



RuSH: Sickle Cell Surveillance and Registry Program

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Centers for Disease Control and Prevention

Outline

- ❑ Background / Goals/ Partners
- ❑ Site Selection
- ❑ Data gathering / Data elements
- ❑ Evaluation of data collected
- ❑ Next Steps: Future expansion of Surveillance and Development of Registry
- ❑ Questions

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Domestic Statistics

- In the United States, SCD affects approx.
 - 72,000 to 84,000¹ (104,000 to 138,900)
 - 89,000²

- Among newborn American infants, SCD occurs in approximately
 - 1 in 400 Blacks
 - 1 in 1,400 to 1 in 36,000 Hispanics
 - 1 in 80,000 Whites

- Over 3 million Americans have sickle cell trait
 - 1 in 12 (or 8%) African Americans

1 Hassell, K (2009) AJPM (in press)

2 Brousseau et. al (2009) American Journal of Hematology

Domestic Statistics

- ❑ The total healthcare cost associated with sickle cell disease is 1.1 billion annually (1)
- ❑ The number of hospitalizations among adults with sickle cell disease (SCD) in 2004 was 83,149 (2)
- ❑ The total hospital costs for hospitalizations principally for SCD were approximately \$488 million (2)
- ❑ Among those hospital stays principally for SCD, 66 percent were paid by Medicaid and 13 percent were paid by Medicare (2)

Current Challenges

- Three decades shorter life expectancy
- Significant pain and other complications
- Growing population
 - More adults
 - Changing demographics

Current Challenges

- ❑ Unknown Prevalence
- ❑ Lack of access to specialty care/quality care especially for adults
- ❑ Lack of understanding of risk factors and complications over the lifespan
- ❑ Lack of understanding the overall impact and barriers to diffusion of effective interventions
- ❑ No national coordination of services
- ❑ Lack of community awareness

Stakeholders

- ❑ Federal Partners
- ❑ Sickle Cell Disease Association of America (SCDAA) and other SCD CBOs
- ❑ Thalassemia CBOs
- ❑ Post-ASPHO SCD Summit Surveillance Action Committee
- ❑ States
- ❑ SCD and Thal Consumer Community



A collaborative effort between NHLBI and CDC working with other Federal and state agencies

- Interagency agreement – CDC and NHLBI
- 4 year pilot project
- Two Phases: Planning and Implementation
- Phased implementation plan.

RuSH Objectives

- Develop a hemoglobinopathy surveillance system that will
 - Fill a need for generating statistically sound estimates and to store bio-specimens for use in genetic and genomic analyses
 - Provide data for population-based and clinical studies, health services planning, and policies
- Create an infrastructure that enables the development of registries to monitor health outcomes of hemoglobinopathy patient populations
- Establish a bio-specimen repository for hemoglobinopathies

Public Health Surveillance, 2009

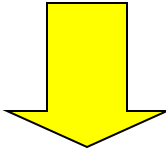
Definition of Public Health Surveillance

“The ongoing systematic collection, analysis, and interpretation, of data on specific health events affecting a population, closely integrated with timely dissemination of these data to those responsible for prevention and control.”

- Centers for Disease Control and Prevention

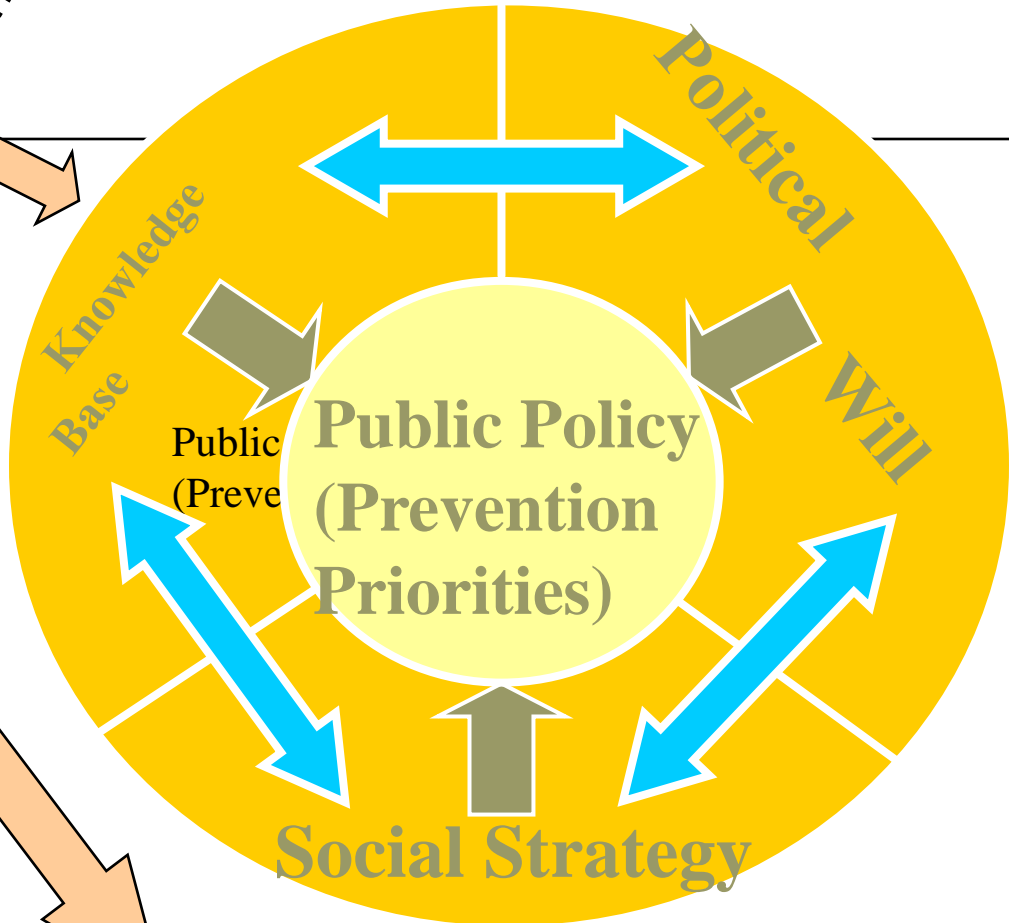
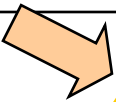


