

SCCRIP

SICKLE CELL CLINICAL RESEARCH AND INTERVENTION PROGRAM

*Establishment of a Longitudinal Clinical Cohort of
Patients with Sickle Cell Disease (SCD)*



St. Jude Children's
Research Hospital

ALSAC • Danny Thomas, Founder

Finding cures. Saving children.

Data Tables and Figures

Data Freeze December 31, 2017

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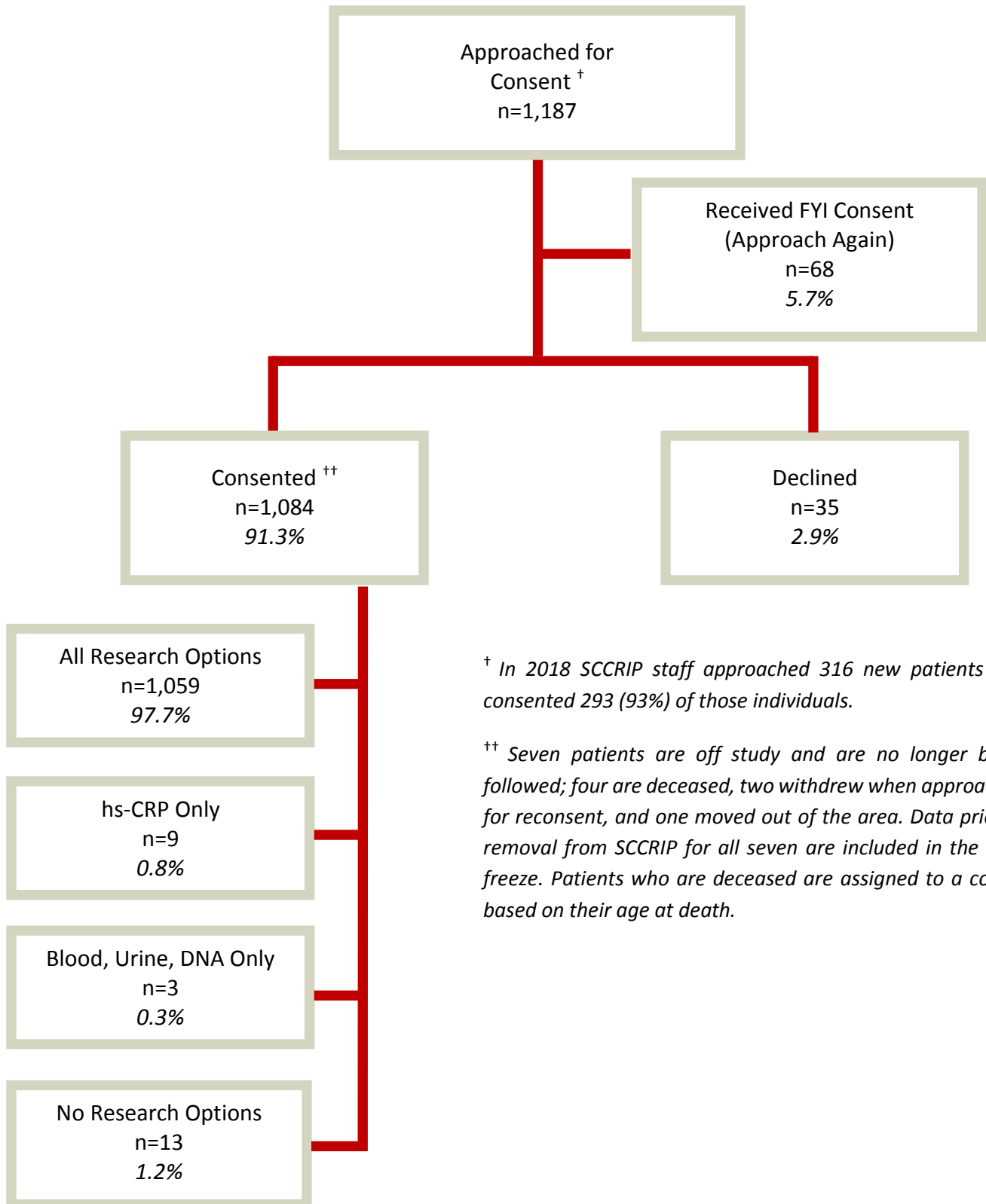
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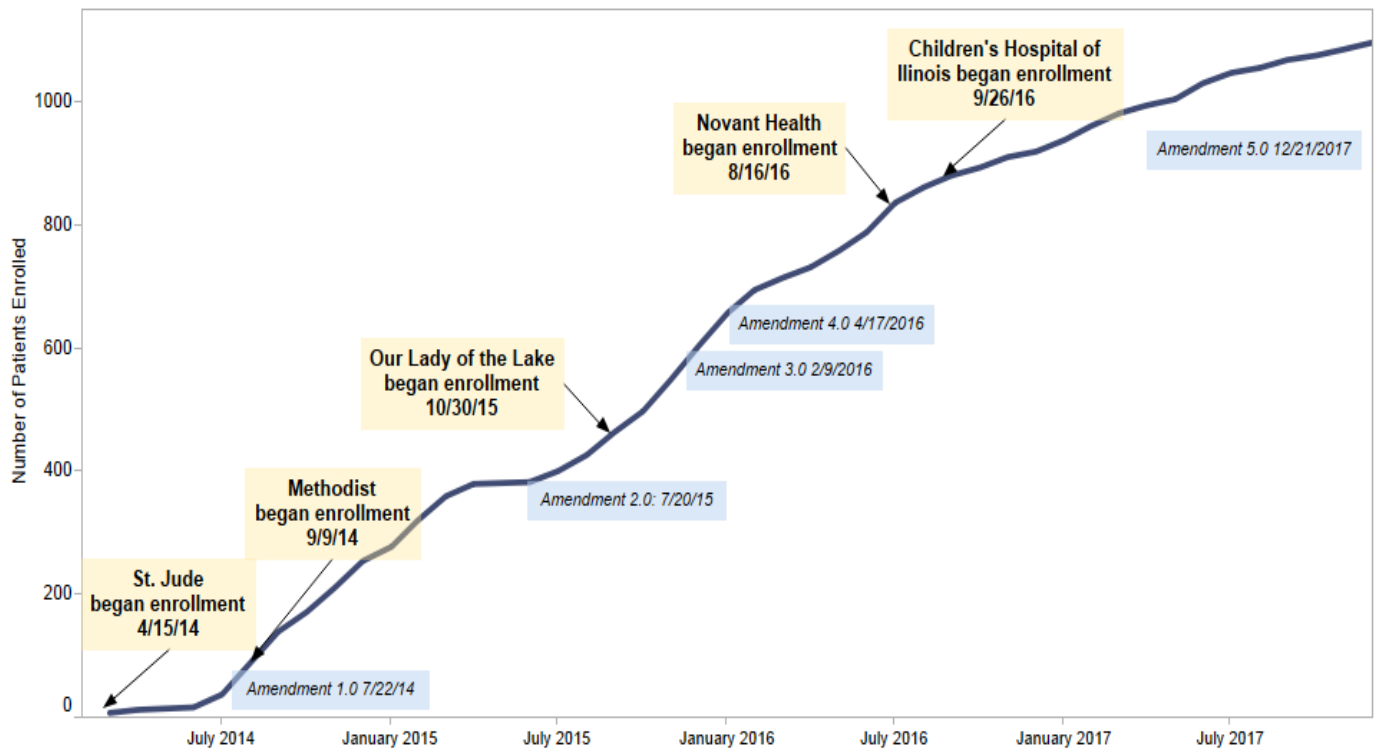
SCCRIP Study Status and Enrollment

Patients under 18 years old who decline are re-approached at age of majority.



SCCRIP Study Status and Enrollment

The initial protocol was approved March 11, 2014. Projected enrollment was 250 patients per year for the first four years. Enrollment for 2018 was 293 patients.



Definitions

Cohort

Cohort is based on patient's age at the freeze date for demographic data and treatment/vaccination history. Cohort is based on patient's age at event for research tests, evaluations, healthcare utilization, and other clinical events, unless otherwise noted. Where cohort is based on age at event, the sum of unique participants by cohort is greater than the total number of unique participants in the dataset because a single participant may have data available at multiple age cohorts.

Cohort	Participant Ages
Infant/Toddler	Birth through 5 years
School Age	6 through 11 years
Teen	12 through 17 years
Young Adult	18 through 24 years
Mature Adult	25+ years

Participants can enroll at any age. Pediatric and young adult participants age into a new cohort every six years. Descriptive data on age by cohort is provided in the Demographics table. Four participants are deceased and have been assigned to a cohort based on their age at death.

Genotype

Sickle cell genotype is based on the diagnosis in the medical record.

SCD	Text in Problem List
SS/Sβ ⁰	SICKLE CELL DISEASE, SS SICKLE CELL DISEASE, FS SICKLE CELL DISEASE, S BETA ZERO THALASSEMIA SICKLE CELL DISEASE, HEMOGLOBIN SS W/ ALPHA THALASSEMIA TRAIT SICKLE CELL ANEMIA
SC	SICKLE CELL DISEASE, SC SICKLE CELL DISEASE, FSC SICKLE CELL DISEASE, HEMOGLOBIN SC W/ALPHA THALASSEMIA TRAIT
Sβ ⁺	SICKLE CELL DISEASE, S BETA PLUS THALASSEMIA
Other SCD	SICKLE CELL DISEASE, S/HPFH SICKLE CELL DISEASE, S/HOPE SICKLE CELL DISEASE, S/WILLIAMETTE SICKLE CELL DISEASE, S/N BALTIMORE SICKLE CELL DISEASE, S/D, LOS ANGELES SICKLE CELL DISEASE, S/LEPORE BOSTON-WASHINGTON SICKLE CELL DISEASE, S/GAMMA DELTA BETA THALASSEMIA SICKLE CELL DISEASE, SD SICKLE CELL DISEASE, SE SICKLE CELL DISEASE, SO ARAB SICKLE CELL DISEASE, TYPE S WITH ANOTHER VARIANT HEMOGLOBIN HEMOGLOBIN SC HARLEM DISEASE

