

**INSTRUCTIONS:** This form must be completed for all subjects at Study Visit 4 (final visit). This form has two functions: 1) **UPDATE** information for all relatives reported on at the Baseline visit (even if a relative has died since the Baseline visit); and 2) determine if any **ADDITIONAL RELATIVES** have been diagnosed with Marfan syndrome since the Baseline visit.

**Section A: KEY IDENTIFYING INFORMATION**

A1. Study Identification Number \_\_\_\_\_ - \_\_\_\_\_ - \_\_\_\_\_ - \_\_\_\_\_ - \_\_\_\_\_

A2. Study visit STUDY VISIT 4 (Month 36) ..... (4)

A3. Date of form completion \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_ Y Y Y Y  
M M D D Y Y Y Y

A4. Name of person completing form \_\_\_\_\_  
PRINT FULL NAME INITIALS

A5. Date of visit \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_ Y Y Y Y  
M M D D Y Y Y Y

A6. Did this subject report any relatives with Marfan syndrome at Baseline (Form A111)?  
YES..... 1  
NO ..... 2 (C1)

Prior to Study Visit 4, run the **A111 REPORT** to view information collected on the Baseline A111 Form. The A111 Report may be run from the Screening and Baseline Event in ADEPT. The order of relatives on this form must match the order in which they appear on the A111 Report and A111 Form.

If question A6 = NO (at Baseline, the subject reported on no biological relatives with known or suspected Marfan syndrome), then **SKIP TO C1**.

If question A6 = YES, then complete and update the family history questions with **NEW** information about first-degree biological relatives and/or grandparents with known or suspected Marfan syndrome that were reported previously on form A111. Indicate the relationship using the code list below. Provide answers for all of the questions (a-g) even if the response is 'UNKNOWN'.

To report age: Round to the nearest full year; Round up to the nearest year if ≥ 6 months; Round down for < 6 months (e.g., if relative is 33 years and 8 months old at the time of aortic surgery, report relative's age as 34 years).

If more than 3 relatives with Marfan syndrome were reported at Baseline, then make additional copies of page 3 or 4 of this form as appropriate and complete the information.

Code	Relationship to Subject	Code	Relationship to Subject
1	Mother	7	Maternal Grandmother
2	Father	8	Paternal Grandmother
3	Sister	9	Maternal Grandfather
4	Brother	10	Paternal Grandfather
5	Son		
6	Daughter		

**Section B: FAMILY HISTORY**

B1. Number of first-degree biological relatives and grandparents reported at BASELINE with known or suspected Marfan syndrome (MUST match information from A111 and A111 Report)

a. Relative #1 code (Code MUST match Code #1 as listed on A111 REPORT)

a1. Was this relative reported as deceased at baseline (A111)? YES..... 1  
 NO ..... 2 (next relative or C1)

b. **New** Aortic aneurysm since Baseline YES..... 1  
 NO ..... 2 (c)  
 UNKNOWN.....-8 (c)

1. Location of aneurysm AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH..... 3  
 UNKNOWN.....-8

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c. **New** Aortic dissection since Baseline YES..... 1  
 NO ..... 2 (d)  
 UNKNOWN.....-8 (d)

1. Location of dissection AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH..... 3  
 UNKNOWN.....-8

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d. **New** Aortic surgery since Baseline YES..... 1  
 NO ..... 2 (e)  
 UNKNOWN.....-8 (e)

1. Location of surgery AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH..... 3  
 UNKNOWN.....-8

2. Age at new surgery \_\_\_\_\_ Years (-8 if unknown)

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e. **New** Mitral valve surgery since Baseline YES..... 1  
 NO ..... 2  
 UNKNOWN.....-8

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f. **New** Ectopia lentis since Baseline YES..... 1  
 NO ..... 2  
 UNKNOWN.....-8

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g. Is this relative alive today? YES..... 1 (a)  
 NO ..... 2  
 UNKNOWN.....-8 (a)

1. Cause of death AORTIC DISSECTION..... 1  
 OTHER CARDIAC ..... 2  
 NON-CARDIAC..... 3  
 SUDDEN, CAUSE UNKNOWN..... 4

