**What is the temporal relationship between drug use and the onset of SJS?**

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

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

- SJS is also known as *erythema multiforme 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

What percent of SJS cases are believed to be drug-induced?
At least 50 (most sources put the number closer to 80)

Which four drugs/drug classes are most commonly implicated?
--NSAIDs
--Anticonvulsants
--Sulfonamides
--Allopurinol

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

What is the temporal relationship between drug use and the onset of SJS? If SJS occurs, it will commence **within 8 weeks of the start of drug use**
- SJS is also known as *erythema multiforme 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

**What percent of SJS cases are believed to be drug-induced?**
**At least** 50 (most sources put the number closer to 80)

**Which four drugs/drug classes are most commonly implicated?**
-- NSAIDs
-- Anticonvulsants
-- Sulfonamides
-- Allopurinol

**There is a possible exception to this—what is it?**
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!

**What is the temporal relationship between drug use and the onset of SJS?**
If SJS occurs, it will commence **within 8 weeks of the start of drug use**
SJS is also known as *erythema multiforme*. \( ^{\text{major}} \) 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

What percent of SJS cases are believed to be drug-induced? At least 50 (most sources put the number closer to 80)

This is all well and good, but does it really matter whether a given drug is at fault?

If SJS occurs, it will commence within 8 weeks of the start of drug use
SJS is also known as *erythema multiforme 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

---

What percent of SJS cases are believed to be drug-induced?

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

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This is all well and good, but does it really matter whether a given drug is at fault? 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?**

If SJS occurs, it will commence within **8 weeks** of the start of drug use.
Re *Stevens-Johnson syndrome*: True, False, or Controversial?

- SJS is also known as *erythema multiforme minor* False
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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

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

- SJS is also known as *erythema multiforme 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

What is the second-most common inciting factor in SJS?
SJS is also known as *erythema multiforme 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

**What is the second-most common inciting factor in SJS?**
Infectious agents
SJS is also known as *erythema multiforme minor* False

Incidence is higher in young people  True

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

**Which infectious agent is most commonly implicated?**
Re Stevens-Johnson syndrome: True, False, or Controversial?

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- Adverse drug reaction is the most common cause True

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.
SJS is also known as *erythema multiforme minor* False

Incidence is higher in young people True

Mortality rate is about 10% True

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Q

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

- SJS is also known as *erythema multiforme 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

Q

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

- SJS is also known as *erythema multiforme major*  **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
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as *erythema multiforme major* 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
Re Stevens-Johnson syndrome: True, False, or Controversial?

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- Incidence is higher in young people True
- Mortality rate is about 10% True
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- Adverse drug reaction is the most common cause True
- Membranous/pseudomembranous conjunctivitis may develop True

What is the difference between a membrane and a pseudomembrane?
SJS is also known as *erythema multiforme maj*or  False
- Incidence is higher in young people  True
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- Systemic (non-skin/mucosal) symptoms are common  True
- Adverse drug reaction is the most common cause  True
- Membranous/pseudomembranous conjunctivitis may develop  True

*What is the difference between a membrane and a pseudomembrane?*
Removal of a membrane leaves the epithelial bed bleeding; removing a pseudomembrane does not
SJS is also known as *erythema multiforme 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**

Chronic ocular complications are usually mild
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Membranous/pseudomembranous conjunctivitis may develop  **True**
Chronic ocular complications are usually mild  **False**

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

How you should think about managing SJS

?  ?
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---

**SJS mgmt**

How you should think about managing SJS

Acute issues  Chronic sequelae
Re **Stevens-Johnson syndrome:** True, False, or Controversial?