SCCRIP 2017 DATA FREEZE

Demographic and Diagnostic Characteristics

<i>Cohort is based on age at freeze.</i>			Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
	Total N (%)	N (%)	N (%)	N (%)	N (%)	N (%)	N (%)
	1,084	266 (24.6)	331 (30.5)	284 (26.2)	185 (17.1)	18 (1.7)	
Sex							
Female	543 (50.1)	139 (52.3)	161 (48.6)	140 (49.3)	92 (49.7)	11 (61.1)	
Male	541 (49.9)	127 (47.7)	170 (51.4)	144 (50.7)	93 (50.3)	7 (38.9)	
Race							
Black	1,072 (98.9)	263 (98.9)	324 (97.9)	284 (100)	183 (98.9)	18 (100)	
Other / Unknown	12 (1.1)	3 (1.1)	7 (2.1)	-	2 (1.1)	-	
Age							
Mean ± SD	11.51 ± 6.39	3.52 ± 1.43	9.15 ± 1.73	14.95 ± 1.74	20.49 ± 1.83	26.68 ± 1.66	
Range	0.67 - 30.33	0.67 - 5.92	6.00 - 11.92	12.00 - 17.92	18.00 - 24.67	25.08 - 30.33	
Site							
St. Jude	697 (64.3)	198 (74.4)	252 (76.1)	230 (81.0)	17 (9.2)	-	
Methodist	162 (14.9)	-	-	-	144 (77.8)	18 (100)	
Novant Health	111 (10.2)	24 (9.0)	45 (13.6)	31 (10.9)	11 (5.9)	-	
Our Lady of the Lake	81 (7.5)	32 (12.0)	29 (8.8)	20 (7.0)	-	-	
Children's Hospital of Illinois	22 (2.0)	12 (4.5)	5 (1.5)	3 (1.1)	2 (1.1)	-	
Non-St. Jude Affiliate ¹	11 (1.0)	-	-	-	11 (5.9)	-	
State of Residence							
Tennessee	551 (50.8)	123 (46.2)	162 (48.9)	149 (52.5)	108 (58.4)	9 (50.0)	
Mississippi	215 (19.8)	46 (17.3)	65 (19.6)	59 (20.8)	39 (21.1)	6 (33.3)	
North Carolina	110 (10.1)	24 (9.0)	44 (13.3)	31 (10.9)	11 (5.9)	-	
Arkansas	81 (7.5)	25 (9.4)	20 (6.0)	15 (5.3)	20 (10.8)	1 (5.6)	
Louisiana	81 (7.5)	32 (12.0)	29 (8.8)	20 (7.0)	-	-	
Illinois	21 (1.9)	11 (4.1)	5 (1.5)	3 (1.1)	2 (1.1)	-	
Other / Outside the USA ²	25 (2.3)	5 (1.9)	6 (1.8)	7 (2.5)	5 (2.7)	2 (11.1)	
Sickle Cell Genotype³							
SS / Sβ0	674 (62.2)	170 (63.9)	192 (58.0)	178 (62.7)	124 (67.0)	10 (55.6)	
SC	277 (25.6)	63 (23.7)	96 (29.0)	67 (23.6)	46 (24.9)	5 (27.8)	
Sβ+	96 (8.9)	20 (7.5)	31 (9.4)	31 (10.9)	13 (7.0)	1 (5.6)	
Other	37 (3.4)	13 (4.9)	12 (3.6)	8 (2.8)	2 (1.1)	2 (11.1)	
TBANK Enrollment							
On TBANK	903 (83.3)	197 (74.1)	297 (89.7)	240 (84.5)	156 (84.3)	13 (72.2)	
Not On TBANK	181 (16.7)	69 (25.9)	34 (10.3)	44 (15.5)	29 (15.7)	5 (27.8)	
Survival Status							
Deceased, SCD-related	3	-	1	1	1	-	
Deceased, not SCD-related	1	-	-	-	1	-	

¹ Includes former St. Jude patients who transitioned to Regional One (N=6), another provider (N=3), or are lost to follow up (N=2).

² Other states include Florida, Georgia, Indiana, Iowa, Missouri, Pennsylvania, South Carolina, and Texas.

³ SS / Sβ0 includes patients with FS, Sickle Cell Anemia, and HbSS with Thalassemia Trait. SC includes FSC and HbSC with Thalassemia Trait. Other SCD includes S/HPFH and less common types such as SD and SE.

Demographic and Diagnostic Characteristics, continued

<i>Cohort is based on age at freeze.</i>		Total ¹ N	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
All Participants		1,062	259	325	278	182	18
Distance from Clinic (Drive Time¹)							
0-30 Minutes		597 (55.1)	148 (55.6)	199 (60.1)	144 (50.7)	97 (52.4)	9 (50.0)
30-60 Minutes		188 (17.3)	43 (16.2)	51 (15.4)	61 (21.5)	30 (16.2)	3 (16.6)
60-120 Minutes		238 (21.9)	61 (22.9)	70 (21.1)	60 (21.1)	44 (23.8)	3 (16.6)
Over 120 Minutes		61 (5.6)	14 (5.3)	11 (3.3)	19 (6.7)	14 (7.6)	3 (16.6)
0-30 Minutes		597 (55.1)	148 (55.6)	199 (60.1)	144 (50.7)	97 (52.4)	9 (50.0)
Stored Biospecimens							
<i>Cohort is based on age at freeze.</i>		Total ² N	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
All Participants		1,062	259	325	278	182	18
Cells							
N		429	95	115	123	87	9
%		40.4	36.7	35.4	44.2	47.8	50.0
Plasma							
N		498	106	127	132	123	10
%		46.9	40.9	39.1	47.5	67.6	55.6
Urine							
N		484	105	121	134	114	10
%		45.6	40.5	37.2	48.2	62.6	55.6
DNA Sequenced ³							
N		535	82	173	168	107	5
%		50.4	31.7	53.2	60.4	58.8	27.8

¹ Drive time distance to clinic was calculated using ArcGIS's Network Analysis tool. Participants' most recent address was geocoded, along with SCD clinic address.

² Includes patients who consented to blood, urine, and DNA banking. An additional 22 patients did not consent to research options, or consented to hs-CRP only.

³ Patients with DNA sequenced may not currently have additional DNA samples in the biorepository.

Demographic Characteristics of Adults

Age of Completion of Socio-Demographic CRF

Total	220
18 years old	101 (45.9%)
20 years old	56 (25.5%)
22 years old	42 (19.1%)
24 years old	14 (6.4%)
26 years old	6 (2.7%)
28 years old	1 (0.5%)

Number of People in Household (Including Respondent)

N	219
Mean $\pm$ SD	3.81 $\pm$ 1.56
Median	4
Range	1 - 10

Living Arrangement

Live with parent(s)	168 (76.4%)
Live with other relative(s)	26 (11.8%)
Live with spouse / partner	10 (4.5%)
Live with roommate(s)	7 (3.2%)
Live alone	6 (2.7%)
Other / Prefer not to answer	3 (1.4%)

Marital Status

Single, never been married	207 (94.1%)
Married / Living with a partner	10 (4.5%)
Other / Prefer not to answer	3 (1.4%)

Total Household Income in the Past Year

Less than \$10,000	38 (17.3%)
\$10,000 - \$29,999	34 (15.5%)
\$30,000 - \$94,999	20 (9.1%)
\$95,000 and over	8 (3.6%)
Don't know	109 (49.5%)
Prefer not to answer	11 (5.0%)

Health Insurance Status

Medicaid or public assistance	92 (41.8%)
Through parent's policy	69 (31.4%)
Medicare	24 (10.9%)
Through healthcare.gov	4 (1.8%)
Through employer	3 (1.4%)
Uninsured	14 (6.4%)
Other / Prefer not to answer	14 (6.4%)

The Adult CRF is given every two years. In addition to the information presented here, data are collected on occupation and place of employment as well as current enrollment in school, vocational rehabilitation, and government assistance programs. Data from the Child CRF will be available in 2019.

Demographic Characteristics of Adults, continued

Employment Status

Not currently working	149 (67.7%)
Full time	47 (21.4%)
Part time	22 (10.0%)
Prefer not to answer	2 (0.9%)

Education

Less than a high school degree	50 (22.7%)
Received high school diploma / GED	86 (39.1%)
Some college / training after HS	62 (28.2%)
College graduate	14 (6.4%)
Other / Prefer not to answer	8 (3.6%)

The Adult CRF is given every two years. In addition to the information presented here, data are collected on occupation and place of employment as well as current enrollment in school, vocational rehabilitation, and government assistance programs. Data from the Child CRF will be available in 2019.

Assessment of Health-Related Quality of Life

The Pediatric Quality of Life Inventory™ (PedsQL™) is administered at specific developmental stages with a parent/caregiver and children over four years of age. Age-appropriate generic and disease-specific scales are administered for each SCCRIP time point as defined by the protocol.

Stage	PedsQL™ Age	Administered for SCCRIP
Infant	Birth to 24 months	Birth to 18 months
Toddler	2 to 4 years	3 years
Young Child	5 to 7 years	5 to 6 years
Child	8 to 12 years	11 to 12 years
Teenager	13 to 17 years	17 to 18 years
Young Adult	18 to 25 years	23 to 24 years
Mature Adult	26 + years	29 to 30 years (and every six years thereafter)

Respondents rate their agreement with items on a 0 to 4 scale which is transformed to reverse scores (0=100, 1=75, 2=50, 3=25, 4=0). Individual items assess functioning on multiple dimensions based on the respondent's experiences in the past month. Scale and subscale scores are computed as the sum of items over the number of items answered (i.e., the mean) if more than 50% of the items in the scale/subscale were answered. Thus, higher scores indicate higher quality of life.

PedsQL™ Infant and Core Scales measure core dimensions of health as delineated by the World Health Organization as well as role (school) functioning. The scales assess Physical, Emotional, Social and Cognitive/School Functioning at all stages as well as Physical Symptoms in infancy. A Total Scale score and Summary scores for Physical Health and Psychosocial Health are calculated. The Physical Functioning Subscale Score is equal to the Physical Health Summary Score for all developmental stages except for Infant, which includes items from the Physical Symptoms Subscale in the summary score calculation. **PedsQL™ Sickle Cell Disease Module** measures nine SCD-specific dimensions related to experiences of physical pain and pain management, emotion and worry about having SCD, SCD treatment, and communication with healthcare providers and others about SCD. **PedsQL™ Fatigue Module** measures three dimensions of fatigue (General, Sleep/Rest, and Cognitive).

