

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 SIGNS8. 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 mean8. 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 EXAM8. 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

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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):
 b.i. Triangular face
 b.ii. Wide nasal bridge
 b.iii. Prominent forehead
 b.iv. Low set ears
 b.v. Deep set eyes
 b.vi. Other (Specify: _____)
 b.vii. No information given
 c. Do these features suggest a known syndrome (check all that apply)?
 c.i. No
 c.ii. Alagille syndrome
 c.iii. Other (Specify: _____)
 c.iv. 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. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E2. Skin	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E3. HEENT	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E4. Neck and thyroid	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E5. Lungs and Chest	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E6. Lymphatic	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E7. Heart	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E8. Abdomen	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E9. Musculoskeletal	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E10. Neurological	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>
E11. Other	1. <input type="checkbox"/>	2. <input type="checkbox"/>	8. <input type="checkbox"/>		1. <input type="checkbox"/>

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

A1. Site/Study ID #: ____ / ____

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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. 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**