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Management of Stroke in Sickle Cell Disease

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Outline of talk

- Definitions
- Epidemiology
- Pathophysiology
- Treatment
- Prevention
- Stroke Guidelines
- Conclusion

Definitions

- Stroke – ‘a neurological deficit of cerebro-vascular cause that persists beyond 24hours or is interrupted by death within 24hours’. (WHO)

- TIA – is the same neurological process with resolution within 24hours. (WHO)

- Stroke-like-episode: is a focal neurological deficit lasting more than 24hours for which a vascular component is unproven but can not be excluded. (WHO)

What is Sickle cell disease

- Sickle cell disease (SCD) is a group of inherited disorders of red blood cells (RBC) characterized by anemia, vaso-occlusive complications, chronic organ damage, and reduced survival
- SCD is caused by the predominant presence in RBC of sickle hemoglobin (Hb S), instead of the regular Hb A
- Hb S results from a mutation in the beta-globin gene:

  Glutamine (GAG) → Valine (GTG)

Epidemiology of Sickle cell disease in Uganda

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Region	Normal	Variant	Trait	Trait (%)	Disease	Disease (%)	TOTAL
Central 1	12187	30	1802	12.80	64	0.45	14083
Central 2	9048	31	1486	13.95	85	0.80	10650
East Central	4911	42	1201	19.24	89	1.43	6243
Kampala	11006	40	1725	13.42	81	0.63	12852
Mid Eastern	3835	35	721	15.52	55	1.18	4646
Mid Northern	9681	121	2359	19.16	153	1.24	12314
Mid Western	10897	31	1358	11.00	59	0.48	12345
North East	3517	30	651	15.36	40	0.94	4238
South Western	12440	16	586	4.48	25	0.19	13067
West Nile	2421	50	394	13.69	13	0.45	2878
Total	79943	426	12283	13.16	664	0.71	**93316**

Epidemiology of Stroke in Sickle Cell Disease

- Worldwide,10% of patients will have a clinical stroke by age 20 (Ohene-Frempong et al 1998)
- In Sub Saharan Africa, prevalence varies between 2.9% to 16.9% (Marks et al, 2018)
- In Uganda, prevalence is 6.8% (Munube et al, 2016)

Types of stroke

- Hemorrhagic stroke is the result of bleeding from a ruptured cerebral artery

- Ischemic stroke is more frequently caused
  - by arterial occlusion
  - by venous occlusion of cerebral veins and sinuses
- Ischemic stroke results from different mechanisms:
  - Embolism
  - Thrombosis
  - Diminished systemic perfusion