Pediatric Quality of Life: Core Assessment Total Scale Scores

	Participant Report	Parent Report
Infant Scale		
N	na	96
Mean $\pm$ SD	na	80.40 $\pm$ 12.22
Median	na	83.96
Range	na	50.56 - 100
Toddler Scale		
N	na	114
Mean $\pm$ SD	na	79.58 $\pm$ 18.47
Median	na	85.22
Range	na	31.94 - 100
Young Child Scale		
N	136	145
Mean $\pm$ SD	74.15 $\pm$ 15.96	71.17 $\pm$ 16.54
Median	76.09	73.91
Range	30.43 - 100	34.78 - 100
Child Scale		
N	155	154
Mean $\pm$ SD	73.94 $\pm$ 13.45	66.77 $\pm$ 17.36
Median	75.00	67.67
Range	25.00 - 97.83	21.59 - 98.86
Teen Scale		
N	134	116
Mean $\pm$ SD	73.30 $\pm$ 14.59	64.24 $\pm$ 15.65
Median	75.00	63.04
Range	40.22 - 100	31.52 - 100
Young Adult Scale		
N	21	4
Mean $\pm$ SD	65.15 $\pm$ 19.04	63.86 $\pm$ 7.07
Median	68.48	61.96
Range	23.81 - 93.48	57.61 - 73.91
Mature Adult Scale		
N	3	0
Mean $\pm$ SD	53.24 $\pm$ 7.34	-
Median	52.94	-
Range	46.05 - 60.71	-

Higher scores indicate higher quality of life. Infant and Core scales include subscales for Physical Symptoms, Physical Health, and Psychosocial Health, as well as Emotional, Social, and School/Cognitive Functioning.

Pediatric Quality of Life: Fatigue Module Total Scale Scores

	Participant Report	Parent Report
Toddler Fatigue Scale		
N	na	114
Mean ± SD	na	82.77 ± 16.50
Median	na	89.58
Range	na	29.55 - 100
Young Child Fatigue Scale		
N	136	145
Mean ± SD	69.76 ± 18.20	74.59 ± 17.52
Median	69.44	75.00
Range	22.22 - 100	31.94 - 100
Child Fatigue Scale		
N	155	154
Mean ± SD	69.01 ± 16.09	70.52 ± 16.57
Median	68.06	70.14
Range	19.44 - 100	13.89 - 100
Teen Fatigue Scale		
N	134	116
Mean ± SD	65.52 ± 17.53	64.73 ± 19.20
Median	67.36	65.28
Range	26.39 - 100	12.50 - 100
Young Adult Fatigue Scale		
N	21	4
Mean ± SD	60.78 ± 18.84	67.36 ± 4.61
Median	61.11	65.97
Range	30.56 - 100	63.89 - 73.61
Mature Adult Fatigue Scale		
N	3	0
Mean ± SD	50.46 ± 4.88	-
Median	50.00	-
Range	45.83 - 55.56	-

Higher scores indicate higher quality of life. The Fatigue Module include subscales for General, Sleep/Rest, and Cognitive Fatigue.

Pediatric Quality of Life: Sickle Cell Disease Module Total Scale Scores

	Participant Report	Parent Report
Toddler SCD Scale		
N	na	114
Mean $\pm$ SD	na	79.60 $\pm$ 20.03
Median	na	84.76
Range	na	19.59 - 100
Young Child SCD Scale		
N	136	145
Mean $\pm$ SD	70.49 $\pm$ 19.54	74.46 $\pm$ 17.52
Median	71.15	76.85
Range	12.50 - 100	28.13 - 100
Child SCD Scale		
N	155	154
Mean $\pm$ SD	67.97 $\pm$ 17.52	68.62 $\pm$ 18.82
Median	66.86	68.60
Range	23.26 - 100	18.60 - 100
Teen SCD Scale		
N	134	116
Mean $\pm$ SD	67.62 $\pm$ 17.22	65.99 $\pm$ 18.26
Median	68.60	64.53
Range	20.93 - 100	27.33 - 98.84
Young Adult SCD Scale		
N	21	4
Mean $\pm$ SD	50.33 $\pm$ 16.38	64.65 $\pm$ 15.30
Median	50.00	65.56
Range	22.67 - 76.16	45.35 - 82.14
Mature Adult SCD Scale		
N	3	0
Mean $\pm$ SD	47.01 $\pm$ 7.88	-
Median	43.29	-
Range	41.67 - 56.06	-

Higher scores indicate higher quality of life. The Sickle Cell Disease Module include subscales for Pain and Hurt, Pain Impact, Pain Management and Control, Worry, Emotions, Treatment, and Communication.

SCCRIP 2017 DATA FREEZE

Screening Assessment of Neuropsychology Skills (SCANS)

Number of Participants Who Have Completed Evaluations

	Developmental	School Age	Teen 1	Teen 2	Young Adult
Demographic	203	76	76	76	-
Caregiver Questionnaire	45	31	29	22	-
BASC-2 Parent	45	21	21	20	-
BASC-2 Self	-	8	15	15	-
BASC-2 Teacher	2	4	5	4	-
Berry VMI	-	2	13	14	-
Brief Parent	59	21	21	21	-
BRIEF Teacher	3	4	5	4	-
CPT II	-	13	16	15	2
DKEFS	-	23	22	22	2
Grooved Pegboard	-	75	22	22	2
WAIS-IV	-	-	-	22	-
WASI-II	-	24	22	22	2
WJ-III ACH	-	20	17	31	2
WRAML-2	-	24	21	22	2
WISC-IV	-	23	22	-	-
BRACKEN	60	-	-	-	-
NEPSY II	35	-	-	-	-
WPPSI-IV	49	-	-	-	-
WRAYNA	-	-	-	-	-
BRIEF Adult Self	-	-	-	-	2
BSI	-	-	-	-	2

SCCRIP 2017 DATA FREEZE

Exposure to Disease-Modifying Treatment¹

<i>Cohort is based on age at freeze.</i>						
	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
All Participants	1,084	266	331	284	185	18

Hydroxyurea²

N	568	110	168	148	131	11
%	52.4	48.7	50.8	52.1	70.8	61.1
Age at Start (Mean ± SD)	4.27 ± 5.44	1.71 ± 1.11	5.32 ± 2.60	8.50 ± 4.09	12.00 ± 5.41	19.41 ± 6.89

HU Maximum Tolerated Dose³

N	314	51	82	97	80	4
% of HU treated with MTD	55.3	46.4	48.9	65.5	61.1	36.4
MTD (mg/kg/day; Mean ± SD)	27.10 ± 5.77	28.76 ± 5.84	29.29 ± 5.60	26.05 ± 5.44	25.32 ± 5.32	21.75 ± 5.56
MTD (mg/kg/day; Range)	7.8 - 35.8	15.3 - 35.8	14.0 - 35.7	7.8 - 35.0	13.0 - 35.0	18.0 - 35.8
Days to MTD (Mean ± SD)	681.76 ± 864.77	419.06 ± 229.54	458.58 ± 356.54	772.82 ± 952.75	928.64 ± 1150.65	1237.5 ± 2049.09
Days to MTD (Range) ⁴	0 - 4389	112 - 1169	54 - 2020	0 - 4366	50 - 4389	196 - 4311

Chronic Transfusion Therapy⁵

N	127	8	33	50	34	2
%	11.7	6.3	26.0	39.4	26.8	1.6
Age at Start (Mean ± SD)	5.31 ± 3.70	1.98 ± 1.32	3.72 ± 1.84	5.31 ± 3.34	7.42 ± 4.62	9.02 ± 0.18

Endari® (L-glutamine)⁶

N	-	-	-	-	-	-
%	-	-	-	-	-	-
Age at Start (Mean ± SD)	-	-	-	-	-	-

Bone Marrow Transplant

N	2	0	1	0	1	0
Age at BMT	-	-	7.98	-	15.88	-

¹ Treatment categories are not mutually exclusive; a patient who has a history of multiple types of treatment will be included in multiple treatment categories.

² Includes patients who were ever prescribed HU, regardless of the length of time on HU. All HU start and stop dates are available in the frozen data.

³ MTD is typically reached several months after HU start. MTD is not available for patients who are still in dose escalation (including most of those who started HU in mid- to late-2017) and it is not available for patients who stopped HU before MTD was reached.

⁴ The lower end of the range on days to MTD is zero (0) because one patient started HU at the MTD.

⁵ Includes patients who ever started chronic transfusion therapy, regardless of the number of transfusions. Data on transfusion events are available in the Blood Transfusion History table in the Clinical Tests, Evaluations, and Events section.

⁶ Endari® (L-glutamine) was first prescribed in 2018. Use of gene therapy, voxelotor, and crizanlizumab will also be tracked should these be prescribed to any patient(s) on SCCRIP.

SCCRIP 2017 DATA FREEZE

Vaccinations

<i>Cohort is based on age at freeze.</i>						
	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
All Participants	1,084	266	331	284	185	18
Influenza Vaccine in 2017						
N	446	122	168	138	18	-
%	41.1	45.9	50.8	48.6	9.7	-
Pneumococcal 23-polyvalent						
N	801	142	250	228	168	13
%	73.9	46.6	75.5	80.3	90.8	72.2
Pneumococcal 13-valent conjugate						
N	892	195	252	224	157	1
%	82.3	73.3	76.1	78.9	84.9	5.6
Pneumococcal 7-valent¹						
N	552	5	196	203	136	12
%	50.9	1.9	59.2	71.5	73.5	66.7
Hepatitis A						
N	105	19	38	31	16	1
%	9.7	7.1	10.6	10.9	8.6	5.6
Hepatitis B						
N	797	194	248	211	133	11
%	73.5	72.9	74.9	74.3	71.9	61.1
DtaP/Tdap						
N	174	112	33	23	6	-
%	16.1	42.1	10.0	8.1	3.2	-
Meningococcal						
N	550	132	189	176	53	-
%	50.7	49.6	57.1	62.0	28.6	-
MMR (Measles, Mumps, & Rubella)						
N	43	9	13	21	-	-
%	4.0	3.4	3.9	11.4	-	-

¹ Pneumococcal 7-valent was given through 2015.

