Caring for Students with Sickle Cell Disease

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Sickle Cell Disease

• Most common inherited disorder in the United States
• Every baby born in the United States is screened via newborn screen and diagnosis is confirmed with hemoglobin electrophoresis
• Inherited in people of ALL races and ethnic backgrounds
• Genetic disorder that alters the DNA of red blood cells (RBC) changing the shape from soft and round to sticky, hard, sickle (crescent moon) shaped cells
• RBC have decreased life span of from 120 days to 10-20 days
• Leads to vaso-occlusion, ischemia, infarcts, and tissue death

Devika Bhatia, 2003

Sickle Cell Disease

• Many different types
  – Most common: hemoglobin SS and hemoglobin SC

<table>
<thead>
<tr>
<th>Genotype</th>
<th>Predicted Severity</th>
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<tbody>
<tr>
<td>HbSS</td>
<td>Moderate to very severe</td>
</tr>
<tr>
<td>HbSC</td>
<td>Mild to severe</td>
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<tr>
<td>HbS-β thal – 0</td>
<td>Moderate to very severe</td>
</tr>
<tr>
<td>HbS-β thal – +</td>
<td>Mild to moderate</td>
</tr>
<tr>
<td>HbSD (Punjab)</td>
<td></td>
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<tr>
<td>HbS-O Arab</td>
<td>Moderate to very severe</td>
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</tbody>
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Everyday Needs at School

• Plenty of fluids
• Flexible bathroom privileges
• Avoid physical exhaustion
• Avoid extreme temperatures
• Transportation to and from school
• Allow positive peer-to-peer interaction

Students with SCD can live a normal life!

Routine Medications

• Penicillin: an antibiotic taken twice daily for infection prophylaxis until about age five or fully vaccinated with meningococcal vaccinations
• Folic acid: a B vitamin taken one daily to help support red blood cells development
• Hydroxyurea: taken daily to decrease sickling and painful episodes for patients with genotypes HgSS or HgSbeta0
• Acetaminophen: taken as needed for pain control for mild pain
• Ibuprofen: taken as needed for mild pain
• Opioids: taken as needed for moderate to severe pain

Support for Students

• Allow accommodations during physical education and recess activities
• Special care of injuries
  – NO COLD PACKS!
• Be aware of emotional well-being
• Maintain open communication with parents
• Create individualized care plans
Fostering Emotional Well Being

- Normal treatment
- Group or partner projects
- Identification of strengths
- Support for self-esteem and competence
- Support after extended absences
- Social skills and peer interactions
- Concerns about mood and behavior

Individualized Care Plans

- Individualized Care Plans should include:
  - preventive measures to keep the child well at school
  - arrangements for giving pain medication to the child
  - non pharmacological pain management techniques
  - what constitutes an emergency and what to do
  - key contacts
- The care plan should be distributed to all teachers/staff that will have interaction with the student
- The plan should be reviewed and redistributed each year

School Performance Challenges

- Students with chronic illnesses often miss vital academic content due to absences
  - Total absences vary drastically depending on genotype and suffered complications
  - Should be given special consideration for missed instruction, assignments, and testing
- Neurocognitive complications due to silent infarcts or strokes hinders student’s executive functioning
  - Annual screening performed by our neuropsychologist
  - May need to assist with planning, provide incentive, or chunk information
- Decrease attention
- Learning and memory difficulties
- Reduced mental processing speed

Common Complications

- Pain
  - Be responsive to complaints of pain!
- Anemia
- Jaundice
- Fever
  - Be alert for signs of fever
- Delayed growth and development
- Stroke
- Avascular necrosis of joints
  - Very painful!
- Cholelithiasis (gallstones)
- Priapism
- Vision

Medical Emergencies!

- Fever over 101°F
- Difficulty breathing
- Chest pain
- Abdominal swelling
- Severe headache
- Sudden weakness or loss of feeling and movement
- Seizure
- Painful erection that last more than 4 hours
- Sudden change in vision

Student should immediately be taken to an ER or 911 should be called
Pain Prevention

- Hydration
- Keep warm and dry
  - Students should remain inside if temperature is 34°F or colder or excessive wind chill that correlates to 34°F or colder
- Check in periodically with student to establish usual levels of pain
- Manage stress
- Reinforce coping strategies to utilize when pain worsens
- Prevention is important, although pain episodes are often unpredictable

Pain Management

- Eliminate any possible contributing factors
  - Cold/wet
  - Dehydration
  - Strenuous exertion
  - Stress
- Refer to patient’s treatment plan
  - Medications
    - Acetaminophen, ibuprofen, opioids
  - Coping strategies
  - Non pharmacologic methods
- Functional pain assessment

Non Pharmacologic Methods for Pain Relief

- Meditation
  - Applications
    - Cain
    - Stop, Breath, and Think Kids
    - Smiling Mind
  - Books
    - A Zebra Like Me
    - Master of Mindfulness: How to be your own Superhero in Times of Stress
- Guided Imagery
- Deep Breathing
- Distraction
- Aromatherapy
  - peppermint or lavender
- Heating pad/hot pack

Guide for Functional Pain Assessment

- 0 = No pain
- 2 = Tolerable pain: able to perform all activities permitted
- 4 = Tolerable pain: able to perform some permitted activities: pain prevents full engagement in rehabilitative activities
- 5 = Tolerable pain that becomes intolerable and interferes with physically demanding activities (e.g. Physical Therapy)
- 6 = Intolerable: interferes with most activities requiring physical exertion, but not passive activities, such as reading, watching TV, talking
- 8 = Intolerable: Interferes with all “active” and most passive activities such speaking about pain
- 10 = Intolerable: patient is unable to do anything or even speak due to their pain

Comprehensive Sickle Cell Clinic

- Patients with sickle cell disease attend our comprehensive clinic at least annually to see our multidisciplinary team and complete routine testing
  - Hematologist
  - Social Work
  - Neuropsychologist
  - Pharmacist
  - Nurse
  - Genetic Counselor
  - Dentist

Helpful Resources

- Our team!
  - Virtual in-service for teachers and staff
  - Discuss the disease with student’s peer/classmates
  - Help with accommodations for individualized health plan
  - Address any concerns that come up

Contacts

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Sickle Cell Social Worker: Abbie Yoder, LMSW
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Helpful Resources

• Online resources
  • Centers for Disease Control and Prevention
    • https://www.cdc.gov/ncbddd/sicklecell/index.html
  • Sickle Cell Information Center
    • http://scinfo.org/2015/02/24/faq-teachers-and-students/
  • St Jude Children’s Research Hospital
    • https://www.stjude.org/treatment/disease/sickle-cell-disease/educational-resources.html
  • U.S. Department of Education (DOE): A Guide to the Individualized Education Plan (IEP)
    • https://www2.ed.gov/parents/needs/speced/iepguide/iepguide.pdf

Questions?

References


Children’s Hospital of Philadelphia.