CLINICAL PROCEDURES FORM

ID NUMBER: ☐☐☐☐☐☐☐ FORM CODE: **CLN** VERSION: **B** 02/19/09 Visit Number ☐0☐1 SEQ # ☐0☐0

Instructions: *This form is to be completed for each patient to be included in the Registry after informed consent is received. Information is obtained through patient interview, or from patient chart or electronic medical record.*

I. PULMONARY FUNCTION

1. Spirometry measurement (recent stable state)?.................................... Y   N →**Go to 2**   U →**Go to 2**

    a. Most recent stable state spirometry date: *(mm/dd/yyyy)*........... ☐☐/☐☐/☐☐☐☐

    Pre-Bronchodilator:

    b. FVC (liters / % predicted):... __.__ __ (L) / __ __ __ %

    c. FEV1 (liters / % predicted): .... __.__ __ (L) / __ __ __ %

    Post-Bronchodilator:

    d. FVC (liters / % predicted):... __.__ __ (L) / __ __ __ %

    e. FEV1 (liters / % predicted): .... __.__ __ (L) / __ __ __ %

II. IMMUNOLOGY AND ALLERGY

2. Has the patient had immunoglobulins measured? .... Y   N →**Go to 3**   U →**Go to 3**

    a. Most recent date immunoglobulins measured: *(mm/dd/yyyy)* ... ☐☐/☐☐/☐☐☐☐

    b. IgG: .... ☐☐☐☐ (mg / dL)

    c. IgM: .... ☐☐☐ (mg / dL)

    d. IgA:... ☐☐☐ (mg / dL)

    e. IgE:... ☐☐☐ (IU / mL)*

      ***NOTE:** IU and U are the same unit. IU/mL = U/mL = kU/L*

3. Has the patient had IgG subclasses measured? .... Y   N →**Go to 4**   U →**Go to 4**

    a. Most recent IgG subclass measurement date: *(mm/dd/yyyy)* ... ☐☐/☐☐/☐☐☐☐

    b. IgG1: .... ☐☐.☐☐ (mg / mL)

    c. IgG2: .... ☐.☐☐☐ (mg / mL)

    d. IgG3: .... ☐.☐☐☐ (mg / mL)

    e. IgG4: .... ☐.☐☐☐ (mg / mL)

4. Has the patient's humoral antibody response been tested? .... Y   N →**Go to 5**   U →**Go to 5**

   a. Most recent date antibody tested *(mm/dd/yyyy)*? .... ☐☐/☐☐/☐☐☐☐

Please use the following codes to report antigens tested: (circle one)
(A=Baseline, P=Post-immunization, B=Both, N=Not Tested, U=Unknown)

   b. Pneumococcal, 23 valent polysaccharide .... A   P   B   N →**Go to c** U →**Go to c**

      b1. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

   c. Pneumococcal, 7 valent protein .... A   P   B   N →**Go to d** U →**Go to d**

      c1. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

   d. Haemophilus influenza b .... A   P   B   N →**Go to e** U →**Go to e**

      d1. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

   e. Tetanus .... A   P   B   N →**Go to f** U →**Go to f**

      e1. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

   f. Other antigen .... A   P   B   N →**Go to 5** U →**Go to 5**

      f1. (please specify) .... _____

      f2. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

   g. Other antigen .... A   P   B   N →**Go to 5** U →**Go to 5**

      g1. (please specify) .... _____

      g2. Results: (N=Normal, A=Abnormal, U=Unknown) .... N   A   U

5. Has the patient had a rheumatoid factor performed? .... Y   N →**Go to 6**   U →**Go to 6**

   a. Most recent date factor performed: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

   b1. Nephlometry: .... __ __ . __ **IU/ml**   **and/or**

   b2. Agglutination:…………………………………………………….. __: __ __ **titer**

6. Has the patient had Allergic Bronchopulmonary Aspergillosis (ABPA) testing done? .... Y   N →**Go to 7**   U →**Go to 7**

   a. Most recent date of ABPA tests: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

   b. Total IgE: .... __ __ __IU/mL

   c. White Blood Cell (WBC) count: .... __ __.__ **x 10^9 L**

   d. % eosinophils: .... __ __.__ %

   e. # eosinophils: .... __ __ __ cells/µL

   f. Aspergillus skin test: .... 1 = Positive   2 = Negative

   g. Serum IgE to *Aspergillus fumigatus*: .... __ __ __ __ __kU/L

   h. Aspergillus precipitins: (1 or more positive) .... Y   N

   i. Serum IgG to *Aspergillus fumigatus*: .... __ __ __. __ mg/mL

III. GENETICS

7. Has the patient had alpha-1 antitrypsin (AAT) tested? .... Y   N →**Go to 8**   U →**Go to 8**

    a. Most recent date AAT tested: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

    b. Level: .... __ __ __ mg/dl

    c. Phenotype: *(circle one)*: .... MM   MZ   ZZ   SZ   MS   SS

8. Has the patient been tested for cystic fibrosis? .... Y   N →**Go to 9**   U →**Go to 9**

    a. Test date: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

      a1. Sweat Cl: .... ☐☐☐ (mmol / L)

    b. Genotype: .... Y   N →**Go to 9**   U →**Go to 9**

      b1. Test date: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

      b2. Lab: Ambry (A); Genzyme (G); Local (L); Other commercial (O):   A   G   L   O

      b3. Mutation 1: Specify or None detected (ND) .... _____   ND

      b4. Mutation 2: Specify or None detected (ND) .... _____   ND

9. Has the patient undergone a genetic study for Primary Cilia Dyskinesia (PCD)? .... Y   N →**Go to 10**   U →**Go to 10**

    a. Most recent date of PCD study *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