SCCRIP 2017 DATA FREEZE

Hematologic Indices

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	869	643	616	404	161	1
Records	47,125	15,642	16,498	14,139	845	1
WBC (x10e³/mm³)						
N	46,797	15,531	16,391	14,050	824	1
Mean ± SD	11.00 ± 5.37	12.15 ± 5.71	10.60 ± 5.19	10.24 ± 4.98	10.26 ± 4.87	12.3
Median (Q1 - Q3)	10 (7.9-13.5)	11 (8.2-14.8)	9.6 (7-13)	9.3 (6.7-12.7)	9.2 (6.6-13.1)	-
Range	0 - 107	0.9 - 107	1.2 - 64.5	0 - 59.6	2.2 - 35.6	-
RBC (x10e⁶/mm³)						
N	46,218	15,148	16,307	13,965	797	1
Mean ± SD	3.15 ± 0.80	3.28 ± 0.79	3.06 ± 0.76	3.09 ± 0.82	3.32 ± 0.95	2.29
Median (Q1 - Q3)	3.02 (2.56-3.62)	3.13 (2.69-3.77)	2.94 (2.51-3.45)	2.98 (2.45-3.63)	3.22 (2.54-3.93)	-
Range	0.41 - 6.81	0.41 - 6.81	0.94 - 6.18	0.52 - 6.18	1.43 - 6.08	-
Hemoglobin (Hb; g/dL)						
N	47,040	15,598	16,479	14,130	832	1
Mean ± SD	9.51 ± 1.56	9.36 ± 1.49	9.41 ± 1.43	9.75 ± 1.67	10.35 ± 2.45	8.9
Median (Q1 - Q3)	9.4 (8.4-10.6)	9.4 (8.3-10.4)	9.3 (8.4-10.4)	9.6 (8.6-10.8)	10.1 (8.85-11.6)	-
Range	1.2-26	1.2 - 26	2.7 - 14.8	1.9 - 18.9	4.7 - 20.7	-
Mean Corpuscular Volume (MCV; fL)						
N	46,610	15,391	16,383	14,000	835	1
Mean ± SD	91.05 ± 14.04	86.12 ± 12.07	92.18 ± 13.74	94.95 ± 14.80	94.18 ± 14.20	112.3
Median (Q1 - Q3)	89 (81.7-100.1)	85.6 (77.794)	89.7 (82.9-102.1)	91.8 (84.9-105.2)	91.9 (85.9-102.3)	-
Range	42 - 168.8	42 - 126.7	52 - 168.8	58.3 - 157.2	56.4 - 135.1	-
Mean Corpuscular Hemoglobin (MCH; pg)						
N	46,550	15,377	16,376	13,986	819	1
Mean ± SD	31.21 ± 5.17	29.36 ± 4.40	31.70 ± 5.03	32.64 ± 5.5	32.12 ± 5.31	38.8
Median (Q1 - Q3)	30.5 (27.8-34.6)	29.1 (26.3-32.2)	30.9 (28.3-35.3)	31.6 (28.8-36.5)	31.3 (29-34.8)	-
Range	13.1 - 54.5	13.1 - 43.7	18.9 - 54.5	19.3 - 51.8	16.3 - 48.4	-
Mean Corpuscular Hemoglobin Concentration (MCHC; g/dL)						
N	46,216	15,146	16,307	13,965	797	1
Mean ± SD	34.23 ± 1.11	34.06 ± 1.09	34.34 ± 1.07	34.31 ± 1.16	34.08 ± 1.19	34.5
Median (Q1 - Q3)	34.2 (33.5-35.0)	34.1 (33.4-34.8)	34.3 (33.7-35.0)	34.3(33.5 - 35.1)	34.1(33.4 - 34.9)	-
Range	27.9 - 42.8	28.3 - 39	27.9 - 42.8	28.5 - 39	28.8 - 37	-
Red Cell Distribution Width (RDW; %)						
N	46,530	15,371	16,360	13,979	819	1
Mean ± SD	19.00 ± 3.61	19.22 ± 3.85	18.72 ± 3.32	19.06 ± 3.60	19.65 ± 4.31	19.3
Median (Q1 - Q3)	18.2 (16.4-20.9)	18.4 (16.5-21.2)	18 (16.3-20.4)	18.1 (16.4-21)	19 (16.2-22.4)	-
Range	0 - 82	0 - 76	12.4 - 48	12.4 - 82.0	12.9 - 37.7	-

SCCRIP 2017 DATA FREEZE

Hematologic Indices, continued

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	869	643	616	404	161	1
Records	47,125	15,642	16,498	14,139	845	1
Platelet Count (mm3)						
N	46,478	15,333	16,331	13,998	815	1
Mean ± SD	352.87 ± 161.00	350.78 ± 148.88	360.12 ± 159.14	347.47 ± 174.26	339.95 ± 174.89	364
Median (Q1 - Q3)	334 (241-439)	338 (246-435)	341 (248-446)	321 (228-436)	312 (208-426)	-
Range	5.0 - 2171	5 - 1593	6.0 - 2171	5.0 - 2011	32 - 1296	-
Mean Platelet Volume (MPV; fL)						
N	45,955	14,997	16,246	13,921	790	1
Mean ± SD	7.95 ± 0.86	7.71 ± 0.79	7.92 ± 0.81	8.19 ± 0.92	8.44 ± 0.96	9.4
Median (Q1 - Q3)	7.9 (7.4-8.5)	7.7(7.2 - 8.2)	7.9 (7.4-8.4)	8.1 (7.6-8.8)	8.3(7.7 - 9)	-
Range	0.2 - 14.2	5.3 - 13.4	5.2 - 13.4	0.2 - 14.2	6.2 - 11.6	-
Absolute Neutrophil Count (ANC; mm3)						
N	45,806	15,128	16,228	13,673	776	1
Mean ± SD	5691.32 ± 4038.46	5225.54 ± 3777.89	5925.94 ± 4222.32	5897.67 ± 4050.44	6226.96 ± 4025.77	7400
Median (Q1 - Q3)	4700 (3000-7300)	4200 (2600-6700)	4900 (3100-7600)	4800 (3100-7500)	5100 (3350-8100)	-
Range	0 - 61700	0 - 45400	0 - 61700	0 - 50500	500 - 30700	-
Absolute Reticulocyte Count (ARC; x10e6/mm3)						
N	11,134	4269	5026	1812	27	-
Mean ± SD	0.19 ± 0.10	0.20 ± 0.10	0.18 ± 0.09	0.18 ± 0.09	0.24 ± 0.14	-
Median (Q1 - Q3)	0.17 (0.12-0.24)	0.18 (0.13-0.26)	0.16 (0.12-0.23)	0.16 (0.12-0.23)	0.22(0.14 - 0.35)	-
Range	0 – 0.853	0.0009 – 0.8533	0 – 0.6729	0.003 - 0.804	0.0196 - 0.6194	-

SCCRIP 2017 DATA FREEZE

Blood Chemistry

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	869	621	602	403	155	1
Records	32399	9493	11952	10312	641	1
Blood Urea Nitrogen (BUN; mg/dL)						
N	30,404	8,896	11,033	9,863	611	1
Mean ± SD	8.38 ± 3.53	8.14 ± 3.82	8.67 ± 3.51	8.07 ± 3.21	8.34 ± 3.52	5
Median (Q1 - Q3)	8 (6 - 10)	8 (6 - 10)	8 (7 - 11)	8 (6 - 10)	8 (6 - 10)	-
Range	1 - 152	1 - 152	2.0 - 51	2.0 - 44	2.0 - 28	-
Creatinine (mg/dL)						
N	30,535	8,977	11,067	9,878	612	1
Mean ± SD	0.4 ± 0.17	0.28 ± 0.16	0.39 ± 0.10	0.52 ± 0.15	0.65 ± 0.23	0.6
Median (Q1 - Q3)	0.4 (0.3 - 0.5)	0.28 (0.2 - 0.3)	0.4 (0.3 - 0.42)	0.5 (0.4 - 0.6)	0.6 (0.5 - 0.74)	-
Range	0.06 - 13.0	0.06 - 13	0.12 - 1.88	0.16 - 2.39	0.1 - 2.45	-
Glucose (mg/dL)						
N	30,291	8,892	10,985	9,813	601	-
Mean ± SD	95.51 ± 18.02	95.51 ± 16.81	94.17 ± 14.89	97.82 ± 21.26	97.29 ± 24.99	-
Median (Q1 - Q3)	93 (86 - 102)	92 (85 - 101)	92 (85 - 100)	94 (88 - 104)	93 (86 - 102)	-
Range	8 - 787	20 - 382	53 - 273	8 - 787	58 - 443	-
Calcium (mg/dL)						
N	30,328	8,900	11,011	9,818	599	-
Mean ± SD	9.41 ± 0.47	9.56 ± 0.53	9.4 ± 0.39	9.26 ± 0.42	9.19 ± 0.58	-
Median (Q1 - Q3)	9.4 (9.2 - 9.7)	9.6 (9.3 - 9.9)	9.4 (9.2 - 9.6)	9.3 (9 - 9.5)	9.2 (9 - 9.5)	-
Range	0.7 - 12.2	0.7 - 12.2	4.4 - 12	2.6 - 11.9	0.8 - 10.8	-
Lactate Dehydrogenase (LDH; units/L)						
N	25,137	6,514	9,834	8,308	481	-
Mean ± SD	489.08 ± 225.47	531.92 ± 247.34	498.72 ± 213.83	449.88 ± 215.11	388.56 ± 176.59	-
Median (Q1 - Q3)	447 (338 - 589)	478 (367 - 631)	458 (353 - 597)	412 (304 - 549)	347 (265 - 466)	-
Range	28 - 4875	28 - 3180	134 - 2774	130 - 4875	161 - 1529	-
Albumin Level (g/dL)						
N	30,305	8,845	10,984	9,876	600	-
Mean ± SD	4.37 ± 0.40	4.38 ± 0.38	4.4 ± 0.39	4.32 ± 0.42	4.3 ± 0.49	-
Median (Q1 - Q3)	4.4 (4.2 - 4.6)	4.4 (4.2 - 4.6)	4.5 (4.3 - 4.6)	4.4 (4.2 - 4.6)	4.4 (4.1 - 4.6)	-
Range	0.9 - 6.4	1 - 6.4	1.1 - 5.5	1.4 - 6.2	0.9 - 5.6	-
Aspartate Aminotransferase: AST (SGOT) (units/L)						
N	30,245	8,864	10,980	9,799	602	-
Mean ± SD	44.69 ± 34.47	49.33 ± 38.61	46.05 ± 36.95	39.55 ± 26.73	35.45 ± 19.32	-
Median (Q1 - Q3)	39 (31 - 51)	43 (35 - 54)	40 (31 - 51)	34 (26 - 46)	31 (24 - 42)	-
Range	8.0 - 2392	13 - 2392	9 - 1525	9 - 587	8 - 208	-

