

10a. Specify the area(s) of all new abnormalities and enter date of earliest abnormality since enrollment.

Area of Aorta	Newly Tortuous	Newly Dilated	Newly Dissected	Date of Earliest Abnormality Since Enrollment
a. Ascending				
b. Arch				
c. Descending				
d. Abdominal				
e. Sinus of Valsalva				
f. Other _____				

11. Since the date of enrollment, indicate whether there has been a new diagnosis or new information obtained for any of the conditions below. If YES, enter the Month and Year of the diagnosis. Otherwise check No/NA.

A. Cardiovascular Conditions	Yes	MM/YY	No/NA
1. Mitral valve prolapse			
2. Mitral valve regurgitation > trace			
3. Calcified mitral annulus			
4. Aortic valve regurgitation			
5. Aortic valve stenosis			
6. Aortic coarctation, current/repared			
7. Bicuspid Aortic Valve (BAV)			
8. Tricuspid regurgitation > trace			
9. Pulmonary stenosis/right ventricular outflow obstruction			
10. Pulmonary regurgitation > trace			
11. Endocarditis			
12. Atrial arrhythmia			
13. Cardiac arrhythmia (not atrial)			
14. Coronary dissection			
15. Coronary ectasia			
16. Carotid dissection			
17. Symptomatic coronary artery disease			
18. Congestive heart failure/left ventricular dysfunction			
19. Left ventricular dilation			
20. Left ventricular hypertrophy			
21. Right ventricular hypertrophy			
22. Right ventricular dysfunction/failure			
23. Dilatation of pulmonary artery			
24. Other pulmonary artery abnormality			
25. Hypertension			
26. New diagnosis of a congenital heart defect, specify:			

B. Musculoskeletal	Yes	MM/YY	No/NA
1. Pectus carinatum			
2. Pectus excavatum			
3. Shield chest, broad with widely spaced nipples			
4. Recurrent dislocations/subluxations			
5. Joint hypermobility in >1 joint			
6. Wrist sign			
7. Scoliosis that is clinically evident			
8. Abnormal kyphoscoliosis/lordosis			
9. Pes planus			
11. Osteopenia/osteoporosis			
12. Congenital bilateral dislocated hips			
15. Joint pain, arthralgia			
16. Thumb sign			
21. Clubfoot			
22. Reduced elbow extension (<170°)			
23. Long fingers, arachnodactyly			
24. Genu recurvatum			
25. Musculoskeletal surgery, Specify _____			
C. Craniofacial	Yes	MM/YY	No/NA
1. High palate/dental crowding			
2. Dolicocephaly			
3. Malar hypoplasia			
4. Enophthalmos			
5. Retrognathia			
6. Down-slanting palpebral fissures			
7. Cleft palate			
8. Bifid or broad-based uvula			
9. Craniosynostosis			

D. Vascular & Circulatory Conditions	Yes	MM/YY	No/NA
1. Early-onset varicose veins, <40 years of age			
2. Arteriovenous malformation			
3. Stroke			
4. Arterial rupture			
5. Lymphedema			
E. Ocular	Yes	MM/YY	No/NA
1. Lens dislocation (ectopia lentis)			
2. Glaucoma			
4. Hypertelorism			
5. Retinal detachment			
6. Amblyopia			
7. Strabismus			
9. Myopia >4 diopters			
10. Cataracts, <50 years of age			
11. Iris Flocculi			
12. Blue/grey sclera			
13. Iridodonesis			
14. Eye surgery, specify:			
F. Gastrointestinal	Yes	MM/YY	No/NA
1. Ulcerative colitis			
2. Ruptured bowel			
3. Crohn's disease			
4. Irritable bowel syndrome			
5. Diverticulosis/diverticulitis			
6. G-tube is present			

G. Skin/integument	Yes	MM/YY	No/NA
1. Skin hyperextensibility			
2. Wide atrophic scars			
3. Poor wound healing			
4. Easy bruising			
5. Soft, velvety or doughy skin			
6. Tissue fragility			
8. Striae atrophicae			
9. Hernia, specify type			
10. Recurrent hernia			
11. Thin translucent skin			
12. Webbed neck			

