ALPHA 1-ANTITRYPSIN DEFICIENCY REGISTRY
PULMONARY FUNCTION TEST RESULTS

Form Completion Instructions: (See also Appendix 9.5)

This form is used to record the results of the pulmonary function tests for the tests performed at the Clinical Center. All the appropriate numerical and graphical data for the test session with each effort labelled and sent to the Clinical Coordinating Center.

QUESTION #	ITEM	INSTRUCTIONS
8-9	Weight, Height	These two items should be recorded in units requested.
10	Tech Number	Use the technicians number assigned by the Clinical Coordinating Center. This number should be used throughout the duration of the Registry.
12-16	Patient Status	These questions are those which are part of the triage letter as well. If any of these answers are Yes, the pulmonary function test should be delayed until later in the day, if possible. Review these questions/guidelines again with the patient to explain their importance in testing lung function. Inform the patient that testing may have to be rescheduled should the patient fail to adhere to these guidelines.
17	Acute Respiratory Illness	Hopefully, if the patient has had this condition, it will be caught using the triage letter and the patient will be reschedule for the test. If the answer to item #17 is "Yes", try to reschedule for a time three weeks or more after the infection clears up. Be sure the rescheduled visit is still within the six month time window for the visit.

ALPHA 1-ANTITRYPSIN DEFICIENCY REGISTRY
PULMONARY FUNCTION TEST RESULT

Form Completion Instructions:

QUESTION #	ITEM	INSTRUCTIONS
20,33,44	Instrument	Use the instrument number assigned by the Clinical Coordinating Center. That number should be used each time Form #03 is completed to indicate the instrument used. Remember, from visit to visit for each particular patient, it is recommended that the same equipment be used. If a test is not done, enter 0000 as the instrument code and skip to the next section or write "NOT DONE" in large print over the section not tested. If a new machine not assigned a code number, is used, (preferably the Clinical Coordinating Center will already be aware of new equipment - prior to its use) leave this space blank AND complete and submit Form #14 for the machine.

Definitions:

SVC = Slow Vital Capacity

FVC = Forced Vital Capacity

Total amount of air that can be expelled in one forced exhalation after a maximal inhalation

FEV_1 = Forced Expiratory Volume in one second

Maximum volume of air exhaled within the first second of exhalation

FRC = Forced Respiratory Capacity

ERV = Expiratory Reserve Volume

RV = Residual Volume

IC = Inspiratory Capacity

TLC = Total Lung Capacity

DLCO = Diffusing Lung Capacity

V_A = Alveolar Volume

ALPHA 1-ANTITRYPSIN DEFICIENCY REGISTRY
PULMONARY FUNCTION TEST RESULTS

Form Completion Instructions:

Form #03 - PFT Results Form accommodates the reporting of spirometry, lung volume and diffusing capacity results. Pre- and post-bronchodilator spirometry are the minimum pulmonary function data requested by the Registry. Lung volumes and diffusion capacity tests are NOT mandatory; space was left on the form for reporting this data if the results are available. In order to assess the technical acceptability of these pulmonary function tests, the Clinical Coordinating Center is requesting the following information. If this information is not available, the data should still be reported on Form #03, PFT Results Form. Be sure to label each effort as reported on Form #03.

SUMMARY OF DATA REQUESTED FOR THE REGISTRY

I. Pulmonary Function Data requested by the Clinical Coordinating Center

A. Spirometry

1. Copy of volume-time tracing of 3 Liter Syringe Calibration
2. Volume-time tracing (or copy) use patient Registry ID#
   a. Volume scale at least 10 mm/L
   b. Time scale at least 20 mm/second
3. Flow-volume tracing (or copy) use patient Registry ID#
   a. Flow scale at least 5 mm/L/sec
   b. Volume scale at least 10 mm/L
4. Form PFT #03 including expiratory times (FET)
5. Printout of numeric parameters showing the three reported efforts (screen dumps are adequate if formal report showing all three efforts is not available)

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PULMONARY FUNCTION TEST RESULTS

SUMMARY OF DATA REQUESTED FOR THE REGISTRY (CONTINUED)

B. Lung Volumes
   1. Helium dilution
      a. Helium-time tracing (or copy)
      b. Tidal volume-time tracing (or copy) during dilution test
      c. Volume-time tracing (or copy) showing tidal volume and SVC during measurement of lung subdivisions.
      d. Form PFT #03
      e. Printout showing lung subdivisions
   2. Nitrogen washout
      a. Expired nitrogen-time tracing (or copy)
      b. Tidal volume-time tracing (or copy) during washout test
      c. Volume-time tracing (or copy) showing tidal volume and SVC during measurement of lung subdivisions
      d. Form PFT #03
      e. Printout showing lung subdivisions
   3. Body plethysmography
      a. Plethysmographic tracings (or copy) including calibration factors

C. Single Breath Diffusing Capacity (S.B. DLCO)
   1. Form PFT #03
   2. Volume-time tracing of maneuvers
   3. Printout showing raw data used to calculate DLCO (He initial, He final, CO initial, CO final, breathholding time, inspiratory vital capacity, calculated V_A, etc.); delete patient name

ALPHA 1-ANTITRYPSIN DEFICIENCY REGISTRY
Pulmonary Function Test Results Form

This form should be completed for each set of Pulmonary Function Tests done. It should be included in the package sent to the Clinical Coordinating Center accompanied by the required printouts and tracings.

