GREETINGS
from
Gujarat

Vignettes from
Sickle Cell Anemia Control Program

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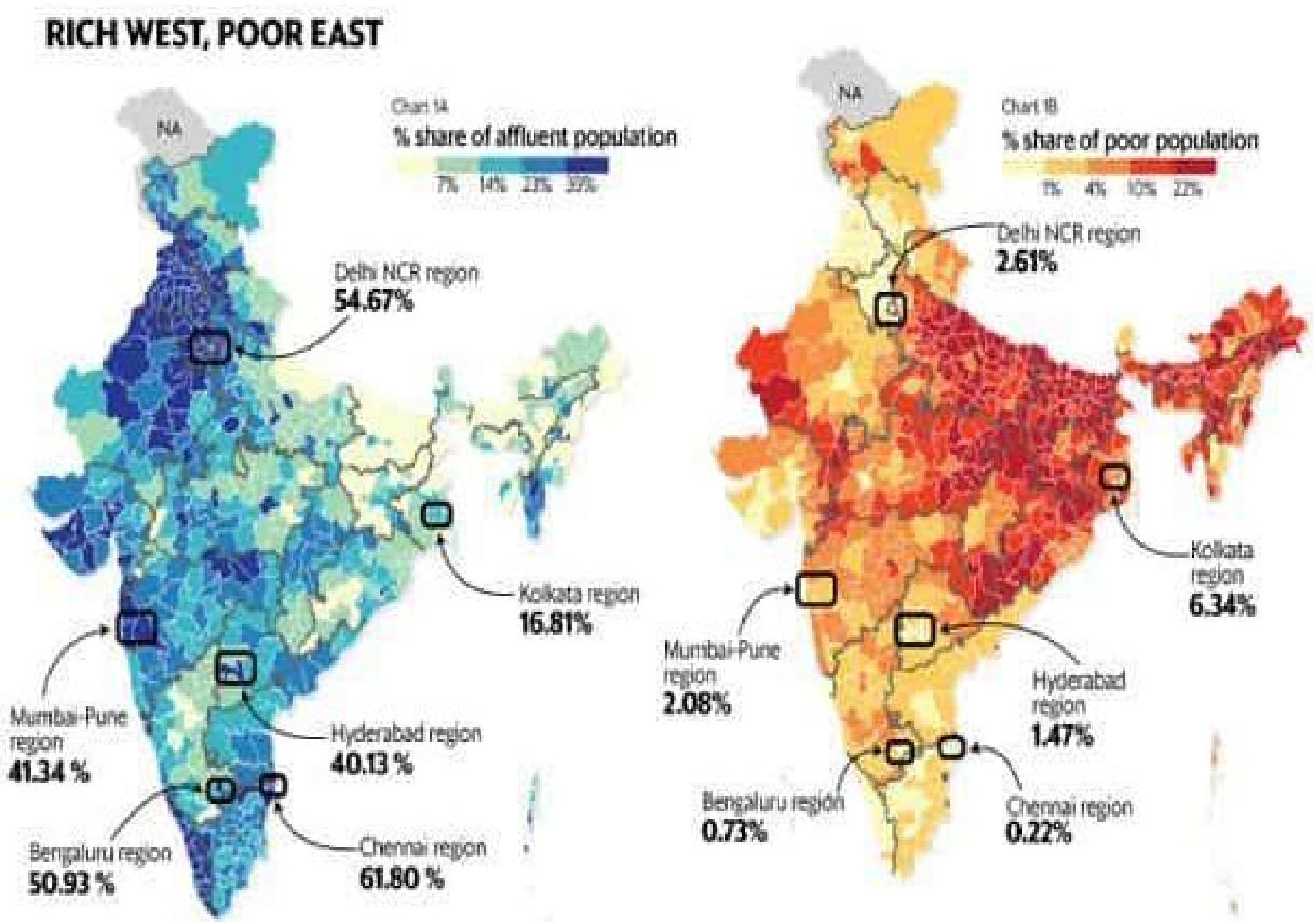
Background



14% population tribal in Gujarat; 45% below the poverty line.

Poor, illiterate tribals residing in underserved remote areas will never visit a pathological lab for their blood tests by spending their own money **and**

Since they are not tested for Sickle Cell, they are ***misdiagnosed and mis-treated*** as the clinicians unaware of the existence of Sickle gene amongst them.

Hence the need for a public health programme for control

Clinical Presentation

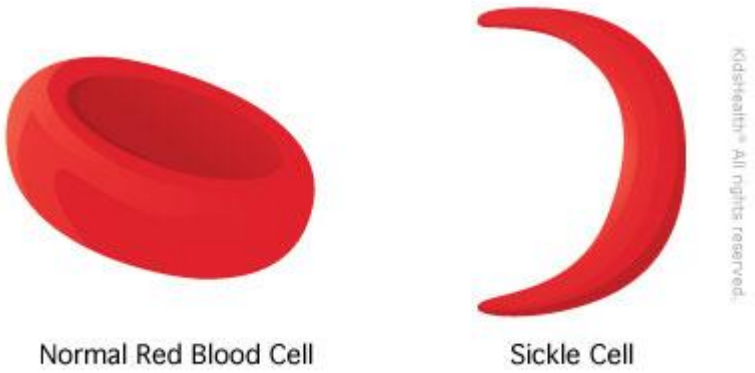
Acute chest syndrome	Aplastic crisis
Hand-foot syndrome	Infection
Priapism	Stroke
Splenic sequestration crises	
leg ulcers, *bone or joint damage*, gallstones, *kidney damage*, eye damage, and *delayed growth*.	