Why do we need public health surveillance?

- Assess burden of disease
 - Monitor trends in health
 - Identify emerging risks
- 
- Develop, implement, and evaluate disease control and prevention programs

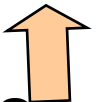


CDC, NIH, HRSA, FDA,
AHRQ, Academia, Others



Richmond and Kotelchuck's Health Policy Model

Communities, CBOs, State and Local Governments



Uses of Public Health Surveillance

- Planning of programs and services for SCD
 - Characterization of the affected population
 - Number of affected individuals
 - Location and types of utilized services/facilities
 - Spectrum of complication and issues experienced/services needed and gaps in services available
 - Nature and number of providers
 - The effectiveness of services, prevention efforts and intervention on populations

Objectives of Surveillance System

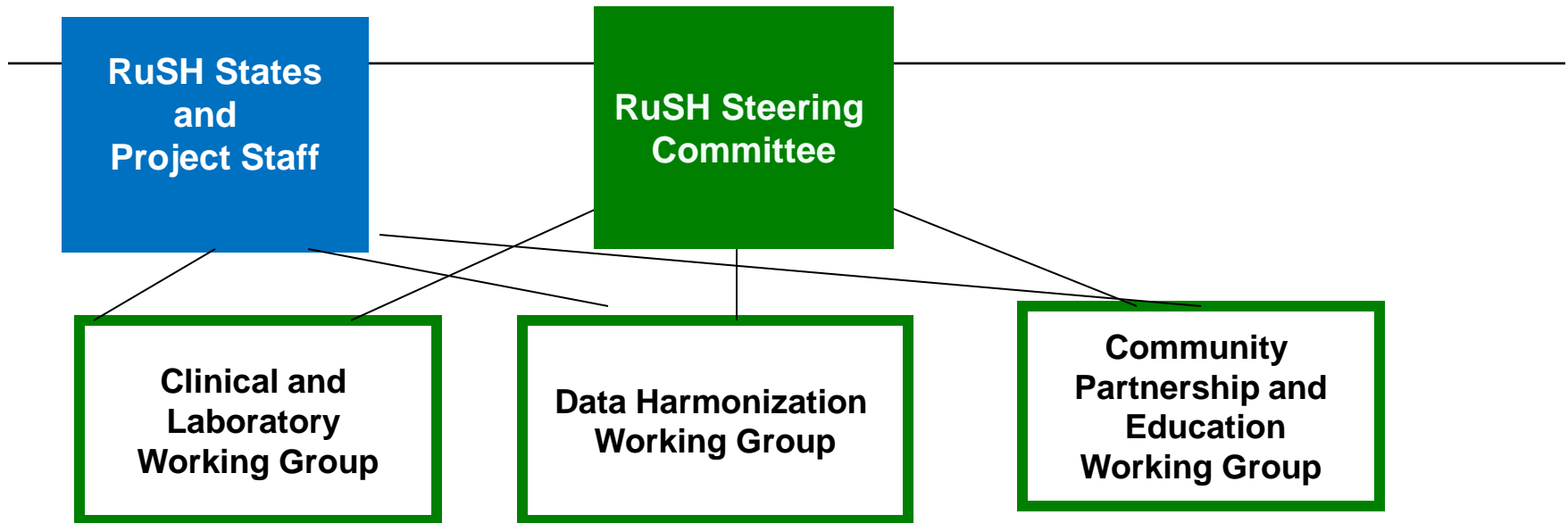
- To describe the ongoing pattern of disease occurrence and to link with public health action
- **Primary**
 1. Prevalence of hemoglobinopathies by genotype including patients not born in US
 2. Incidence of hemoglobinopathies using NBS data plus immigrants
 3. Demographics characteristics and geographic distribution

Objectives of Surveillance System

Secondary

1. Disease severity, co-morbidities, and chronic disease complications of persons with hemoglobinopathies;
2. Disease and treatment-related infections;
3. Reproductive and pregnancy outcomes of hemoglobinopathy patient populations;
4. Mortality rates, including case fatality rates for hemoglobinopathies and complications;
5. Health care utilization, costs of care, and the geographic variation of specific services

