SPARCo STANDARDS OF CARE FOR SICKLE CELL DISEASE IN SUB-SAHARAN AFRICA

HOME BASED VERSION

September,2023

SPARCo
Sickle African
Research Consortium

SADaCC
Sickle Africa Data
Coordinating Centre

FOREWORD

Sickle cell disease (SCD) is the commonest clinically significant haemoglobinopathy. It was first reported in literature in November 1910 by James Herrick when he saw “peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia”. (1) The sickling of RBCs contributes to Vaso-occlusion and intravascular hemolysis which leads to the symptomatology these patients present with. (2) They are at increased risk of coming with several complications which can lead to mortality if the appropriate management schedule is not instituted. The absence of a structured clinical management guideline can lead to lots of morbidity and mortality for the patient with SCD.

This clinical management of Sickle Cell Disease (SCD) guidelines were developed by a standard of Care Working group composed of healthcare personnel with expertise in paediatric and adult haematology and clinical psychology. This initiative was started after the establishment of a “Sickle Cell Disease in Sub-Saharan Africa: Collaborative Consortium after being awarded the National Heart, Lung and Blood Institute (NHLBI) of the National Institute of Health (NIH) of the United States of America grant(1U24HL135881-01). This grant was awarded to a multinational collaboration among Muhimbili University of Health and Allied Sciences (MUHAS), Dar es Salaam, Tanzania as hub and three sites; MUHAS, Dar es Salaam, Tanzania, University of Abuja, Abuja, Nigeria and The Kwame Nkrumah University of Science and Technology, Kumasi, Ghana.

Its first goal was to reduce morbidity and mortality in SCD in Africa through implementation research demonstrating the feasibility of introducing newborn screening (NBS) and providing comprehensive care to prevent, identify, treat, and manage acute and chronic complications. Specific aim 2 of the proposal was to develop, implement and evaluate a resource-based, multilevel, “Guidelines for Management of SCD in SSA”.

This work was done under the esteemed leadership of Prof Kwaku Ohene Frempong (KOF), (Sickle Cell Foundation of Ghana) who demonstrated exceptional dedication and unwavering commitment to improving the lives of SCD patients. The innovative design approach developed by the KOF and the Foundation using a multi-referral level guideline approach formed the blueprint for the creation of these guidelines, empowering healthcare professionals across the region to provide evidence-based and compassionate care to those in need. Both African and non-African SCD guidelines were used to draft locally appropriate SCD management recommendations.

The purpose of these guidelines is to guide healthcare professionals at the last referral level to be able to help people living with sickle cell disease (SCD) so that they receive appropriate care. The target audience are clinicians, nurses, and staff who provide emergency or continuity care to individuals with SCD at the last referral hospitals.

The guidelines focus on recommendations for Diagnosis of Sickle Cell Disease (SCD) and
Related Conditions, Health Maintenance and Preventive Therapy, screening for Specific
Complications of Sickle Cell Disease, Management of Acute Complications of SCD,
Management of Chronic Complications of Sickle Cell Disease and Special Management Protocols including hydroxyurea protocol, blood transfusion among a few. The recommendations address the care of infants, children, adolescents, and adults with SCD, with the goal of facilitating highquality and appropriate care for all individuals with this disease no matter where they find themselves.

As we embrace these guidelines within the SPARCo consortium, we acknowledge the immense responsibility bestowed upon us to carry forward the legacy of Professor Kwaku Ohene Frempong.

Together, we embark on this transformative journey, equipped with the wisdom of the past and the hope of the future, guided by the vision of Professor Kwaku Ohene Frempong, and committed to making a lasting impact on the lives of SCD patients in Sub-Saharan Africa.

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ACKNOWLEDGEMENT

We sincerely thank everyone who contributed significantly to the development and improvement of the guidelines. Their invaluable suggestions and insights have significantly raised the caliber and efficacy of guidelines.

For internal reviewers, despite their demanding schedules and numerous responsibilities, they generously devoted their time and effort to thoroughly review the guidelines. Their meticulous attention to detail and comprehensive feedback significantly contributed to the overall improvement of the document. We are truly indebted to their expertise and commitment to patient care.

