ALPHA 1-ANTITRYPSIN DEFICIENCY REGISTRY
ADVERSE REACTION FORM

Form Completion Instructions:

This form should be completed for each individual where an adverse reaction has occurred from receipt of augmentation therapy. Complete one form per reaction.

Be sure to record the Prolastin Lot Number and the dosage administered.

Refer to question 38a of Form #05A as well as the "Problem" column on Form 10 to gather information to complete this form.

If further information is required, contact the local caregiver.

This form is completed for each date therapy given where at least one adverse reaction has occurred. If no problems have arisen, do not complete form.

1. Date form completed: F11Q01_f2d (fuzzed) ___ ___/ ___ ___/ ___ ___ (month / day / year)
2. Patient Registry ID: NewID (scrambled) ___ ___ ___ ___ ___
3. Patient name code: namecode (censored) ___ ___ ___ ___
4. Clinical Center: clinic (censored) ___ ___
5. Date of augmentation therapy: Aug.Date_f2d (fuzzed) ___ ___/ ___ ___/ ___ ___ (month / day / year)
   a. Location of therapy: F11Q05a
      ___(1)Registry Clinical Center ___(4)Home
      ___(2)Local physician's office
      ___(3)Other (Specify) F11Q05a3 ___(9)Unknown
6. Prolastin lot number: F11Q06 ___ ___ ___ ___ ___ ___
   a. Additional Prolastin lot number used: F11Q06a ___ ___ ___ ___ ___ ___
   b. Additional Prolastin lot number used: F11Q06b
7. Dosage (grams): F11Q07 ___ ___ ___.___
   a. Frequency of therapy: F11Q07a ___(1)Weekly ___(2)Monthly ___(3)Other (specify): Never entered

Note: Coded by the CCC as the interval (days) between infusions. Code '98' represents "intermittent" or occasional use.

8a. Type of therapy: F11Q08 ___(1)Infusion ___(2)Other (specify): Never entered

b. When did the reaction occur? F11Q8B
   ___(1)During the infusion ___(4)Other (Specify) F11Q8B4
   ___(2)Immediately after the infusion but within 24 hours ___(9)Unknown
   ___(3)Greater than 24 hours after the infusion

c. Was hospitalization or emergency room treatment required?: F11Q8C ___(1)Yes ___(2)No

d. Was medication and/or contact with physician required?: F11Q8D ___(1)Yes ___(2)No

e. Was therapy discontinued permanently?: F11Q8E ___(1)Yes ___(2)No

f. Date therapy permanently discontinued: F11Q8F_f2d (fuzzed) ___ ___/ ___ ___/ ___ ___ (month / day / year)

g. What frequency did reaction occur? F11Q8G
   specify F11Q8G5

h. Was patient pre-medicated? F11Q8H
   specify F11Q8H1

See explanation pages following this form

White/Yellow: Clinical Coordinating Center, Pink: Clinical Center

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

Check each sign/symptom that occurred at the time of this therapy. Use the following codes for "Action Taken":

1=Flow rate decreased
2=Infusion temporarily ceased, then resumed
3=Infusion ceased, not resumed
4=Infusion continued, no change
5=Other (specify in comments)

		REACTION (check)	ACTION TAKEN (indicate code from above)
9. a.	Anaphylactic Shock:	F11Q09A1	F11Q09A2
b.	Anxiety:	F11Q09B1	F11Q09B2
c.	Chest Tightness:	F11Q09C1	F11Q09C2
d.	Chills:	F11Q09D1	F11Q09D2
e.	Dizziness/Fainting:	F11Q09E1	F11Q09E2
f.	Dyspnea/Shortness of Breath:	F11Q09F1	F11Q09F2
g.	Emesis:	F11Q09G1	F11Q09G2
h.	Fever:	F11Q09H1	F11Q09H2
i	Flushing:	F11Q09I1	F11Q09I2
j.	Headache:	F11Q09J1	F11Q09J2
k.	Hives:	F11Q09K1	F11Q09K2
l.	Hypotension:	F11Q09L1	F11Q09L2
m.	Muscle Cramps:	F11Q09M1	F11Q09M2
n.	Nausea:	F11Q09N1	F11Q09N2
o.	Pain:	F11Q09O1	F11Q09O2
p.	Rash:	F11Q09P1	F11Q09P2
q.	Tachycardia:	F11Q09Q1	F11Q09Q2
r.	Wheezing:	F11Q09R1	F11Q09R2
s.	Other	F11Q09S1	F11Q09S2

(Specify): F11Q09S3 (see next page)

Comments: COMMENTS1
COMMENTS2

Form Completed By (Name): never entered

Physician Signature: never entered

White/Yellow: Clinical Coordinating Center, Pink: Clinical Center

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Notes to Form 11, Question 9

Based on the value in F11Q09S3 ["Other ....(Specify)"] the following variables were also coded into the dataset.

