Demographic Information

Physician

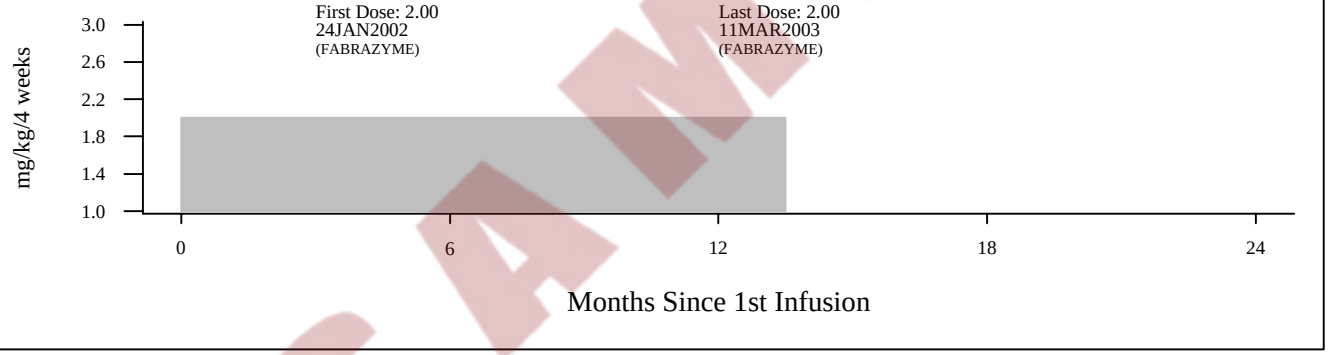
ID:	10002
Name:	Fabry

Patient

ID:	999999	Date of First Infusion:	24JAN2002
Initials:	AAA	Genotype:	
Date of Birth:	25DEC1945	Blood type:	O-
Current Age:	58	Leukocyte a−GAL Activity:	0.41 NMOL/MIN/MG PROTEIN
Sex:	MALE	Plasma a−GAL Activity:	NOT DONE

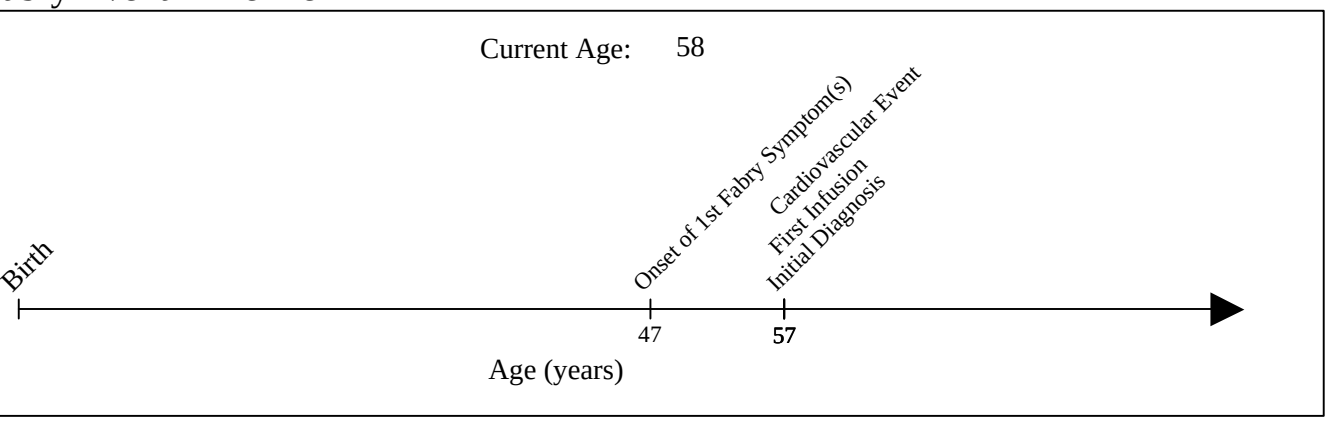
For patients Not on ERT or ERT status is unknown, First Assessment is based on the earliest recorded serum creatinine date if available, or the earliest recorded date from Labs or Vitals.