Committee and Working Groups for RuSH



CDC/Division of Blood Disorders

NIH/NHLBI

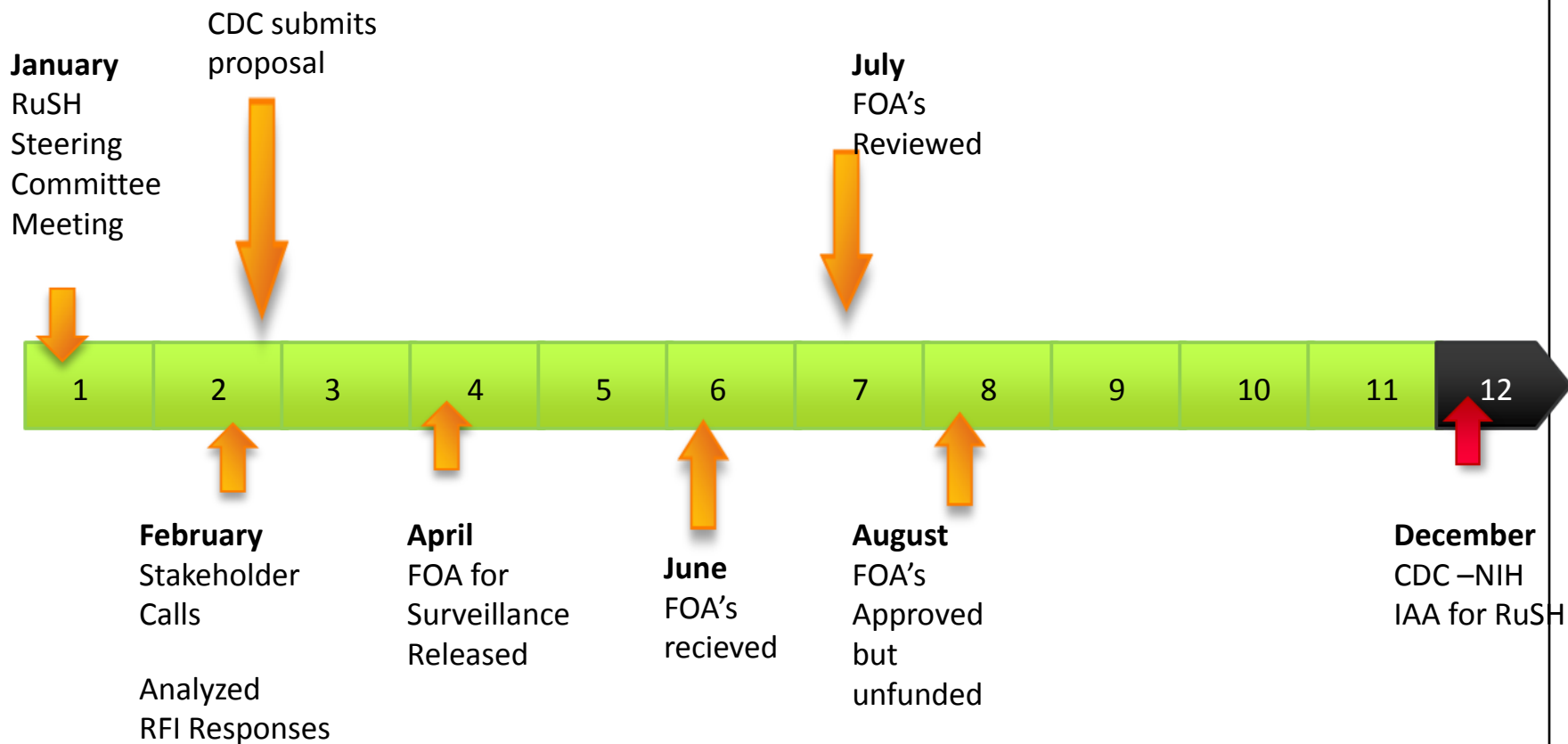
CDC RuSH Oversight Committee

NIH RuSH Monitoring Committee



RUSH TIMELINE 2009 –

Planning Phase



Ongoing

- Monthly CROC Meetings
- CDC Weekly RuSH Team Meetings
- Montly CDC-NHLBI Meetings

RUSH Steering Committee

January 13-14, 2009

- Key Recommendations
 - Learn from experience
 - Avoid duplication
 - Modify list of conditions
 - Distinguish surveillance from registry approaches
 - Considering state qualifications and readiness
 - Consider and be responsive to the ethical, legal, and social issues (ELSI).

Community-Based Organizations (CBOs)/Advocacy Groups Panel - Themes

- ❑ CBOs should be involved early to start educational process and build trust with clients. They should also be apart of process throughout project.
- ❑ CDC needs to clearly articulate how RuSH will benefit the patient and their family
- ❑ Avoid a paternalistic approach - patients need to be empowered and feel that their participation can make a difference in their families' outcomes

Local and State Health Partners Panel - Themes

- ❑ State infrastructure and capacity varies from state to state
- ❑ Lack of continuity of care makes accessing adult population difficult
- ❑ Participants emphasized that success of RuSH will rely heavily on CBO involvement and their ability to build trust between patients and providers

Request for Information Summary

- 6 responses
 - 4 from clinical care institutions,
 - 1 from a private non-profit business organization, and
 - 1 from a blood center.
- 5 respondents described 10 existing data sets
- 1 respondent described a database in the development process.