Symptoms	%
Painful crisis	90%
Severe anemia	83%
Fever	37.5%
Hematuria	5.0%
Neurological complain	5.2%

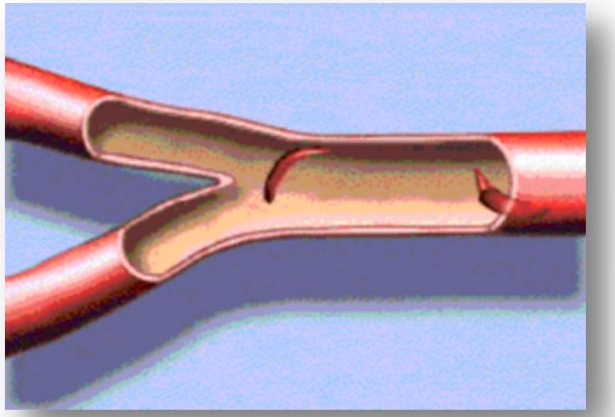
Signs	%
Pallor	100 %
Spleenomegaly	78%
Hepatomegaly	70%
Joint swelling and bony tenderness	55%
Gall stones	10 %
Osteomyelitis	5%
Avascular necrosis of hip	3%
Retinopathy	38%
Transient visual loss	2.5%
Stroke	4.5%

What Is Sickle Cell Disease?

Shape of cells



- Sickle cell disease is a condition in which red blood cells are not shaped as they should be. Red blood cells look like round discs. But in sickle cell disease, they're shaped like sickles, or crescent moons, instead.



- These sickle shaped cells get stuck together and block small blood vessels. This stops blood from moving as it should, causing pain and organ damage.

Malaria and sickle cell disease connected?

- **How does sickle cell trait protect against malaria?**
- Malarial parasites invade normal red blood cells and rearrange their content. The parasites breed and produce proteins that make red blood cells sticky. These blood cells explode, releasing parasites capable of infecting other red blood cells.
- The misshapen hemoglobin of SCT affects a parasite's ability to complete this cycle. The parasite triggers the SCT hemoglobin to sickle. The immune system then clears the infected red blood cells before the parasite can complete its life cycle and infect other red blood cells. This means fewer parasites and milder illness.
- People with SCT are not as affected by malaria compared to those with normal hemoglobin. During infection, those with SCT have 50 to 90 percent fewer parasites in their blood than people with normal hemoglobin. People with SCT also get rid of the parasites faster.
- People SCT have:
    - milder cases of malaria –
    - lower hospital admissions
    - Have lower rates of blood transfusions
    - Are less likely to die from malaria

What Causes Sickle Cell Disease?

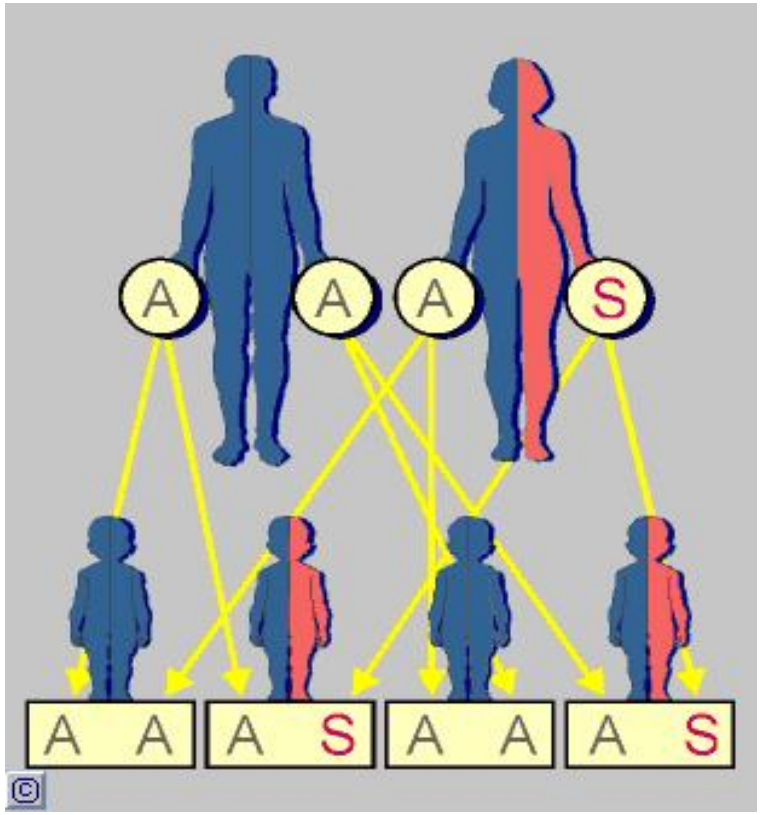
- Sickle cell disease is a genetic condition. ***People who have it inherited certain hemoglobin genes from their parents***. Hemoglobin is the protein inside of red blood cells that carries oxygen. *Abnormal hemoglobin makes the red blood cells sickle shaped*.

