

# Sickle Cell Disease

## A to Z

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# Sickle Cell Disease A to Z

## What is Sickle Cell Disease?

Sickle Cell Disease is an inherited disorder of abnormally shaped hemoglobin.

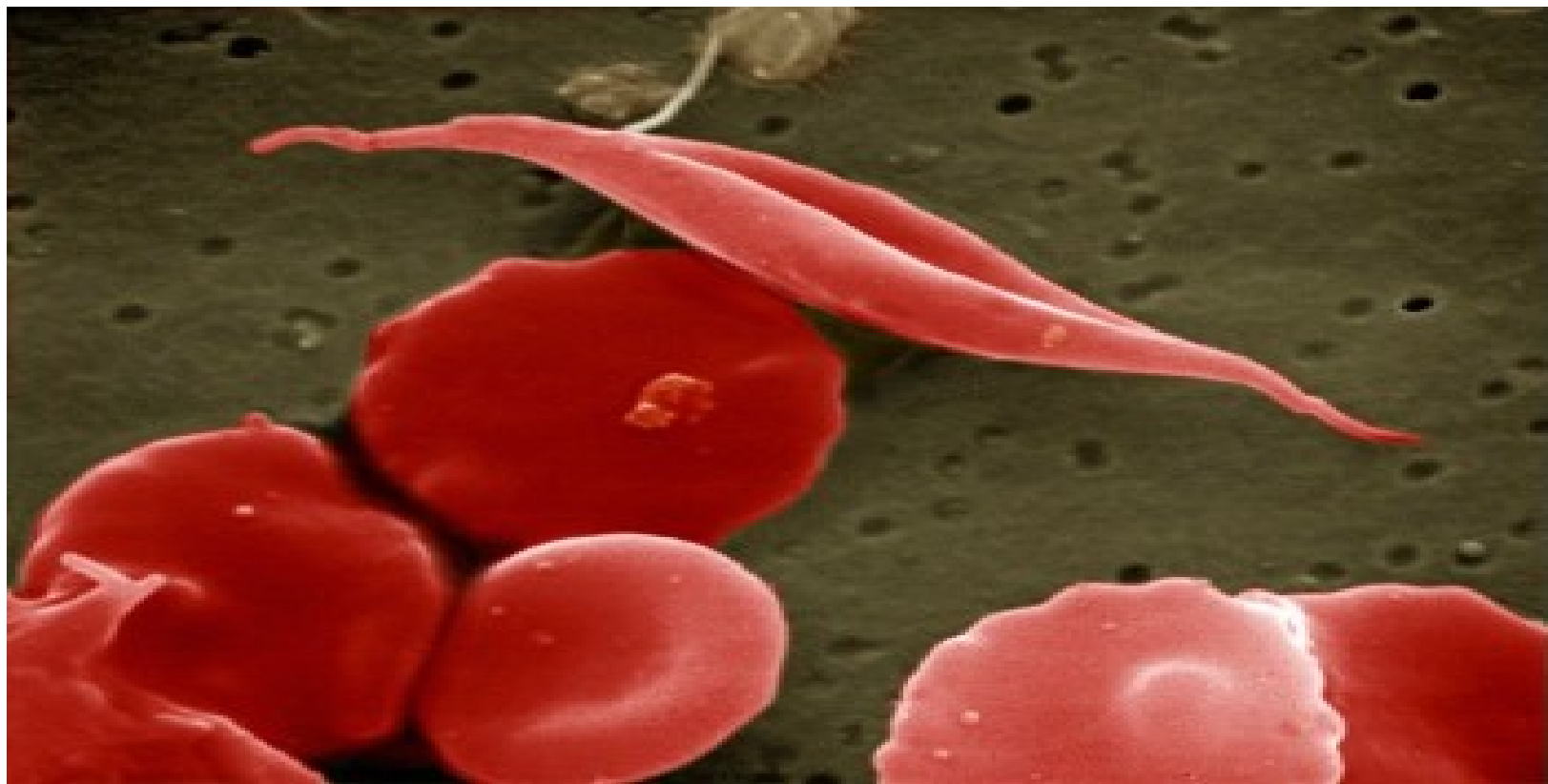