Pathophysiology of Stroke in Sickle Cell Disease

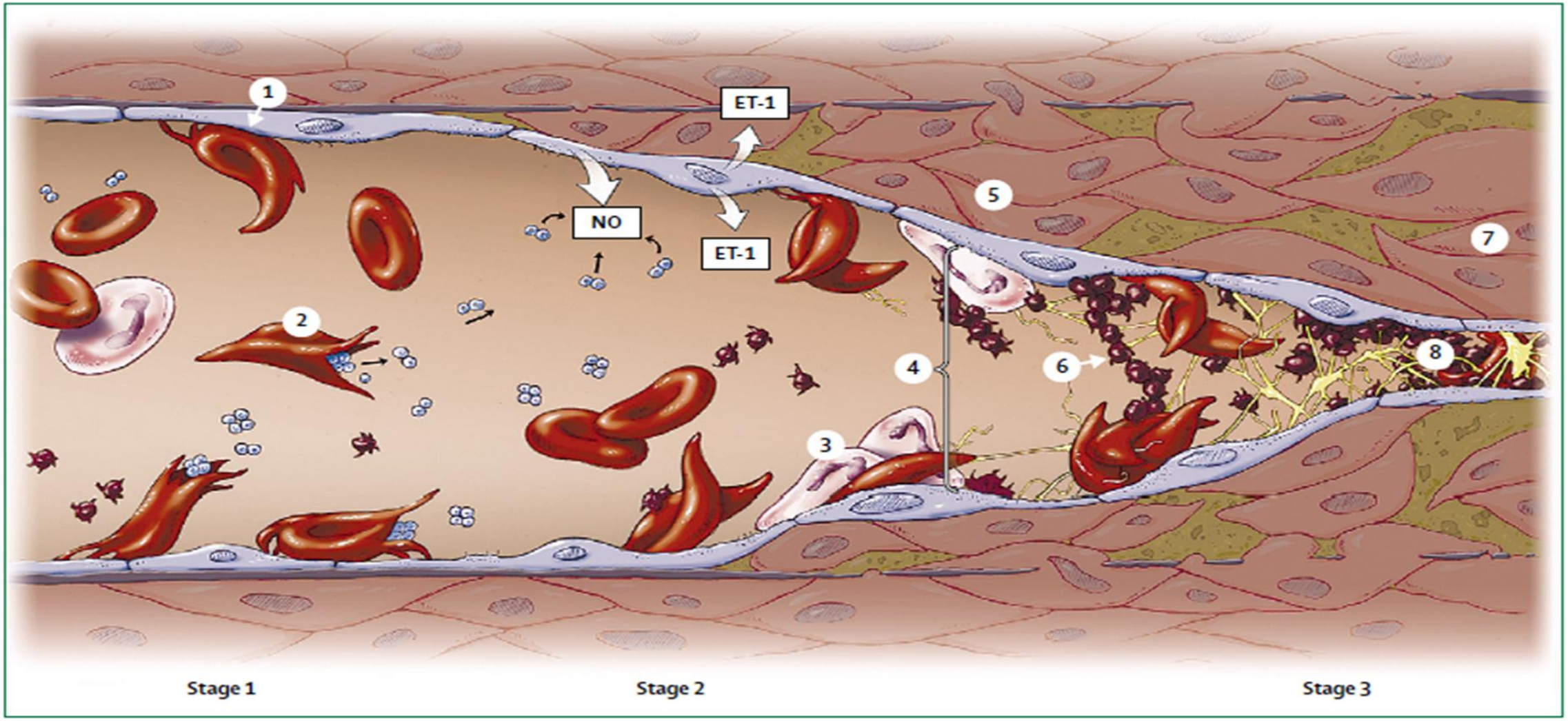

Figure 4: Vasculopathy and stroke in sickle-cell disease

Clinical Features of stroke in SCD

- Headaches
- Seizures
- Visual loss
- Prior fever/ History of fever
- Altered mental state
- Weakness of one side of the body (upper and lower limbs)
- Reduced level of consciousness
- Coma
- Treatment for Acute Chest Syndrome
- Acute Anaemia
- History of surgery
- Previous history of Transient ischaemic attack

Diagnosis of stroke in SCD

- History of SCD
- Symptoms and signs of an acute neurological event

Investigation

- Need to have urgent Complete blood count (CBC) – Haemoglobin level
- Screen for underlying triggers- ACS etc
- Brain imaging – CT Scan , CT – Angiogram , MRI/ MRA
- Haemoglobin Electrophoresis (HbS and HbF levels)
- Other tests based on the clinical presentation

Treatment of stroke in SCD

- Assess Airway (A), Breathing (B), Circulation (C) .
- Give Oxygen – avert hypoxia
- Blood transfusion – simple or exchange (if possible)
- Treat other underlying triggers e.g. Acute Chest Syndrome
- Initiate on Hydroxyurea or escalate dose to achieve MTD (maximum tolerated dose)
- Plan for chronic transfusion (continued monthly transfusions)
- Stroke rehabilitation: Physiotherapy, Occupational Therapy, Speech and Language Therapy etc

Prevention of Stroke in SCD

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Primary Prevention

- Transcranial Doppler screening
  - Screening to begin from 2 years
  - Done yearly up to 19 years
  - Identifies at risk patients

Secondary Prevention

- Chronic blood transfusion
  - Monthly
  - Monitor iron levels
  - Monitor HbF levels
- Hydroxyurea
  - Daily for life
  - Monitor adherence
  - Clinical response

American Society of Haematology 2020 guidelines for prevention, diagnosis and treatment of Cerebrovascular disease in SCD

- 1. Use of transcranial Doppler ultrasound screening
- 2. Use of hydroxyurea
- 3. Surveillance for developmental delay, cognitive impairment and neurodevelopmental disorders in children

Sickle Cell Pan African Consortium (SPARCO) – Standards of Care for Stroke

- Stroke screening using Transcranial Doppler ultrasound for children at risk for stroke
- Initiating hydroxyurea for children from 9 months of age
- Acute stroke management – immediate blood transfusion

Uganda Clinical Guidelines 2022- Stroke in Sickle Cell Disease

- Care at the level of Regional Referral Hospital
- Provide oxygen therapy
- Blood transfusion
- Fluid maintenance
- Refer for advanced care

National Guidelines for the Management and Prevention of Sickle Cell Disease 2020

- Acute Stroke
  - General principles of care
  - Guidance by paediatrician or physician
  - Urgent blood transfusion

- Prevention of stroke
  - Initiate hydroxyurea
  - Transcranial Doppler screening
  - Chronic blood transfusion

Conclusion

- Sickle cell disease is a common cause of stroke in children
- Acute management of stroke requires immediate blood transfusion
- Chronic management requires monthly blood transfusions
- Initiation of hydroxyurea can present occurrence of both primary and secondary stroke
- Transcranial Doppler ultrasound screening is a key modality to detect risk for stroke

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Thank you for listening

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Acknowledgements

SPAN Sickle Pan African Network

SPARCO Sickle Pan African Research Consortium

SADaCC Sickle Africa Data Coordinating Center

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