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Projeto Atenção Especializada

Workshop:

Projeto Atenção Especializada para Doença Falciforme

REALIZAÇÃO:

CEHMOB-MG · NUPAD Faculdade de Medicina UFMG · HC UFMG · UFMG · Fundação HEMOMINAS

APOIO:

Prefeitura Belo Horizonte · dreminas · Governo de Minas Saúde · SUS · Ministério da Saúde · Governo Federal Brasil País rico é país sem pobreza

Newborn Screening and the Healthcare System for Sickle Cell Disease in Minas Gerais

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Federal University of Minas Gerais - Brazil

Minas Gerais - Brazil

Minas Gerais - Brazil

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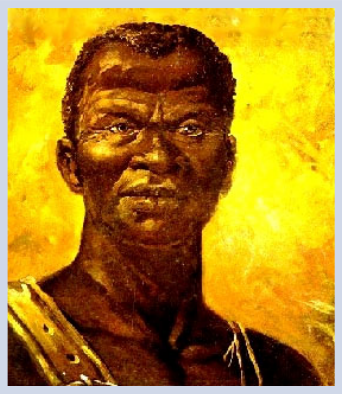
The implementation of newborn screening programs for sickle cell disease in Brazil is strongly connected to the struggle of the African descent for the their health rights, mainly since 1995



Zumbi

Leader of the movement for slave freedom in Brazil (1655-1695).

Background

- **The decision to implement the program in Minas Gerais (1998), the first state universal program for SCD in Brazil, was supported on :**
- The results of the United States SCD experience
- **It also relied on:**
- The previous existence of a well-established state NSP program for PKU and Congenital Hypothyroidism since 1994
- The existence of a hematologic care service for sickle cell disease provided at 12 regional facilities of the State Blood Centers (HEMOMINAS)

Directive No. 822 - June 2001
-Ministry of Health-

- Owing to the significant incidence of sickle cell disease in Brazil, as detected by means of the newborn screening programs carried out in the State of Minas Gerais (1998) and later in the State of Rio de Janeiro (2000) , the Brazilian Ministry of Health introduced a National Screening Program - including newborn screening of hemoglobinopathies - within the scope of the Brazilian Universal Health System (SUS).

- In this context the diagnosis and the treatment are free of charge for all patients with SCD.

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National Brazilian Newborn Screening Program

Implemented in <u>2001</u> for:

- Phenylketonuria
- Congenital Hypothyroidism
- Sickle Cell Disease
- Cystic Fibrosis

Expanded in 2013 for:

- Biotinidase Deficiency
- Congenital Adrenal Hyperplasia

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Programa Nacional de Triagem Neonatal

