

INSTRUCTIONS: Screen all patients who 1) have a single ventricle and 2) are ≤45 days old at the time screening begins.

Section A: KEY IDENTIFYING INFORMATION

Study Identification Number _____ - _____ - _____ - _____

Replaced by blinded subject ID and blinded site ID

blind_id	Blinded ID
blind_site	Blinded site ID

A2. Acrostic Identifier _____

Removed to protect privacy

A3. a. Is patient enrolled in SVR Trial? YES..... 1 NO2 (A4)

SVR_ID_YN	[Added Version E] A3a. Is patient enrolled in SVR
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b. SVR Identification Number _____ - _____ - _____ - _____

Removed to protect privacy

A4. Start Inclusion/Exclusion screening _____ / _____ / _____ / _____ / _____ / _____ / _____ / _____
M M D D Y Y Y Y

Replaced by age at start of screening

screen_age	A4. <created var>Age at start of inclusion/exclusion screening, days
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A5. Complete Inclusion/Exclusion screening _____ / _____ / _____ / _____ / _____ / _____ / _____ / _____
M M D D Y Y Y Y

Replaced by age at completion of screening

comp_age	A5. <created var>Age at completion of inclusion/exclusion screening, days
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A6. Name of person completing form _____ PRINT FULL NAME _____ INITIALS _____

Removed to protect privacy

Section B: PATIENT INFORMATION

B1. Date of birth _____ / _____ / _____
 M M / D D / Y Y Y Y

Removed to protect privacy

B2. Gender MALE 1 FEMALE 2

GENDER	B2. Patient gender
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B3. Race WHITE 1
 BLACK OR AFRICAN AMERICAN 2
 ASIAN 3
 AMERICAN INDIAN OR ALASKAN NATIVE 4
 NATIVE HAWAIIAN OR OTHER PACIFIC ISLANDER 5
 MORE THAN ONE RACE 6
 a. Specify _____
 OTHER 99
 b. Specify _____
 UNKNOWN -8

RACE	B3. Race
RACEMR_S	B3a. More than one race: specify
RACEOT_S	B3b. Other: specify

B4. Is patient of Hispanic or Latino origin? YES..... 1 NO 2 UNKNOWN.. -8

HISPANIC	B4. Is patient of Hispanic or Latino origin?
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Use this definition of Hypoplastic Left Heart Syndrome (HLHS) to answer question B5 below.

HLHS is a spectrum of cardiac malformations, characterized by a severe underdevelopment of the left heart-aorta complex, consisting of aortic and/or mitral valve atresia, stenosis, or hypoplasia with marked hypoplasia or absence of the LV, and hypoplasia of the ascending aorta and aortic arch.

Other anomalies that should not lead to a diagnosis of HLHS include:

- severely unbalanced AV septal defect,
- double-outlet right ventricle with LV hypoplasia,
- tricuspid atresia with transposition, or
- univentricular hearts with LV morphology, with or without aortic arch obstruction.

B5. Does patient have hypoplastic left heart syndrome (**HLHS**)? YES..... 1 NO..... 2

HLHS	B5. Does patient have hypoplastic left heart syndrome
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Section C: INCLUSION and EXCLUSION CRITERIA

At time of screening or death, did patient meet the following criteria?		YES	NO
C1.	Is patient ≤ 45 days old? (Date of birth = day 0)	1	2

AGELE45D	C1. Is patient less than or equal to 45 days old?
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C2.	Does patient have stable systemic & pulmonary blood flow?	1	2
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BLOODFLW	[Added Versions D] C2. Does patient have stable systemic and pulmonary blood flow?
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C3.	Is a Glenn shunt or hemi-Fontan surgery part of the future treatment plan for this patient?	1	2
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GLENNPLN	C3. Glenn shunt or hemi-Fontan surgery part of future treatment?
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C4.	Does the patient have an aortic oxygen saturation < 65%?	1	2
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O2SAT	[Added Versions B] C4. Does the patient have an aortic O2 Sat < 65%?
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C5.	a. Gestational age (enter -3 if "full term" is noted in medical record)	___ weeks	
	b. Gestational age < 35 weeks	1	2

GESTAGE	C5a. Gestational age
GALT36	[Versions A-C only] C5b. Gestational age < 36 weeks
GALT35	[Added Versions D] C5b. Gestational age < 35 weeks

C6.	a. Birth weight	___ g		If gestational age <35 wks, go to C7
	b. Birth weight < 10 th percentile for gestational age (35 weeks ≤ 1950g; 36 ≤ 2125g; 37 ≤ 2325g; Full term/38 ≤ 2500g)	1	2	

BWT	C6a. Birth weight
BWT2500	[Versions A-C only] C4b. Birth weight ≤ 2500g
BWT10TH	[Added Versions D] C6b. Birth weight < 10th percentile for gestational age
_ZWEI	<created var>WHO weight-for-age z-score, at birth
wazpct	<created var>WHO percentile for weight-for-age, at birth

C7.	Patient have pulmonary atresia with intact ventricular septum?	1	2
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ATRESIA	C7. Does patient have pulmonary atresia?
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C8.	a. Date of most recent cardiac surgery	___ / ___ / ___ M M D D Y Y Y Y	
	b. Cardiac surgical procedure performed in last 3 days? (Day of surgery = day 1)	1	2

Date of most recent cardiac surgery replaced by age at most recent surgery

csurg_age	C8a. <created var>Age at most recent cardiac surgery, days
CSURG3	C8b. Cardiac surgical procedure performed in last 3 days?