Enzyme Replacement Therapy History

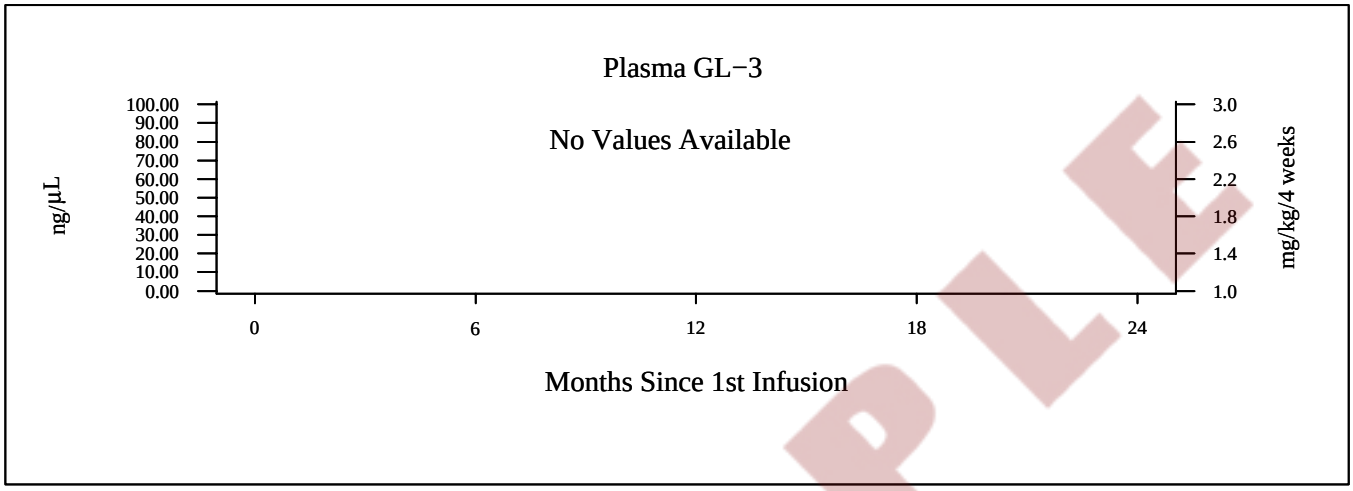


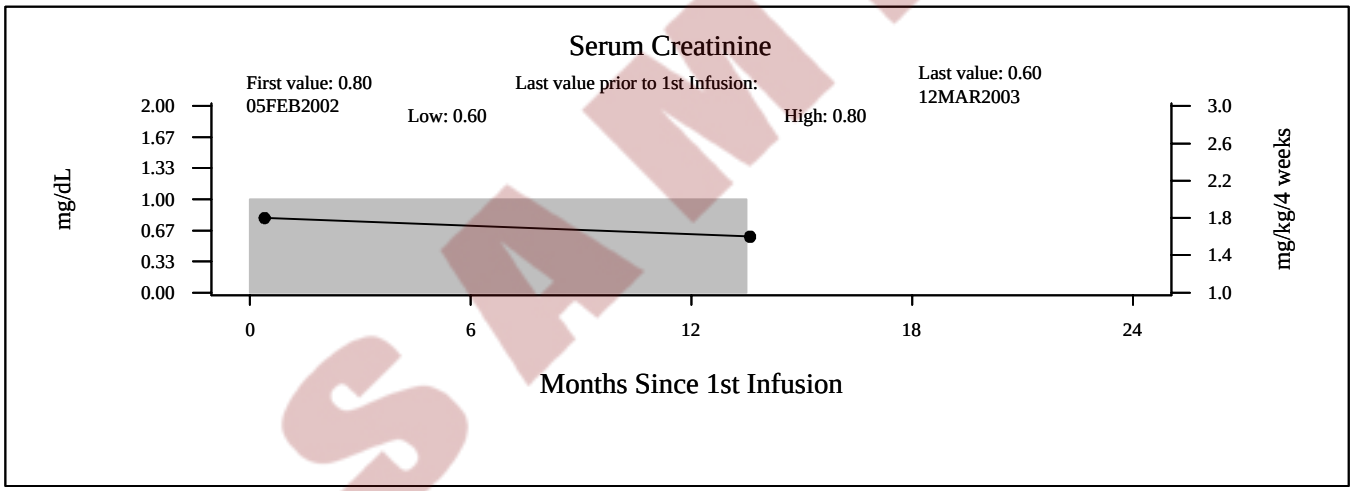
− Dose is presented as mg/kg/4 weeks, which was calculated as follows: ((dose in mg/kg x dose frequency)/frequency interval) x 4.
− The previously recorded dose value is carried forward when an interim dose value is missing; Last Dose: 0.0 signifies a treatment interruption.