MINISTRY OF HEALTH POLICY FOR NEWBORN SCREENING PROGRAM

2011 **2013** **2014**

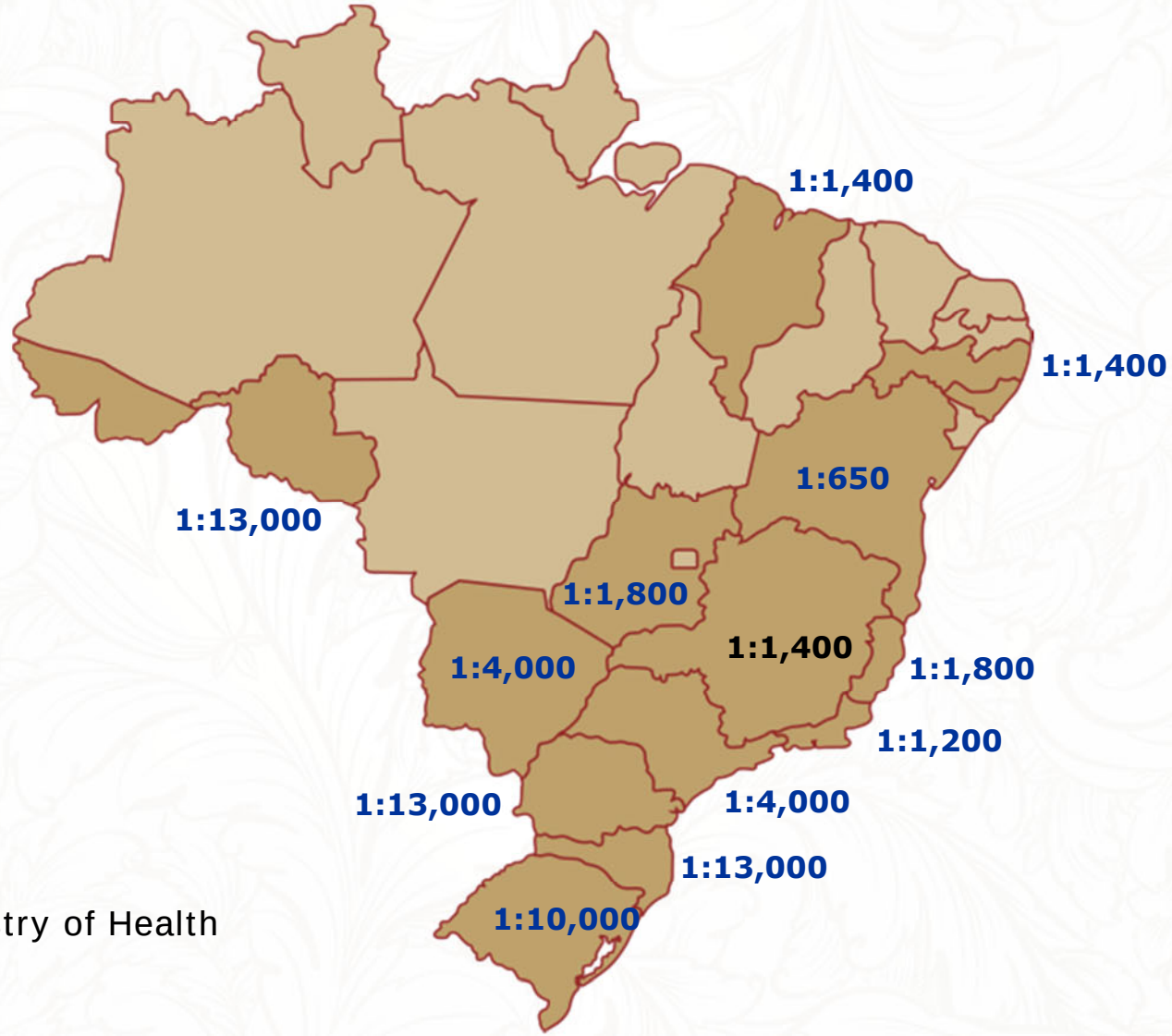
- CH + PKU
- CH + PKU + SCD
- CH + PKU + SCD + CF
- CH + PKU + SCD + CF + BD + CAH

SUS

Ministério da Saúde

GOVERNO FEDERAL BRASIL PAÍS RICO É PAÍS SEM POBREZA

Sickle Cell Disease in Brazil
Incidence in newborns per State



Source: Ministry of Health

SCD Minas Gerais Program

Program sponsored by:

Ministry of Health – Universal Health System (SUS)

Minas Gerais State Health Department

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20 million inhabitants

590,000 km^2

853 counties

Live Births (LB) 2009

CALIFORNIA
527,000 LB

TEXAS
402,000 LB

NEW YORK STATE
248,000 LB

MINAS GERAIS
260,000 LB

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Screened babies sept/1993 to sept/2013		
Congenital hypothyroidism	4,9 million	(1993)
Phenilketonuria	4,9 million	(1993)
Sickle Cell Disease and other hemoglobinopathies	4,0 million	(1998)
Cystic Fibrosis	2,5 million	(2003)
Biotinidase Deficiency	85,000	(2013)
Congenital Adrenal Hyperplasia	85,000	(2013)

