

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	<input type="checkbox"/> FF10AA1	<input type="checkbox"/> FF10AA2	<input type="checkbox"/> FF10AA3	FF10AAM / FF10AAY
b. Arch	<input type="checkbox"/> FF10AB1	<input type="checkbox"/> FF10AB2	<input type="checkbox"/> FF10AB3	FF10ABM / FF10ABY
c. Descending	<input type="checkbox"/> FF10AC1	<input type="checkbox"/> FF10AC2	<input type="checkbox"/> FF10AC3	FF10ACM / FF10ACY
d. Abdominal	<input type="checkbox"/> FF10AD1	<input type="checkbox"/> FF10AD2	<input type="checkbox"/> FF10AD3	FF10ADM / FF10ADY
e. Sinus of Valsalva	<input type="checkbox"/> FF10AE1	<input type="checkbox"/> FF10AE2	<input type="checkbox"/> FF10AE3	FF10AEM / FF10AEY
f. Other ___ FF10AFSP	<input type="checkbox"/> FF10AF1	<input type="checkbox"/> FF10AF2	<input type="checkbox"/> FF10AF3	FF10AFM / FF10AFY

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 1	MM/YY	No/ NA ⁰
1. Mitral valve prolapse FF11A01	<input type="checkbox"/>	FF11A01M FF11A01Y	<input type="checkbox"/>
2. Mitral valve regurgitation > trace FF11A02	<input type="checkbox"/>	FF11A02M FF11A02Y	<input type="checkbox"/>
3. Calcified mitral annulus FF11A03	<input type="checkbox"/>	FF11A03M FF11A03Y	<input type="checkbox"/>
4. Aortic valve regurgitation FF11A04	<input type="checkbox"/>	FF11A04M FF11A04Y	<input type="checkbox"/>
5. Aortic valve stenosis FF11A05	<input type="checkbox"/>	FF11A05M FF11A05Y	<input type="checkbox"/>
6. Aortic coarctation, current/repai red FF11A06	<input type="checkbox"/>	FF11A06M FF11A06Y	<input type="checkbox"/>
7. Bicuspid Aortic Valve (BAV) FF11A07	<input type="checkbox"/>	FF11A07M FF11A07Y	<input type="checkbox"/>
8. Tricuspid regurgitation > trace FF11A08	<input type="checkbox"/>	FF11A08M FF11A08Y	<input type="checkbox"/>
9. Pulmonary stenosis/right ventricular outflow obstruction FF11A09	<input type="checkbox"/>	FF11A09M FF11A09Y	<input type="checkbox"/>
10. Pulmonary regurgitation > trace FF11A10	<input type="checkbox"/>	FF11A10M FF11A10Y	<input type="checkbox"/>
11. Endocarditis FF11A11	<input type="checkbox"/>	FF11A11M FF11A11Y	<input type="checkbox"/>
12. Atrial arrhythmia FF11A12	<input type="checkbox"/>	FF11A12M FF11A12Y	<input type="checkbox"/>
13. Cardiac arrhythmia (not atrial) FF11A13	<input type="checkbox"/>	FF11A13M FF11A13Y	<input type="checkbox"/>
14. Coronary dissection FF11A14	<input type="checkbox"/>	FF11A14M FF11A14Y	<input type="checkbox"/>
15. Coronary ectasia FF11A15	<input type="checkbox"/>	FF11A15M FF11A15Y	<input type="checkbox"/>
16. Carotid dissection FF11A16	<input type="checkbox"/>	FF11A16M FF11A16Y	<input type="checkbox"/>
17. Symptomatic coronary artery disease FF11A17	<input type="checkbox"/>	FF11A17M FF11A17Y	<input type="checkbox"/>
18. Congestive heart failure/left ventricular dysfunction FF11A18	<input type="checkbox"/>	FF11A18M FF11A18Y	<input type="checkbox"/>
19. Left ventricular dilation FF11A19	<input type="checkbox"/>	FF11A19M FF11A19Y	<input type="checkbox"/>
20. Left ventricular hypertrophy FF11A20	<input type="checkbox"/>	FF11A20M FF11A20Y	<input type="checkbox"/>

22. Right ventricular dysfunction/failure FF11A22	<input type="checkbox"/>	FF11A22M FF11A22Y	<input type="checkbox"/>
23. Dilatation of pulmonary artery FF11A23	<input type="checkbox"/>	FF11A23M FF11A23Y	<input type="checkbox"/>
24. Other pulmonary artery abnormality FF11A24	<input type="checkbox"/>	FF11A24M FF11A24Y	<input type="checkbox"/>
25. Hypertension FF11A25	<input type="checkbox"/>	FF11A25M FF11A25Y	<input type="checkbox"/>
26. New diagnosis of a congenital heart defect, specify: ___ FF11A26S FF11A26	<input type="checkbox"/>	FF11A26M FF11A26Y	<input type="checkbox"/>
B. Musculoskeletal	Yes 1	MM/YY	No/NA 0
1. Pectus carinatum FF11B01	<input type="checkbox"/>	FF11B01M FF11B01Y	<input type="checkbox"/>
2. Pectus excavatum FF11B02	<input type="checkbox"/>	FF11B02M FF11B02Y	<input type="checkbox"/>
3. Shield chest, broad with widely spaced nipples FF11B03	<input type="checkbox"/>	FF11B03M FF11B03Y	<input type="checkbox"/>
4. Recurrent dislocations/subluxations FF11B04	<input type="checkbox"/>	FF11B04M FF11B04Y	<input type="checkbox"/>
5. Joint hypermobility in >1 joint FF11B05	<input type="checkbox"/>	FF11B05M FF11B05Y	<input type="checkbox"/>
6. Wrist sign FF11B06	<input type="checkbox"/>	FF11B06M FF11B06Y	<input type="checkbox"/>
7. Scoliosis that is clinically evident FF11B07	<input type="checkbox"/>	FF11B07M FF11B07Y	<input type="checkbox"/>
8. Abnormal kyphoscoliosis/lordosis FF11B08	<input type="checkbox"/>	FF11B08M FF11B08Y	<input type="checkbox"/>
9. Pes planus FF11B09	<input type="checkbox"/>	FF11B09M FF11B09Y	<input type="checkbox"/>
11. Osteopenia/osteoporosis FF11B10	<input type="checkbox"/>	FF11B11M FF11B11Y	<input type="checkbox"/>
12. Congenital bilateral dislocated hips FF11B12	<input type="checkbox"/>	FF11B12M FF11B12Y	<input type="checkbox"/>
15. Joint pain, arthralgia FF11B13	<input type="checkbox"/>	FF11B15M FF11B15Y	<input type="checkbox"/>
16. Thumb sign FF11B14	<input type="checkbox"/>	FF11B16M FF11B16Y	<input type="checkbox"/>
21. Clubfoot FF11B21	<input type="checkbox"/>	FF11B21M FF11B21Y	<input type="checkbox"/>
22. Reduced elbow extension (<170°) FF11B22	<input type="checkbox"/>	FF11B22M FF11B22Y	<input type="checkbox"/>
23. Long fingers, arachnodactyly FF11B23	<input type="checkbox"/>	FF11B23M FF11B23Y	<input type="checkbox"/>
24. Genu recurvatum FF11B24	<input type="checkbox"/>	FF11B24M FF11B24Y	<input type="checkbox"/>
25. Musculoskeletal surgery, Specify _ FF11B25S FF11B25	<input type="checkbox"/>	FF11B25M FF11B25Y	<input type="checkbox"/>

C. Craniofacial	Yes 1	MM/YY	No/NA 0
1. High palate/dental crowding FF11C01	<input type="checkbox"/>	FF11C01M FF11C01Y	<input type="checkbox"/>
2. Dolicocephaly FF11C02	<input type="checkbox"/>	FF11C02M FF11C02Y	<input type="checkbox"/>
3. Malar hypoplasia FF11C03	<input type="checkbox"/>	FF11C03M FF11C03Y	<input type="checkbox"/>
4. Enophthalmos FF11C04	<input type="checkbox"/>	FF11C04M FF11C04Y	<input type="checkbox"/>
5. Retrognathia FF11C05	<input type="checkbox"/>	FF11C05M FF11C05Y	<input type="checkbox"/>
6. Down-slanting palpebral fissures FF11C06	<input type="checkbox"/>	FF11C06M FF11C06Y	<input type="checkbox"/>
7. Cleft palate FF11C07	<input type="checkbox"/>	FF11C07M FF11C07Y	<input type="checkbox"/>
8. Bifid or broad-based uvula FF11C08	<input type="checkbox"/>	FF11C08M FF11C08Y	<input type="checkbox"/>
9. Craniosynostosis FF11C09	<input type="checkbox"/>	FF11C09M FF11C09Y	<input type="checkbox"/>

D. Vascular & Circulatory Conditions		Yes 1	MM/YY	No/NA 0
1. Early-onset varicose veins, <40 years of age	FF11D01	<input type="checkbox"/>	FF11D01M FF11D01Y	<input type="checkbox"/>
2. Arteriovenous malformation	FF11D02	<input type="checkbox"/>	FF11D02M FF11D02Y	<input type="checkbox"/>
3. Stroke	FF11D03	<input type="checkbox"/>	FF11D03M FF11D03Y	<input type="checkbox"/>
4. Arterial rupture	FF11D04	<input type="checkbox"/>	FF11D04M FF11D04Y	<input type="checkbox"/>
5. Lymphedema	FF11D05	<input type="checkbox"/>	FF11D05M FF11D05Y	<input type="checkbox"/>
E. Ocular		Yes	MM/YY	No/NA
1. Lens dislocation (ectopia lentis)	FF11E01	<input type="checkbox"/>	FF11E01M FF11E01Y	<input type="checkbox"/>
2. Glaucoma	FF11E02	<input type="checkbox"/>	FF11E02M FF11E02Y	<input type="checkbox"/>
4. Hypertelorism	FF11E04	<input type="checkbox"/>	FF11E04M FF11E04Y	<input type="checkbox"/>
5. Retinal detachment	FF11E05	<input type="checkbox"/>	FF11E05M FF11E05Y	<input type="checkbox"/>
6. Amblyopia	FF11E06	<input type="checkbox"/>	FF11E06M FF11E06Y	<input type="checkbox"/>
7. Strabismus	FF11E07	<input type="checkbox"/>	FF11E07M FF11E07Y	<input type="checkbox"/>
9. Myopia >4 diopters	FF11E09	<input type="checkbox"/>	FF11E09M FF11E09Y	<input type="checkbox"/>
10. Cataracts, <50 years of age	FF11E10	<input type="checkbox"/>	FF11E10M FF11E10Y	<input type="checkbox"/>
11. Iris Flocculi	FF11E11	<input type="checkbox"/>	FF11E11M FF11E11Y	<input type="checkbox"/>
12. Blue/grey sclera	FF11E12	<input type="checkbox"/>	FF11E12M FF11E12Y	<input type="checkbox"/>
13. Iridodonesis	FF11E13	<input type="checkbox"/>	FF11E13M FF11E13Y	<input type="checkbox"/>
14. Eye surgery, specify:	FF11E14S FF11E14	<input type="checkbox"/>	FF11E14M FF11E14Y	<input type="checkbox"/>
F. Gastrointestinal		Yes	MM/YY	No/NA
1. Ulcerative colitis	FF11F01	<input type="checkbox"/>	FF11F01M FF11F01Y	<input type="checkbox"/>
2. Ruptured bowel	FF11F02	<input type="checkbox"/>	FF11F02M FF11F02Y	<input type="checkbox"/>
3. Crohn's disease	FF11F03	<input type="checkbox"/>	FF11F03M FF11F03Y	<input type="checkbox"/>
4. Irritable bowel syndrome	FF11F04	<input type="checkbox"/>	FF11F04M FF11F04Y	<input type="checkbox"/>
5. Diverticulosis/diverticulitis	FF11F04	<input type="checkbox"/>	FF11F05M FF11F05Y	<input type="checkbox"/>
6. G-tube is present	FF11F06	<input type="checkbox"/>	FF11F06M FF11F06Y	<input type="checkbox"/>

G. Skin/integument	Yes	MM/YY	No/NA
1. Skin hyperextensibility	FF11G01	<input type="checkbox"/>	FF11G01M FF11G01Y
2. Wide atrophic scars	FF11G02	<input type="checkbox"/>	FF11G02M FF11G02Y
3. Poor wound healing	FF11G03	<input type="checkbox"/>	FF11G03M FF11G03Y
4. Easy bruising	FF11G04	<input type="checkbox"/>	FF11G04M FF11G04Y
5. Soft, velvety or doughy skin	FF11G05	<input type="checkbox"/>	FF11G05M FF11G05Y
6. Tissue fragility	FF11G06	<input type="checkbox"/>	FF11G06M FF11G06Y
8. Striae atrophicae	FF11G08	<input type="checkbox"/>	FF11G08M FF11G08Y
9. Hernia, specify type	FF11G09	<input type="checkbox"/>	FF11G09M FF11G09Y
10. Recurrent hernia	FF11G10	<input type="checkbox"/>	FF11G10M FF11G10Y
11. Thin translucent skin	FF11G11	<input type="checkbox"/>	FF11G11M FF11G11Y
12. Webbed neck	FF11G12	<input type="checkbox"/>	FF11G12M FF11G12Y

H. Reproductive (WOMEN ONLY)	Yes	MM/YY	No/NA
1. Uterine rupture	FF11H01	<input type="checkbox"/>	FF11H01M FF11H02Y
2. Ovarian failure	FF11H02	<input type="checkbox"/>	FF11H02M FF11H02Y
3. Endometriosis	FF11H03	<input type="checkbox"/>	FF11H03M FF11H03Y
4. Uterine fibroids	FF11H04	<input type="checkbox"/>	FF11H04M FF11H04Y
5. Premature rupture of membranes	FF11H05	<input type="checkbox"/>	FF11H05M FF11H05Y

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

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 FF12A	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
b. Losartan FF12B	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
c. Other ARB FF12C	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
d. ACE-inhibitor FF12D	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
e. Statins FF12E	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
f. Verapamil or Diltiazem FF12F	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
g. Other Ca+ channel blocker FF12G	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
h. Other BP lowering drug FF12H	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>

13. Since enrollment, has the subject undergone any surgical procedures listed on the next page and related to treatment of their cardiovascular genetic disorder? 1 Yes
0 No → Go To Q14

FF13

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 FF13AA	<input type="checkbox"/>	FF13AAM FF13AAY
b. Aortic valve repair FF13AB	<input type="checkbox"/>	FF13ABM FF13ABY
c. Isolated aortic valve replacement (non-root) FF13AC	<input type="checkbox"/>	FF13ACM FF13ACY
d. Aortic root replacement (valve-replacing) FF13AD	<input type="checkbox"/>	FF13ADM FF13ADY
e. Valve-sparing aortic root replacement FF13AE	<input type="checkbox"/>	FF13AEM FF13AEY
f. Ascending aortic replacement FF13AF	<input type="checkbox"/>	FF13AFM FF13AFY
g. Aortic arch replacement FF13AG	<input type="checkbox"/>	FF13AGM FF13AGY
h. Mitral valve repair or replacement FF13AH	<input type="checkbox"/>	FF13AHM FF13AHY
i. Coarctation repair/intervention FF13AI	<input type="checkbox"/>	FF13AIM FF13AIY
j. Pacemaker/ICD implantation FF13AJ	<input type="checkbox"/>	FF13AJM FF13AJY

Surgical Intervention	Yes	MM/YY
k. Descending thoracic aortic replacement FF13AK	<input type="checkbox"/>	FF13AKM FF13AKY
l. Thoracoabdominal aortic replacement FF13AL	<input type="checkbox"/>	FF13ALM FF13ALY
m. Open arterial bypass FF13AM	<input type="checkbox"/>	FF13AMM FF13AMY
o. Percutaneous/endovascular intervention, non-coronary FF13AO	<input type="checkbox"/>	FF13AOM FF13AOY
p. Aorta to pulmonary shunt FF13AP	<input type="checkbox"/>	FF13APM FF13APY
q. TOF repair FF13AQ	<input type="checkbox"/>	FF13AQM FF13AQY
r. Pulmonary valve replacement FF13AR	<input type="checkbox"/>	FF13ARM FF13ARY
s. Tricuspid valve repair/replacement FF13AS	<input type="checkbox"/>	FF13ASM FF13ASY
t. Other, specify: FF13ATS	<input type="checkbox"/>	FF13ATM FF13ATY
u. Other, specify: FF13AUS	<input type="checkbox"/>	FF13AUM FF13AUY

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 FF14

15. Sources for information provided on this form. **(CHECK ALL THAT APPLY.)** FF15 (checkbox list)

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

16. Did the subject have a pregnancy since enrollment? FF16

- 2 N/A, child or male → **END**
- 0 No, female → **END**
- 1 Yes → Complete table below

	Pregnancy 1	Pregnancy 2
16a. What was the outcome of this pregnancy? (IF PREGNANT WITH MULTIPLES, CHECK ALL THAT APPLY)	1 <input type="checkbox"/> Live birth 4 <input type="checkbox"/> Abortion FF16A_1 2 <input type="checkbox"/> Still birth 5 <input type="checkbox"/> Tubal, ectopic 3 <input type="checkbox"/> Miscarriage 6 <input type="checkbox"/> Currently pregnant	1 <input type="checkbox"/> Live birth 4 <input type="checkbox"/> Abortion FF16A_2 2 <input type="checkbox"/> Still birth 5 <input type="checkbox"/> Tubal, ectopic 3 <input type="checkbox"/> Miscarriage 6 <input type="checkbox"/> Currently pregnant
16b. After this pregnancy was over, was the subject told the baby had a birth defect, chromosome abnormality, or other serious medical problem?	0 <input type="checkbox"/> <input type="checkbox"/> No FF16B_1 1 <input type="checkbox"/> <input type="checkbox"/> Yes → What condition? _____ FF16B_1S	0 <input type="checkbox"/> <input type="checkbox"/> No FF16B_2 1 <input type="checkbox"/> <input type="checkbox"/> Yes → What condition? _____ FF16B_2S
16c. While the subject was pregnant, which of these conditions did the subject have?	1 <input type="checkbox"/> High blood pressure, toxemia 2 <input type="checkbox"/> Maternal diabetes FF16C_1 3 <input type="checkbox"/> Premature rupture of membranes 4 <input type="checkbox"/> Change in aortic dimension 5 <input type="checkbox"/> Aortic dissection or rupture 6 <input type="checkbox"/> None	1 <input type="checkbox"/> High blood pressure, toxemia 2 <input type="checkbox"/> Maternal diabetes FF16C_2 3 <input type="checkbox"/> Premature rupture of membranes 4 <input type="checkbox"/> Change in aortic dimension 5 <input type="checkbox"/> Aortic dissection or rupture 6 <input type="checkbox"/> None

Comments:

_____ **FFCOMM**

Initials for Investigator Approval **FFINIT**



Gentac

FORM 1
ENROLLMENT CLINICAL
EVALUATION

Version 1.0, 10/01/2007

DIAGNOSIS

1. Patient is being enrolled in the GenTAC Registry because of the following condition(s) (Check all that apply):

a.	1	Marfan syndrome	F101A
b.	1	Turner syndrome	F101B
c.	1	Ehlers-Danlos syndrome, vascular type	F101C
d.	1	Ehlers-Danlos syndrome, other type and with aortic enlargement ¹	F101D
e.	1	Loeys-Dietz syndrome	F101E
f.	1	FBN1, TGFBR1 or TGFBR2 genetic mutation	F101F
g.	1	Bicuspid aortic valve with aortic enlargement ¹ and without known family history	F101G
h.	1	Bicuspid aortic valve with family history of BAV	F101H
i.	1	Shprintzen-Goldberg syndrome	F101I
j.	1	Familial Thoracic Aortic Aneurysm and Dissections with aortic enlargement ¹	F101J
k.	1	Other aneurysms/dissections of the thoracic aorta not due to trauma and in person < 40 years of age.	F101K
l.	1	Other congenital heart disease with aortic enlargement ¹ and family history (i.e., Tetralogy of Fallot)	F101L
m.	1	1st degree family member of proband already enrolled in the GenTAC registry	F101M

¹2 SD above normal based on nomographs OR prior dissection OR significant enlargement compared to adjacent area

2. Approximate age at earliest diagnosed above condition (physician confirmed) _____ years **F102**

3. Date of enrollment into Registry |_|_|-|_|_|-|_|_|_|_|
 Month Day Year
 F103MM F103DD F103YY

4. Has the patient ever been diagnosed with a thoracic aortic aneurysm, dissection/rupture, or marked tortuosity? **F104**

- 1 Yes
- 0 No → **GO TO Q6**
- 2 Unknown → **GO TO Q6**

5. Specify the current status of each aortic area (check all that apply in each row) and enter date of earliest abnormal:

Area of Aorta	Always Normal	Ever Tortuous	Ever Dilated	Ever Dissected	Unknown	Date of Earliest Abnormality
a. Ascending	1 F105A1	1 F105A2	1 F105A3	1 F105A4	1 F105A5	F105AMM F105ADD F105AYY
b. Arch	1 F105B1	1 F105B2	1 F105B3	1 F105B4	1 F105B5	F105BMM F105BDD F105BYY
c. Descending	1 F105C1	1 F105C2	1 F105C3	1 F105C4	1 F105C5	F105CMM F105CDD F105CYY
d. Abdominal	1 F105D1	1 F105D2	1 F105D3	1 F105D4	1 F105D5	F105DMM F105DDD F105DYY
e. Sinus of Valsalva	1 F105E1	1 F105E2	1 F105E3	1 F105E4	1 F105E5	F105EMM F105EDD F105EYY

DEMOGRAPHIC DATA

6. Date of birth |_|_|-|_|_|-|_|_|_|_|
 Month Day Year
 F106MM F106DD F106YY

7. Gender 1 Male
 F107 2 Female

8. Race (check all 1 White F1081
that apply) 1 Black or African American F1082
 1 Asian F1083
 1 American Indian or Alaska Native F1084
 1 Native Hawaiian or Other Pacific Islander F1085
 1 Unknown F1086

9. Hispanic/Latino origin? 1 Yes
 F109 0 No → **GO TO Q10**
 2 Don't Know → **GO TO Q10**

9a. What is the country of origin of the patient's family (e.g., Cuba, Mexico)? _____ F109A

BASIC MEASUREMENTS (at time of enrollment)

10. Height F110 |_|_|_|_| cm

11. Weight F111 |_|_|_|_|. |_| kg

12. Arm Span F112 |_|_|_|_| cm

13. Lower Segment F113 |_|_|_|_| cm

14. Heart Rate F114 |_|_|_|_| Beats/Minute

15. Blood Pressure F115S |_|_|_|_| systolic / |_|_|_|_| diastolic F115D

16. Karnofsky Performance 100 Normal, no complaints, no evidence of disease (100)
 F116 (Check one) 90 Able to carry on normal activity, minor symptoms of disease (90)
 80 Normal activity with effort, some symptoms of disease (80)
 70 Able to care for self, unable to do normal activity or work (70)
 60 Requires occasional assistance but is able to care for needs (60)
 50 Requires considerable assistance and frequent medical care (50)
 40 Disabled, requires special care and assistance (40)
 30 Severely disabled, hospitalization is indicated (30)
 20 Very sick, hospitalization and active treatment are necessary (20)
 10 Fatal processes progressing rapidly (10)

DIAGNOSTIC CRITERIA AND ORGAN SYSTEM REVIEW

17. Indicate the historical status of each condition with a check in the appropriate column (Yes, No, Don't Know)

A. Musculoskeletal			Yes	No	DK				Yes	No	DK
1.	Pectus carinatum	F117A01	1	0	2	14.	Tendon/muscle rupture	F117A14	1	0	2
2.	Pectus excavatum	F117A02	1	0	2	15.	Joint pain, arthralgia	F117A15	1	0	2
3.	Shield chest, broad with widely spaced nipples	F117A03	1	0	2	16.	Thumb sign (bony part of thumb protrudes from within clenched fist)	F117A16	1	0	2
4.	Recurrent dislocations/subluxations	F117A04	1	0	2	17.	Hypoplastic or hyperconvex nails	F117A17	1	0	2
5.	Joint hypermobility in >1 joint	F117A05	1	0	2	18.	Low back pain	F117A18	1	0	2
6.	Wrist sign (at least one phalanx of little finger overlaps with thumb)	F117A06	1	0	2	19.	Cubitus valgus (increased carrying angle of elbow)	F117A19	1	0	2
7.	Scoliosis that is clinically evident	F117A07	1	0	2	20.	Short fourth metacarpal or metatarsal	F117A20	1	0	2
8.	Abnormal kyphoscoliosis/lordosis	F117A08	1	0	2	21.	Clubfoot	F117A21	1	0	2
9.	Pes planus	F117A09	1	0	2	22.	Reduced elbow extension (<170°)	F117A22	1	0	2
10.	Protrusio acetabulae	F117A10	1	0	2	23.	Long fingers, arachnodactyly	F117A23	1	0	2
11.	Osteopenia/osteoporosis	F117A11	1	0	2	24.	Other, specify _____	F117A24 F117A24S	1	0	2
12.	Congenital bilateral dislocated hips	F117A12	1	0	2	25.	Other, specify _____	F117A25 F117A25S	1	0	2
13.	Hypotonia	F117A13	1	0	2	26.	Other, specify _____	F117A26 F117A26S	1	0	2

B. Pulmonary			Yes	No	DK				Yes	No	DK
1.	Pneumothorax	F117B01	1	0	2	3.	Pulmonary hypertension	F117B02	1	0	2
2.	Asthma	F117B02	1	0	2	4.	Other, specify _____	F117B04 F117B04S	1	0	2

C. Cardiovascular			Yes	No	DK				Yes	No	DK
1.	Patent ductus arteriosus	F117C01	1	0	2	18.	Endocarditis	F117C18	1	0	2
2.	Atrial septal defect (ASD)/PFO	F117C02	1	0	2	19.	Atrial arrhythmia	F117C19	1	0	2
3.	Ventricular septal defect (VSD)	F117C03	1	0	2	20.	Cardiac arrhythmia (not atrial)	F117C20	1	0	2
4.	Tetralogy of Fallot	F117C04	1	0	2	21.	Coronary dissection	F117C21	1	0	2
5.	Other congenital heart defect, specify _____	F117C05 F117C05S	1	0	2	22.	Coronary ectasia	F117C22	1	0	2
6.	Mitral valve prolapse	F117C06	1	0	2	23.	Carotid dissection	F117C23	1	0	2
7.	Mitral valve stenosis	F117C07	1	0	2	24.	Symptomatic coronary artery disease	F117C24	1	0	2
8.	Mitral valve regurgitation > trace	F117C08	1	0	2	25.	Left cardiac hypertrophy	F117C25	1	0	2
9.	Calcified mitral annulus	F117C09	1	0	2	26.	Congestive heart failure/left ventricular function	F117C26	1	0	2
10.	Aortic valve regurgitation	F117C10	1	0	2	27.	Left ventricular dilation	F117C27	1	0	2
11.	Aortic valve stenosis	F117C11	1	0	2	28.	Left ventricular hypertrophy	F117C28	1	0	2

12.	Aortic coarctation, current/repaired	F117C12	1	0	2	29.	Right ventricular hypertrophy	F117C29	1	0	2
13.	Bicuspid valve regurgitation	F117C13	1	0	2	30.	Right ventricular dysfunction/failure	F117C30	1	0	2
14.	Tricuspid regurgitation > trace	F117C14	1	0	2	31.	Dilatation of the pulmonary artery	F117C31	1	0	2
15.	Pulmonary stenosis/right ventricular outflow obstruction	F117C15	1	0	2	32.	Other pulmonary artery abnormality	F117C32	1	0	2
16.	Pulmonary regurgitation > trace	F117C16	1	0	2	33.	Other, specify _____	F117C33 F117C33S	1	0	2
17.	Pulse deficits	F117C17	1	0	2	34.	Other, specify _____	F117C34 F117C34S	1	0	2

D. Craniofacial			Yes	No	DK				Yes	No	DK
1.	High palate/dental crowding	F117D01	1	0	2	6.	Down-slanting palpebral fissures	F117D06	1	0	2
2.	Dolicocephaly	F117D02	1	0	2	7.	Cleft palate	F117D07	1	0	2
3.	Malar hypoplasia	F117D03	1	0	2	8.	Bifid or broad-based uvula	F117D08	1	0	2
4.	Enophthalmos	F117D04	1	0	2	9.	Craniosynostosis	F117D09	1	0	2
5.	Retrognathia	F117D05	1	0	2	10.	Other, specify _____	F117D10 F117D10S	1	0	2

E. Ocular		Examiner Type: F117EXMN	1 Ophthalmologist	2 Optometrist	3 Unknown						
		Slit lamp Exam: F117LAMP	0 NO	1 YES							
			Yes	No	DK				Yes	No	DK
1.	Lens dislocation (ectopia lentis)	F117E01	1	0	2	8.	Scleral fragility or rupture of globe	F117E08	1	0	2
2.	Glaucoma	F117E02	1	0	2	9.	Myopia >4 diopters	F117E09	1	0	2
3.	Microcornea	F117E03	1	0	2	10.	Cataracts, <50 years of age	F117E10	1	0	2
4.	Hypertelorism	F117E04	1	0	2	11.	Iris Flocculi	F117E11	1	0	2
5.	Retinal detachment	F117E05	1	0	2	12.	Blue/grey sclera	F117E12	1	0	2
6.	Amblyopia	F117E06	1	0	2	13.	Other, specify _____	F117E13 F117E13S	1	0	2
7.	Strabismus	F117E07	1	0	2	14.	Other, specify _____	F117E14 F117E14S	1	0	2

F. Skin/integument			Yes	No	DK				Yes	No	DK
1.	Skin hyperextensibility	F117F01	1	0	2	8.	Striae atrophicae	F117F08	1	0	2
2.	Wide atrophic scars	F117F02	1	0	2	9.	Hernia, specify type _____	F117F09 F117F09S	1	0	2
3.	Poor wound healing	F117F03	1	0	2	10.	Recurrent hernia	F117F10	1	0	2
4.	Easy bruising	F117F04	1	0	2	11.	Thin translucent skin	F117F11	1	0	2
5.	Soft, velvety or doughy skin	F117F05	1	0	2	12.	Webbed neck	F117F12	1	0	2
6.	Tissue fragility	F117F06	1	0	2	13.	Other, specify _____	F117F13 F117F13S	1	0	2
7.	Lack of subcutaneous fat	F117F07	1	0	2	14.	Other, specify _____	F117F14 F117F14S	1	0	2

