TEXAS
Health and Human Services

Texas Department of State Health Services

Newborn Screening Clinical Care Coordination

Sickle Cell Task Force Meeting

Karen Hess, Director

Newborn Screening Unit

August 18, 2023

Goals of Texas Newborn Screening (NBS) Program

- Two screening tests for each baby born in Texas at 24 – 48 hours of age and at 1 – 2 weeks of age.
- Notify Primary Care Provider (PCP)/Parents of abnormal screens same day or within 24 hours.
- Provide recommendations for appropriate follow-up including confirmatory testing.
- Follow patients for up to 18 years of age.

Clinical Care Coordination (CCC) Genetics Branch Organization

- Three Disorder Specific Teams
  - Hemoglobin (HGB) / Cystic Fibrosis (CF) / Severe Combined Immunodeficiency (SCID)
  - Endocrine / Biotinidase
  - Metabolic / X-linked Adrenoleukodystrophy (XALD) / Spinal Muscular Atrophy (SMA)
- Long Term Team within each group
- Hemoglobinopathies Team:
  - Manager - Selda Rios
  - Team Lead - Julianna Ybarbo, RN
  - Hemoglobin Nurse - Virginia “Star” McNeely, RN
  - Trait specialist - Michel Olivier
  - Long term specialist - Jess Fields

Newborn Screening/Hemoglobin Workload 2019-2022

Specimens reported with *Presumptive Positive* results that required follow-up by Clinical Care Coordination

Determination	2019	2020	2021	2022
HGB Non-Sickle	70	80	75	61
HGB Other	28	9	14	15
HGB Sickle	425	362	353	413
Subtotal	523	451	442	489
Sickle Cell Trait	11,278	11,222	11,397	11,808
Grand Total	**11,801**	**11,673**	**11,839**	**12,297**

Presumptive positives are specimens

Abnormal Hemoglobin CCC Notification Process

- Abnormal Hemoglobin screen from the laboratory
  - CCC staff call and fax physician same day
  - Send letter to parent
  - Wait for DNA
- DNA results released from laboratory (usually 1- 2 weeks)
  - CCC call and fax DNA letter to physician
  - Send parent packet (letter and brochure)
- Enter diagnosis information and upload forms
- Move to Long Term (LT) team

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Diagnosed Hemoglobinopathies, January 2017-2022

Diagnosis*	2017	2018	2019	2020	2021	2022
HGB – S Beta Plus Thalassemia	19	17	8	26	17	15
HGB – S Beta Zero Thalassemia	2	1	2	3	3	3
HGB – SC Sickle C Disease	45	55	54	57	46	54
HGB – Sickle Cell Disease with HPFH	1	1				
HGB – SS Sickle Cell Anemia	126	123	143	90	101	127
Total Core Recommended Uniform Screening Panel (RUSP)	**193**	**197**	**207**	**176**	**167**	**199**
HGB – C Beta Thalassemia	5	6	8	9	5	6
HGB – CC Hemoglobin C Disease	13	5	13	9	9	8
HGB – D/Beta Thalassemia	1			1	1	
HGB – DD Disease				1	1	
HGB – E Beta Thalassemia			2	2	5	1
HGB – EE Hemoglobin E Disease	8	9	6	7	9	6
HGB – H Disease	7	6	12	4	6	5
HGB – Homozygous Beta Thalassemia	1	5		6	2	
HGB – Other	13	8	4	3	5	5
HGB – Other Sickle	1	4	5	2	9	6
HGB – Sickle E disease	1	1	1			
Total	**243**	**241**	**258**	**220**	**219**	**236**

*Diagnosed case numbers may change as new cases are diagnosed or when diagnoses are updated.

Hemoglobinopathies By Race/Ethnicity

Race/Ethnicity*	Non-Sickling	Sickling	Total
African American	43%	85%	78%
American Indian	1%	0.1%	0.2%
Asian	28%	0.3%	4%
Hispanic	5%	5%	5%
Other	17%	8%	9%
White	6%	2%	2%
Total	**100%**	**100%**	**100%**

*Diagnosis entered January 2017 – August 14, 2023

Short Term Requirements

- Before moving to Long Term Follow Up (LTFU) or closing case
  - Referred and seen by hematologist at least once
  - Confirm Diagnosis (Dx Form)
  - Capture treatments start dates and other information
  - Non-Sickling (generally not followed LT) seen once by hematologist before closing case

Long Term Follow Up

- Follow conditions certain conditions up to 18 years
- Long term follow up elements identified by specialists or by national recommendation for each condition
- Periodic contact in first year of life
  - Currently send LTFU forms:
    - ~ 3 months post-diagnosis
    - at one year of age
    - every one to five years depending on condition and age
- Forms returned by specialist or PCP