    b. Result: .... _____

IV. OTHER TESTING

10. Has the patient had nasal potential difference measured? .... Y   N →**Go to 11**   U →**Go to 11**

    a. Most recent date of nasal difference: (mm/dd/yyyy) .... ☐☐/☐☐/☐☐☐☐

    b. Diagnostic for: .... CF = 1   Normal = 2

    c: Other diagnostic: .... _____

11. Has the patient had GERD testing? .... Y   N →**Go to 12**   U →**Go to 12**

    a. Date of most recent GERD testing: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

    b. Esophagram test? .... Y   N →**Go to 11c**   U →**Go to 11c**

      b1. Reflux noted on esophagram? .... Y   N   U

      b2. Other findings: (please specify) .... _____

    c. Ambulatory pH monitoring? .... Y   N →**Go to 11d**   U →**Go to 11d**

      c1. Reflux noted on esophagram? .... Y   N   U

11 cont.

d. Endoscopy? .... Y   N →**Go to 11e**   U →**Go to 11e**

    d1. Findings consistent with reflux? .... Y   N   U

    d2. Other findings: (please specify)..................................... _____

e. Empiric treatment trial (PPI or H_2 blocker)? .... Y   N →**Go to 12**   U →**Go to 12**

    e1. Improvement in symptoms (cough, chest pain)?: .... Y   N   U

12. Has the patient had an echocardiogram?... Y   N →**Go to 13**   U →**Go to 13**

    a. Date of most recent echocardiogram: *(mm/dd/yyyy)*................. [ ][ ]/[ ][ ]/[ ][ ][ ][ ]

<u>**Chambers:**</u>

    b. Left ventricular size and function abnormal?............................ Y   N →**Go to 12c**   U →**Go to 12c**

      Findings (please specify)... _____

    c. Left atrial size and appearance abnormal? .... Y   N →**Go to 12d**   U →**Go to 12d**

      Findings (please specify)... _____

    d. Right ventricular size and function abnormal? .... Y   N →**Go to 12e**   U →**Go to 12e**

      Findings (please specify)... _____

    e. Right atrial size and appearance abnormal? .... Y   N →**Go to 12f**   U →**Go to 12f**

      Findings (please specify)... _____

    f. Was Right Ventricular Systolic Pressure (RVSP) estimated?.... Y   N →**Go to 12g**   U →**Go to 12g**

      f1. RVSP... __ __ __ mmHg

<u>**Valves:**</u>

NB: "Regurgitation" refers to greater than "trace" or "physiologic" amounts of regurgitant flow.

    g. Was mitral regurgitation noted?.. Y   N   U

    h. Was mitral stenosis noted? .... Y   N   U

    i. Was mitral valve prolapse noted?... Y   N   U

    j. Was aortic regurgitation noted? .... Y   N   U

    k. Was aortic stenosis noted? .... Y   N   U

    l. Was tricuspid regurgitation noted? .... Y   N   U

    m. Was tricuspid stenosis noted?... Y   N   U

    n. Was pulmonic regurgitation noted?.. Y   N   U

    o. Was pulmonic stenosis noted?... Y   N   U

12. cont.

Other:

   p. Was the pericardium abnormal or was an effusion present? .... Y   N →**Go to 12q**   U →**Go to 12q**

       Findings (please specify).. _____

   q. Were the great vessels abnormal?... Y   N →**Go to 12r**   U →**Go to 12r**

       Findings (please specify).. _____

   r. Were other abnormalities noted? .... Y   N →**Go to 13**   U →**Go to 13**

       Findings (please specify).. _____

13. Has the patient had a bronchoscopy?... Y   N →**Go to 14**   U →**Go to 14**

   a. Most recent bronchoscopy date: *(mm/dd/yyyy)*........................ ☐☐/☐☐/☐☐☐☐

   b. Were airway abnormalities present? .... Y   N →**Go to 14**   U →**Go to 14**

       Finding(s): Please specify: .... _____

14. Has the patient been tested for exhaled breath condensates? ....Y   N →**Go to 15**   U →**Go to 15**

   a. Most recent test for breath condensates: *(mm/dd/yyyy)*............ ☐☐/☐☐/☐☐☐☐

   b. What type?... _____

   c. Results: .... _____

15. Has the patient had a mucosal biopsy (for cilia)? .... Y   N →**Go to 16**   U →**Go to 16**

   a. Most recent mucosal biopsy date: *(mm/dd/yyyy)* .... ☐☐/☐☐/☐☐☐☐

   b. Normal electron microscopic ultrastructure: .... Y   N   U

   c. Outer dynein arm defect: .... Y   N   U

   d. Inner dynein arm defect: .... Y   N   U

   e. Central apparatus defect:... Y   N   U

   f. Other abnormality: .... Y   N →**Go to 15g**   U →**Go to 15g**

      f1. Please specify: .... _____

   g. Light microscopy or ciliary beat frequency assessment done? . Y   N →**Go to 16**   U →**Go to 16**

      g1. Was it normal?... Y   N   U

16. Has the patient had nasal nitric oxide measured?................................ Y   N →**Go to 17**   U →**Go to 17**

   a. Most recent date nitric oxide measured: *(mm/dd/yyyy)*............. ☐☐/☐☐/☐☐☐☐

   b. Result:... ☐☐☐.☐ (nL / min)

V. ADMINSTRATIVE INFORMATION:

17. Exam / Interview / medical records date: *(mm/dd/yyyy)*........................ ☐☐/☐☐/☐☐☐☐

18. Source of Data:
    a. Participant interview.. Y   N
    b. Participant exam .... Y   N
    c. Medical records review/abstraction .... Y   N

19. Form date: *(mm/dd/yyyy)*... ☐☐/☐☐/☐☐☐☐

20. Interviewer's / recorder's initials:... ☐☐☐