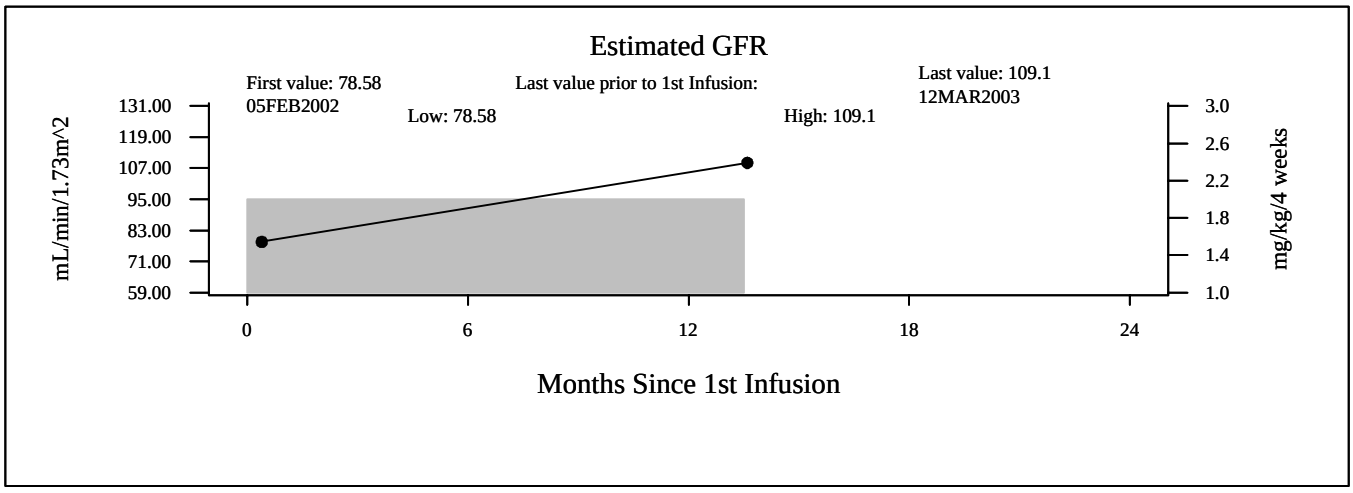
Fabry Event Timeline



Events shown on the timeline represent the first event for each body system (i.e., first cerebrovascular event, first cardiovascular event, first renal event).