Furthermore, we would like to express our appreciation to the principal investigators (Prof Julie Makani and Prof. Ambroise Wonkam) for their unwavering support throughout the development process. Their guidance, expertise, and encouragement were instrumental in steering the project in the right direction. Their commitment to evidence-based medicine and patient-centered care has been a driving force behind the success of this endeavor.

We also want to thank the administrative and technical staff at the SPARCO Hub in Tanzania and SADaCC, in Cape Town who were instrumental in ensuring the smooth functioning of the entire process. Their technical expertise and tireless efforts in managing data, coordinating meetings, and overseeing the logistics of the process were invaluable. Without their dedication and proficiency, these guidelines would not have been executed with such precision and efficiency.

Last but not least, we thank the National Institutes of Health for funding this project. Their generous support and belief in our vision have been instrumental in bringing this project to fruition.

Their financial support enabled us to do significant research, engage professional reviewers, and promote the guidelines to a large audience. We are deeply appreciative of their commitment to improving disease management and healthcare outcomes.

Finally, we want to underline that the successful completion of these management standards was a joint endeavor that required the dedication, knowledge, and assistance of numerous individuals. Each and every contributor, whether internal or external, played an integral role in shaping the final outcome. Their commitment to quality and patient care has made a lasting impact on the healthcare community. Thank you once again for your invaluable contributions and unwavering support.

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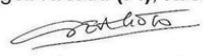

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LIST OF ACRONYMS

ACN	Acute Chest Syndrome
ASS	Acute Splenic Sequestration
AVN	Avascular Necrosis
CE	Capillary Electrophoresis
CAE	Cellulose Acetate Electrophoresis
CAGE	Citrate Agar Gel Electrophoresis
FBC	Full Blood Count
Hb	Haemoglobin
HE	Haemoglobin electrophoresis
HLPC	High Performance Liquid Chromatography
HSCT	Haematopoietic Stem Cell Transplantation
IEF	Isoelectric Focusing
IPD	Invasive Pneumococcal Disease
MSOF	Multisystem organ failure
MTD	Maximal Tolerated Dose
NHLBI	National Heart, Lung and Blood Institute
NSAIDS	Non-Steroidal Anti-Inflammatory Drugs
POC	Point-of-Care
RBC	Red Blood Cells
SCD	Sickle Cell Disease
TCD	Transcranial Doppler
TD	Therapeutic Dose
WHO	World Health Organization

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INTRODUCTION

Sickle cell disease (SCD) is the commonest clinically significant haemoglobinopathy. It was first reported in literature in November 1910 by James Herrick when he saw “peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anaemia. (1)The sickling of RBCs contributes to vaso-occlusion and intravascular hemolysis which leads to the symptoms these patients present with. (2)They are at increased risk of coming with several complications which can lead to mortality if the appropriate management schedule is not instituted. The absence of a structured clinical management guideline can lead to lots of morbidity and mortality for the patient with SCD.

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providing comprehensive care to prevent, identify, treat and manage acute and chronic complications. Specific aim 2 of the proposal was to develop, implement and evaluate a resource-based, multilevel," Guidelines for Management of SCD in SSA". The specific objectives included reviewing existing guidelines and setting minimum standards for management of SCD based on available institutional and human resources.

The purpose of these guidelines is to guide patients, parents and guardians to be able to help people living with sickle cell disease (SCD) so that they receive appropriate care.

The guidelines focus on recommendations for Diagnosis of Sickle Cell Disease (SCD) and
Related Conditions, Health Maintenance and Preventive Therapy, Screening for Specific Complications of Sickle Cell Disease, Management of Acute Complications of SCD, and Management of Chronic Complications of Sickle Cell Disease. The recommendations address the care of infants, children, adolescents, and adults with SCD, with the goal of facilitating highquality and appropriate care for all individuals with this disease no matter where they find themselves.

CHAPTER 1

DIAGNOSIS OF SICKLE CELL DISEASE (SCD) AND RELATED CONDITIONS

When doctors are trying to figure out if someone has sickle cell disease, they do tests to look at the type of hemoglobin in the blood. Hemoglobin is a part of the blood that carries oxygen. They can do this by separating the different types of hemoglobin or by looking at the DNA to confirm the diagnosis if needed.