	REACTION (check)	ACTION TAKEN (indicate code from above)
9. (continued)		
t. Asthma attack	F11Q09T1	F11Q09T2
u. Arthralgia, body ache	F11Q09U1	F11Q09U2
v. Flu-like symptoms	F11Q09V1	F11Q09V2
w. Itching	F11Q09W1	F11Q09W2
x. Cyanosis	F11Q09X1	F11Q09X2
y. Indigestion/diarrhea	F11Q09Y1	F11Q09Y2
z. Warmth/sweating	F11Q09Z1	F11Q09Z2
aa. Tremors-shaking	F11Q9AA1	F11Q9AA2
ab. Edema-fluid retention	F11Q9AB1	F11Q9AB2
ac. Weakness-malaise	F11Q9AC1	F11Q9AC2
ad. miscellaneous	F11Q9AD1	F11Q9AD2

Variable: SEVERITY

This is an overall severity score:

(1) Severe	Presence of one or more of the following: anaphylactic shock, dyspnea, shortness of breath, wheezing, hypotension. Also included were asthma attack and cyanosis, both subject to physician review.
(2) Moderate	Absence of any "severe" symptoms, and presence of one or more of the following: chest tightness, chills, dizziness/fainting, emesis, fever, headache, hives, rash, tachycardia.
(3) Mild	Absence of any "severe" or "moderate" symptoms.

Notes on Coding:

Question 5a. Location of Therapy.

The codes shown on the form are those used to enter data. Codes appearing on previous versions of the forms differed from these.

Question 7a. Frequency of Therapy:

The interval in days between infusions is entered, e.g., 7, 14, 30. Code "98" is used to represent "intermittent" or "occasional" use.

Question 9a-s.

For each question, there are two variables, coded to capture information on whether the reaction occurred (1=yes, 0=blank), and what the action taken was (using codes indicated on the form, with 9=unknown).

9a. Anaphylactic shock:
F11Q09A1 - Anaphylactic shock, reaction
F11Q09A2 - Anaphylactic shock, action

. . .

9s. Other
F11Q09S1 - Other, reaction
F11Q09S2 - Other, action

Additional variables coded at the CCC:

Additional Signs and Symptoms:
The following additional variables were created to code other specific adverse reaction signs and symptoms not included as specific questions on the original form:

	Variable Names	
Adverse Reaction	Reaction	Action
Asthma attack	F11Q09T1	F11Q09T2
Arthralgia, body ache	F11Q09U1	F11Q09U2
Flu-like symptoms	F11Q09V1	F11Q09V2
Itching	F11Q09W1	F11Q09W2
Cyanosis	F11Q09X1	F11Q09X2
Indigestion-diarrhea	F11Q09Y1	F11Q09Y2
Warmth-sweating	F11Q09Z1	F11Q09Z2
Tremors-shaking	F11Q9AA1	F11Q9AA2
Edema-fluid retention	F11Q9AB1	F11Q9AB2
Weakness-malaise	F11Q9AC1	F11Q9AC2
Miscellaneous	F11Q9AD1	F11Q9AD2

Additional variables coded at the CCC (continued):

Severity of Reaction was coded at the CCC using the following algorithm:

SEVERITY = 1 Severe -	Defined as the presence of one or more of the following "severe" symptoms: dyspnea, shortness of breath, wheezing, hypotension
2 Moderate -	Absence of any "severe" symptom, and presence of one or more "moderate" symptoms
3. Mild -	Absence of any "severe" or "moderate" symptoms

Classification of symptoms in Question 9a:

Question	Severity
a. Anaphylactic shock	Severe - none in database
b. Anxiety	Mild
c. Chest tightness	Moderate
d. Chills	Moderate
e. Dizziness/fainting	Moderate
f. Dyspnea/SOB	Severe
g. Emesis	Moderate
h. Fever	Moderate
i. Flushing	Mild
j. Headache	Moderate
k. Hives	Moderate
l. Hypotension	Severe
m. Muscle cramps	Mild
n. Nausea	Mild
o. Pain	Mild
p. Rash	Moderate
q. Tachycardia	Moderate
r. Wheezing	Severe
t. Asthma attack	Severe*
u. Arthralgia, body ache	Mild
v. Flu-like symptoms	Mild
w. Itching	Mild
x. Cyanosis	Severe*
y. Indigestion/diarrhea	Mild
z. Warmth/sweating	Mild
aa. Tremors-shaking	Mild
ab. Edema-fluid retention	Mild
ac. Weakness-malaise	Mild
ad. Miscellaneous	

Other symptoms classified as "mild": "Flaky spells", "Mild depression", "Cold sensation", "Nasal congestion", "Gas", "Numbness around mouth".

* These symptoms were classified as indicated, subject to physician review

Additional variables coded at the CCC (continued):

Variable	Description
F11Q8G	With what frequency did the reaction(s) occur? Codes used: 1 Single episode 2 two or three times 3 greater than 3 times 4 with every infusion 5 Other 9 Unknown
F11Q8G5	If other frequency, specify Format: character $25
F11Q8H	Was patient pre-medicated? Codes used: 1 Yes 2 No
F11Q8H1	Was patient premedicated? Specify: Format: character $25
BADLOT	Was the adverse reaction associated with a known bad lot of Prolastin? Codes used 1 Yes 0 No