Attachment F

SICKLE CELL DISEASE CHRONIC CARE MANAGEMENT PLAN

SICKLE CELL DISEASE OVERVIEW

Sickle cell disease is an inherited disorder manifested by hemolytic anemia, unpredictable acute crisis that can rapidly become life-threatening, and development of chronic organ damage. The severity of anemia, the frequency of crisis, and the scope of organ damage are variable among sickle cell disease patients and also differ depending on the genotype of sickle cell disease present. Four genotypes account for most sickle cell disease seen in the United States. These include sickle cell anemia with hemoglobin SS (65%), sickle-hemoglobin C disease with hemoglobin SC (25%), and two types of sickle β-thallasemia (10%). Unfortunately, most afflicted students will not know which genotype they have and may get sickle cell disease confused with sickle cell trait. Sickle cell trait is a generally benign and asymptomatic genetic carrier state, and students with sickle cell trait can be managed by a care plan designed for well students.

Diagnosis
The health and wellness staff should establish the diagnosis of sickle cell disease as quickly as possible, either from the information on the student's application to Job Corps, the student's report after arrival on center, and/or very low hemoglobin on entry to Job Corps. A hemoglobin electrophoresis from the student's physician will establish the diagnosis of and genotype of sickle cell disease.

Plan
Obtaining Past Records
1. Obtain the baseline CBC and reticulocyte counts from the student's physician.
2. Obtain the most recent liver function tests, renal function tests, chest X-ray results, EKG and/or echocardiogram, pulmonary function tests, and blood pressure measurements if available. Also request information on the results of abdominal ultrasound or history of cholecystectomy.
3. Interview student and obtain medical records to determine the medical problems the student has experienced related to his or her sickle cell disease.
4. Interview student and obtain medical records to determine the medications being prescribed.
5. Interview student and obtain medical records to determine the administration of immunizations, including pneumococcal and meningococcal vaccines.

Patient Education
1. Review with the student the physician's plan for pain management on center.
2. Review with the student the center's plan for management of urgent medical evaluation and treatment of febrile illnesses.
3. Discuss with the student the availability of contraception and possibility of transmission of sickle cell disease.
4. Discuss the management of priapism with male sickle cell students.
5. Discuss the caution regarding excessive physical activity, including athletics, on center and the need to avoid temperature extremes and assure adequate hydration.
6. Discuss the importance of avoiding drugs that may precipitate or exacerbate complications of sickle cell disease, such as alcohol, tobacco, and cocaine.

Acute Illness
1. Develop a plan for round-the-clock access to a facility that can provide urgent evaluation for and treatment of acute illness characterized by fever, pallor, lethargy, abdominal distention or enlarging spleen size, tachypnea or other signs of respiratory illness, prolonged (more than 2 to 4 hours) priapism, or any neurologic sign or symptom.
2. Arrange immediate access at the acute care facility of baseline information about the student in the medical record at the center.

Monitoring/Treatment at Health and Wellness Center
1. CBC and reticulocyte count at quarterly intervals
2. Interim history and physical assessment by center physician or his or her designee at monthly intervals
3. Update of basic immunizations to include pneumococcal and meningococcal vaccines
4. Provide prophylactic penicillin V potassium 250 mg PO BID
5. Provide folic acid 1 mg PO daily
6. Provide annual influenza vaccination

Reference

"Health Supervision for Children with Sickle Cell Disease," *Pediatrics,* 109/3 (March 2002).

Name: _____

Student ID#: _____ **DOB:** _____

SICKLE CELL DISEASE CHRONIC CARE MANAGEMENT PLAN

OUTREACH AND ADMISSIONS PERIOD

Please provide us with the following information.

1. Date of diagnosis: _____
2. Age of onset: _____
3. List current medications and/or treatment including dosage and frequency prescribed.
   _____
   _____
4. Has applicant been compliant with medications and treatment? If no, please explain.
   _____
   _____
5. List past hospitalizations including dates, reason for admission, and discharge summaries.
   _____
   _____
   _____
6. What is current status and prognosis?
   _____
7. When was last appointment? _____
8. Will the applicant need to continue follow-up under your care? If yes, please list the date and/or frequency of follow-up appointments.
   _____
   _____
9. In your opinion, will the applicant be able to self-manage his or her medications unsupervised and participate in a vocational training program? If no, please explain.
   _____
   _____
10. In your opinion, will the applicant be appropriate to reside in a dormitory style residence with minimal supervision? If no, please explain.
    _____
    _____
11. Are there any restrictions or limitations related to this specific illness?
    _____
    _____
12. List any precipitants to a crisis for this applicant.
    _____
13. List any allergies this applicant may experience.
    _____

Name: _____

Student ID#: _____ **DOB:** _____

14. What is the applicant's smoking history?

    _____

15. Does the applicant use contraception?

    _____

16. Does the applicant have health insurance documentation?

    _____

Please sign below and return the form in the attached addressed envelope.

_____ _____

Print Name and Title **Signature**

_____ _____

Phone **Date**

For any questions, please call _____

Admission Counselor/Health and Wellness Staff

_____

Phone

Name: _____

Student ID#: _____ **DOB:** _____

SICKLE CELL DISEASE CHRONIC CARE MANAGEMENT PLAN

CAREER PREPARATION PERIOD, CAREER DEVELOPMENT PERIOD, CAREER TRANSITION PERIOD

Goals:

1. Enhance employability by optimizing control of sickle cell disease symptoms.
2. Educate the student regarding recognition of symptoms and self-management.
3. Reduce exposure to precipitants or triggers of sickle cell crises.
4. Optimize pharmacotherapy and immunizations utilizing national guidelines.
5. Implement regularly scheduled follow-up visits.