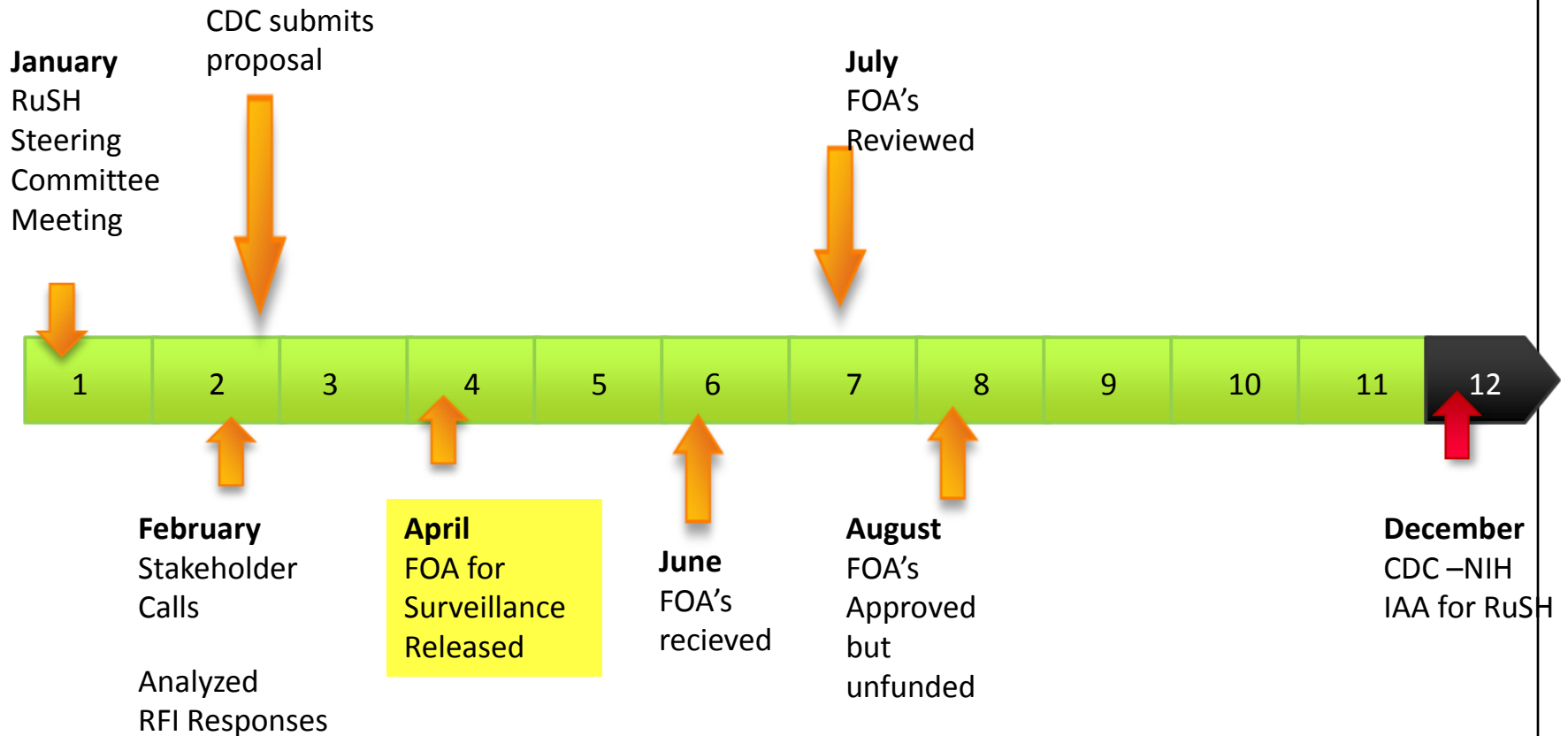
Request for Information Summary

- Populations:
 - SCD, SC trait, and/or abnormal hemoglobinopathies
 - one database included information related to family members.
- 8 data bases contained clinical information.
- 3 databases with linked biospecimens
- The biospecimen collections linked to data bases had specimens from 200-600 patients.
 - The data set with the largest no. of patients had information on approximately 3,500 patients.

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RUSH TIMELINE 2009



Ongoing

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Funding Opportunity Announcement