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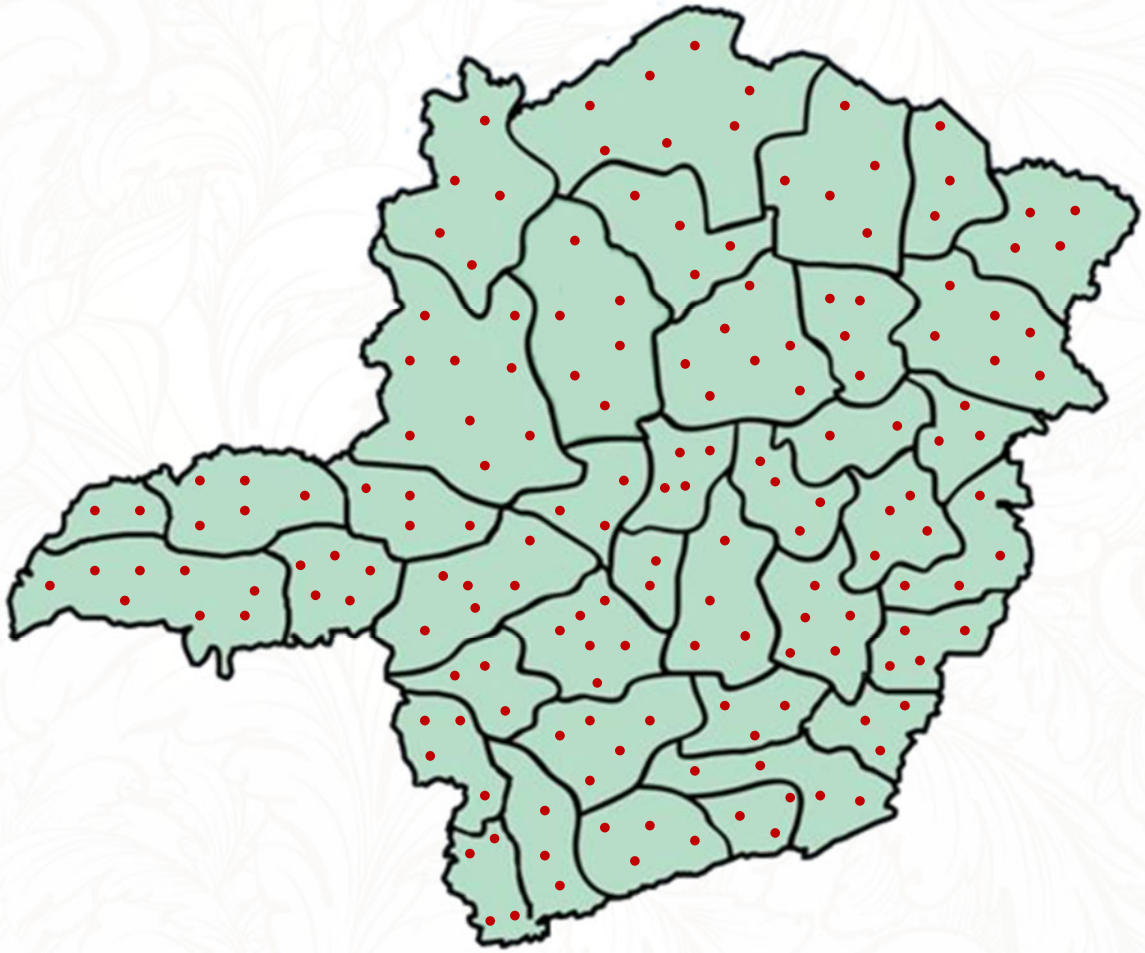
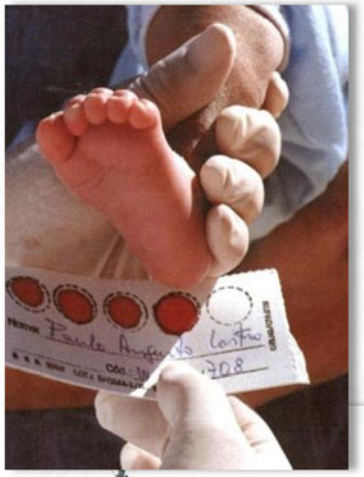
Collection of specimens at the 3th-5th day of life in 2,468 primary care centers throughout the state



Collection (free of charge)



Collections are made in

853 counties

Tests for:

PKU

Congenital Hypothyroidism

Sickle Cell Disease

Cystic Fibrosis

Mail

Single Laboratory - School of Medicine

Laboratory Performance

- A single reference laboratory for all state (20 million inhabitants)
- 1,100 samples/day
- 250,000 samples (children)/year
- Isoelectric Focusing- First Test
- Molecular techniques available at the Genetics Section

School of Medicine - UFMG

Fundação Hemominas

Minas Gerais State Center of Hematology and Blood Transfusion

- **Founded in 1985**
- **23 facilities in Minas Gerais State**
- **260,000 units of blood collection/year**
- **Referral State Service for the treatment of sickle cell disease and hemophilia**

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FUNDAÇÃO HEMOMINAS

Hemominas network: regional units

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Protocols

- Transfusion: phenotype matching of donor red blood cell for red cells antigens C, E and K leukodepletion of red cells
- Hydroxyurea: protocol of Brazilian Ministry of Health
- Stroke: chronic transfusion
- Chelation
- TCD

Teaching Hospital - UFMG

Total capacity: 466 beds

A leader in treatment of medium-and high-complexity diseases, HC/UFMG stands out in several activities, such as oncologic treatments, graft, hemodialisis, chemotherapy, high-risk maternity and nursery, and human reproduction

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Sickle Cell Disease and other Hemoglobynophaties - Minas Gerais

