# Sickle Cell Disease in Arkansas

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# Introduction

## Overview

- Purpose of meeting
- Origin of disease

#### Epidemiology (World)

Origins of the Sickle Cell Gene Greece Italy Albania India urkey Senegal Abitotion Previoushill Benin CAR



# Epidemiology (U.S.)

- Population as of 3-22-16 323,214,999
- Estimate of 40,078,287.88 African Americans which shows 66,797 with disease based on 1 in 600 with disease and 4,007,828 with disease trait based on 1 in 10

## Epidemiology (Arkansas)

 2.966 million as of 2014 population which gives an estimate of 40,000 plus with trait and an estimate of 1,107 plus with disease based on 15.4% African Americans

### Pathophysiology

- Inherited Disease (genetic)
- Results from abnormal hemoglobin which differs from normal hemoglobin by the 6<sup>th</sup> amino acid position of the globulin component by having valine in that position rather than glutamic acid which is from a substitution
- When the red blood cell is deprived of oxygen it becomes irregularly shaped or sickled and can lead to occlusion of the capillary blood vessel
- Normal life span of red blood cell can be up to 120 days but the sickle cell may last only 30-60 days.
- Sickle cell trait results when only half of the gene pool for hemoglobin is abnormal
- Sickle cell disease results when more than half of gene pool for hemoglobin is abnormal

#### **Disease Management**



- Medical
- Social
- Political
- Economic

#### Treatment



Preventive
Acute care
Chronic care

#### Prevention



- Education
- Genetic counseling

#### Future Management

Bone marrow transplant Stem Cell transplant Gene therapy

#### Future Recommendations in Arkansas

- Screening Programs
- Medication Reform
- Registry
- Provider Education



Thank You!