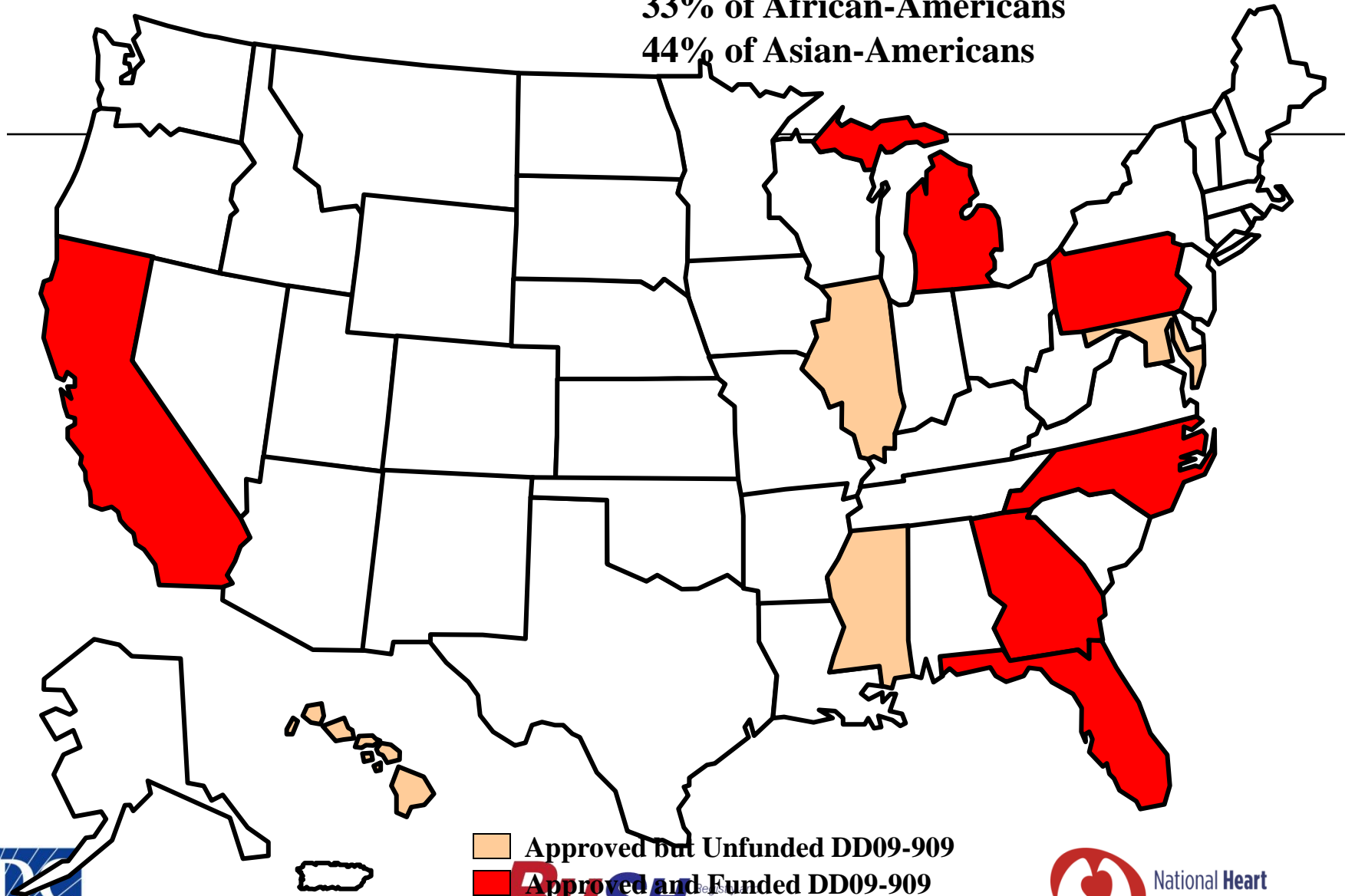
- Two Modules
 - Module A: Surveillance of Hemoglobinopathies in States
 - Module B: Capacity Building and Surveillance of Hemoglobinopathies in States with a High Historically Underserved Population
- Eligibility:
 - State governments, territories, NYC and DC
 - Module A: All states
 - Module B: States with
 - 14% or more of population below U.S. poverty level
 - At least 20% or more racial/ethnic minorities
 - 14% or more of the population who are black or African American

FOA Activities

- ❑ **Data collection and reporting**
- ❑ **Collaboration**
- ❑ **Data integration**
- ❑ **Dissemination of information**
- ❑ **Evaluation and progress reports**