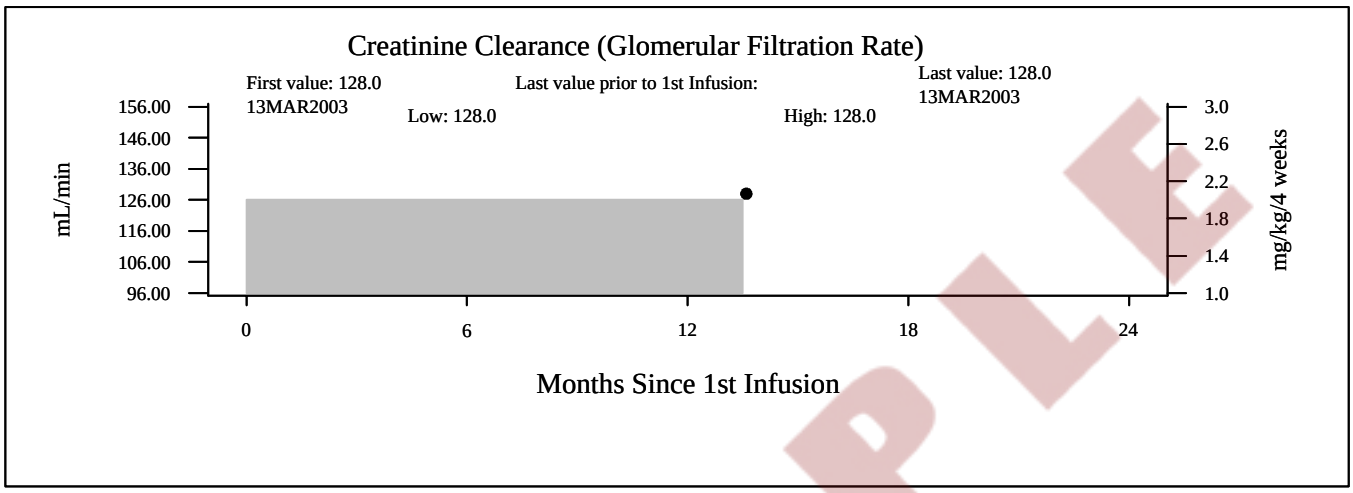


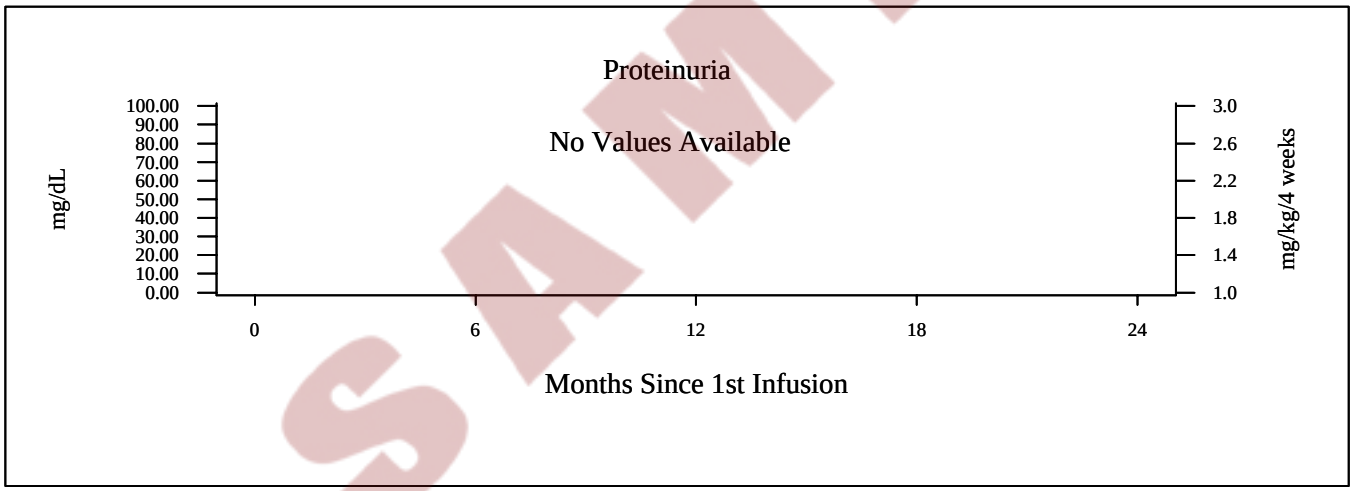




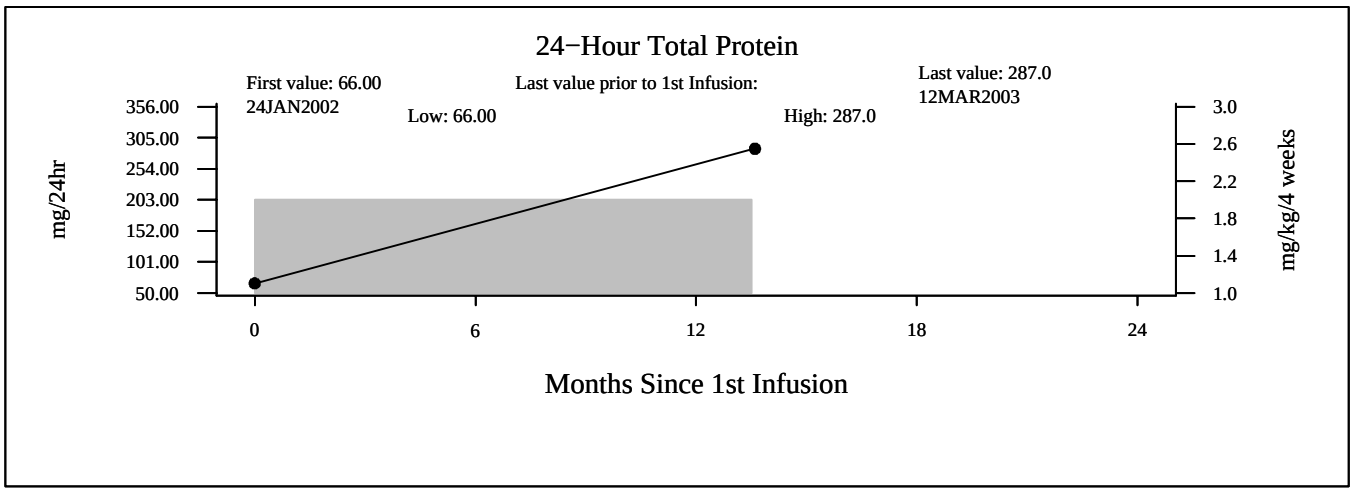
Estimated GFR=186 X (Serum Creatinine (mg/dL))^ −1.154 X (age)^ −0.203 X (0.742, if female) X (1.212, if black).

