INSTRUCTIONS: All questions on this form refer to the time since the last annual follow-up and cover the same 12-month period each year based on the subject's birthday. DO NOT *include* information recorded on the Stage III Fontan Hospitalization form. In addition, if a subject is withdrawn from the trial, complete this form at the time of withdrawal.

Section A: KEY IDENTIFYING INFORMATION

A1. Study Identification Number ___ ___ - ___ ___ ___ ___ ___ - ___

A2. Acrostic Identifier ___ ___ ___ ___ ___

A3. Study visit

STUDY VISIT 5 (2 years).....5
STUDY VISIT 6 (3 years).....6
STUDY VISIT 7 (4 years).....7
STUDY VISIT 8 (5 years).....8
STUDY VISIT 9 (6 years).....9
STUDY VISIT 10 (7 years).....10
STUDY VISIT 11 (8 years).....11
STUDY VISIT 12 (9 years).....12
STUDY VISIT 13 (10 years).....13
STUDY VISIT 14 (11years).....14

A4. Were any of the data derived from the Medical Record? YES……..1 NO…....2

A5. Was there an interview component? YES……1 NO……2 **(A7)**

How is the person interviewed related to the patient?

a. Mother YES……1 NO……2

b. Father YES……1 NO……2

c. Other YES……1 NO……2 **(A6)**

1. If other, specify: _____

A6. Date of most recent interview ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

A7. Date of form completion ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

A8. Name of person completing form _____ ___ ___ ___
PRINT FULL NAME INITIALS

Section B: ANNUAL MEDICAL and SURGICAL HISTORY

B1. Since the last annual form completion, has the patient undergone heart transplant or died? YES.....1 NO..... 2

IF YES, COMPLETE PRIMARY OUTCOME FORM R108

B2. Number of cardiac catheterization interventions since last form completion. ___ ___ (0-10) (If 0, skip to **B3**)

[DO NOT include procedures performed during the Fontan hospitalization or diagnostic catheterizations]

Cardiac Catheterization Intervention Code
(**See Code List F**)
[code required for data entry]

	1. Level 1	2. Level 2	3. Level 3	4. Level 4	5. Level 5	
a.	___ ___ -	___ ___ -	___ ___ -	___ ___ -	___ ___	Specify other (Code List F) _____
	6. Date of Catheterization Intervention ___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y					**7. Primary indication for intervention (See code list below)** ___ ___ a. If other, specify_____
b.	___ ___ -	___ ___ -	___ ___ -	___ ___ -	___ ___	Specify other (Code List F) _____
	6. Date of Catheterization Intervention ___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y					**7. Primary indication for intervention (See code list below)** ___ ___ a. If other, specify_____

Catheterization Intervention Indication Code List

Code	Primary indication for catheterization intervention	Code	Primary indication for catheterization intervention
01	Stenosis	08	Ventricular tachycardia
02	Low cardiac output	09	Thrombosis
03	Protein-losing enteropathy	10	Infection
04	Cyanosis	11	AP collaterals or excessive pulmonary blood flow
05	High output failure	12	Signs of increased SVP, such as pleural effusions or ascites
06	Bradycardia		
07	Intra-atrial reentrant tachycardia	99	Other

If >2 interventional cardiac catheterizations, please make a copy of this page.

B3. Number of other cardio-pulmonary surgical procedures since last form completion (last 12 month period) ___ ___ (0-5) (If 0, skip to **B4**)

[DO NOT include procedures performed during the Fontan hospitalization or procedures listed previously]

Surgical Procedure Code List			
Code	**Procedure Name**	**Code**	**Procedure Name**
01	Patch repair of pulmonary artery stenosis or other procedures to treat pulmonary artery stenosis (e.g. balloon dilation or stent placement in OR)	14	Extracorporeal membrane oxygenation
02	Atrioventricular valvuloplasty or repair for regurgitation	15	Pericardial window
03	Atrial septectomy	16	Pleurodesis
04	Revision of superior vena cava connection	17	Thrombectomy
05	Ligation of main pulmonary artery	18	Thoracic duct ligation
06	Division of main pulmonary artery	19	Tracheostomy
07	Atrio-ventricular valve oversewn	20	EFE resection
08	Atrio-ventricular valve replacement	21	Semilunar valve repair or valvuloplasty
09	Semilunar valve replacement	22	Placement of permanent pacemaker wires
10	Aortic arch repair	23	ORL surgical procedure
11	Pacemaker insertion*	24	Thoracentesis
12	Revision of Fontan pathway	25	Thoracostomy tube
13	Fontan Fenestration	99	Other surgical procedure

