

B4. Primary cause of dilated cardiomyopathy **CAUSE**

- METABOLIC DISORDER 1
- MITOCHONDRIAL DISORDER 2
- NEUROMUSCULAR DISEASE ASSOCIATED WITH CARDIOMYOPATHY 3
- MALFORMATION SYNDROME ASSOCIATED WITH CARDIOMYOPATHY 4
- SINGLE GENE DEFECT 5
- ADRIAMYCIN-ASSOCIATED CARDIOTOXICITY 6
- IDIOPATHIC 7
- OTHER 99

a. Specify _____ **CAUSESPEC**

B5. Known or suspected familial cardiomyopathy **FAMCM** YES.....1 NO..... 2

[Definition: Familial cardiomyopathy is any form of cardiomyopathy that has been confirmed to be present in more than one genetically-related family member.]

B6. Was the patient hospitalized within the 6 months prior to the baseline study echocardiogram? **HOSPINYR** YES.....1 NO.....2 (**B7**)

a. Number of hospitalizations **related to heart failure** ___ ___ (0-10) **HOSPNUM**

B7. Did the patient experience any of the following symptoms or events within the 6 months prior to the baseline study echocardiogram?

	YES	NO	NA	UNKNOWN
a. Failure to thrive BFAILTHR	1	2	-1	-8
b. Feeding difficulties (tachypnea, retractions, grunting, diaphoresis with feeds) BFEDDIFF	1	2	-1	-8
c. Dyspnea (including orthopnea, paroxysmal nocturnal dyspnea) BDYSPNEA	1	2		-8
d. Exercise intolerance BEXINTOL	1	2	-1	-8
e. Edema BEDEMA	1	2		-8
f. Syncope BSYNCOPE	1	2		-8
g. Palpitations BPALPIT	1	2		-8
h. Chest pain BCHSTPAIN	1	2		-8
i. Thrombo-embolic event BTHROEMB	1	2		-8
j. Other BOTHSYMP	1	2		-8

1. If Other is YES, specify: _____ **BSYMPSPC**

B8. Within the 6 months prior to the baseline study echocardiogram, was the patient treated with

a. Intravenous inotropic medications? YES 1 NO 2 **(B8b)**

BINOTROP

1. Last date of IV inotrope treatment: _____ / _____ / _____
 REMOVED AGE_BINO_LD M M D D Y Y Y Y

b. Left ventricular assist device (LVAD)? YES 1 NO2 **(B8c)**

BLVAD

1. Last date of LVAD treatment: _____ / _____ / _____
 REMOVED AGE_BLVAD_LD M M D D Y Y Y Y

c. Extracorporeal membrane oxygenation (ECMO)? YES 1 NO2 **(C1)**

BECMO

1. Last date of ECMO treatment:: _____ / _____ / _____
 REMOVED AGE_BECMO_LD M M D D Y Y Y Y

Section C: CARDIAC MEDICATION HISTORY WITHIN THE 6 MONTHS PRIOR TO THE BASELINE ECHOCARDIOGRAM

INSTRUCTIONS:

- Record only cardiac-related oral medications that the patient received within the 6 months prior to the baseline echocardiogram (or since diagnosis if the patient was diagnosed less than 6 months before the baseline echocardiogram).
- If the patient was treated with intravenous inotropic medications, LVAD, or ECMO within the 6 months preceding the baseline echocardiogram, only list cardiac-related oral medications given during the time period since all three of these treatments were discontinued – i.e., the start date for listing cardiac-related oral medications should be the day following the last day that the patient received any of the 3 treatments. The medication status for drugs that the patient was receiving by mouth on the last date of any of these treatments and continued to receive after discontinuation should be coded as 2 (“Dose change”) in column c.
- If the patient received no cardiac-related medications in the 6 months prior to the baseline echocardiogram, enter -1 in Column a for C1.
- If a medication at the dose specified in Column d was started more than 6 months before the baseline echocardiogram, do not fill in column e.
- Enter dates in MM/DD/YYYY date format.

