

Managing Medical Emergencies in Childhood and Adulthood

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Faculty

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General-Complications

- **Essentially all organs can be affected by SCD due to chronic anemia and pain**
- **Manifested differently in children and adults**

Complications – Aplastic Event

- **Aplastic event**
 - **Severe drop in the hemoglobin, usually due to infection, causes the bone marrow to stop making RBCs**
 - **Symptoms include:**
 - **Headache**
 - **Irritability**

Complications – Aplastic Event

- **Poor appetite**
- **More than usual jaundice**
- **Unusual tiredness**
- **Rapid heart beat**
- **Change in complexion**

Complications - Pain

- **Common precipitators or none**
 - **Adults usually can give good history, children cannot**
 - **Unusual stress or anxiety**
 - **Dehydration**
 - **Extreme cold**

Complications - Pain

- If pain is severe, may require hospitalization if other treatments do not work
- Often confused with other diagnosis in children and with drug addiction in adults
 - That is why diagnosis and proper medical and family history are so important

Complications - Children Pain

- Children often cannot communicate pain directly
- Usually manifested in children through other signs:
 - Fever
 - Infection
 - N&V
 - Severe abdominal pain

Complications - Children Pain

- Bone pain
- Difficulty breathing
- May be manifested as swelling in area of the pain
- Smaller children
 - Usually hands and/or feet
 - May be related to bone infection

Complications - Kidney/Bladder

- Kidney and Bladder Infections
 - Bladder infections fairly common for children and adults
 - If they are not treated promptly, can move from bladder to kidneys to cause kidney damage

Complications - Kidney/Bladder

- Symptoms:
 - Irritating or painful urination
 - Hematuria
 - Polyuria

Complications - Children

- Enuresis
 - Children should be managed closely

Complications – Splenic Sequestration

- Possible complication of SCD commonly occurs in children under the age of 5
- Occurs when sickling occurs in the vessels of the spleen, enlarging the spleen, causing anemia and potentially shock
 - Medical emergency

Complications – Splenic Sequestration

- Treated with hydration, analgesia, and red cell transfusion
- Sometimes splenectomy warranted

Complications – Splenic Sequestration

- Signs and symptoms of complications of spleen:
 - Sudden weakness
 - Pallor
 - Especially of lips, gums and nails
 - Abdominal pain

Complications – Splenic Sequestration

- Increase in abdominal girth
- Enlargement of the spleen
- Fever

Common Complications - Children

- Delay in skeletal maturation
- Delay in onset of puberty
 - 2 years
- Gallstones
- Strokes

Complications - Gallstones

- About a third of children with SCD have gallstones by the age of seven
- Excess bilirubin caused by RBC destruction which leads to gallstone formation
- Surgery is often needed
- This is the most common surgery in children with SCD

Complications - Gallstones

- **Signs and symptoms:**
 - Right upper abdominal pain
 - Shoulder pain
 - N&V

Complications - Stroke

- **Sudden and severe complication of SCA**
- **Affects from 6-12 percent of patients**
 - Usually between the ages of 5-10
- **May or may not have a precipitating event**
- **Major episodes can cause:**

Complications - Stroke

- Brain damage
- Paralysis requiring rehabilitation
- Coma
- Death
- **Minor strokes can cause learning disabilities or affect cognitive learning skills**
- **Risk in adults is increased**

Complications - Stroke

- **Signs and symptoms:**
 - Difficulty with memory
 - Difficulty speaking or understanding others
 - Defective or absent language
 - Difficulty with balance
 - Muscular weakness on one half of body

Complications - Stroke

- Sudden loss of, or blurred vision
- Persistent headaches, or sudden strong headache
- Fainting
- Dizziness
- Sudden weakness or tingling of an arm, leg, or the whole body

Other Complications

- **Priapism**
 - Painful swelling of the penis due to trapped RBCs, Sustained erection
 - Usually occurs without relation to sexual excitement and can occur in young children as well as adults
 - Prolonged episodes-several days, hospitalization

Case Study

- The EMS is called to the home of an AA boy 10 years old, recently adopted by parents
- He complains of severe abdominal pain
- The family history is limited, although you know that he was born in Africa
- What could be the differential diagnosis?

Summary/Bottom Lines

- SCD affects most organ systems and can result in serious medical emergencies
 - Can ultimately cause death
- Important to have early diagnosis, prevention and maintenance of SCD
 - Key to limiting some emergencies

Summary/Bottom Lines

- Important that sharing of diagnosis should be made with all HCPs, teachers, employers, etc
- Important to recognize common emergencies and be able to differentiate them between children and adults

References

- Wethers, DL “Sickle Cell Disease in Childhood” Accessed on 09/26/11 www.aafp.org/afp/20000902/1013.html
- Steinberg MH. “In the Clinic: Sickle Cell Disease” Ann Intern Med (2011) Sept 6; 155 (5): ITC31

References

- Understanding the Child with Sickle Cell Disease: A Handbook for School Personnel, Virginia Department of Health, Accessed on 09/26/11, www.vahealth.org/sicklecell/docs/SchoolHandbook_SickleCellChild_PDF%5B1%5D.pdf