G. Central Nervous and Neurologic			Yes	No	DK				Yes	No	DK
1.	Lumbosacral dural ectasia	F117G01	1	0	2	5.	Abnormal junction of the brain and medulla (Arnold-Chiari malformation)	F117G05	1	0	2
2.	Hemorrhagic stroke	F117G02	1	0	2	6.	Migraines	F117G06	1	0	2

3.	Ischemic stroke	F117G03	1	0	2	7.	Cerebral aneurysms, specify location _____	F117G07 F117G07S	1	0	2
4.	Stroke, type unknown	F117G04	1	0	2	8.	Other, specify _____	F117G08 F117G08S	1	0	2

H. Vascular and Circulatory			Yes	No	DK				Yes	No	DK
1.	Early-onset varicose veins, <40 years of age	F117H01	1	0	2	4.	Lymphedema	F117H04	1	0	2
2.	Arteriovenous malformation	F117H02	1	0	2	5.	Other, specify _____	F117H05 F117H05S	1	0	2
3.	Arterial rupture	F117H03	1	0	2	6.	Other, specify _____	F117H06 F117H06S	1	0	2

I. Gastrointestinal			Yes	No	DK				Yes	No	DK
1.	Ulcerative colitis	F117I01	1	0	2	4.	Irritable bowel syndrome	F117I04	1	0	2
2.	Ruptured bowel	F117I02	1	0	2	5.	Diverticulosis/diverticulitis	F117I05	1	0	2
3.	Crohn's disease	F117I03	1	0	2	6.	Other, specify _____	F117I06 F117I06S	1	0	2

J. Reproductive			Yes	No	DK				Yes	No	DK
1.	Uterine rupture	F117J01	1	0	2	4.	Uterine fibroids	F117J04	1	0	2
2.	Ovarian failure	F117J02	1	0	2	5.	Premature rupture of membranes	F117J05	1	0	2
3.	Endometriosis	F117J03	1	0	2	6.	Other, specify _____	F117J06 F117J06S	1	0	2

K. Cognitive			Yes	No	DK				Yes	No	DK
1.	Learning disability	F117K01	1	0	2	3.	Other, specify _____	F117K03 F117K03S	1	0	2
2.	ADHD	F117K02	1	0	2	4.	Other, specify _____	F117K04 F117K04S	1	0	2

L. Familial History			Yes	No	DK				Yes	No	DK
1.	Immediate family history (parent, child, sibling) of TAA or other major vessel dilatation or dissection	F117L01	1	0	2	2.	Secondary family history (cousin, aunt, uncle, grandparent) of TAA or other major vessel dilatation or dissection	F117L02	1	0	2

18. Has this patient ever been diagnosed with diabetes? 1 Yes
 F118 0 No → GO TO Q19
2 Don't know → GO TO Q19

18a. What was the age at diagnosis? _____ years F118A

18b. Does the patient receive insulin? 1 Yes
F118B 0 No
2 Don't know

19. Has the patient ever been diagnosed with cancer? 1 Yes
F119 0 No → GO TO Q20
2 Don't know → GO TO Q20

For each primary cancer diagnosed, complete a row in the following table:

Cancer #	Primary Cancer Type	Cancer Location	Age at Diagnosis	Metastatic?
First	F1191A	F1191B	F1191C	F1191D 1 Yes 0 No 2 Unknown
Second	F1192A	F1192B	F1192C	F1192D 1 Yes 0 No 2 Unknown
Third	F1193A	F1193B	F1193C	F1193D 1 Yes 0 No 2 Unknown

20. Have any family members of this patient been enrolled in this Registry? **1** Yes
F120 (here or at any other participating center) **0** No → **GO TO Q21**
2 Don't Know → **GO TO Q21**

20a.
IF YES,

	GenTAC Registry ID Number	Relationship Code	Code List	
a.	Removed from dataset		01 = Maternal Grandparent	07 = Maternal Aunt
b.	See separate pedigree file		02 = Paternal Grandparent	08 = Paternal Aunt
c.			03 = Parent	09 = Maternal Uncle
d.			04 = Whole Sibling	10 = Paternal Uncle
e.			05 = Half Sibling	11 = Maternal Cousin
f.			06 = Child	12 = Paternal Cousin

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

Drug Treatment		Never Prescribed	Past Use Only	Currently Using	Unknown	Earliest Start Date Month/Year
a. Beta-Blocker	F121A	1	2	3	4	F121AMM / F121AYY
b. Losartan	F121B	1	2	3	4	F121BMM / F121BYY
c. Other ARB	F121C	1	2	3	4	F121CMM / F121CYY
d. ACE-Inhibitor	F121D	1	2	3	4	F121DMM / F121DYY
e. Statins	F121E	1	2	3	4	F121EMM / F121EYY
f. Verapamil or Diltiazem	F121F	1	2	3	4	F121FMM / F121FYY
g. Other Ca+ channel blocker	F121G	1	2	3	4	F121GMM / F121GY
h. Other BP lowering drug	F121H	1	2	3	4	F121HMM / F121HYY

22. Has the patient ever undergone a surgical procedure related to treatment of their cardiovascular genetic disorder?

F122

1 Yes

0 No → GO TO Q23

22a. Please indicate the procedures the patient received by checking the “YES” column. Remember to complete a Surgical Intervention Form for each procedure that was performed.

Surgical Intervention	Yes
a. Coronary artery bypass or PTCA grafting F122A	1
b. Aortic valve repair or replacement F122B	1
c. Isolated aortic valve replacement F122C	1
d. Aortic root replacement F122D	1
e. Valve-sparing aortic root replacement F122E	1
f. Ascending aortic replacement F122F	1
g. Aortic arch replacement F122G	1
h. Mitral valve repair or replacement F122H	1
i. Coarctation repair F122I	1
j. Pacemaker/ICD implantation F122J	1

Surgical Intervention	Yes
k. Descending thoracic aortic replacement F122K	1
L Thoracoabdominal aortic replacement F122L	1
m. Open arterial bypass F122M	1
n. Repair of pectus deformity F122N	1
o. Percutaneous/endovascular intervention, non-coronary PTCA F122O	1
p. Aorta to pulmonary shunt F122P	1
q. TOF repair F122Q	1
r. Pulmonary valve replacement F122R	1
s. Tricuspid valve surgery F122S	1
t. Other, specify _____ F122T F122TSP	1

23. Status and follow-up potential for this patient (*Check one.*) F122TSP

1 Patient new to institution and likely to have documentation of follow-up care

2 Patient new to institution and NOT likely to have documentation of follow-up care

3 Previously diagnosed patient with most of their follow-up done at our institution

4 Previously diagnosed patient with most of their follow-up done somewhere else

24. Source of information for answers recorded on this form (*Check all that apply.*)

1 Medical record of exams and tests done at your institution F1241

1 Medical record of exams and tests done elsewhere F1242

1 Medical record of patient interviews and clinical history F1243

1 Direct patient interview specifically for this study F1244

25. Year this patient was first seen at your institution: |__|__|__|__| F125

Comments on this form: _____ F1COMM

Initials of abstractor: |__|__|__| F1INIT

FORM COMPLETE	<ul style="list-style-type: none"> KEY DATA ON-LINE AT http://gentac.rti.org PUT FORM IN SUBJECT'S STUDY FILE.
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Gentac

FORM 1
ENROLLMENT CLINICAL
EVALUATION

Version 1.2, 3/27/2009

DIAGNOSIS

1. Patient is being enrolled in the GenTAC Registry because of the following condition(s) (*Check all that apply*):

a.	1	Marfan syndrome	F101A
b.	1	Turner syndrome	F101B
c.	1	Ehlers-Danlos syndrome, vascular type	F101C
d.	1	Ehlers-Danlos syndrome, other type and with aortic enlargement ¹	F101D
e.	1	Loeys-Dietz syndrome	F101E
f.	1	FBN1, TGFBR1 or TGFBR2, ACTA2 or MYH11 genetic mutation	F101F
g.	1	Bicuspid aortic valve with aortic enlargement ¹ and without known family history	F101G
h.	1	Bicuspid aortic valve with family history of BAV	F101H
i.	1	Bicuspid aortic valve with coarctation	F101N
j.	1	Shprintzen-Goldberg syndrome	F101I
k.	1	Familial Thoracic Aortic Aneurysm and Dissections with aortic enlargement ¹	F101J
l.	1	Other aneurysms/dissections of the thoracic aorta not due to trauma and in person < 40 years of age.	F101K
m.	1	Other congenital heart disease with aortic enlargement ¹ and family history (i.e., Tetralogy of Fallot)	F101L
n.	1	1st degree family member of proband already enrolled in the GenTAC registry	F101M

¹2 SD above normal based on nomographs OR prior dissection OR significant enlargement compared to adjacent area

2. Approximate age at earliest diagnosed above condition (physician confirmed) _____ years **F102**

3. Date of enrollment into Registry |_|_|_|-|_|_|_|-|_|_|_|_|_|_|
 Month Day Year
 F103MM F103DD F103YY

4. Has the patient ever been diagnosed with a thoracic aortic aneurysm, dissection/rupture, or marked tortuosity? **F104**

- 1** Yes
- 0** No → **GO TO Q6**
- 2** Unknown → **GO TO Q6**

5. Specify the current status of each aortic area (check all that apply in each row) and enter date of earliest abnormal:

Area of Aorta	Always Normal	Ever Tortuous	Ever Dilated	Ever Dissected	Unknown	Date of Earliest Abnormality
a. Ascending	1 F105A1	1 F105A2	1 F105A3	1 F105A4	1 F105A5	F105AMM F105ADD F105AYY
b. Arch	1 F105B1	1 F105B2	1 F105B3	1 F105B4	1 F105B5	F105BMM F105BDD F105BYY
c. Descending	1 F105C1	1 F105C2	1 F105C3	1 F105C4	1 F105C5	F105CMM F105CDD F105CYY
d. Abdominal	1 F105D1	1 F105D2	1 F105D3	1 F105D4	1 F105D5	F105DMM F105DDD F105DYY
e. Sinus of Valsalva	1 F105E1	1 F105E2	1 F105E3	1 F105E4	1 F105E5	F105EMM F105EDD F105EYY

DEMOGRAPHIC DATA

6. Date of birth |_|_|-|_|_|-|_|_|_|_|
 Month Day Year
 F106MM F106DD F106YY

7. Gender 1 Male
 F107 2 Female

8. Race (check all that apply) 1 White F1081
 1 Black or African American F1082
 1 Asian F1083
 1 American Indian or Alaska Native F1084
 1 Native Hawaiian or Other Pacific Islander F1085
 1 Unknown F1086

9. Hispanic/Latino origin? 1 Yes
 F109 0 No → **GO TO Q10**
 2 Don't Know → **GO TO Q10**

9a. What is the country of origin of the patient's family (e.g., Cuba, Mexico)? _____ F109A

BASIC MEASUREMENTS (at time of enrollment)

10. Height F110 |_|_|_|_| cm

11. Weight F111 |_|_|_|_|. |_| kg

12. Arm Span F112 |_|_|_|_| cm

13. Lower Segment F113 |_|_|_|_| cm

14. Heart Rate F114 |_|_|_|_| Beats/Minute

15. Blood Pressure F115S |_|_|_|_| systolic / |_|_|_|_| diastolic F115D

16. Karnofsky Performance 100 Normal, no complaints, no evidence of disease (100)
 F116 (Check one) 90 Able to carry on normal activity, minor symptoms of disease (90)
 80 Normal activity with effort, some symptoms of disease (80)
 70 Able to care for self, unable to do normal activity or work (70)
 60 Requires occasional assistance but is able to care for needs (60)
 50 Requires considerable assistance and frequent medical care (50)
 40 Disabled, requires special care and assistance (40)
 30 Severely disabled, hospitalization is indicated (30)
 20 Very sick, hospitalization and active treatment are necessary (20)
 10 Fatal processes progressing rapidly (10)

DIAGNOSTIC CRITERIA AND ORGAN SYSTEM REVIEW

17. Indicate the historical status of each condition with a check in the appropriate column (Yes, No, Don't Know)

A. Musculoskeletal			Yes	No	DK				Yes	No	DK
1.	Pectus carinatum	F117A01	1	0	2	14.	Tendon/muscle rupture	F117A14	1	0	2
2.	Pectus excavatum	F117A02	1	0	2	15.	Joint pain, arthralgia	F117A15	1	0	2
3.	Shield chest, broad with widely spaced nipples	F117A03	1	0	2	16.	Thumb sign (bony part of thumb protrudes from within clenched fist)	F117A16	1	0	2
4.	Recurrent dislocations/subluxations	F117A04	1	0	2	17.	Hypoplastic or hyperconvex nails	F117A17	1	0	2
5.	Joint hypermobility in >1 joint	F117A05	1	0	2	18.	Low back pain	F117A18	1	0	2
6.	Wrist sign (at least one phalanx of little finger overlaps with thumb)	F117A06	1	0	2	19.	Cubitus valgus (increased carrying angle of elbow)	F117A19	1	0	2
7.	Scoliosis that is clinically evident	F117A07	1	0	2	20.	Short fourth metacarpal or metatarsal	F117A20	1	0	2
8.	Abnormal kyphoscoliosis/lordosis	F117A08	1	0	2	21.	Clubfoot	F117A21	1	0	2
9.	Pes planus	F117A09	1	0	2	22.	Reduced elbow extension (<170°)	F117A22	1	0	2
10.	Protrusio acetabulae	F117A10	1	0	2	23.	Long fingers, arachnodactyly	F117A23	1	0	2
11.	Osteopenia/osteoporosis	F117A11	1	0	2	24.	Genu recurvatum	F117A25	1	0	2
12.	Congenital bilateral dislocated hips	F117A12	1	0	2	25.	Other, specify _____	F117A25 F117A25S	1	0	2
13.	Hypotonia	F117A13	1	0	2	26.	Other, specify _____	F117A26 F117A26S	1	0	2

B. Pulmonary			Yes	No	DK				Yes	No	DK
1.	Pneumothorax	F117B01	1	0	2	3.	Pulmonary hypertension	F117B02	1	0	2
2.	Asthma	F117B02	1	0	2	4.	Other, specify _____	F117B04 F117B04S	1	0	2

C. Cardiovascular			Yes	No	DK				Yes	No	DK
1.	Patent ductus arteriosus	F117C01	1	0	2	18.	Endocarditis	F117C18	1	0	2
2.	Atrial septal defect (ASD)/PFO	F117C02	1	0	2	19.	Atrial arrhythmia	F117C19	1	0	2
3.	Ventricular septal defect (VSD)	F117C03	1	0	2	20.	Cardiac arrhythmia (not atrial)	F117C20	1	0	2
4.	Tetralogy of Fallot	F117C04	1	0	2	21.	Coronary dissection	F117C21	1	0	2
5.	Other congenital heart defect, specify _____	F117C05 F117C05S	1	0	2	22.	Coronary ectasia	F117C22	1	0	2
6.	Mitral valve prolapse	F117C06	1	0	2	23.	Carotid dissection	F117C23	1	0	2
7.	Mitral valve stenosis	F117C07	1	0	2	24.	Symptomatic coronary artery disease	F117C24	1	0	2
8.	Mitral valve regurgitation > trace	F117C08	1	0	2	25.	Left cardiac hypertrophy	F117C25	1	0	2
9.	Calcified mitral annulus	F117C09	1	0	2	26.	Congestive heart failure/left ventricular function	F117C26	1	0	2
10.	Aortic valve regurgitation	F117C10	1	0	2	27.	Left ventricular dilation	F117C27	1	0	2
11.	Aortic valve stenosis	F117C11	1	0	2	28.	Left ventricular hypertrophy	F117C28	1	0	2

12.	Aortic coarctation, current/repai red	F117C12	1	0	2	29.	Right ventricular hypertrophy	F117C29	1	0	2
13.	Bicuspid valve regurgitation	F117C13	1	0	2	30.	Right ventricular dysfunction/failure	F117C30	1	0	2
14.	Tricuspid regurgitation > trace	F117C14	1	0	2	31.	Dilatation of the pulmonary artery	F117C31	1	0	2
15.	Pulmonary stenosis/right ventricular outflow obstruction	F117C15	1	0	2	32.	Other pulmonary artery abnormality	F117C32	1	0	2
16.	Pulmonary regurgitation > trace	F117C16	1	0	2	33.	Other, specify _____	F117C33 F117C33S	1	0	2
17.	Pulse deficits	F117C17	1	0	2	34.	Other, specify _____	F117C34 F117C34S	1	0	2

D. Craniofacial			Yes	No	DK				Yes	No	DK
1.	High palate/dental crowding	F117D01	1	0	2	6.	Down-slanting palpebral fissures	F117D06	1	0	2
2.	Dolicocephaly	F117D02	1	0	2	7.	Cleft palate	F117D07	1	0	2
3.	Malar hypoplasia	F117D03	1	0	2	8.	Bifid or broad-based uvula	F117D08	1	0	2
4.	Enophthalmos	F117D04	1	0	2	9.	Craniosynostosis	F117D09	1	0	2
5.	Retrognathia	F117D05	1	0	2	10.	Other, specify _____	F117D10 F117D10S	1	0	2

E. Ocular		Examiner Type: F117EXMN	1	Ophthalmologist	2	Optometrist	3	Unknown				
		Slit lamp Exam: F117LAMP	0	NO	1	YES				Yes	No	DK
			Yes	No	DK				Yes	No	DK	
1.	Lens dislocation (ectopia lentis)	F117E01	1	0	2	8.	Scleral fragility or rupture of globe	F117E08	1	0	2	
2.	Glaucoma	F117E02	1	0	2	9.	Myopia >4 diopters	F117E09	1	0	2	
3.	Microcornea	F117E03	1	0	2	10.	Cataracts, <50 years of age	F117E10	1	0	2	
4.	Hypertelorism	F117E04	1	0	2	11.	Iris Flocculi	F117E11	1	0	2	
5.	Retinal detachment	F117E05	1	0	2	12.	Blue/grey sclera	F117E12	1	0	2	
6.	Amblyopia	F117E06	1	0	2	13.	Iridodonesis	F117E15	1	0	2	
7.	Strabismus	F117E07	1	0	2	14.	Other, specify _____	F117E14 F117E14S	1	0	2	

F. Skin/integument			Yes	No	DK				Yes	No	DK
1.	Skin hyperextensibility	F117F01	1	0	2	8.	Striae atrophicae	F117F08	1	0	2
2.	Wide atrophic scars	F117F02	1	0	2	9.	Hernia, specify type _____	F117F09 F117F09S	1	0	2
3.	Poor wound healing	F117F03	1	0	2	10.	Recurrent hernia	F117F10	1	0	2
4.	Easy bruising	F117F04	1	0	2	11.	Thin translucent skin	F117F11	1	0	2
5.	Soft, velvety or doughy skin	F117F05	1	0	2	12.	Webbed neck	F117F12	1	0	2
6.	Tissue fragility	F117F06	1	0	2	13.	Other, specify _____	F117F13 F117F13S	1	0	2
7.	Lack of subcutaneous fat	F117F07	1	0	2	14.	Other, specify _____	F117F14 F117F14S	1	0	2

G. Central Nervous and Neurologic			Yes	No	DK				Yes	No	DK
1.	Lumbosacral dural ectasia	F117G01	1	0	2	5.	Abnormal junction of the brain and medulla (Arnold-Chiari malformation)	F117G05	1	0	2
2.	Hemorrhagic stroke	F117G02	1	0	2	6.	Migraines	F117G06	1	0	2
3.	Ischemic stroke	F117G03	1	0	2	7.	Cerebral aneurysms,	F117G07	1	0	2

				specify location _____	F117G07S						
4.	Stroke, type unknown	F117G04	1	0	2	8.	Other, specify _____	F117G08 F117G08S	1	0	2

H. Vascular and Circulatory			Yes	No	DK				Yes	No	DK
1.	Early-onset varicose veins, <40 years of age	F117H01	1	0	2	4.	Lymphedema	F117H04	1	0	2
2.	Arteriovenous malformation	F117H02	1	0	2	5.	Other, specify _____	F117H05 F117H05S	1	0	2
3.	Arterial rupture	F117H03	1	0	2	6.	Other, specify _____	F117H06 F117H06S	1	0	2

I. Gastrointestinal			Yes	No	DK				Yes	No	DK
1.	Ulcerative colitis	F117I01	1	0	2	4.	Irritable bowel syndrome	F117I04	1	0	2
2.	Ruptured bowel	F117I02	1	0	2	5.	Diverticulosis/diverticulitis	F117I05	1	0	2
3.	Crohn's disease	F117I03	1	0	2	6.	Other, specify _____	F117I06 F117I06S	1	0	2

J. Reproductive			Yes	No	DK				Yes	No	DK
1.	Uterine rupture	F117J01	1	0	2	4.	Uterine fibroids	F117J04	1	0	2
2.	Ovarian failure	F117J02	1	0	2	5.	Premature rupture of membranes	F117J05	1	0	2
3.	Endometriosis	F117J03	1	0	2	6.	Other, specify _____	F117J06 F117J06S	1	0	2

K. Cognitive			Yes	No	DK				Yes	No	DK
1.	Learning disability	F117K01	1	0	2	3.	Other, specify _____	F117K03 F117K03S	1	0	2
2.	ADHD	F117K02	1	0	2	4.	Other, specify _____	F117K04 F117K04S	1	0	2

L. Familial History			Yes	No	DK				Yes	No	DK
1.	Immediate family history (parent, child, sibling) of TAA or other major vessel dilatation or dissection	F117L01	1	0	2	2.	Secondary family history (cousin, aunt, uncle, grandparent) of TAA or other major vessel dilatation or dissection	F117L02	1	0	2

18. Has this patient ever been diagnosed with diabetes? 1 Yes
 F118 0 No → GO TO Q19
2 Don't know → GO TO Q19

18a. What was the age at diagnosis? _____ years F118A

18b. Does the patient receive insulin? 1 Yes
F118B 0 No
2 Don't know

19. Has the patient ever been diagnosed with cancer? 1 Yes
F119 0 No → GO TO Q20
2 Don't know → GO TO Q20

For each primary cancer diagnosed, complete a row in the following table:

Cancer #	Primary Cancer Type	Cancer Location	Age at Diagnosis	Metastatic?
First	F1191A	F1191B	F1191C	F1191D 1 Yes 0 No 2 Unknown
Second	F1192A	F1192B	F1192C	F1192D 1 Yes 0 No 2 Unknown
Third	F1193A	F1193B	F1193C	F1193D 1 Yes 0 No 2 Unknown

20. Have any family members of this patient been enrolled in this Registry? **1** Yes
F120 (here or at any other participating center) **0** No → **GO TO Q21**
2 Don't Know → **GO TO Q21**

20a.
IF YES,

	GenTAC Registry ID Number	Relationship Code	Code List	
a.	Removed from transferred dataset		01 = Maternal Grandparent	07 = Maternal Aunt
b.	See separate pedigree file		02 = Paternal Grandparent	08 = Paternal Aunt
c.			03 = Parent	09 = Maternal Uncle
d.			04 = Whole Sibling	10 = Paternal Uncle
e.			05 = Half Sibling	11 = Maternal Cousin
f.			06 = Child	12 = Paternal Cousin

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

Drug Treatment	Never Prescribed	Past Use Only	Currently Using	Possible use, clinical trial	Unknown	Earliest Start Date Month/Year
a. Beta-Blocker F121A	1	2	3	5	4	F121AMM / F121AYY
b. Losartan F121B	1	2	3	5	4	F121BMM / F121BYY
c. Other ARB F121C	1	2	3	5	4	F121CMM / F121CYY
d. ACE-Inhibitor F121D	1	2	3	5	4	F121DMM / F121DYY
e. Statins F121E	1	2	3	5	4	F121EMM / F121EYY
f. Verapamil or Diltiazem F121F	1	2	3	5	4	F121FMM / F121FYY
g. Other Ca+ channel blocker F121G	1	2	3	5	4	F121GMM / F121GY Y
h. Other BP lowering drug F121H	1	2	3	5	4	F121HMM / F121HYY

22. Has the patient ever undergone a surgical procedure related to treatment of their cardiovascular genetic disorder?

F122

1 Yes

0 No → GO TO Q23

22a. Please indicate the procedures the patient received by checking the “YES” column. Remember to complete a Surgical Intervention Form for each procedure that was performed.

Surgical Intervention	Yes
a. Coronary artery bypass or PTCA grafting F122A	1
b. Aortic valve repair or replacement F122B	1
c. Isolated aortic valve replacement F122C	1
d. Aortic root replacement F122D	1
e. Valve-sparing aortic root replacement F122E	1
f. Ascending aortic replacement F122F	1
g. Aortic arch replacement F122G	1
h. Mitral valve repair or replacement F122H	1
i. Coarctation repair F122I	1
j. Pacemaker/ICD implantation F122J	1

Surgical Intervention	Yes
k. Descending thoracic aortic replacement F122K	1
L Thoracoabdominal aortic replacement F122L	1
m. Open arterial bypass F122M	1
n. Repair of pectus deformity F122N	1
o. Percutaneous/endovascular intervention, non-coronary PTCA F122O	1
p. Aorta to pulmonary shunt F122P	1
q. TOF repair F122Q	1
r. Pulmonary valve replacement F122R	1
s. Tricuspid valve surgery F122S	1
t. Other, specify _____ F122T F122TSP	1

22b. How many different times has the patient had the above surgeries? F122B1 ____

23. Status and follow-up potential for this patient (*Check one.*) F122TSP

- 1 Patient new to institution and likely to have documentation of follow-up care
- 2 Patient new to institution and NOT likely to have documentation of follow-up care
- 3 Previously diagnosed patient with most of their follow-up done at our institution
- 4 Previously diagnosed patient with most of their follow-up done somewhere else

24. Source of information for answers recorded on this form (*Check all that apply.*)

- 1 Medical record of exams and tests done at your institution F1241
- 1 Medical record of exams and tests done elsewhere F1242
- 1 Medical record of patient interviews and clinical history F1243
- 1 Direct patient interview specifically for this study F1244

25. Year this patient was first seen at your institution: |__|__|__|__| F125

Comments on this form: _____ F1COMM

Initials of abstractor: |__|__|__| F1INIT



CLINICAL EVALUATION – FORM 1

Subject ID Label

1. Enrollment Diagnosis: (YOU MUST CHECK 1 PRIMARY CONDITION; YOU MAY ALSO CHECK 1 SECONDARY CONDITION.)

Diagnosis	1°	2°	Diagnosis	1°	2°
a. Marfan syndrome	F101A	F101A_S	h. BAV with family history	F101H	F101H_S
b. Turner syndrome	F101B	F101B_S	i. BAV with coarctation	F101N	F101N_S
c. Ehlers-Danlos syndrome, vascular type	F101C	F101C_S	j. Shprintzen-Goldberg syndrome	F101I	F101I_S
d. Ehlers-Danlos syndrome, other type and with aortic enlargement	F101D	F101D_S	k. Familial Thoracic Aortic Aneurysm and Dissections with aortic enlargement	F101J	F101J_S
e. Loeys-Dietz syndrome	F101E	F101E_S	l. Other aneurysms/dissections of the thoracic aorta in persons ≤ 50 years of age	F101K	F101K_S
f. FBN1, TGFBR1, TGFBR2, ACTA2 or MYH11 genetic mutation	F101F	F101F_S	m. Other congenital heart disease with aortic enlargement	F101L	F101L_S
g. BAV with aortic enlargement, family history not required	F101G	F101G_S	n. 1 st degree family member of proband already enrolled in the registry (with approval)	F101M	F101M_S

2. Approximate age at earliest diagnosed above condition (physician confirmed): _____ YEARS F102

3. Ever diagnosed with a thoracic aortic aneurysm, dissection/rupture, or marked tortuosity? Yes F104
 No → Go To Q5
 Unknown → Go To Q5

4. Specify the current status of each aortic area (CHECK ALL THAT APPLY IN EACH ROW) and enter date of earliest abnormal:

Area of Aorta	Always Normal	Ever Tortuous	Ever Dilated	Ever Dissected	Unknown	Date of Earliest Abnormality
a. Ascending	1 F105A1	1 F105A2	1 F105A3	1 F105A4	1 F105A5	F105AMM F105ADD F105AYY
b. Arch	1 F105B1	1 F105B2	1 F105B3	1 F105B4	1 F105B5	F105BMM F105BDD F105BYY
c. Descending	1 F105C1	1 F105C2	1 F105C3	1 F105C4	1 F105C5	F105CMM F105CDD F105CYY
d. Abdominal	1 F105D1	1 F105D2	1 F105D3	1 F105D4	1 F105D5	F105DMM F105DDD F105DYY
e. Sinus of Valsalva	1 F105E1	1 F105E2	1 F105E3	1 F105E4	1 F105E5	F105EMM F105EDD F105EYY

BASIC MEASUREMENTS (at time of enrollment)

5. Height F110 |__|__|__| CM F110A|__| FT F110B |__|__| INCHES
6. Weight F111 |__|__|__|. |__| KG F111A|__|__|__| POUNDS
7. Arm Span F112 |__|__|__| CM F112A|__|__| INCHES
8. Lower Segment F113 |__|__|__| CM F113A|__|__| INCHES
9. Heart Rate F114|__|__|__| BEATS/MINUTE
10. Blood Pressure F115S|__|__|__| SYSTOLIC / F115D |__|__|__| DIASTOLIC

DIAGNOSTIC CRITERIA AND ORGAN SYSTEM REVIEW

11. Indicate the historical status of each condition with a check in the appropriate column (YES, NO). Check NO if the patient is not known to have the condition.