Phenotype	Number
Hb SS	3.319
Hb SC	2.128
Hb Sβ-thal	366
Hb CC	307
Outras	194
Talassemia	123
TOTAL	6.437

Source: Hemominas – Mar/2012

Performance measures

Step	Days (median)
Age at collection	5
Sample transportation (mail)	4
Laboratory processing	2
Age at first appointment for SCD	35

Source: Nupad/UFMG

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SCD follow-up results (1998-2009) (all positive lab results were traced – no lost)

Follow-up features (*)	Number
Under public program treatment	1,841
Under private treatment	23
Transfer to other state program	17
Change of residence (outside Minas Gerais)	44
Treatment interrupted by family	06
Death	145
TOTAL	2,076

Source: Nupad/UFMG

0021-7557/10/86-04/279
Jornal de Pediatria
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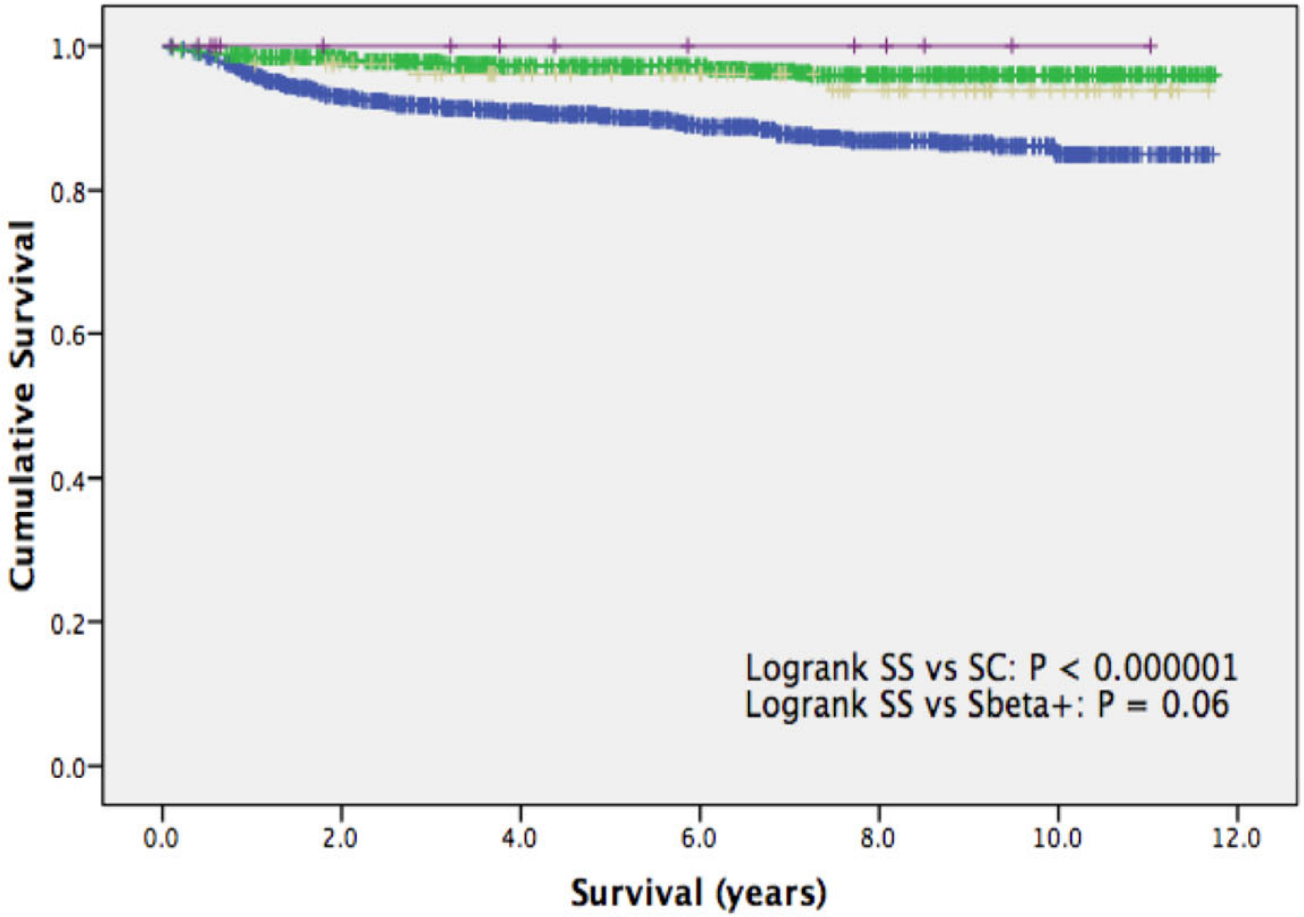
ORIGINAL ARTICLE

