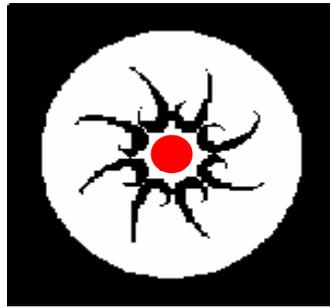


COOPERATIVE STUDY OF SICKLE CELL DISEASE

PHASE II: 09/01/1989 – 09/30/1993

DOCUMENTATION FOR CSSCD PHASE II ADULT COHORT DATASETS



COOPERATIVE STUDY OF SICKLE CELL DISEASE
Study Period: 1 September 1989 – 30 September 1993

ADULT COHORT

DOCUMENTATION FOR CSSCD ADULT DATASETS
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SECTION 1: OVERVIEW

This manual provides documentation for the SAS datasets which contain data collected for an adult cohort of Cooperative Study of Sickle Cell Disease (CSSCD) patients who were born before 1/1/56 and participated in the CSSCD Phase II study between 9/89 and 9/93.

Description of Cohort:

Eight hundred seventy sickle cell disease (SCD) patients from 16 CSSCD centers, who were born before 1/1/56, were enrolled in the original (Phase I) CSSCD study between 3/79 and 5/81 and followed through 9/88. Patients from 11 of the centers were eligible for participation in the Phase II study which began in September 1989. The list of the center codes, number enrolled at each during Phase II, and the number who died during Phase I are presented in Table 1.

TABLE 1
PHASE 1 CSSCD PARTICIPANTS WHO WERE BORN BEFORE 1/1/56

Center Letter	# enrolled in Phase I	# who died before 10/88	# Eligible for participation in Phase II
AA*	22	7	--
B*	1	0	--
C	127	22	105
G	39	5	34
I*	26	2	--
K*	31	7	--
L	27	4	23
M	74	11	63
N	56	10	46
S	60	7	53
T*	45	8	--
V	63	9	54
X	82	14	68
Y	73	15	58
Z	69	14	55
BB	75	14	61
TOTAL	870	149	620

*center did not participate in Phase II

Patients eligible for Phase II were enrolled in the Phase II study between 9/1/89 and 6/30/91. Entry status statistics for each center are presented in Table 2. The entry status is stored in the SAS dataset ROST.SD2 (variable: **P2ENTRY**)

TABLE 2
ENTRY STATUS OF PATIENTS ELIGIBLE FOR PARTICIPATION IN PHASE II STUDY

Center # - Letter	#eligible ¹	# not entered who died between 9/89 & 7/91 (% of eligible)	# entered I Phase II (% of eligible)
C	105	8 (8%)	70 (67%)
G	34	2 (6%)	19 (56%)
L	23	3 (13%)	10 (43%)
M	63	2 (3%)	33 (52%)
N	46	3 (7%)	27 (59%)
S	53	3 (6%)	21 (40%)
V	54	3 (6%)	37 (69%)
X	68	7 (10%)	40 (59%)
Y	58	5 (9%)	36 (62%)
Z	55	3 (5%)	26 (47%)
BB	61	4 (7%)	40 (66%)
TOTAL	620	43 (7%)	359 (58%)

¹ not known to be dead at end of Phase I study (9/30/88)

Study Objective for Phase II:

To determine the progression and outcome of organ damage, specifically involving the heart, lung, kidneys, and liver.

Study Follow-up Period: 9/1/89 through 9/30/93

Patients were enrolled in the Phase II study between 9/1/89 and 6/30/91. Exit visits began in September 1991 and continued through September 1993.

Study Requirements:

Study requirements are summarized in Table 3.

Special Notes:

Local Laboratory Requirements:

Although 24 hour urine samples for creatinine clearance and protein were required at the entry and exit visits, these data are of poor quality and should **NOT** be included in analyses. All other local laboratory data are considered to be of good quality.

TABLE 3
STUDY REQUIREMENTS – ADULT COHORT
COOPERATIVE STUDY OF SICKLE CELL DISEASE
1989 –1993

TYPE OF DATA	STUDY VISIT							
	ENTRY	ANNUAL 1	ANNUAL 2 (EXIT)					
	♂ ♀		♂ ♀					
History	♂ ♀	♂ ♀	♂ ♀					
Physical & Neurological Exam	♂ ♀	♂ ♀	♂ ♀					
Local Laboratory								
CBC	♂ ♀	♂ ♀	♂ ♀					
Platelet Count	♂ ♀	♂ ♀	♂ ♀					
Serum Chemistries (Alk phos, SGOT, SGPT, Bilirubin, Bun, Creatinine, Uric Acid)	♂ ♀	♂ ♀	♂ ♀					
Creatinine Clearance	♂ ♀		♂ ♀					
24 hour Urine Protein	♂ ♀		♂ ♀					
Central Laboratory								
Haplotyping	♂ ♀							
Serum Bank	♂ ♀		♂ ♀					
Special Studies								
PFT with arterial blood gases	♂ ♀		♂ ♀					
Cardiovascular Studies								
MUGA	♂ ♀		♂ ♀					
Echocardiography	♂ ♀		♂ ♀					
ECG/Treadmill	♂ ♀		♂ ♀					
Clinical events	♂ ♀	♂ ♀	♂ ♀	♂ ♀	♂ ♀	♂ ♀	♂ ♀	♂ ♀