A. Musculoskeletal		Yes	No/Unk			Yes	No/Unk
1. Pectus carinatum	F117A01	<input type="checkbox"/>	<input type="checkbox"/>	11. Osteopenia/osteoporosis	F117A11	<input type="checkbox"/>	<input type="checkbox"/>
2. Pectus excavatum	F117A02	<input type="checkbox"/>	<input type="checkbox"/>	12. Congenital bilateral dislocated hips	F117A12	<input type="checkbox"/>	<input type="checkbox"/>
3. Shield chest, broad with widely spaced nipples	F117A03	<input type="checkbox"/>	<input type="checkbox"/>	15. Joint pain, arthralgia	F117A15	<input type="checkbox"/>	<input type="checkbox"/>
4. Recurrent dislocations/subluxations	F117A04	<input type="checkbox"/>	<input type="checkbox"/>	16. Thumb sign	F117A16	<input type="checkbox"/>	<input type="checkbox"/>
5. Joint hypermobility in >1 joint	F117A05	<input type="checkbox"/>	<input type="checkbox"/>	21. Clubfoot	F117A21	<input type="checkbox"/>	<input type="checkbox"/>
6. Wrist sign	F117A06	<input type="checkbox"/>	<input type="checkbox"/>	22. Reduced elbow extension (<170°)	F117A22	<input type="checkbox"/>	<input type="checkbox"/>
7. Scoliosis that is clinically evident	F117A07	<input type="checkbox"/>	<input type="checkbox"/>	23. Long fingers, arachnodactyly	F117A23	<input type="checkbox"/>	<input type="checkbox"/>
8. Abnormal kyphoscoliosis/lordosis	F117A08	<input type="checkbox"/>	<input type="checkbox"/>	24. Genu recurvatum	F117A27	<input type="checkbox"/>	<input type="checkbox"/>
9. Pes planus	F117A09	<input type="checkbox"/>	<input type="checkbox"/>				
B. Pulmonary		Yes	No/Unk			Yes	No/Unk
1. Pneumothorax	F117B01	<input type="checkbox"/>	<input type="checkbox"/>	2. Asthma	F117B02	<input type="checkbox"/>	<input type="checkbox"/>
C. Cardiovascular		Yes	No/Unk			Yes	No/Unk
1. Patent ductus arteriosus	F117C01	<input type="checkbox"/>	<input type="checkbox"/>	18. Endocarditis	F117C18	<input type="checkbox"/>	<input type="checkbox"/>
2. Atrial septal defect (ASD)/PFO	F117C02	<input type="checkbox"/>	<input type="checkbox"/>	19. Atrial arrhythmia	F117C19	<input type="checkbox"/>	<input type="checkbox"/>
3. Ventricular septal defect (VSD)	F117C03	<input type="checkbox"/>	<input type="checkbox"/>	20. Cardiac arrhythmia (not atrial)	F117C20	<input type="checkbox"/>	<input type="checkbox"/>
4. Tetralogy of Fallot	F117C04	<input type="checkbox"/>	<input type="checkbox"/>	21. Coronary dissection	F117C21	<input type="checkbox"/>	<input type="checkbox"/>
5. Other congenital heart defect, specify:	F117C05 F117C05S	<input type="checkbox"/>	<input type="checkbox"/>	22. Coronary ectasia	F117C22	<input type="checkbox"/>	<input type="checkbox"/>
6. Mitral valve prolapse	F117C06	<input type="checkbox"/>	<input type="checkbox"/>	23. Carotid dissection	F117C23	<input type="checkbox"/>	<input type="checkbox"/>
8. Mitral valve regurgitation > trace	F117C08	<input type="checkbox"/>	<input type="checkbox"/>	24. Symptomatic coronary artery disease	F117C24	<input type="checkbox"/>	<input type="checkbox"/>
9. Calcified mitral annulus	F117C09	<input type="checkbox"/>	<input type="checkbox"/>	26. Congestive heart failure/left ventricular function	F117C26	<input type="checkbox"/>	<input type="checkbox"/>
10. Aortic valve regurgitation	F117C10	<input type="checkbox"/>	<input type="checkbox"/>	27. Left ventricular dilation	F117C27	<input type="checkbox"/>	<input type="checkbox"/>
11. Aortic valve stenosis	F117C11	<input type="checkbox"/>	<input type="checkbox"/>	28. Left ventricular hypertrophy	F117C28	<input type="checkbox"/>	<input type="checkbox"/>
12. Aortic coarctation, current/repared	F117C12	<input type="checkbox"/>	<input type="checkbox"/>	29. Right ventricular hypertrophy	F117C29	<input type="checkbox"/>	<input type="checkbox"/>
13. Bicuspid Aortic Valve (BAV)	F117C13	<input type="checkbox"/>	<input type="checkbox"/>	30. Right ventricular dysfunction/failure	F117C30	<input type="checkbox"/>	<input type="checkbox"/>
14. Tricuspid regurgitation > trace	F117C14	<input type="checkbox"/>	<input type="checkbox"/>	31. Dilatation of the pulmonary artery	F117C31	<input type="checkbox"/>	<input type="checkbox"/>
15. Pulmonary stenosis/right ventricular outflow obstruction	F117C15	<input type="checkbox"/>	<input type="checkbox"/>	32. Other pulmonary artery abnormality	F117C32	<input type="checkbox"/>	<input type="checkbox"/>
16. Pulmonary regurgitation > trace	F117C16	<input type="checkbox"/>	<input type="checkbox"/>	33. Hypertension	F117C33	<input type="checkbox"/>	<input type="checkbox"/>
D. Craniofacial		Yes	No/Unk			Yes	No/Unk
1. High palate/dental crowding	F117D01	<input type="checkbox"/>	<input type="checkbox"/>	6. Down-slanting palpebral fissures	F117D06	<input type="checkbox"/>	<input type="checkbox"/>
2. Dolicocephaly	F117D02	<input type="checkbox"/>	<input type="checkbox"/>	7. Cleft palate	F117D07	<input type="checkbox"/>	<input type="checkbox"/>
3. Malar hypoplasia	F117D03	<input type="checkbox"/>	<input type="checkbox"/>	8. Bifid or broad-based uvula	F117D08	<input type="checkbox"/>	<input type="checkbox"/>
4. Enophthalmos	F117D04	<input type="checkbox"/>	<input type="checkbox"/>	9. Craniosynostosis	F117D09	<input type="checkbox"/>	<input type="checkbox"/>
5. Retrognathia	F117D05	<input type="checkbox"/>	<input type="checkbox"/>				
E. Ocular		Examiner Type: F117EXMN <input type="checkbox"/> Ophthalmologist <input type="checkbox"/> Optometrist <input type="checkbox"/> Unknown Slit lamp Exam: F117LAMP <input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Unknown					
		Yes	No/Unk			Yes	No/Unk
1. Lens dislocation (ectopia lentis)	F117E01	<input type="checkbox"/>	<input type="checkbox"/>	9. Myopia >4 diopters	F117E09	<input type="checkbox"/>	<input type="checkbox"/>

2. Glaucoma	F117E02	<input type="checkbox"/>	<input type="checkbox"/>	10. Cataracts, <50 years of age	F117E10	<input type="checkbox"/>	<input type="checkbox"/>
4. Hypertelorism	F117E04	<input type="checkbox"/>	<input type="checkbox"/>	11. Iris Flocculi	F117E11	<input type="checkbox"/>	<input type="checkbox"/>
5. Retinal detachment	F117E05	<input type="checkbox"/>	<input type="checkbox"/>	12. Blue/grey sclera	F117E12	<input type="checkbox"/>	<input type="checkbox"/>
6. Amblyopia	F117E06	<input type="checkbox"/>	<input type="checkbox"/>	13. Iridodonesis	F117E15	<input type="checkbox"/>	<input type="checkbox"/>
7. Strabismus	F117E07	<input type="checkbox"/>	<input type="checkbox"/>				
F. Skin/integument		Yes No/Unk				Yes No/Unk	
1. Skin hyperextensibility	F117F01	<input type="checkbox"/>	<input type="checkbox"/>	8. Striae atrophicae	F117F08	<input type="checkbox"/>	<input type="checkbox"/>
2. Wide atrophic scars	F117F02	<input type="checkbox"/>	<input type="checkbox"/>	9. Hernia, specify type	F117F09 F117F09S	<input type="checkbox"/>	<input type="checkbox"/>
3. Poor wound healing	F117F03	<input type="checkbox"/>	<input type="checkbox"/>	10. Recurrent hernia	F117F10	<input type="checkbox"/>	<input type="checkbox"/>
4. Easy bruising	F117F04	<input type="checkbox"/>	<input type="checkbox"/>	11. Thin translucent skin	F117F11	<input type="checkbox"/>	<input type="checkbox"/>
5. Soft, velvety or doughy skin	F117F05	<input type="checkbox"/>	<input type="checkbox"/>	12. Webbed neck	F117F12	<input type="checkbox"/>	<input type="checkbox"/>
6. Tissue fragility	F117F06	<input type="checkbox"/>	<input type="checkbox"/>			<input type="checkbox"/>	<input type="checkbox"/>
G. Central Nervous and Neurologic		Yes No/Unk				Yes No/Unk	
1. Lumbosacral dural ectasia	F117G01	<input type="checkbox"/>	<input type="checkbox"/>	5. Abnormal junction of the brain and medulla (Arnold-Chiari malformation)	F117G05	<input type="checkbox"/>	<input type="checkbox"/>
2. Hemorrhagic stroke	F117G02	<input type="checkbox"/>	<input type="checkbox"/>	6. Migraines	F117G06	<input type="checkbox"/>	<input type="checkbox"/>
3. Ischemic stroke	F117G03	<input type="checkbox"/>	<input type="checkbox"/>	7. Cerebral aneurysms, specify location:	F117G07 F117G07S	<input type="checkbox"/>	<input type="checkbox"/>
4. Stroke, type unknown	F117G04	<input type="checkbox"/>	<input type="checkbox"/>				
H. Vascular and Circulatory		Yes No/Unk				Yes No/Unk	
1. Early-onset varicose veins, <40 years of age	F117H01	<input type="checkbox"/>	<input type="checkbox"/>	3. Arterial rupture	F117H03	<input type="checkbox"/>	<input type="checkbox"/>
2. Arteriovenous malformation	F117H02	<input type="checkbox"/>	<input type="checkbox"/>	4. Lymphedema	F117H04	<input type="checkbox"/>	<input type="checkbox"/>
I. Gastrointestinal		Yes No/Unk				Yes No/Unk	
1. Ulcerative colitis	F117I01	<input type="checkbox"/>	<input type="checkbox"/>	4. Irritable bowel syndrome	F117I04	<input type="checkbox"/>	<input type="checkbox"/>
2. Ruptured bowel	F117I02	<input type="checkbox"/>	<input type="checkbox"/>	5. Diverticulosis/diverticulitis	F117I05	<input type="checkbox"/>	<input type="checkbox"/>
3. Crohn's disease	F117I03	<input type="checkbox"/>	<input type="checkbox"/>			<input type="checkbox"/>	<input type="checkbox"/>
J. Reproductive (WOMEN ONLY)		Yes No/Unk				Yes No/Unk	
1. Uterine rupture	F117J01	<input type="checkbox"/>	<input type="checkbox"/>	4. Uterine fibroids	F117J04	<input type="checkbox"/>	<input type="checkbox"/>
2. Ovarian failure	F117J02	<input type="checkbox"/>	<input type="checkbox"/>	5. Premature rupture of membranes	F117J05	<input type="checkbox"/>	<input type="checkbox"/>
3. Endometriosis	F117J03	<input type="checkbox"/>	<input type="checkbox"/>			<input type="checkbox"/>	<input type="checkbox"/>
K. Other Important Health Problem Potentially Related to Diagnosis		Yes No/Unk				Yes No/Unk	
1. Learning disability	F117K01	<input type="checkbox"/>	<input type="checkbox"/>	3. Other, specify:	F117K03 F117K03S	<input type="checkbox"/>	<input type="checkbox"/>
2. ADHD	F117K02	<input type="checkbox"/>	<input type="checkbox"/>	4. Other, specify:	F117K04 F117K04S	<input type="checkbox"/>	<input type="checkbox"/>
L. Familial History		Yes No/Unk				Yes No/Unk	
1. Immediate family history (parent, child, sibling) of TAA or other major vessel dilatation or dissection	F117L01	<input type="checkbox"/>	<input type="checkbox"/>	2. Secondary family history (cousin, aunt, uncle, grandparent) of TAA or other major vessel dilatation or dissection	F117L02	<input type="checkbox"/>	<input type="checkbox"/>

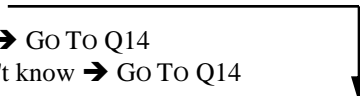
12. Has patient ever been diagnosed with diabetes?

- F118 Yes →
 No → Go To Q13
 Don't know → Go To Q13

F118AUN – Age Unknown
 12a. What was the age at diagnosis? F118A _____ YEARS

12b. Does the patient receive insulin? Yes
 F118B Don't know

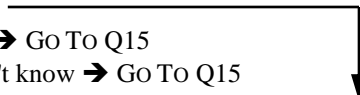
13. Patient ever been diagnosed with cancer?

- F119** Yes 
 No → Go To Q14
 Don't know → Go To Q14

13a. IF YES: For each primary cancer, complete a row in the table:

	Primary Cancer Type	Cancer Location	Age at Diagnosis	Metastatic?
1.	F1191A	F1191B	F1191C	<input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unknown F1191D
2.	F1192A	F1192B	F1192C	<input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unknown F1192D

14. Have any family members of this patient been enrolled in this Registry (here or at any other participating center)?

- F120** Yes 
 No → Go To Q15
 Don't know → Go To Q15

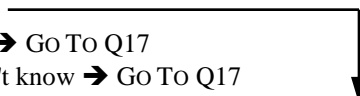
14a. IF YES: For each family member enrolled in this Registry, complete a row in the table:

	GenTAC ID Number	Code	Relationship Code List		
1.	Removed from dataset		00 = Grandchild	07 = Maternal Aunt	13 = Maternal Niece
2.	See separate pedigree file		01 = Maternal Grandparent	08 = Paternal Aunt	14 = Paternal Niece
3.			02 = Paternal Grandparent	09 = Maternal Uncle	15 = Maternal Nephew
4.			03 = Parent	10 = Paternal Uncle	16 = Paternal Nephew
5.			04 = Whole Sibling	11 = Maternal Cousin	17 = Other
6.			05 = Half Sibling	12 = Paternal Cousin	
			06 = Child		

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

Drug Treatment	Never Prescribed	Past Use Only	Currently Using	Possible Use, Clinical Trial	Unknown	Earliest Start Date Month/Year
a. Beta-Blocker F121A	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121AMM / F121AYY
b. Losartan F121B	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121BMM / F121BYY
c. Other ARB F121C	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121CMM / F121CYY
d. ACE-Inhibitor F121D	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121DMM / F121DYY
e. Statins F121E	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121EMM / F121EYY
f. Verapamil or Diltiazem F121F	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121FMM / F121FYY
g. Other Ca+ channel blocker F121G	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121GMM / F121GYG
h. Other BP lowering drug F121H	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	F121HMM / F121HYG

16. Has patient ever had a surgical procedure related to treatment of their cardiovascular genetic disorder?

- F122** Yes 
 No → Go To Q17
 Don't know → Go To Q17

16a. IF YES: Indicate the procedures received 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 Repair of pectus deformity.

Surgical Intervention	Yes	MM/YY	Surgical Intervention	Yes	MM/YY
1. Coronary artery bypass or PTCA grafting F122A	<input type="checkbox"/>	F122AMM/ F122AYY	11. Descending thoracic aortic replacement F122K	<input type="checkbox"/>	F122KMM/ / F122KYY
2. Aortic valve repair F122B	<input type="checkbox"/>	F122BMM/ F122BYY	12. Thoracoabdominal aortic replacement F122L	<input type="checkbox"/>	F122LMM/ F122LYY
3. Isolated aortic valve replacement (non-root) F122C	<input type="checkbox"/>	F122CMM/ F122CYY	13. Open arterial bypass F122M	<input type="checkbox"/>	F122MMM/ / F122MY Y
4. Aortic root replacement (valve-replacing) F122D	<input type="checkbox"/>	F122DMM/ F122DYY	14. Repair of pectus deformity F122N	<input type="checkbox"/>	F122NMM/ / F122NYY
5. Valve-sparing aortic root replacement F122E	<input type="checkbox"/>	F122EMM/ F122EYY	15. Percutaneous/endovascular intervention, non-coronary PTCA F122O	<input type="checkbox"/>	F122OMM/ / F122OYY
6. Ascending aortic replacement F122F	<input type="checkbox"/>	F122FMM/ F122FYY	16. Aorta to pulmonary shunt F122P	<input type="checkbox"/>	F122PMM/ F122PYY
7. Aortic arch replacement F122G	<input type="checkbox"/>	F122GMM/ F122GY Y	17. TOF repair F122Q	<input type="checkbox"/>	F122QMM/ / F122QYY
8. Mitral valve repair or replacement F122H	<input type="checkbox"/>	F122HMM/ F122HY Y	18. Pulmonary valve replacement F122R	<input type="checkbox"/>	F122RMM/ F122RY Y
9. Coarctation repair F122I	<input type="checkbox"/>	F122IMM/ F122IYY	19. Tricuspid valve surgery F122S	<input type="checkbox"/>	F122SMM/ F122SY Y
10. Pacemaker/ICD implantation F122J	<input type="checkbox"/>	F122JMM/ F122JYY	20. Other, specify: F122T F122TSP	<input type="checkbox"/>	F122TMM/ F122TY Y

16b. How many different Surgical Intervention Forms will be completed? F122B1 _____ Unknown F122B2

17. Source of information for answers recorded on this form (*Check all that apply.*)

- Medical record of exams and tests done at your institution F1241
- Medical record of exams and tests done elsewhere F1242
- Medical record of patient interviews and clinical history F1243
- Direct patient interview specifically for this study F1244

18. Year this patient was first seen at your institution: |__|_|_|_|_| F125 Patient not seen at this institution F125UN

Initials of Abstractor _____ F11INIT

Comments: _____ F1COMM



GenTAC

FORM 2
ENROLLMENT PATIENT
QUESTIONNAIRE

Version 1.0, 10/01/2007

We are interested in learning more about people who are thought to have a genetic condition that may cause a problem with their thoracic aorta. As you complete this form, answer the questions as best as you can. If you don't know the answer or do not want to answer a question, you may leave it blank. If you are completing this form for a child or other dependent that is enrolled in this study, any references to "you" or "your" refer to that person.

Today's Date: |__|_|_|-|__|_|_|-|__|_|_|_|_|_|
 Month Day Year
 F2_MM F2_DD F2_YY

SECTION A: DEMOGRAPHIC DATA

1. What is your date of birth? |__|_|_|-|__|_|_|-|__|_|_|_|_|_|
 Month Day Year
 F2A01MM F2A01DD F2A01YY

2. What is your gender? F2A02

- 1 Male
- 2 Female

3. Which of the following groups describes your race? (*You may choose more than one*)

- 1 White F2A031
- 1 Black or African American F2A032
- 1 Asian F2A033
- 1 American Indian or Alaskan Native F2A034
- 1 Native Hawaiian or Other Pacific Islander F2A035

4. Are you of Hispanic or Latino origin? F2A04

- 0 No → GO TO Q5
- 1 Yes

4a. What is your country of origin? _____ F2A04SP

5. What is the highest level of schooling you have completed? (*Choose only one.*) F2A05

- 1 1-8 years
- 2 Some high school
- 3 High school graduate
- 4 GED (high school equivalency)
- 5 1-3 years vocational education beyond high school
- 6 Some college
- 7 College graduate
- 8 One or more years of graduate school or professional school
- 9 Something else (*Please describe below*)

_____ F2A05SP

6. What is your current marital status? **F2A06**
- 1 Married or living as married (including living with a partner)
 - 2 Divorced or separated
 - 3 Widowed
 - 4 Never married
7. Are you covered by health insurance or some other kind of health care plan? **F2A07**
- 0 No → **GO TO Q8**
 - 1 Yes
- 7a. What kind of health insurance or health care coverage do you have? (*Choose only one.*) **F2A07A**
- 1 Private health insurance plan from employer
 - 2 Medicare
 - 3 Medicaid
 - 4 Other, specify _____ **F2A07ASP**
8. What is your approximate yearly household income? Include income from all sources. **F2A08**
- 1 \$25,000 and under
 - 2 \$25,001 - \$50,000
 - 3 \$50,001 - \$75,000
 - 4 \$75,001 - \$100,000
 - 5 \$100,001 or more
9. What is your primary current employment status? (*Choose only one.*) **F2A09**
- 1 Employed full-time at a job or business
 - 2 Employed part-time at a job or business
 - 3 Self-employed
 - 4 Unable to work due to disability
 - 5 Student
 - 6 Homemaker
 - 7 Retired
 - 8 Unemployed
 - 9 Something else (*Please describe below*)
-
- F2A09A**

IF YOU ARE UNDER 18 YEARS OF AGE, CHECK THIS BOX AND GO TO SECTION E.

SECTION B: ALCOHOL USE

The next two questions ask about your general habits concerning alcoholic beverages. For these questions, **a drink is defined as one beer, a glass of wine, or a shot of hard liquor.**

1. During the past 12 months, how often did you usually drink any kind of alcoholic beverage? **F2B01**

- 1 Never → **GO TO SECTION C**
- 2 Less than one time a month
- 3 1-3 times a month
- 4 1 time a week
- 5 2-4 times a week
- 6 Almost every day
- 7 Every day

2. During the past 12 months, about how many drinks would you have on a day when you drank? **F2B02**

- 1 1 or 2 drinks
- 2 3 or 4 drinks
- 3 5-8 drinks
- 4 9 or more drinks

SECTION C: SMOKING HISTORY

1. During your lifetime, have you smoked at least 100 cigarettes? **F2C01**

- 0 No → **GO TO SECTION D**
- 1 Yes

2. Do you smoke cigarettes now? **F2C02**

- 0 No
- 1 Yes

3. On average, how much do you or did you smoke each day? (*1 pack = 20 cigarettes*) **F2C03**

- 1 10 cigarettes or less
- 2 11-20 cigarettes
- 3 21-40 cigarettes
- 4 More than 40 cigarettes

4. What is the total number of years you smoked cigarettes? (*Do not count years you did not smoke.*)

_____ Total number of years you smoked cigarettes **F2C04**

5. Other than cigarettes, which of the following tobacco products have you used on a regular basis for six months or longer? (*Choose all the tobacco products that you used.*)

1	Pipe	F2C051
1	Cigars	F2C052
1	Cigarillos	F2C053
1	Chewing tobacco	F2C054
1	Snuff	F2C055
1	Never used any of these tobacco products for six months or longer	F2C056

SECTION D: RECREATIONAL DRUG USE

The next questions are about certain drugs you might have used on a recreational basis. Place an X in Column A if you've never used the drug. Place an X in Column B if you used the drug in the past 12 months. Place an X in Column C if you've used it but not in the past 12 months. If you don't know if you've use it, place an X in the last column. Give only 1 answer for each drug, and remember that your answers will be kept strictly confidential.

	<u>Col A</u> No, I've never used this drug	<u>Col B</u> Yes, I've used this drug in the past 12 months	<u>Col C</u> Yes, I've used this drug but not in the past 12 months	<u>Col D</u> I don't know if I've used this drug
1. Crack cocaine F2D01	1	2	3	4
2. Other types of cocaine, like powder F2D02	1	2	3	4
3. Stimulants, like amphetamines, methamphetamines, or ecstasy F2D03	1	2	3	4
4. Narcotics, like heroin, codeine, or Morphine F2D04	1	2	3	4
5. Hallucinogens or psychedelics, like LSD, PCP, or mushrooms F2D05	1	2	3	4
6. Cannabis, like marijuana or hash F2D06	1	2	3	4
7. Sedatives (benzodiazepines), like Valium, Xanax, or Avitan F2D07	1	2	3	4
8. Depressants or downers (barbiturates), like Phenobarbital, Seconal, Nembutal, Amytal, or Placidyls F2D08	1	2	3	4
9. Inhalants, like amyl nitrate, ether, or sniffing glue or lighter fluid F2D09	1	2	3	4
10. Psychotropic drugs or anti-depressants, like Prozac or Zoloft F2D10	1	2	3	4
11. Anabolic steroids F2D11	1	2	3	4
12. 2 or more drugs taken together, like cocaine and heroin, specify: _____ F2D12	1	2	3	4
13. Other, specify: _____ F2D13 F2D13SP	1	2	3	4
14. Other, specify: _____ F2D14 F2D14SP	1	2	3	4

SECTION E: HEALTH AND WELL-BEING

1. In general, would you say your health is... F2E01

- 1 Excellent
- 2 Very good
- 3 Good
- 4 Fair
- 5 Poor

2. Compared to one year ago, how would you rate your health in general now? F2E02

- 1 Much better now than one year ago
- 2 Somewhat better now than one year ago
- 3 About the same as one year ago
- 4 Somewhat worse now than one year ago
- 5 Much worse now than one year ago

3. In the following table, record the number of hours you spend doing each activity in an average week.

Activity	# of hours per week
a. Strenuous activity such as aerobics, running, swimming, active sports, shoveling, lifting heavy objects, etc. F2E03A	
b. Moderate activity such as walking for exercise, cleaning house, gardening, carpentry, etc. F2E03B	
c. Slight activity , such as walking on level ground around the house, office, shopping, etc. F2E03C	

4. The following questions are about activities you might do during a typical day. Does your health now limit you in these activities? If so, how much? **If your activities are limited a lot**, please indicate whether they are restricted on the advice of your doctor.

Activity	No, not limited at all	Yes, limited a little	Yes, limited a lot →	Due to a doctor's recommendation?	
				Yes	No
Example: riding a bike			X		X
a. Vigorous activities, such as running, lifting heavy objects, participating in strenuous sports	1 F2E04A1	2	3	1 F2E04A2	0
b. Moderate activities, such as moving a table, pushing a vacuum cleaner, bowling, or playing golf	1 F2E04B1	2	3	1 F2E04B2	0
c. Lifting or carrying groceries	1 F2E04C1	2	3	1 F2E04C2	0
d. Climbing several flights of stairs	1 F2E04D1	2	3	1 F2E04D2	0
e. Climbing one flight of stairs	1 F2E04E1	2	3	1 F2E04E2	0
f. Bending, kneeling, or stooping	1 F2E04F1	2	3	1 F2E04F2	0
g. Walking more than a mile	1 F2E04G1	2	3	1 F2E04G2	0
h. Walking several hundred yards	1 F2E04H1	2	3	1 F2E04H2	0
i. Walking one hundred yards	1 F2E04I1	2	3	1 F2E04I2	0
j. Bathing or dressing yourself	1 F2E04J1	2	3	1 F2E04J2	0

5. During the past 4 weeks, how much of the time have you had any of the following problems with your work or other regular daily activities as a result of your physical health?

	All of the time	Most of the time	Some of the time	A little of the time	None of the time
a. Cut down on the <u>amount of time</u> you spent on work or other activities F2E05A	1	2	3	4	5
b. <u>Accomplished less than</u> you would like F2E05B	1	2	3	4	5
c. Were limited in the <u>kind</u> of work or other activities F2E05C	1	2	3	4	5
d. Had <u>difficulty</u> performing the work or other activities (for example, it took extra effort) F2E05D	1	2	3	4	5

6. How much bodily pain have you had during the past 4 weeks? F2E06

- 1 None
- 2 Very mild
- 3 Mild
- 4 Moderate
- 5 Severe
- 6 Very Severe

7. During the past 4 weeks, how much did pain interfere with your normal work (including both work outside the home and housework)? F2E07

- 1 Not at all
- 2 Slightly
- 3 Moderately
- 4 Quite a bit
- 5 A lot

8. How TRUE or FALSE is each of the following statements for you?

	Definitely true	Mostly true	Don't know	Mostly false	Definitely false
a. I seem to get sick a little easier than other people F2E08A	1	2	3	4	5
b. I am as healthy as anybody I know F2E08B	1	2	3	4	5
c. I expect my health to get worse F2E08C	1	2	3	4	5
d. My health is excellent F2E08D	1	2	3	4	5

SECTION F: SLEEPING

1. On an average night, how many hours do you sleep? F2F01

_____ Number of hours of sleep on an average night

2. Do you usually have trouble falling asleep? F2F02

- 0 No
- 1 Yes

3. Except to use the bathroom, do you usually wake up several times a night? F2F03

- 0 No
- 1 Yes

4. Has your spouse or other housemate(s) complained about your loud snoring? F2F04

- 0 No
- 1 Yes

SECTION G: HEARING/VISION

1. Have you ever worn a hearing aid? **F2G01**
0 No
1 Yes

2. Does a hearing problem cause you difficulty when listening to television or radio? **F2G02**
0 No
1 Yes

3. Do you wear eyeglasses or contact lenses? **F2G03**
0 No
1 Yes

4. How would you rate your vision, **without** eyeglasses or contact lenses if you wear them? **F2G04**
1 Excellent
2 Good
3 Fair
4 Poor

5. Have you ever had any surgical procedure to correct or improve your vision? **F2G05**
1 No
2 Yes, LASIK or laser correction
3 Yes, lens extraction and replacement
4 Yes, implanted contact lens
5 Yes, cataract removal
6 Yes, other

IF YOU ARE A MALE, CHECK THIS BOX AND GO TO SECTION J.

SECTION H: PREGNANCIES

The next questions are about any pregnancies you have had.

1. Have you ever been pregnant? **F2H01**
0 No → **GO TO SECTION J**
1 Yes

2. Are you currently pregnant? **F2H02**
0 No
1 Yes

3. How many times have you been pregnant? Please be sure to include any pregnancies that ended in a live birth, miscarriage, stillbirth, or abortion, as well as a tubal or molar pregnancy. **F2H03**

_____ Number of times you have been pregnant

As you answer the following questions, please think about each of your pregnancies. Start with your very first pregnancy, listing it in the column labeled “1st pregnancy”. From there, work forward until you have provided information about all of your pregnancies, ending with the most recent. If you have been pregnant more than 6 times, please contact the coordinator for additional forms on which to record this information.