SCCRIP 2017 DATA FREEZE

Blood Chemistry, continued

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	869	621	602	403	155	1
Records	32399	9493	11952	10312	641	1
Alanine Aminotransferase: ALT (SGPT) (units/L)						
N	30,167	8,827	10,979	9,762	599	-
Mean ± SD	23.46 ± 30.35	21.03 ± 27.03	25.34 ± 37.24	23.59 ± 24.23	22.82 ± 18.61	-
Median (Q1 - Q3)	17 (13 - 25)	17 (14 - 22)	18 (14 - 26)	17 (12 - 26)	17 (12 - 25)	-
Range	4.0 - 1901	4 - 1324	4.0 - 1901	4 - 432	5 - 220	-
Alkaline Phosphatase (units/L)						
N	30,060	8,742	10,953	9,778	587	-
Mean ± SD	176.22 ± 76.39	197.54 ± 85.76	188.48 ± 61.68	148.57 ± 72.55	90.75 ± 32.79	-
Median (Q1 - Q3)	170 (130 - 212)	188 (154 - 225)	178 (148 - 216)	134 (93 - 188)	86 (65 - 108)	-
Range	6.0 - 2985	6.0 - 2985	50 - 968	31 - 1046	33 - 244	-
Bilirubin, Total (mg/dL)						
N	30,259	8,853	10,990	9,812	604	-
Mean ± SD	2.15 ± 1.89	1.54 ± 1.20	2.19 ± 2.10	2.63 ± 1.99	2.62 ± 2.10	-
Median (Q1 - Q3)	1.7 (1 - 2.7)	1.2 (0.8 - 2.0)	1.7 (1.1 - 2.8)	2.1 (1.3 - 3.4)	2 (1.1 - 3.5)	-
Range	0.1 - 43.8	0.1 - 21.3	0.2 - 43.8	0.2 - 21.7	0.3 - 20.7	-
Bilirubin, Direct (mg/dL)						
N	2,0641	4,779	7,540	7,861	461	-
Mean ± SD	0.38 ± 0.96	0.31 ± 0.29	0.42 ± 1.43	0.37 ± 0.58	0.45 ± 0.86	-
Median (Q1 - Q3)	0.3 (0.2 - 0.4)	0.3 (0.2 - 0.4)	0.3 (0.2 - 0.4)	0.3 (0.2 - 0.4)	0.3 (0.2 - 0.4)	-
Range	0 - 35.9	0 - 15.4	0 - 35.9	0 - 16.6	0.1 - 13.2	-
Ferritin (ng/mL)						
N	7,686	1,151	3,671	2,689	174	1
Mean ± SD	2385.63 ± 2037.69	1505.11 ± 1470.03	2668 ± 2132.53	2415.36 ± 2006.27	1798.44 ± 1962.98	1522
Median (Q1 - Q3)	1957 (805 - 3429)	1197 (332 - 2092)	2320 (1080 - 3770)	2053 (844 - 3443)	853.5 (261 - 3115)	-
Range	4 - 20799	4 - 12677	9 - 20799	6.0 - 17362	12.0 - 7178	-
Cystatin C (mg/L)						
N	2,445	225	1,012	1,073	135	-
Mean ± SD	0.83 ± 0.18	0.78 ± 0.14	0.8 ± 0.17	0.85 ± 0.19	0.88 ± 0.28	-
Median (Q1 - Q3)	0.81 (0.71 - 0.91)	0.78 (0.7 - 0.85)	0.8 (0.7 - 0.88)	0.84 (0.73 - 0.94)	0.84 (0.72 - 0.95)	-
Range	0.41 - 3.06	0.46 - 1.25	0.41 - 2.17	0.41 - 2.22	0.46 - 3.06	-

SCCRIP 2017 DATA FREEZE

Hemoglobin Fractionation

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	862	708	509	326	109	-
Records	22,227	5,620	8,662	7,560	385	-
Hb S (%)						
N	22,069	5,597	8,534	7,554	384	-
Mean $\pm$ SD	52.72 $\pm$ 22.68	54.64 $\pm$ 20.27	52.03 $\pm$ 22.46	52.19 $\pm$ 24.41	50.32 $\pm$ 24.16	-
Median (Q1 - Q3)	60.0 (34.1-71.6)	60.3 (41.4-70.1)	59 (33.7-70.9)	61.3 (30.2-73)	47.8 (30.7 - 73)	-
Range	0 - 91.3	0 - 90.3	0 - 91.3	0.3 - 90.7	0.3 - 89.8	-
Hb F (%) ¹						
N	22,164	5,573	8,657	7,551	383	-
Mean $\pm$ SD	15.0 $\pm$ 14.3	24.16 $\pm$ 19.4	12.95 $\pm$ 10.87	10.95 $\pm$ 9.86	7.77 $\pm$ 8.59	-
Median (Q1 - Q3)	12.9 (3-22.7)	20.7 (10.8-31.2)	11 (2.7-21.3)	8 (1.9 - 18.9)	3.9 (1.0-13.3)	-
Range	0 - 96.1	0 - 96.1	0 - 87.9	0 - 49.6	0 - 41.6	-
Hb A (%)						
N	21,963	5,505	8,534	7,541	3,83	-
Mean $\pm$ SD	20.26 $\pm$ 27.83	10.06 $\pm$ 20.31	22.67 $\pm$ 28.35	24.53 $\pm$ 29.93	29.1 $\pm$ 29.51	-
Median (Q1 - Q3)	0 (0-47.3)	0 (0-3.7)	0 (0-50.7)	0 (0-55.1)	24.9 (0-57.3)	-
Range	0 - 91.8	0 - 87.2	0 - 91.8	0 - 84.9	0 - 88.2	-
Hb C (%)						
N	21,952	5,499	8,530	7,539	384	-
Mean $\pm$ SD	2.68 $\pm$ 10.12	3.07 $\pm$ 10.5	2.54 $\pm$ 9.97	2.54 $\pm$ 9.97	2.87 $\pm$ 10.79	-
Median (Q1 - Q3)	0 (0 - 0)	0 (0 - 0)	0 (0 - 0)	0 (0 - 0)	0 (0 - 0)	-
Range	0 - 48.3	0 - 48.3	0 - 47.3	0 - 45.8	0 - 45.3	-

¹ Includes Acetylated Hb F if it was reported separately.

SCCRIP 2017 DATA FREEZE

Endocrinology

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	758	248	364	354	107	-
Records	3,815	513	1342	1,807	153	-

25-OH Vitamin D (ng/mL)

N	3687	498	1280	1765	144	-
Mean ± SD	23.21 ± 11.27	28.38 ± 13.32	25.27 ± 12.07	20.44 ± 9.11	20.99 ± 10.04	-
Median (Q1 - Q3)	21 (15-29)	26 (20-34)	23 (17-31)	19 (14-26)	20.5 (12.5-27)	-
Range	3.8 - 104	6 - 101	5 - 104	3.8 - 56	5.0 - 60	-

Total Calcium PTH (mg/dL)

N	157	18	64	48	27	-
Mean ± SD	9.53 ± 0.43	9.52 ± 0.39	9.61 ± 0.46	9.52 ± 0.43	9.35 ± 0.38	-
Median (Q1 - Q3)	9.5 (9.2-9.8)	9.4 (9.2-9.8)	9.6 (9.3-10)	9.45 (9.2-9.8)	9.4 (9-9.7)	-
Range	8.2 - 10.6	9 - 10.4	8.2 - 10.6	8.8 - 10.5	8.7 - 10.0	-

Parathyroid Hormone Intact Serum (pg/mL)

N	157	18	64	48	27	-
Mean ± SD	42.10 ± 27.97	32.82 ± 18.01	41.24 ± 21.84	41.96 ± 17.82	50.59 ± 51.14	-
Median (Q1 - Q3)	35.3 (27-49.1)	29.45 (20.3-41.1)	34.3 (25.6-49.3)	38.9 (30.25-50.7)	36.9 (26.2-52.6)	-
Range	12.2 - 290.8	13.3 - 88.4	13.3 - 126.5	12.2 - 9.7	17.1 - 290.8	-

Erythropoietin Level (units/mL)

N	716	71	377	254	14	-
Mean ± SD	109.25 ± 138.47	138.13 ± 128.2	104.07 ± 105.18	110.57 ± 179.82	77.98 ± 105.19	-
Median (Q1 - Q3)	76.2 (43-134.5)	114 (72.9-152)	73.8 (43-130)	73.55 (39-132)	43.15 (33-96)	-
Range	6.0 - 1972	27.5 - 953	6.0 - 957	14.0 - 1972	9.0 - 425	-

Viral Serology

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	719	339	383	153	33	2
Records	1,395	551	603	203	36	2

Parvovirus IgG

N	1352	524	587	203	36	2
Mean ± SD	1.77 ± 2.37	0.98 ± 1.61	2.17 ± 2.58	2.53 ± 2.74	2.49 ± 2.68	3.27 ± 3.92
Median (Q1 - Q3)	0.4 (0.28-3.16)	0.4 (0.21-0.70)	0.42 (0.30-4.90)	0.53 (0.30-5.40)	0.59 (0.39-5.10)	3.27 (0.49-6.04)
Range	0.05 - 8.48	0.1 - 7.8	0.05 - 8.25	0.1 - 8.48	0.1 - 7.12	0.49 - 6.04

Parvovirus IgM

N	1219	481	541	166	29	2
Mean ± SD	1.05 ± 2.91	0.84 ± 2.59	1.29 ± 3.19	0.99 ± 2.96	0.45 ± 1.26	0.13 ± 0.01
Median (Q1 - Q3)	0.2 (0.10-0.30)	0.2 (0.10-0.28)	0.2 (0.11-0.30)	0.18 (0.10-0.20)	0.2 (0.15-0.30)	0.13 (0.12-0.14)
Range	0.06 - 19.76	0.06 - 19.76	0.07 - 18.06	0.08 - 15.59	0.1 - 6.96	0.12 - 0.14