Mortality of children with sickle cell disease: a population study

Ana Paula Pinheiro Chagas Fernandes,[1] José Nélio Januário,[2] Camila Blanco Cangussu,[3] Daniela Lino de Macedo,[3] Marcos Borato Viana[4]

Probability survival at 5 years age (includes ALL positive confirmed lab results)



SD = 100%

SC/Sβ0 = 97.7%

$S\beta^+$ = 94.7%

$SS/S\beta^0$ = 89.4%

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Incidence for all types of SCD out of 2.9 million children (newborn screening)

HbS group-specific data	Number	Incidence
SS or Sβ^{o}thal	1,151	1 : 2,530 (0,04%)
Hemoglobinopathy SC	825	1 : 3,530 (0,03%)
Hemoglobinopathy SD	12	--
Probable Sβ^{+}thal	75	--
ALL TYPES SCD	2,072	1 : 1,400

Source: Nupad/UFMG

Center of Education and Aid on Hemoglobinopathies

CEHMOB·MG

Centro de Educação e Apoio para Hemoglobinopatias

Projeto Aninha

Cuidando da Gestante com Doença Falciforme

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CEHMOB
ATENDE

Service provided for all state

ANY DOUBTS ON SCD?

Call:

0800 · 722 · 6500

Toll free

24 hours - 7 days

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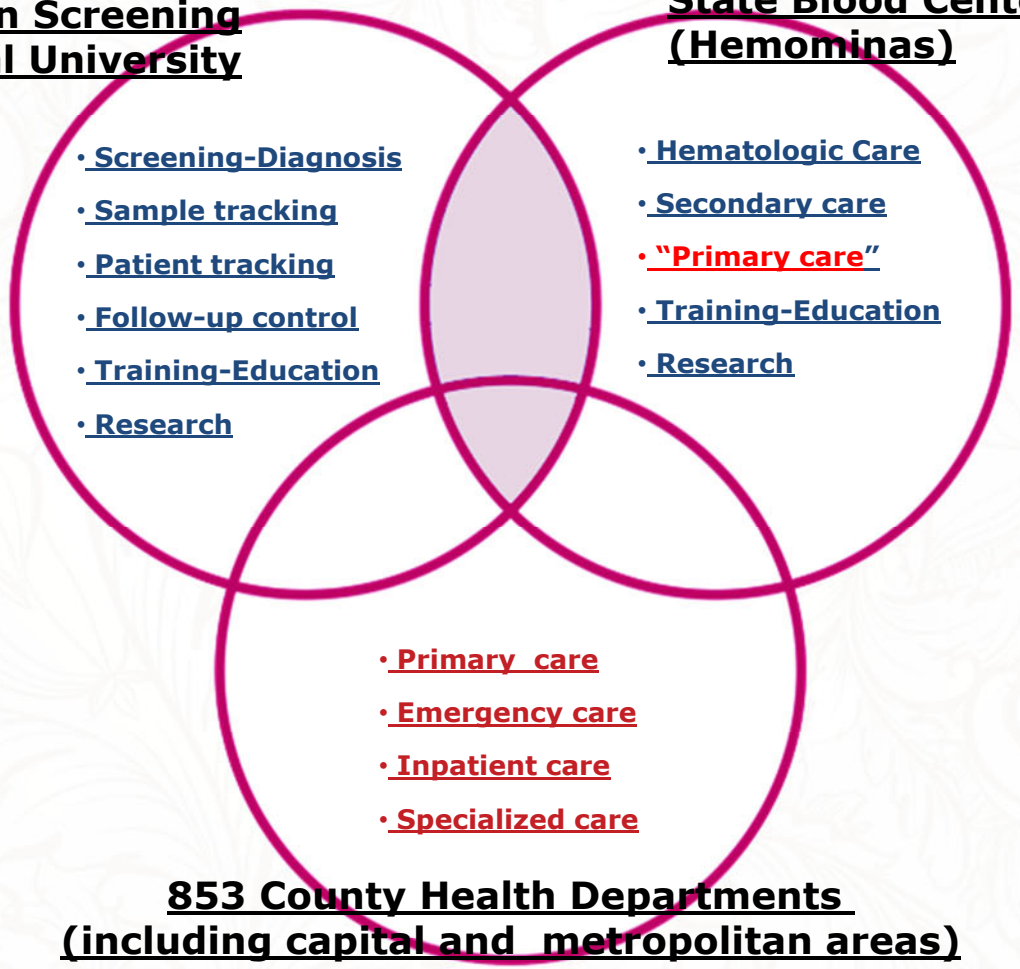
Brazil-Africa Cooperation Project