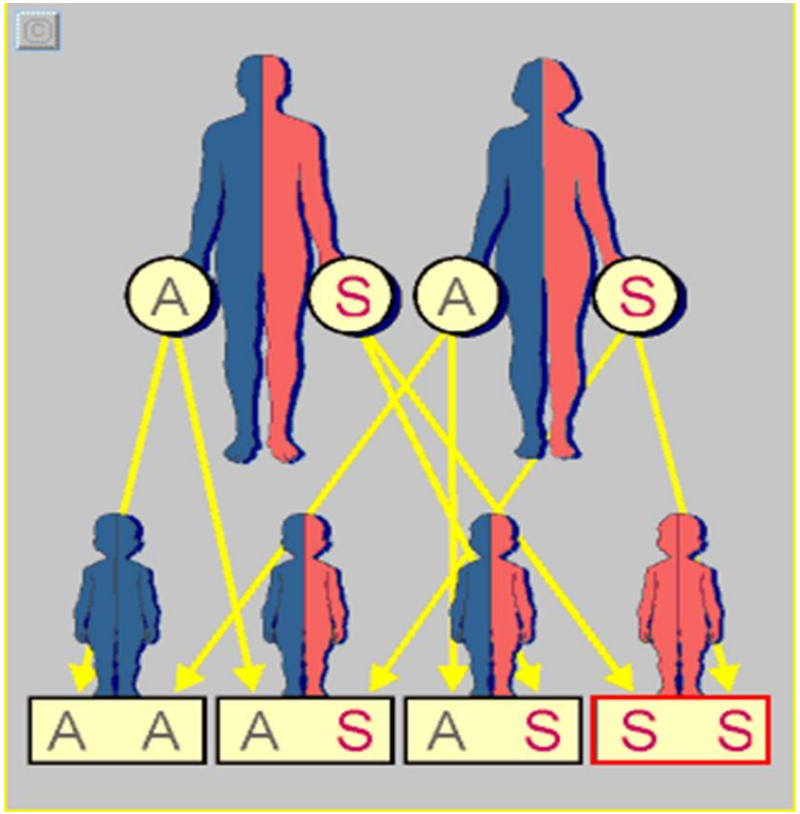
- Someone who inherits a sickle cell gene from each parent has **sickle cell disease**.

- Someone who inherits a sickle cell gene from one parent and a normal hemoglobin gene from the other has ***a sickle cell trait,*** rather than sickle cell disease. Most people with sickle cell trait don't have symptoms, but can pass the gene to their children.

- Someone who inherits a sickle cell gene from one parent and another kind of abnormal gene from the other parent may have a different form of sickle cell disease, such as hemoglobin SC disease or ***sickle beta thalassemia***.

Inheritance





Why is it critical for us to understand this condition?

Estimated Total Population of India (2021)	Total Tribal Population of India @ 8.6%	Suspected Sickle Cell Trait -Carriers @ 7.46 %	Suspected Sickle Cell Disease Patients @ 0.30 %
139 Cr.	11.95 Cr.	**89.70 Lakhs**	**3.58 Lakhs**
Approx. New Births Every year in India @17.64 % decadal growth			
	New Tribal Births 21.08 Lakhs	**New Sickle Cell Trait Birth 1.57 Lakhs**	**New Sickle Cell Disease Births 6,324**

Estimated Prevalence of SCA in India considering Mass Sickle Cell Screening data of Gujarat

How Is Sickle Cell Disease Treated?

- People with sickle cell disease can lead fairly normal lives if they follow their treatment plan, which involves;
- Immunizations and daily doses of penicillin to help prevent infection. Kids with sickle cell disease should get all recommended vaccinations, including the pneumococcal, flu, and meningococcal vaccines.
- Folic acid supplements, which can help kids make new red blood cells.
- Hydroxyurea, a medicine that makes the cells less sticky. This helps decrease the frequency and intensity of painful episodes and other complications. Hydroxyurea is taken every day.
- L-glutamine, another medicine. It's used if hydroxyurea isn't working well or someone still has pain crises even with hydroxyurea.
- Medicines to help manage pain when it does happen.
- Blood transfusions for severe anemia & other complications.
- Stem cell transplant (also called bone marrow transplant) is the only known cure for sickle cell disease. Transplants are complex and risky, and for now are an option only for some patients.
- Scientists are studying gene therapy for sickle cell anemia. One day, it's hoped that doctors can stop the disease by changing or replacing the abnormal gene that causes it.