	1 st pregnancy	2 nd pregnancy	3 rd pregnancy
4. In what month and year did this pregnancy end? (If currently pregnant, enter your due date.)	_____/_____ Month / Year F2H04M F2H04Y	_____/_____ Month / Year	_____/_____ Month / Year
5. Was this a pregnancy with multiples?	F2H05 0 <input type="checkbox"/> No 1 Yes → How many? ____ F2H05N	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> <input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____
6. What medicines or procedures did you have to help you get pregnant? <i>(Check all that apply)</i>	1 None F2H061 1 Pills or shots F2H062 1 Artificial insemination F2H063 1 In vitro fertilization F2H064 1 Donor egg F2H065 1 Donor sperm F2H066 1 Other F2H067	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other
7. <u>While you were pregnant</u> , did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	F2H07 0 No 1 Yes → What condition? _____ F2H07SP	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____
8. What was the outcome of this pregnancy? <i>(If you were pregnant with multiples, check all that apply and enter the number of babies with that outcome.)</i>	1 Live birth #___ F2H081 F2H081N 1 Still birth #___ F2H082 F2H082N 1 Miscarriage #___ F2H083 F2H083N 1 Abortion (elective, induced) #___ F2H084 F2H084N 1 Tubal, ectopic #___ F2H085 F2H085N 1 Other #___ F2H086 F2H086S #___ F2H086N 1 Currently pregnant #___ F2H087 F2H087N	<input type="checkbox"/> Live birth #___ <input type="checkbox"/> Still birth #___ <input type="checkbox"/> Miscarriage #___ <input type="checkbox"/> Abortion (elective, induced) #___ <input type="checkbox"/> Tubal, ectopic #___ <input type="checkbox"/> Other #___ <input type="checkbox"/> Currently pregnant #___	<input type="checkbox"/> Live birth #___ <input type="checkbox"/> Still birth #___ <input type="checkbox"/> Miscarriage #___ <input type="checkbox"/> Abortion (elective, induced) #___ <input type="checkbox"/> Tubal, ectopic #___ <input type="checkbox"/> Other #___ <input type="checkbox"/> Currently pregnant #___
9. <u>After this pregnancy</u> was over, did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	F2H09 0 No 1 Yes → What condition? _____ F2H09SP	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____

The following are questions about your health during and after each of your pregnancies.

	1 st pregnancy	2 nd pregnancy	3 rd pregnancy
10. <u>While you were pregnant</u> , which of these conditions did you have?	<p>1 None F2H101</p> <p>1 High blood pressure, toxemia F2H102</p> <p>1 Maternal diabetes F2H103</p> <p>1 Premature rupture of membranes F2H104</p> <p>1 Change in aortic dimension F2H105</p> <p>1 Aortic dissection F2H106</p> <p>1 Aortic rupture F2H107</p>	<p><input type="checkbox"/> None</p> <p><input type="checkbox"/> High blood pressure, toxemia</p> <p><input type="checkbox"/> Maternal diabetes</p> <p><input type="checkbox"/> Premature rupture of membranes</p> <p><input type="checkbox"/> Change in aortic dimension</p> <p><input type="checkbox"/> Aortic dissection</p> <p><input type="checkbox"/> Aortic rupture</p>	<p><input type="checkbox"/> None</p> <p><input type="checkbox"/> High blood pressure, toxemia</p> <p><input type="checkbox"/> Maternal diabetes</p> <p><input type="checkbox"/> Premature rupture of membranes</p> <p><input type="checkbox"/> Change in aortic dimension</p> <p><input type="checkbox"/> Aortic dissection</p> <p><input type="checkbox"/> Aortic rupture</p>
11. Were you on bed rest during this pregnancy?	<p>F2H11</p> <p>0 No</p> <p>1 Yes→ Why? For how long? _____ F2H11SP</p>	<p><input type="checkbox"/> No</p> <p><input type="checkbox"/> Yes→ Why? For how long? _____</p>	<p><input type="checkbox"/> No</p> <p><input type="checkbox"/> Yes→ Why? For how long? _____</p>
12. <u>After this pregnancy</u> , which of these conditions did you have?	<p>1 None F2H121</p> <p>1 Change in aortic dimension F2H122</p> <p>1 Aortic dissection F2H123</p> <p>1 Aortic rupture F2H124</p>	<p><input type="checkbox"/> None</p> <p><input type="checkbox"/> Change in aortic dimension</p> <p><input type="checkbox"/> Aortic dissection</p> <p><input type="checkbox"/> Aortic rupture</p>	<p><input type="checkbox"/> None</p> <p><input type="checkbox"/> Change in aortic dimension</p> <p><input type="checkbox"/> Aortic dissection</p> <p><input type="checkbox"/> Aortic rupture</p>

	4th pregnancy	5th pregnancy	6th pregnancy
4. In what month and year did this pregnancy end? (If currently pregnant, enter your due date.)	_____/_____ Month / Year	_____/_____ Month / Year	_____/_____ Month / Year
5. Was this a pregnancy with multiples?	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____
6. What medicines or procedures did you have to help you get pregnant? (Check all that apply)	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other
7. <u>While you were pregnant</u> , did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____ _____
8. What was the outcome of this pregnancy? (If you were pregnant with multiples, check all that apply and enter the number of babies with that outcome.)	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____
9. <u>After this pregnancy</u> was over, did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____ _____

The following are questions about your health during and after each of your pregnancies.

	4th pregnancy	5th pregnancy	6th pregnancy
10. <u>While you were pregnant</u> , which of these conditions did you have?	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture
11. Were you on bed rest during this pregnancy?	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____
12. <u>After this pregnancy</u> , which of these conditions did you have?	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture

SECTION J: MEDICAL CONDITIONS

1. Has a doctor ever told you that you had any of the following conditions? Place a check in the appropriate column (No, Yes or Don't Know). For each **YES** answer, please provide your age at the time of diagnosis or procedure.

Condition		No	Yes →	Age at Diagnosis	Don't Know
1	Marfan syndrome	0 F2J0101	1	F2J0101A	2
2	Turner syndrome	0 F2J0102	1	F2J0102A	2
3	Ehlers-Danlos syndrome	0 F2J0103	1	F2J0103A	2
4	Loeys-Dietz syndrome	0 F2J0104	1	F2J0104A	2
5	FBN1, TGFBR1 or TGFBR2 genetic mutations	0 F2J0105	1	F2J0105A	2
6	Bicuspid aortic valve	0 F2J0106	1	F2J0106A	2
7	Shprintzen-Goldberg syndrome	0 F2J0107	1	F2J0107A	2
8	Heart murmur	0 F2J0108	1	F2J0108A	2
9	Heart rhythm palpitations	0 F2J0109	1	F2J0109A	2
10	Angina	0 F2J0110	1	F2J0110A	2
11	Heart attack	0 F2J0111	1	F2J0111A	2
12	Atherosclerosis, clogged arteries	0 F2J0112	1	F2J0112A	2
13	Heart, blood vessel infection (endocarditis)	0 F2J0113	1	F2J0113A	2
14	Cardiomyopathy or heart failure	0 F2J0114	1	F2J0114A	2
15	Congenital heart disease, specify: _____ F2J0115S	0 F2J0115	1	F2J0115A	2
16	Other heart disease, specify: _____ F2J0116S	0 F2J0116	1	F2J0116A	2
17	Hypertension, high blood pressure	0 F2J0117	1	F2J0117A	2
18	Stroke	0 F2J0118	1	F2J0118A	2
19	Aneurysms outside the aorta	0 F2J0119	1	F2J0119A	2
20	Asthma	0 F2J0120	1	F2J0120A	2
21	Emphysema	0 F2J0121	1	F2J0121A	2
22	Pneumonia	0 F2J0122	1	F2J0122A	2
23	Pneumothorax or collapsed lung	0 F2J0123	1	F2J0123A	2
24	Other pulmonary or lung disease, specify: _____ F2J0124S	0 F2J0124	1	F2J0124A	2
25	Cancer, specify site: _____ F2J0125S	0 F2J0125	1	F2J0125A	2

Condition		No	Yes →	Age at Diagnosis	Don't Know
26	Cancer, specify site: _____ F2J0126S	0 F2J0126	1	F2J0126A	2
27	Diabetes	0 F2J0127	1	F2J0127A	2
28	Bleeding disorder	0 F2J0128	1	F2J0128A	2
29	Blood clotting disorder	0 F2J0129	1	F2J0129A	2
30	Endometriosis (<i>leave blank if male</i>)	0 F2J0130	1	F2J0130A	2
31	Uterine fibroids (<i>leave blank if male</i>)	0 F2J0131	1	F2J0131A	2
32	Raynaud's syndrome (fingers turn purple when cold)	0 F2J0132	1	F2J0132A	2
33	Liver disease or cirrhosis	0 F2J0133	1	F2J0133A	2
34	Problems with digestive system, specify: _____ F2J0134S	0 F2J0134	1	F2J0134A	2
35	Rheumatoid arthritis	0 F2J0135	1	F2J0135A	2
36	Ankylosing spondylitis	0 F2J0136	1	F2J0136A	2
37	Arthritis, other	0 F2J0137	1	F2J0137A	2
38	Lupus	0 F2J0138	1	F2J0138A	2
39	Scleroderma	0 F2J0139	1	F2J0139A	2
40	Other autoimmune disease, specify type: _____ F2J0140S	0 F2J0140	1	F2J0140A	2
41	Joint dislocations, specify sites: _____ F2J0141S	0 F2J0141	1	F2J0141A	2
42	Other joint disease, specify: _____ F2J0142S	0 F2J0142	1	F2J0142A	2
43	Kidney disease	0 F2J0143	1	F2J0143A	2
44	Nerve disease, specify: _____ F2J0144S	0 F2J0144	1	F2J0144A	2
45	Memory loss/problems, specify: _____ F2J0145S	0 F2J0145	1	F2J0145A	2
46	Learning problems, specify: _____ F2J0146S	0 F2J0146	1	F2J0146A	2
47	Depression	0 F2J0147	1	F2J0147A	2
48	Other mental health concern, specify: _____ F2J0148S	0 F2J0148	1	F2J0148A	2
49	Other important health condition, specify: _____ F2J0149S	0 F2J0149	1	F2J0149A	2

2. Have you ever had surgery or been hospitalized for any therapeutic or medical procedure? This includes all vascular or heart surgeries or procedures, including those on your heart valves.
F2J02

0 No → **GO TO SECTION K**
 1 Yes

Type of surgery or procedure	Year or age at time of surgery or procedure
a. F2J02A	F2J02AYR F2J02AAG
b. F2J02B	F2J02BYR F2J02BAG
c. F2J02C	F2J02CYR F2J02CAG
d. F2J02D	F2J02DYR F2J02DAG
e. F2J02E	F2J02EYR F2J02EAG
f. F2J02F	F2J02FYR F2J02FAG
g. F2J02G	F2J02GYR F2J02GAG

SECTION K: PRESCRIPTION MEDICATIONS

1. These next questions are about prescription medications. Place an X in Column A if you've never taken the medication. Place an X in Column B if you are currently taking the medication. Place an X in Column C if you've used it in the past but are not using it now. If you don't know if you've use it, place an X in the last column. When thinking about medications you are taking now, include those that you usually take at home. **Do not include those you may have been given only during a recent hospitalization or emergency room visit.**

		<u>Column A</u> No, I've never taken this	<u>Column B</u> Yes, I'm taking this now	<u>Column C</u> Yes, I've taken this in the past but not now	<u>Column D</u> I don't know if I've taken this
A. ACE-inhibitors					
1. benazepril (Lotensin)	F2K1A01	1	2	3	4
2. captopril (Capoten)	F2K1A02	1	2	3	4
3. enalapril (Vasotec)	F2K1A03	1	2	3	4
4. fosinopril (Monopril)	F2K1A04	1	2	3	4
5. isinopril (Prinivil, Zestril)	F2K1A05	1	2	3	4
6. moexipril (Univasc)	F2K1A06	1	2	3	4
7. perindopril (Aceon)	F2K1A07	1	2	3	4
8. quinapril (Accupril)	F2K1A08	1	2	3	4
9. ramipril (Altace)	F2K1A09	1	2	3	4
10. randolapril (Mavik)	F2K1A10	1	2	3	4
11. Other _____	F2K1A11 F2K1A11S	1	2	3	4
12. Other _____	F2K1A12 F2K1A12S	1	2	3	4
B. Angiotensin Receptor Blockers					
1. candesartan (Atacand)	F2K1B01	1	2	3	4
2. eprosartan (Tevetan)	F2K1B02	1	2	3	4
3. irbesartan (Avapro)	F2K1B03	1	2	3	4
4. losartan (Cozaar)	F2K1B04	1	2	3	4
5. telmisartan (Mycardis)	F2K1B05	1	2	3	4
6. valsartan (Diovan)	F2K1B06	1	2	3	4
7. Other _____	F2K1B07 F2K1B07S	1	2	3	4
8. Other _____	F2K1B08 F2K1B08S	1	2	3	4
C. Beta Blockers					
1. acebutolol (Sectral)	F2K1C01	1	2	3	4
2. atenolol (Tenormin)	F2K1C02	1	2	3	4
3. betaxolol (Kerlone)	F2K1C03	1	2	3	4
4. bisoprolol fumarate (Zebeta)	F2K1C04	1	2	3	4
5. carteolol hydrochloride (Cartro)	F2K1C05	1	2	3	4
6. carvedilol (Coreg)	F2K1C06	1	2	3	4
7. labetalol (Trandate)	F2K1C07	1	2	3	4
8. metoprolol tartrate (Lopressor)	F2K1C08	1	2	3	4
9. metoprolol succinate (Toprol-XL)	F2K1C09	1	2	3	4
10. nadolol (Corgard)	F2K1C10	1	2	3	4
11. penbutolol sulfate (Levatol)	F2K1C11	1	2	3	4
12. pindolol (Visken)	F2K1C12	1	2	3	4

		<u>Column A</u> No, I've never taken this	<u>Column B</u> Yes, I'm taking this now	<u>Column C</u> Yes, I've taken this in the past but not now	<u>Column D</u> I don't know if I've taken this
13. propranolol hydrochloride (Inderal, Inderal LA)	F2K1C13	1	2	3	4
14. Other _____	F2K1C14 F2K1C14S	1	2	3	4
15. Other _____	F2K1C15 F2K1C15S	1	2	3	4
D. Diuretics					
1. bumetanide (Bumex)	F2K1D01	1	2	3	4
2. chlorothiazide or CTZ (Diuril)	F2K1D02	1	2	3	4
3. eplerone (Inspra)	F2K1D03	1	2	3	4
4. furosemide (Lasix)	F2K1D04	1	2	3	4
5. hydrochlorothiazide or HCTZ (Hydrodiuril)	F2K1D05	1	2	3	4
6. indapamide (Lozol)	F2K1D06	1	2	3	4
7. metolazone (Mykrox, Zaroxolyn)	F2K1D07	1	2	3	4
8. spironolactone (Aldactone)	F2K1D08	1	2	3	4
9. torsemide (Demadex)	F2K1D09	1	2	3	4
10. Other _____	F2K1D10 F2K1D10S	1	2	3	4
11. Other _____	F2K1D11 F2K1D11S	1	2	3	4
E. Calcium Channel Blockers					
1. amlodipine (Norvasc)	F2K1E01	1	2	3	4
2. bepridil (Vasacor)	F2K1E02	1	2	3	4
3. diltiazem (Cardizem)	F2K1E03	1	2	3	4
4. felodipine (Plendil)	F2K1E04	1	2	3	4
5. isradipine (Dynacirc)	F2K1E05	1	2	3	4
6. neifedipine (Adalat, Procardia)	F2K1E06	1	2	3	4
7. nicardipine (Cardene)	F2K1E07	1	2	3	4
8. nimodipine (Nimotop)	F2K1E08	1	2	3	4
9. nisoldipine (Sular)	F2K1E09	1	2	3	4
10. verapamil (Calan, Isoptin)	F2K1E10	1	2	3	4
11. Other _____	F2K1E11 F2K1E11S	1	2	3	4
12. Other _____	F2K1E12 F2K1E12S	1	2	3	4
F. Statins					
1. atorvastatin (Lipitor)	F2K1F01	1	2	3	4
2. fluvastatin (Lescol)	F2K1F02	1	2	3	4
3. lovastatin (Mevacor)	F2K1F03	1	2	3	4
4. pravastatin (Pravachol)	F2K1F04	1	2	3	4
5. rosuvastatin (Crestor)	F2K1F05	1	2	3	4
6. simvastatin (Zocor)	F2K1F06	1	2	3	4
7. Other _____	F2K1F07 F2K1F07	1	2	3	4

		<u>Column A</u> No, I've never taken this	<u>Column B</u> Yes, I'm taking this now	<u>Column C</u> Yes, I've taken this in the past but not now	<u>Column D</u> I don't know if I've taken this
8. Other _____	F2K1F08 F2K1F08S	1	2	3	4
G. Alpha Blockers					
1. alfuzosin (Uroxatral)	F2K1G01	1	2	3	4
2. doxazosin (Cardura)	F2K1G02	1	2	3	4
3. prazosin (Minipress)	F2K1G03	1	2	3	4
4. tamsulosin (Flomax)	F2K1G04	1	2	3	4
5. terazosin (Hytrin)	F2K1G05	1	2	3	4
6. Other _____	F2K1G06 F2K1G06S	1	2	3	4
7. Other _____	F2K1G07 F2K1G07S	1	2	3	4
H. Other types of medications					
1. clonidine (Catapres)	F2K1H01	1	2	3	4
2. clopidogrel (Plavex)	F2K1H02	1	2	3	4
3. enoxaparin (Lovenox)	F2K1H03	1	2	3	4
4. hydralizine (Hydra-Zide)	F2K1H04	1	2	3	4
5. Hypoglycemics, oral	F2K1H05	1	2	3	4
6. Insulin	F2K1H06	1	2	3	4
7. methyl dopa (Aldomet)	F2K1H07	1	2	3	4
8. methylphenidate (Ritalin)	F2K1H08	1	2	3	4
9. minoxidil (Loniten)	F2K1H09	1	2	3	4
10. Nitrates	F2K1H10	1	2	3	4
11. warfarin (Coumadin)	F2K1H11	1	2	3	4
12. Other _____	F2K1H12 F2K1H12S	1	2	3	4
13. Other _____	F2K1H13 F2K1H13S	1	2	3	4
I. Record any other prescription medications you have ever or are currently taking.					
1.	F2K1I01 F2K1I01S	1	2	3	4
2.	F2K1I02 F2K1I02S	1	2	3	4
3.	F2K1I03 F2K1I03S	1	2	3	4

2. Do you regularly take aspirin? F2K2

0 No → GO TO Q3
1 Yes

2a. On average, about how many aspirin tablets do you take each week? F2K2A

____ Number taken each week

3. Not including aspirin, do you regularly take any non-steroidal anti-inflammatory medications (NSAIDS)? This includes ibuprofen (Motrin), naproxen (Naprosyn), piroxicam (Feldene), and nabumetone (Relafen). **F2K3**

0 No → **GO TO Q4**
1 Yes

- 3a. On average, about how many NSAID tablets do you take each week? **F2K3A**

____ Number taken each week

4. Do you regularly take Celebrex? **F2K4**

0 No → **FORM IS COMPLETE**
1 Yes

- 4a. On average, about how many Celebrex tablets do you take each week? **F2K4A**

____ Number taken each week



GenTAC

FORM 2
ENROLLMENT PATIENT
QUESTIONNAIRE

Version 2.0 - 3/27/2009

This study has a clinical exemption under 24USC 285b and 285d

We are interested in learning more about people who are thought to have a genetic condition that may cause a problem with their thoracic aorta. As you complete this form, answer the questions as best as you can. If you don't know the answer or do not want to answer a question, you may leave it blank. If you are completing this form for a child or other dependent that is enrolled in this study, any references to "you" or "your" refer to that person.

Today's Date: |__|__|_|-|__|__|_|-|__|__|__|__|
Month Day Year
F2_MM F2_DD F2_YY

SECTION A: DEMOGRAPHIC DATA

1. What is your date of birth? |__|__|_|-|__|__|_|-|__|__|__|__|
Month Day Year
F2A01MM F2A01DD F2A01YY

2. What is your gender? F2A02
1 Male
2 Female

3. Which of the following groups describes your race? (*You may choose more than one*)
1 White F2A031
1 Black or African American F2A032
1 Asian F2A033
1 American Indian or Alaskan Native F2A034
1 Native Hawaiian or Other Pacific Islander F2A035

4. Are you of Hispanic or Latino origin? F2A04
0 No → GO TO Q5
1 Yes

4a. What is your country of origin? _____ F2A04SP

5. What is the highest level of schooling you have completed? (*Choose only one.*) F2A05
1 1-8 years
2 Some high school
3 High school graduate
4 GED (high school equivalency)
5 1-3 years vocational education beyond high school
6 Some college
7 College graduate
8 One or more years of graduate school or professional school
9 Something else (*Please describe below*)
_____ F2A05SP

6. What is your current marital status? **F2A06**
- 0 Not Applicable (subject is a child)
 - 1 Married or living as married (including living with a partner)
 - 2 Divorced or separated
 - 3 Widowed
 - 4 Never married
7. Are you covered by health insurance or some other kind of health care plan? **F2A07**
- 0 No → **GO TO Q8**
 - 1 Yes
- 7a. What kind of health insurance or health care coverage do you have? (*Choose only one.*) **F2A07A**
- 1 Private health insurance plan from employer
 - 2 Medicare
 - 3 Medicaid
 - 4 Other, specify _____ **F2A07ASP**
8. What is your approximate yearly household income? Include income from all sources. **F2A08**
- 1 \$25,000 and under
 - 2 \$25,001 - \$50,000
 - 3 \$50,001 - \$75,000
 - 4 \$75,001 - \$100,000
 - 5 \$100,001 or more
9. What is your primary current employment status? (*Choose only one.*) **F2A09**
- 10 Not Applicable (subject is a child)
 - 1 Employed full-time at a job or business
 - 2 Employed part-time at a job or business
 - 3 Self-employed
 - 4 Unable to work due to disability
 - 5 Student
 - 6 Homemaker
 - 7 Retired
 - 8 Unemployed
 - 9 Something else (*Please describe below*)
-
- F2A09A**

IF YOU ARE UNDER 18 YEARS OF AGE, CHECK THIS BOX AND GO TO SECTION E.

SECTION B: ALCOHOL USE

The next two questions ask about your general habits concerning alcoholic beverages. For these questions, **a drink is defined as one beer, a glass of wine, or a shot of hard liquor.**

1. During the past 12 months, how often did you usually drink any kind of alcoholic beverage? **F2B01**

- 1 Never → **GO TO SECTION C**
- 2 Less than one time a month
- 3 1-3 times a month
- 4 1 time a week
- 5 2-4 times a week
- 6 Almost every day
- 7 Every day

2. During the past 12 months, about how many drinks would you have on a day when you drank? **F2B02**

- 1 1 or 2 drinks
- 2 3 or 4 drinks
- 3 5-8 drinks
- 4 9 or more drinks

SECTION C: SMOKING HISTORY

1. During your lifetime, have you smoked at least 100 cigarettes? **F2C01**

- 0 No → **GO TO SECTION D**
- 1 Yes

2. Do you smoke cigarettes now? **F2C02**

- 0 No
- 1 Yes

3. On average, how much do you or did you smoke each day? (*1 pack = 20 cigarettes*) **F2C03**

- 1 10 cigarettes or less
- 2 11-20 cigarettes
- 3 21-40 cigarettes
- 4 More than 40 cigarettes

4. What is the total number of years you smoked cigarettes? (*Do not count years you did not smoke.*)

_____ Total number of years you smoked cigarettes **F2C04**

5. Other than cigarettes, which of the following tobacco products have you used on a regular basis for six months or longer? (*Choose all the tobacco products that you used.*)

- 1 Pipe F2C051
- 1 Cigars F2C052
- 1 Cigarillos F2C053
- 1 Chewing tobacco F2C054
- 1 Snuff F2C055
- 1 Never used any of these tobacco products for six months or longer F2C056

SECTION D: RECREATIONAL DRUG USE

The next questions are about certain drugs you might have used on a recreational basis. Place an X in Column A if you used the drug in the past 12 months. Place an X in Column B if you've used it but not in the past 12 months. Give only 1 answer for each drug, and remember that your answers will be kept strictly confidential.

	<u>Column A</u>	<u>Column B</u>
	Yes, I've used this drug in the past 12 months	Yes, I've used this drug but not in the past 12 months
1. Crack cocaine F2D01	1 <input type="checkbox"/>	2 <input type="checkbox"/>
2. Other types of cocaine, like powder F2D02	1 <input type="checkbox"/>	2 <input type="checkbox"/>
3. Stimulants, like amphetamines, methamphetamines, or ecstasy F2D03	1 <input type="checkbox"/>	2 <input type="checkbox"/>
4. 2 or more drugs taken together, like cocaine and heroin, specify: F2D12		
_____ F2D12SP	1 <input type="checkbox"/>	2 <input type="checkbox"/>

SECTION E: HEALTH AND WELL-BEING

1. In general, would you say your health is... F2E01

- 1 Excellent
- 2 Very good
- 3 Good
- 4 Fair
- 5 Poor

2. Compared to one year ago, how would you rate your health in general now? F2E02

- 1 Much better now than one year ago
- 2 Somewhat better now than one year ago
- 3 About the same as one year ago
- 4 Somewhat worse now than one year ago
- 5 Much worse now than one year ago

3. In the following table, record the number of hours you spend doing each activity in an average week.

Activity	# of hours per week
a. Strenuous activity such as aerobics, running, swimming, active sports, shoveling, lifting heavy objects, etc. F2E03A	
b. Moderate activity such as walking for exercise, cleaning house, gardening, carpentry, etc. F2E03B	
c. Slight activity , such as walking on level ground around the house, office, shopping, etc. F2E03C	

4. The following questions are about activities you might do during a typical day. Does your health now limit you in these activities? If so, how much? **If your activities are limited a lot**, please indicate whether they are restricted on the advice of your doctor.

Activity	No, not limited at all	Yes, limited a little	Yes, limited a lot →	Due to a doctor's recommendation?	
				Yes	No
Example: riding a bike			X		X
a. Vigorous activities, such as running, lifting heavy objects, participating in strenuous sports	1 F2E04A1	2	3	1 F2E04A2	0
b. Moderate activities, such as moving a table, pushing a vacuum cleaner, bowling, or playing golf	1 F2E04B1	2	3	1 F2E04B2	0
c. Lifting or carrying groceries	1 F2E04C1	2	3	1 F2E04C2	0
d. Climbing several flights of stairs	1 F2E04D1	2	3	1 F2E04D2	0
e. Climbing one flight of stairs	1 F2E04E1	2	3	1 F2E04E2	0
f. Bending, kneeling, or stooping	1 F2E04F1	2	3	1 F2E04F2	0
g. Walking more than a mile	1 F2E04G1	2	3	1 F2E04G2	0
h. Walking several hundred yards	1 F2E04H1	2	3	1 F2E04H2	0
i. Walking one hundred yards	1 F2E04I1	2	3	1 F2E04I2	0
j. Bathing or dressing yourself	1 F2E04J1	2	3	1 F2E04J2	0

5. During the past 4 weeks, how much of the time have you had any of the following problems with your work or other regular daily activities as a result of your physical health?

	All of the time	Most of the time	Some of the time	A little of the time	None of the time
a. Cut down on the <u>amount of time</u> you spent on work or other activities F2E05A	1	2	3	4	5
b. <u>Accomplished less than</u> you would like F2E05B	1	2	3	4	5
c. Were limited in the <u>kind of</u> work or other activities F2E05C	1	2	3	4	5
d. Had <u>difficulty</u> performing the work or other activities (for example, it took extra effort) F2E05D	1	2	3	4	5

6. How much bodily pain have you had during the past 4 weeks? **F2E06**

- 1 None
- 2 Very mild
- 3 Mild
- 4 Moderate
- 5 Severe
- 6 Very Severe

7. During the past 4 weeks, how much did pain interfere with your normal work (including both work outside the home and housework)? **F2E07**

- 1 Not at all
- 2 Slightly
- 3 Moderately
- 4 Quite a bit
- 5 A lot

8. How TRUE or FALSE is each of the following statements for you?

	Definitely true	Mostly true	Don't know	Mostly false	Definitely false
a. I seem to get sick a little easier than other people F2E08A	1	2	3	4	5
b. I am as healthy as anybody I know F2E08B	1	2	3	4	5
c. I expect my health to get worse F2E08C	1	2	3	4	5
d. My health is excellent F2E08D	1	2	3	4	5

SECTION F: HEARING/VISION

1. Have you ever worn a hearing aid? **F2G01**
 - 0** No
 - 1** Yes

2. Does a hearing problem cause you difficulty when listening to television or radio? **F2G02**
 - 0** No
 - 1** Yes

3. Do you wear eyeglasses or contact lenses? **F2G03**
 - 0** No
 - 1** Yes

4. How would you rate your vision, **without** eyeglasses or contact lenses if you wear them? **F2G04**
 - 1** Excellent
 - 2** Good
 - 3** Fair
 - 4** Poor

5. Have you ever had any surgical procedure to correct or improve your vision? **F2G05**
 - 1** No
 - 2** Yes, LASIK or laser correction
 - 3** Yes, lens extraction and replacement
 - 4** Yes, implanted contact lens
 - 5** Yes, cataract removal
 - 6** Yes, other

SECTION G: SURGERY

1. Have you ever had any of the vascular or heart surgeries or procedures listed below? This includes all types of heart surgeries, including those on your heart valves. **F2S01**
 - No → **GO TO SECTION H**
 - Yes

Please indicate the procedures you had by recording the year it was done. If you don't remember exactly when it was done, record your best guess.

Surgical Intervention	Year
Heart bypass or coronary bypass	F2S01A
Aortic root replacement	F2S01B
Aortic arch replacement	F2S01C
Ascending aortic replacement	F2S01D
Aortic valve repair (valve-replacing)	F2S01E
Aortic valve repair (non-root)	F2S01F
Valve-sparing aortic root replacement	F2S01G
Mitral valve repair or replacement	F2S01H
Pulmonary valve replacement	F2S01I
Pacemaker or defibrillator implantation	F2S01J

Descending thoracic aortic replacement	F2S01K
Thoracoabdominal aortic replacement	F2S01L
Aorto-femoral bypass surgery	F2S01M
Repair of pectus deformity	F2S01N
Angioplasty	F2S01O
Aorta to pulmonary shunt	F2S01P
Tetralogy repair	F2S01Q
Coarctation repair	F2S01R
Other heart surgery, specify: F2S01S_SP	F2S01S

SECTION H: MEDICAL CONDITIONS

1. Has a doctor ever told you that you had any of the following conditions? Place a check in the appropriate column (No, Yes or Don't Know). For each **YES** answer, please provide your age at the time of diagnosis or procedure.