− Data points in the above graphs are presented for last reported assessment to 100 months back. All data points are presented in the tables which follow.
− For normal range information, please refer to your institution's Lab Normal Ranges.
− Laboratory results are presented in conventional units for the above graphs. Conversion to conventional units may have been necessary. Values not able to be converted are not presented in the graphs, but are shown in the supporting tables that follow. All originally reported results and units are shown in the supporting tables that follow.
− Data under review are not presented in the above graphs.





Proteinuria results measured by dipstick (e.g., Trace, 1+, 2+) are not presented in the above graph. All proteinuria results are shown in Table 2d: Urinalysis Parameters.



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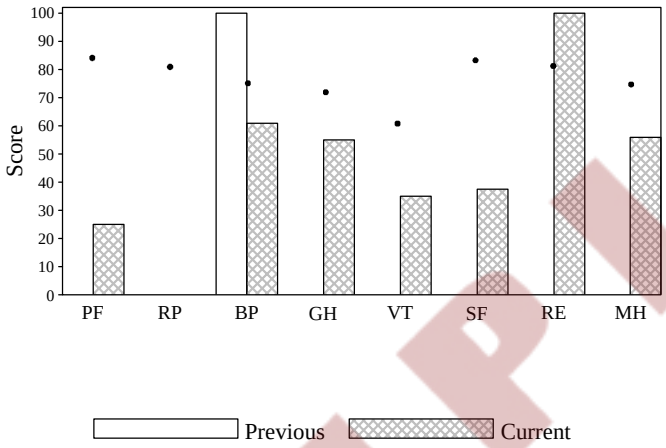
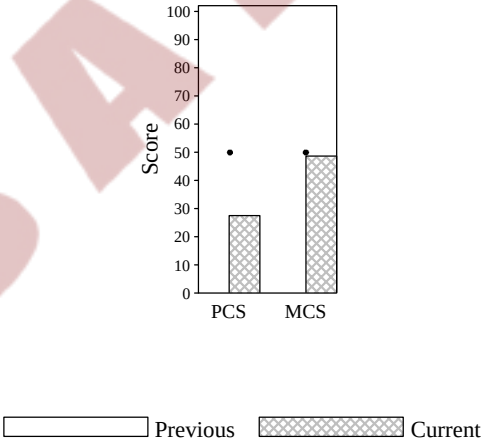
− For normal range information, please refer to your institution's Lab Normal Ranges.
− Laboratory results are presented in conventional units for the above graphs. Conversion to conventional units may have been necessary.
Values not able to be converted are not presented in the graphs, but are shown in the supporting tables that follow.
All originally reported results and units are shown in the supporting tables that follow.
− Data under review are not presented in the above graphs.

SF−36®

Health Status Profile



Health Status Profile Summary Scores



Legend

SF−36 Visit	SF−36 Visit Date	Months Since 1st Infusion
= Previous (prior to Current and >1 mo. from 1st Infusion/Assessment)	07NOV2002	9.4
= Current (>1 mo. from 1st Infusion/Assessment)	11MAR2003	13.5

- U.S. general population norm (NSFHS, 1990).

PF=Physical Functioning
RP=Role Physical
BP=Bodily Pain
GH=General Health
VT=Vitality
SF=Social Functioning
RE=Role Emotional
MH=Mental Health
PCS=Physical Component Summary
MCS=Mental Component Summary

Appendix: Supporting Tables

Section 1: ENZYME REPLACEMENT THERAPY

Table 1: Treatment History

Visit Date	Months Since 1st Infusion	Age (years)	Therapy	Dose (mg/kg)	Frequency/ 4 weeks	Calculated Dose (mg/kg/4 weeks)	Total Amount Infused (mg)	Time Required per Infusion	Reason for Change
24JAN2002	0	57	FABRAZYME	1.00	2	2	70.00	2 HOURS TO <4 HOURS	
11MAR2003	13.5	58	FABRAZYME	1.00	2	2	70.00	2 HOURS TO <4 HOURS	

− Calculated Dose (mg/kg/4 weeks)=Dose (mg/kg) multiplied by Frequency/4 weeks.