To start, they use a method to separate the different types of hemoglobin. There are different ways to do this, like using a special gel or a machine that uses electricity. These methods help identify the type of hemoglobin someone has. However, sometimes these methods can't give a clear answer, so they might need to use a different test to be sure.

For babies younger than 6 months, they use specific methods to check their hemoglobin type, and if needed, they refer them to a special program called Newborn Screening (NBS).

If someone doesn't show any unusual types of hemoglobin in the first tests, but the doctors still think there might be a problem, they do more tests. These tests look at things like the number of red blood cells and the size of those cells. This helps them see if there's a genetic condition called thalassemia. They also look at the amounts of different types of hemoglobin using machines that can measure them. This is important, especially when thinking about genetic conditions. If a certain test isn't available, they send the person to a place where they can get that test done. Overall, these tests help doctors understand if someone has sickle cell disease or other blood related conditions, so they can provide the right treatment and advice.

CHAPTER 2

HEALTH MAINTENANCE AND PREVENTIVE THERAPY

Infection Prevention: General

Seek immediate medical attention for a person with SCD who has developed a high body temperature (fever) greater than 38.3°C or other signs of infection like poor feeding, vomiting and irritability. If a child with SCD has fever, rush the child to the nearest hospital or clinic. Do not waste time trying to bring down the high temperature.

Prevention of invasive pneumococcal disease (IPD)

If your child is diagnosed with SCD after birth, the child should be on oral penicillin V from 2 Months of age. The dose is 125 mg every 12 hours (twice daily) by mouth until age 3 years when it should increase to 250 mg twice daily, every day. Erythromycin, another antibiotic will be prescribed instead if your child is allergic to penicillin. You may stop giving penicillin when the child reaches 5 years of age. Otherwise, only stop if your child's doctor advises you to. Do not discontinue penicillin at age 5 years if your child has had a previous pneumococcal infection like pneumonia, septicemia and meningitis or has had surgical removal of the spleen. As Patients/caregivers, you should ensure that the pneumococcal vaccination schedule is completed. Seek immediate medical attention whenever fever (temperature greater than 101.3°F or 38.5°C) occurs, due to the risk for severe bacterial infections.

Prevention of malaria

Talk to your doctor/nurse about you or your child's medications to prevent malaria. Protect your Child from malaria by using Insecticide-Treated bed Net (ITN), indoor Residual spraying, and proper clothes to cover the child in the evenings and early morning to help reduce malaria infection.

Prevention of enteric gram-negative organisms (Salmonella, E. coli, Klebsiella, etc.)

Wash your hands with soap and water whenever you change your child, before eating, and after your use of the toilet. Children should be taught to do the same as they get older.

Immunization

Make sure that the person with SCD receives all the vaccination required for children and adults based on age, plus all the special vaccines that people with SCD need for extra protection from serious infection. In addition to all the regular vaccines, make sure the child or adult with SCD receives vaccination for three bacteria that are very dangerous for people with SCD: Vaccinate ALL people with SCD against pneumococcus, Hemophilus influenzae type b (Hib), and meningococcus. Always check with your doctor or nurse in your clinic to see if the child or adult with SCD needs any additional vaccines.

Nutrition

Breastfeed your child with SCD exclusively for the first 6 months of life. Start introducing other feeds in addition to breast milk from 6 months. These feeds should contain adequate proteins (e.g., Red meat, fish and chicken), starches (yam, rice, potatoes) fruits and green leafy vegetables.

Thereafter, they should eat the same foods as the rest of the family. Children with sickle cell disease do not need special food. Each day include items from the various food groups i.e. Starches (e.g., yam, potato, rice, maize); Protein (e.g., red meat, fish, chicken); fruits (e.g., mango, oranges, banana, tomato); Vegetables (e.g., cabbage, carrot, spinach); dairy products (e.g., milk, cheese); and Fats (e.g., margarine, butter). Foods such as eggs and chicken should be thoroughly cooked to prevent salmonella infections in SCD patients. Drink oral

fluids as much as can be tolerated to reduce risk of dehydration caused by passing large amounts of dilute urine.

Plain water and diluted juice is preferable, avoid fizzy drinks. Talk to your doctor/nurse if your child eats things which are non-nutritious like chalk, clay, paper and coal or you have any nutritional concerns.