# Sickle Cell Disease A to Z



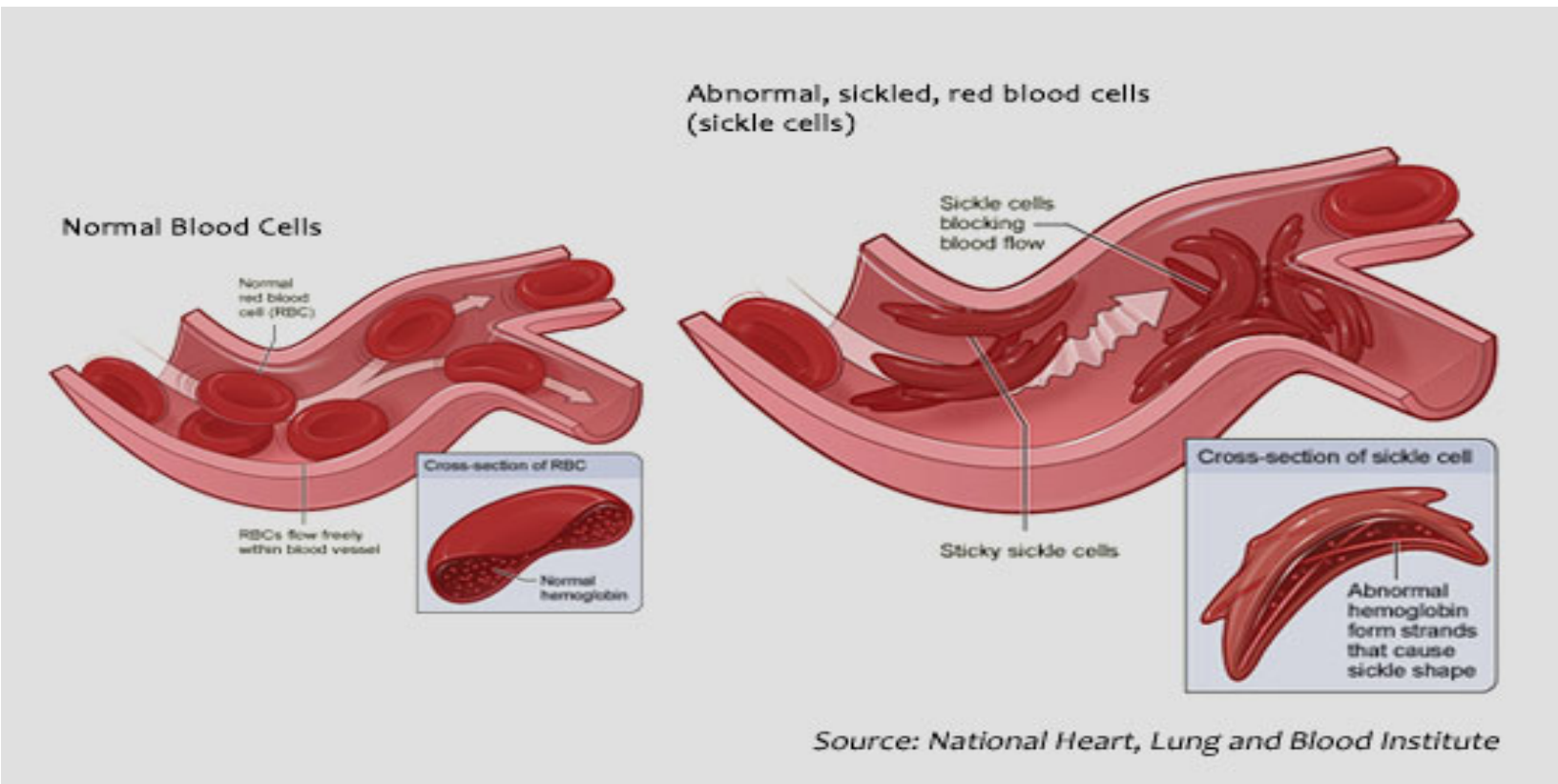


# Sickle Cell Disease A to Z





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# Sickle Cell Disease

## A to Z

### Diagnosis

- Newborn state screening in Texas currently screens for 28 disorders including Sickle Cell Disease.