	a. Medication Code (See Code List D) MEDCODE_0- MEDCODE_24	b. Medication Name [If code ends in .99, medication name will be entered into ADEPT] BMEDNAM_0- BMEDNAM_24	c. Medication Status NEW DRUG 1 DOSE CHANGE.... 2 ON > 6 MONTHS.. 3 BMEDSTAT_0- BMEDSTAT_24	d. Dose (mg/day) BMEDDOSE_0- BMEDDOSE_24	e. Start Date or Dose Change Date REMOVED AGE_BSTRCHNG_0- AGE_BSTRCHNG_24	f. Still on? YES.. 1 NO.... 2 BSTLONMD_0- BSTLONMD_24	g. Stop Date REMOVED AGE_BSTOP_0- AGE_BSTOP_24
C1.	___ . ___		___	___ . ___	___ / ___ / ___	___	___ / ___ / ___
C2.	___ . ___		___	___ . ___	___ / ___ / ___	___	___ / ___ / ___
... C25.	___ . ___		___	___ . ___	___ / ___ / ___	___	___ / ___ / ___

<created var> Days on Medication: **BMEDDAY_0- BMEDDAY_24**

**Section D: ASSESSMENT OF SEVERITY OF HEART FAILURE
AT THE TIME OF THE BASELINE ECHOCARDIOGRAM VISIT**

D1. Age of patient at time of the baseline echocardiogram visit **BECHAGE**
 < 5 YEARS..... 1 (D2) ≥ 5 YEARS.....2 (D3)

D2. Ross Classification of Congestive Heart Failure (Circle one) **BROSCLAS**
 COMPLETE IF CURRENT AGE IS < 5 YEARS

The PHN Pediatric Nurse should determine the patient’s classification at the time of the baseline echocardiogram visit.

CLASS I (NO LIMITATIONS OR SYMPTOMS) 1 (D4)

CLASS II (SYMPTOMS BUT NO GROWTH FAILURE*) 2 (D2a)

CLASS III (GROWTH FAILURE* AND PROLONGED FEEDING TIME IN INFANTS) 3 (D2b)

CLASS IV (GROWTH FAILURE* AND SYMPTOMATIC AT REST 4 (D2c)
 WITH ≥1 SYMPTOM LISTED IN D2c BELOW)

* Growth failure is defined as weight-for-age < 5th percentile

a. If Class II, indicate signs and symptoms present (Answer **ALL** questions 1-3)

		YES	NO	NA
1.	Mild tachypnea with feeds in infants BR2TACHY	1	2	-1
2.	Mild diaphoresis with feeds in infants BR2DIAPH	1	2	-1
3.	Dyspnea on exercise in older children BR2DYSPEX	1	2	-1
GO TO D4				

b. If Class III, indicate signs and symptoms present (Answer **ALL** questions 1-2)

		YES	NO
1.	Marked tachypnea with feeds or exertion BR3TACHY	1	2
2.	Marked diaphoresis with feeds or exertion BR3DIAPH	1	2
GO TO D4			

c. If Class IV, indicate signs and symptoms present (Answer **ALL** questions 1-4)

		YES	NO
1.	Tachypnea BR4TACHY	1	2
2.	Retractions BR4RETRC	1	2
3.	Grunting BR4GRUNT	1	2
4.	Diaphoresis BR4DIAPH	1	2
GO TO D4			

D3. New York Heart Association (NYHA) Classification of Congestive Heart Failure
 COMPLETE IF CURRENT AGE IS ≥ 5 YEARS **BNYHCLAS**

The PHN Pediatric Nurse should determine the patient’s classification at the time of the baseline echocardiogram visit.

- CLASS I** 1
 NO LIMITATION OF ACTIVITIES; NO SYMPTOMS FROM ORDINARY ACTIVITIES
- CLASS II** 2
 SLIGHT, MILD LIMITATION OF ACTIVITY; COMFORTABLE WITH REST OR WITH MILD EXERTION
- CLASS III** 3
 MARKED LIMITATION OF ACTIVITY; COMFORTABLE ONLY AT REST
- CLASS IV** 4
 SHOULD BE AT COMPLETE REST, CONFINED TO BED OR CHAIR; ANY PHYSICAL ACTIVITY BRINGS ON DISCOMFORT AND SYMPTOMS OCCUR AT REST

D4. Does the patient have rhythm abnormalities?