***If code 11 is selected, question B4 must be YES**

Surgical Code (See codes above)

a. ___ ___ 1. If Other (99), specify _____

b. ___ ___ 1. If Other (99), specify:_____

c. ___ ___ 1. If Other (99), specify _____

d. ___ ___ 1. If Other (99), specify _____

e. ___ ___ 1. If Other (99), specify _____

B4. Implantable electronic device placed (IED)? YES ……1 NO …… 2 **(B7)** UNKNOWN …… -8 **(B7)**

a. Date of placement ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

b. Type of device
EPICARDIAL ATRIAL ……1
EPICARDIAL VENTRICULAR ……2
EPICARDIAL DUAL CHAMBER ……3
EPICARDIAL BIVENTRICULAR ……4
IMPLANTABLE CARDIOVERTER DEFIBRILLATOR ……5
OTHER ……99

1. OTHER, specify _____

B5. Classification indication for Implantable Electronic Device (IED):

CLASS I……1

CLASS IIa……..…2

CLASS IIb…….3

CLASS III……..4

UNKNOWN……....-8

B6. Indication for IED placement:

		YES	NO
a.	Sinus node dysfunction	1	2
b.	Atrio-ventricular block	1	2
c.	Atrial tachycardia	1	2
d.	Ventricular tachycardia	1	2
e.	Ventricular dysynchrony	1	2
f.	Inducible arrhythmia	1	2
g.	Unknown	1	2
h.	Other	1	2 **(B7)**

1. Specify: _____

B7. Was 24 hour ECG holter monitor data obtained before Fontan surgery? YES……..1 NO…...2 **(B8)** N/A..…-1 **(B8)**

Note: **If YES, please use N/A for subsequent R131 Forms**

IF YES, COMPLETE Clinical 24 hr. ECG Form R209

a. Date of 24 hour ECG: (Starting Date) ___ ___ / ___ ___ / ___ ___ ___ ___ (M M D D Y Y Y Y)

B8. Arrhythmia requiring medication or intervention since the last annual contact? YES……1 NO..... 2 **(B9)**

[**DO NOT include arrhythmias that occurred during the Fontan hospitalization or arrhythmias that resulted in IED implantation per above**]

		YES	NO	UNKNOWN
a.	Atrial tachycardia/ flutter	1	2	-8
b.	Supraventricular tachycardia- other	1	2	-8
c.	Atrial fibrillation	1	2	-8
d.	Junctional ectopic tachycardia	1	2	-8
e.	Ventricular tachycardia	1	2	-8

Cont. B8.

		YES	NO	UNKNOWN
f.	Ventricular fibrillation	1	2	-8
g.	Bradycardia requiring therapy	1	2	-8
h.	2nd degree AV block	1	2	-8
i.	3rd degree AV block (complete heart block)	1	2	-8
j.	Indeterminate type	1	2	
k.	Other	1	2 **(B9)**	

1. Specify:_____

B9. Thrombosis since last contact YES……..1 NO……2 (**B10**) UNKNOWN……..-8 **(B10)**

a. Date of thrombosis diagnosis ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

b. Number of locations _____ (1-5)

c. Location of thrombosis (**See code list below**)

Code list for thrombosis locations			
01	Inferior vena cava (IVC)	11	Hepatic veins (Vein hep)
02	Lateral Tunnel (Lat Tun)	12	Innominate vein (Vein inn)
03	Right atrium (RA)	13	Right subclavian vein (Vein RSC)
04	Left Atrium (LA)	14	Left subclavian vein (Vein LSC)
05	Extracardiac conduit (ECC)	15	Azygous vein (Vein azy)
06	Pulmonary artery (PA) main	16	Homozygous vein (Vein Hemi Azy)
07	Pulmonary artery (PA) right	17	Right ventricle (RV)
08	Pulmonary artery (PA) left	18	Left ventricle (LV)
09	Superior vena cava (SVC) right	19	Aortic valve (AoV)
10	Superior vena cava (SVC) left	20	Aorta (Ao)
		99	Other

1. ___ ___ i. If Other (99), specify_____
2. ___ ___ i. If Other (99), specify_____
3. ___ ___ i. If Other (99), specify_____
4. ___ ___ i. If Other (99), specify_____
5. ___ ___ i. If Other (99), specify_____

B9. (cont.)

d. Method of diagnosis	YES	NO	UNKNOWN
1. TEE	1	2	-8
2. TTE	1	2	-8
3. MRI	1	2	-8
4. Cardiac catheterization	1	2	-8
5. Clinical	1	2	-8