1. Date form completed: F03Q01_fzd (fuzzed) ___ ___/ ___ ___/ ___ ___ (month / day / year)
2. Patient Registry ID: Newid (scrambled) ___ ___ ___ ___ ___
3. Patient name code: namecode (censored) ___ ___ ___ ___
4. Clinical Center code number: clinic (censored) ___ ___
5. Date of tests: F03Q05_fzd (fuzzed) ___ ___/ ___ ___/ ___ ___ (month / day / year)
   visit number vsno
6. Visit type: F03Q06 ___(1)Initial ___(2)Follow-Up
7. Time of day (24-hour clock): F03Q07 ___ ___ : ___ ___
8. Patient weight (kg): F03Q08 ___ ___ ___ . ___
9. Patient height (cm): F03Q09 ___ ___ ___ . ___

NOTE: If the machine being used is not the primary or secondary machine be sure to use Form #14 to inform the CCC of changes in equipment.

10. Technician study number: F03Q10 Not Research Related
11. a. Ambient temperature (°C): F03Q11A ___ ___ . ___
    b. Barometric pressure (mmHg): F03Q11B ___ ___ ___
    c. Conversion factor (ATPS to BTPS): F03Q11C ___ . ___ ___ ___

A copy of the 3L syringe calibration should be attached to this form.

PATIENT STATUS

12. Did patient use a prescription or non-prescription inhaled bronchodilator within 8 hours prior to testing? F03Q12 ___(1)Yes ___(2)No
13. Did patient smoke any tobacco (cigarettes, cigars, pipes, etc.) within 2 hours prior to testing? F03Q13 ___(1)Yes ___(2)No
14. Did patient use caffeine-containing products (coffee, tea, colas, Dr. Pepper, Mountain Dew, No-Doz, Vivarin, Midol, Anacin, Excedrin, etc.) within 6 hours prior to testing? F03Q14 ___(1)Yes ___(2)No
15. Did patient eat a large meal within 1 hour prior to testing? F03Q15 ___(1)Yes ___(2)No

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Patient Registry ID: ___ ___ ___ ___ ___
Date of Tests: ___ ___/ ___ ___/ ___ ___
month day year

16. Has the patient taken a Theophylline preparation recently? F03Q16 ___(1)Yes ___(2)No
If NO, skip to Question 17.

If YES,
a. Specify drug taken: F03Q16A (never entered)

b. How many hours before first spirometry was last dose taken? F03Q16B ___ . ___

17. Has patient had an acute respiratory infection (including colds, influenza, acute bronchitis, pneumonia, pleurisy or abdominal and/or chest surgery) within 3 weeks prior to the visit? F03Q17 ___(1)Yes ___(2)No

If YES, specify: never entered

If YES to Question 17, try to reschedule PFT testing for at least three weeks post surgery or after the infection has cleared.

a. Was patient rescheduled? F03Q17A ___(1)Yes ___(2)No

If NO, why not: never entered

18. Has patient had any other co-morbid condition that may affect the PFT? F03Q18

___(1)Yes (specify): ___(2)No ___(9)Unknown

PRE-BRONCHODILATOR SPIROMETRY: (Record the three BEST of up to eight efforts)

19. Patient position: F03Q19 ___(1)Sitting ___(2)Standing

20. Instrument used: F03Q20 (not research related) ___ ___ ___ ___

	TEST 1	TEST 2	TEST 3
21. SVC (L) (BTPS):	F03Q21A ___	F03Q21B	F03Q21C
22. FVC (L) (BTPS):	F03Q22A ___	F03Q22B	F03Q22C
a. Forced expiratory time ($FET_{100\%}$)(sec)	F03Q22AA	F03Q22AB	F03Q22AC
23. FEV_1 (L) (BTPS):	F03Q23A	F03Q23B	F03Q23C
24. FEV_1/FVC (%):	F03Q24A	F03Q24B	F03Q24C

25. If all three tests are not recorded here, state reason:

F03Q25

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Patient Registry ID: ___ ___ ___ ___ ___
Date of Tests: ___ ___/ ___ ___/ ___ ___
month day year

POST-BRONCHODILATOR SPIROMETRY: (Record the three BEST of up to eight efforts)