RuSH States , February 2010

33% of African-Americans
44% of Asian-Americans

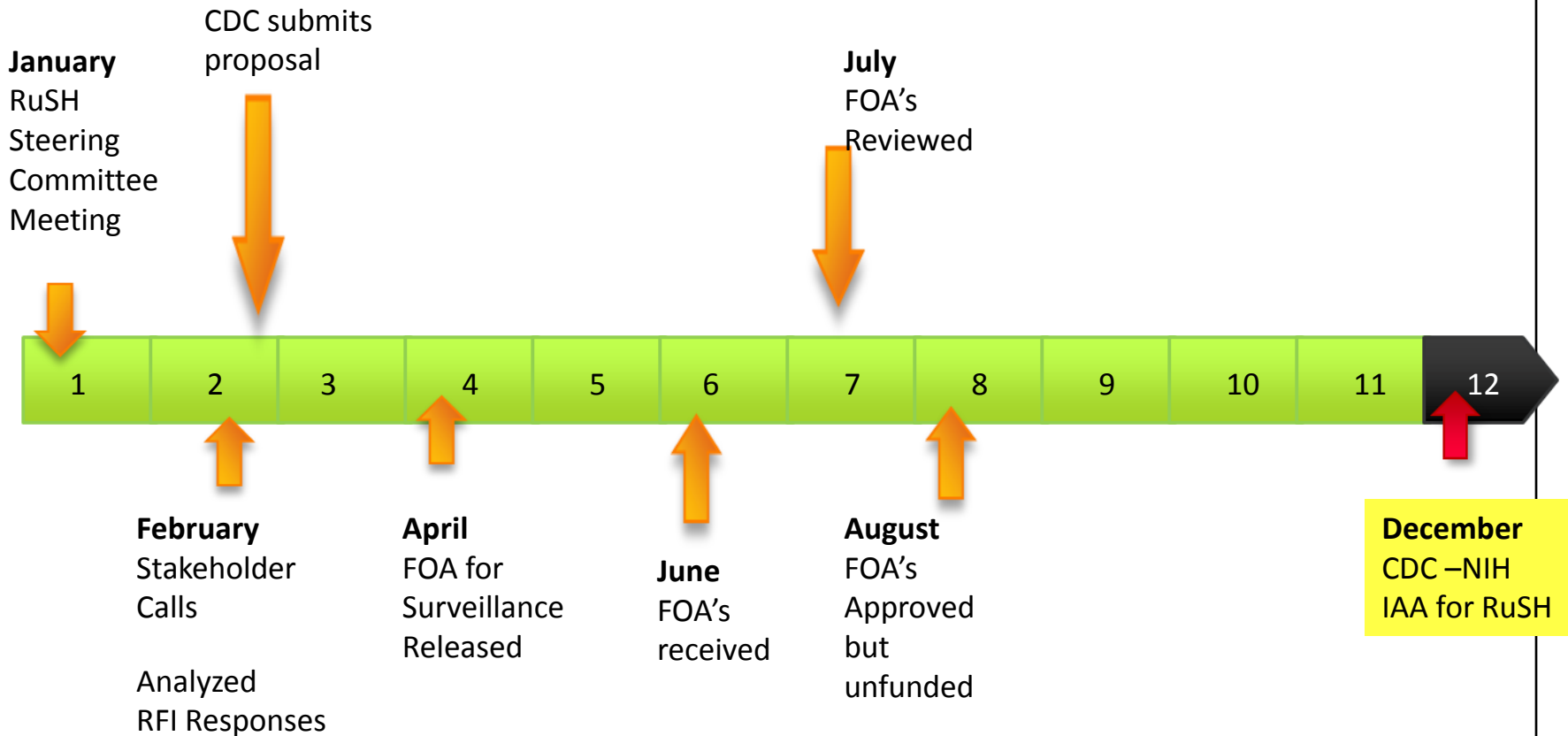


Approved but Unfunded DD09-909

Approved and Funded DD09-909