Condition		No	Yes →	Age at Diagnosis	Don't Know
1	Marfan syndrome	0 F2J0101	1	F2J0101A	2
2	Turner syndrome	0 F2J0102	1	F2J0102A	2
3	Ehlers-Danlos syndrome	0 F2J0103	1	F2J0103A	2
4	Loeys-Dietz syndrome	0 F2J0104	1	F2J0104A	2
5	FBN1, TGFBR1 or TGFBR2 genetic mutations	0 F2J0105	1	F2J0105A	2
6	Bicuspid aortic valve	0 F2J0106	1	F2J0106A	2
6a	Coarctation	0 F2J0151	1	F2J0151A	2
7	Shprintzen-Goldberg syndrome	0 F2J0107	1	F2J0107A	2
7a	Thoracic aortic aneurysm or dissection	0 F2J0150	1	F2J0150A	2
8	Heart murmur	0 F2J0108	1	F2J0108A	2
9	Heart rhythm palpitations	0 F2J0109	1	F2J0109A	2
10	Angina	0 F2J0110	1	F2J0110A	2
11	Heart attack	0 F2J0111	1	F2J0111A	2
12	Atherosclerosis, clogged arteries	0 F2J0112	1	F2J0112A	2
13	Heart, blood vessel infection (endocarditis)	0 F2J0113	1	F2J0113A	2
14	Cardiomyopathy or heart failure	0 F2J0114	1	F2J0114A	2
15	Congenital heart disease, specify: _____ F2J0115S	0 F2J0115	1	F2J0115A	2

Condition		No	Yes →	Age at Diagnosis	Don't Know
16	Other heart disease, specify: _____ F2J0116S	0 F2J0116	1	F2J0116A	2
17	Hypertension, high blood pressure	0 F2J0117	1	F2J0117A	2
18	Stroke	0 F2J0118	1	F2J0118A	2
19	Aneurysms outside the aorta	0 F2J0119	1	F2J0119A	2
20	Asthma	0 F2J0120	1	F2J0120A	2
23	Pneumothorax or collapsed lung	0 F2J0123	1	F2J0123A	2
24	Other pulmonary or lung disease, specify: _____ F2J0124S	0 F2J0124	1	F2J0124A	2
25	Cancer, specify site: _____ F2J0125S	0 F2J0125	1	F2J0125A	2
27	Diabetes	0 F2J0127	1	F2J0127A	2
28	Bleeding disorder	0 F2J0128	1	F2J0128A	2
29	Blood clotting disorder	0 F2J0129	1	F2J0129A	2
32	Raynaud's syndrome (fingers turn purple when cold)	0 F2J0132	1	F2J0132A	2
34	Problems with digestive system, specify: _____ F2J0134S	0 F2J0134	1	F2J0134A	2
37	Arthritis, other	0 F2J0137	1	F2J0137A	2
39	Scleroderma	0 F2J0139	1	F2J0139A	2
40	Other autoimmune disease, specify type: _____ F2J0140S	0 F2J0140	1	F2J0140A	2
41	Joint dislocations, specify sites: _____ F2J0141S	0 F2J0141	1	F2J0141A	2
45	Memory loss/problems, specify: _____ F2J0145S	0 F2J0145	1	F2J0145A	2
46	Learning problems, specify: _____ F2J0146S	0 F2J0146	1	F2J0146A	2
47	Depression	0 F2J0147	1	F2J0147A	2

SECTION I: PRESCRIPTION MEDICATIONS

1. These next questions are about prescription medications. Place an X in Column A if you've never taken the medication, Column B if you are currently taking the medication, or Column C if you've used it in the past but are not using it now. If you don't know if you've use it, place an X in the last column. Include only those medications that you usually take at home. **Do not include those you may have been given only during a recent hospitalization or emergency room visit.**

<u>Column A</u>	<u>Column B</u>	<u>Column C</u>	<u>Column D</u>	<u>I don't know if I've taken this</u>
	No, I've never taken this	Yes, I've Yes, I'm taking this now	taken this in the past but not now	
a. ACE-inhibitors Such as: benazepril (Lotensin), captopril (Capoten), enalapril (Vasotec), fosinopril (Monopril), lisinopril (Prinivil, Zestril), moexipril (Univasc), perindopril (Aceon), quinapril (Accupril), ramipril (Altace), trandolapril (Mavik) F2K1A01	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
b. Angiotensin Receptor Blockers Such as: candesartan (Atacand), eprosartan (Tevetan), irbesartan (Avapro), losartan (Cozaar), telmisartan (Mycardis), valsartan (Diovan) F2K1A02	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
c. Beta Blockers Such as: acebutolol (Sectral), atenolol (Tenormin), betaxolol (Kerlone), bisoprolol fumarate (Zebeta), carteolol hydrochloride (Cartrol), carvedilol (Coreg), labetalol (Trandate), metoprolol tartrate (Lopressor), metoprolol succinate (Toprol-XL), nadolol (Corgard), penbutolol sulfate (Levatol), pindolol (Visken), propranolol hydrochloride (Inderal, Inderal LA) F2K1A03	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
d. Diuretics Such as: bumetanide (Bumex), chlorothiazide or CTZ (Diuril), eplerone (Inspra), furosemide (Lasix), hydrochlorothiazide or HCTZ (Hydrodiuril), indapamide (Lozol), metolazone (Mykrox, Zaroxolyn), spironolactone (Aldactone), torsemide (Demadex) F2K1A04	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
e. Calcium Channel Blockers Such as: amlodipine (Norvasc), bepridil (Vascor), diltiazem (Cardizem), felodipine (Plendil), isradipine (Dynacirc), neifedipine (Adalat, Procardia), nicardipine (Cardene), nimodipine (Nimotop), nisoldipine (Sular), verapamil (Calan, Isoptin) F2K1A05	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
f. Statins Such as: atorvastatin (Lipitor), fluvastatin (Lescol), lovastatin (Mevacor), pravastatin (Pravachol), rosuvastatin (Crestor), simvastatin (Zocor) F2K1A06	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
g. Alpha Blockers Such as: alfuzosin (Uroxatral), doxazosin (Cardura), prazosin (Minipress), tamsulosin (Flomax), terazosin (Hytrin) F2K1A07	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
h. Other Medications Insulin F2K1A08	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>
Warfarin (Coumadin) F2K1A09	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>

2. Do you regularly take aspirin? **F2K2**

- 0** No → **GO TO Q3**
- 1** Yes

2a. On average, about how many aspirin tablets do you take each week? **F2K2A**

_____ Number taken each week

IF YOU ARE A MALE, CHECK THIS BOX AND GO TO SECTION J. **MaleQ**

SECTION J: PREGNANCIES

The next questions are about any pregnancies you have had.

1. Have you ever been pregnant? **F2H01**

- 0** No → **GO TO SECTION J**
- 1** Yes

2. Are you currently pregnant? **F2H02**

- 0** No
- 1** Yes

3. How many times have you been pregnant? Please be sure to include any pregnancies that ended in a live birth, miscarriage, stillbirth, or abortion, as well as a tubal or molar pregnancy. **F2H03**

_____ Number of times you have been pregnant

As you answer the following questions, please think about each of your pregnancies. Start with your very first pregnancy, listing it in the column labeled “1st pregnancy”. From there, work forward until you have provided information about all of your pregnancies, ending with the most recent. If you have been pregnant more than 6 times, please contact the coordinator for additional forms on which to record this information.

	1 st pregnancy	2 nd pregnancy	3 rd pregnancy
4. In what month and year did this pregnancy end? (If currently pregnant, enter your due date.)	____/____ Month / Year F2H041M F2H041Y	____/____ Month / Year	____/____ Month / Year
5. Was this a pregnancy with multiples?	F2H05 0 <input type="checkbox"/> No 1 Yes → How many? ____ F2H05N	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> <input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____
6. What medicines or procedures did you have to help you get pregnant? <i>(Check all that apply)</i>	1 None F2H0611 1 Pills or shots F2H0612 1 Artificial insemination F2H0613 1 In vitro fertilization F2H0614 1 Donor egg F2H0615 1 Donor sperm F2H0616 1 Other F2H0617	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other
7. While you were pregnant, did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	F2H07 0 No 1 Yes → What condition? _____ F2H07SP	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____
8. What was the outcome of this pregnancy? <i>(If you were pregnant with multiples, check all that apply and enter the number of babies with that outcome.)</i>	1 Live birth #____ F2H0811 #____ F2H0811N 1 Still birth #____ F2H0812 #____ F2H0812N 1 Miscarriage #____ F2H0813 #____ F2H0813N 1 Abortion (elective, induced) #____ F2H0814 #____ F2H0814N 1 Tubal, ectopic #____ F2H0815 #____ F2H0815N 1 Other #____ F2H0816 #____ F2H086S #____ F2H0816N 1 Currently pregnant #____ F2H0817 #____ F2H0817N	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other #____ <input type="checkbox"/> Currently pregnant #____	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other #____ <input type="checkbox"/> Currently pregnant #____
9. After this pregnancy was over, did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	F2H09 0 No 1 Yes → What condition? _____ F2H09SP	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____ _____

The following are questions about your health during and after each of your pregnancies.

	1 st pregnancy	2 nd pregnancy	3 rd pregnancy
10. <u>While you were pregnant</u> , which of these conditions did you have?	<ul style="list-style-type: none"> 1 None F2H101 1 High blood pressure, toxemia F2H102 1 Maternal diabetes F2H103 1 Premature rupture of membranes F2H104 1 Change in aortic dimension F2H105 1 Aortic dissection F2H106 1 Aortic rupture F2H107 	<ul style="list-style-type: none"> <input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture 	<ul style="list-style-type: none"> <input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture
11. Were you on bed rest during this pregnancy?	<p>F2H11</p> <ul style="list-style-type: none"> 0 No 1 Yes→ Why? For how long? _____ F2H11SP 	<ul style="list-style-type: none"> <input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____ 	<ul style="list-style-type: none"> <input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____
12. <u>After this pregnancy</u> , which of these conditions did you have?	<ul style="list-style-type: none"> 1 None F2H121 1 Change in aortic dimension F2H122 1 Aortic dissection F2H123 1 Aortic rupture F2H124 	<ul style="list-style-type: none"> <input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture 	<ul style="list-style-type: none"> <input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture

	4th pregnancy	5th pregnancy	6th pregnancy
4. In what month and year did this pregnancy end? (If currently pregnant, enter your due date.)	_____/_____ Month / Year	_____/_____ Month / Year	_____/_____ Month / Year
5. Was this a pregnancy with multiples?	<input type="checkbox"/> <input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____	<input type="checkbox"/> <input type="checkbox"/> No <input type="checkbox"/> Yes → How many? ____
6. What medicines or procedures did you have to help you get pregnant? <i>(Check all that apply)</i>	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other	<input type="checkbox"/> None <input type="checkbox"/> Pills or shots <input type="checkbox"/> Artificial insemination <input type="checkbox"/> In vitro fertilization <input type="checkbox"/> Donor egg <input type="checkbox"/> Donor sperm <input type="checkbox"/> Other
7. <u>While you were pregnant</u> , did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____
8. What was the outcome of this pregnancy? <i>(If you were pregnant with multiples, check all that apply and enter the number of babies with that outcome.)</i>	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____	<input type="checkbox"/> Live birth #____ <input type="checkbox"/> Still birth #____ <input type="checkbox"/> Miscarriage #____ <input type="checkbox"/> Abortion (elective, induced) #____ <input type="checkbox"/> Tubal, ectopic #____ <input type="checkbox"/> Other _____ #____ <input type="checkbox"/> Currently pregnant #____
9. <u>After this pregnancy</u> was over, did a doctor tell you the baby had a birth defect, chromosome abnormality, or other serious medical problem?	<input type="checkbox"/> No <input type="checkbox"/> Yes → What condition? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ What condition? _____

The following are questions about your health during and after each of your pregnancies.

	4th pregnancy	5th pregnancy	6th pregnancy
10. <u>While you were pregnant</u> , which of these conditions did you have?	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> High blood pressure, toxemia <input type="checkbox"/> Maternal diabetes <input type="checkbox"/> Premature rupture of membranes <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture
11. Were you on bed rest during this pregnancy?	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____	<input type="checkbox"/> No <input type="checkbox"/> Yes→ Why? For how long? _____
12. <u>After this pregnancy</u> , which of these conditions did you have?	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture	<input type="checkbox"/> None <input type="checkbox"/> Change in aortic dimension <input type="checkbox"/> Aortic dissection <input type="checkbox"/> Aortic rupture



FAMILY HISTORY OF DISEASE – Form 3

Version 1.0, 10/01/2007

Subject ID Label

The GenTAC Registry is interested in collecting information about the families of enrolled subjects.

1. What is today's date? |__|__| |__|__| |__|__|__|__|
Month Day Year
F3A01MM F3A01DD F3A01YY
2. How many whole-blood brothers and sisters do/did you have?
These siblings have the same mother and father that you do.
____ whole brothers F3A02B 1 Don't know F3A02BD
____ whole sisters F3A02S 1 Don't know F3A02SD
3. How many paternal half-blood brothers and sisters do/did you have?
These siblings have the same father that you do.
____ half brothers, same father F3A03B 1 Don't know F3A03BD
____ half sisters, same father F3A03S 1 Don't know F3A03SD
4. How many maternal half-blood brothers and sisters do/did you have?
These siblings have the same mother that you do.
____ half brothers, same mother F3A04B 1 Don't know F3A04BD
____ half sisters, same mother F3A04S 1 Don't know F3A04SD
5. How many biological children have you had?
____ sons F3A05S
____ daughters F3A05D
6. Are you adopted? F3A06
0 NO
1 YES → On the following pages, please provide information only on blood relatives, if known. *If you do not know any of your biologic relatives, you do not need to complete this form.*

INSTRUCTIONS FOR COMPLETING THIS FORM

We are interested in knowing about your family's medical history and whether any of your family members have enrolled in the GenTAC Registry. The clinical centers that are currently enrolling patients into GenTAC include:

COR =	Weill Cornell Medical College in New York City
JHU =	Johns Hopkins Medical Center in Baltimore, Maryland
OHS =	Oregon Health & Science University in Portland
PEN =	University of Pennsylvania in Philadelphia
UTH =	University of Texas in Houston
BAY =	Baylor College of Medicine in Houston, Texas

We would like to know the medical history of your mother, father, grandparents, siblings and children. Please answer the questions about each of your family members listed at the top of each column. Collection of this information has been approved by the human subjects committee at each institution.

Place a check in the appropriate box to indicate diseases or conditions that have been diagnosed in the family members that are listed at the top of each column. For every "✓" mark you make, enter the approximate age of that relative at the time the condition was diagnosed. If you are unsure of the age at diagnosis, provide your best guess, and if you don't know, enter a question mark "?".

If a relative did not have any of the conditions listed, check the box at the bottom of the column to indicate **NONE OF THE ABOVE**.

If you have any questions about how to complete this form, please ask the study coordinator. The study coordinator can also give you additional pages for siblings or children if you need them. Give your completed form to the study coordinator when you are done.

**THANK YOU VERY MUCH FOR YOUR CONTRIBUTION TO THIS
IMPORTANT RESEARCH PROJECT!**

Blood Relative →	Mother	Maternal Grandmother	Maternal Grandfather
1. Enter the first name of each relative in the box F3B01	Removed from dataset		
2. Enter the relative's State of current residence (eg, NY) F3B02			
3. Circle the Gender of each sibling (M= Male F= Female) F3B03	2 F	2 F	1 M
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX) F3B04	Removed from dataset		
5. Place a √ in the box if the person is deceased F3B05			
6. Enter the relatives current age or age at death F3B06			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below F3B07			
8. How well do you know the medical history of each relative? F3B08	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

EXAMPLE: Diabetes

√

59

√

?

9	Marfan syndrome	F3B09	F3B09A				
10	Ehlers-Danlos syndrome	F3B10	F3B10A				
11	Loeys-Dietz syndrome	F3B11	F3B11A				
12	TGF beta (TGF-β) receptor or FBN1 mutation	F3B12	F3B12A				
13	Shprintzen-Goldberg Syndrome	F3B13	F3B13A				
14	Aortic aneurysm	F3B14	F3B14A				
15	Aortic dissection	F3B15	F3B15A				
16	Coarctation or narrowing of the aorta	F3B16	F3B16A				
17	Bicuspid aortic valve disease	F3B17	F3B17A				
18	Other valvular (heart valve) disease	F3B18	F3B18A				
19	Congenital heart disease, specify type in the box →	F3B19	F3B19A F3B19S				
20	Heart attack	F3B20	F3B20A				
21	Angina	F3B21	F3B21A				
22	Angioplasty of coronary artery	F3B22	F3B22A				
23	Bypass surgery	F3B23	F3B23A				
24	Hypertension, high blood pressure	F3B24	F3B24A				
25	Congestive heart failure	F3B25	F3B25A				
26	Other heart disease, specify type in the box to the right	F3B26	F3B26A F3B26S				
27	Varicose veins	F3B27	F3B27A				
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection	F3B28	F3B28A F3B28S F3B28L				
29	Stroke	F3B29	F3B29A				
30	Joint disease	F3B30	F3B30A				
31	Blood clotting or bleeding disorder	F3B31	F3B31A				
32	Renal or kidney disease	F3B32	F3B32A				

33	Cancer, specify the type in the box to the right	F3B33	F3B33D1 F3B33A1 F3B33D2 F3B33A2 F3B33D3 F3B33A3				
34	Developmental disability or mental retardation	F3B34	F3B34A				
35	Sudden or unexplained death	F3B35	F3B35A				
36	If NONE OF THE ABOVE, check the box →	F3B36	1	<input type="checkbox"/>		<input type="checkbox"/>	

Blood Relative →	Father	Paternal Grandmother	Paternal Grandfather
1. Enter the first name of each relative in the box F3B01			
2. Enter the relative's State of current residence (eg, NY) F3B02			
3. Circle the Gender of each sibling (M= Male F= Female) F3B03	1 M	2 F	1 M
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX) F3B04			
5. Place a √ in the box if the person is deceased F3B05			
6. Enter the relatives current age or age at death F3B06			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below F3B07			
8. How well do you know the medical history of each relative? F3B08	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

EXAMPLE: Diabetes

√ 79 √ ?

9	Marfan syndrome	F3B09	F3B09A				
10	Ehlers-Danlos syndrome	F3B10	F3B10A				
11	Loeys-Dietz syndrome	F3B11	F3B11A				
12	TGF beta (TGF-β) receptor or FBN1 mutation	F3B12	F3B12A				
13	Shprintzen-Goldberg Syndrome	F3B13	F3B13A				
14	Aortic aneurysm	F3B14	F3B14A				
15	Aortic dissection	F3B15	F3B15A				
16	Coarctation or narrowing of the aorta	F3B16	F3B16A				
17	Bicuspid aortic valve disease	F3B17	F3B17A				
18	Other valvular (heart valve) disease	F3B18	F3B18A				
19	Congenital heart disease, specify type in the box →	F3B19	F3B19A F3B19S				
20	Heart attack	F3B20	F3B20A				
21	Angina	F3B21	F3B21A				
22	Angioplasty of coronary artery	F3B22	F3B22A				
23	Bypass surgery	F3B23	F3B23A				
24	Hypertension, high blood pressure	F3B24	F3B24A				
25	Congestive heart failure	F3B25	F3B25A				
26	Other heart disease, specify type in the box to the right	F3B26	F3B26A F3B26S				
27	Varicose veins	F3B27	F3B27A				
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection	F3B28	F3B28A F3B28S F3B28L				
29	Stroke	F3B29	F3B29A				
30	Joint disease	F3B30	F3B30A				
31	Blood clotting or bleeding disorder	F3B31	F3B31A				
32	Renal or kidney disease	F3B32	F3B32A				

33	Cancer, specify the type in the box to the right	F3B33	F3B33D1 F3B33A1 F3B33D2 F3B33A2 F3B33D3 F3B33A3				
34	Developmental disability or mental retardation	F3B34	F3B34A				
35	Sudden or unexplained death	F3B35	F3B35A				
36	If NONE OF THE ABOVE, check the box →	F3B36	1	<input type="checkbox"/>		<input type="checkbox"/>	

Blood Relative →	Child 1	Child 2	Child 3
1. Enter the first name of each relative in the box			
2. Enter the relative's State of current residence (eg, NY)			
3. Circle the Gender of each child (M= Male F= Female)	1 M 2 F	1 M 2 F	1 M 2 F
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX)			
5. Place a √ in the box if the person is deceased			
6. Enter the relatives current age or age at death			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below			
8. How well do you know the medical history of each relative?	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

EXAMPLE: Diabetes

√

14

9	Marfan syndrome	F3B09	F3B09A			
10	Ehlers-Danlos syndrome	F3B10	F3B10A			
11	Loeys-Dietz syndrome	F3B11	F3B11A			
12	TGF beta (TGF-β) receptor or FBN1 mutation	F3B12	F3B12A			
13	Shprintzen-Goldberg Syndrome	F3B13	F3B13A			
14	Aortic aneurysm	F3B14	F3B14A			
15	Aortic dissection	F3B15	F3B15A			
16	Coarctation or narrowing of the aorta	F3B16	F3B16A			
17	Bicuspid aortic valve disease	F3B17	F3B17A			
18	Other valvular (heart valve) disease	F3B18	F3B18A			
19	Congenital heart disease, specify type in the box →	F3B19	F3B19A F3B19S			
20	Heart attack	F3B20	F3B20A			
21	Angina	F3B21	F3B21A			
22	Angioplasty of coronary artery	F3B22	F3B22A			
23	Bypass surgery	F3B23	F3B23A			
24	Hypertension, high blood pressure	F3B24	F3B24A			
25	Congestive heart failure	F3B25	F3B25A			
26	Other heart disease, specify type in the box to the right	F3B26	F3B26A F3B26S			
27	Varicose veins	F3B27	F3B27A			
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection	F3B28	F3B28A F3B28S F3B28L			
29	Stroke	F3B29	F3B29A			
30	Joint disease	F3B30	F3B30A			
31	Blood clotting or bleeding disorder	F3B31	F3B31A			
32	Renal or kidney disease	F3B32	F3B32A			

33	Cancer, specify the type in the box to the right	F3B33	F3B33D1 F3B33A1 F3B33D2 F3B33A2 F3B33D3 F3B33A3				
34	Developmental disability or mental retardation	F3B34	F3B34A				
35	Sudden or unexplained death	F3B35	F3B35A				
36	If NONE OF THE ABOVE, check the box →	F3B36	1	<input type="checkbox"/>		<input type="checkbox"/>	

Blood Relative →	Child 4	Child 5	Child 6
1. Enter the first name of each relative in the box			
2. Enter the relative's State of current residence (eg, NY)			
3. Circle the Gender of each child (M= Male F= Female)	1 M 2 F	1 M 2 F	1 M 2 F
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX)			
5. Place a √ in the box if the person is deceased			
6. Enter the relatives current age or age at death			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below			
8. How well do you know the medical history of each relative?	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

EXAMPLE: Diabetes

√ 14

9	Marfan syndrome				
10	Ehlers-Danlos syndrome				
11	Loeys-Dietz syndrome				
12	TGF beta (TGF-β) receptor or FBN1 mutation				
13	Shprintzen-Goldberg Syndrome				
14	Aortic aneurysm				
15	Aortic dissection				
16	Coarctation or narrowing of the aorta				
17	Bicuspid aortic valve disease				
18	Other valvular (heart valve) disease				
19	Congenital heart disease, specify type in the box →				
20	Heart attack				
21	Angina				
22	Angioplasty of coronary artery				
23	Bypass surgery				
24	Hypertension, high blood pressure				
25	Congestive heart failure				
26	Other heart disease, specify type in the box to the right				
27	Varicose veins				
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection				
29	Stroke				
30	Joint disease				
31	Blood clotting or bleeding disorder				
32	Renal or kidney disease				
33	Cancer, specify the type in the box to the right				
34	Developmental disability or mental retardation				
35	Sudden or unexplained death				
36	IF NONE OF THE ABOVE, check the box →	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Blood Relative →	Sibling 1	Sibling 2	Sibling 3
Check the box which indicates your sibling's relationship to you. F3BLD	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad
1. Enter the first name of each relative in the box F3B01			
2. Enter the relative's State of current residence (eg, NY) F3B02			
3. Circle the Gender of each sibling (M= Male F= Female) F3B03	1 M 2 F	1 M 2 F	1 M 2 F
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX) F3B04			
5. Place a √ in the box if the person is deceased F3B05			
6. Enter the relatives current age or age at death F3B06			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below F3B07			
8. How well do you know the medical history of each relative? F3B08	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

EXAMPLE: Diabetes

√

14

9	Marfan syndrome	F3B09	F3B09A				
10	Ehlers-Danlos syndrome	F3B10	F3B10A				
11	Loeys-Dietz syndrome	F3B11	F3B11A				
12	TGF beta (TGF-β) receptor or FBN1 mutation	F3B12	F3B12A				
13	Shprintzen-Goldberg Syndrome	F3B13	F3B13A				
14	Aortic aneurysm	F3B14	F3B14A				
15	Aortic dissection	F3B15	F3B15A				
16	Coarctation or narrowing of the aorta	F3B16	F3B16A				
17	Bicuspid aortic valve disease	F3B17	F3B17A				
18	Other valvular (heart valve) disease	F3B18	F3B18A				
19	Congenital heart disease, specify type in the box →	F3B19	F3B19A F3B19S				
20	Heart attack	F3B20	F3B20A				
21	Angina	F3B21	F3B21A				
22	Angioplasty of coronary artery	F3B22	F3B22A				
23	Bypass surgery	F3B23	F3B23A				
24	Hypertension, high blood pressure	F3B24	F3B24A				
25	Congestive heart failure	F3B25	F3B25A				
26	Other heart disease, specify type in the box to the right	F3B26	F3B26A F3B26S				
27	Varicose veins	F3B27	F3B27A				
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection	F3B28	F3B28A F3B28S F3B28L				
29	Stroke	F3B29	F3B29A				
30	Joint disease	F3B30	F3B30A				
31	Blood clotting or bleeding disorder	F3B31	F3B31A				
32	Renal or kidney disease	F3B32	F3B32A				

33	Cancer, specify the type in the box to the right	F3B33	F3B33D1 F3B33A1 F3B33D2 F3B33A2 F3B33D3 F3B33A3				
34	Developmental disability or mental retardation	F3B34	F3B34A				
35	Sudden or unexplained death	F3B35	F3B35A				
36	If NONE OF THE ABOVE, check the box →	F3B36	1	<input type="checkbox"/>		<input type="checkbox"/>	

Blood Relative →	Sibling 4	Sibling 5	Sibling 6
Check the box which indicates your sibling's relationship to you.	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad	1 whole sibling 2 half sibling, same Mom 3 half sibling, same Dad
1. Enter the first name of each relative in the box			
2. Enter the relative's State of current residence (eg, NY)			
3. Circle the Gender of each sibling (M= Male F= Female)	1 M 2 F	1 M 2 F	1 M 2 F
4. If this relative is enrolled in the GenTAC Registry, enter his/her center code from page 2 (e.g., TEX)			
5. Place a √ in the box if the person is deceased			
6. Enter the relatives current age or age at death			
7. If deceased, enter the cause of death in text or enter the numeric code from the list of conditions below			
8. How well do you know the medical history of each relative?	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all	1 Good 2 Fair 3 Not at all

INSTRUCTION: Place a √ for each condition ever present and enter age at diagnosis. If age is unknown, estimate or enter a "?"

9	Marfan syndrome				
10	Ehlers-Danlos syndrome				
11	Loeys-Dietz syndrome				
12	TGF beta (TGF-β) receptor or FBN1 mutation				
13	Shprintzen-Goldberg Syndrome				
14	Aortic aneurysm				
15	Aortic dissection				
16	Coarctation or narrowing of the aorta				
17	Bicuspid aortic valve disease				
18	Other valvular (heart valve) disease				
19	Congenital heart disease, specify type in the box →				
20	Heart attack				
21	Angina				
22	Angioplasty of coronary artery				
23	Bypass surgery				
24	Hypertension, high blood pressure				
25	Congestive heart failure				
26	Other heart disease, specify type in the box to the right				
27	Varicose veins				
28	Artery disease, specify location and type in the box For type, O= occlusive, A=aneurysm, D=dissection				
29	Stroke				
30	Joint disease				
31	Blood clotting or bleeding disorder				
32	Renal or kidney disease				
33	Cancer, specify the type in the box to the right				
34	Developmental disability or mental retardation				
35	Sudden or unexplained death				
36	If NONE OF THE ABOVE, check the box →	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	



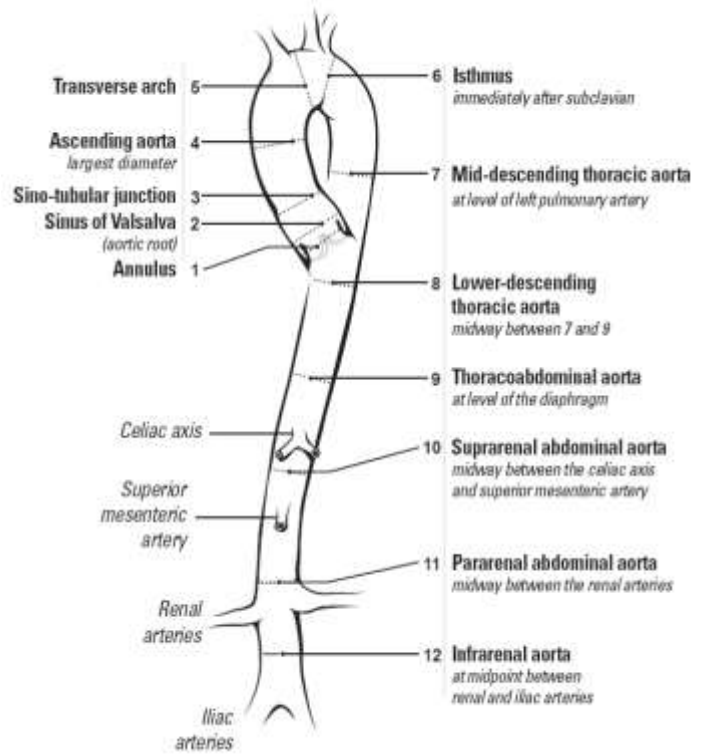
Imaging Evaluation - Form 4

Version 1.0, 10/01/2007

Subject ID Label

Complete this form for the most recent imaging study available for each modality.