C9.	Is the patient currently on mechanical ventilatory support?	1	2
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ONVENT	C9. Is the patient currently on mechanical ventilatory support?
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C10.	Is patient currently receiving intravenous inotropic support?	1	2	NOT ASSESSED
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INOTROPE	[Added Versions B] C10. Is patient currently receiving intravenous inotropic support
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C11.	a. Is most recent serum creatinine >1.0 mg/dL?	1	2	3
	b. Serum creatinine (screening sample taken >3 days after surgery in C8a and NOT be >3 days old)	___ . ___ mg/dL		

CRTNGT1	C11a. Is most recent serum creatinine >1.0 mg/dL?
CREAT	[Added Versions D] C11b. Serum creatinine

C12.	a. Is most recent absolute neutrophil count <1000 cells/μL?	1	2	3
	b. Absolute neutrophils (screening sample taken >3 days after surgery in C8a and NOT be >3 days old)	_____ cells/mL		

NEUTRO	C12a. Is most recent absolute neutrophil count <1000 cells/mL?
ABSNEUTR	[Added Versions D] C12b. Absolute neutrophils

C13.	Does the patient have a chromosomal or recognizable phenotypic syndrome of non-cardiac congenital abnormalities associated with growth failure (including but not limited to Trisomy 21, Noonan Syndrome, Turner's Syndrome)?	1	2	3
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CHROMSYN	C13. Does patient have chrom. or recognizable phenotypic syndrome
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C14.	Is patient currently taking an ACE inhibitor?	1	2 (C15)
	a. If YES, start date of ACE inhibitor use	_ _ / _ _ / _ _ _ _ M M D D Y Y Y Y	

Date of start of ACE inhibitor use replaced by age at start of use

ACECURR	C14. Is patient currently taking an ACE inhibitor?
acest_age	C14a. <created var>Age at start of ACE inhibitor use, days

C15.	Has the patient ever received an ACE inhibitor for >7 consecutive days?	1	2 (C16)
	a. If YES, start date of ACE inhibitor use	_ _ / _ _ / _ _ _ _ M M D D Y Y Y Y	
	b. End date of ACE inhibitor use (Record 01/01/0101 if still taking)	_ _ / _ _ / _ _ _ _ M M D D Y Y Y Y	

Date of start of ACE inhibitor use replaced by age at start of use

Date of end of ACE inhibitor use replaced by age at end of use

ACE7DAYS	C15. Has patient ever received ACE inhibitor for > 7 days
acest7_age	C15a. <created var>Age at start of <7 consecutive days of ACE inhibitor use, days
aceend_age	C15b. <created var>Age at end of >7 consecutive days of ACE inhibitor use, days

C16.	Contraindication to ACE inhibitor therapy (e.g., renal disease)	1	2
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ACECONTR	[Added Versions C] C16. Contraindication to ACE inhibitor therapy?
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C17.	Patient not approached due to distance of residence from PHN center	1	2
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DISTANCE	[Added Versions C] C17. Patient not approached due to distance of residence
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C18.	Is patient alive?	1 (C19)	2
	a. If NO, date of death	___ / ___ / ___ ___ ___ M M D D Y Y Y Y	

Date of death replaced by age at death.

PTALIVE	[Added Versions B] C18. Is patient alive?
death_age	C18a. <created var>Age at death, days

C19.	Any other reason patient is unable to successfully complete trial requirements? (If YES, contact ISV Project Director at NERI, prior to finalizing the form, to confirm eligibility.)	1	2 (D1)
	a. Specify other reason _____		

UNABLE	[Added Versions C] C19. Any other reason patient unable to complete trial?
UNABLE_S	[Added Versions C] C19a. Specify other reason patient unable to complete trial

Patient is NOT ELIGIBLE for the trial if:

- Born at 35 weeks gestational age and has not been alive for at least 7 days.
- Any answers in shaded boxes: If C1-C3 or C18=NO or C4-C17 or C19=YES or C11 and C13=NOT ASSESSED

Section D: TRIAL ELIGIBILITY

D1. Is patient eligible for the trial? YES..... 1 NO..... 2 (END)

ELIGIBLE	D1. Is patient eligible for the trial?
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Section E: PATIENT CONSENT

E1. Did the parent/legal guardian sign the informed consent document for enrollment? YES..... 1 NO..... 2 (E2)

a. Date consent signed _____ / _____ / _____ (END)
 M M D D Y Y Y Y

Date of consent replaced with age at consent

CONSENT	E1. Did parent/guardian sign the informed consent document?
cnst_age	E1a. <created var>Age at signing of consent, days

E2. Reason for NOT signing Informed Consent Form: (Circle the **first** applicable choice)

- OPEN LABEL USE OF ACE INHIBITOR IS PLANNED 1 **(END)**
- FAMILY UNWILLING TO PARTICIPATE IN RESEARCH STUDY 2
- LANGUAGE BARRIER 3 **(END)**
- OTHER 99
- i. Specify other reason _____ **(END)**

NOCNST	E2. Reason for NOT signing Informed Consent Form:
NOCNST_S	E2i. No consent: Other: specify

a. If family is unwilling to participate, specify primary reason:

- BECAUSE OF TIME COMMITMENT 1
- BECAUSE OF DISTANCE FROM CENTER 2
- NO REASON GIVEN 3
- OTHER.....99
- i. Specify other reason _____

NOFAM	E2a. If family is unwilling to participate, specify reason:
NOFAM_S	E2ai. Family unwilling: Other: specify

**STOP — End of form for patients not signing Informed Consent Document.
For eligible and consenting patients, complete randomization now.**