The Government of Gujarat initiative

Sickle Cell Anemia – Gujarat Model

Valsad Rakt Dan Kendra at the forefront of this Movement

Catalysts for this initiative

Transforming Projects Into Programmatic Mode.

A journey of 16 years from 2006 to 2022

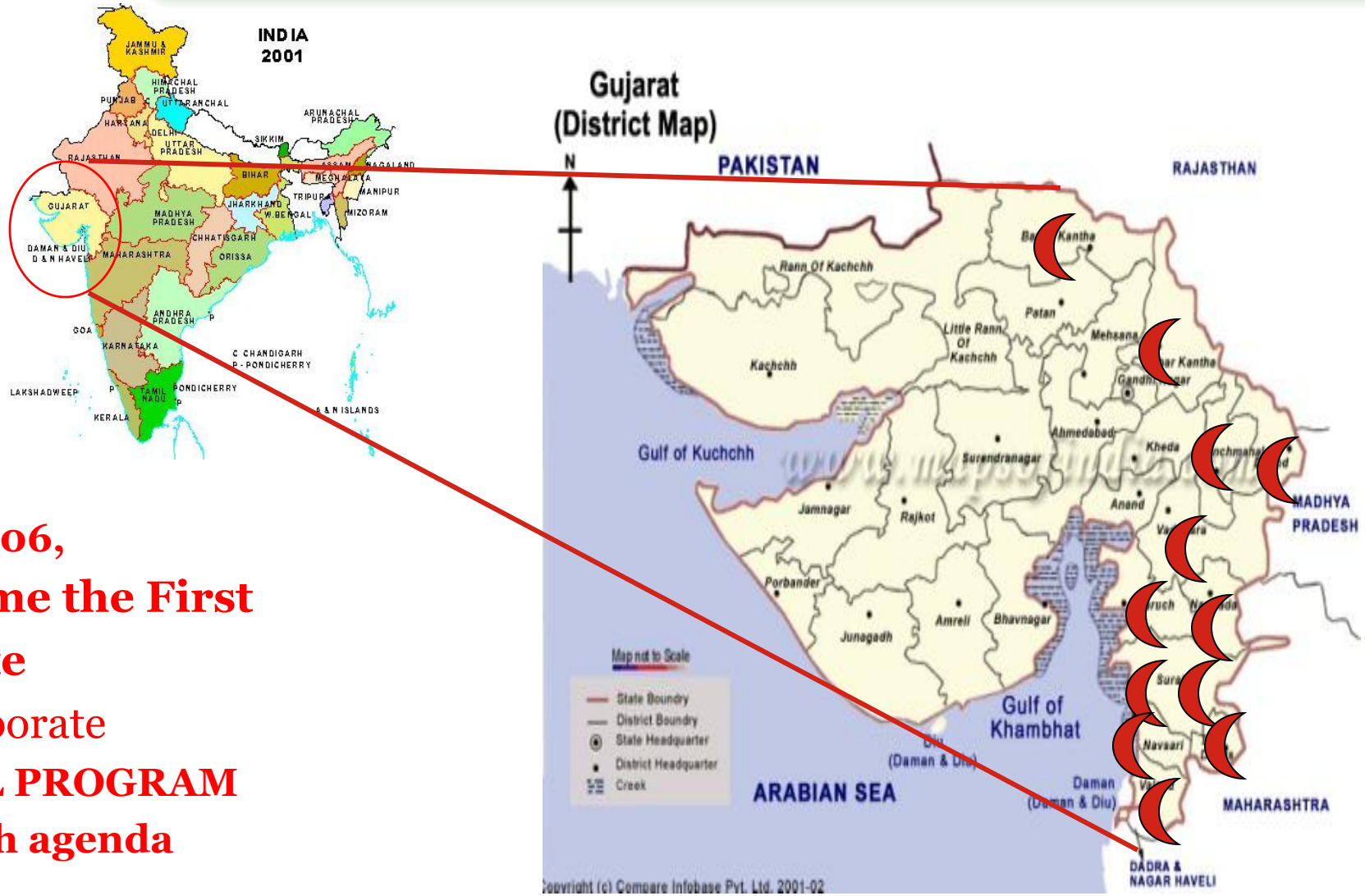
- **1910 : James Heric**
- 1952 Lehmann & Cutbush in Nilgiris tea estates
- **1978 - Dr Italia VRK identified / diagnosed SCD patient at Valsad,**
- **2006: Sickle Cell Anemia Control Program of Gujarat State in PPP mode with VRK**
- 2022

Bringing Science to the Door Step of Tribal Community.....

Sickle Cell Anemia – Gujarat Model

Sickle Cell Anemia is a major tribal health issue in tribal areas of Gujarat.