BRHYTHAB YES 1 NO..... 2 (D5)

Type of rhythm abnormality	YES	NO
a. Atrial tachyarrhythmia BABATTAC	1	2
b. Ventricular tachyarrhythmia BABVNTAC	1	2
c. Bradycardia BABBRADY	1	2
d. 2 nd or 3 rd degree heart block BABHTBLK	1	2

D5. Does the patient have an AICD? YES 1 NO..... 2
BAICD

D6. Is the patient on a heart transplant waiting list?
 YES..... 1 NO..... 2 UNKNOWN-8
BTRNWAIT

**Section E: LABORATORY ASSESSMENT OF SEVERITY OF HEART FAILURE
IN THE 6 MONTHS PRIOR TO THE BASELINE ECHOCARDIOGRAM**

E1.	Echocardiogram ejection fraction obtained	BECHOEF YES 1	NO.....2 (E2)	UNKNOWN.....-8 (E2)
	a. Date of most recent echocardiogram	___ ___ / ___ ___ / ___ ___	REMOVED	AGE_BECHOEFD
	b. LV ejection fraction	___ ___ %	BECHLVEF	
E2.	MUGA ejection fraction obtained	BMUGADON YES..... 1	NO.....2 (E3)	UNKNOWN.....-8 (E3)
	a. Date of most recent MUGA	___ ___ / ___ ___ / ___ ___	REMOVED	AGE_BMUGA_D
	b. LV ejection fraction	___ ___ %	BMUGALVEF	
E3.	Cardiac MRI ejection fraction obtained	BMRIDONE YES..... 1	NO.....2 (E4)	UNKNOWN.....-8 (E4)
	a. Date of most recent MRI	___ ___ / ___ ___ / ___ ___	REMOVED	AGE_BMRIDATE
	b. LV ejection fraction	___ ___ %	BMRILVEF	
E4.	Cardiac catheterization	YES..... 1 BCATHDON	NO.....2 (E5)	UNKNOWN.....-8 (E5)
	a. Date of most recent catheterization	___ ___ / ___ ___ / ___ ___	REMOVED	AGE_BCATH_D
	b. LVEDP	___ ___ mmHg	BLVEDP	
	c. PCWP	___ ___ mmHg	BPCWP	
	d. Cardiac index	___ . ___ L/min/m ²	BCARDIN	
E5.	Exercise Testing	YES 1 BEXTEST	NO.....2 (E6)	UNKNOWN.....-8 (E6)
	a. Date of most recent exercise test	___ ___ / ___ ___ / ___ ___	REMOVED	AGE_BEXDATE

Form V102: Medical Record Review and Baseline Clinical Assessment

- b. Type of exercise test **BEXTYPE** BIKE 1 TREADMILL 2
- c. Height at exercise test **BHT_ECHO** ___ ___ . ___ cm
- d. Weight at exercise test **BWT_ECHO** ___ ___ . ___ kg
- e. Did patient achieve maximum effort? **BMAXEFFT**
 YES..... 1 NO..... 2 UNKNOWN -8
- f. Peak VO₂ **BPEAKVO2** ___ ___ . ___ ml/kg/min INDETERMINATE -8
- g. VO₂ at anaerobic threshold **BATVO2** ___ ___ . ___ ml/kg/min INDETERMINATE -8

- E6. Was BNP level obtained within 3 days before the baseline echocardiogram? **BBNPDONE**
 YES..... 1 NO 2 (END) UNKNOWN -8 (END)
- a. Standard BNP assay performed? **BBNPSTND**
 YES..... 1 NO 2 (E6b)
 - 1. Date of most recent BNP level
 _____ / _____ / _____
 REMOVED **AGE_BBNP_D**
 - 2. Assay result **BBNPRSLT** _____ . _____
 - a. Units **BSTNDU** PG/ML..... 1 PMOL/L... 2
 - b. N-terminal proBNP assay performed? YES..... 1 NO 2 (END) **BNTPBNP**
 - 1. Date of most recent NT-proBNP level
 _____ / _____ / _____
 REMOVED **AGE_BNTP_D**
 - 2. Assay result **BNTPRSLT** _____ . _____
 - a. Units PG/ML..... 1 PMOL/L..... 2 **BNTPU**