- SJS is also known as *erythema multiforme minor* — False
- Incidence is higher in young people — True
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- Adverse drug reaction is the most common cause — True
- Membranous/pseudomembranous conjunctivitis may develop — True
- Chronic ocular complications are usually mild — False

SJS mgmt

Acute issues

Chronic sequelae

*In a nutshell, how should acute SJS be managed?*
SJS is also known as *erythema multiforme 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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Chronic ocular complications are usually mild False

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

**Acute issues**

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

**Chronic sequelae**

SJS mgmt

How you should think about managing SJS
**Q**

- SJS is also known as *erythema multiforme minor* False
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---

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

- SJS mgmt
  - Acute issues
  - Chronic sequelae

- In a nutshell, how should you think about managing SJS?
  - Like a *thermal burn*

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

- SJS is also known as **erythema multiforme minor**  False
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**SJS mgmt**

- **Acute issues**
  - In a nutshell, how should you think about managing SJS?
  - **Like a thermal burn**

- **Chronic sequelae**
  - What are the two overarching goals in thermal-burn management?
    - Maintain adequate…
    - Prevent…
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Incidence is higher in young people  True
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Systemic (non-skin/mucosal) symptoms are common True
Adverse drug reaction is the most common cause  True
Membranous/pseudomembranous conjunctivitis may develop  True
Chronic ocular complications are usually mild  **False**

**SJS mgmt**

**Acute issues**

- Like a **thermal burn**

**How you should think about managing SJS**

**Chronic sequelae**

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

--Maintain adequate...**hydration**
--Prevent...**superinfection**
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**Stevens-Johnson syndrome:**
- **True, False, or Controversial?**

**SJS mgmt**

- Acute issues
- **Chronic sequelae**

*How likely are SJS pts to develop chronic complications?*
• SJS is also known as *erythema multiforme minor* False
• Incidence is higher in young people True
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• Systemic (non-skin/mucosal) symptoms are common True
• Adverse drug reaction is the most common cause True
• Membranous/pseudomembranous conjunctivitis may develop True
• Chronic ocular complications are usually mild False

How likely are SJS pts to develop chronic complications? Quite. The rate for children is %; for adults, %.
Re **Stevens-Johnson syndrome:** True, False, or Controversial?

- SJS is also known as *erythema multiforme minor*  **False**
- Incidence is higher in young people  **True**
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- Membranous/pseudomembranous conjunctivitis may develop  **True**
- Chronic ocular complications are usually mild  **False**

---

**SJS mgmt**

- Acute issues
- Chronic sequelae

*How likely are SJS pts to develop chronic complications?*

Quite. The rate for children is ~25%; for adults, ~50%.
Re Stevens-Johnson syndrome: True, False, or Controversial?

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- Incidence is higher in young people  True
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What are some of the chronic ocular complications of SJS?
- Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- ...
Re Stevens-Johnson syndrome: True, False, or Controversial?

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What are some of the chronic ocular complications of SJS?
- Symblephara formation
- Entropion
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- Corneal neovascularization
- Lid margin keratinization

SJS mgmt

Chronic sequelae

How you should think
- SJS is also known as *erythema multiforme minor* \(^\text{False}\)
- Incidence is higher in young people \(^\text{True}\)
- Mortality rate is about 10% \(^\text{True}\)
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- Chronic ocular complications are usually mild \(^\text{False}\)

### Stevens-Johnson syndrome: True, False, or Controversial?

- What are symblephara?
- Acute issues
- Chronic sequelae

### Chronic sequelae

- Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization

### What are symblephara?

Adhesions between conjunctival surfaces

- 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

### Extensive symblephara can lead to what dreaded complication?

A 'frozen' globe
Re *Stevens-Johnson syndrome*: True, False, or Controversial?

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What are *symblephara*?
Adhesions between conjunctival surfaces

What are some of the chronic ocular complications of SJS?
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Stevens-Johnson syndrome:

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Adhesions between conjunctival surfaces

Adhesions between which two conj surfaces are involved?

