

A1. Site/Study ID #: _____ / _____

A2. Date of Exam: _____ / _____ / _____
Month Day Year

A3. Staff Initials: _____

A4. Follow-up visit (month): 1 ☐ 2 ☐ 3 ☐ 6 ☐ OR Age: _____ mo/yr To DCC ☐A5. Source of data (check all that apply): a. ☐ Attending physician b. ☐ Medical record**SECTION B: VITAL SIGNS**8. ☐ ND → Complete Form 40 Protocol Deviation if P004

B1. Blood Pressure _____ / _____ mm Hg

B2. Heart rate _____ beats/min

B4. Temperature _____ °

B4a. Temperature taken: 1. ☐ Axillary 2. ☐ Rectal 3. ☐ Tympanic 4. ☐ Temporal artery 5. ☐ OralB5. Oxygen saturation (upright position): _____ % 8. ☐ ND**SECTION C: ANTHROPOMETRICS – Measure skinfold in triplicate and record mean**8. ☐ ND → Complete Form 40 Protocol Deviation

C1. Weight: _____ . _____ kg OR _____ lbs _____ oz

C2. Height or length: _____ . _____ cm OR _____ . _____ inches

C3. Head circumference: _____ . _____ cm OR _____ . _____ inches 8. ☐ NDC4. Right mid arm circumference: _____ . _____ cm 8. ☐ NDC5. Right triceps skinfold thickness: _____ . _____ mm 8. ☐ ND**SECTION D: PHYSICAL EXAM**8. ☐ ND → Complete Form 40 Protocol Deviation

D2. Pruritus:

1. ☐ None
2. ☐ Mild scratching when undistracted
3. ☐ Active scratching without abrasion
4. ☐ Active scratching with abrasions
5. ☐ Cutaneous mutilation with bleeding and scarring

D3. Xanthoma 1. ☐ Absent 2. ☐ PresentD4. Peripheral edema 1. ☐ Absent 2. ☐ Present

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D5. Cyanosis (check all that apply):

- a. ☐ Central (e.g., lips)
b. ☐ Peripheral (e.g., fingers, toes)
c. ☐ None
d. ☐ No information given

D6. Clubbing 1. ☐ Absent 2. ☐ Present

D7. Facial features (check all that apply):

- a. ☐ Normal → **Go to D9**
b. ☐ Dysmorphic facial features (check all that apply below):
 bi. ☐ Triangular face
 bii. ☐ Wide nasal bridge
 biii. ☐ Prominent forehead
 biv. ☐ Low set ears
 bv. ☐ Deep set eyes
 bvi. ☐ Other (Specify: _____)
 bvii. ☐ No information given
c. Do these features suggest a known syndrome (check all that apply)?
 ci. ☐ No
 cii. ☐ Alagille syndrome
 ciii. ☐ Other (Specify: _____)
 civ. ☐ No information given
d. ☐ Other facial anomalies (Specify: _____)

LIVER, SPLEEN & ASCITES8. ☐ ND → **Complete Form 40 Protocol Deviation**

D9. Liver:

- b. Liver span: ____ cm at right (left) mid-clavicular line
c. Liver edge: ____ cm below right (left) costal margin 1. ☐ Liver edge not palpable
d. ____ cm below xiphoid 1. ☐ Liver edge not palpable
e. Liver texture: 1. ☐ Soft 2. ☐ Firm 3. ☐ Hard
4. ☐ Nodular and hard 5. ☐ Not palpable

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D10. Spleen:

b. Spleen size ____ cm below the left (right) costal margin 4. ☐ Not palpable

D11. Ascites:

1. ☐ Absent2. ☐ Present

SECTION E: ANOMALIES AND ABNORMALITIES

8. ☐ ND

Review each of the following items below and check the appropriate box.

Body System	Normal	Abnormal	Not Done	If abnormal, specify abnormality	If abnormality may be related to P004 treatment (Complete Form 45)
E1. Appearance	1. ☐	2. ☐	8. ☐		1. ☐
E2. Skin	1. ☐	2. ☐	8. ☐		1. ☐
E3. HEENT	1. ☐	2. ☐	8. ☐		1. ☐
E4. Neck and thyroid	1. ☐	2. ☐	8. ☐		1. ☐
E5. Lungs and Chest	1. ☐	2. ☐	8. ☐		1. ☐
E6. Lymphatic	1. ☐	2. ☐	8. ☐		1. ☐
E7. Heart	1. ☐	2. ☐	8. ☐		1. ☐
E8. Abdomen	1. ☐	2. ☐	8. ☐		1. ☐
E9. Musculoskeletal	1. ☐	2. ☐	8. ☐		1. ☐
E10. Neurological	1. ☐	2. ☐	8. ☐		1. ☐
E11. Other	1. ☐	2. ☐	8. ☐		1. ☐

OTHER EVALUATIONS

D15. Tanner Score (if child is 8 years or older or if precocious puberty is suspected)

9. ☐ Refused8. ☐ NAa. Development ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5b. Pubic hair ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5

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Month Day YearD16. Was the child previously listed for transplant? 1. ☐ No → **Go to D17** 2. ☐ Yes → **If new listing, complete Form 25L**

a. If yes, was the child removed from the transplant list since the last research visit?

1. ☐ No → **Go to D17**2. ☐ Yes3. ☐ Yes, but relisted

b. If removed, why?

1. ☐ Improved2. ☐ Too ill for transplant3. ☐ Family wishes4. ☐ Other (Specify: _____)

D17. Biopsy to be/was performed at BARC site?

1. ☐ No2. ☐ Yes → **Complete Forms 48 and 30**

D18. Transplant to be/was performed?

1. ☐ No2. ☐ Yes → **Complete Forms 48 and 30; and Form 35 if end of study**

D19. Was there a change in the diagnosis?

1. ☐ No2. ☐ Yes → **Complete Form 29**