Ghanaian technicians training at Minas Gerais Program

Diagnosis, tracking and follow-up control



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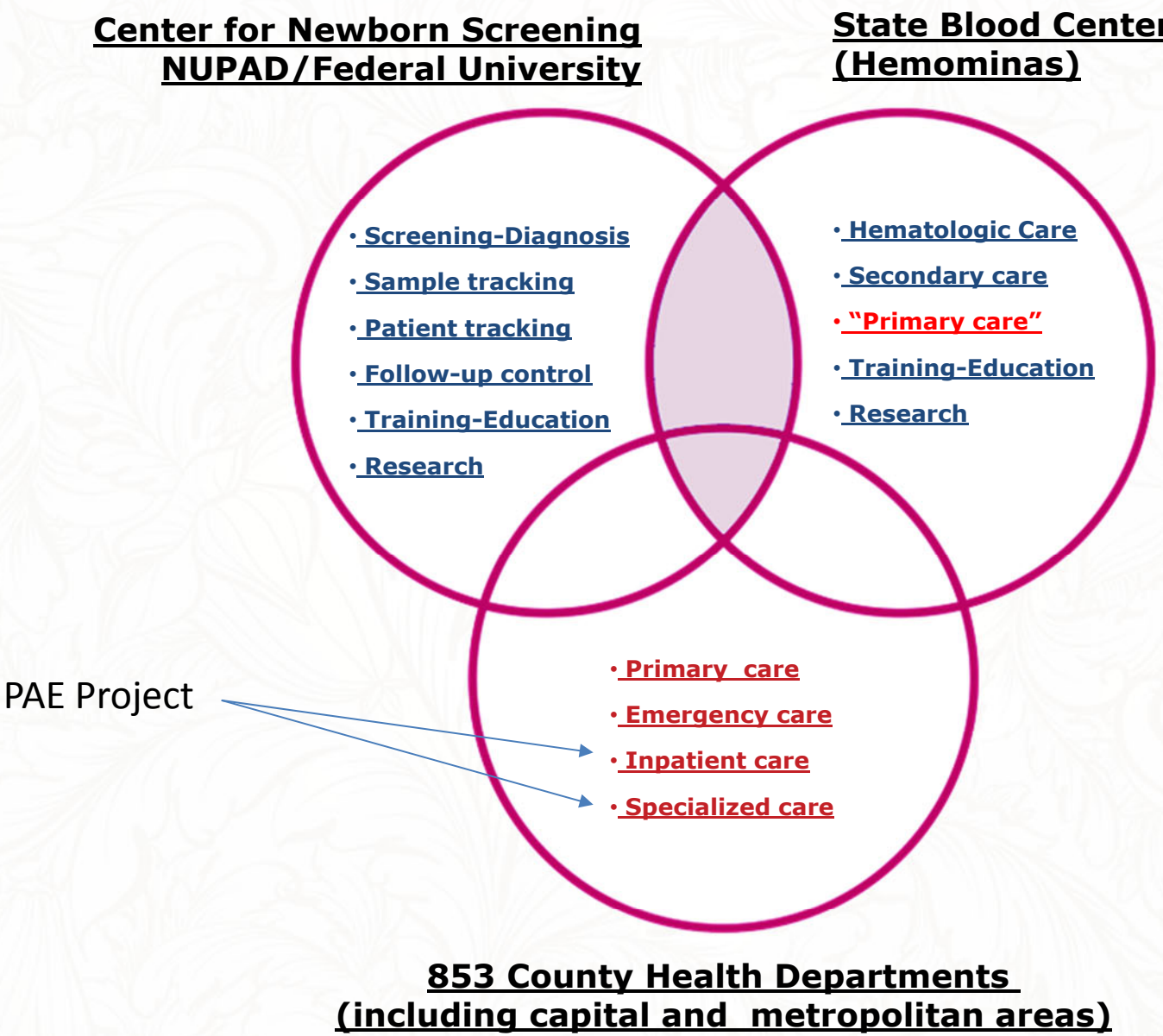
Specialized Care Project for Sickle Cell Disease

Why?