14 Tribal districts are affected by the Sickle Cell Anemia issue



On 30th July 2006, Gujarat became the First State to incorporate SCA CONTROL PROGRAM in the health agenda

GOAL:

- Prevention of Sickle Cell Disease Child Birth
- Prevention of Death from Sickle Cell Crises
- Improve Health Status and Quality of Life of Sickle Cell Disease patients

OBJECTIVES: Screening; Counseling; IEC program & Convergence with other programs

The Gujarat Model of Sickle Cell Anemia Control Programme

Gujarat State SCA Control Society formed

- Every district EMO is a Nodal Officer for SCA Control Program.
- 180 Sickle Cell Counselors: (Almost one counsellor amongst two Primary Health Centers) in tribal districts.
- Telephonic contact with each SCD patients and monthly visits by counsellors.

Good rapport between the patients and counselors

During hospital visits & crises situations

- Training and re-training of health care personnel.
- IEC in targeted population.
- PPP and NGO involvement.
- Free Mass Screening of Population - Diagnosis, Counseling & treatment.
- Routine medicines like Folic Acid, Analgesics and Hydroxyurea are provided at the door steps through MPHWs in the field.
- Routine investigations are carried out at CHCs and General Hospitals free of cost
- Marriage Counseling and prevention by PND.
- 108 Free Ambulance services made available.

Progress of Sickle Cell Anemia Control Program - Gujara

2006	• Program started in 5 districts of South Gujarat on PPP basis (**Surat, Tapi, Navsari, Valsad and Dang)**
2008	• Extended to remaining 9 tribal districts of Gujarat (Dahod, Narnada, Chhotaudepur , Panchmahal ,Bharuch, Sabarkantha, Banaskantha, Aravalli, Mahisagar)
2011	• Gujarat Sickle Cell Anemia Control Society formed to integrate efforts by different departments
2012	• Screening outsourced to 7 qualified, competent & dedicated competitive agencies – VRK, Red Cross, Indu blood bank*
2015	• Prenatal Diagnosis initiated
2017	• Pneumococcal Vaccine given to all SCD patients
2021	• Treatment care center started at **Surat & Valsad**
2022	• Sickle Cell Day Care Center Started at **Valsad, Navsari & Dang**

* 2015-2016 entire tribal population screened

Journey of SCACP- Gujarat over Decade

(Facilities made available)

No.	Facilities available	2007-08	2010	2022
1	Primary Screening for Sickle Cell (DTT*) test, Counseling and Treatment	78	419	2771
2	HPLC based Hb Variant system for quantitative estimation of different hemoglobin	2	3	7
3	Day Care Center (Dedicated Treatment Centers)	0	0	3
4	Molecular Lab for prenatal diagnosis and Genetic Counseling Center	0	1	3
5	Dedicated Sickle Cell Counselor	0	5	180

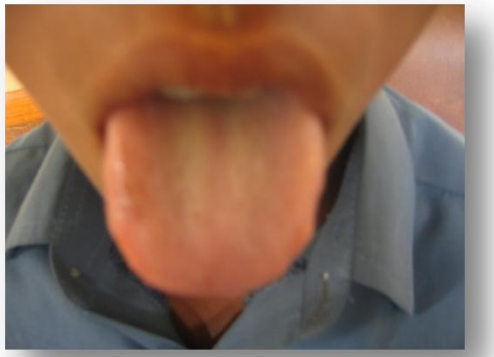
- **Convergence with Tribal and other line department… Training of teachers and students in Ashram Shalas and College in tribal blocks**
- **Rs 500 PM for each SCD patient and disability certificate issued; all complications treated under PMJAY along with free blood transfusion**

All services at Comprehensive Sickle Cell Clinic (CSCC) provide free of cost to tribal/rural patients at 4 CHCs.

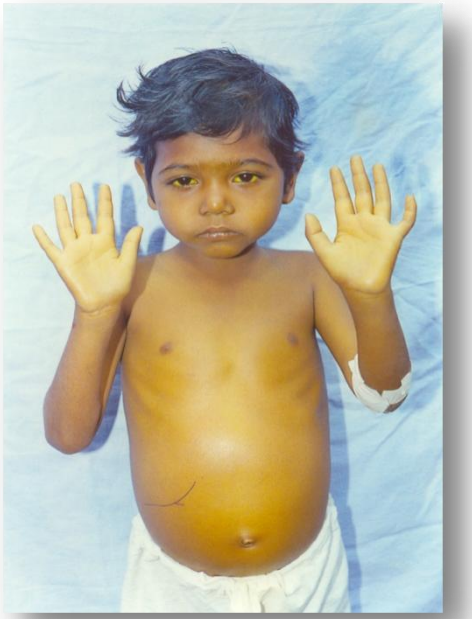
- Primary Screening
- Marriage counseling,
- Counseling of Diseased patients,
- Treatment
- Routine follow up
- Clinical Evaluation by MOs
- High end management of Sickle cell crisis and chronic complications.
- Referral Services
- Pre Natal Diagnosis (PND)
- New Born Screening (NBS) Collection
- Free Blood Transfusion
- Expert doctors & yearly Medical Camps for SCD patients.
- Monthly Visit of Hematologist at each center.

