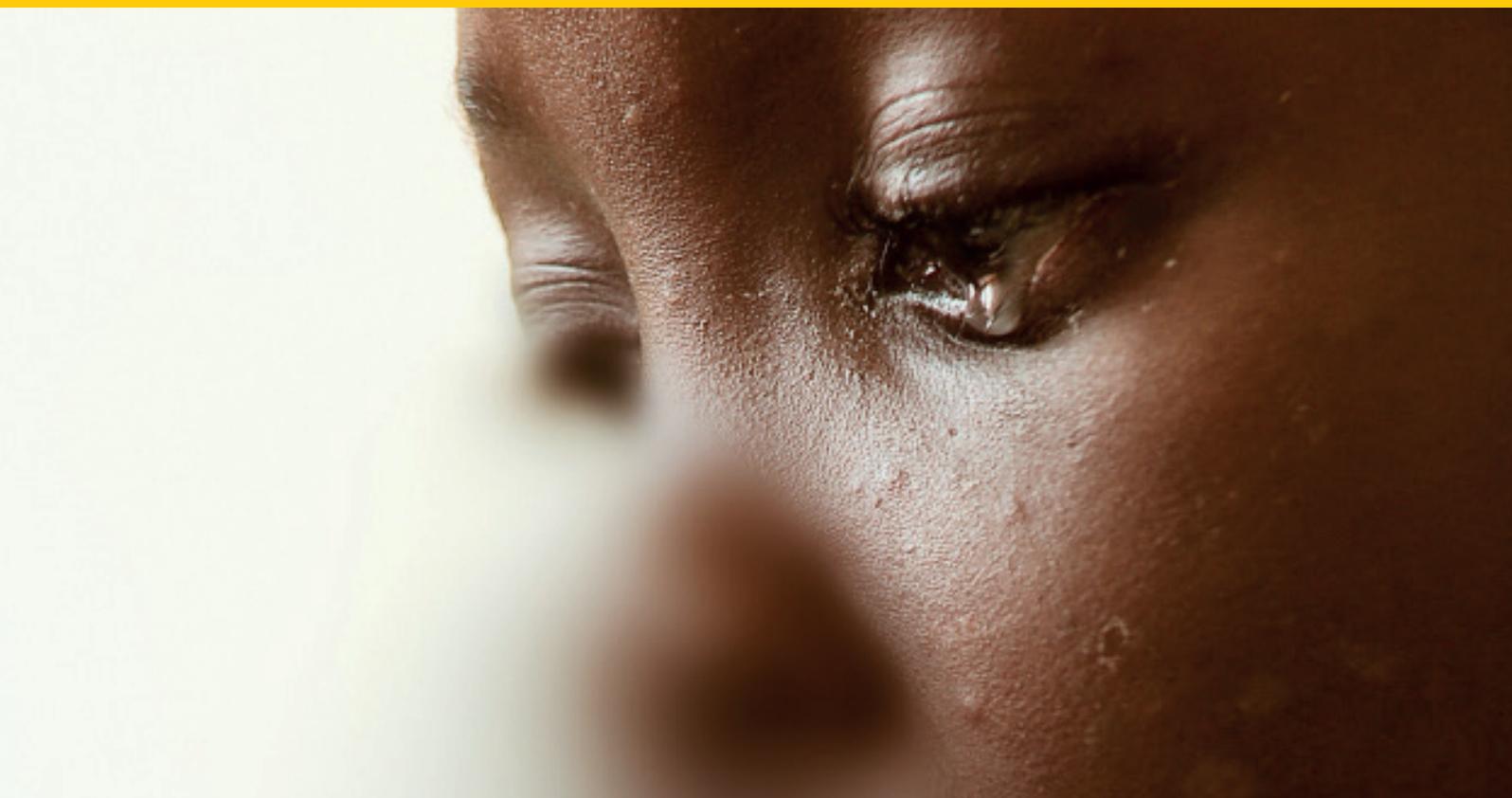


HIV Paediatric case 9 – Shukran



This is the story of 8 year old Shukran. He is an only child who now lives with his father. He's come into hospital with a rash. The skin is blistering and peeling off. The rash is all over his body, but it is particularly bad on his mouth, eyes, and genitals.....

STOP POINT

WHAT DOES THIS LOOK LIKE?