Diagnosis, tracking and follow-up control



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PAE Project (first step:2 years) Goals:

- Critical mass on SCD
- Sharing knowledge on SCD – Electronic Database Support
- Performing Brazilian Protocols
- Comprehensive Care
- Contribute to establish referral services network in Minas Gerais
- Sharing single SCD Database for researching
- Research
- Difusion SCD knowledge at Medical Schools

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PAE Project (first step:2 years) Medical Specialties Selected

- Women's health
- Orthopedics
- Ophthalmology
- Nephrology (October, 2013 workshop)
- Cardiology
- Pulmonology (October, 2013 workshop)
- Neurology (October, 2013 workshop)

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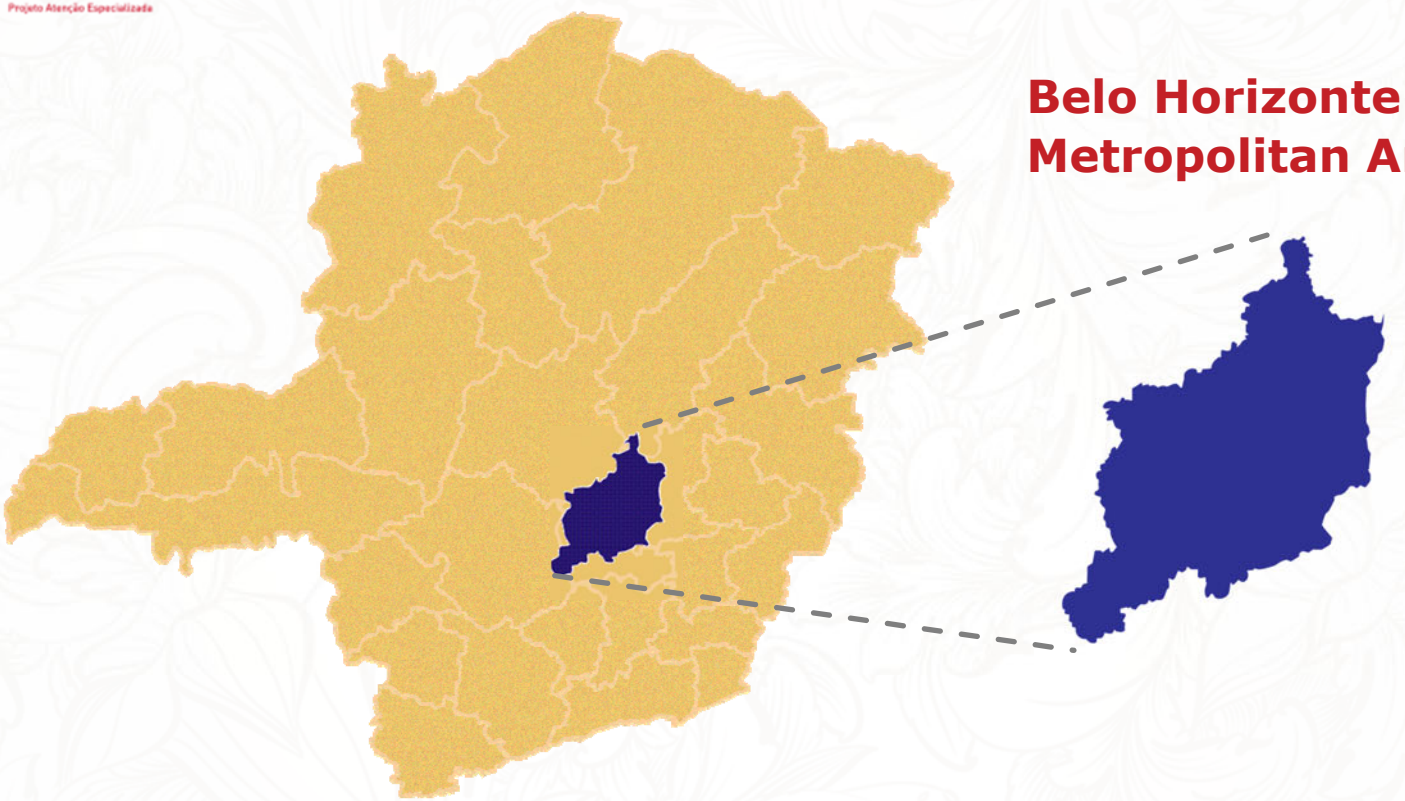