H. Reproductive (WOMEN ONLY)	Yes	MM/YY	No/NA
1. Uterine rupture			
2. Ovarian failure			
3. Endometriosis			
4. Uterine fibroids			
5. Premature rupture of membranes			

I. Familial History	Yes	No/NA
1. Immediate family history (parent, child, sibling) of TAA or other major vessel dilatation or dissection		
2. Secondary family hx (cousin, aunt, uncle, grandparent) of TAA or other major vessel dilatation or dissection		

12. For each drug or category listed, check one box in each row which best describes the current status of the drug treatment.

Drug Treatment	Never prescribed	Stopped use since enrollment	Currently Using	Blinded Clinical Trial	Unknown
a. Beta-blocker					
b. Losartan					
c. Other ARB					
d. ACE-inhibitor					
e. Statins					
f. Verapamil or Diltiazem					
g. Other Ca+ channel blocker					
h. Other BP lowering drug					

13. Since enrollment, has the subject undergone any surgical procedures listed on the next page and related to treatment of their cardiovascular genetic disorder? Yes No → Go To Q14

13a. Indicate the procedures received since enrollment by checking the "YES" column and entering the corresponding Month and Year, **if readily available** (OTHERWISE LEAVE BLANK). Complete a Surgical Intervention Form for each procedure *except* for Pacemaker/ICD implantation and any "other" types of surgeries.

Surgical Intervention	Yes	MM/YY
a. Coronary artery bypass grafting or PTCA		
b. Aortic valve repair		
c. Isolated aortic valve replacement (non-root)		
d. Aortic root replacement (valve-replacing)		
e. Valve-sparing aortic root replacement		
f. Ascending aortic replacement		
g. Aortic arch replacement		
h. Mitral valve repair or replacement		
i. Coarctation repair/intervention		
j. Pacemaker/ICD implantation		

Surgical Intervention	Yes	MM/YY
k. Descending thoracic aortic replacement		
l. Thoracoabdominal aortic replacement		
m. Open arterial bypass		
o. Percutaneous/endovascular intervention, non-coronary		
p. Aorta to pulmonary shunt		
q. TOF repair		
r. Pulmonary valve replacement		
s. Tricuspid valve repair/replacement		
t. Other, specify:		
u. Other, specify:		

14. Is there an actual imaging study (not an imaging report) available for review by a Core Imaging Lab for this follow-up evaluation? Yes No

15. Sources for information provided on this form. (CHECK ALL THAT APPLY.)

- Medical record of exam by a geneticist done at your institution
- Medical record of other exams and tests done at your institution
- Medical record of exams and tests done elsewhere
- Medical record of patient interviews and clinical history
- Direct patient interview specifically for this study

16. Did the subject have a pregnancy since enrollment? N/A, child or male è **END**
 No, female è **END**
 Yes è Complete table below

	Pregnancy 1		Pregnancy 2	
16a. What was the outcome of this pregnancy? (IF PREGNANT WITH MULTIPLES, CHECK ALL THAT APPLY)	Live birth	Abortion	Live birth	Abortion
	Still birth	Tubal, ectopic	Still birth	Tubal, ectopic
	Miscarriage	Currently pregnant	Miscarriage	Currently pregnant
16b. <u>After this pregnancy</u> was over, was the subject told the baby had a birth defect, chromosome abnormality, or other serious medical problem?	No Yes è What condition? _____		No Yes è What condition? _____	
16c. <u>While the subject was pregnant</u> , which of these conditions did the subject have?	High blood pressure, toxemia Maternal diabetes Premature rupture of membranes Change in aortic dimension Aortic dissection or rupture None		High blood pressure, toxemia Maternal diabetes Premature rupture of membranes Change in aortic dimension Aortic dissection or rupture None	

Comments: _____