People should be able to recognise that this is Stevens Johnston Syndrome.(SJS). It is very distinctive. So the first question to ask would be what treatment is he on. Note that his tongue is very white - which also looks like oral candidiasis.

HISTORY:



Shukran's parents have been separated since he was born. His mother was not tested for HIV during her pregnancy. Indeed it seems that she never found out her status. Just over a year ago, when Shukran was seven, she became suddenly ill and died. Shukran's father on the other hand, has known he is HIV positive for more than ten years. Shukran's first recorded height and weight was when he was almost six and a half, before his mother died. He was already very thin, with a BMI right at the bottom of the WHO percentile chart. This is the same reading plotted on an alternative z score BMI chart with a greater range which categorizes Shukran as severely thin. Over the next 18 months, there was no improvement. It was only when he was eight that his father, concerned about his weight, took him to a health centre where he was finally given an HIV test. That was five weeks ago. The test came out positive.

STOP POINT

WHAT MISTAKES HAVE BEEN MADE? WITH AVAILABLE INFORMATION, IN WHICH WHO STAGE WOULD YOU JUDGE SHUKRAN TO BE? WOULD YOU HAVE STARTED ART THEN?

1) Shukran should have been tested earlier,. Height and weight should have been measured through childhood. He would certainly have been picked up before. The loss of one or both parents should also prompt people to think about HIV.

2) WHO Stage III

3) Yes. WHO Stage III – unexplained wasting is a reason for initiation on ART. Also for his age, Shukran can start ART.



Shukran was started straight away on co-trimoxazole prophylaxis. Then two weeks later he was initiated on ART. He was given a starter pack with a two week supply of medicines. Unfortunately, this was not recorded in his health passport. The health passport does show that two weeks after initiation, he went back to the health centre to get more medicines. At the review he complained of having sore, itchy eyes and eye discharge.

STOP POINT

Shukran Age: 8

5 WEEKS AGO: INITIATED CO-TRIMOXAZOLE

3 WEEKS AGO: INITIATED ART (AZT + 3TC + NVP)

1 WEEK AGO: PRESENTED AT HEALTH CENTRE WITH SORE, ITCHY EYES AND EYE DISCHARGE.

WOULD YOU GIVE HIM A REFILL? WHAT WOULD YOU DO?

1) This could be the start of a drug reaction. Stevens-Johnson syndrome normally affects the mucus membranes first. Also liver often affected. Look for the start of a rash...or skin becoming wrinkly....

2) Refer to hospital straight away. This is a very serious condition which needs specialised care. Stop ARVs and co-trimoxazole.



Fortunately Shukran was not given a refill. Rather he was referred to the hospital where has now arrived. Unfortunately the father did not realise the urgency. So it has taken a week for him to be brought here.

STOP POINT

WHAT WOULD YOU LOOK FOR IN A PHYSICAL EXAMINATION? WHAT TESTS WOULD YOU ORDER?

1) The mucus membranes are particularly affected (eyes, mouth and genitals. SJS also often affects the liver; so signs of jaundice or liver swelling. Symptoms are: Diffuse rash, fever, conjunctivitis, erythematous lesions in mouth and GU area. Also he appears to have candida. Look for other opportunistic infections

which could complicate his treatment and recovery.

2) CD4 would be useful as he did not have a baseline cd4 done. And it would give some idea as to how susceptible to infection he is likely to be.



By now the sore eyes and mouth are much worse and a rash has developed all over his body. The skin has blistered and is in some places peeling, leaving sores which are open to infection. In a few places the sores have already healed with new skin forming. He complains that it is painful to urinate because of sores on his penis. Shukran also has a nasty cough and his breathing appears slightly laboured. He has a fever.

The father says he has been having fevers and night sweats on and off for a month. He tests positive for malaria.

STOP POINT

- **Temperature: 38.2**
- **Malaria +ve**
- **Oral sores**
- **FBC - Hb: 9,4g/dl**
- **Coughing since a week ago**
- **Fevers for the last month**

WHAT WOULD BE YOUR DIFFERENTIAL DIAGNOSIS?

HOW WOULD BE YOUR TREATMENT AND MANAGEMENT?

1) Stevens-Johnson Syndrome. Most often this is a drug reaction, but in about 25% cases it happens to children not on medication. NVP or Co-trimoxazole can give this kind of reaction. But Efavirenz (EFV) and a number of anti-biotics also cause SJS. It is not a common reaction. Indeed it is quite rare. But when it happens it is life threatening.