2. Age at death \_\_\_\_\_ Years (-8 if unknown)

**Section B: FAMILY HISTORY**

a. Relative #2 code	_____ (Code <b>MUST</b> match Code #2 as listed on A111 REPORT)
a1. Was this relative reported as deceased at baseline (A111)?	YES..... 1 <div style="text-align: right;"><b>(next relative or C1)</b></div> NO ..... 2
b. <b>New</b> Aortic aneurysm since Baseline	YES..... 1 NO ..... 2 <b>(c)</b> UNKNOWN.....-8 <b>(c)</b>
1. Location of aneurysm	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH..... 3 UNKNOWN.....-8
c. <b>New</b> Aortic dissection since Baseline	YES..... 1 NO ..... 2 <b>(d)</b> UNKNOWN.....-8 <b>(d)</b>
1. Location of dissection	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH..... 3 UNKNOWN.....-8
d. <b>New</b> Aortic surgery since Baseline	YES..... 1 NO ..... 2 <b>(e)</b> UNKNOWN.....-8 <b>(e)</b>
1. Location of surgery	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH..... 3 UNKNOWN.....-8
2. Age at new surgery	_____ Years <b>(-8 if unknown)</b>
e. <b>New</b> Mitral valve surgery since Baseline	YES..... 1 NO ..... 2 UNKNOWN.....-8
f. <b>New</b> Ectopia lentis since Baseline	YES..... 1 NO ..... 2 UNKNOWN.....-8
g. Is this relative alive today?	YES..... 1 <b>(a)</b> NO ..... 2 UNKNOWN.....-8 <b>(a)</b>
1. Cause of death	AORTIC DISSECTION..... 1 OTHER CARDIAC ..... 2 NON-CARDIAC..... 3 SUDDEN, CAUSE UNKNOWN..... 4
2. Age at death	_____ Years <b>(-8 if unknown)</b>

**Section B: FAMILY HISTORY**

a. Relative #3 code	_____ (Code <b>MUST</b> match Code #3 as listed on A111 REPORT)
a1. Was this relative reported as deceased at baseline (A111)?	YES..... 1 <span style="float: right;"><b>(next relative or C1)</b></span> NO ..... 2
b. <b>New</b> Aortic aneurysm since Baseline	YES..... 1 NO ..... 2 <b>(c)</b> UNKNOWN ..... -8 <b>(c)</b>
1. Location of aneurysm	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN ..... -8
c. <b>New</b> Aortic dissection since Baseline	YES..... 1 NO ..... 2 <b>(d)</b> UNKNOWN ..... -8 <b>(d)</b>
1. Location of dissection	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN ..... -8
d. <b>New</b> Aortic surgery since Baseline	YES..... 1 NO ..... 2 <b>(e)</b> UNKNOWN ..... -8 <b>(e)</b>
1. Location of surgery	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN ..... -8
2. Age at new surgery	_____ Years <b>(-8 if unknown)</b>
e. <b>New</b> Mitral valve surgery since Baseline	YES..... 1 NO ..... 2 UNKNOWN ..... -8
f. <b>New</b> Ectopia lentis since Baseline	YES..... 1 NO ..... 2 UNKNOWN ..... -8
g. Is this relative alive today?	YES..... 1 <b>(a)</b> NO ..... 2 UNKNOWN ..... -8 <b>(a)</b>
1. Cause of death	AORTIC DISSECTION ..... 1 OTHER CARDIAC..... 2 NON-CARDIAC ..... 3 SUDDEN, CAUSE UNKNOWN ..... 4
2. Age at death	_____ Years <b>(-8 if unknown)</b>

In Section C below, please report on any first-degree relatives who were newly identified (i.e. NOT previously reported on the Baseline A111 Form) as having, or possibly having, Marfan syndrome during this subject's 3-year trial period.

To report age: Round to the nearest full year; Round up to the nearest year if  $\geq 6$  months; Round down for  $< 6$  months (e.g., if relative is 33 years and 8 months old at the time of aortic surgery, report relative's age as 34 years).

If more than 3 NEW relatives must be included, then make additional copies of page 7 or 8 of this form as appropriate and complete the information.

**Section C: RELATIVES WITH NEW MARFAN DIAGNOSES**

C1. Does this subject have any first-degree biological relatives and grandparents who were NEWLY diagnosed with or suspected of having Marfan syndrome?  
(Report only on new diagnoses that occurred between the subject's Baseline Visit and 36-month Visit)

YES ..... 1  
NO ..... 2 (D1)

If question C1 = NO (during the subject's 3-year trial period, there were no newly diagnosed biological relatives who were not known at the time of the Baseline visit), then SKIP TO D1.

**Section C: RELATIVES WITH NEW MARFAN DIAGNOSES**

C2. In the previous 3 years, how many biological relatives were NEWLY diagnosed with or suspected of having Marfan syndrome? \_\_\_\_\_

a. **NEW** relative #1 code \_\_\_\_\_ (use Code List on page 1 of this form)

b. Aortic aneurysm YES..... 1  
 NO ..... 2 (c)  
 UNKNOWN ..... -8 (c)

1. Location of aneurysm AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH ..... 3  
 UNKNOWN ..... -8

c. Aortic dissection YES..... 1  
 NO ..... 2 (d)  
 UNKNOWN ..... -8 (d)

1. Location of dissection AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH ..... 3  
 UNKNOWN ..... -8

d. Aortic surgery YES..... 1  
 NO ..... 2 (e)  
 UNKNOWN ..... -8 (e)