B10. Stroke since last contact YES…….1 NO……..2 (**B11**) UNKNOWN … -8 **(B11)**

a. Date of stroke ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

B11. Seizure since last contact YES ……. 1 NO…….2 (**B12**) UNKNOWN …-8 **(B12)**

a. Date of seizure ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

b. Does the patient have a chronic seizure disorder? YES ..... 1 NO ..... 2 UNKNOWN…-8

B12. Protein-losing enteropathy diagnosis since last contact YES..... 1 NO……...2**(B13)** UNKNOWN…-8 **(B13)**

a. Date of diagnosis ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

b. Clinical and laboratory findings at presentation

	YES	NO	UNKNOWN
1. Hypoalbuminemia	1	2	-8
2. Ascites	1	2	-8
3. Edema	1	2	-8
4. Elevated stool α-1 antitrypsin	1	2	-8

B13. Cirrhosis diagnosis since last contact YES..... 1 NO……..2 **(B14)** UNKNOWN ...-8 **(B14)**

a. Date of diagnosis ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

B14. Plastic bronchitis diagnosis since last contact YES..... 1 NO……..2 **(B15)** UNKNOWN ...-8 **(B15)**

B14. (cont.)

a. Date of diagnosis ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

B15. Number of other significant complications since last annual contact **(exclude complications identified in B8-B14 and complications on the Fontan Hospitalization Form)** ___ (0-8) (If 0, skip to **C1**)

Complications Code (See Code List M) [Code required for data entry]	**Specify** [Use spaces below to record dates and write complications]
a1. Date of onset a2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
b1. Date of onset b2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
c1. Date of onset c2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
d1. Date of onset d2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
e1. Date of onset e2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
f1. Date of onset f2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____
g1. Date of onset g2. ___ - ___ ___ ___ ___	___ ___ / ___ ___ / ___ ___ ___ ___ M M D D Y Y Y Y Specify other:_____

If >7 complications, please make a copy of this page

B16. Diagnosed with a genetic syndrome since last annual contact?

YES ……..…..1

NO ……..……2 **(C1)**

SUSPECTED ……3

UNKNOWN ……..…….-8 **(C1)**

a. If YES, what is the diagnosis? **(Refer to Code List below)** ____ ____

1. Specify other (99): _____

Identified Syndromes			
Code	**Name**	**Code**	**Name**
01	Alagille Syndrome	09	Noonan's Syndrome
02	CHARGE association	10	Smith-Lemle-Opitz
03	DiGeorge Syndrome	11	Treacher Collins Syndrome
04	Down Syndrome	12	Turner Syndrome
05	Ellis –Van Creveld Syndrome	13	VATER/VACTERL Association
06	Goldenhar Syndrome	14	Williams Syndrome
07	Holt-Oram Syndrome		
08	Jacobsen's Syndrome	99	Other, identifiable

Section C: CURRENT MEDICATIONS

C1. How many medications is the patient currently receiving? _____ (0-10) (If 0, skip to **D1**)

Please refer to Code List D.

	1. Code **[Code required for data entry]**	**Name of Drug** **(Specify the name of drug if the code is 99)**
a.	___ ___ . ___ ___	_____
b.	___ ___ . ___ ___	_____
c.	___ ___ . ___ ___	_____
d.	___ ___ . ___ ___	_____
e.	___ ___ . ___ ___	_____
f.	___ ___ . ___ ___	_____
g.	___ ___ . ___ ___	_____
h.	___ ___ . ___ ___	_____

If > 8 medications, please make a copy of this page.

Section D: CLINICAL ASSESSMENT

Record the most recent growth measurements obtained. Enter the physical exam data that is closest to the subject's current birthday, and does not exceed 30 days beyond that birthday.

D1. Date of physical exam ___ ___ / ___ ___ / ___ ___ ___ ___
M M D D Y Y Y Y

D2. Length/ height ___ ___ ___ . ___ cm

D3. Weight ___ ___ . ___ ___ kg

D4. Has the patient had a Fontan procedure since the last annual contact? YES……1 NO……2 **(D5)**

a. Date of Fontan procedure ___ ___ / ___ ___ / ___ ___ ___ ___ **(Go to E1 if visit=5; else E9)**
M M D D Y Y Y Y

D5. Is the Fontan procedure planned? YES……1 **(E1 or E9)** NO……2 **(D6)** UNKNOWN ……-8 **(E1 or E9)**

D6. If no, specify reason: _____

Go to E1 if visit=5 (24 mo.); else go to E9

Section E: DEVELOPMENTAL ASSESSMENT

IN THE FIRST YEAR OF LIFE, DID YOUR CHILD RECEIVE ANY OF THE FOLLOWING SERVICES*?