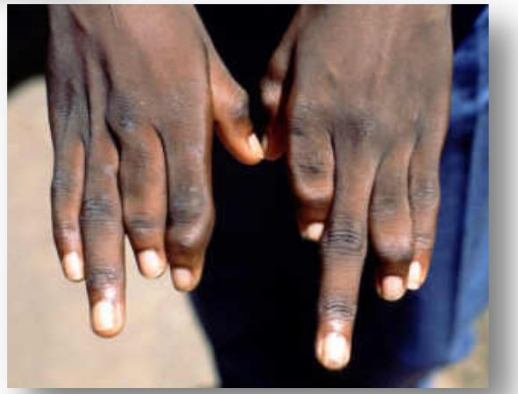
Awareness generation

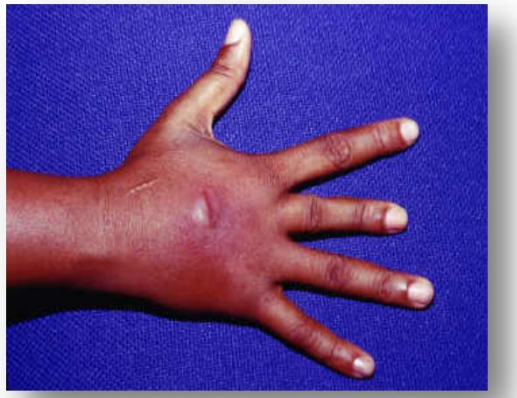
Posters on Common Symptoms of Sickle Cell Anemia



- Pallor
- Frequent jaundice
- Bone & Body ache
- Enlarged Spleen
- Retarded Growth
- Frequent Infections
- Dectilytis







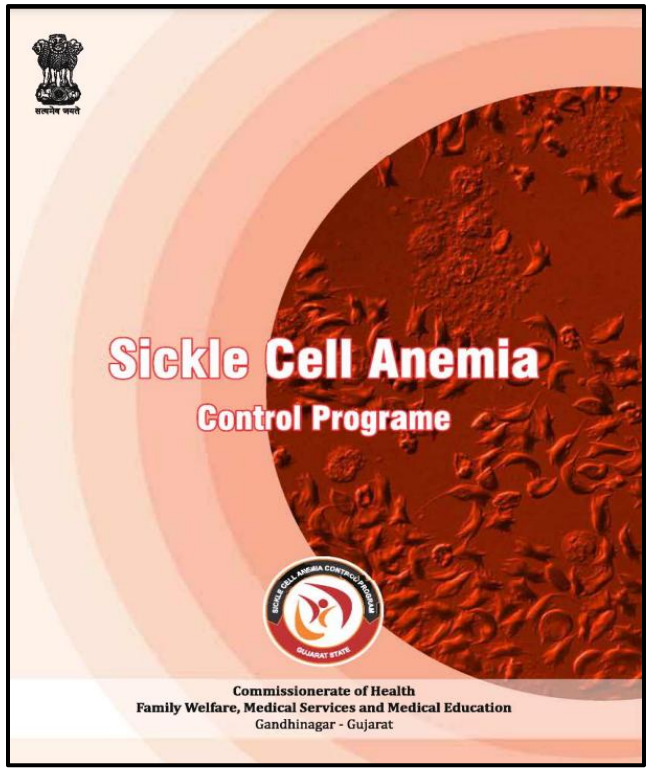
સિકલ સેલના દર્દીએ લેવાની કાળજી

શું કરવું જોઈએ ?

શું ન કરવું જોઈએ ?

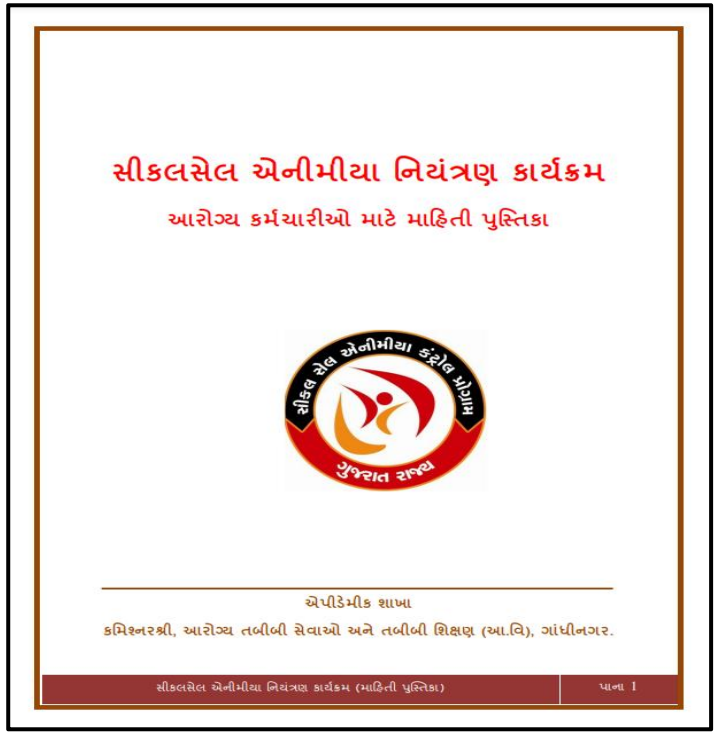
સિકલ સેલ ધરાવનાર વ્યક્તિ પણ લાંબુ આયુષ્ય ભોગવી શકે છે.

