

Does not use table PHY5, the correct table and variable name is PHY2

A1. Site/Study ID #: _____ / **G** _____ / _____ A2. Visit Date: _____ / _____ / _____ A3. Staff Initials: _____
Month Day Year

A4. Visit: 1. Baseline 2. Year 1 3. Year 2 4. Year 3 5. Year 4 6. Year 5 7. LT/ABD
 8. Year 6 9. Year 7 10. Year 8 11. Year 9 12. Year 10 To DCC

A5. Source of data: a. Physician/designee PHY2A05A b. Medical record PHY2A05B V2(3)

SECTION B: PHYSICAL EXAM

B1. VITAL SIGNS:

- | | | |
|--|---|--|
| 1. Blood Pressure | PHY2B011M V2(3)/ _ PHY2B011HG V2(3)_ mm Hg | 88. <input type="checkbox"/> ND PHY2B011ND V2(3) |
| 2. Heart Rate | PHY2B012 V2(3) beats/min | 88. <input type="checkbox"/> ND PHY2B012ND V2(3) |
| 3. Respiratory rate | PHY2B013 V2(3) respirations per minutes | 88. <input type="checkbox"/> ND PHY2B013ND V2(3) |
| 4. Temperature | PHY2B014 V2(4) °C PHY2B014F °F | 88. <input type="checkbox"/> ND PHY2B014ND V2(3) |
| 5. Oxygen saturation (upright position): | PHY2B015 V2(4) % | 88. <input type="checkbox"/> ND PHY2B015ND V2(3) |

B2. ANTHROPOMETRICS:

- | | | | | | |
|--------------------------------------|---------------------|----|-------------------------|---------------------|--|
| 1. Weight: | PHY2B021KG V2(5)kg | OR | PHY2B021LB V2(4) lbs | PHY2B021OZ V2(4) oz | 88. <input type="checkbox"/> ND PHY2B021ND V2(3) |
| 2. Height or length: | PHY2B022CM V2(5) cm | OR | PHY2B022IN V2(5) inches | | 88. <input type="checkbox"/> ND PHY2B022ND V2(3) |
| 3. Head circumference (Age < 3yrs) | PHY2B023CM V2(4) cm | OR | PHY2B023IN V2(5) inches | | 88. <input type="checkbox"/> ND PHY2B023ND V2(3) |
| 4. Right mid arm circumference | PHY2B024CM V2(4) cm | | | | 88. <input type="checkbox"/> ND PHY2B024ND V2(3) |
| 5. Right triceps skinfold thickness: | PHY2B025M V2(4) mm | | | | 88. <input type="checkbox"/> ND PHY2B025ND V2(3) |
- (Measure skinfold in triplicate and record mean)

B3. PHYSICAL FINDINGS: 1. Done 88. Not Done – END PHY2B03 V2(3)

B4. SKIN EXAM: 1. Done 88. Not Done – Go to B5 PHY2B04 V2(3)

1. Jaundice:

- a. None PHY2B041A V2(3)
- b. Sclera PHY2B041B V2(3)
- c. Skin PHY2B041C V2(3)

2. Cyanosis: PHY2B042 V2(3)

- 1. None PHY2B042A
- 2. Peripheral (e.g., fingers, toes) PHY2B042B
- 3. Central (e.g. lips) PHY2B042C
- 4. No information given PHY2B042D
- 5. Peripheral (e.g., fingers, toes) and Central (e.g. lips)

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3. Pruritus PHY2B043 V2(3)

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

4. Xanthoma: PHY2B044V2(3)

- 1. None
- 2. Mild/moderate
 - a. Location: (Specify: PHY2B442SP)
- 3. Extensive
 - Location: (specify: PHY2B443SP V2(300))

B5. FACIAL FEATURES: 1. Done 88. Not Done – Go to B6 PHY2B05 V2(3)

1. Facial Features:

- 1. Normal PHY2B051 V2(3)
- 2. Dysmorphic facial features (check all that apply below):
 - a. Triangular face PHY2B51A V2(3)
 - b. Wide nasal bridge PHY2B51B V2(3)
 - c. Prominent forehead PHY2B51C V2(3)
 - d. Low set ears PHY2B51D V2(3)
 - e. Deep set eyes PHY2B51EV2(3)
 - f. Other (specify: PHY2B51F V2(3) _____ PHY2B51FSP V2(300) _____)
- 3. No information given

2. Do these features suggest a known syndrome? PHY2B052 V2(3)

- 1. No
- 2. Alagille syndrome
- 3. Other (specify: _____ PHY2B52SP V2(300) _____)
- 4. No information given