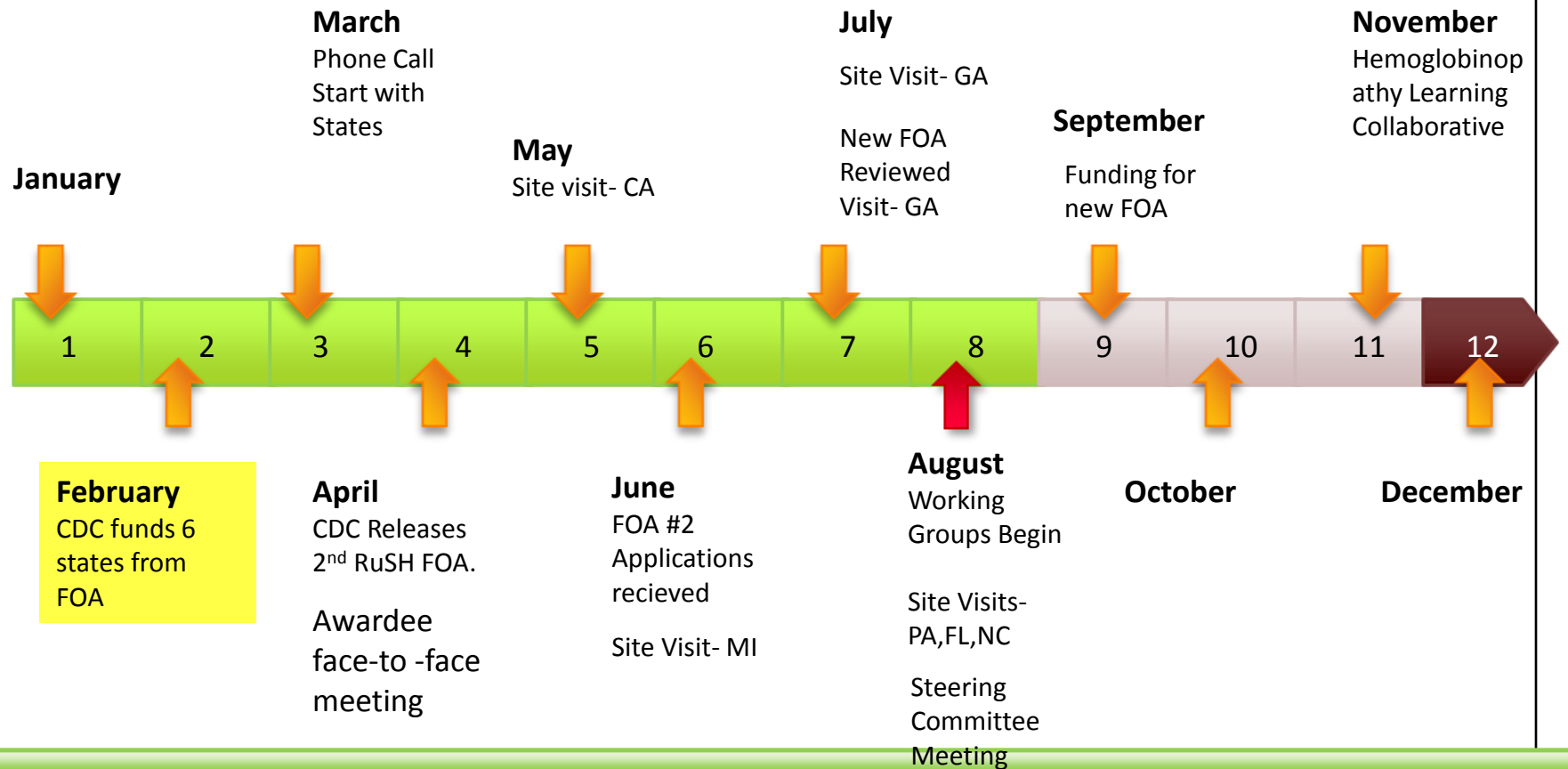
RUSH TIMELINE 2009



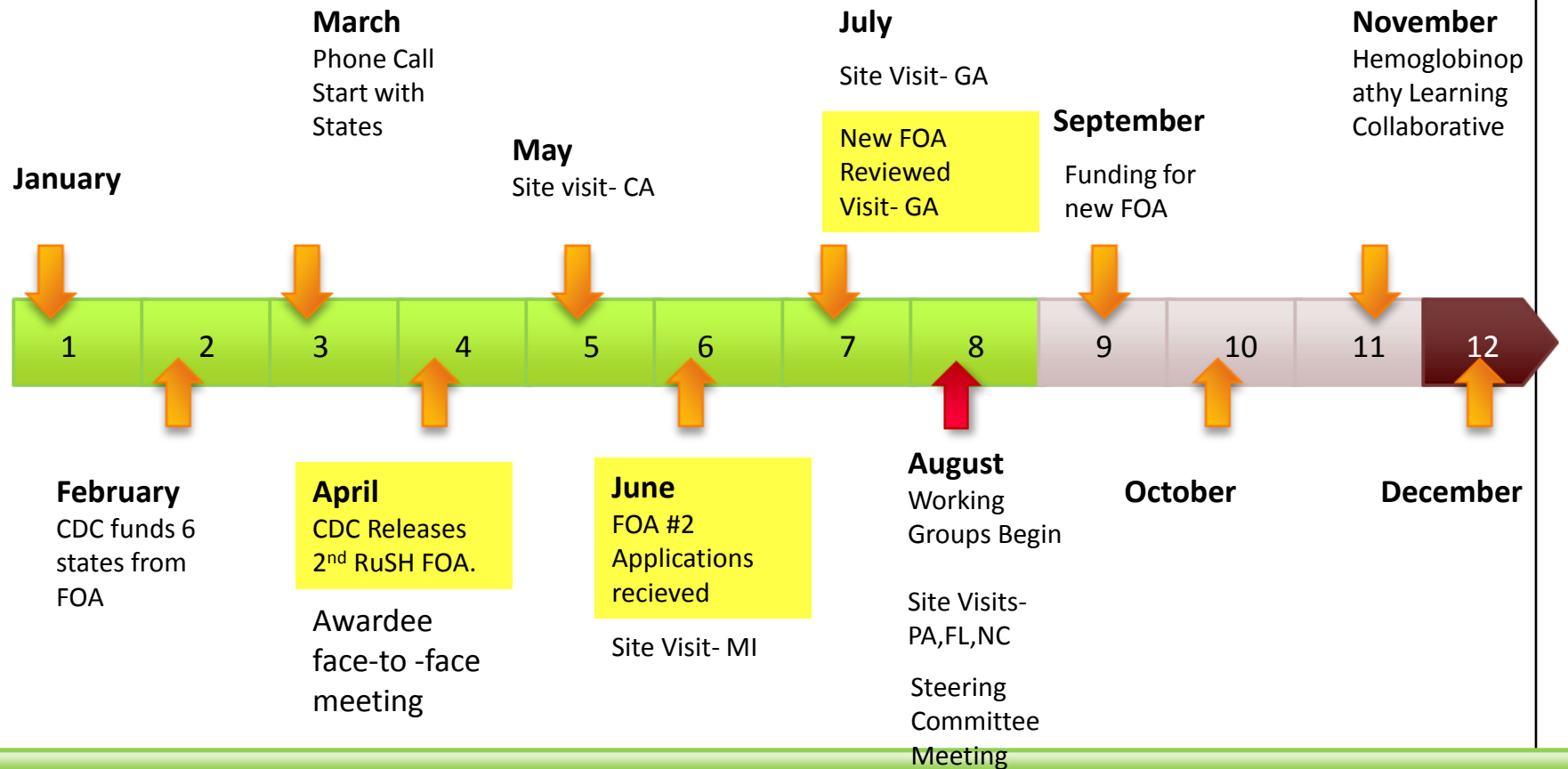
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RuSH TIMELINE 2010