પ્રકાશક : કમિશ્નર, આરોગ્ય અને પરિવાર કલ્યાણ વિભાગ, ગાંધીનગર

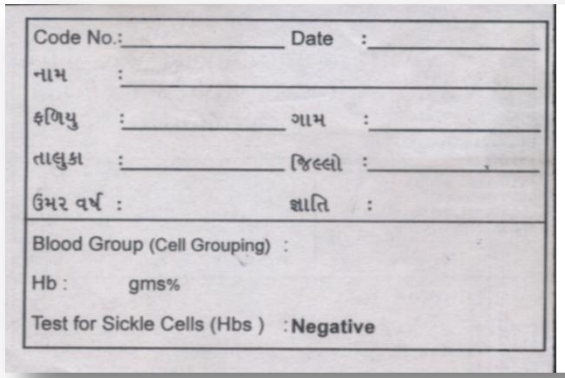


Program Manuals

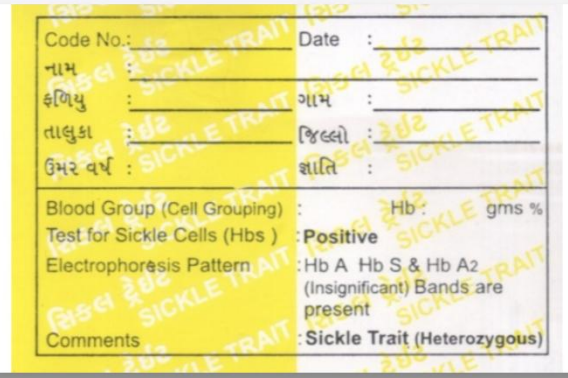
- Treatment Guideline for Medical Officers at different levels PHC/ CHC and DH.
- Manuals for HCWs/ ASHA in vernacular language, for home management.
- Management of perinatal care of SCA pregnant women



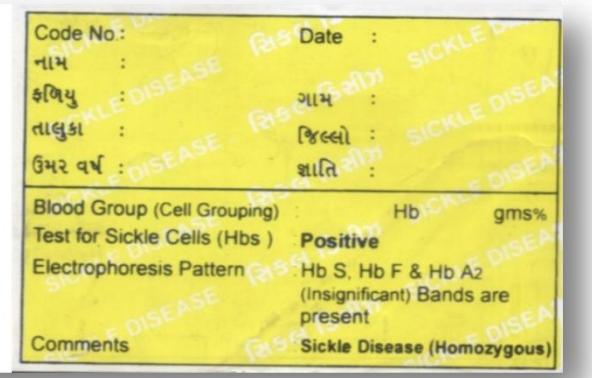
Sickle Cell Anemia – Gujarat Model



Normal Haemoglobin



Sickle Cell Trait



Sickle Disease

Laminated color coded cards were given to all screened persons.

These color coded cards are further used for marriage counseling that the two yellow card people should avoid marriage. Even priests advise this

Pink colored card given for Thalassemia

Genetic Counseling

- Any family, who had come across with Sickle Cell Disease Child, does not wish to have another such child in their family.
- They do come forward for marriage counseling for other family members.
- The majority of population in South Gujarat, i.e. Dhodia Patel, Gamit & Choudhary ***understand the gravity of the Sickle Cell Disease*** and if explained in friendly manner, willing to improve their health status.
- Willingly follow advice.

Programme achievements

Sickle Cell Anemia – Gujarat Model

Results of *Mass Sickle Cell Screening* in 14 Tribal Districts of Gujarat up to March 2022

Total Tribal Population	Population Screened	Sickle Cell Trait	%	Sickle Cell Disease	%
1,40,26,563	96,81,695	7,22,645	7.46	29,555	0.3

Overall Sickle gene incidence in tribal population is 7.76 % down from 10-12% 2010-11.

Severity of the disease reduced down from 1 – 0.3%

Screening Output- 2 Screening of Antenatal Mothers

	2011	2021- 2022
No. of Antenatal Mothers Screened for Sickle Cell Anemia	**2,13,779**	**1,50,581**
No. of Antenatal Mothers found Sickle Cell Trait	**15367 (7.18%)**	**2545 (1.69%)**
No. of Antenatal Mothers found Sickle Cell Disease	**848 (0.44%)**	**169 (0.11%)**

Screening Output- 3 Counselling

	Upto March- 2022
No. Prenatal Diagnosis Done	4008
No. Antenatal eligible for MTP after PND	1227
No. of MTP done (SCD child birth prevented)	613

Sickle cell Pneumococcal Vaccine
(Up to March – 22)