- He has Malaria.
- His cough could be explained by the SJS or a chest infection. But he has signs of TB. So TB needs to be ruled out. But it is more urgent to treat him for the SJS and malaria.

2) Make sure NVP + other ARVs have been stopped immediately!!!

- Stop CPT
- Admit and isolate
- Treat skin as burns , daily clean wounds, apply Gentian Violet, avoid secondary infection, cover child in sterile linen, replace lost fluids (Parkland formula => 4cc x Kg x BodySurfaceArea; 1/2 over 1st 8 hours; 1/2 over last 16 hours)
- Antihistamine, antibiotics, Paracetamol
- Treat for Malaria with Artesimate or quinine.
- Eye - tetracycline eye ointment
- Oral sores - Nystatin
- High protein diet



By co-incidence, Shukran is not the only child to have come in with Stevens-Johnson syndrome. Indeed three children have come in at the same time. This is Janie, she has skin peeling off on a number of places on her body. This is very typical of Stevens Johnson syndrome. The skin first gets a strange rash with lines and



small folds, where it almost looks as if it has grown a size too big for the body. After that it starts to peel off. The areas where this happens it looks almost like severe burns. Once again Janie has been particularly affected around the mouth.

The third child is 4 year old Chikonde. She is in the worst condition. A very large area of skin has already peeled off, leaving oozing open wounds. She is in a critical condition and is in a great deal of pain. Again the mouth and the eyes are badly affected. Unlike Shukran both of the girls test HIV negative. And their guardians both say they were not on any medication when the rash started. Steven's Johnson's syndrome is a severe immune reaction which is normally provoked by medicines. It is not just Nevirapine. Co-trimoxazole and some other anti-biotics can also cause it. And it can also be triggered by other things. In the case of these two girls, who are both HIV negative, it is not possible to find out the cause,

For all three children the most important first step for SJS is to stop all medication or other possible cause. Fortunately Shukran's ARVs were stopped at the health centre a week ago. After that successful treatment depends of good nursing care.....

INTERVIEW: The nursing care is so important because as the condition of the skin is open, it is not closed as the normal skin, so when you see that condition it is good to isolate the person from other conditions, and the environment should be clean.

Shukran and the two girls were isolated in a room which was first scrubbed from top to bottom with disinfectant. All linen was sterilised.

INTERVIEW: We are cleaning the room each and every day with chlorine and even changing the linens.....we are changing them each and every day..

The children were treated as if they had severe burns, where large areas of skin are also lost. Initially they were given fluids – using the same calculation used for burns.

INTERVIEW: I was also monitoring the vital signs to the kids, because the condition can also complicate to other conditions. So I was doing the vital signs two hourly, especially temperature, because the fever can come out if there is an infection.

The eyes and the mouth, need to be kept moist. The eyes in particular must be treated with an anti-biotic ointment and must not be allowed to dry out. Otherwise complications can damage the eyes and even lead to blindness. Finally the children were put onto a high protein diet to try to help the healing process....in this case lots of milk and eggs. They were also given strong pain killers.

INTERVIEW: The lips and the eyes also are so painful. So for the pain management they were prescribed with morphine – or any analgesic – because the wounds are so painful.

7 DAYS LATER:

It is a week later, and the careful nursing has paid off. The two girls are also now much better. Janie is hardly recognizable. On her body, many of the wounds are no longer there. A week ago this was an open sore. Now it has healed and most of the skin has grown back. Elsewhere the dead layer of skin has dried up and is peeling off in sheets. But new skin has already grown to replace it underneath. Her lips have almost completely healed when compared to a week ago – and her eyes also look so much better.

Chikonde still has some way to go in comparison. She is still in a lot of pain, and the massive wound on her



back is still extensive. But it is not infected and you can see the raw patches starting to form new skin and heal. As long as the good nursing care continues and she avoids infection, she is past the danger point. The change is most obvious in her face, especially the eyes and lips which are healing well. This is what it looked like a week ago. And once again this is today.

And finally Shukran. After a course of anti-malarials, he no longer has a fever. The skin rash has also improved a great deal. All the open sores and blisters which were on the trunk of his body have now healed up. He still has lesions on his legs, but they look like patches of dry skin which will soon peel off. On his arms this has already happened. It no longer hurts for him to urinate. And the sores on his lips have dried up. However he still has ulcers in his mouth. More worrying Shukran still has some problem with his eyes. They look sore and are still watering. Long term eye problems are one of the main complications of Steven's Johnson syndrome which is why constant application of ointment is so important.