	<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
1. Date performed	____/____/____ F4E01MM F4E01DD F4E01YY	____/____/____ F4C01MM F4C01DD F4C01YY	____/____/____ F4M01MM F4M01DD F4M01YY
2. Height	____ ____ cm F4E02	____ ____ cm F4C02	____ ____ cm F4M02
3. Weight	____ ____ .____ kg F4E03	____ ____ .____ kg F4C03	____ ____ .____ kg F4M03
4. Blood pressure	____/____ F4E04SY F4E04DS	____/____ F4C04SY F4C04DS	____/____ F4M04SY F4M04DS
5. Data source	1 Medical record F4E051 1 Imaging report F4E052 1 Hard <input type="checkbox"/> copy or digital images F4E053 1 Other F4E054 F4E054SP	1 Medical record F4C051 1 Imaging report F4C052 1 Hard <input type="checkbox"/> copy or digital images F4C053 1 Other F4C054 F4C054SP	1 Medical record F4M051 1 Imaging report F4M052 1 Hard <input type="checkbox"/> copy or digital images F4M053 1 Other F4M054 F4M054SP
6. Dimensions taken (ECHO only)	F4E06		
	1 Diastolic (leading edge to leading edge) 2 Systolic (inner edge to inner edge) 3 Unknown		



Using the diagram of the thoracic aorta for reference, complete the measurements requested in the table below.

Measurement	Reference	<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
7. Measurement Convention		1 TEE 2 TTE F4E07	1 Axial 2 Double oblique F4C07A	1 Axial 2 Double oblique F4M07A
			1 Non-contrast 2 Contrast F4C07B	1 MRI 2 MRA F4M07B
8. Aortic valve annulus (1)	From parasternal long axis	____ .____ cm F4E08	____ .____ cm F4C08	____ .____ cm F4M08
9. Sinus of Valsalva (aortic root) (2)	From parasternal long axis	____ .____ cm F4E09	____ .____ cm F4C09	____ .____ cm F4M09
10. Sino-tubular junction (3)	From parasternal long axis	____ .____ cm F4E10	____ .____ cm F4C10	____ .____ cm F4M10
11. Ascending aorta (largest) (4)	From parasternal or high parasternal long axis	____ .____ cm F4E11	____ .____ cm F4C11	____ .____ cm F4M11
12. Transverse arch (5)	From suprasternal notch	____ .____ cm F4E12	____ .____ cm F4C12	____ .____ cm F4M12
13. Isthmus (6)	From suprasternal notch	____ .____ cm F4E13	____ .____ cm F4C13	____ .____ cm F4M13
14. Mid-descending thoracic aorta (7)	From suprasternal notch or high parasternal long axis	____ .____ cm F4E14	____ .____ cm F4C14	____ .____ cm F4M14
15. Lower-descending thoracic aorta (8)	From parasternal or subxiphoid short axis	____ .____ cm F4E15	____ .____ cm F4C15	____ .____ cm F4M15

Measurement		Reference	<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
16.	Thoracoabdominal aorta (9)	From subxiphoid short axis	____.____ cm F4E16	____.____ cm F4C16	____.____ cm F4M16
17.	Suprarenal abdominal aorta (10)			____.____ cm F4C17	____.____ cm F4M17
18.	Pararenal abdominal aorta (11)	From maximal renal anterior-posterior	____.____ cm F4E18	____.____ cm F4C18	____.____ cm F4M18
19.	Infrarenal aorta (12)			____.____ cm F4C19	____.____ cm F4M19
20.	Coronary aneurysms	Right coronary artery	1 Present 2 Absent F4E20	1 Present 2 Absent F4C20	1 Present 2 Absent F4M20
21.		Left coronary artery	1 Present 2 Absent F4E21	1 Present 2 Absent F4C21	1 Present 2 Absent F4M21
22.	Coronary fistulae		1 Present 2 Absent F4E22		

Measurement	ECHO	CT	MRI
Using the Diagram: Enter the location (start and end), length and widest diameter in the column for the appropriate mode			
Evidence of Aortic Aneurysm			
23.	0 None → skip to Q27 1 1 2 2 3 3 F4E23	0 None → skip to Q27 1 1 2 2 3 3 F4C23	0 None → skip to Q27 1 1 2 2 3 3 F4M23
24.	<i>Aneurysm 1</i> Loc: Start ____ End ____ F4E24S F4E24E Length: ____ cm F4E24L Diameter: ____ cm F4E24D	<i>Aneurysm 1</i> Loc: Start ____ End ____ F4C24S F4C24E Length: ____ cm F4C24L Diameter: ____ cm F4C24D	<i>Aneurysm 1</i> Loc: Start ____ End ____ F4M24S F4M24E Length: ____ cm F4M24L Diameter: ____ cm F4M24D
25.	<i>Aneurysm 2</i> Loc: Start ____ End ____ F4E25S F4E25E Length: ____ cm F4E25L Diameter: ____ cm F4E25D	<i>Aneurysm 2</i> Loc: Start ____ End ____ F4C25S F4C25E Length: ____ cm F4C25L Diameter: ____ cm F4C25D	<i>Aneurysm 2</i> Loc: Start ____ End ____ F4M25S F4M25E Length: ____ cm F4M25L Diameter: ____ cm F4M25L
26.	<i>Aneurysm 3</i> Loc: Start ____ End ____ F4E26S F4E26E Length: ____ cm F4E26L Diameter: ____ cm F4E26D	<i>Aneurysm 3</i> Loc: Start ____ End ____ F4C26S F4C26E Length: ____ cm F4C26L Diameter: ____ cm F4C26D	<i>Aneurysm 3</i> Loc: Start ____ End ____ F4M26S F4M26E Length: ____ cm F4M26E Diameter: ____ cm F4M26D
Evidence of Aortic Dissection			
27.	0 None → skip to Q30 1 1 2 2 F4E27	0 None → skip to Q30 1 1 2 2 F4C27	0 None → skip to Q30 1 1 2 2 F4M27

28.	<i>Dissection 1</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E28S F4E28E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E28L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E28D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C28S F4C28E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C28L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C28D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M28S F4M28E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M28L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M28D
29.	<i>Dissection 2</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E29S F4E29E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E29L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E29D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C29S F4C29E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C29L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C29D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M29S F4M29E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M29L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M29D

Evidence of Aortic Graft

30.		0 None → skip to Q35 1 1 2 2 3 3 4 4 F4E30	0 None → skip to Q35 1 1 2 2 3 3 4 4 F4C30	0 None → skip to Q35 1 1 2 2 3 3 4 4 F4M30
31.	<i>Graft 1</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E31S F4E31E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E31L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E31D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C31S F4C31E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C31L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C31D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M31S F4M31E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M31L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M31D
32.	<i>Graft 2</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E32S F4E32S Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E32L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E32D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C32S F4C32E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C32L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C32D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M32S F4M32E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M32L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M32D
33.	<i>Graft 3</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E33S F4E33E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E33L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E33D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C33S F4C33E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C33L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C33D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M33S F4M33E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M33L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M33D
34.	<i>Graft 4</i>	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4E34S F4E34E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E34L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E34D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4C34S F4C34E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C34L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C34D	Loc: Start <input type="text"/> <input type="text"/> End <input type="text"/> <input type="text"/> F4M34S F4M34E Length: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M34L Diameter: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M34D

Measurement		Reference	ECHO	CT	MRI
Image Acquisition					
35.	Mitral valve annular diameter	From apical 4 chamber view	<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4E35	<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4C35	<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> cm F4M35
36.	Mitral regurgitation	Trivial = 1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E36	1 1 2 2 3 3 4 4 F4C36	1 1 2 2 3 3 4 4 F4M36

Measurement		Reference	ECHO	CT	MRI
37.	Mitral valve prolapse	Trivial =1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E37	1 1 2 2 3 3 4 4 F4C37	1 1 2 2 3 3 4 4 F4M37
38.	Tricuspid regurgitation	Trivial =1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E38	1 1 2 2 3 3 4 4 F4C38	1 1 2 2 3 3 4 4 F4M38
39.	Tricuspid valve diameter	From apical 4 chamber view	_ _ _ _ . _ cm F4E39	_ _ _ _ . _ cm F4C39	_ _ _ _ . _ cm F4M39
40.	Tricuspid regurgitation velocity	Peak Doppler velocity	_ _ _ _ . _ m/sec F4E40		
41.	Tricuspid valve prolapse	Trivial =1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E41	1 1 2 2 3 3 4 4 F4C41	1 1 2 2 3 3 4 4 F4M41
42.	Pulmonary valve annular diameter	From parasternal imaging, long or short	_ _ _ _ . _ cm F4E42	_ _ _ _ . _ cm F4C42	_ _ _ _ . _ cm F4M42
43.	Pulmonary regurgitation	Trivial =1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E43	1 1 2 2 3 3 4 4 F4C43	1 1 2 2 3 3 4 4 F4M43
44.	Pulmonary stenosis	Peak Doppler velocity	_ _ _ _ . _ m/sec F4E44		_ _ _ _ . _ m/sec F4M44
45.	Main pulmonary artery diameter	From parasternal short axis	_ _ _ _ . _ cm F4E45	_ _ _ _ . _ cm F4C45	_ _ _ _ . _ cm F4M45
46.	Right pulmonary artery diameter	From parasternal short axis	_ _ _ _ . _ cm F4E46	_ _ _ _ . _ cm F4C46	_ _ _ _ . _ cm F4M46
47.	Left pulmonary artery diameter	From parasternal short axis	_ _ _ _ . _ cm F4E47	_ _ _ _ . _ cm F4C47	_ _ _ _ . _ cm F4M47
48.	Branch pulmonary stenosis	<i>Check all that apply</i>	1 Left 1 Right F4E48L F4E48R	1 Left 1 Right F4C48L F4C48R	1 Left 1 Right F4M48L F4M48R
49.	Aortic valve morphology	Bi-commissural = 1 Unicommissural = 2 Fusion = 3	1 1 2 2 3 3 F4E49	1 1 2 2 3 3 F4C49	1 1 2 2 3 3 F4M49
50.	<i>Type of fusion</i>	Right-left = 1 Right-non = 2 Left-non = 3 Unicuspid/ Unicommissural = 4 Partial = 5	1 1 2 2 3 3 4 4 5 5 F4E50	1 1 2 2 3 3 4 4 5 5 F4C50	1 1 2 2 3 3 4 4 5 5 F4M50
51.	Aortic regurgitation	Trivial =1 Mild = 2 Moderate = 3 Severe = 4	1 1 2 2 3 3 4 4 F4E51	1 1 2 2 3 3 4 4 F4C51	1 1 2 2 3 3 4 4 F4M51
52.	Peak aortic gradient	Peak Doppler velocity	_ _ _ _ . _ m/sec F4E52		_ _ _ _ . _ m/sec F4M52
53.	Mean aortic gradient	Mean Doppler velocity	_ _ _ _ . _ m/sec F4E53		_ _ _ _ . _ m/sec F4M53
54.	Arch sidedness		1 Left 2 Right F4E54	1 Left 2 Right F4C54	1 Left 2 Right F4M54

55.	Ventricular Function: Quantitative Methodology		1 Parasternal short M-mode 2 Parasternal long M-mode 3 Simpson's biplane 4 Unknown F4E55	1 Quantitative Planimetry 2 Visual Estimate 3 Unknown F4C55	1 Quantitative Planimetry 2 Visual Estimate 3 Unknown F4M55
56.	Left ventricular end <u>diastolic</u> volume		____.____ mL F4E56	____.____ mL F4C56	____.____ mL F4M56
57.	Left ventricular end <u>systolic</u> volume		____.____ mL F4E57	____.____ mL F4C57	____.____ mL F4M57
58.	Left ventricular end <u>diastolic</u> dimension		____.____ cm F4E58	____.____ cm F4C58	____.____ cm F4M58
59.	Left ventricular end <u>systolic</u> dimension		____.____ cm F4E59	____.____ cm F4C59	____.____ cm F4M59
60.	Ejection fraction	Left ventricle	____.____ % F4E60	____.____ % F4C60	____.____ % F4M60
61.	Shortening fraction	Left ventricle	____.____ % F4E61	____.____ % F4C61	____.____ % F4M61
62.	Right ventricular function	Qualitative Normal = 1 Mildly depressed = 2 Moderately depressed = 3 Severely depressed = 4	1 1 2 2 3 3 4 4 F4E62	1 1 2 2 3 3 4 4 F4C62	1 1 2 2 3 3 4 4 F4M62
63.	Right ventricular end <u>diastolic</u> volume		____.____ mL F4E63	____.____ mL F4C63	____.____ mL F4M63
64.	Right ventricular end <u>systolic</u> volume		____.____ mL F4E64	____.____ mL F4C64	____.____ mL F4M64
65.	Ejection fraction	Right ventricle	____.____ % F4E65	____.____ % F4C65	____.____ % F4M65
66.	Pulmonary regurgitant fraction	Right ventricle	____.____ % F4E66	____.____ % F4C66	____.____ % F4M66
Cardiac Anomalies:					
67.	Patent Foramen Ovale (PFO)/ Atrial Septal Defect (ASD)		1 Yes 0 No F4E67	1 Yes 0 No F4C67	1 Yes 0 No F4M67
68.	Ventricular Septal Defect		1 Yes 0 No F4E68	1 Yes 0 No F4C68	1 Yes 0 No F4M68
69.	Subaortic membrane		1 Yes 0 No F4E69	1 Yes 0 No F4C69	1 Yes 0 No F4M69
70.	Aortic Coarctation		1 Yes 0 No F4E70	1 Yes 0 No F4C70	1 Yes 0 No F4M70
71.	Patent Ductus Arteriosus (PDA)		1 Yes 0 No F4E71	1 Yes 0 No F4C71	1 Yes 0 No F4M71
72.	Other cardiac anomaly: _____		1 Yes 0 No F4E72 F472SP	1 Yes 0 No F4C72 F472SP	1 Yes 0 No F4M72 F04M72SP
Supplemental Measurements			CT		MRI
73.	Arterial tortuosity (vessels)		1 Yes 0 No F4C73		1 Yes 0 No F4M73

74.	Aneurysms Located Outside the Aorta	<i>Check all that apply:</i> Cerebral=1; Carotid=2; Subclavian=3; Vertebral=4; Iliac=5; Renal=6; Infrarenal=7; Superior mesenteric=8; Inferior mesenteric=9; Pelvic=10; Femoral=11; None=12	F4C741 F4C742 F4C743 F4C744 F4C745 F4C746	F4C747 F4C748 F4C749 F4C7410 F4C7411 F4C7412	F4M741 F4M742 F4M743 F4M744 F4M745 F4M746	F4M747 F4M748 F4M749 F4M7410 F4M7411 F4M7412
75.	Dissections Located Outside the Aorta	<i>Check all that apply:</i> Cerebral=1; Carotid=2; Subclavian=3; Vertebral=4; Iliac=5; Renal=6; Infrarenal=7; Superior mesenteric=8; Inferior mesenteric=9; Pelvic=10; Femoral=11; None=12	F4C751 F4C752 F4C753 F4C754 F4C755 F4C756	F4C757 F4C758 F4C759 F4C7510 F4C7511 F4C7512	F4M751 F4M752 F4M753 F4M754 F4M755 F4M756	F4M757 F4M758 F4M759 F4M7510 F4M7511 F4M7512
76.	Scoliosis		1 Yes 0 No F4C76		1 Yes 0 No F4M76	
77.	Spondylolisthesis		1 Yes 0 No F4C77		1 Yes 0 No F4M77	
78.	Pectus deformity excavatum		1 Yes 0 No F4C78		1 Yes 0 No F4M78	
79.	Pectus deformity carinatum		1 Yes 0 No F4C79		1 Yes 0 No F4M79	
80.	Emphysema		1 Yes 0 No F4C80		1 Yes 0 No F4M80	
81.	Dural ectasia\Tarlof cyst		1 Yes 0 No F4C81		1 Yes 0 No F4M81	
82.	Pulmonary bullae		1 Yes 0 No F4C82		1 Yes 0 No F4M82	

Comments: _____ Initials of Abstractor: |__|_|_|_|
F4COMM F4INIT

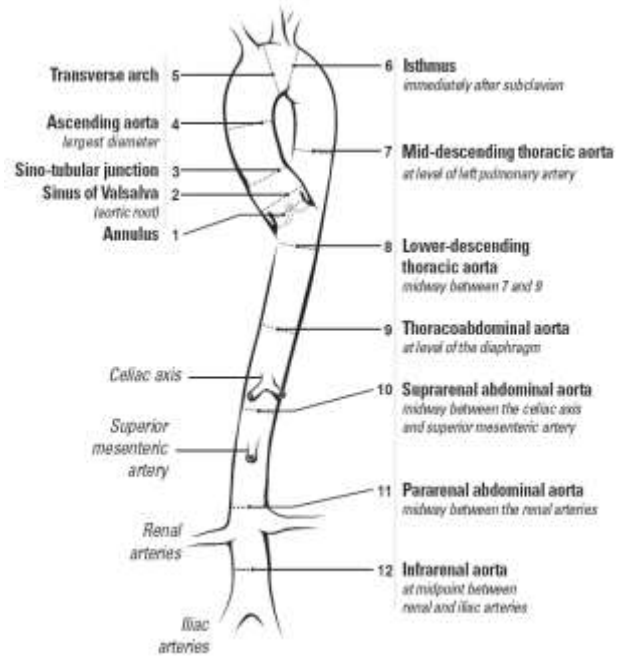
FORM COMPLETE	<ul style="list-style-type: none"> • KEY DATA ON-LINE AT http://gentac.rti.org • PUT FORM IN SUBJECT'S STUDY FILE.
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Subject ID

Complete this form for the most recent imaging study available for each modality.

	<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
1. Date test performed	____/____/____ F4E01MM F4E01DD F4E01YY	____/____/____ F4C01MM F4C01DD F4C01YY	____/____/____ F4M01MM F4M01DD F4M01YY
2. Height	____ cm F4E02	____ cm F4C02	____ cm F4M02
3. Weight	____.____ kg F4E03	____.____ kg F4C03	____.____ kg F4M03
4. Blood pressure	____/____ F4E04SY F4E04DS	____/____ F4C04SY F4C04DS	____/____ F4M04SY F4M04DS
1. Data source	1 Medical record F4E051 1 Imaging report F4E052 1 Direct review of image F4E053	1 Medical record F4C051 1 Imaging report F4C052 1 Direct review of image F4C053	1 Medical record F4M051 1 Imaging report F4M052 1 Direct review of image F4M053
5a. Are the actual images available for review and transmission to DCC?	<input type="checkbox"/> Yes, video <input type="checkbox"/> Yes, digital (DICOM) <input type="checkbox"/> No F4E05A	<input type="checkbox"/> Yes <input type="checkbox"/> No F4C05A	<input type="checkbox"/> Yes <input type="checkbox"/> No F4M05A
6. Dimensions taken (ECHO only)	<input type="checkbox"/> Diastolic (leading edge to leading edge) <input type="checkbox"/> Systolic (inner edge to inner edge) <input type="checkbox"/> Unknown F4E06		



Using the diagram of the thoracic aorta for reference, complete the measurements requested in the table below.

Measurement		Reference			<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
7. Measurement Convention			Check if evidence of:		1 <input type="checkbox"/> TEE 2 <input type="checkbox"/> TTE F4E07	1 <input type="checkbox"/> Axial 2 <input type="checkbox"/> Double oblique F4C07A	1 <input type="checkbox"/> Axial 2 <input type="checkbox"/> Double oblique F4m07A
			Dissection	Graft		1 <input type="checkbox"/> Non-contrast 2 <input type="checkbox"/> Contrast F4C07B	<input type="checkbox"/> MRI <input type="checkbox"/> MRA
8.	Aortic valve annulus (1)	From parasternal long axis	<input type="checkbox"/> F4E08A F4C08A F4M08A	<input type="checkbox"/> F4E08B F4C08B F4M08B	____.____ cm F4E08	____.____ cm F4C08	____.____ cm F4M08
9.	Sinus of Valsalva (aortic root) (2)	From parasternal long axis	<input type="checkbox"/> F4E09A F4C09A F4M09A	<input type="checkbox"/> F4E09B F4C09B F4M09B	____.____ cm F4E09	____.____ cm F4C09	____.____ cm F4M09
10.	Sino-tubular junction (3)	From parasternal long axis	<input type="checkbox"/> F4E10A F4C10A F4M10A	<input type="checkbox"/> F4E10B F4C10B F4M10B	____.____ cm F4E10	____.____ cm F4C10	____.____ cm F4M10
11.	Ascending aorta (largest) (4)	From parasternal or high parasternal long axis	<input type="checkbox"/> F4E11A F4C11A F4M11A	<input type="checkbox"/> F4E11B F4C11B F4M11B	____.____ cm F4E11	____.____ cm F4C11	____.____ cm F4M11

Measurement		Reference			<input type="checkbox"/> ECHO	<input type="checkbox"/> CT	<input type="checkbox"/> MRI
12.	Transverse arch (5)	From suprasternal notch	<input type="checkbox"/> F4E12A F4C12A F4M12A	<input type="checkbox"/> F4E12B F4C12B F4M12B	_ _ . _ cm F4E12	_ _ . _ cm F4C12	_ _ . _ cm F4M12
13.	Isthmus (6)	From suprasternal notch	<input type="checkbox"/> F4E13A F4C13A F4M13A	<input type="checkbox"/> F4E13B F4C13B F4M13B	_ _ . _ cm F4E13	_ _ . _ cm F4C13	_ _ . _ cm F4M13
14.	Mid-descending thoracic aorta (7)	From suprasternal notch or high parasternal long axis	<input type="checkbox"/> F4E14A F4C14A F4M14A	<input type="checkbox"/> F4E14B F4C14B F4M14B	_ _ . _ cm F4E14	_ _ . _ cm F4C14	_ _ . _ cm F4M14
15.	Lower-descending thoracic aorta (8)	From parasternal or subxiphoid short axis	<input type="checkbox"/> F4E15A F4C15A F4M15A	<input type="checkbox"/> F4E15B F4C15B F4M15B	_ _ . _ cm F4E15	_ _ . _ cm F4C15	_ _ . _ cm F4,15
16.	Thoracoabdominal aorta (9)	From subxiphoid short axis	<input type="checkbox"/> F4E16A F4C16A F4M16A	<input type="checkbox"/> F4E16B F4C16B F4M16B	_ _ . _ cm F4E16	_ _ . _ cm F4C16	_ _ . _ cm F4M16
17.	Suprarenal abdominal aorta (10)		<input type="checkbox"/> F4E17A F4C17A F4M17A	<input type="checkbox"/> F4E17B F4C17B F4M17B		_ _ . _ cm F4C17	_ _ . _ cm F417
18.	Pararenal abdominal aorta (11)	From maximal renal anterior-posterior	<input type="checkbox"/> F4E18A F4C18A F4M18A	<input type="checkbox"/> F4E18B F4C18B F4M18B	_ _ . _ cm F4E18	_ _ . _ cm F4C18	_ _ . _ cm F4M18
19.	Infrarenal aorta (12)		<input type="checkbox"/> F4E19A F4C19A F4M19A	<input type="checkbox"/> F4E19B F4C19B F4M19B		_ _ . _ cm F4C19	_ _ . _ cm F4M19
20.	Coronary aneurysms	Right coronary artery			1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4E20	1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4C20	1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4M20
21.		Left coronary artery			1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4E21	1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4C21	1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4M21
22.	Coronary fistulae				1 <input type="checkbox"/> Present 2 <input type="checkbox"/> Absent F4E22		

Measurement	ECHO	CT	MRI
Using the Diagram: Enter the location (start and end), length and widest diameter in the column for the appropriate mode			
Evidence of Aortic Aneurysm			
5.	Number of: 0 None → skip to Q27 1 1 2 2 3 3 F4E23	0 None → skip to Q27 1 1 2 2 3 3 F4C23	0 None → skip to Q27 1 1 2 2 3 3 F4M23
6.	Aneurysm 1 Loc: Start ___ End ___ F4E24S F4E24E Length: ___ . ___ cm F4E24L Diameter: ___ . ___ cm F4E24D	Aneurysm 1 Loc: Start ___ End ___ F4C24S F4C24E Length: ___ . ___ cm F4C24L Diameter: ___ . ___ cm F4C24D	Aneurysm 1 Loc: Start ___ End ___ F4M24S F4M24E Length: ___ . ___ cm F4M24L Diameter: ___ . ___ cm F4M24D
7.	Aneurysm 2 Loc: Start ___ End ___ F4E25S F4E25E Length: ___ . ___ cm F4E25L Diameter: ___ . ___ cm F4E25D	Aneurysm 2 Loc: Start ___ End ___ F4C25S F4C25E Length: ___ . ___ cm F4C25L Diameter: ___ . ___ cm F4C25D	Aneurysm 2 Loc: Start ___ End ___ F4M25S F4M25E Length: ___ . ___ cm F4M25L Diameter: ___ . ___ cm F4M25L
8.	Aneurysm 3 Loc: Start ___ End ___ F4E26S F4E26E Length: ___ . ___ cm F4E26L Diameter: ___ . ___ cm F4E26D	Aneurysm 3 Loc: Start ___ End ___ F4C26S F4C26E Length: ___ . ___ cm F4C26L Diameter: ___ . ___ cm F4C26D	Aneurysm 3 Loc: Start ___ End ___ F4M26S F4M26E Length: ___ . ___ cm F4M26E Diameter: ___ . ___ cm F4M26D

Measurement	Reference	ECHO	CT	MRI
34a.	Presence of prosthetic valves	<input type="checkbox"/> Aortic <input type="checkbox"/> Tricuspid <input type="checkbox"/> Mitral <input type="checkbox"/> Pulmonic F4E34A	<input type="checkbox"/> Aortic <input type="checkbox"/> Tricuspid <input type="checkbox"/> Mitral <input type="checkbox"/> Pulmonic F4C34A	<input type="checkbox"/> Aortic <input type="checkbox"/> Tricuspid <input type="checkbox"/> Mitral <input type="checkbox"/> Pulmonic F4M34A
35	Mitral valve annular diameter From apical 4 chamber view	___ . ___ cm F4E35	___ . ___ cm F4C35	___ . ___ cm F4M35
36	Mitral regurgitation None= 0 Trivial=1 Mild=2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E36		<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M36
37	Mitral valve prolapse None= 0 Trivial=1 Mild=2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E37	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4C37	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M37
38	Tricuspid regurgitation None= 0 Trivial=1 Mild=2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E38		<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M38
39	Tricuspid valve diameter From apical 4 chamber view	___ . ___ cm F4E39	___ . ___ cm F4C39	___ . ___ cm F4M39
40	Tricuspid regurgitation velocity Peak Doppler velocity	___ . ___ m/sec F4E40 mmHG		
41	Tricuspid valve prolapse None= 0 Trivial=1 Mild=2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E41	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4C41	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M41
42	Pulmonary valve annular diameter From parasternal imaging, long or short	___ . ___ cm F4E42	___ . ___ cm F4C42	___ . ___ cm F4M42
43	Pulmonary regurgitation None= 0 Trivial=1 Mild=2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E43		<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M43

44	Pulmonary stenosis	Peak Doppler velocity	_ _ _ . _ _ F4E44	m/sec mmHG		_ _ _ . _ _ F4M44	m/sec mmHG
45	Main pulmonary artery diameter	From parasternal short axis	_ _ _ . _ _ cm F4E45		_ _ _ . _ _ cm F4C45	_ _ _ . _ _ cm F4M45	
46	Right pulmonary artery diameter	From parasternal short axis	_ _ _ . _ _ cm F4E46		_ _ _ . _ _ cm F4C46	_ _ _ . _ _ cm F4M46	
47	Left pulmonary artery diameter	From parasternal short axis	_ _ _ . _ _ cm F4E47		_ _ _ . _ _ cm F4C47	_ _ _ . _ _ cm F4M47	
48	Branch pulmonary stenosis	<i>Check all that apply</i>	1 Left 1 Right F4E48L F4E48R		1 Left 1 Right F4C48L F4C48R	1 Left 1 Right F4M48L F4M48R	