SCCRIP 2017 DATA FREEZE

C-Reactive Protein, High Sensitivity

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	522	191	133	147	54	2
Records	604	222	149	175	56	2

hs-CRP (mg/L)

	N	604	222	149	175	56	2
Mean ± SD	3.92 ± 8.54	3.56 ± 7.61	4.57 ± 12.58	3.85 ± 6.29	3.89 ± 3.74	2.6 ± 0.99	
Median (Q1 - Q3)	1.7 (0.6 - 4.4)	1.5 (0.6 - 3.9)	1.5 (0.6 - 4.6)	1.9 (0.7 - 4.8)	2.4 (0.9 - 5.8)	2.6 (1.9 - 3.3)	
Range	0.1 - 131.0	0.1 - 69.5	0.1 - 131.0	0.1 - 45.7	0.1 - 13.2	1.9 - 3.3	

Nuclear Medicine Tc99

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	236	80	120	92	41	-
Records	443	105	157	139	42	-

GFR Result (mL / min / 1.73 sqm)

	N	443	105	157	139	42	-
Mean ± SD	134.46 ± 30.24	136.52 ± 34.87	142.11 ± 25.48	132.29 ± 28.33	107.90 ± 25.13	-	
Median (Q1 - Q3)	135 (114-151)	136 (112-152)	141 (124-156)	134 (110-147)	109.5 (98-120)	-	
Range	28 - 308	67 - 308	89 - 247	58 - 225	28 - 158	-	

Uncorrected GFR Result (mL / min / sqm)

	N	442	104	157	139	42	-
Mean ± SD	77.47 ± 16.83	77.95 ± 17.72	82.08 ± 14.74	76.45 ± 16.36	62.38 ± 14.55	-	
Median (Q1 - Q3)	78 (66-87)	78.5 (65-88)	81 (71-90)	77 (64-85)	63 (57-69)	-	
Range	16 - 143	39 - 124	52 - 143	34 - 130	16 - 91	-	

SCCRIP 2017 DATA FREEZE

Iron Liver MRI Results

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	81	15	52	45	13	1
Records	301	22	137	125	16	1

R2* (Hz)

	N	300	22	137	125	15	1
Mean ± SD	265.56 ± 161.79	246.14 ± 146.47	288.72 ± 168.26	256.35 ± 153.31	167.07 ± 160.25	149	
Median (Q1 - Q3)	224.5 (147.5-343)	205 (135-303)	233 (161-386)	224 (138-312)	89 (55-241)	-	
Range	32 - 936	102 - 729	67 - 936	38 - 750	32 - 577	-	

Liver Iron Content (LIC; mg/g of liver dry weight)

	N	300	22	137	125	15	1
Mean ± SD	7.68 ± 5.02	7.05 ± 4.54	288.72 ± 168.26	7.40 ± 4.76	4.60 ± 4.97	4.04	
Median (Q1 - Q3)	6.43 (4.05-10.05)	5.78 (3.6-8.82)	233 (161-386)	6.54 (3.69-9.09)	2.16 (1.12-6.88)	-	
Range	0.41 - 28.44	2.59 - 22.01	67 - 936	0.59 - 22.88	0.41 - 17.30	-	

Iron Heart MRI Results

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	52	11	27	28	7	-
Records	151	17	69	56	9	-

T2* (ms)

	N	151	17	69	56	9	-
Mean ± SD	37.21 ± 6.17	39.69 ± 3.92	36.19 ± 6.54	37.19 ± 6.20	40.5 ± 4.62	-	
Median (Q1 - Q3)	38.0 (35.1-41.0)	38.6 (37.4-42.9)	38.3 (33.7-40.9)	37.3 (35.0-41.0)	39.9 (38.8-42.6)	-	
Range	17.5 - 52.8	34.1 - 48.3	18.1 - 45.8	17.5 - 52.8	32.7 - 49.3	-	

SCCRIP 2017 DATA FREEZE

Urine Chemistry and Urinalysis

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	854	588	578	399	123	-
Records	13,803	3,501	5,570	4,455	277	-
Creatinine Urine Random (mg/dL)						
N	3,441	397	1,214	1,693	137	-
Mean ± SD	81.21 ± 39.64	51.84 ± 25.31	67.21 ± 27.99	95.98 ± 40.38	107.86 ± 53.93	-
Median (Q1 - Q3)	73.3 (54.04-101.21)	49.82 (35.0-64.0)	64.0 (48.0-81.04)	89.2 (67.2-119.0)	100.0 (73.3-128.0)	-
Range	2.2 - 381	2.2 - 190	4.0 - 223	4.0 - 284.99	23.0 - 381.0	-
Protein Urine Random (mg/dL)						
N	2,829	289	982	1,441	117	-
Mean ± SD	16.64 ± 57.31	10.22 ± 5.36	20.59 ± 93.78	15.12 ± 18.98	18.03 ± 29.46	-
Median (Q1 - Q3)	9.0 (7.0 - 13.0)	9.0 (7.0 - 12.0)	9.0 (7.0-13.0)	10.0 (7.0-15.0)	9.0 (7.0-14.0)	-
Range	2.0 -2365	2.0 - 42	2.0 - 2365	2.0 - 262	3.0 - 210	-
Protein Creatinine Ratio (mg/g Creatinine)						
N	2,819	289	974	1,439	117	-
Mean ± SD	241.10 ±1083.73	198.17 ± 68.75	370.50 ± 1809.92	165.58 ± 234.24	198.75 ± 380.54	-
Median (Q1 - Q3)	133.0 (98.0-180.0)	187.0 (154.0-226.0)	148.5 (117-183)	114 (82.0-156.0)	95.0 (67.0-145.0)	-
Range	26.0 - 29403	68.0 - 633	52.0 - 29403	26.0 - 4389	33.0 - 2360	-
Microalbumin Urine Random (mg/dL)						
N	967	96	410	440	21	-
Mean ± SD	9.63 ± 66.91	2.98 ± 7.70	4.24 ± 16.93	15.38 ± 97.0	24.74 ± 42.23	-
Median (Q1 - Q3)	1.1 (0.4-4.0)	0.5 (0.4-1.5)	0.7 (0.4-2.3)	1.8 (0.6 - 5.6)	2.9 (1.1-18.5)	-
Range	0.2 - 1376	0.2 - 61.4	0.2 - 270	0.2 - 1376	0.4 - 156.4	-
Microalbumin Creatinine Ratio (mg/g Creatinine)						
N	900	83	372	424	21	-
Mean ± SD	64.28 ± 234.68	42.55 ± 247.26	42.78 ± 175.27	75.67 ± 247.79	300.86 ± 533.28	-
Median (Q1 - Q3)	11.0 (6.0-33.0)	8.0 (6.0-14.0)	8.0 (5.0-22.0)	17.0 (7.0-49.5)	23.0 (9.0-185.0)	-
Range	1.0 - 4117	3.0 - 2557	2.0 - 2771	1.0 - 4117	4.0 - 1777	-
Urine Specific Gravity						
N	13,180	3,394	5,327	4,195	264	-
Mean ± SD	1.01 ±0.004	1.01 ± 0.005	1.01 ± 0.004	1.009 ± 0.003	1.01 ± 0.003	-
Median (Q1 - Q3)	1.01 (1.008-1.013)	1.01 (1.008-1.013)	1.01 (1.009-1.013)	1.01 (1.008-1.012)	1.01 (1.008-1.012)	-
Range	1.0 - 1.042	1.0 - 1.032	1.0 - 1.042	1.0 - 1.029	1.002 - 1.020	-
Urine RBC						
N	1874	415	727	678	54	-
Mean ± SD	4.88 ±15.13	2.80 ± 6.02	2.89 ± 6.48	8.04 ± 22.42	8.11 ± 24.29	-
Median (Q1 - Q3)	1.0 (1.0-3.0)	1.0 (1.0 - 2.0)	1.0 (1.0-2.0)	1.0 (1.0-4.0)	2.0 (1.0-6.0)	-
Range	0 - 182	0 - 77	0 - 65	0 - 182	1.0 - 177	-

SCCRIP 2017 DATA FREEZE

Transcranial Doppler Ultrasound

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	515	406	376	131	5	-
Records	3,408	1,438	1,655	310	5	-
Overall Result						
Normal	2,284	861	1,180	240	3	-
Low Conditional	438	194	225	19	-	-
High Conditional	273	125	138	9	1	-
Abnormal	177	86	65	25	1	-
Left M1 (cm/sec)						
N	3,075	1,245	1,533	292	5	-
Mean ± SD	92.58 ± 22.76	95.55 ± 23.92	91.48 ± 20.10	86.10 ± 28.24	70.20 ± 22.24	-
Range	12.0 - 260.0	12.0 - 209.0	29.0 - 191.0	24.0 - 260.0	48.0 - 107.0	-
Right M1 (cm/sec)						
N	3,127	1,285	1,547	290	5	-
Mean ± SD	92.29 ± 122.73	93.60 ± 23.52	92.53 ± 21.30	85.49 ± 25.22	76.00 ± 26.46	-
Range	11.0 - 254.0	24.0 - 185.0	29.0 - 239.0	11.0 - 254.0	47.0 - 111.0	-
Left MCA (cm/sec)						
N	3,277	1,354	1,621	297	5	-
Mean ± SD	139.11 ± 28.18	141.84 ± 27.34	138.42 ± 26.54	130.81 ± 37.34	118.20 ± 27.77	-
Range	9.0 - 310.0	11.0 - 234.0	17.0 - 285.0	9.0 - 310.0	84.0 - 152.0	-
Right MCA (cm/sec)						
N	3,324	1,386	1,632	304	5	-
Mean ± SD	104.55 ± 29.04	141.68 ± 28.63	140.92 ± 27.63	133.68 ± 36.49	121.80 ± 35.93	-
Range	60.0 - 322.0	60.0 - 243.0	72.0 - 304.0	62.0 - 322.0	75.0 - 163.0	-
Left Bifurcation (cm/sec)						
N	3,097	1,261	1,537	294	5	-
Mean ± SD	141.11 ± 28.52	145.20 ± 27.09	139.48 ± 27.16	132.45 ± 37.34	119.00 ± 26.12	-
Range	11.0 - 346.0	71.0 - 246.0	11.0 - 284.0	64.0 - 346.0	81.0 - 144.0	-
Right Bifurcation (cm/sec)						
N	3,137	1,282	1,552	298	5	-
Mean ± SD	142.11 ± 28.86	144.33 ± 28.40	141.89 ± 27.81	133.86 ± 34.09	128.00 ± 41.10	-
Range	55.0 - 297.0	55.0 - 240.0	72.0 - 287.0	62.0 - 297.0	79.0 - 187.0	-
Left ACA (cm/sec)						
N	2,888	1,076	1,520	287	5	-
Mean ± SD	97.03 ± 24.05	101.45 ± 24.38	96.01 ± 22.84	86.44 ± 24.86	65.20 ± 17.58	-
Range	7.0 - 211.0	22.0 - 190.0	7.0 - 211.0	32.0 - 204.0	49.0 - 91.0	-