34 counties

4,9 million inhabitants

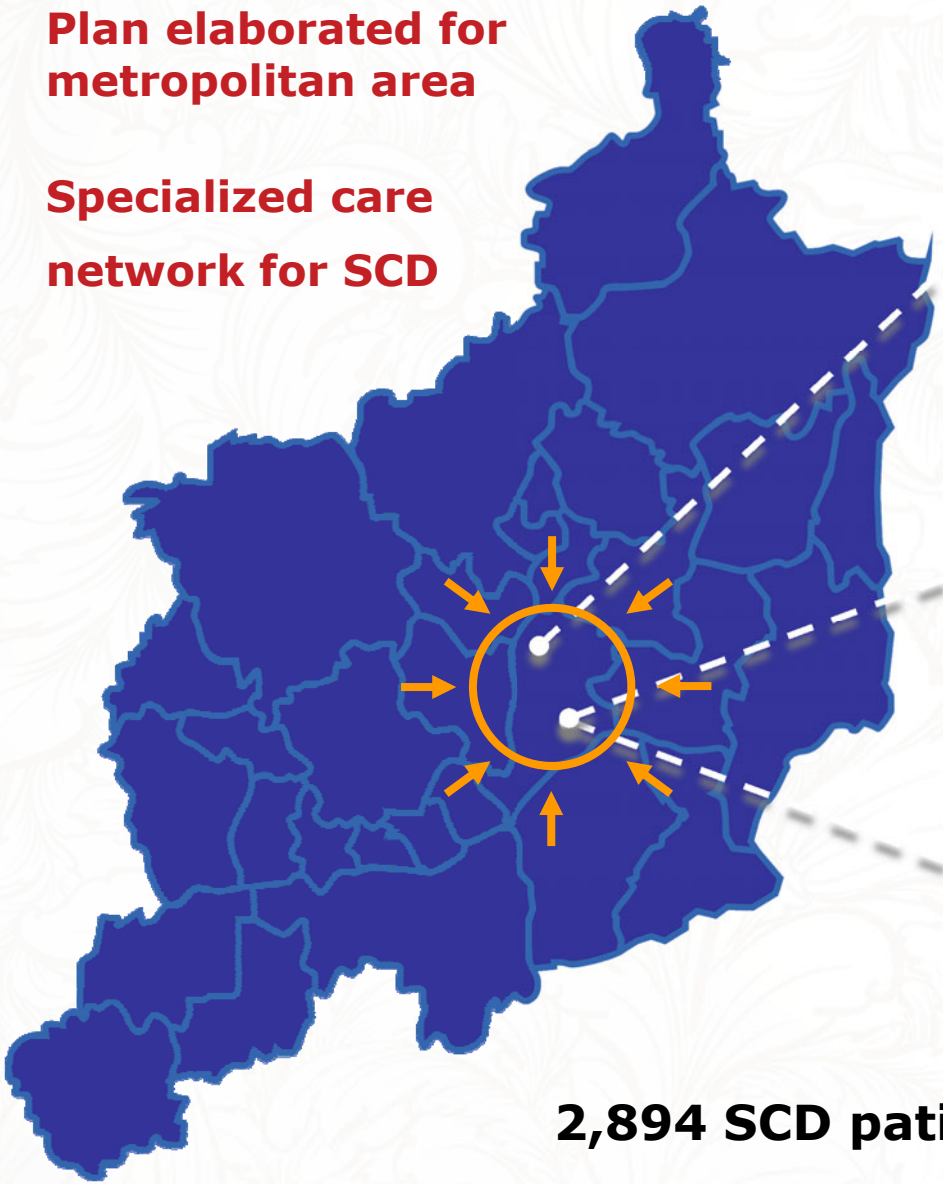
40,000 Km²

(Massachusetts + New Jersey area)

FIRST STEP

Plan elaborated for metropolitan area

Specialized care network for SCD



2,894 SCD patients

SECOND STEP

Expanding experience to whole Minas Gerais state

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EXPLORING EUA-BRAZIL COLLABORATIVES OPPORTUNITIES ON RESEARCH

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FUNDAÇÃO HEMOMINAS

Background

- **Large number of infants diagnosed yearly with sickle cell disease by Newborn Screening Program in Minas Gerais (around 180; 120 SS)**
- **Regular follow-up for almost 100%; half at the state capital (Belo Horizonte)**
- **Mixed ethnic background different from American Afro-descendents**
- **Laboratory facilities for molecular and genetic studies**
- **On going translation and dissemination to community through educational programs and practical guidelines**

Welcome to the Brazilian Symposium for Sickle Cell Disease and Other Hemoglobinopaties

Salvador – Bahia. November, 2013

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REALIZAÇÃO:

APOIO:

29 e 30 de Outubro, 2013.

Belo Horizonte, MG, Brasil.

http://www.cehmob.org.br