Measurement		Reference	ECHO		CT		MRI	
49	Aortic valve morphology	Bi-commissural = 1 Unicommissural = 2 Normal/Unknown = 3	<input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 F4E49	<input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 F4C49	<input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 F4M49			
50	Type of fusion Bi-commissural only	Right-left = 1 Right-non = 2 Left-non = 3 Partial = 5	1 1 2 2 3 3 4 4 5 5 F4E50	1 1 2 2 3 3 4 4 5 5 F4C50	1 1 2 2 3 3 4 4 5 5 F4M50			
51	Aortic regurgitation	None=0 Trivial=1 Mild= 2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E51			<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M51		
51a.	Aortic stenosis	None=0 Trivial=1 Mild= 2 Moderate = 3 Severe = 4	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4E51A	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4C51A	<input type="checkbox"/> 0 <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4 F4M51A			
52.	Peak aortic gradient	Peak Doppler velocity	. . m/sec F4E52 mmHG			. . m/sec F4EM52 mmHG		
53.	Mean aortic gradient	Mean Doppler velocity	. . m/sec F4E53 mmHG			. . m/sec F4M53 mmHG		
54	Arch sidedness		1 Left 2 Right F4E54	1 Left 2 Right F4C54	1 Left 2 Right F4M54			
55	Ventricular Function: Quantitative Methodology		1 Parasternal short M-mode 2 Parasternal long M-mode 3 Simpson's biplane 4 Unknown F4E55	1 Quantitative Planimetry 2 Visual Estimate 3 Unknown F4C55	1 Quantitative Planimetry 2 Visual Estimate 3 Unknown F4M55			
56	Left ventricular end <u>diastolic</u> volume		. . mL F4E56 mL/m ²	. . mL F4C56 mL/m ²	. . mL F4M56 mL/m ²			
57	Left ventricular end <u>systolic</u> volume		. . mL F4E57 mL/m ²	. . mL F4C57 mL/m ²	. . mL F4M57 mL/m ²			
58	Left ventricular end <u>diastolic</u> dimension		. . cm F4E58	. . cm F4C58	. . cm F4M58			
59	Left ventricular end <u>systolic</u> dimension		. . cm F4E59	. . cm F4C59	. . cm F4M59			
60	Ejection fraction	Left ventricle	. . % F4E60	. . % F4C60	. . % F4M60			
61	Shortening fraction	Left ventricle	. . % F4E61	. . % F4C61	. . % F4M61			
62	Right ventricular function	Qualitative Normal =1 Mildly depressed = 2 Moderately depressed = 3 Severely depressed = 4	1 1 2 2 3 3 4 4 F4E62	1 1 2 2 3 3 4 4 F4C62	1 1 2 2 3 3 4 4 F4M62			
63	Right ventricular end <u>diastolic</u> volume		. . mL F4E63	. . mL F4C63	. . mL F4M63			
64	Right ventricular end <u>systolic</u> volume		. . mL F4E64	. . mL F4C64	. . mL F4M64			
65	Ejection fraction	Right ventricle	. . % F4E65	. . % F4C65	. . % F4M65			
66	Pulmonary regurgitant fraction	Right ventricle	. . % F4E66	. . % F4C66	. . % F4M66			
Cardiac Anomalies:								

67	Patent Foramen Ovale (PFO)/ Atrial Septal Defect (ASD)	1 Yes 0 No F4E67	1 Yes 0 No F4C67	1 Yes 0 No F4M67
68	Ventricular Septal Defect	1 Yes 0 No F4E68	1 Yes 0 No F4C68	1 Yes 0 No F4M68
69	Subaortic membrane	1 Yes 0 No F4E69	1 Yes 0 No F4C69	1 Yes 0 No F4M69
70	Aortic Coarctation	1 Yes 0 No F4E70	1 Yes 0 No F4C70	1 Yes 0 No F4M70
71	Patent Ductus Arteriosus (PDA)	1 Yes 0 No F4E71	1 Yes 0 No F4C71	1 Yes 0 No F4M71
72	a. Other cardiac anomaly: _____	1 Yes 0 No F4E72 F472SP	1 Yes 0 No F4C72 F472SP	1 Yes 0 No F4M72 F04M72SP
	b. Other cardiac anomaly: _____	1 Yes 0 No F4E67	1 Yes 0 No F4C67	1 Yes 0 No F4M67

Supplemental Measurements		CT	MRI
73	Arterial tortuosity (vessels)	1 Yes 0 No F4C73	1 Yes 0 No F4M73
74	Aneurysms Located Outside the Aorta <i>Check all that apply:</i> Cerebral=1; Carotid=2; Subclavian=3; Vertebral=4; Iliac=5; Renal=6; Infrarenal=7; Superior mesenteric=8; Inferior mesenteric=9; Pelvic=10; Femoral=11; None=12	F4C741 F4C747 F4C742 F4C748 F4C743 F4C749 F4C744 F4C7410 F4C745 F4C7411 F4C746 F4C7412	F4M741 F4M747 F4M742 F4M748 F4M743 F4M749 F4M744 F4M7410 F4M745 F4M7411 F4M746 F4M7412
75	Dissections Located Outside the Aorta <i>Check all that apply:</i> Cerebral=1; Carotid=2; Subclavian=3; Vertebral=4; Iliac=5; Renal=6; Infrarenal=7; Superior mesenteric=8; Inferior mesenteric=9; Pelvic=10; Femoral=11; None=12	F4C751 F4C757 F4C752 F4C758 F4C753 F4C759 F4C754 F4C7510 F4C755 F4C7511 F4C756 F4C7512	F4M751 F4M757 F4M752 F4M758 F4M753 F4M759 F4M754 F4M7510 F4M755 F4M7511 F4M756 F4M7512
76	Scoliosis	1 Yes 0 No F4C76	1 Yes 0 No F4M76
77	Spondylolisthesis	1 Yes 0 No F4C77	1 Yes 0 No F4M77
78	Pectus deformity excavatum	1 Yes 0 No F4C78	1 Yes 0 No F4M78
79	Pectus deformity carinatum	1 Yes 0 No F4C79	1 Yes 0 No F4M79
80	Emphysema	1 Yes 0 No F4C80	1 Yes 0 No F4M80
81	Dural ectasia\Tarlov cyst	1 Yes 0 No F4C81	1 Yes 0 No F4M81
82	Pulmonary bullae	1 Yes 0 No F4C82	1 Yes 0 No F4M82

Comments: _____ Initials of Abstractor: |_|_|_|_|

F4COMM

F4INIT



Non-Thoracic Imaging Evaluation - Form 5

Version 1.0, 10/01/2007

Subject ID: _____

Complete this form for the most recent non-thoracic imaging study available for each modality.

		<input type="checkbox"/> CT	<input type="checkbox"/> MRI
1.	Date performed	__ __ __ __ __ Month Day Year F5C01MM F5C01DD F5C01YY	__ __ __ __ __ Month Day Year F5M01MM F5M01DD F5M01YY
2.	Height	__ __ __ __ cm F5C02	__ __ __ __ cm F5M02
3.	Weight	__ __ __ __ kg F5C03	__ __ __ __ kg F5M03
4.	Blood pressure	__ __ / __ __ F5C04SY F5C04DS	__ __ / __ __ F05M04SY F05M04DS
5.	Data source	1 Medical record F5C051 1 Imaging report F5C052 1 Hard copy or digital images F5C053 1 Other F5C054 _____ F5C054SP	1 Medical record F5M051 1 Imaging report F5M052 1 Hard copy or digital images F5M053 1 Other F5M054 _____ F5M054SP
6.	Craniosynostosis: sutures and/or fontanelles (Check all that apply)	1 Coronal suture F5C061 1 Sagittal suture F5C062 1 Metopic suture F5C063 1 Anterolateral fontanelle F5C064 1 Postlateral fontanelle F5C065 1 Anterior fontanelle F5C066 1 None F5C067	1 Coronal suture F5M061 1 Sagittal suture F5M062 1 Metopic suture F5M063 1 Anterolateral fontanelle F5M064 1 Postlateral fontanelle F5M065 1 Anterior fontanelle F5M066 1 None F5M067
7.	Arnold-Chiari malformation	1 Yes 0 No → go to Q9 F5C07	1 Yes 0 No → go to Q9 F5M07
8.	Arnold-Chiari (mm of displacement below foramen magnum)	__ __ __ __ mm F5C08	__ __ __ __ mm F5M08
9.	Hydrocephalus	1 Yes 0 No F5C09	1 Yes 0 No F5M09
10.	Congenital cervical spine abnormalities	1 Yes 0 No F5C10	1 Yes 0 No F5M10
11.	Dural ectasia/Tarlov cyst	1 Yes 0 No F5C11	1 Yes 0 No F5M11

Comments: _____

F5COMM

Initials of Abstractor: |__| |__| |__|

F5INIT

FORM COMPLETE	<ul style="list-style-type: none"> KEY DATA ON-LINE AT http://gentac.rti.org PUT FORM IN SUBJECT'S STUDY FILE.
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Genetics and Blood Chemistry – Form 6

Enrollment

Version 1.0, 10/01/2007

Subject ID Label

Section A. Genetics

1. Was **karyotyping** done on this patient?

F6A1KT

0 No

1 Yes → Results: _____
F6A1RS

Date: |__|_| / |__|_|_|_|_|
Month Year
F6A1MM F6A1YY

→ Resolution: _____
F6A1RN

2. Was **FISH** analysis done on this patient?

F6A2FS

0 No

1 Yes → Results: _____
F6A2RS

Date: |__|_| / |__|_|_|_|_|
Month Year
F6A2MM F6A2YY

→ Probe: _____
F6A2PB

3. Was **genetic testing** done on this patient?

F6A3GT

0 No

1 Yes → Complete the table below for the appropriate gene tested, and specify any mutation identified. If any information is unknown, please leave the column blank.

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/ Location Test Conducted	Month & Year Tested
	Positive	Negative	Not Done		Familial	De Novo	Unknown			
a. FBN1	1 F6A3ARS	2	3	_____ F6A3AMT	1 F6A3AMD	2	3	1 Clinical 2 Research F6A3ATT	_____ F6A3AOR	_ _ / _ _ _ _ F6A3AMM F6A3AYY
b. FBN2	1 F6A3BRS	2	3	_____ F6A3BMT	1 F6A3BMD	2	3	1 Clinical 2 Research F6A3BTT	_____ F6A3BOR	_ _ / _ _ _ _ F6A3BMM F6A3BY Y
c. TGFBR1	1 F6A3CRS	2	3	_____ F6A3CMT	1 F6A3CMD	2	3	1 Clinical 2 Research F6A3CTT	_____ F6A3COR	_ _ / _ _ _ _ F6A3CMM F6A3CYY
d. TGFBR2	1 F6A3DRS	2	3	_____ F6A3DMT	1 F6A3DMD	2	3	1 Clinical 2 Research F6A3DTT	_____ F6A3DOR	_ _ / _ _ _ _ F6A3DMM F6A3DY Y
e. COL5A1	1 F6A3ERS	2	3	_____ F6A3EMT	1 F6A3EMD	2	3	1 Clinical 2 Research F6A3ETT	_____ F6A3EOR	_ _ / _ _ _ _ F6A3EMM F6A3EYY

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/L ocation Test Conducted	Month & Year Tested
	Positive <i>Positive</i>	Negative <i>Negative</i>	Not Done <i>Not Done</i>		Familial <i>Familial</i>	De Novo <i>De Novo</i>	Unknown <i>Unknown</i>			
f. COL5A2	1 F6A3FRS	2	3	_____ F6A3FMT	1 F6A3FMD	2	3	1 Clinical 2 Research F6A3FTT	_____ F6A3FOR	____/____ F6A3FMM F6A3FYY
g. COL3A1	1 F6A3GRS	2	3	_____ F6A3GMT	1 F6A3GMD	2	3	1 Clinical 2 Research F6A3GTT	_____ F6A3GOR	____/____ F6A3GMM F6A3GY Y
h. Tenascin X	1 F6A3HRS	2	3	_____ F6A3HMT	1 F6A3HMD	2	3	1 Clinical 2 Research F6A3HTT	_____ F6A3HOR	____/____ F6A3HMM F6A3HY Y
j. Other, specify: _____ _____	1	2	3	Specify mutation: _____ _____	1	2	3	1 Clinical 2 Research F6A3JTT	_____ F6A3JOR	____/____ F6A3JMM F6A3JYY
F6A3JSP	F6A3JRS			F6A3JMU	F6A3JMD					F6A3JMM F6A3JYY

Section B. Blood Chemistry

Test Name	Units	Date of Most Recent
1. Hemoglobin	_ _ _ _ . _ _ g/dL F6B1HG	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B1MM F6B1DD F6B1YY
2. Hematocrit	_ _ _ _ . _ _ % F6B2HT	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B2MM F6B2DD F6B2YY
3. Serum Creatinine	_ _ _ _ . _ _ mg/dL F6B3SC	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B3MM F6B3DD F6B3YY
4. Total Cholesterol	_ _ _ _ _ mg/dL F6B4TC	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B4MM F6B4DD F6B4YY
5. Non-Fasting HDL	_ _ _ _ _ mg/dL F6B5NH	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B5MM F6B5DD F6B5DD
6. Fasting HDL	_ _ _ _ _ mg/dL F6B6FH	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B6MM F6B6DD F6B6YY
7. Non-Fasting LDL	_ _ _ _ _ mg/dL F6B7NL	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B7MM F6B7DD F6B7YY
8. Fasting LDL	_ _ _ _ _ mg/dL F6B8FL	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B8MM F6B8DD F6B8YY
9. Non-Fasting Blood Glucose	_ _ _ _ _ mg/dL F6B9NG	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B9MM F6B9DD F6B9YY
10. Fasting Blood Glucose	_ _ _ _ . _ _ mg/dL F6B10FG	_ _ _ / _ _ _ / _ _ _ _ _ Month Day Year F6B10MM F6B10DD F6B10YY

Comments: _____ F6COMM

Initials of Abstractor: |_|_|_|_|_| F6INIT

FORM COMPLETE

- KEY DATA ON-LINE AT <http://gentac.rti.org>
- PUT FORM IN SUBJECT'S STUDY FILE.



Genetics and Blood Chemistry – Form 6

Enrollment

Version 1.1, 6/30/2008

Subject ID Label

Section A. Genetics

1. Was **karyotyping** done on this patient?

F6A1KT

0 No

1 Yes → Results: _____
F6A1RS

Date: |__|_| / |__|_|_|_|

Month Year
F6A1MM F6A1YY

→ Resolution: _____
F6A1RN

2. Was **FISH** analysis done on this patient?

F6A2FS

0 No

1 Yes → Results: _____
F6A2RS

Date: |__|_| / |__|_|_|_|

Month Year
F6A2MM F6A2YY

→ Probe: _____
F6A2PB

3. Was **genetic testing** done on this patient?

F6A3GT

0 No

1 Yes → Complete the table below for the appropriate gene tested, and specify any mutation identified. If any information is unknown, please leave the column blank.

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/ Location Test Conducted	Month & Year Tested
	Positive	Negative	Not Done		Familial	De Novo	Unknown			
a. FBN1	1 F6A3ARS	2	3	_____ F6A3AMT	1 F6A3AMD	2	3	1 Clinical 2 Research F6A3ATT	_____ F6A3AOR	_ _ / _ _ _ _ F6A3AMM F6A3AYY
b. FBN2	1 F6A3BRS	2	3	_____ F6A3BMT	1 F6A3BMD	2	3	1 Clinical 2 Research F6A3BTT	_____ F6A3BOR	_ _ / _ _ _ _ F6A3BMM F6A3BY Y
c. TGFBR1	1 F6A3CRS	2	3	_____ F6A3CMT	1 F6A3CMD	2	3	1 Clinical 2 Research F6A3CTT	_____ F6A3COR	_ _ / _ _ _ _ F6A3CMM F6A3CYY
d. TGFBR2	1 F6A3DRS	2	3	_____ F6A3DMT	1 F6A3DMD	2	3	1 Clinical 2 Research F6A3DTT	_____ F6A3DOR	_ _ / _ _ _ _ F6A3DMM F6A3DY Y
e. COL5A1	1 F6A3ERS	2	3	_____ F6A3EMT	1 F6A3EMD	2	3	1 Clinical 2 Research F6A3ETT	_____ F6A3EOR	_ _ / _ _ _ _ F6A3EMM F6A3EYY

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/L ocation Test Conducted	Month & Year Tested
	Positive	Negative	Not Done		Familial	De Novo	Unknown			
f. COL5A2	1 F6A3FRS	2	3	_____	1 F6A3FMD	2	3	1 Clinical 2 Research F6A3FTT	_____	____/____ F6A3FMM F6A3FYY
g. COL3A1	1 F6A3GRS	2	3	_____	1 F6A3GMD	2	3	1 Clinical 2 Research F6A3GTT	_____	____/____ F6A3GMM F6A3GY
h. Tenascin X	1 F6A3HRS	2	3	_____	1 F6A3HMD	2	3	1 Clinical 2 Research F6A3HTT	_____	____/____ F6A3HMM F6A3HY
i. ACTA2	1 F6A3IRS	2	3	_____	1 F6A3IMD	2	3	1 Clinical 2 Research F6A3ITT	_____	____/____ F6A3IMM F6A3IYY
j. MYH11	1	2	3	_____	1	2	3	1 Clinical 2 Research	_____	____/____
	F6A3JRS			F6A3JMT	F6A3JMD			F6A3JTT	F6A3JOR	F6A3JMM F6A3JYY
z. Other, specify: _____ _____	1	2	3	Specify mutation: _____ _____	1	2	3	1 Clinical 2 Research	_____	____/____ F6A3ZMM F6A3ZYY
F6A3ZSP	F6A3ZRS			F6A3ZMU	F6A3ZMD			F6A3ZTT	F6A3ZOR	

Section B. Blood Chemistry

Test Name	Units	Date of Most Recent
1. Hemoglobin	_____ _____ ._____ g/dL F6B1HG	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B1MM F6B1DD F6B1YY
2. Hematocrit	_____ _____ ._____ % F6B2HT	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B2MM F6B2DD F6B2YY
3. Serum Creatinine	_____ _____ ._____ mg/dL F6B3SC	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B3MM F6B3DD F6B3YY
4. Total Cholesterol	_____ _____ _____ mg/dL F6B4TC	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B4MM F6B4DD F6B4YY
5. Non-Fasting HDL	_____ _____ _____ mg/dL F6B5NH	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B5MM F6B5DD F6B5DD
6. Fasting HDL	_____ _____ _____ mg/dL F6B6FH	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B6MM F6B6DD F6B6YY
7. Non-Fasting LDL	_____ _____ _____ mg/dL F6B7NL	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B7MM F6B7DD F6B7YY
8. Fasting LDL	_____ _____ _____ mg/dL F6B8FL	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B8MM F6B8DD F6B8YY
9. Non-Fasting Blood Glucose	_____ _____ _____ mg/dL F6B9NG	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B9MM F6B9DD F6B9YY
10. Fasting Blood Glucose	_____ _____ ._____ mg/dL F6B10FG	_____ _____ / ____ ____ / ____ ____ ____ Month Day Year F6B10MM F6B10DD F6B10YY

Comments: _____ F6COMM

Initials of Abstractor: _____ F6INIT

FORM COMPLETE

- KEY DATA ON-LINE AT <http://gentac.rti.org>
- PUT FORM IN SUBJECT'S STUDY FILE.



Genetics – Form 6

Version 2.0, 3/27/2009

Subject ID Label

Section A. Genetics

1. Was **karyotyping** done on this patient?

F6A1KT

0 No

1 Yes → Results: _____
F6A1RS

Date: |__|_| / |__|_| / |__|_|

Month Year
F6A1MM F6A1YY

→ Resolution: _____
F6A1RN

2. Was **FISH** analysis done on this patient?

F6A2FS

0 No

1 Yes → Results: _____
F6A2RS

Date: |__|_| / |__|_| / |__|_|

Month Year
F6A2MM F6A2YY

→ Probe: _____
F6A2PB

3. Was **genetic testing** done on this patient?

F6A3GT

0 No

1 Yes → Complete the table below for the appropriate gene tested, and specify any mutation identified. If any information is unknown, please leave the column blank.

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/ Location Test Conducted	Month & Year Tested
	Positive	Negative	Not Done		Familial	De Novo	Unknown			
a. FBN1	1 F6A3ARS	2	3	_____ F6A3AMT	1 F6A3AMD	2	3	1 Clinical 2 Research F6A3ATT	_____ F6A3AOR	_ _ / _ _ _ _ F6A3AMM F6A3AYY
b. FBN2	1 F6A3BRS	2	3	_____ F6A3BMT	1 F6A3BMD	2	3	1 Clinical 2 Research F6A3BTT	_____ F6A3BOR	_ _ / _ _ _ _ F6A3BMM F6A3BY Y
c. TGFBR1	1 F6A3CRS	2	3	_____ F6A3CMT	1 F6A3CMD	2	3	1 Clinical 2 Research F6A3CTT	_____ F6A3COR	_ _ / _ _ _ _ F6A3CMM F6A3CYY
d. TGFBR2	1 F6A3DRS	2	3	_____ F6A3DMT	1 F6A3DMD	2	3	1 Clinical 2 Research F6A3DTT	_____ F6A3DOR	_ _ / _ _ _ _ F6A3DMM F6A3DYY
e. COL5A1	1 F6A3ERS	2	3	_____ F6A3EMT	1 F6A3EMD	2	3	1 Clinical 2 Research F6A3ETT	_____ F6A3EOR	_ _ / _ _ _ _ F6A3EMM F6A3EYY

Gene/Protein	Result (Check One):			Mutation	Mode (Check One):			Test Type	Organization/ Location Test Conducted	Month & Year Tested
	Positive	Negative	Not Done		Familial	De Novo	Unknown			
f. COL5A2	1 F6A3FRS	2	3	_____	1 F6A3FMD	2	3	1 Clinical 2 Research F6A3FTT	_____	____/____ F6A3FMM F6A3FYY
g. COL3A1	1 F6A3GRS	2	3	_____	1 F6A3GMD	2	3	1 Clinical 2 Research F6A3GTT	_____	____/____ F6A3GMM F6A3GY
h. Tenascin X	1 F6A3HRS	2	3	_____	1 F6A3HMD	2	3	1 Clinical 2 Research F6A3HTT	_____	____/____ F6A3HMM F6A3HY
i. ACTA2	1 F6A3IRS	2	3	_____	1 F6A3IMD	2	3	1 Clinical 2 Research F6A3ITT	_____	____/____ F6A3IMM F6A3IYY
j. MYH11	1 F6A3JRS	2	3	_____	1 F6A3JMD	2	3	1 Clinical 2 Research F6A3JTT	_____	____/____ F6A3JMM F6A3JYY
z. Other, specify: _____ _____	1 F6A3ZRS	2	3	Specify mutation: _____ _____	1 F6A3ZMD	2	3	1 Clinical 2 Research F6A3ZTT	_____	____/____ F6A3ZMM F6A3ZYY

Comments: _____ F6COMM

Initials of Abstractor: _____ F6INIT



Surgical and Percutaneous Interventions Form – Form 7

Version 1.1, 6/30/2008

Use this form to document any surgical or percutaneous interventions on the heart, aorta, or peripheral arteries that this subject has had. This includes interventions performed prior to and since enrollment in GenTAC. Complete one form for each intervention.

SECTION A. General Information

1. On what date was the procedure performed? |__|_|_|-|__|_|_|-|__|_|_|_|_|_|
F7A1MM F7A1DD F7A1YY

2. What type of procedure(s) was performed? (*Check all that apply. Information recorded here should correspond with Form 1, Q22.*)

Surgical Intervention	Yes
F7A2A a. Coronary artery bypass or PTCA grafting	1
F7A2B b. Aortic valve repair	1
F7A2C c. Isolated aortic valve replacement (non-root)	1
F7A2D d. Aortic root replacement (valve-replacing)	1
F7A2E e. Valve-sparing aortic root replacement	1
F7A2F f. Ascending aortic replacement	1
F7A2G g. Aortic arch replacement	1
F7A2H h. Mitral valve repair or replacement	1
F7A2I i. Coarctation repair	1
F7A2J j. Pacemaker/ICD implantation	1

Surgical Intervention	Yes
F7A2K k. Descending thoracic aortic replacement	1
F7A2L l. Thoracoabdominal aortic replacement	1
F7A2M m. Open arterial bypass	1
F7A2N n. Repair of pectus deformity	1
F7A2O o. Percutaneous/endovascular intervention, non-coronary PTCA	1
F7A2P p. Aorta to pulmonary shunt	1
F7A2Q q. TOF repair	1
F7A2R r. Pulmonary valve replacement	1
F7A2S s. Tricuspid valve surgery	1
F7A2T t. Other, specify _____ F7A2TS	1

3. Where was the procedure performed? Name of institution: _____ **F7A31**
 City: ___ **F7A32** State: ___ **F7A33** Country: ___ **F7A34**

4. What sources of information were used to complete this form? (*Check all that apply*)

1 History from patient/family **F7A41** 1 Operative report **F7A42**

1 Discharge summary **F7A43** 1 Other medical records **F7A44**

5. Was a tissue sample obtained during this procedure? **F7A5**

0 No

1 Yes

2 Unknown

6. Do you have additional information to report about the procedure(s) performed?

0 No → **FORM COMPLETE**

1 Yes → **CONTINUE BELOW**

SECTION B. Indications

1. What were the indications for the intervention? (Check all that apply.)

- | | |
|--|---------|
| 1 Aneurysm → Complete Col A in table below | F7B11 |
| 1 Ruptured aorta/artery → Complete Col B in table below | F7B12 |
| 1 Dissection → Complete Col C in table below and table at Q3 | F7B13 |
| 1 Malperfusion Complete table at Q4 | F7B14 |
| 1 Valve dysfunction (congenital and acquired) → Complete table at Q5 | F7B15 |
| 1 Aortic repair failure → Complete table at Q6 | F7B16 |
| 1 Aortic coarctation → Continue to Section C unless other indications checked | F7B17 |
| 1 Congenital heart disease → Continue to Section C unless other indications checked | F7B18 |
| 1 Other indication: | F7B19 |
| _____ → Continue to Section C unless other indications checked | F7B19SP |

2. For each indication identified above, place a check mark in the row that describes the location involved by the disorder.

Location		A. Aneurysm	B. Ruptured Aorta/Artery	C. Dissection
a. Aortic root		1 F7B2AA	1 F7B2BA	1 F7B2CA
b. Coronary artery		1 F7B2AB	1 F7B2BB	1 F7B2CB
c. Ascending aorta		1 F7B2AC	1 F7B2BC	1 F7B2CC
d. Aortic arch		1 F7B2AD	1 F7B2BD	1 F7B2CD
e. Descending thoracic aorta		1 F7B2AE	1 F7B2BE	1 F7B2CE
f. Suprarenal abdominal aorta		1 F7B2AF	1 F7B2BF	1 F7B2CF
g. Infrarenal abdominal aorta		1 F7B2AG	1 F7B2BG	1 F7B2CG
h. Innominate artery		1 F7B2AH	1 F7B2BH	1 F7B2CH
i. Subclavian artery	Right	1 F7B2AIR	1 F7B2BIR	1 F7B2CIR
	Left	1 F7B2AIL	1 F7B2BIL	1 F7B2CIL
j. Common carotid artery	Right	1 F7B2AJR	1 F7B2BJR	1 F7B2CJR
	Left	1 F7B2AJL	1 F7B2BJL	1 F7B2CJL
k. Celiac trunk		1 F7B2AK	1 F7B2BK	1 F7B2CK
l. Superior mesenteric artery		1 F7B2AL	1 F7B2BL	1 F7B2CL
m. Renal artery	Right	1 F7B2AMR	1 F7B2BMR	1 F7B2CMR
	Left	1 F7B2AML	1 F7B2BML	1 F7B2CML
n. Common iliac artery	Right	1 F7B2ANR	1 F7B2BNR	1 F7B2CNR
	Left	1 F7B2ANL	1 F7B2BNL	1 F7B2CNL
o. Common femoral artery	Right	1 F7B2AOR	1 F7B2BOR	1 F7B2COR
	Left	1 F7B2AOL	1 F7B2BOL	1 F7B2COL
p. Other artery, specify: _____		1 F7B2AP F7B2APSP	1 F7B2BP F7B2BPSP	1 F7B2CP F7B2CPSP

3. DISSECTION

a. Acuity (check one)	1 Acute (operated within 14 days of onset)	
	2 Chronic (operated more than 14 days from onset)	
	3 Unknown F7B3A	
b. Site of primary tear (all that apply)	1 None	F7B3B1
	1 Aortic root	F7B3B2
	1 Ascending aorta	F7B3B3
	1 Aortic arch	F7B3B4
	1 Descending thoracic aorta	F7B3B5
	1 Abdominal aorta	F7B3B6
	1 Peripheral artery	F7B3B7
	1 Unknown	F7B3B8

4. MALPERFUSION

End organ (all that apply)	1 Coronary	F7B41
	1 Cerebral	F7B42
	1 Upper extremity	F7B43
	1 Spinal	F7B44
	1 Mesenteric	F7B45
	1 Renal	F7B46
	1 Lower extremity	F7B47

c. Type of dissection (check one)	1 Classic dissection 2 Intramural hematoma 3 Penetrating aortic ulcer 4 Iatrogenic dissection F7B3C
--------------------------------------	--

5. VALVE DYSFUNCTION

Which valve(s) was affected?
(all that apply)

- 1** Aortic valve → **Complete Col A below** **F7B51**
- 1** Mitral valve → **Complete Col B below** **F7B52**
- 1** Tricuspid valve → **Complete Col C below** **F7B53**
- 1** Pulmonic valve → **Complete Col D below** **F7B54**

Pathology		A. Aortic Valve		B. Mitral Valve		C. Tricuspid Valve		D. Pulmonic Valve			
a. Stenosis	Mild	1	F7B5AA	1	F7B5BA	1	F7B5CA	1	F7B5DA		
	Moderate	2		2		2		2			
	Severe	3		3		3		3			
	Severity unknown	4		4		4		4			
b. Regurgitation	Mild	1	F7B5AB	1	F7B5BB	1	F7B5CB	1	F7B5DB		
	Moderate	2		2		2		2			
	Severe	3		3		3		3			
	Flail leaflet	4		4		4		4			
	Severity unknown	5		5		5		5			
c. Endocarditis, organism if known: _____		F7B5AC F7B5ACSP		F7B5BC F7B5BCSP		F7B5CC F7B5CCSP		F7B5DC F7B5DCSP			
d. Failed repair		F7B5AD		F7B5BD		F7B5CD		F7B5DD			
e. Prolapse		F7B5AE		F7B5BE		F7B5CE		F7B5DE			
f. Prosthetic dysfunction		F7B5AF		F7B5BF		F7B5CF		F7B5DF			
g. Bicuspid or unicuspid		Valve morphology (check one): F7B5AG1 1 Bicuspid 2 Unicuspid/ unicommisural Type of fusion (all that apply): 1 Right-left F7B5AG21 1 Right-non F7B5AG22 1 Left-non F7B5AG23 1 Partial F7B5AG24 1 Unknown F7B5AG25									
h. Paravalvular leak		F7B5AH		F7B5BH		F7B5CH		F7B5DH			
i. Related operative findings (all that apply)		1 Leaflet fenestration F7B5AI1 1 Torn leaflet F7B5AI2 1 Pannus F7B5AI3									

6. AORTIC REPAIR FAILURE (Check all that apply.)

<p>1 Pseudoaneurysm → F7B61</p>	<p>Location: 1 Aortic valve annulus F7B611 2 Coronary reattachment site 3 Aortic anastomosis 4 Other: _____ 5 Unknown</p>
<p>1 Aortic graft infection → F7B62</p>	<p>Organism, if known: _____ F7B62SP</p>
<p>1 Patch aneurysm → (all that apply) F7B63</p>	<p>1 Brachiocephalic patch F7B631 1 Intercostal or lumbar artery patch F7B632 1 Visceral patch F7B633 1 Other: F7B634 _____ F7B634SP</p>
<p>1 Endoleak → F7B64</p>	<p>Type: 1 I F7B6411 → 1 IA F7B64111 (all that apply, 1 IB F7B64112 see definitions 1 IC F7B64113 page 11) 1 II F7B6412 → 1 IIA F7B64121 1 IIB F7B64122 1 III F7B6413 → 1 IIIA F7B64131 1 IIIB F7B64132 1 IV F7B6414 1 V F7B6415 1 Unknown F7B6416</p> <p>Location: 1 Ascending aorta F7B6421 (all that apply) 1 Aortic arch F7B6422 1 Descending thoracic aorta F7B6423 1 Suprarenal abdominal aorta F7B6424 1 Infrarenal abdominal aorta F7B6425 1 Unknown F7B6426</p>

SECTION C. Types of Procedures

In this section, indicate which type(s) of procedures were performed during this surgery or intervention. For each type performed, provide the procedural details. For all aortic aneurysm and dissection repairs, indicate the largest diameter of the repaired segment based on preoperative imaging or the operative report.