Section 2: VITALS AND LABORATORY MEASUREMENTS

Table 2a: Vitals − Weight, Height and Blood Pressure

Exam Date	Months Since 1st Infusion	Age (years)	Weight (kg)	Height (cm)	Arterial Blood Pressure (systolic/diastolic)
24JAN2002	0.0	57	85.00		
23JAN2002	0.0	57	70.00	164.00	110/70
11MAR2003	13.5	58	68.00	164.00	122/70

− Weight values reported in pounds (lb) were converted to kilograms (kg) by dividing by 2.2046.
− Height values reported in inches (in) were converted to centimeters (cm) by multiplying by 2.54.

Table 2b: Serology Parameters (Hemoglobin, Triglycerides, Serum Creatinine, BUN)

			Hemoglobin (HGB)		Triglycerides		Serum Creatinine		BUN	
Sample Date	Months Since 1st Infusion	Age (years)	Reported Value (unit)	Converted Value (g/dL)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/dL)
05FEB2002	0.4	57	13.3 (G/DL)	13.30	131 (MG/DL)	131.00	0.8 (MG/DL)	0.80		
12MAR2003	13.6	58	13 (G/DL)	13.00	142 (MG/DL)	142.00	0.6 (MG/DL)	0.60		

− HGB values reported in mmol/L were converted to g/dL by multiplying by 1.650.
− Tryglyceride values reported in mmol/L were converted to mg/dL by multiplying by 88.54.
− Serum Creatinine values reported in µmol/L were converted to mg/dL by multiplying by 0.01131.
− BUN values reported in mmol/L were converted to mg/dL by multiplying by 2.801.

Table 2c: Serology Parameters (HDL, LDL, Total Cholesterol)

			HDL		LDL		Total Cholesterol	
Sample Date	Months Since 1st Infusion	Age (years)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/dL)
05FEB2002	0.4	57	52 (MG/DL)	52.00	116 (MG/DL)	116.00	194 (MG/DL)	194.00
12MAR2003	13.6	58	59 (MG/DL)	59.00	103 (MG/DL)	103.00	190 (MG/DL)	190.00

− HDL, LDL and Total Cholesterol values reported in mmol/L were converted to mg/dL by multiplying by 36.67.

Table 2d: Urinalysis Parameters

			Microalbumin		Proteinuria		24−Hour Total Protein	
Sample Date	Months Since 1st Infusion	Age (years)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/dL)	Reported Value (unit)	Converted Value (mg/24 hr)
24JAN2002	0.0	57					66 (MG/24HR)	66.00
25JAN2002	0.0	57	51 (MG/L)	5.10				
12MAR2003	13.6	58					287 (MG/24HR)	287.00

− Proteinuria values reported as measured by dipstick (e.g., Trace, 1+, 2+) were not converted.

Appendix: Supporting Tables

Table 2e: Specialized Test Parameters

					Glomerular Filtration Rate					
			Plasma GL3		Creatinine Clearance		Inulin Clearance		Iothalamate Clearance	
Sample Date	Months Since 1st Infusion	Age (years)	Reported Value (unit)	Converted Value (ng/ μL)	Reported Value (unit)	Converted Value (mL/min)	Reported Value (unit)	Converted Value (mL/min)	Reported Value (unit)	Converted Value (mL/min)
13MAR2003	13.6	58			128 (ML/MIN)	128.00				

Section 3: CEREBROVASCULAR ASSESSMENTS & EVENTS

Table 3a: Transient Ischemic Attack

HISTORY

Visit Date	Age (years)	Transient Ischemic Attack	# Attacks in the Past Year
07NOV2002	57		

FOLLOW−UP (Since Last Assessment)

Visit Date	Months Since 1st Infusion	Age (years)	Transient Ischemic Attack	# Attacks Since Last Assessment
11MAR2003	13.5	58	N	