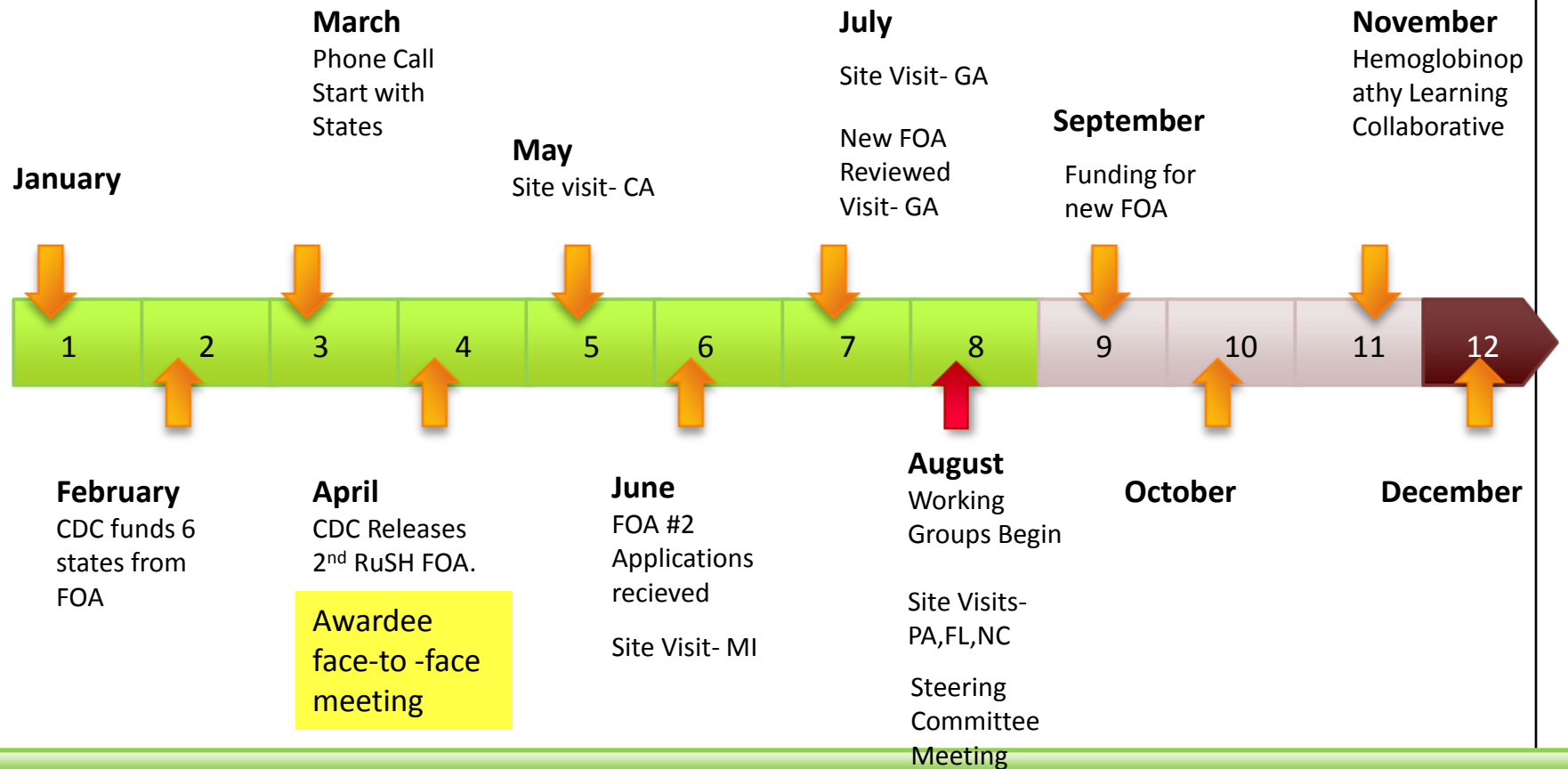
RuSH TIMELINE 2010



FOA 2010

- **State-based Surveillance for Hemoglobinopathies**
- **Applicants**
 - New York
 - Ohio
 - Louisiana
 - Hawaii

RuSH TIMELINE 2010



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Collaborative Activities Year 1

- ❑ Refine state surveillance strategies
- ❑ Indicators and Case definition Discussion
- ❑ Minimal Dataset Development
- ❑ Data Linkage
- ❑ Develop Data Sharing and Dissemination Plan

Common Data Sources

- ❑ Newborn Screening
- ❑ Vital statistics birth and death files
- ❑ Medicaid claims
- ❑ Hospital discharge data
- ❑ Emergency department data
- ❑ Clinic-based data for individuals ever in care
- ❑ Program Service Data
- ❑ Registries
 - Immunization, Stroke , Birth Defects, Cancer

Unique data sources

- ❑ WIC
- ❑ Medicare
- ❑ Other Payers
- ❑ Data from CBOs
- ❑ Blood Banks
- ❑ School Health
- ❑ Registries
- Immunization, Stroke , Birth Defects, Cancer

Challenges for RuSH surveillance

- Case definition (who's in the dataset?)
 - Thalassemia
 - Sickle cell
- Datasharing/data access
- ICD coding validity
 - Need for validation study
- Non-NBS population
 - Thalassemia in most states
 - Adults
- What indicators can you measure ?

Working Group Composition

- 10-12 members
- Chaired by 1 member of RSC
- 1 member from each site/awardee
- Additional members selected by CDC and NHLBI based on expertise
 - **Data Collection and Harmonization WG**
 - **Clinical and Laboratory WG**
 - **Community Partnerships and Health Education WG**

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Collaborative Activities Year 2

- ❑ Plan Program Evaluation
- ❑ Surveillance Evaluation
- ❑ Data Validation

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Next Steps

- Fund next 2010 FOA
- NHLBI Nutrition and Diet in Surveillance and Registry Studies of Hemoglobinopathies Meeting
- Hemoglobinopathies Learning Collaborative
- Minimal Dataset Development
- Working Groups
 - Address Challenges
 - Development and refinement of case definition
 - Refinement of indicators

Questions?

The screenshot shows a Windows Internet Explorer browser window displaying the CDC website. The address bar shows the URL <http://www.cdc.gov/ncbddd/sicklecell/index.html>. The page title is "CDC - Sickle Cell Disease, Home - NCBDDD". The main content area is titled "Sickle Cell Disease (SCD)" and features a navigation menu on the left with links such as "SCD Homepage", "Facts", "Symptoms & Treatment", and "Research". The main text area is titled "What Should You Know?" and includes a photograph of three people (two men and one woman) smiling. The text below the photo states: "Sickle cell disease (SCD) is a group of inherited red blood cell disorders. In SCD, the red blood cells become hard and sticky and look like a C-shaped farm tool called a 'sickle.'" and "People with SCD can live full lives and enjoy most of the activities that other people do. If you have SCD, or know someone who does, it's important to learn how to stay as healthy as possible." There is also a "More SCD facts >>" link. On the right side, there are utility links like "Text size: S M L XL", "Email page", "Print page", and "Bookmark and share". At the bottom of the browser window, the taskbar shows the system clock as 9:57 AM on 2/18/2010.

<http://www.cdc.gov/ncbddd/sicklecell/>