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B6. EYE EXAM (by investigator): 1. Done 88. Not Done – Go to B7 PHY2B06 V2(3)

1. Sclera: Icterus 1. Yes 2. No 88. Not Done PHY2B061 V2(3)

2. Iris: PHY2B062 V2(3) 1. Normal 2. Coloboma 3. Other abnormality (specify: PHY2B62SP V2(300)) 88. Not Done

3. Posterior embryotoxon seen (without slit lamp): 1. Yes 2. No 88. Not Done PHY2B063 V2(3)

4. Fundus: PHY2B064 1. Normal 2. Hyperpigmented Macula 3. Papilledema
 88. Not Done 4. Other (specify: _____ PHY2B64SP 2(300)_____)

5. Other Abnormality: (specify: _____ PHY2B65SP V2(300)_____)

6. Did the participant receive any eye exam by an ophthalmologist? 1. Yes 2. No PHY2B066 V2(3)

a. Results: PHY2B066A V2(3) 1. Normal 2. Abnormal (specify: PHY2B66ASP V2(300)_

B7. CHEST/LUNG Exam: 1. Done 88. Not Done - Go to B8 PHY2B07 V2(3)

1. Right side: PHY2B071 V2(3) 1. Normal 2. Wheezing 3. Other (Specify: PHY2B71SP V2(300))

2. Left side: PHY2B072 V2(3) 1. Normal 2. Wheezing 3. Other (Specify: __ PHY2B72SP V2(300))

3. Hyperexpanded chest: 1. Yes 2. No PHY2B073 V2(3)

B8. CARDIAC Exam: 1. Done 88. Not Done – Go to B9 PHY2B08 V2(3)

1. Heart Murmur: (check all that apply)

a. Absent PHY2B81A b. Functional Murmur PHY2B81B c. Peripheral pulmonary artery stenosis murmur PHY2B81C d. Other pathologic murmur PHY2B81D (Specify: _____PHY2B81SP_____)

B9. LIVER, SPLEEN & ASCITES Exam: 1. Done 88. Not Done - Go to B10 PHY2B09 V2(3)

1. Liver:

a. Liver location: 1. Normal (right side) 2. Midline 3. Left side PHY2B091A V2(3)

b. Liver span: PHY2B91BCM cm at right (left) mid-clavicular line OR 88. Not Done PHY2B91BND

c. Liver edge: PHY2B91CCM cm below right (left) costal margin OR 1. Liver edge not palpable PHY2B91C1 OR 88. Not Done PHY2B91CND
 PHY2B91CXC cm below xiphoid: OR 1. Liver edge not palpable PHY2B91CX1

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OR 88. Not Done PHY2B91CXND

PHY2B091D V2(3)d. Liver texture: 1. Soft 2. Firm 3. Hard 4. Nodular and hard 5. Not palpable

2. Spleen:

a. Spleen location: (check all that apply)

a. Normal (left side) PHY2B092AA b. Midline(wandering) PHY2B092AB
 c. Right Side PHY2B092AC d. Not palpable – Go to 3 PHY2B092AD

b. Spleen size: PHY2B92BCM V2(3) cm below the left (right) costal margin 1. Not Done _ PHY2B92BND V2(3)

c. Spleen texture: 1. Soft 2. Firm 3. Hard PHY2B092C V2(3)

3. Ascites: 1. Absent 2. Present PHY2B093 V2(3)

4. Ascites confirmed by ultrasound: 1. Yes 2. No 3. No information given PHY2B094 V2(3)

B10. GENITALIA Exam:

1. Done 88. Not Done - Go to B11 PHY2B10 V2(3)

1. Anomaly of the genitalia present: PHY2B101 V2(3) 1. Yes (Specify: __PHY2B101SP V2(300)____) 2. No

2. Tanner Stage (if participant is 8 years or older or if precocious puberty is suspected) 1. NA PHY2B102 V2(3)

9. Refused PHY2B102R

a. Development: 1. 1 2. 2 3. 3 4. 4 5. 5 PHY2B102A V2(3)

b. Pubic hair: 1. 1 2. 2 3. 3 4. 4 5. 5 PHY2B102B V2(3)

B11. EXTREMITIES EXAM:

1. Done 88. Not Done – skip to B12 PHY2B11 V2(3)

IF DONE,

1. All are normal? 1. Yes 88. No PHY2B111 V2(3)

IF all are NOT Normal, complete each of the following:

- a. Joints: PHY2B11A V2(3) 1. Normal 2. Abnormal (Specify: PHY2B11ASP V2(300))
- b. Fingers: PHY2B11B V2(3) 1. Normal 2. Short 3. Other Abnormality: (Specify: PHY2B11BSP V2(300))
- c. Toes: PHY2B11C V2(3) 1. Normal 2. Short 3. Other Abnormality: (Specify: PHY2B11CSP V2(300))
- d. Peripheral edema: PHY2B11D V2(3) 1. Absent 2. Present
- e. Clubbing: PHY2B11E V2(3) 1. Absent 2. Present
- f. Malformation of an extremity? PHY2B11F V2(3) 1. Yes (Specify: PHY2B11FSP V2(300)) 2. No

B12. NEUROLOGIC Exam:

1. Done 88. Not Done - END PHY2B12 V2(3)

1. Muscle tone: 1. Normal 2. Abnormal 88. Not Done PHY2B121 V2(3)

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a. If abnormal: 1. Hypotonic 2. Hypertonic 3. Flaccid PHY2B121A V2(3)
 4. Other (Specify: _____ PHY2B121ASP V2(300)_____)

2. Muscle strength: 1. Normal 2. Abnormal 88. Not Done PHY2B122 V2(3)
 PHY2B122A V2(3) a. If abnormal: 1. Weakness 2. Other (Specify: _____ PHY2B122ASP V2(300)_____)

3. Knee: deep tendon reflexes: PHY2B123 V2(3)
 1. Normal
 2. Present but diminished
 3. Absence
 4. Increased
 5. Markedly hyperactive
 88. Not Done

4. Ataxia: 1. Yes 2. No PHY2B124 V2(3)
 1. If Yes: 1. Mild 2. Moderate 3. Severe PHY2B1241 V2(3)

2. Check all involved:
 a. Upper extremities PHY2B1242A V2(3)
 b. Lower extremities PHY2B1242B V2(3)
 c. Truncal _____ PHY2B1242C V2(3) PHY2B1242CSP V2(300)

5. Extraocular movements: PHY2B125 V2(3)
 1. Normal
 2. Decreased upward gaze
 3. Decreased lateral gaze
 4. Other: (Specify: _____ PHY2B125SP V2(300)_____)

6. Other neurological abnormalities: (Specify: _____ PHY2B126SP V2(300)_____)

PHY2INSIG V2(3)

PHY2SIGMM V2(3)/ PHY2SIGDD V2(3)/ PHY2SIGYY V2(3)/ PHY2SIGDT

Investigator/Designee Signature

Date (MM/DD/YYYY)

PHY2CMMNT V2(800) Comment

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GIRLS SELECT ONE FROM EACH SET OF DRAWINGS BELOW.

SET ONE: The drawings below show 5 different stages of how the breasts grow. A girl can go through each of the 5 stages as shown. Please look at each drawing and read the sentences that match the drawings. Then, mark an "X" in the box above the drawing that you think is closest to your stage of breast growth.

Name _____
 D.O.B. _____ Age _____
 Medical Record No. _____

Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>
The nipple is raised a little. The rest of the breast is still flat.	This is the breast bud stage. In this stage, the nipple is raised more than in stage 1. The breast is a small mound. The areola is larger than stage 1.	The breast and areola are both larger than in stage 2. The areola does not stick out away from the breast.	The areola and the nipple make up a mound that sticks up above the shape of the breast. NOTE: This stage may not happen at all for some girls. Some girls develop from stage 3 to stage 5 with no stage 4.	This is the mature adult stage. The breasts are fully developed. Only the nipple sticks out in this stage. The areola has moved back in the general shape of the breast.

SET TWO: The drawings below show 5 different stages of female pubic hair growth. A girl goes through each of the 5 stages as shown. Please look at each drawing and read the sentences below that match each drawing. Then, mark an "X" in the box above the drawing that you think is closest to the amount of your pubic hair growth.

Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>	Stage <input type="checkbox"/>
There is no pubic hair at all.	There is a little soft, long, lightly-colored hair. This hair may be straight or a little curly.	The hair is darker in this stage. It is coarser and more curly. It has spread out and thinly covers a bigger area.	The hair is now as dark, curly, and coarse as that of an adult female. The area that the hair covers is not as big as that of an adult female. The hair has NOT spread out to the legs.	The hair is now like that of an adult female. It covers the same area as that of an adult female. The hair usually forms a triangular (∇) pattern as it spreads out to the legs.