CAREER PREPARATION PERIOD		
YES	**NO**	
		Establish a Sickle Cell Disease Action Plan for student
		Offer the student a Medical Identification bracelet/necklace/anklet
		Conduct clinical evaluation • Refer to comprehensive Sickle Cell Program if available • Routine medical evaluations every 2-6 months • Laboratory evaluation every 6-12 months • Annual ophthalmology examination for retinopathy, increased ocular pressure, refraction • Routine dental care
		Monthly visits initially, then visits every 2-3 months
		Conduct laboratory evaluation • Complete blood count with differential • Reticulocyte count • Hemoglobin F % • Renal function (BUN, creatinine, urinalysis • Hepatobiliary function (ALT, bilirubin) • Ferritin (if transfused) • Pulmonary function (transcutaneous O2 sat)
		Assess vocational training match
		Mandatory TUPP/smoking cessation enrollment
		Reduce or eliminate exposure to other precipitants
		Annual influenza vaccination in October or November
		Emergency response plan
		Educate student about sickle cell disease complications • Early death in adults with sickle cell disease associated with painful crises, anemia, acute chest syndrome, chronic renal failure, and pulmonary disease • Frequency of painful crises may increase in early adulthood • Proliferative retinopathy predisposes to retinal hemorrhage, retinal detachment, glaucoma • Renal glomerular disease may lead to chronic renal failure • Chronic pulmonary disease and pulmonary hypertension may contribute to morbidity • Leg ulcers and oseonecrosis of the hips and shoulders may lead to disability
		Educate student about lifestyle choices • Weight management • Encourage whole fruits, vegetables, low fat milk, increased fiber • Avoid soda and fruit juices • Encourage aerobic physical activity (exercise 30 minutes per day, 5 days per week) • Avoid sedentary lifestyle (limit TV) • Avoid smoking • Limit alcohol use
		Educate student on sickle cell disease management as it relates to employment

Name: _____

Student ID#: _____ **DOB:** _____

CAREER DEVELOPMENT PERIOD		
YES	**NO**	
		Monitor adherence issues • Medication regimen • Medication refills • Routine medical care • Urgent medical care • Environmental control • Self-monitoring • Physiotherapy • Rest • Exercise • Nutrition • Tobacco, alcohol, drug use
CAREER TRANSITION PERIOD		
		Conduct a Wellness Center exit interview approximately 2 weeks before program completion.
		Identify potential sources of primary health care, and specialty care if needed, in the work community.
		Obtain signed HIPAA authorizations for the transfer of student health records to identified health care providers.
		Assist the student in enrolling or maintaining enrollment in a public or private health insurance program.
		Provide the student with a copy of the SF-93, SF-88, immunization records, and chronic care management plan, including flowsheets.
		Provide the student with an adequate amount of medication(s) and supplies at departure.

Asplenic Adolescents and Adults
Increased risk of fulminant bacteremia associated with high mortality:
- Pneumococcal conjugate vaccine
- Pneumococcal polysaccharide vaccine
- *Haemophilus influenzae b* vaccine
- Meningococcal vaccine (MCV4)
- Daily penicillin prophylaxis for at least 1 year after splenectomy—may discontinue at age 5 in sickle cell disease

Pneumococcal Vaccine
If no previous immunization, give **both**:
- Pneumococcal conjugate vaccine (PCV7)
  - 2 doses of PCV7 6-8 weeks apart
- Pneumococcal polysaccharide vaccine (PS23)
  - 1 dose of PS23 6-8 weeks after the last dose of PCV 7
  - 1 dose of PS23 vaccine 3-5 years after the first dose of PS23

Supportive Care
- Regular, slowly progressive exercise is encouraged
- Adequate hydration with water
- Minimize cold exposure
- Avoid high altitude exposure—travel in pressurized aircraft permitted
- Promptly seek medical care with onset of symptoms
- Availability of genetic counseling and prenatal diagnosis

See Sickle Cell Disease Flow Sheet for tracking patient visits.
See Treatment Guideline for Sickle Cell Disease for additional guidance.

Comment [r1]: Please tell where this is, or delete the lsentence. Don't find it in this CCMP.

SICKLE CELL DISEASE CHRONIC CARE MANAGEMENT PLAN FLOWSHEET

Student Name:										
Sex: M or F		**Date of Birth:**		**Date of Entry:**						
Co-Morbid Conditions:										
SICKLE CELL MAINTENANCE										
Date										
Annual or as indicated	Complete physical examination									
	Complete blood count (CBC), reticulocyte count, hemoglobin phenotype, liver profile									
	If transfused, consider ferritin, hepatitis serologies, and red cell phenotype									
	Urinalysis									
	Microalbuminuria									
	Chest X-ray									
	Ophthalmology evaluation for retinopathy									
SOCIAL PROFILE										
Every visit	Smoking history									
	Alcohol and drugs of abuse									
	Sexual history (including birth control and safe-sex measures)									
CHARACTERISTICS OF PAIN EPISODES										
Frequency										
Duration										
Usual home treatment										
Usual emergency department treatment										
Number and duration of hospitalizations										