Central Laboratories Requirements:

Haplotyping: This test was performed centrally at the Core Laboratory of the CSSCD at San Francisco General Hospital, under the direction of Dr. Stephen Embury.

DNA samples are stored at : BBI-Biotech Research Laboratories
ATTN: NHLBI REPOSITORY (CSSCD Study)
217 Perry Parkway
Gaithersburg, MD 20877

Serum Repository: All samples collected for the serum repository are also stored at BBI.

Special Studies:

Pulmonary Function Tests: Pulmonary function test forms and reports were forwarded to either Dr. Wanda Huff or Dr. Victor Tapson for central reading. Dr. Huff read 25% of the pulmonary function tests, and Dr. Victor Tapson read the remaining 75%. Dr. Tapson served as the pulmonary function consultant for the CSSCD and chair of the CSSCD Adult Pulmonary Function Writing Committee.

Cardiac Studies: Echocardiograms, echocardiography forms, ECGs, and exercise testing forms were forwarded to Dr. Wesley Covitz at Bowman Gray School of Medicine for central review and reading. All echo results and ECG J point results entered on the database were from central readings by Dr. Covitz and quality control reviews by Dr. Dalane Kitzman, also at Bowman Gray. Dr. Covitz served as the cardiac consultant for the CSSCD and Chair of the CSSCD Cardiac Writing Committee.

Clinical Events:

An Event form was to be completed for all clinical events for which a patient was seen at a CSSCD Center between **6/1/86** and 9/30/93. During Phase 1, clinical event data were collected for adult patients through 5/31/86 ONLY. As a result, centers were asked to complete Event forms for all events that occurred after 6/86 in order to have complete documentation of all clinical events that occurred since patients were enrolled in the original study.

Study forms:

A list of forms used to collect the Phase II adult data are included in Table 4. A separate binder containing copies of all form versions was submitted to NIH with this manual.

Special Notes:

Although a neurological examination was required at the time of the entry visit and each annual visit, submission of a Neurological Evaluation form was required ONLY IF the neurological exam was reported as abnormal on the physical exam form (i.e., PEABNE=2).

SAS Datasets & Documentation Manual for Datasets:

Table 5 provides the following information about the data and documentation files that are on the CD which accompany this manual:

- Source of data (form)
- Name and location of SAS dataset
- # of records in the dataset
- name and location of format file containing value labels for categorical variables
- name and location of the documentation of each form/dataset which is provided in this manual (WORD 2.0 for the majority; WORD 97 for a few that were revised in June 2001).

Codes for missing values: The following codes were used for missing values for numerical variables:

- or **-9**: value missing, no reason given; **.A** or **-1**: value missing, reason given
- **.B** or **-8**: value missing, out of range; **.C** or **-7**: value missing, not applicable

The following codes were used for missing values for date variables:

- 090909**: date missing, no reason given; **010101**: date missing, reason given;
- 080808**: date missing, out of range; **111111**: date missing , not applicable

NOTE: The documentation manual for the adult datasets was produced in 1995.

The SAS datasets were recreated (SAS version: 6.12) in August 2000 so that the CSSCD ID#s (CASEID) of patients were replaced with anonymized ID#s (ANONID). Records within each dataset were resorted by ANONID. In addition,

- **ACROSTIC (TACROST)** was removed from all datasets when they were recreated;
- **CLINIC #** was replaced with an anonymized clinic letter code in datasets which included a clinic # variable;
- **Clinic name, city, and state variables** in datasets which included these variables were removed when the datasets were recreated. Some text variables with identifying information about the location of the patient were also removed. Affected datasets and variables removed are listed below;

SAS Dataset	Variables Removed
ROST.SD2	CLINIC, OTHRREAS
STATDATA.SD2	STESTATE, STCLINN, STCLINC
DTHADATA	CITY, COUNTY, STATE, DTHCLIN, DTHPNAME
DUADATA	DUASTATE, DUACITY, DUAOCTR, DUAINTOR, DUARSNRE