[*Complete questions E1-E8 only at 24 month follow-up (Visit 5)]

Service	a. Yes/No	b. Frequency	c. Duration (mo/wk)	d. Discontinued?	e. Reason
E1. Speech or language therapy	YES….1 NO …..2 **(E2)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E2)**	
E2. Physical therapy	YES….1 NO …..2 **(E3)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E3)**	
E3. Occupational therapy	YES….1 NO …..2 **(E4)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E4)**	
E4. Early intervention for a cognitive disorder	YES….1 NO …..2 **(E5)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E5)**	
E5. Treatment for a psychological or behavioral disorder	YES….1 NO …..2 **(E6)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E6)**	
E6. Other	YES….1 NO …..2 **(E7)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E7)**	
E6a1. (specify)					

E7. In the first year of life, has your child had any problems with his/her hearing for which he/she has seen a specialist? YES……1 NO……2 **(E8)**

a. If yes, has any intervention been needed such as a hearing aid or hearing tubes? YES……1 NO……2 **(E8)**

b. If yes, identify the intervention:_____

E8. In the first year of life, has your child had any problems with his/her vision for which he/she has seen a specialist? YES……1 NO……2 **(E9)**

a. If yes, has any intervention been required such as eye glasses, a patch or an operation? YES……1 NO……2 **(E9)**

b. If yes, identify the intervention:_____

IN THE PAST 12 MONTHS, DID YOUR CHILD RECEIVE ANY OF THE FOLLOWING SERVICES*?
(*Complete questions E9-E16 at each annual contact for the 12 month period only)

Service	a. Yes/No	b. Frequency	c. Duration (mo/wk)	d. Discontinued?	e. Reason
E9. Speech or language therapy	YES….1 NO …..2 **(E10)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E10)**	
E10. Physical therapy	YES….1 NO …..2 **(E11)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E11)**	
E11. Occupational therapy	YES….1 NO …..2 **(E12)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E12)**	
E12. Early intervention for a cognitive disorder	YES….1 NO …..2 **(E13)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E13)**	
E13. Treatment for a psychological or behavioral disorder	YES….1 NO …..2 **(E14)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E14)**	
E14. Other	YES….1 NO …..2 **(E15)**	___ ___ . ___ / wk	___ ___. ___ ___	YES….1 NO …..2 **(E15)**	
E14a1. (specify)					

E15. In the past 12 months, has your child had any problems with his/her hearing for which he/she has seen a specialist? YES……1 NO…….2 **(E16)**

a. If yes, has any intervention been needed such as a hearing aid or hearing tubes? YES……1 NO…….2 **(E16)**

b. If yes, identify the intervention:_____

E16. In the past 12 months, has your child had any problems with his/her vision for which he/she has seen a specialist? YES……1 NO…….2 **(F1)**

a. If yes, has any intervention been required such as eye glasses, a patch or an operation? YES……1 NO…….2 **(F1)**

b. If yes, identify the intervention:_____

Section F: FEEDING

F1. What type of diet does your child eat?

		YES	NO
a.	Regular Texture Diet	1	2
b.	Low Fat Diet	1	2
c.	Low Salt Diet	1	2
d.	Pureed/Soft Diet	1	2
e.	Thickened Liquids	1	2
f.	Elemental Formula	1	2
g.	Caloric Augmentation or Supplementation	1	2
h.	Other	1	2 **(F2)**

i. Specify:_____

F2. How is your child fed?
MOUTH ONLY……1 **(F4)**
MOUTH WITH SOME TUBE FEEDING SUPPLEMENTS……..2
TUBE FEEDING ONLY.……3

F3. Tube type?
G-TUBE (gastrostomy tube)……..1
NG-TUBE (nasogastric tube) ……...2
GJ-TUBE (gastrostomy-jejunostomy tube)…….3

F4. Has your child experienced any feeding issues in the past 12 months? YES .....…1 NO……2 **(F5)**

a. Types of problems:
1. Oral aversion YES .....…1 NO….....2
2. Swallowing disorders YES .....…1 NO….....2
3. Reflux YES .....…1 NO….....2
4. Gagging YES .....…1 NO….....2
5. Frequent vomiting YES .....…1 NO….....2 **(F4a.6)**
i. Frequency
1-3 TIMES PER WEEK ……..…..1
4-6 TIMES PER WEEK ……...…..2
> 6 TIMES PER WEEK…. ……..…..3
6. Other YES ..... …1 NO.....…….2 **(F5)**
i. specify: _____

F5. Does your child receive therapy for feeding or swallowing issues? YES .....…1 NO.....…….2 **(END)**

END OF FORM