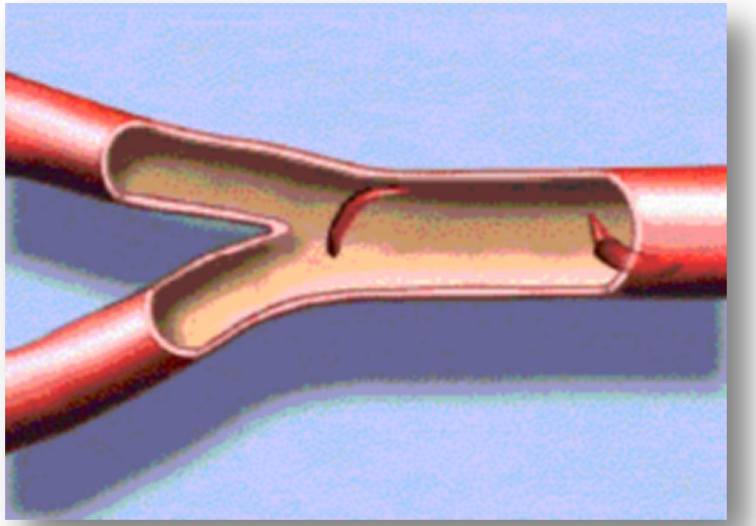
Sr.No.	District	Total Disease Patient	Vaccine Given
1	Surat	2507	2159
2	Dahod	4878	4693
3	Narmada	1565	1411
4	Tapi	3271	2617
5	Valsad	2519	2415
6	Navsari	1761	1761
7	Chhotaudepur	6090	5960
8	Panchmahal	2771	2671
9	Dang	385	361
10	Bharuch	1276	1091
11	Sabarkantha	973	725
12	Banaskantha	386	323
13	Arvalli	272	271
14	Mahisagar	339	334
	Total	28993	26792

Lessons from the SCA program

- Intensive IEC
- Active involvement of NGOs.
- Community involvement.
- Need to reach the most vulnerable tribal population
- Lifecycle approach.
- Capacity building of both Government and NGO staff
- Key role of counsellors
- Integration with Van Bandhu programme.

Way forward: **Centre of excellence for SCA at GMC Surat; Comprehensive day care center in each taluka under Van Bandhu Yojna – 1000 crore set aside;**

NHM has adopted Gujarat model



Our Sicklers are

Born with Pain

Live with Pain

&

Die with Pain.

But they hardly Complain.

Thank you

for paying attention to them.

What Problems Can Happen?

- **Acute chest syndrome**: Caused by inflammation , infection, and blockages of small blood vessels of the lung. Signs include chest pain, coughing, trouble breathing, and fever.
- **Aplastic crisis**: This is when the body temporarily does not make enough red blood cells, and can cause ***severe anemia***. Signs include paleness, extreme tiredness, and a fast heartbeat.
- **Hand-foot syndrome**: This painful swelling of the fingers and toes (also called ***dactylitis***) is the first sign of sickle cell anemia in some infants.
- **Infection**: Kids with sickle cell disease *are at risk for some bacterial infections*. It's important to watch for fevers of 101°F (38°C) or higher, which can be signs of an infection.
- **Priapism**: Males with sickle cell disease can have painful, long-lasting erections. If it's not treated quickly, damage can cause problems with getting erections later on.
- **Splenic sequestration crises**: The spleen traps the abnormal red blood cells and gets very large. This can lead to a ***serious, quick drop in the number of red blood cells in the bloodstream***. Signs include paleness, weakness or extreme tiredness, an enlarged spleen, and belly pain.
- **Stroke**: Sickle-shaped cells can block small blood vessels in the brain, causing a stroke. Signs include headache, seizures, weakness in the arms and legs, speech problems, a facial droop, or loss of consciousness.
- People with sickle cell disease are also at risk for problems such as **leg ulcers, *bone or joint damage*, gallstones, *kidney damage*, eye damage, and *delayed growth***.

SCA TOTAL SCREENING UPTO MARCH 2022

SR NO	DISTRICT	TRIBAL POPULATION	COVERED POPUPATION	COVERED POPUPATION %	SCD	SCT
1	SURAT	771813	745674	96.61	2627	19376
2	DAHOD	2355496	2544195	108.01	4878	229055
3	NARMADA	549030	549030	100.00	1562	40811
4	TAPI	810796	750324	92.54	3271	65526
5	VALSAD	999639	956237	95.66	2519	80947
6	NAVASARI	749887	775307	103.39	1761	45391
7	CHHOTAUDEPUR	957938	889792	92.89	6090	92375
8	PANCHMAHAL	762444	790039	103.62	3277	63074
9	DANG	289061	292990	101.36	385	11102
10	BHARUCH	325768	270554	83.05	1276	15795
11	SABARKANTHA	351739	338274	96.17	973	27147
12	BANASKANTHA	191184	214772	112.34	384	17229
13	ARAVALLI8	265477	271124	102.13	333	4007
14	MAHISAGAR	394239	238702	60.55	339	10810
	TOTAL	**9754511**	**9681695**	**99.25**	**29555**	**722645**