TABLE 4
CSSCD STUDY FORMS FOR ADULT COHORT
1989-1993

FORM	Version Date	Version
<u>ROUTINE STUDY VISIT FORMS</u>		
History	08/04/89	G
	09/19/90	A
	10/01/91	E
Physical & Neurological Exam	08/04/89	G
Neurological Evaluation Form	08/17/89	C
	10/01/91	E
Laboratory Test Results	08/04/89	G
Laboratory Test Results-Revised	08/08/90	A
	10/01/91	E
Annual Status Report	09/24/90	A
	10/01/91	E
<u>SPECIAL STUDY FORMS</u>		
Pulmonary Function Test		
Echocardiography		
MUGA – Nuclear Imaging		
Exercise Testing		
<u>CLINICAL EVENT FORMS</u>		
Event Form	08/04/89	G
	03/01/91	H
Cause of Death Form	10/20/89	B
<u>CENTRAL LABORATORIES</u>		
Serum Storage Log		
<u>OTHER</u>		
Disposition Update Form-Adult	05/91	A
	01/92	B

**TABLE 5
 DIRECTORY OF CSSCD ADULT DATA FORMS AND FILES**

TOPIC: Form Name/Form Abbreviation		Subdirectory : ADLTSD2S Dataset Name	# of Records	Subdirectory: ADLTFORM File Name	Subdirectory: ADLTFMTS Format File Name	Subdirectory: ADLTDOC Documentation File Name
Overview (no form)	-----	-----	-----	-----	-----	OVERVIEW.DOC
Roster (no form)	-----	ROST.SD2	620	-----	ROST.FMT	ROST_DESCRIPTION.DOC ROST_CODEBOOK.DOC
Adult History	HXA	HXADATA.SD2	935	HXA.DOC	HXAC.FMT	HXA_DESCRIPTION.DOC HXA_CODEBOOK.DOC
Adult Physical Exam	PEA	PEADATA.SD2	942	PEA.DOC	PEA.FMT	PEA_DESCRIPTION.DOC PEA_CODEBOOK.OUT
Neurological Evaluation	NE	NEADATAA.SD2	322	NEA.DOC	NEA.FMT	NEA_DESCRIPTION.DOC NEA_CODEBOOK.DOC
Adult Laboratory Test Results	LAB	LABADATA.SD2	926	LABA.DOC	LABA.FMT	LABA_DESCRIPTION.DOC LABA_CODEBOOK.DOC
Adult Status Report	STAT-A STAT-E	STATDATA.SD2	581	STAT.DOC	CSTAT.FMT	STAT_DESCRIPTION.DOC STAT_CODEBOOK.DOC
Pulmonary Function Test	PFT	PFTDATA.SD2	530	PFT.DOC	PFT.FMT	PFT_DESCRIPTION.DOC PFT_CODEBOOK.DOC
Echocardiography	ECHO	ECHODATA.SD2	492	ECHO.DOC	ECHO.FMT	ECHO_DESCRIPTION.DOC ECHO_CODEBOOK.DOC
MUGA – Nuclear imaging	MUGA	MUGADATA.SD2	456	MUGA.DOC	MUGA.FMT	MUG_DESCRIPTION.DOC MUGA_CODEBOOK.DOC
Exercise Testing	EXERCISE	EXERDATA.SD2	449	EXER.DOC	EXER.FMT	EXER_DESCRIPTION.DOC EXER_CODEBOOK.DOC EXER_FREQ_COUNTS1.DOC EXER_FREQ_COUNTS2.DOC EXER_FREQ_COUNTS3.DOC EXER_FREQ_COUNTS4.DOC

**TABLE 5 (continued)
 DIRECTORY OF CSSCD ADULT DATA FORMS AND FILES**

TOPIC: Form Name/Form Abbreviation		Subdirectory : ADLTS2S Dataset Name	# of Records	Subdirectory: ADLTFORM File Name	Subdirectory: ADLTFMTS Format File Name	Subdirectory: ADLTDOC Documentation File Name
β ^S Haplotyping (no form)		HAPDATA.SD2 *	248	-----	-----	HAP_DESCRIPTION.DOC HAP_CODEBOOK.DOC
Event Form	EV	EVTADATA.SD2	5054	EVTA.DOC	EVTA.FMT	EVTA_DESCRIPTION.DOC EVTA_CODEBOOK.DOC
Cause of Death	DEATH	DTHADATA.SD2	68	DEATH.DOC	DTHA.FMT	DTHA_DESCRIPTION.DOC DTHA_CODEBOOK.DOC
Disposition Update Form - Adult	DUA	DUADATA.SD2	266	DUA.DOC	DUA.FMT	DUA_DESCRIPTION.DOC DUA_CODEBOOK.DOC

* Also stored in Phase 1 dataset: R04.SD2