Shukran is given a nutritional assessment. A month ago, he was already very thin, with of BMI just above 12. Since then his weight has gone down by almost a kilo. And his BMI has dropped to 11.5. On admission Shukran was coughing badly and had had recurrent fevers. So TB needs to be investigated. On the other hand all these symptoms could be explained by the malaria and Stevens Johnson syndrome – which often causes a serious cough. A week later his cough is quite a bit better. A chest x-ray is ordered.

STOP POINT

SHUKRAN AGE 8 ON ADMISSION

- Coughing badly
- Night sweats
- Repeated fevers
- Prolonged weight loss

SEVEN DAYS LATER

- Mild cough
- No fever

WOULD YOU TREAT FOR TB?

WHAT WOULD YOU DO ABOUT HIS ART?

WOULD A CD4 OR VIRAL LOAD BE USEFUL?

1) No the x-ray looks quite clear and the cough has almost cleared up. The other symptoms, weight loss and fevers could be attributed to the malaria and SJS.

2) NVP is the likely cause of SJS. He needs to recover before starting a different regimen without NVP. Patients who developed severe toxicity to any specific ARV (eg Stevens-Johnson Syndrome or Hepatitis from NVP or EFV, severe Anaemia from AZT,...) must NEVER AGAIN be given a regimen containing the responsible ARVs.

3) A viral load will show high levels of the virus – only telling us what we already know. He has only just started ART – and stopped again almost immediately because of the SJS. A CD4 count could be useful in setting a baseline – and also seeing how damaged his immune system is – and how prone to opportunistic infections he is likely to be.



The chest x-ray is judged to be non-suspect of TB – so Shukran is not started on TB treatment.

TWO WEEKS AFTER ADMISSION:

Shukran is now being discharged. A referral letter is being sent to the health centre for him to be re-initiated on an alternative first line ART regimen once the rash has completely resolved. He is told he can start Co-trimoxazole again because he had been treated with this previously without side effects. He is given eye ointment and drops to continue treatment until the eyes are completely healed. The importance of keeping the eyes moist is stressed.....

6 MONTHS LATER...

6 months later Shukran has come for a follow up visit to the hospital.

STOP POINT

WHAT WOULD YOU DO TODAY?

- Check weight
- Adherence
- Disclosure done?
- ART side effects
- Review at eye clinic



He looks much better. His skin doesn't show scars but there are hyperpigmented spots in a few places. His legs have completely healed. He no longer has any sores on his mouth or lips. Stevens-Johnson syndrome is a life-threatening reaction. But once the acute phase has passed the skin rash does heal up almost completely. Over the last five months since starting ART Shukran has put on more than two kilos. While he is still small for his age, he is noticeably less thin.

However Shukran's eyes are sore. This is a very common long term complication. At an eye clinic a diagnosis of symblepharon is made. This is where the layer of eyelid, closest to the eye, and the eyeball stick together. Because of the constant discomfort, Shukran's eyes are watering permanently. This may have happened because the eyedrops were not continued for enough time after the rash on the rest of his body cleared up. The good news is that it has not affected his vision and it is possible to treat with an operation on the eye, so Shukran is referred to an eye centre where this can be done.

Finally Shukran's ARVs need to be checked. The last time he went to pick up medicines at the health centre was two weeks ago. But once again this was not noted in his health passport. So his father is asked to show the pills that he was given.

STOP POINT

WHAT WOULD YOU DO?

- He had to stop NVP because of the SJS and should never again be given!
- Stop current ARTs and start the ART which was ordered before: AZT/3TC+EFV
- Make a CLEAR note in Shukran's health passport that NVP should NOT BE GIVEN ANY MORE!!!



It turns out that two weeks ago Shukran was put back onto the old regimen containing Nevirapine because they had run out of alternative first line at the health centre. So today he is switched back to the regimen not containing Nevirapine. It was again written in his health passport that he must never be given Nevirapine because he had a life threatening reaction. The father is given counselling on the danger of Nevirapine for Shukran as well as adherence. They are told that if there are no medicines at the health centre he can come to the hospital clinic to pick them up in future.