# Sickle Cell Disease

## A to Z

## Genetics

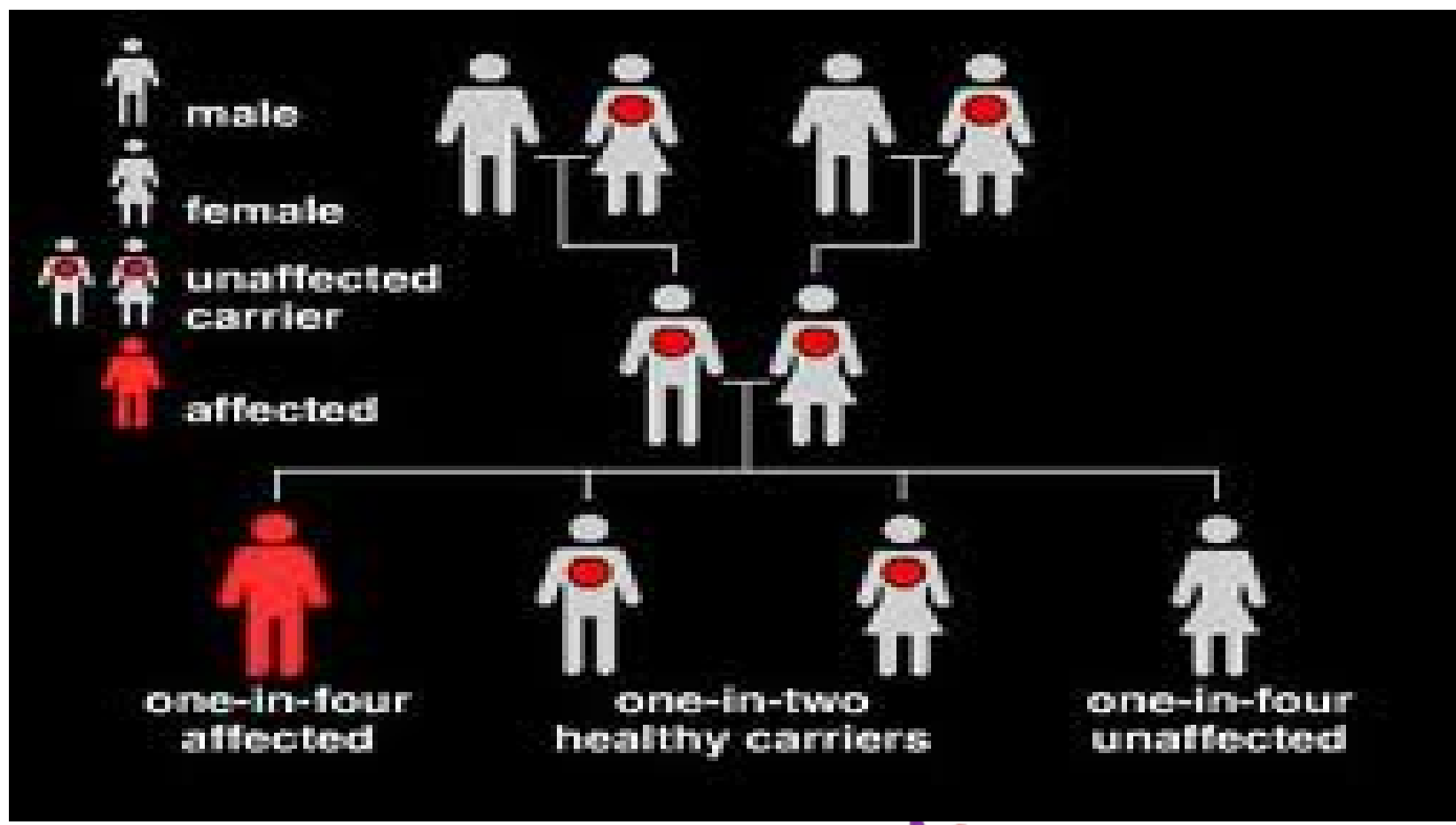
There are several different genotypes of Sickle Cell Disease. The specific genotype plays a big role in what symptoms one might develop or experience:

- Hbg SS
- Hgb SC
- Hgb S / Beta + Thal
- Hgb S / Beta Thal

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# Sickle Cell Disease A to Z





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## Survival

- Sickle Cell Patients used to have a very poor prognosis and not usually survive past their 30's or 40's.
- These numbers are improving due to:
  - Comprehensive care
  - Immunizations
  - Antibiotics
  - Better treatment of complications



# Sickle Cell Disease A to Z

## Major Clinical Manifestations

- Painful episodes
- Neurological injury
- Psychosocial Issues
- Growth and development
- Infection
- Cerebrovascular events



# Sickle Cell A to Z

## Painful Episodes

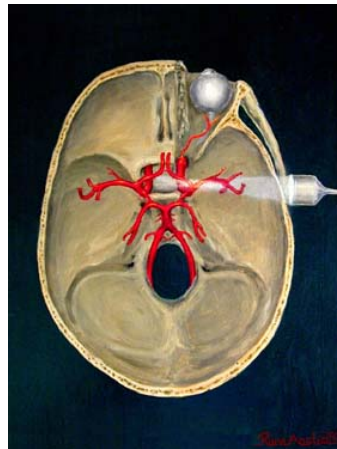
- Previously called “Sickle Cell Crisis”, now referred to as “Vaso-occlusive Episode”
- This is the most frequent symptom experienced
- Painful episodes can oftentimes be managed at home



# Sickle Cell A to Z

## Neurologic Injury

- 25% of children with Sickle Cell Anemia have silent strokes which may impair neurocognitive function and be a predictor of progressive neurologic disease
- Screening studies, Transcranial Dopplers (TCDs) are done once a year to look for predictors of stroke





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## Psychological Issues

- Most children with Sickle Cell Disease are well adjusted
- There are some issues involving:
  - Low self esteem
  - Social isolation
  - Poor family relationships



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### Growth and Development

- Growth failure and delayed puberty are common
- Normal height is often achieved by adulthood but weight remains lower
- Both boys and girls have delayed sexual maturation
- Girls often have delayed menarche
- Poor ability to concentrate urine; prolonged bedwetting



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### Infection

- Major cause of hospitalization and death in Sickle Cell patients
- Sickle Cell patients are more susceptible to life-threatening infections
- Sickle Cell patients lose the functionality of their spleen very early in life (around 2-4 years) due to sickling cells clogging the microvessels of the spleen.



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### Infection

- Penicillin VK prophylaxis
- Pneumococcal vaccination
- Menactra
- Flu shots



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## How Do We Treat Sickle Cell Disease?

### Comprehensive Medical Care

- Early diagnosis
- Extensive health maintenance
- Prophylactic measures
- Parental education
- Psychosocial support
- Periodic medical assessment and monitoring



# Sickle Cell Disease

## A to Z

## How Do We Treat Sickle Cell Disease?

- Medical Intervention
  - Hydroxyurea
  - Chronic Transfusion
  - Pain Control



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## What Do You Need To Know?

- More frequent absences than other students
- Necessity of providing make-up work to the student
- Help avoid vaso-occlusive events by:
  - Allowing the child to keep well hydrated with water
  - Allowing for increased bathroom breaks
  - Encouraging PE participation
  - Avoiding exposure to extreme temperatures hot/cold
- Sickle Cell patients may tire before others; allow for rest periods



# Sickle Cell Disease A to Z

## When to Seek Medical Attention

- Fever of 101 or greater
- Chest Pain
- Abdominal pain
- Extreme pain
- Headache, asymmetry, drooling, speech difficulty
- Numbness or weakness
- Prolonged erection (priapism)



# Sickle Cell Disease A to Z





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## References

- [www.scinfo.org](http://www.scinfo.org)
- [www.sicklecellkids.org](http://www.sicklecellkids.org)
- [www.uptodate.com](http://www.uptodate.com)