26. Number of minutes spirometry done post-bronchodilator treatment: F03Q26 ___ ___

27. Type of bronchodilator treatment: F03Q27

___(1)Albuterol ___(2)Isoproteronol ___(3)Other (Specify): _____

	TEST 1	TEST 2	TEST 3
28. SVC (L) (BTPS):	F03Q28A	F03Q28B	F03Q28C
29. FVC (L) (BTPS):	F03Q29A	F03Q29B	F03Q29C
a. Forced expiratory time ($FET_{100\%}$)(sec)	F03Q29AA	F03Q29AB	F03Q29AC
30. FEV_1 (L) (BTPS):	F03Q30A	F03Q30B	F03Q30C
31. FEV_1/FVC (%):	F03Q31A	F03Q31B	F03Q31C

32. If all three tests are not recorded here, state reason:

F03Q32

LUNG COMPARTMENTS (Please attach all available numeric and graphic data relating to lung compartments and DLCO. Columns for both pre- and post-bronchodilators are provided if the tests are done at all. If either pre- OR post is done, simply complete the applicable column and mark the other column "not done".)

	PRE BRONCHODILATOR	POST BRONCHODILATOR
33. Instrument used:	F03Q33A Not Research Related →	F03Q33B ___ ___
34. Patient position:	F03Q34A ___(1)Sit ___(2)Stand	F03Q34B ___(1)Sit ___(2)Stand
35. FRC (L) (BTPS):	F03Q35A ___ ___ . ___ ___	F03Q35B ___
36. ERV (L):	F03Q36A ___.___ ___	F03Q36B
37. RV (L):	F03Q37A ___ ___.___ ___	F03Q37B
38. SVC (L):	F03Q38A ___.___ ___	F03Q38B
39. IC (L):	F03Q39A ___.___ ___	F03Q39B

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Patient Registry ID: ___ ___ ___ ___ ___
Date of Tests: ___ ___/ ___ ___/ ___ ___
month day year

	PRE BRONCHODILATOR	POST BRONCHODILATOR
40. TLC (by gas dilution) (L):	F03Q40A ___.___ ___	F03Q40B
41. TLC (by body box) (L):	F03Q41A ___.___ ___	F03Q41B
42. TGV measured at:	F03Q42A (1)TLC ___(2)FRC	F03Q42B ___(1)TLC ___(2)FRC
43. TLC (by x-ray planimetry) (L):	F03Q43A ___ ___	F03Q43B

DLCO: (To be measured twice at least four minutes apart)

44. Instrument used: F03Q44 (not research related) ___ ___ ___ ___

45. Patient position: F03Q45 ___(1)Sitting ___(2)Standing

46. a. When was test performed? F03Q46A
___(1)Pre-Bronchodilator ___(2)Post-Bronchodilator

For the following: Indicate two individual values if available OR, if not, indicate average value from printout. Do not correct for hemoglobin.

	VALUE 1	VALUE 2	OR	AVERAGE OF 2 TESTS
b. DLCO (mlCO/min/mmHg):	F03Q46B1	F03Q46B2	OR	F06Q46B3

c. Hemoglobin (g/dl) at time of DLCO: F03Q46C ___ ___.___

47. DL/VA (mlCO/min/mmHg/L) (BTPS):

F03Q471 ___ F03Q472 ___ OR F03Q473 ___

Comments: never entered

Form Completed By (Name): never entered

Physician Signature: never entered

The additional variables are listed on the following page.

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Notes on Coding:

Additional Calculated Variables included in the Form 3 database:

Variable Name	Description
PREDFEV1[1]	Predicted FEV1 (liters)
PREDFVC[1]	Predicted FVC (liters)
PREDDLCO[2]	Predicted DLCO (mlCO/min/mmHg)
PREFEV1	Maximal Pre-BD FEV1 (liters) = Max(F03Q23A,F03Q23B,F03Q23C)
PREFVC	Maximal Pre-BD FVC (liters) = Max(F03Q22A,F03Q22B,F03Q22C)
POSTFEV1	Maximal Post-BD FEV1 (liters) = Max(F03Q30A,F03Q30B,F03Q30C)
POSTFVC	Maximal Post-BD FVC (liters) = Max(F03Q29A,F03Q29B,F03Q29C)
PFEV1_CR	Pre-BD FEV1 % predicted
PFVC_CR	Pre-BD FVC % predicted
OPFEV_CR	Post-BD FEV1 % predicted
OPFVC_CR	Post_BD FVC % predicted
DLCO1	Average DLCO from two tests = F03Q46B3
DLCO2	Average of DLCO values 1 and 2 = Ave(F03Q46B1,F03Q46B2)
PFTDLCO	Max(DLCO1,DLCO2) =Value of DLCO used in analyses
PERCDLCO	DLCO % predicted = 100*(PFTDLCO/PREDDLCO)

(1) Predicted normal values for FEV1 and FVC were calculated using the predictive equations of Crapo et al. *Am Rev Respir Dis* 123: 659-664, 1981.

(2) Predicted normal values for DLCO were calculated using the predictive equations of Crapo et al. *Am Rev Respir Dis* 123: 185-189, 1981.