Growth and Development monitoring in children

At regular intervals, your doctor will assess your child's growth to ensure he/she is growing normally. Children with sickle cell disease may be thinner and slightly shorter than children who do not have SCD. Talk to your doctor/nurse if you have any concerns about your child's growth and development.

Genetic and reproductive counseling

Attend SCD clinic for genetic and reproductive counseling.

Female reproductive health (Pregnancy, Contraception and Fertility)

Attend reproductive health care clinics for counseling on the use of contraceptives. Seek medical advice at SCD clinic before conception. Attend antenatal clinic for proper follow up during pregnancy.

Male reproductive health

Attend the closest high level or last referral hospital for counseling on male reproductive health.

Education and psychosocial counseling

Inquire about SCD care during each visit. The healthcare provider will educate all children from school going age with their parents/caregivers and adults routinely about SCD at each visit by using available materials e.g., brochures, videos, and apps. The healthcare provider will talk to patients and

parents, encourage you to talk about daily problems including finance and poverty, and will offer practical solutions.

Patients are encouraged to talk about stigma and their experience with how others relate to them. Example; Supernatural beliefs such as witchcraft and reincarnation, religion, society's attitudes due to lack of knowledge, understanding, and burden. Healthcare provider will educate parents/caregivers about importance of schooling, encourage their children to attend school when they are well and to keep a record of the number of school days lost. Adults are encouraged to find employment (paid or voluntary). If an adolescent / youth or adult SCD patient is accompanied by parent/caregiver, healthcare provider may consider to request privacy so that a patient may be free to express or ask the health care provider especially on psychosocial issue. Patients are encouraged to participate in social activities and not feel isolated. Participation in support groups where available is encouraged. Self-management and health maintenance are encouraged. Ensure understanding of the importance of adherence to medical advice and treatment.

Organizing support groups

Healthcare provider will suggest to patients and parents about setting up a support group if one is not available for them, they should also consider social media support groups. Patients and parents are encouraged to organize their own groups but consider what healthcare or other professional support is required. Help identify organizers/leaders for the group but emphasize that everyone has a role to play and can take turns, and be open to suggestions and contributions.

Consider how the group is structured e.g., parents of newborns and infants, teenagers only, group size and scope of support. Consider the time and place to meet, frequency, length, cost of venue (free or cheap) e.g., school, church, library or other public venue suitable and accessible for everyone, and available transportation. Consider how patients or parents will be encouraged to join e.g., clinic flyers, personal invitations, social media. Consider some ground rules and how to solve problems e.g., confidentiality, a person talking too much, someone who is upsetting others within the group, someone not well enough to attend and has responsibilities. Consider fundraising and/or contribution from members towards sustainable running costs, refreshments, and social activities. Consider whether to formalize the group after a while to become a registered charitable organization.

Transition of adolescents to adult care.

At a designated age usually from 16-18 years children are moved from pediatric clinic to adult clinic. Preparation usually starts earlier, and you and your child will be supported by your doctor/nurse through this period. Discuss any concerns you and your child will have with the health care team.

Travel management

Seek travel advice and accept all the offered immunizations relevant to the area to which you are travelling; this includes live vaccines like yellow fever. Receive malaria prophylaxis when travelling to malarial areas from a non-endemic country in line with general guidance.

CHAPTER 3

SCREENING FOR SPECIFIC COMPLICATIONS OF SICKLE CELL DISEASE

Screening for Stroke Risk.

Starting from 2years to 16 years of age, your child should have a scan of the head which is called a Transcranial Doppler (TCD) Ultrasound to check for the risk for stroke. Attend SCD clinic for medical advice and ask from the doctor or nurses about how to have this screening done at a facility with a TCD machine.

Screening for Kidney Disease

Starting from 5 years of age, the child should have urine test to look for too much protein. If the test is negative, it needs to be repeated only once a year. If it is abnormal, the child should have more urine and blood tests. Whenever caregivers or people with sickle cell disease go for a checkup with their doctors or nurses, they should ask about urine test for kidney disease.

Screening for Eye complication

Starting from 10 years of age, SCD patients should be screened for eye complications (retinopathy). Attend SCD clinic for medical advice and ask Your doctor or nurse about the referral to the eye clinic for the eye to be screened.