1. Location of surgery AORTIC ROOT/ASCENDING AORTA .. 1  
 DESCENDING AORTA/ABDOMINAL ... 2  
 BOTH ..... 3  
 UNKNOWN ..... -8

2. Age at first surgery \_\_\_\_\_ Years (-8 if unknown)

e. Mitral valve surgery YES..... 1  
 NO ..... 2  
 UNKNOWN ..... -8

f. Ectopia lentis YES..... 1  
 NO ..... 2  
 UNKNOWN ..... -8

g. Is this relative alive today? YES..... 1 (a)  
 NO ..... 2  
 UNKNOWN ..... -8 (a)

1. Cause of death AORTIC DISSECTION ..... 1  
 OTHER CARDIAC..... 2  
 NON-CARDIAC ..... 3  
 SUDDEN, CAUSE UNKNOWN ..... 4

2. Age at death \_\_\_\_\_ Years (-8 if unknown)

**Section C: RELATIVES WITH NEW MARFAN DIAGNOSES**

a.	NEW relative #2 code	_____ (use Code List on page 1 of this form)
b.	Aortic aneurysm	YES..... 1 NO ..... 2 (c) UNKNOWN .....-8 (c)
	1. Location of aneurysm	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
c.	Aortic dissection	YES..... 1 NO ..... 2 (d) UNKNOWN .....-8 (d)
	1. Location of dissection	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
d.	Aortic surgery	YES..... 1 NO ..... 2 (e) UNKNOWN .....-8 (e)
	1. Location of surgery	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
	2. Age at first surgery	_____ Years (-8 if unknown)
e.	Mitral valve surgery	YES..... 1 NO ..... 2 UNKNOWN .....-8
f.	Ectopia lentis	YES..... 1 NO ..... 2 UNKNOWN .....-8
g.	Is this relative alive today?	YES..... 1 (a) NO ..... 2 UNKNOWN .....-8 (a)
	1. Cause of death	AORTIC DISSECTION ..... 1 OTHER CARDIAC..... 2 NON-CARDIAC ..... 3 SUDDEN, CAUSE UNKNOWN ..... 4
	2. Age at death	_____ Years (-8 if unknown)

**Section C: RELATIVES WITH NEW MARFAN DIAGNOSES**

a.	NEW relative #3 code	_____ (use Code List on page 1 of this form)
b.	Aortic aneurysm	YES..... 1 NO ..... 2 <b>(c)</b> UNKNOWN .....-8 <b>(c)</b>
	1. Location of aneurysm	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
<hr/>		
c.	Aortic dissection	YES..... 1 NO ..... 2 <b>(d)</b> UNKNOWN .....-8 <b>(d)</b>
	1. Location of dissection	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
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d.	Aortic surgery	YES..... 1 NO ..... 2 <b>(e)</b> UNKNOWN .....-8 <b>(e)</b>
	1. Location of surgery	AORTIC ROOT/ASCENDING AORTA .. 1 DESCENDING AORTA/ABDOMINAL ... 2 BOTH ..... 3 UNKNOWN .....-8
	2. Age at first surgery	_____ Years <b>(-8 if unknown)</b>
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e.	Mitral valve surgery	YES..... 1 NO ..... 2 UNKNOWN .....-8
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f.	Ectopia lentis	YES..... 1 NO ..... 2 UNKNOWN .....-8
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g.	Is this relative alive today?	YES..... 1 <b>(C1)</b> NO ..... 2 UNKNOWN .....-8 <b>(C1)</b>
	1. Cause of death	AORTIC DISSECTION ..... 1 OTHER CARDIAC..... 2 NON-CARDIAC ..... 3 SUDDEN, CAUSE UNKNOWN ..... 4
	2. Age at death	_____ Years <b>(-8 if unknown)</b>



**Section D: RELATIVES SCREENED**

D1. Does this subject have parents, children, and/or siblings (first-degree relatives) who were screened for the Marfan Trial at this site?  
 YES ..... 1  
 NO ..... 2 (Z1)  
 (Report on ALL relatives who were given a Study ID Number, regardless of their final eligibility or enrollment status.)

D2. How many first-degree relatives (parents, children, and or siblings) were screened for the Marfan Trial? \_\_\_\_\_

- |                | 1. Study ID Number | 2. Relationship to Subject |
|----------------|--------------------|----------------------------|
| a. Relative #1 | _____              | _____                      |
| b. Relative #2 | _____              | _____                      |
| c. Relative #3 | _____              | _____                      |
| d. Relative #4 | _____              | _____                      |
| e. Relative #5 | _____              | _____                      |

Code	Relationship to Subject
1	Mother
2	Father
3	Sister
4	Brother
5	Son
6	Daughter

**Section Z: TIME TO COMPLETE FORM AND SIGNATURE**

Z1. How long did it take to complete this form? \_\_\_\_\_ minutes  
 Signature of PI: \_\_\_\_\_ Date: \_\_\_\_\_

**END OF FORM**