SCCRIP 2017 DATA FREEZE

Transcranial Doppler Ultrasound, continued

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	515	406	376	131	5	-
Records	3,408	1,438	1,655	310	5	-
Right ACA (cm/sec)						
N	2,942	1,097	1,548	292	5	-
Mean ± SD	99.53 ± 26.82	103.34 ± 26.57	97.92 ± 23.96	93.67 ± 37.37	102.20 ± 64.54	-
Range	14.0 - 274.0	20.0 - 196.0	14.0 - 205.0	20.0 - 274.0	41.0 - 204.0	-
Left dICA (cm/sec)						
N	2,913	1,081	1,536	291	5	-
Mean ± SD	114.09 ± 25.29	120.28 ± 25.14	111.94 ± 23.82	102.85 ± 27.37	87.80 ± 23.32	-
Range	16.0 - 287.0	18.0 - 219.0	16.0 - 260.0	54.0 - 287.0	63.0 - 113.0	-
Right dICA (cm/sec)						
N	2,932	1,088	1,544	295	5	-
Mean ± SD	114.98 ± 25.84	120.47 ± 25.35	113.23 ± 24.75	104.00 ± 24.75	107.20 ± 45.38	-
Range	9.0 - 296.0	39.0 - 192.0	9.0 - 296.0	24.0 - 237.0	64.0 - 179.0	-
Left PCA (cm/sec)						
N	2,874	1,056	1,522	291	5	-
Mean ± SD	78.73 ± 14.66	80.64 ± 14.94	78.24 ± 14.18	74.55 ± 15.03	69.80 ± 12.85	-
Range	8.0 - 154.0	25.0 - 148.0	8.0 - 154.0	32.0 - 154.0	52.0 - 83.0	-
Right PCA (cm/sec)						
N	2,910	1,069	1,540	296	5	-
Mean ± SD	79.51 ± 15.41	81.58 ± 16.00	78.75 ± 14.02	76.21 ± 18.87	65.20 ± 13.65	-
Range	7.0 - 167.0	41.0 - 160.0	31.0 - 145.0	7.0 - 167.0	50.0 - 77.0	-
Prox BA (cm/sec)						
N	2,664	839	1,525	295	5	-
Mean ± SD	89.54 ± 18.55	91.69 ± 18.19	89.42 ± 18.45	84.33 ± 18.86	70.80 ± 20.14	-
Range	10.0 - 166.0	12.0 - 166.0	10.0 - 163.0	37.0 - 139.0	51.0 - 100.0	-
Dist BA (cm/sec)						
N	2,646	829	1,517	295	5	-
Mean ± SD	91.10 ± 18.51	93.14 ± 18.15	91.19 ± 18.31	85.13 ± 19.19	76.00 ± 21.13	-
Range	16.0 - 177.0	16.0 - 177.0	30.0 - 156.0	36.0 - 138.0	50.0 - 105.0	-

SCCRIP 2017 DATA FREEZE

Brain MRI and MRA Results

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	285	123	217	156	19	-
Records	1811	366	810	597	38	-

Brain MRI Result

Normal	437	98	195	138	6	-
Abnormal	497	87	225	172	13	-

Brain MRA Result

Normal	384	85	171	119	9	-
Abnormal	493	96	219	168	10	-

Hip MRI Results

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	63	9	19	38	3	-
Records	116	9	25	79	3	-

Hip MRI Result

AVN Not Present	68	1	14	50	3	-
No AVN	25	5	4	16	-	-
Pending Review	23	3	7	13		

CT Dual Energy X-ray Absorptiometry

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	392	3	124	249	38	0
Records	452	3	133	277	39	0

SCCRIP 2017 DATA FREEZE

Pulmonary Function Test

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants	388	19	144	295	35	-
Records	760	21	234	464	41	-
FVC % Predicted Pre-Bronch						
N	760	21	234	464	41	-
Mean ± SD	90.65 ± 14.34	94.57 ± 16.93	89.97 ± 15.18	91.17 ± 13.93	86.61 ± 11.90	-
Median (Q1 - Q3)	91.0 (81-99)	94.0 (82-106)	90.0 (82-99)	91.0 (82-99.5)	87.0 (77-93)	-
Range	36.0 - 141	65.0 - 119	45.0 - 141.0	36.0 - 139.0	58.0 - 111.0	-
FVC % Predicted Post-Bronch						
N	407	6	102	270	29	-
Mean ± SD	95.49 ± 13.42	89.67 ± 14.19	95.53 ± 14.52	95.87 ± 13.16	93.03 ± 11.7	-
Median (Q1 - Q3)	95.0 (86-104)	92.0 (81-102)	96.0 (88-107)	95.0 (86-105)	92.0 (86-100)	-
Range	53.0 - 142.0	67.0 - 104.0	53.0 - 131.0	67.0 - 142.0	63.0 - 115.0	-
FEV1 % Predicted Pre-Bronch						
N	752	21	227	463	41	-
Mean ± SD	87.22 ± 15.04	96.24 ± 18.32	86.14 ± 16.30	87.71 ± 14.27	83.07 ± 12.47	-
Median (Q1 - Q3)	87.0 (79-97)	99.0 (82-107)	87.0 (78-97)	88.0 (79-97)	83.0 (76-91)	-
Range	22.0 - 138.0	70.0 - 129.0	30.0 - 122.0	22.0 - 138.0	51.0 - 108.0	-
FEV1 % Predicted Post-Bronch						
N	408	6	102	271	29	-
Mean ± SD	95.31 ± 14.29	90.83 ± 16.63	94.62 ± 15.56	95.85 ± 14.04	93.66 ± 11.60	-
Median (Q1 - Q3)	95.0 (86.5-104.5)	91.0 (73-106)	94.0 (87-104)	96.0 (86-105)	93.0 (87-101)	-
Range	43.0 - 144.0	72.0 - 112.0	43.0 - 128.0	52.0 - 144.0	63.0 - 118.0	-
TLC (pleth) % Predicted Pre-Bronch						
N	723	7	216	459	41	-
Mean ± SD	88.26 ± 12.90	99.86 ± 19.05	93.08 ± 14.65	86.20 ± 11.45	83.90 ± 8.75	-
Median (Q1 - Q3)	87.0 (80-95)	100 (78-118)	91.0 (83-101)	85.0 (79-93)	84.0 (78-89)	-
Range	56.0 - 146.0	75.0 - 122.0	64.0 - 146.0	56.0 - 127.0	61.0 - 103.0	-
TLC (pleth) % Predicted Post-Bronch						
N	397	2	97	269	29	-
Mean ± SD	86.95 ± 12.44	106 ± 14.14	92.69 ± 13.48	85.25 ± 11.51	82.17 ± 9.98	-
Median (Q1 - Q3)	86.0 (79-93)	106 (96-116)	90.0 (85-97)	85.0 (78-91)	82.0 (74-87)	-
Range	58.0 - 135.0	96.0 - 116.0	63.0 - 135.0	58.0 - 128.0	64.0 - 107.0	-
DLCOcor % Predicted Pre-Bronch						
N	684	2	188	453	41	-
Mean ± SD	102.54 ± 23.30	104.0 ± 41.01	103.49 ± 21.47	104.06 ± 23.34	81.32 ± 20.56	-
Median (Q1 - Q3)	102.0 (85-117.5)	104.0 (75-133)	103.0 (89-117)	104.0 (87-120)	80.0 (68-90)	-
Range	44.0 - 188.0	75.0 - 133.0	54.0 - 179.0	55.0 - 188.0	44.0 - 141.0	-

SCCRIP 2017 DATA FREEZE

Blood Transfusion History

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Unique Participants ¹	522	61	159	163	129	10
Records ²	11,498	373	2,595	4,938	3,544	48

Blood Product

Red Blood Cells	9,606	235	1,994	3,848	3,484	45
Platelets	41	1	1	11	28	-
FFP / Cryo	4	na	1	2	1	-
Unknown	1,847	137	599	1,077	31	3

Mode of Transfusion

Simple	6,587	139	1,684	3,224	1,523	17
Exchange	2,154	1	86	369	1,698	-
Unknown	2,757	233	825	1,345	323	31

Age at First PBRC Transfusion

Mean ± SD	5.59 ± 4.55	1.70 ± 1.21	3.73 ± 2.46	6.34 ± 4.21	8.38 ± 5.44	10.76 ± 5.16
Median (Q1 - Q3)	4.48 (1.83 - 7.93)	1.35 (0.73 - 2.52)	3.27 (1.52 - 5.84)	5.75 (2.79 - 9.15)	7.31 (3.85 - 13.38)	10.67 (7.77 - 14.39)
Range	0 - 19.22	0.008 - 4.89	0 - 11.19	0 - 16.18	0.08 - 19.22	0.27 - 17.71

Alloimmunization

	Total	Infant / Toddler	School Age	Teen	Young Adult	Mature Adult
Anitbody						
Anti-M	12	10	2	0	0	0
Warm	22	1	9	8	4	0
Low Frequency	8	0	1	7	0	0
Cold	4	0	1	3	0	0
Anti-C	9	0	0	7	2	0
Non Specific	12	0	2	10	0	0
Anti-D	1	0	0	0	1	0
Anti-S	4	0	3	1	0	0
Anti-D	2	0	1	1	0	0
Anti-Jkb	2	0	0	2	0	0

¹ Number of patients who had at least one blood transfusion in the age cohort.