Screening for Lung disease/ complication

Talk to your doctor if you have breathing difficulties, wheezing, heavy snoring or irregular breathing during sleep. You should be referred to a sickle cell center at a Regional or Tertiary Hospital to have your lungs and breathing checked.

Screening for Heart Disease.

Attend an SCD clinic for medical advice.

Screening for Hypertension

Make sure blood pressure is measured at every routine visit in children and adults with SCD

Screening for SCD complications during pregnancy

Attend antenatal visits as scheduled once you get pregnant.

CHAPTER 4

MANAGEMENT OF ACUTE COMPLICATIONS OF SCD.

Acute Anemia

Seek immediate medical attention whenever patient with SCD has sudden onset of headache, easy fatigue, and increased awareness of heartbeats with paleness.

Acute Chest Syndrome

Seek immediate medical attention for a person with SCD who has developed difficulty in breathing, chest pain, and chest tightness with or without high temperature (fever) greater than 38.3°C or other signs of infection, cough, and wheeze. Encourage person with SCD to blow either balloons or a spirometer (Incentive spirometer).

Acute SCD pain (Vaso occlusive pain episode, VOPE, or "pain crisis").

Learn to take care of mild sickle cell pain at home. Go to the hospital or clinic when there is something else such as, fever, breathing difficulty, severe headache, vomiting happening, or if the pain is different from the usual sickle cell pain you normally have.. If you or your child is uncomfortable, pain medication and other comfort measures should be used to help reduce the pain. If the pain is mild, you may try comfort measure such as drinking fluids, local heat, massage, and other diversional therapy like video games etc, before using medication. You can use acetaminophen (Paracetamol, Tylenol, etc.) plus anti-inflammatory medications such as ibuprofen (Brufen, Calprofen, Nurofen, etc.) If pain doesn't resolve and person with SCD needs an opioid, he/she will need to be taken to the hospital. If someone is taking opioids such as morphine, check frequently to make sure that the medication is not making the person too sleepy because deep sleepiness can cause the person to stop breathing. Wake the person up to make sure he or she is alert, can talk, and stay awake.

If you cannot wake the person up easily, rush him or her to the hospital to be checked so that the sleepiness can be reversed with another medication. Encourage the person in pain to walk, if possible, sit up, do deep breathing exercises, using tools such as, incentive spirometer, balloon blowing, or equivalent methods, as soon as possible, to help prevent collapse of the lungs

Spleen complications

Seek immediate medical attention whenever patient with SCD has abdominal swelling with pain at the left upper part of the abdomen, easy fatigue, and increased awareness of heartbeats. Parents/caregivers of children with SCD should be taught how to palpate for the spleen of their children to notice any change in size and report to the hospital accordingly. Ask your doctor or nurse to show you how to palpate your child's spleen.

Acute Stroke

Seek immediate medical attention whenever patient with SCD has unilateral weakness, loss of consciousness, headaches or seizures. A painless limp might be the only symptom for a stroke and so be alert and ask your doctor or nurse about any abnormal signs.

Fever and other signs of infection

A person with SCD and a temperature of 38.3°C or higher (fever), should see a doctor or nurse right away. If your child has fever, rush the child to the hospital or clinic. Do not waste time trying to bring down the high temperature with medicines. Other signs of infection apart from fever to look out for in SCD patients include poor feeding, unusual sleepiness, and vomiting.

Bone disease

Inform your doctor or nurse about any unusual sickle cell pain is it can be an infection of the bone. Your health care provider may ask for an x-ray and other tests. You will be asked to take antibiotics for about 6 weeks if infection of the bone is confirmed.

Multisystem Organ Failure

Seek immediate attention if patient with pain exhibit deterioration of symptoms.

Priapism

Seek immediate attention if person with SCD presents with persistent painful penile erection. It can lead to impotence. You are encourage to take fluid and antipain medication such as paracetamol. You can have a warm bath or undertake some physical exercise when priapism occurs.

Acute Liver and Gallbladder Complications

Seek immediate attention if patient presents with deepening of yellowish coloration of the eyes, abdominal swellings, itching, and pain on the right side of the abdomen.

