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. ☐ Oral

B5. 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. ☐ ND

C4. Right mid arm circumference: ___ ___ . ___ cm 8. ☐ ND

C5. 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. ☐ Present

D4. Peripheral edema 1. ☐ Absent 2. ☐ Present

A1. Site/Study ID #:___ ___ / ___ ___ ___ ___ A2. Date of Exam:___ ___ / ___ ___ / ___ ___ ___ ___
Month Day Year

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 & ASCITES 8. ☐ 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

A1. Site/Study ID #:___ ___ / ___ ___ ___ ___ A2. Date of Exam:___ ___ / ___ ___ / ___ ___ ___ ___
Month Day Year

D10. Spleen:

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

D11. Ascites: 1. ☐ Absent

2. ☐ 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. ☐ Refused 8. ☐ NA

a. Development	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
b. Pubic hair	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

A1. Site/Study ID #:___ ___ / ___ ___ ___ ___ A2. Date of Exam:___ ___ / ___ ___ / ___ ___ ___ ___
Month Day Year

D16. 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. ☐ Yes 3. ☐ Yes, but relisted

b. If removed, why?

1. ☐ Improved 2. ☐ Too ill for transplant 3. ☐ Family wishes

4. ☐ Other (Specify:_____)

D17. Biopsy to be/was performed at BARC site? 1. ☐ No 2. ☐ Yes → **Complete Forms 48 and 30**

D18. Transplant to be/was performed? 1. ☐ No 2. ☐ Yes → **Complete Forms 48 and 30; and Form 35 if end of study**

D19. Was there a change in the diagnosis? 1. ☐ No 2. ☐ Yes → **Complete Form 29**