Adapted from: Morris, N.M., and Udry, J.R., (1980). Validation of a Self-Administered Instrument to Assess Stage of Adolescent Development. *Journal of Youth and Adolescence*, Vol. 9, No. 3: 271-280.

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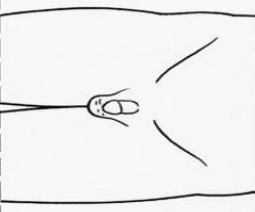
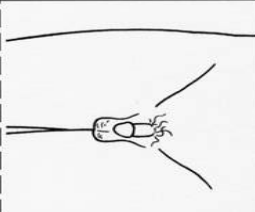
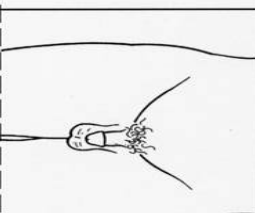
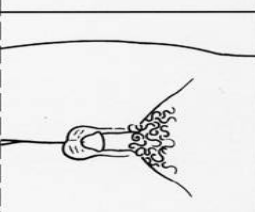
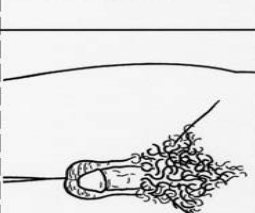
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BOYS

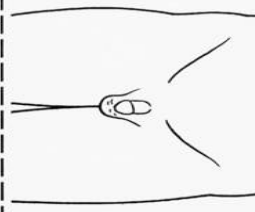
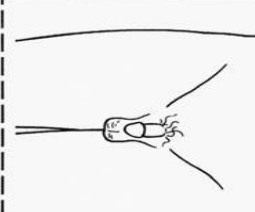
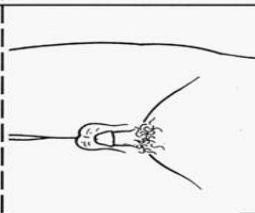
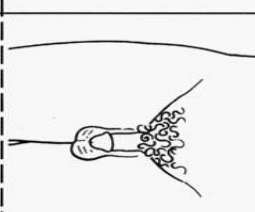
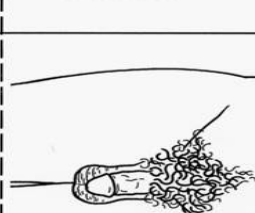
SELECT ONE FROM EACH SET OF DRAWINGS BELOW.

SET ONE: The drawings below show 5 different stages of testes, scrotum, and penis growth. A boy can go through each of the 5 stages as shown. Please look at each drawing and read the sentences that match the drawings. Then, mark an "X" in the box above the drawing that you think is closest to your stage of penis, scrotum, and testes growth. Do not look at or select for pubic hair growth with this set of drawings.

Name _____
 D.O.B. _____ Age _____
 Medical Record No. _____

 The testes, scrotum and penis are about the same size and shape as they were when you were a child.	<input type="checkbox"/> Stage 1	 The testes and scrotum are bigger. The skin of the scrotum has changed. The scrotum, the sack holding the testes, has gotten lower. The penis has gotten only a little bigger.	<input type="checkbox"/> Stage 2	 The penis has grown in length. The testes and scrotum have grown and dropped lower than in drawing 2.	<input type="checkbox"/> Stage 3	 The penis has gotten even bigger. It is wider. The glans (the head of the penis) is bigger. The scrotum is darker than before. It is bigger because the testes are bigger.	<input type="checkbox"/> Stage 4	 The penis, scrotum, and testes are the size and shape of an adult man.	<input type="checkbox"/> Stage 5
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SET TWO: The drawings below show 5 different stages of male pubic hair growth. A boy can pass through each of the 5 stages as shown. Please look at each drawing and read the sentences below that match each drawing. Then, mark an "X" in the box above the drawing that you think is closest to your stage of your pubic hair growth. Do not look at or select for penis size with this set of drawings.

 There is no pubic hair at all.	<input type="checkbox"/> Stage 1	 There is a little soft, long, lightly-colored hair. Most of the hair is at the base of the penis. This hair may be straight or a little curly.	<input type="checkbox"/> Stage 2	 The hair is darker in this stage. It is more curled. It has spread out and thinly covers a bigger area.	<input type="checkbox"/> Stage 3	 The hair is now as dark, curly, and coarse as that of an adult man. The area that the hair covers is not as big as that of an adult man. The hair has NOT spread out to the legs.	<input type="checkbox"/> Stage 4	 The hair has spread out to the legs. The hair is now like that of an adult man. It covers the same area as that of an adult man.	<input type="checkbox"/> Stage 5
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