Acute Ocular Complications

Seek immediate medical attention whenever patient with SCD has sudden onset of reduced vision and/or eye pain.

Acute Renal Failure (Kidney failure)

Seek immediate medical attention whenever patient with SCD has sudden onset of reduced or no urine output despite adequate intake of fluids.

CHAPTER 5

MANAGEMENT OF CHRONIC COMPLICATIONS OF SICKLE CELL DISEASE

Bone complication

Seek immediate medical attention whenever patient with SCD has joint pain or inability to walk. Your health care provider may ask for an x-ray and other tests. Minimize movement and bearing heavy weight on affected joint. Take the prescribed antipain medication.

Heart complications

Seek immediate medical attention whenever patient with SCD has increased awareness of heartbeats, easy fatigability.

Chronic hypersplenism (Spleen complication)

Attend SCD clinic regularly as scheduled. If abdominal swelling increases, seek medical advice immediately.

Chronic Pain

Attend SCD clinic to be assessed for chronic pain annually or more often as needed. Health Care Practitioner (HCP) will use a combination of the person's response to treatment—including pain relief, side effects, and functional outcomes—to guide the long-term use of opioids. You are encouraged to use deep tissue/deep pressure massage therapy, muscle relaxation therapy, and self-hypnosis as indicated. Use long- and short-acting opioids to manage chronic pain that is not relieved by nonopioids. If the use of long-term opioids is indicated for your treatment, we will

collaborate with you to create an individualized plan. This plan will outline the potential risks, benefits, and side effects, empowering you to make informed decisions about your care.

To help Health care provider better understand your pain patterns, you are also encouraged to keep a Pain Diary. This will allow HCP to track your pain episodes more accurately and adjust your treatment plan accordingly.

Endocrine complications

Seek medical attention if patient has delayed puberty

Gastrointestinal complications

Seek medical attention if patient presents with sudden onset abdominal pain and/or tenderness, distension, vomiting, constipation, deep jaundice

Leg Ulcers

SCD patients should go to clinic for proper assessment and care of the ulcers. Adhere to medications and treatment protocol as advised by medical personnel. Elevate feet when sitting to improve blood circulation to the ulcer. Treat the pain with adequate and appropriate pain medication as given by your doctor.

Nocturnal enuresis (Bed Wetting)

Report enuresis during clinic visits if bed wetting is present in a child aged 6 years and older

Ophthalmologic Complications

Report to hospital anytime you have vision problems. Seek medical help early.

Psychological complications

When there are mood and emotional problems with inability to cope and reduced quality of life, seek medical help from your primary SCD doctor who will refer to the appropriate level of care for psychological assessment and interventions.

Pulmonary Hypertension

Seek medical attention for SCD patients present with symptoms such as shortness of breath on exertion, exertional chest pain, and awareness of heartbeats, leg swelling, and abdominal swelling

Kidney Complications

SCD patients with nocturnal enuresis should be taken to the doctor for further evaluation. Attend SCD clinics and nephrology clinic as scheduled. Adhere to medications as prescribed by healthcare providers. Do not abuse medications.

Seizures under neurological complications

SCD patients with seizure disorders should attend neurology clinic as scheduled and make sure to take anticonvulsant medications as prescribed by a doctor.

Stuttering/Recurrent Priapism

Seek immediate attention if patient presents with persistent penile erection. Encourage fluid intake and give antipain medication such as paracetamol.

CHAPTER 6

SPECIAL MANAGEMENT PROTOCOLS

Hematopoietic stem cell transplantation

Attend SCD clinic for more information on stem cell transplantation and Counselling.

Hydration guide

Patients with SCD are at risk of dehydration due to impaired renal concentrating power and poor fluid intake. Encourage intake of oral fluids first, it should be used whenever possible, and you are **encouraged liberal use of oral fluids**

Transfusion Therapy in Sickle Cell Disease/Iron chelation

Attend SCD clinic for evaluation and advice on blood transfusion

Hydroxyurea therapy in SCD

Attend SCD clinic for medical advice
Adhere to Hydroxyurea dosage as prescribed by the healthcare provider.

Peri-operative care and surgery

Follow instructions given by clinicians before and after surgery.

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