Type of procedure	Procedural details	
1 Coronary artery bypass grafting F7C01		
1 Aortic valve repair <i>(all that apply)</i> F7C02	1 Resuspension F7C021 1 Annuloplasty/commissural placation F7C022 1 Unknown F7C023	
1 Isolated aortic valve replacement (non-root replacement) <i>(check one)</i> F7C03	1 Mechanical valve 2 Bioprosthetic valve 3 Unknown F7C031	
1 Aortic root replacement <i>(check one)</i> F7C04	1 Mechanical valved conduit 2 Stentless tissue aortic root 3 Homograft aortic root 4 Autograft aortic root (Ross procedure) 5 Unknown F7C041 ----- 1 Maximum aortic diameter ___ cm F7C043 2 Unknown F7C042	
1 Valve-sparing aortic root replacement <i>(check one)</i> F7C05	1 Remodeling 1 Reimplantation 1 Other, specify _____ F7C051SP 1 Unknown F7C051 ----- 1 Maximum aortic diameter ___ cm F7C053 2 Unknown F7C052	
1 Ascending aortic replacement F7C06		
1 Aortic arch replacement <i>(check one)</i> F7C07	1 Hemi-arch (beveled) 2 Full arch <i>(all that apply)</i> → 3 Other, specify _____ F7C071SP 4 Unknown F7C071 ----- 1 Maximum aortic diameter ___ cm F7C073 2 Unknown F7C072	1 Elephant trunk F7C0711 1 Branched brachiocephalic reattachment F7C0712

Type of procedure		Procedural details
1 Coarctation repair <i>(all that apply)</i> F7C08		1 Reconstruction of hypoplastic arch F7C081 1 Subclavian flap F7C082 1 Direct end-to-end anastomosis F7C083 1 Graft replacement F7C084 1 Patch aortoplasty F7C085 1 Other, specify _____ F7C086 1 Unknown F7C087
1 Mitral valve repair <i>(all that apply)</i> F7C09		1 Annuloplasty F7C091 1 Leaflet resection, anterior F7C092 1 Leaflet resection, posterior F7C093 1 Artificial chordae implantation F7C094 1 Other, specify _____ F7C095 _____ F7C095SP _____ 1 Unknown F7C096
1 Mitral valve replacement <i>(check one)</i> F7C10		1 Mechanical 2 Bioprosthetic F7C101
1 Tricuspid valve repair <i>(all that apply)</i> F7C11		1 Tricuspid valve annuloplasty F7C112 1 Other, specify _____ F7C113 _____ F7C113SP 1 Unknown F7C114
1 Tricuspid valve replacement F7C12		
1 Pulmonary valve repair <i>(all that apply)</i> F7C13		1 Pulmonary valvuloplasty F7C132 1 Pulmonary valvotomy F7C133 1 Other, specify _____ F7C134 _____ F7C134SP 1 Unknown F7C135
1 Pulmonary valve replacement F7C14		
1 Permanent pacemaker or defibrillator insertion F7C15		1 Permanent pacemaker 1 Defibrillator 1 Unknown F7C151
1 Descending thoracic aortic replacement F7C16		1 Reversed elephant trunk F7C161 1 Maximum aortic diameter _____ cm F7C163 2 Unknown F7C162
1 Thoracoabdominal aortic replacement F7C17	Extent F7C171 <i>(check one, see definitions page 11)</i> 1 I 2 II 3 III 4 IV 5 V 6 Unknown	<i>(all that apply)</i> 1 Reversed elephant trunk 2 Branched visceral reattachment F7C172 1 Maximum aortic diameter _____ cm F7C174 2 Unknown F7C173
1 Removal of stent graft F7C18		

Type of procedure	Procedural details	
1 Open arterial bypass <i>(all that apply)</i> F7C19	1 Innominate artery F7C1901 1 Subclavian artery, right F7C1902 1 Subclavian artery, left F7C1903 1 Subclavian artery, unknown F7C1904 1 Common carotid artery, right F7C1905 1 Common carotid artery, left F7C1906 1 Common carotid artery, unknown F7C1907 1 Celiac trunk F7C1908 1 Superior mesenteric artery F7C1909 1 Renal artery, right F7C1910 1 Renal artery, left F7C1911 1 Renal artery, unknown F7C1912 1 Common iliac artery, right F7C1913 1 Common iliac artery, left F7C1914 1 Common iliac artery, unknown F7C1915 1 Common femoral artery, right F7C1916 1 Common femoral artery, left F7C1917 1 Common femoral artery, unknown F7C1918 1 Other artery, specify: _____ F7C1919 F7C1920	
1 Repair of pectus deformity <i>(check one)</i> F7C20	1 Pectus excavatum 2 Pectus carinatum 3 Other, specify _____ F7C201SP 4 Unknown F7C201	
1 Percutaneous/endovascular intervention <i>(all that apply)</i> F7C21	1 Coronary angioplasty F7C211 → 1 Balloon dilatation of aortic valve F7C212 1 Balloon dilatation of aortic coarctation F7C213 → 1 Fenestration for dissection F7C214 1 Converted to open operation F7C215	With stent? 1 Yes 0 No F7C2111 With stent? 1 Yes 0 No F7C2131

Type of procedure	Procedural details
	<p data-bbox="589 138 1076 170">□ Endovascular graft repairs F7C216 →</p> <p data-bbox="1089 138 1409 170"><u>Segments/vessels excluded</u></p> <p data-bbox="1089 176 1227 203"><i>(all that apply):</i></p> <p data-bbox="1089 207 1511 235">1 Ascending aorta F7C21611</p> <p data-bbox="1089 239 1511 266">1 Aortic arch F7C21612</p> <p data-bbox="1089 270 1511 333">1 Descending thoracic aorta F7C21613</p> <p data-bbox="1089 338 1511 401">1 Suprarenal abdominal aorta F7C21614</p> <p data-bbox="1089 405 1511 468">1 Infrarenal abdominal aorta F7C21615</p> <p data-bbox="1089 472 1511 535">1 Other arteries: F7C21616 _____ F7C21617</p> <hr/> <p data-bbox="1089 577 1495 640">1 Maximum aortic diameter __ cm F7C2163</p> <p data-bbox="1089 644 1235 707">2 Unknown F7C2162</p> <hr/> <p data-bbox="1089 714 1357 741"><u>Proximal landing zone</u></p> <p data-bbox="1089 745 1403 772"><i>(check one, see definitions page 11):</i></p> <p data-bbox="1089 777 1357 829">0 0 1 1 2 2 3 3 4 4 F7C2164</p> <hr/> <p data-bbox="1089 861 1317 888"><i>(check one, if applicable))</i></p> <p data-bbox="1089 892 1187 945">1 C 2 E F7C2165</p> <hr/> <p data-bbox="1089 955 1442 1018">Number of grafts used: _____ F7C2166</p> <hr/> <p data-bbox="1089 1024 1495 1052"><u>Debranching procedure?</u> F7C2167</p> <p data-bbox="1089 1056 1219 1083">1 Yes 0 No</p> <p data-bbox="1089 1087 1105 1115">↓</p> <p data-bbox="1089 1140 1219 1167"><i>(all that apply)</i></p> <p data-bbox="1089 1171 1511 1234">1 Carotid-subclavian bypass or transposition F7C21681</p> <p data-bbox="1089 1239 1511 1266">1 Arch debranching F7C21682</p> <p data-bbox="1089 1270 1511 1297">1 Visceral debranching F7C21683</p>

Type of procedure	Procedural details	
	<p>1 Branch vessel stenting F7C217 → (all that apply)</p> <p>1 Other percutaneous procedure F7C218 _____</p> <p>F7C218SP</p>	<p>1 Innominate artery F7C21701</p> <p>1 Subclavian artery, right F7C21702</p> <p>1 Subclavian artery, left F7C21703</p> <p>1 Subclavian artery, unk F7C21704</p> <p>1 Common carotid artery, right F7C21705</p> <p>1 Common carotid artery, left F7C21706</p> <p>1 Common carotid artery, unk F7C21707</p> <p>1 Celiac trunk F7C21708</p> <p>1 Superior mesenteric artery F7C21709</p> <p>1 Renal artery, right F7C21710</p> <p>1 Renal artery, left F7C21711</p> <p>1 Renal artery, unk F7C21712</p> <p>1 Common iliac artery, right F7C21713</p> <p>1 Common iliac artery, left F7C21714</p> <p>1 Common iliac artery, unk F7C21715</p> <p>1 Common femoral artery, right F7C21716</p> <p>1 Common femoral artery, left F7C21717</p> <p>1 Common femoral artery, unk F7C21718</p> <p>1 Other artery, specify: _____ F7C21719 F7C21720</p>
<p>1 Non-aortic congenital cardiac or vascular procedure (all that apply) F7C22</p>	<p>1 Shunt, specify type: _____ F7C2201 F7C2201S</p> <p>1 Tetralogy repair (TOF) → (check one) F7C2202</p> <p>1 Tetralogy with pulmonary atresia F7C2203</p> <p>1 Truncus repair, specify: _____ F7C2204 F7C2204S</p> <p>1 Double outlet right ventricle, specify: _____ F7C2205 F7C2205S</p> <p>1 Replacement of right ventricle to pulmonary artery (RV to PA) conduit, specify type, size: _____ F7C2206 F7C2206S</p> <p>1 Balloon dilation of branched pulmonary artery, location: _____ F7C2207 F7C2207S</p> <p>1 Stent placement, location: _____ F7C2208 F7C2208S</p>	<p>F7C22021</p> <p>1 Repair with transannular patch</p> <p>2 Repair without transannular patch</p> <p>3 Repair with right ventricle to pulmonary artery conduit, specify type, size: _____</p> <p>4 Other, specify: _____ F7C2202S</p>

Type of procedure	Procedural details	
	1 Ventricular septal defect closure F7C2209 1 Atrial septal defect closure F7C2210 1 Patent foramen ovale (PFO) closure F7C2211 1 Patent ductus arteriosus closure F7C2212 1 Other, specify: _____ F7C2213 F7C2213S 1 Unknown F7C2214	
1 Other cardiac or vascular procedure: _____ F7C23 F7C23SP		

SECTION D. Perfusion and Adjuncts

In the table below, indicate whether perfusion and adjuncts were used. If yes, provide the additional information as necessary.

	1 Yes	0 No	2 Unknown
1. Cardiopulmonary bypass F7D1	Time in minutes: ____ F7D1TM		
2. Hypothermic circulatory arrest F7D2	Time in minutes: ____ F7D2TM <i>(all that apply)</i> 1 With antegrade cerebral perfusion F7D21 1 With retrograde cerebral perfusion F7D22		
3. Left heart bypass F7D3			
4. Cold renal perfusion F7D4			
5. Selective visceral perfusion F7D5			
6. Cerebrospinal fluid drainage F7D6			
7. Evoked potential monitoring F7D7	<i>(all that apply)</i> 1 Motor F7D71 1 Somatosensory F7D72		

SECTION E. Outcomes

- How many nights in total was this patient hospitalized for this intervention? Include time spent both pre- and post-intervention. _____ # nights F7E1
1 Unknown F7E1UNK
- How many nights was the patient hospitalized post-intervention only? _____ # nights F7E2
1 Unknown F7E2UNK
- Did the patient die within 30 days of this intervention? F7E3
0 No
1 Yes → **COMPLETE OFF-STUDY FORM**
- Did the patient die during this hospitalization? F7E4
0 No
1 Yes → **COMPLETE OFF-STUDY FORM**
- Did the patient receive transfusion of any blood products during the procedure or the post-intervention hospital stay? F7E5
0 No
1 Yes

6. Did the patient have any of the following major complications as a result of this intervention?

		1 Yes	0 No	2 Unknown
a. Myocardial infarction	F7E6A			
b. Stroke	F7E6B			
c. Paraplegia/paraparesis	F7E6C			
d. Prolonged intubation	F7E6D			
e. Acute renal failure	F7E6E			
f. Bleeding requiring reoperation	F7E6F			
g. Vocal cord paralysis	F7E6G			
h. Other, specify _____	F7E6H F7E6HSP			

7. What is the current status of this repair? F7E7

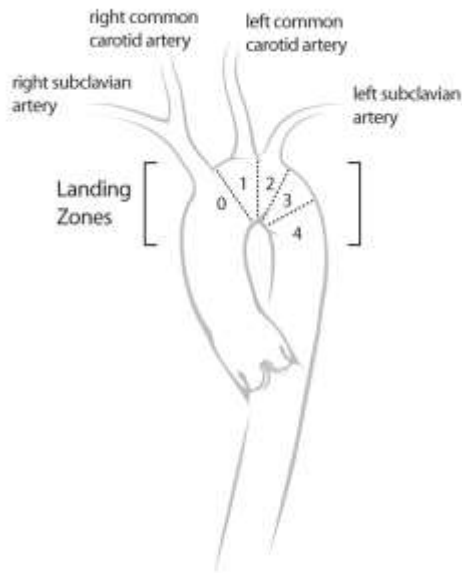
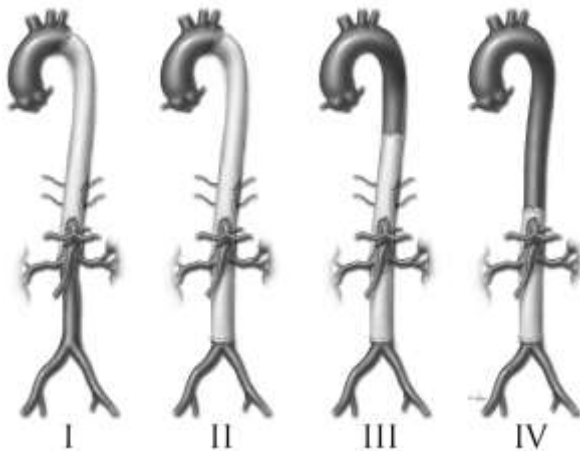
1 Intact		
2 Failure <i>(all that apply)</i>	1 Infection of graft or valve prosthesis F7E71 →	Organism, if known: _____ F7E71SP
	1 Patch aneurysm F7E72	
	1 Pseudoaneurysm F7E73	
	1 Valve dysfunction after repair/replacement <i>(all that apply)</i> F7E74 →	1 Stenosis F7E741 1 Regurgitation F7E742 1 Unknown F7E743
	1 Endoleak <i>(see definitions page 11)</i> F7E75 →	Type: 1 I F7E751 → 1 IA F7E7511 1 IB F7E7512 1 IC F7E7513 <i>(all that apply)</i> 1 II F7E752 → 1 IIA 2 IIB F7E7521 1 III F7E753 → 1 IIIA F7E7531 1 IIIB F7E7532
	1 Reintervention → COMPLETE ANOTHER INTERVENTION FORM F7E76	1 IV F7E754 1 V F7E755 1 Unknown F7E756
	1 Other, specify: _____ F7E77 F7E77SP	

Comments on this form: _____ F7COMM

Initials of Abstractor: |_|_|_|_| F7INIT

FORM COMPLETE	<ul style="list-style-type: none"> KEY DATA ON-LINE AT http://gentac.rti.org PUT FORM IN SUBJECT'S STUDY FILE.
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DEFINITIONS

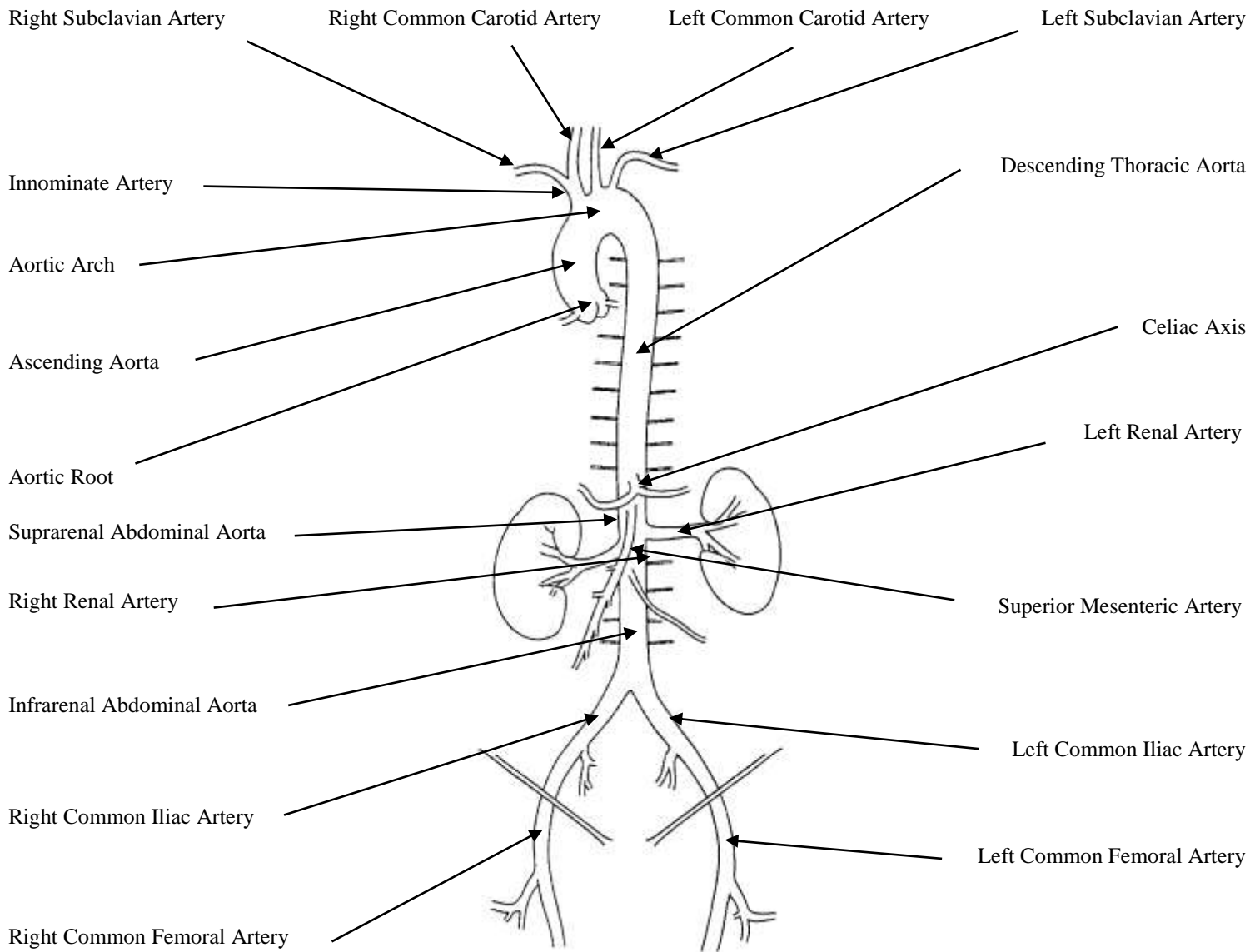


Extent of Thoracoabdominal Aortic Aneurysm Repair (Crawford Classification)	
Repairs are categorized according to the location and extent of aorta replaced in continuity.	
Extent I	Repair extends from the upper descending thoracic aorta (above the 6 th rib) to the suprarenal abdominal aorta
Extent II	Repair extends from the upper descending thoracic aorta (above the 6 th rib) to the infrarenal abdominal aorta
Extent III	Repair involves the distal half or less of the descending thoracic aorta (below the 6 th rib) and any portion of the abdominal aorta
Extent IV	Repair begins near the diaphragm and involves most or all of the abdominal aorta
Extent V*	Repair extends from the distal half of the descending thoracic aorta (below the 6 th rib) to the suprarenal abdominal aorta (repair does not involve infrarenal abdominal aorta).
* The original Crawford Classification is used by most centers and includes extents I – IV. Some centers use a modified Crawford Classification, which adds extent V.	

Proximal Landing Zones for Thoracic Stent Grafts	
Zone 0	Proximal to the origin of the innominate artery
Zone 1	Between the origins of the innominate and left common carotid arteries
Zone 2	Between the origins of the left common carotid and left subclavian arteries
Zone 3	Within 2 cm distal to the origin of the left subclavian artery
Zone 4	More than 2 cm distal to the origin of the left subclavian artery
Zone C	Inside a previously placed conventional surgical graft
Zone E	Inside a previously placed endograft
<i>Adapted from Criado et al. J Vasc Surg 2002;36:1121-8 and Zipfel et al. Ann Thorac Surg 2007; 83:441-9</i>	

Endoleaks	
Type I	Leak at attachment site
A	Proximal attachment site
B	Distal attachment site
C	Iliac occlusion site
Type II	Flow from patent branch vessels (not at attachment sites)
A	Simple (single branch involvement)
B	Complex (two or more branch vessels involved)
Type III	Graft defect
A	Leak at junction or disconnect of graft
B	Disruption of graft fabric (hole)
Type IV	Graft wall porosity
Type V	Increase in maximum aneurysm diameter without detectable endoleak (endotension)
<i>From Veith et al. J Vasc Surg 2002;35:1029-35.</i>	

Use this diagram as a reference to identify the areas of the aorta referred to on this form.





Form 8 - Aortic Tissue Collection

Version 1.0, 02/07/2008

Subject ID Label

For each piece of tissue collected, complete a separate form.

1. Type of aortic sample:

- Normal aorta **F801NA**
- Abnormal aorta → Check one item from list below. **F801AA**
 - Aneurysm without dissection, penetrating aortic ulcer or intramural hematoma

F801A

- Dissection (check one of the following) →

<input type="checkbox"/> With Aneurysm	<input type="checkbox"/> Without Aneurysm	<input type="checkbox"/> Unknown
F801B1	F801B2	F801B3

F801B

- Penetrating aortic ulcer (check only one) →

<input type="checkbox"/> With Aneurysm	<input type="checkbox"/> Without Aneurysm	<input type="checkbox"/> Unknown
F801C1	F801C2	F801C3

F801C

- Intramural hematoma (check only one) →

<input type="checkbox"/> With Aneurysm	<input type="checkbox"/> Without Aneurysm	<input type="checkbox"/> Unknown
F801D1	F801D2	F801D13

F801D

- Coarctation

F801E

- Transition zone between aneurysm and normal aorta

F801F

- Other: _____

F801G

F801G1

2. Date tissue collected:

_ _	_ _	_ _ _ _
Month	Day	Year
F802MM	F802DD	F802YY

3. Affix sample ID# label 017

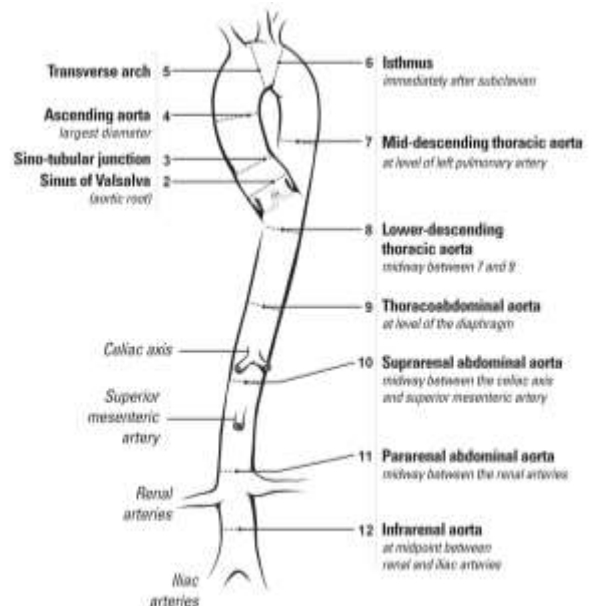
Sample ID label F803

Aortic Tissue Information

4. Based on the reference points in the figure, indicate the region where the sample was obtained.

Check only one. **F804**

- Sinus of Valsalva (region 2) **1**
- Sino-tubular junction (region 3) **2**
- Ascending aorta (region 4) **3**
- Ascending aorta – region unspecified **4**
- Transverse arch (region 5) **5**
- Isthmus (region 6) **6**
- Mid-descending thoracic aorta (region 7) **7**
- Lower descending thoracic aorta (region 8) **8**
- Thoracoabdominal aorta (region 9) **9**
- Descending thoracic aorta – region unspecified **10**
- Suprarenal abdominal aorta (region 10) **11**
- Pararenal abdominal aorta (region 11) **12**
- Infrarenal abdominal aorta (region 12) **13**
- Abdominal aorta – region unspecified **14**



5. Indicate the diameter of the aorta at the sample site:

mm
F805mm

Unknown
F805unk

6. Source of measurement (*check only one*): F806

CT Scan 1
 Echo 2

MRI/MRA 3
 Direct measurement in operating room 4

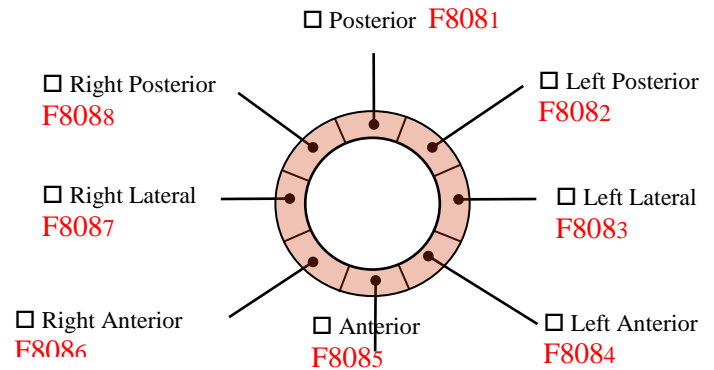
7. For samples from the Sinus of Valsalva, indicate the sinus from which the sample was obtained (*check only one*): F807

Right coronary sinus 1
 Non-coronary sinus 2

Left coronary sinus 3
 Unknown 4

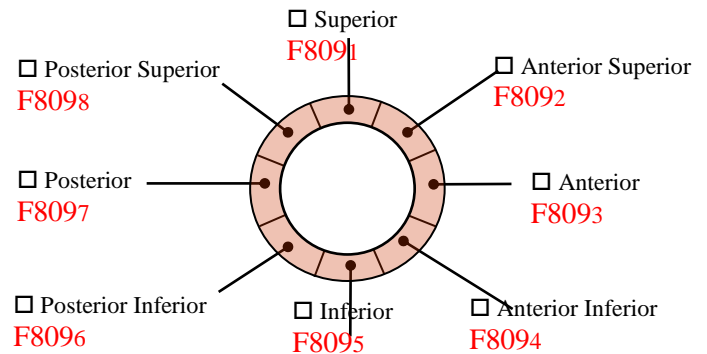
8. For samples from regions 3, 4 and 7 through 12, indicate the portion(s) of the aortic circumference from which the sample was obtained (*check all that apply on the image to the right*): F808

Unknown
F808unk



9. For samples from regions 5 and 6, indicate the portion(s) of the aortic circumference from which the sample was obtained (*check all that apply on the image to the right*): F809

Unknown
F809unk



10. Was the aorta tissue collected adjacent to a previously placed endograft? F810

Yes 0
 No 1
 Unknown 2

11. Was there an *adjacent thrombus*? F811

F811

Yes (*check all that apply*) 0 →
 No 1
 Unknown 2

Chronic thrombus F8110A1
 Liquefied F8110A2
 Fresh clot F8110A3



Subject ID: _____

OFF STUDY FORM (Version 1.0, 3/27/2009)

Complete this form to document significant off-study events that have occurred to enrolled subjects. By reporting a significant event, the subject will not be considered for any scheduled follow-up activities.

- 1. Event 1 Death → **GO TO 4**
- F1401** 2 Duplicate enrollment ID# to delete: _____ **F1401A** → **STOP**
- 3 Ineligibility (data and biospecimens will be destroyed) → **STOP**
- 4 Withdrawal from study Reason: _____ **F1401B** → **GO TO Q2**
- 5 Loss to follow-up Reason: _____ **F1401C** → **GO TO Q3**

2. Do samples and data need to be destroyed? **F1402** 1 Yes 0 No → **STOP**

3. Date last known alive: | _ **F1403MM** _ | | _ **F1403DD** _ | | _ **F1403YY** _ | → **STOP**
Month Day Year

4. Date of death: | _ **F1404MM** _ | | _ **F1404DD** _ | | _ **F1404YY** _ |
Month Day Year

- 5. Cause of death **F1405** 1 Non-cardiovascular
- 2 Cardiovascular

Primary cause _____ **F1405A**
 Secondary cause _____ **F1405B**
 Secondary cause _____ **F1405C**
 Secondary cause _____ **F1405D**

- 6. Was an autopsy performed? 1 Yes 0 No 2 Unknown
- F1406** Location: _____ **F1406A**
- Date: _____ **F1406MM** **F1406DD** **F1406YY**
- Cardiovascular findings: _____ **F1406B**

7. Source of death information: _____ **F1407**

