

Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis



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Case presentation

- Ms NB, a 28 year old woman
- HIV +ve on ART; 18/40 pregnant
- Presented to GSH casualty with:
 - 5 day history of burning eyes
 - 2 day history of a generalised itchy rash with mucosal ulceration

Rash decision?

- On ART since February 2008:
 - stavudine
 - lamivudine
 - efavirenz
- Efavirenz changed to **nevirapine** 4 weeks before presentation (because of pregnancy)

On examination

- Good general condition, BP 117/68 mmHg, HR 91, T° 36.1°C
- Skin: scattered papules – erythematous with central necrosis (predominantly on back and arms); Nikolsky's sign negative
- Mucosa: lips swollen and ulcerated; slough with some bleeding
- Eyes: red sclera; no discharge
- Other systems: NAD

Special investigations

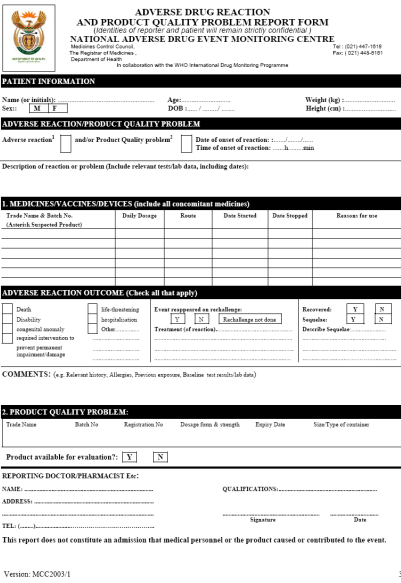
- Liver function tests
- Renal function tests
- Blood culture
- Pus swab

- No abnormalities in her case

Management

- Nevirapine stopped (changed to lopinavir/ritonavir)
- Eyes: artificial tears & lubricating ointment 2 hourly
- Mouth: glycothymol mouthwash 4 hourly
soft paraffin gauze with emulsifying ointment to lips 2 hourly
- Skin: emulsifying ointment

- Discharged after 5 days



ADVERSE DRUG REACTION AND PRODUCT QUALITY PROBLEM REPORT FORM

PATIENT INFORMATION

Name (or initials): Age: Weight (kg):
 Sex: DOB: Height (cm):

ADVERSE REACTION/PRODUCT QUALITY PROBLEM

Adverse reaction? and/or Product Quality problem? Date of onset of reaction:
 Time of onset of reaction: h min

Description of reaction or problem (include relevant test/lab data, including dates):

1. MEDICINES/VACCINES/DEVICES (include all concomitant medicines)

Trade Name & Brand Name	Strength	Route	Dose Interval	Dose Shipped	Reason for use
(Insert Suspended Product)					

ADVERSE REACTION OUTCOME (Check all that apply)

Death Life-threatening Case managed as rechallenge Referred: Y N
 Disability Hospitalisation Exchange not done Supplier: Y N
 Hospitalisation Other: Treatment of reaction: Device supplier: Y N
 Required intervention to prevent permanent impairment/damage:

COMMENTS: (e.g. Relevant history, Allergies, Previous exposure, BioRxn test results/lab data)

2. PRODUCT QUALITY PROBLEM:

Trade Name: Batch No: Expiry Date: Storage Area & strength: Expiry Date: Year Type of container:

Product available for evaluation?: Y N


REPORTING DOCTOR/PHARMACIST Etc:

NAME: QUALIFICATIONS:
 ADDRESS: Signature: Date:
 TEL:

This report does not constitute an admission that medical personnel or the product caused or contributed to the event.

Version: MCC2003/1 3

- Adverse Drug Reaction form
- MedicAlert bracelet



Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN)

- Life-threatening mucocutaneous diseases
 - Epidermal necrosis
 - Mucosal involvement
 - ± Systemic involvement (elevated liver enzymes)
- Usually drug-related

	SJS	SJS-TEN overlap	TEN
BSA* involved	<10%	10-30%	>30%
Mortality	1-3%		~30%

*BSA: body surface area

Common causes

- Antiretrovirals (nevirapine)
- Anticonvulsants
- Sulphonamide antibiotics (cotrimoxazole)
- Non-steroidal anti-inflammatory drugs
- Antituberculosis drugs
- Allopurinol

Risk factors

- Immunocompromise: especially **HIV infection**

Pathogenesis

- Thought to be a Type IV delayed hypersensitivity reaction
- T-cell mediated: CD4/CD8

Diagnosis

- Clinical features:
 - Prodromal phase: malaise, fever, stinging eyes
 - Purpuric rash → blistering and epidermal detachment
 - Mucosa involved: buccal, ocular and genital erosions
- Identify cause
- Exclude differential diagnoses

Management

- **Stop all drugs**
- Supportive treatment
- Drugs: analgesia

Supportive management

- Refer to highest level of care available
- Fluids: preferably oral
- Skin: daily baths; sterile non-adhesive dressings
- Mouth: mouthwash; lubricating cream; paraffin gauze
- Eye care: lubricant drops/ointment 1-2 hourly; ophthalmology referral if possible
- Nutrition
- Monitor vital signs and input/output

Drugs

- Analgesia: opioids
- Systemic steroids: avoid
- IVIg: no conclusive evidence of efficacy

- ADR form
www.mccza.com
- MedicAlert bracelet

Complications

- Eyes: corneal scarring
- GIT: adhesions/stenosis
- Genital: strictures
- Nails: dystrophy

Re-challenge?

- Don't do it!
- Seek advice:
National HIV HCW Hotline
0800 212 506
Medicines Information Centre, UCT
021 406 6782
- Refer if no alternative drugs available
- Remember cross-reactions

In a nutshell

- SJS/TEN are life-threatening conditions
- Early recognition important
- Stop all drugs
- Supportive management (at specialist centre if possible)
- Seek advice

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