Table 3b: Infarct Stroke

No Data for this Patient

Table 3c: Hemorrhagic Stroke

No Data for this Patient

Table 3d: Cranial Brain Image

No Data for this Patient

Section 4: RENAL EVENTS

Table 4a: Event − Dialysis Initiated

No Data for this Patient

Table 4b: Event − Transplant Received

No Data for this Patient

Appendix: Supporting Tables

Section 5: CARDIOVASCULAR ASSESSMENTS & EVENTS

Table 5a: Cardiovascular Assessments

HISTORY

Visit Date	Age (years)	Arrhythmia	Angina Pectoris	Myocardial Infarction	Congestive Heart Failure	Significant Cardiac Procedure	Hypertension	Hypertension Medication Required
07NOV2002	57	Y	Y	N	Y	Y	Y	

FOLLOW−UP (Since Last Assessment)

Visit Date	Months Since 1st Infusion	Age (years)	New Diagnosis or Change in Arrhythmia (Status)	New Diagnosis or Change in Angina Pectoris (Status)	Myocardial Infarction	New Diagnosis or Change in Congestive Heart Failure (Status)	Significant Cardiac Procedure	New Diagnosis or Change in Hypertension (Status)	Hypertension Medication Required
11MAR2003	13.5	58		Y (WORSE)	N	Y	Y	Y	Y

Table 5b: Event − Arrhythmia

Event Date	Months Since 1st Infusion	Age (years)	Sinus Rhythm Abnormalilty	Arrhythmia Type	Medication Required	Conduction Abnormality
21AUG2002	6.9	57	Y	UNKNOWN	Y	Y
15FEB2003	12.7	58	Y	UNKNOWN	Y	Y

Table 5c: Event − Angina Pectoris

Event Date	Months Since 1st Infusion	Age (years)	Angina Type	Medication Required
21AUG2002	6.9	57	INCREASING ANGINA	Y
15FEB2003	12.7	58	INCREASING ANGINA	Y

Table 5d: Event − Myocardial Infarction

No Data for this Patient

Table 5e: Event − Congestive Heart Failure

			Valvular Dysfunction		Wall Motion Abnormality		Hypertrophy				
Event Date	Months Since 1st Infusion	Age (years)	Finding	Valve	Finding	Type	Finding	LVH Present	Left Posterior Wall Thickness (mm)	Estimated Ejection Fraction (%)	Medication Required
21AUG2002	6.9	57	Y	MITRAL	N		Y	Y	12.00	55−70	Y
15FEB2003	12.7	58	Y	MITRAL	N		Y	Y	11.00	55−70	Y

Table 5f: Event − Significant Cardiac Procedure

Event Date	Months Since 1st Infusion	Age (years)	Procedure
21AUG2002	6.9	57	EVENT RECORDER; MYOCARDIAL BIOPSY
15FEB2003	12.7	58	EXPLANATION OF THE EVENT RECORDER

Appendix: Supporting Tables

Section 6: RESPIRATORY ASSESSMENTS

Table 6a: Respiratory Disease Parameters

HISTORY

Visit Date	Age (years)	Pulmonary Involvement	Obstructive Disease	Restrictive Disease
07NOV2002	57	Y	Y	N

FOLLOW−UP (Since Last Assessment)

Visit Date	Months Since 1st Infusion	Age (years)	New Diagnosis or Change in Pulmonary Involvement	Obstructive Disease (Status)	Restrictive Disease (Status)
11MAR2003	13.5	58	N		

Table 6b: Respiratory Exam Parameters

Visit Date	Months Since 1st Infusion	Age (years)	FEV_1(L)	FEV_1 % Predicted	FVC(L)	FVC % Predicted
25JAN2002	0.0	57	1.88	76.00	2.54	84.00
11MAR2003	13.5	58	1.70	70.20	2.73	91.80

Section 7: OPHTHALMOLOGY ASSESSMENTS

Table 7: Ophthalmology Exam Parameters

No Data for this Patient

Section 8: QUALITY OF LIFE MEASUREMENTS

Table 8a: SF−36® Health Status Profile Scales

Visit Date	Months Since 1st Infusion	Age (years)	SF−36 Visit	PF	RP	BP	GH	VT	SF	RE	MH
07NOV2002	9.4	57	PREVIOUS			100					
11MAR2003	13.5	58	CURRENT	25	0	61	55	35	37.5	100	56

− Refer to footnote in Table 8b: SF36® Health Status Profile Summary Scores.