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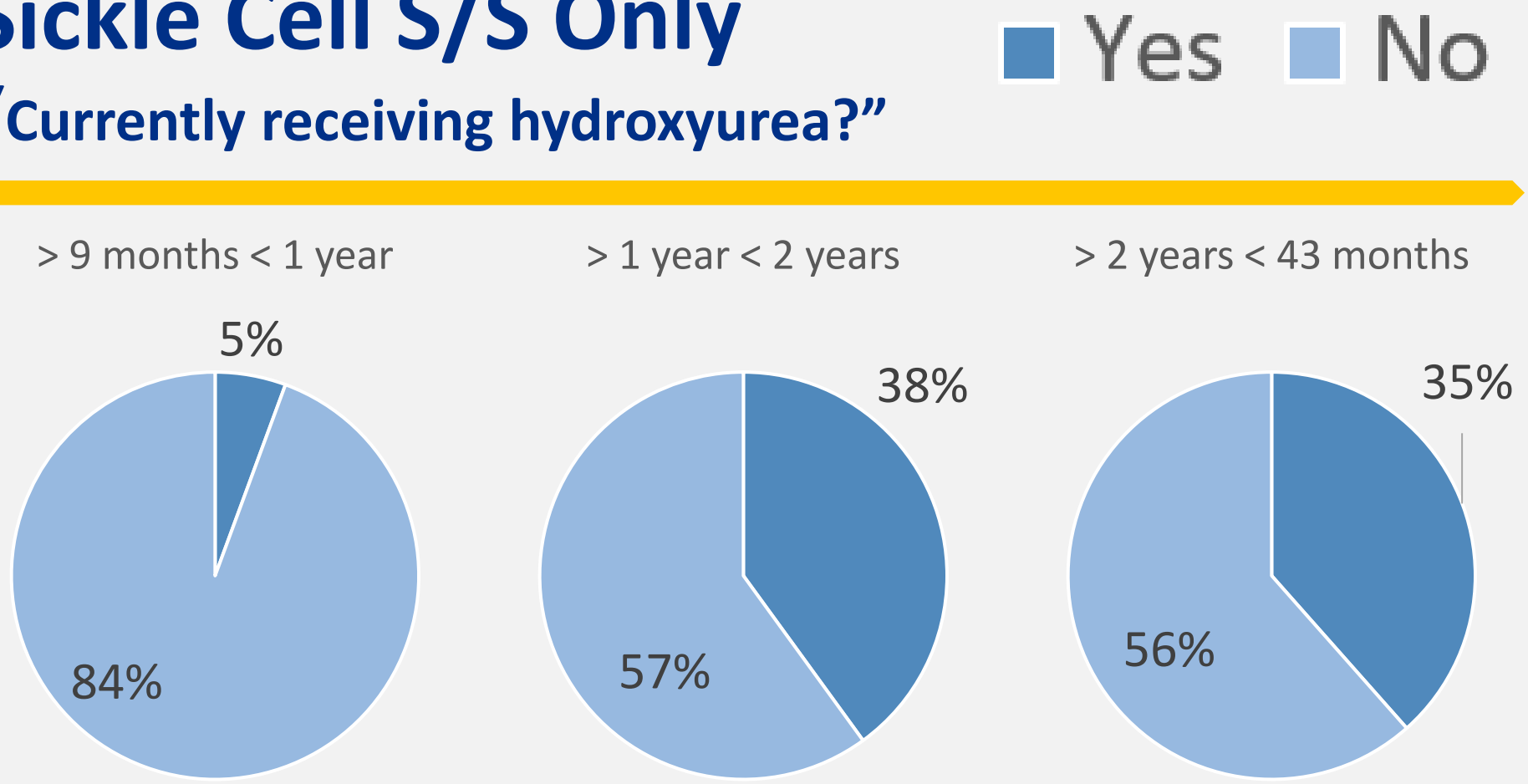
Long-Term Program - Goals

- Ensure children are under the care of a specialist and/or PCP
- Find children who are lost to follow-up
- Connect families with Texas Department of State Health Services (DSHS) social work to identify and address barriers to care
- Develop a better understanding of the long-term health outcomes of disorders found through NBS

Sickle Cell S/S Only

"Currently receiving hydroxyurea?"



# of forms = 317	315	84

*Number of Forms may not reflect number of patients

Hospitalizations

Number of Hospitalizations	Non-Sickling	Sickling
1	11	1468
2	2	620
3		281
4		158
5		70
6		39
7		27
8		13
9+	1	29

“Had A Trans Cranial Doppler in the past 12 months?”

Sickling Only	0 - 9 months		9 months - 3 years		3 - 6 years	
	#	%	#	%	#	%
Yes	5	1%	249	14%	830	52%
No	510	99%	1,538	86%	766	48%
Total Responses	**515**		**1787**		**1596**	

Sickle Trait Notification Process

- Required to screen by Texas Health and Safety Code, Section 33.0021
- DSHS lab notifies submitter/physician by mailer
- CCC does not contact providers
- CCC notifies parents by letter, including informational brochure (Sickle Cell Trait) and resource list
- If letter returned to CCC, the address researched again, and packet resent

Sickle Cell Trait By Race/Ethnicity

Race/Ethnicity	2019	2020	2021	2022	Total	Percent
African American	3,593	3,592	3,511	3,602	14,298	60%
American Indian	6	7	13	6	32	0.1%
Asian	32	25	22	37	116	0.5%
Hispanic	1,184	1,147	1,177	1,313	4,821	20%
Other	555	485	556	680	2,276	9%
White	359	340	412	404	1,515	6%
(blank)	208	280	212	242	942	4%
Total	**5,937**	**5,876**	**5,903**	**6,284**	**24,000**	**100%**

NBS Clinical Care Coordination Contacts

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Thank you!

Newborn Screening Clinical Care Coordination

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