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Chronic sequelae

How you should think

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

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**Adhesions between which two conj surfaces are involved?**
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**What are some of the chronic ocular complications of SJS?**
-- *Symblephara formation*
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--- **Symblephara formation**
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--- Ectropion
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**True, False, or Controversial?**

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A 'frozen' globe

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**Major**

- Systemic (non-skin/mucosal) symptoms are common True
- Adverse drug reaction is the most common cause True
- Membranous/pseudomembranous conjunctivitis may develop True
- Chronic ocular complications are usually mild False

**Acute issues**

**Chronic sequelae**

What are symblephara?
Adhesions between conjunctival surfaces

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**You should think**
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Should the ophthalmologist surveil SJS pts for the onset of symblephara, and lyse them prophylactically?

Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization

Chronic sequelae
A

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Should the ophthalmologist surveil SJS pts for the onset of symblephara, and lyse them prophylactically?
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.

- Symblephara formation
  --Entropion
  --Ectropion
  --Trichiasis
  --Corneal neovascularization
  --Lid margin keratinization

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

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Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization

Chronic sequelae

Under what circumstances would symblephara necessitate surgical redress?
Stevens-Johnson syndrome:

- True, False, or Controversial?

- SJS is also known as erythema multiforme minor: **False**
- Incidence is higher in young people: **True**
- Mortality rate is about 10%: **True**
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Should the ophthalmologist surveil SJS pts for the onset of symblephara, and lyse them prophylactically?
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.

---Symblephara formation---
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization

**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

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

- SJS is also known as erythema multiforme minor False
- Incidence is higher in young people True

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

Extensive symblephara can lead to what dreaded complication?
A ‘frozen’ globe

What non-SJS conditions are associated with symblepharon formation?

--Symblephara formation
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--Corneal neovascularization
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Extensive symblephara can lead to what dreaded complication? A ‘frozen’ globe

What non-SJS conditions are associated with symblepharon formation? The one that should come to mind first is ocular cicatricial pemphigoid. Others include atopic keratoconjunctivitis (AKC), adenoviral conjunctivitis (especially epidemic keratoconjunctivitis, EKC), and trachoma. (There are a number of other causes to boot.)

What are some of the chronic sequelae?
- Symblephara formation
- Entropion
- Ectropion
- Trichiasis
- Corneal neovascularization
- Lid margin keratinization
SJS is also known as *erythema multiforme 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
Chronic ocular complications are usually mild False

**SJS mgmt**

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

**Chronic sequelae**

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**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**

- Symblephara
- Entropion
- Ectropion
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- Lid margin keratinization
Q

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- Systemic steroids have a role in treatment
Re Stevens-Johnson syndrome: True, False, or Controversial?

- SJS is also known as *erythema multiforme minor* \(^\text{major}\) False
- Incidence is higher in young people \(^\text{severe}\) True
- Mortality rate is about 10% \(^\text{severe}\) True
- Systemic (non-skin/mucosal) symptoms are common \(^\text{severe}\) True
- Adverse drug reaction is the most common cause \(^\text{severe}\) True
- Membranous/pseudomembranous conjunctivitis may develop \(^\text{severe}\) True
- Chronic ocular complications are usually mild \(^\text{severe}\) False
- Systemic steroids have a role in treatment \(^\text{major}\) Controversial
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Why is the use of systemic steroids controversial?

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

(Recall this box…)
Q

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Systemic steroids have a role in treatment  Controversial
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Immunosuppressive drugs have a role in treatment  True
Supportive therapy is the mainstay of acute treatment
SJS is also known as *erythema multiforme major*  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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Systemic steroids have a role in treatment  Controversial
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Immunosuppressive drugs have a role in treatment  True
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SJS is also known as *erythema multiforme 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**

Chronic ocular complications are usually mild **False**

Systemic steroids have a role in treatment **Controversial**

Topical steroids have a role in treatment **Controversial**

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*Early* amniotic-membrane grafting shows promise as a therapy
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**In early amniotic-membrane grafting, which surfaces are grafted?**
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- *Early* amniotic-membrane grafting shows promise as a therapy True

In early amniotic-membrane grafting, which surfaces are grafted? The entire ocular surface (note that this includes the lid margins as well)