

[FSC]  
**Hemoglobin SC Disease (HbSC)**

**Differential Diagnosis:** Hemoglobin SC disease most likely.

**Condition Description:** A red cell disorder characterized by the presence of fetal hemoglobin (F) and hemoglobins S and C in the absence of Hb A. The hemoglobins are listed in order of the amount of hemoglobin present (F>S>C). This result is different from FAS, which is consistent with sickle carrier.

**You Should Take the Following Actions**

- **Contact the family to inform them of the screening result.**
- **Refer to a pediatric hematologist (See attached list.)**
- **Evaluate infant and assess for splenomegaly.**
- **Repeat newborn screen if second screen has not yet been done.**
- **Initiate timely confirmatory/diagnostic testing as recommended by consultant.**
- **Initiate treatment as recommended by the consultant.**
- **Educate parents/caregivers regarding the risk of sepsis, the need for urgent evaluation for fever of  $\geq 101.5^{\circ}$  F, and signs and symptoms of splenic sequestration.**
- **Report findings to newborn screening program.**

**Confirmation of Diagnosis:** The NBS program confirms the diagnosis by DNA studies.

**Clinical Expectations:** Newborn infants are usually well. Hemolytic anemia and vaso-occlusive complications develop during infancy or early childhood. Complications include life-threatening infection, splenic sequestration, pneumonia, acute chest syndrome, pain episodes, aplastic crisis, dactylitis, priapism, and stroke. Comprehensive care, including family education, immunizations, prophylactic penicillin and prompt treatment of acute illness, reduces morbidity and mortality.

**Additional Information:**

**Grady Comprehensive Sickle Cell Center**  
<http://scinfo.org/hemoglb.htm#SICKLE%20HEMOGLOBINS>

**American Academy of Pediatrics**  
<http://pediatrics.aappublications.org/cgi/content/full/109/3/526>

**Management and Therapy of Sickle Cell Disease**  
<http://www.nhlbi.nih.gov/health/prof/blood/sickle/index.htm>

**Sickle Cell Disease Association**  
<http://www.sicklecelldisease.org/>

**Sickle Cell Disease in Children and Adolescents:  
Diagnosis, Guidelines for Comprehensive Care, and  
Care Paths and Protocols for Management of Acute  
and Chronic Complications**  
<http://www.dshs.state.tx.us/newborn/pdf/sedona02.pdf>

**Referral (local, state, regional and national):  
Comprehensive Sickle Cell Center Directory**  
<http://www.rhofed.com/sickle/index.htm>

**Sickle Cell Information Center**  
<http://www.scinfo.org/clinics.htm>