² Number of unique transfusion events. Multiple units of blood may be given for a single blood transfusion event. Where possible, transfusion data (e.g., volume, blood type) are collected by unit; otherwise, information is collected by transfusion event, with the total number of units transfused collected separately, if available.

SCCRIP 2017 DATA FREEZE

Acute Care Utilization

	Total N (%)	Infant / Toddler N (%)	School Age N (%)	Teen N (%)	Young Adult N (%)	Mature Adult N (%)
All Participants	1,084	266 (24.6)	331 (30.5)	284 (26.2)	185 (17.1)	18 (1.7)

Frequency of Utilization

Mean ± SD	13.25 ± 19.99	7.91 ± 7.46	10.9 ± 9.47	17.20 ± 14.63	23.35 ± 42.8	6.67 ± 3.54
Median (Q1 - Q3)	9 (4 - 16)	6 (3 - 11)	9 (4 - 14)	13 (7 - 25)	13 (6 - 27)	7 (6 - 8)
Range	0 - 412	0 - 42	0 - 57	1 - 72	1 - 412	1 - 13

Reason for ED Visit or Admission

Fever	3140	2595	418	123	4	-
VOC / Pain Crisis	2633	960	752	762	153	6
Viral Infection	603	424	128	51	-	-
Acute Chest Syndrome	647	388	164	85	10	-
Surgery	300	145	100	51	4	-
Adenoidectomy	14	9	3	2	-	-
Cholecystectomy	19	1	9	9	-	-
Splenectomy	62	48	11	2	1	-
Tonsillectomy	43	17	21	5	-	-
Other Surgery	162	70	56	33	3	-
Splenic Sequestration	178	158	15	5	-	-
Dental Procedure	58	25	18	15	-	-
Aplastic Crisis	79	44	29	6	-	-
Asthma exacerbation	88	36	42	10	-	-
Dactylitis	95	93	1	1	-	-
Ischemic Stroke / TIA	23	7	12	4	-	-
Urinary Tract Infection	97	62	26	7	2	-
Bacteremia	86	55	20	10	1	-
Cholecystitis	11	-	6	2	3	-
Accident / Injury	119	64	34	20	1	-
Pneumonia	52	39	7	4	2	-
Osteomyelitis	29	23	5	1	-	-
Seizure	7	3	3	1	-	-
Pre-sedation IV hydration	118	52	56	10	-	-
Others	1099	650	257	178	14	-

¹Collection of healthcare utilization data are ongoing. The number of unique participants is the number of charts that have been reviewed; cohort is based on cohort assignment at freeze date. For acute care utilization, 374 records are pending review. Of the 710 that have been reviewed, 12 patients have never had a hospital or emergency department visit; patients with no acute care history are included in the N for their cohort at the freeze date.

SCCRIP Dashboards

Data tables and figures are intended to be broad and descriptive. More detailed information can be found in dashboards showing clinical results/events at age of event rather than within cohort. The dashboards utilize interactive filters allowing results to be viewed within SCD genotype, for a specific patient by SCCRIP-ID, or events that occurred on/off HU or within a specific timeframe following blood transfusion. As an example the dashboard below shows CBC results plotted by age at collected, with the total number of results for a specific test shown on each plot. Dashboards do not contain protected health information. They are designed to facilitate exploration of the data to inform the development of research questions.

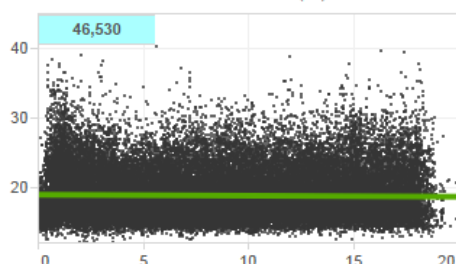
CBC results by age in years

Use the filters to the right to view results by genotype, length of time since blood transfusion prior to CBC, and Hydroxyurea status at the time of CBC. Hover over a data point to see patient information including SCCRIP ID. Enter the SCCRIP ID to view all results for a single patient. Ns are in blue.

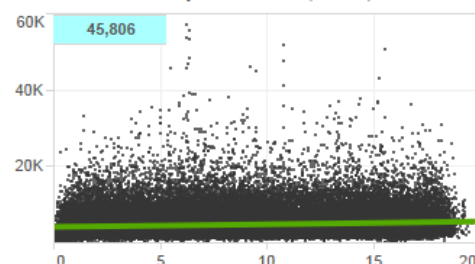
Genotype	Days Since Transfusion	On HU	SCCRIP ID
☒ SS/SB0 ☒ SC ☒ SB+ ☒ Other	☒ 90+ days or never transfused ☒ 61-90 days ☒ 31-60 days ☒ Past 30 days	☒ No ☒ Yes	

Data Source:
SCCRIP 2017 Data Freeze

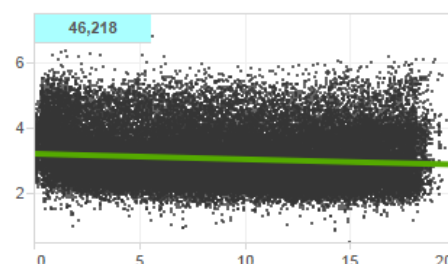
Red Cell Distribution Width (%)



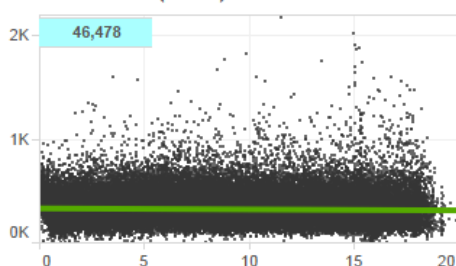
Absolute Neutrophil Count (mm3)



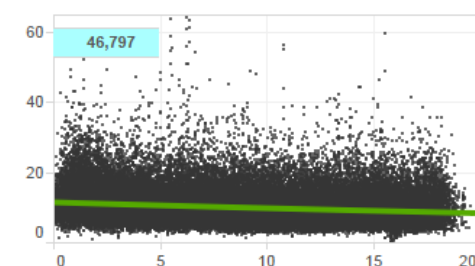
RBC x10e6/mm3



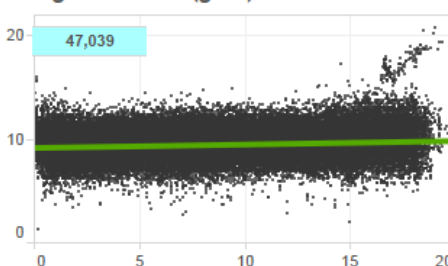
Platelet Count (mm3)



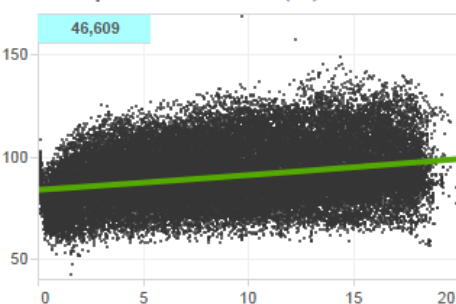
WBC x10e3/mm3



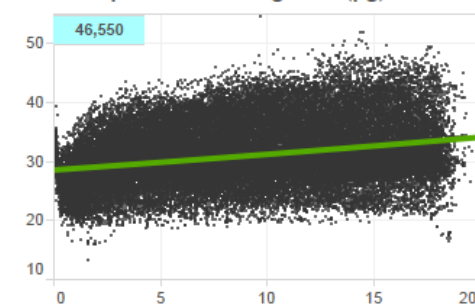
Hemoglobin Level (g/dL)



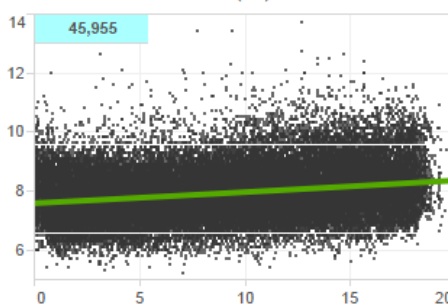
Mean Corpuscular Volume (fL)



Mean Corpuscular Hemoglobin (pg)



Mean Platelet Volume (fL)



Dashboards can be accessed on the SCCRIP Data Management and Reports SharePoint site:

<https://collaborate.stjude.org/sites/HematologyProjects/sccrip/DMR/layouts/15/start.aspx#/SitePages/Home.aspx>

Dashboards are viewed in Tableau Reader which can be downloaded for free:

<https://www.tableau.com/products/reader/download>

Requesting and Using SCCRIP Data

Preparatory to Research

In some cases, information not provided in the data tables and dashboards may be needed to develop research questions and concepts. For example, an investigator may need to know how many patients have MRI results as well as lab values within a specific timeframe (e.g., a three-month period) for a specific group of patients (e.g., SS/S β 0 patients on chronic transfusions). This information is considered preparatory to research and can help determine whether or not there are sufficient data for analysis.

For these requests, email HematologyData@StJude.org.

SCCRIP Concepts

The SCCRIP protocol requires that all research or manuscript proposals involving SCCRIP data be summarized in a brief page concept proposal containing the project title, first/senior author, collaborating authors with their affiliations and contact information, and proposal objective, background/rationale, framework, and planned analysis. All of the individuals listed as authors should have reviewed and approved the concept before its submission to the SCCRIP Steering Committee. Following receipt, a project number will be assigned and tracking from review to publication will begin.

Each concept is reviewed by the SCCRIP Steering Committee and given one of the following assessments: 1) Approved as is, 2) Approved with minor revisions, 3) Deferred pending major revisions, and 4) Not approved. In some circumstances, the Committee may request an ad hoc review from a sub-specialist or someone other than a Steering Committee member who is especially familiar with the subject matter. Concepts that have not been fully developed into protocols/manuscripts within twelve months of receipt of data may be reassigned to another individual who agrees to complete the project in a timely manner.

Approval of a concept effectively initiates the data request process. Data for each concept will be pulled from the most recent SCCRIP data freeze and provided to the investigator(s) and/or statistician on the project. A data summary will be provided to others involved in the project. Only data elements requested in the proposal that are necessary to address research questions will be provided. Protected health information will be provided only to SCCRIP investigators; otherwise data will be de-identified.

The SCCRIP Concept Proposal and Template can be accessed on the SCCRIP SharePoint site:

<https://collaborate.stjude.org/sites/hematologyprojects/sccrip/SitePages/Home.aspx>