Table 8b: SF−36® Health Status Summary Scores

Visit Date	Months Since 1st Infusion	Age (years)	SF−36 Visit	PCS	MCS
07NOV2002	9.4	57	PREVIOUS		
11MAR2003	13.5	58	CURRENT	27.5	48.6

− Initial = the visit −8/+1 months from 1st Infusion or −8/+6 from 1st Assessment.
− Previous = the visit prior to Current and >1 month from 1st Infusion/1st Assessment.
− Current = the most recent visit >1 month from 1st Infusion/1st Assessment.
− Refer to SF36 Manual and Interpretation Guide, Ware JE, et al,1993.

Appendix: Supporting Tables

Table 8c: Brief Pain Inventory (Short Form): Questions 1−6

Visit Date	Months Since 1st Infusion	Age (years)	Unusual Pain Today	Location(s) of Pain	Worst Pain in 24 HR*	Least Pain in 24 HR*	Average Pain*	Pain Right Now*
07NOV2002	9.4	57	N	NOT DONE	0	0	0	0
11MAR2003	13.5	58	N	NOT DONE	0	0	0	0

* Based on a scale of 0−10 where 0=No Pain and 10=Pain as bad as you can imagine.

Table 8d: Brief Pain Inventory (Short Form): Questions 7−9

					Interference from Pain in Last 24HR**						
Visit Date	Months Since 1st Infusion	Age (years)	Medications	Relief in 24HR*	General Activity	Mood	Walking Ability	Normal Work	Relations	Sleep	Enjoyment of Life
07NOV2002	9.4	57		NOT DONE	0	0	0	0	0	0	0
11MAR2003	13.5	58		NOT DONE	5	4	5	6	6	6	3

* Based on a scale of 0−100% where 0%=No Relief and 100%=Complete Relief.
** Based on a scale of 0−10 where 0=Does not interfere and 10=Completely interferes.

Section 9: NEUROLOGY ASSESSMENTS

Table 9a: Autonomic Parameters

HISTORY

Visit Date	Age (years)	Sweating	Heat Tolerance	Cold Tolerance
07NOV2002	57	NORMAL	NORMAL	NORMAL

FOLLOW−UP (Since Last Assessment)

Visit Date	Months Since 1st Infusion	Age (years)	Sweating	Heat Tolerance	Cold Tolerance
11MAR2003	13.5	58	NORMAL	ABNORMAL	NORMAL

Table 9b: Peripheral Pain

HISTORY

		Chronic Pain		Current Pain Therapy		
Visit Date	Age (years)	Finding	Frequency	Finding	Type	Frequency
07NOV2002	57	N		N		

FOLLOW−UP (Since Last Assessment)

			Chronic Pain		Current Pain Therapy			
Visit Date	Months Since 1st Infusion	Age (years)	Finding	Frequency	Finding	Type	Frequency	Change in Therapy Regimen
11MAR2003	13.5	58	Y	UNKNOWN	N			

Appendix: Supporting Tables

Table 9c: Pain Crises and Clinical Depression

HISTORY

		Acute Pain Crisis in the Past Year		Clinical Depression	
Visit Date	Age (years)	Finding	# of Crises in the Past Year	Finding	Medical Treatment Required
07NOV2002	57	N		N	

FOLLOW−UP (Since Last Assessment)

			Acute Pain Crisis Since Last Assessment		New Diagnosis of Clinical Depression	
Visit Date	Months Since 1st Infusion	Age (years)	Finding	# of Crises since the Last Assessment	Finding	Medical Treatment Required
11MAR2003	13.5	58	N		N	

Section 10: OTHER CLINICAL ASSESSMENTS

Table 10a: Gastroenterology Parameters

HISTORY

		Abdominal Pain			Diarrhea	
Visit Date	Age (years)	Finding	Frequency	Intensity	Finding	Frequency
07NOV2002	57	NO			NO	

FOLLOW−UP (Since Last Assessment)

			Abdominal Pain			Diarrhea	
Visit Date	Months Since 1st Infusion	Age (years)	Finding	Frequency	Intensity	Finding	Frequency
11MAR2003	13.5	58	NO			NO	

Table 10b: Skin

HISTORY

Visit Date	Age (years)	Angiokeratoma
07NOV2002	57	N

FOLLOW−UP (Since Last Assessment)

Visit Date	Months Since 1st Infusion	Age (years)	Angiokeratoma (Status)
11MAR2003	13.5	58	N

Table 10c: Additional Medical Information

No Data for this Patient

Appendix: Supporting Tables

Section 11: DATA POINTS UNDER REVIEW

Table 11: Data Points under Review

No Data for this Patient