



Demographic Information

Patient

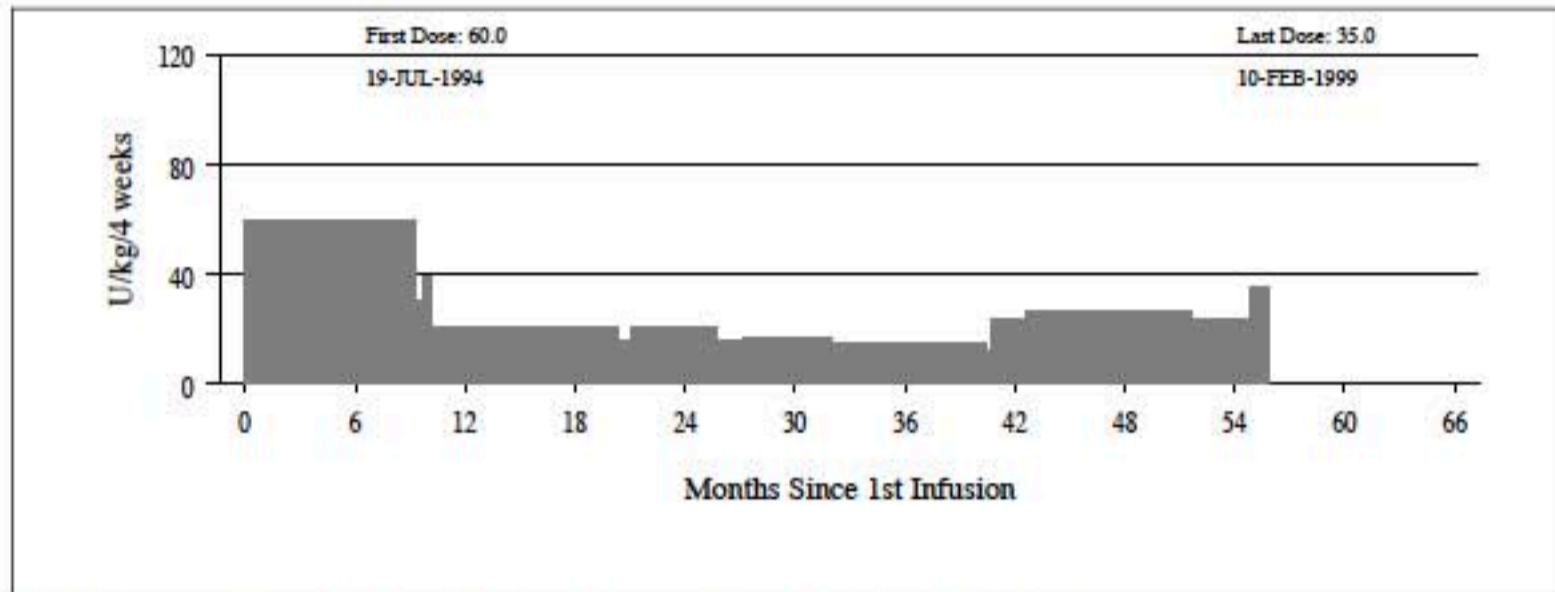
ID:	Date of First Infusion:
Initials:	Splenectomy Status:
Date of Birth:	Date of Splenectomy:
Age:	Disease Type:
Sex:	Genotype:

Physician

ID:
Name:

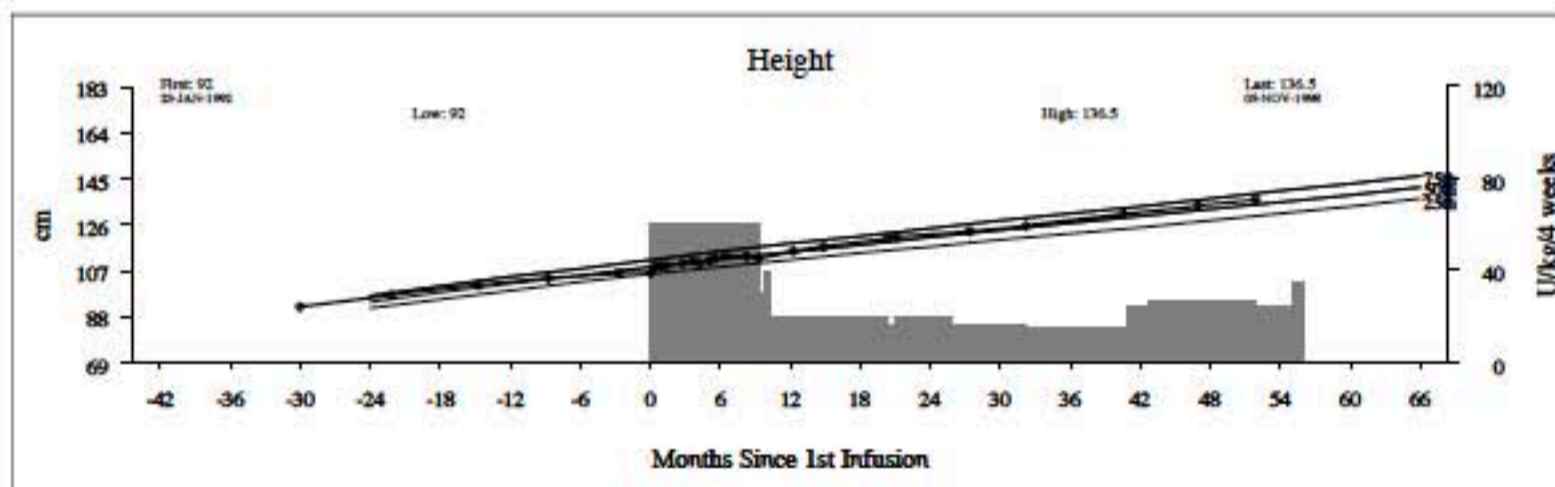
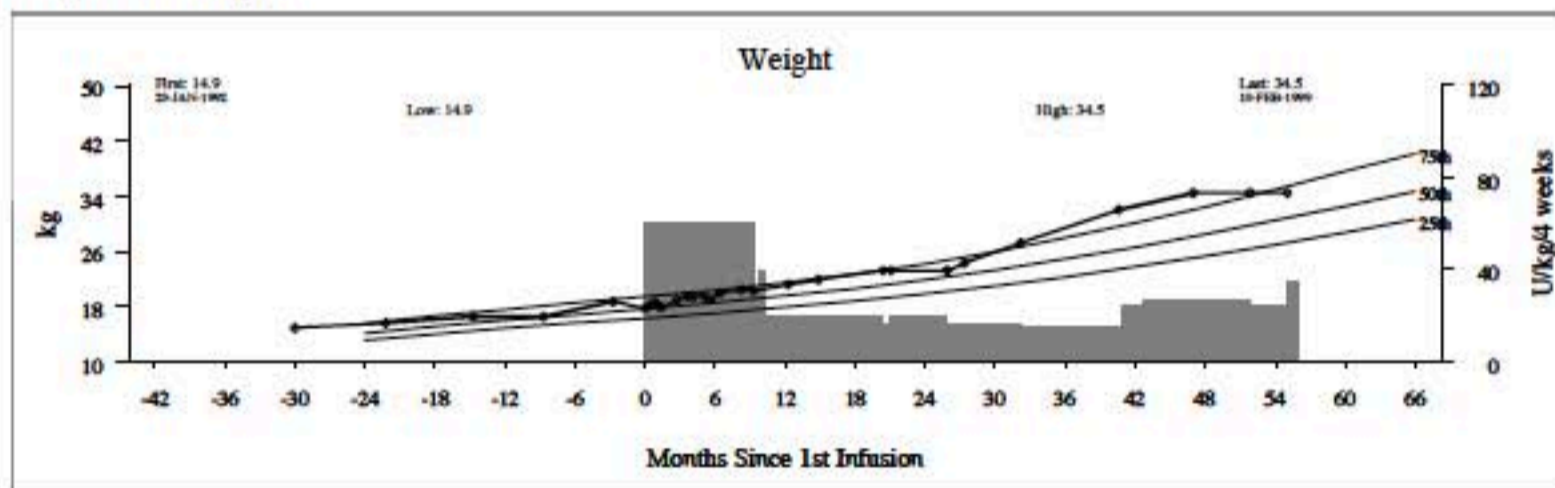
For patients not on ERT, first assessment is based on the earliest recorded date from Hematology, Visceral Organs, or Vital Signs.

Enzyme Replacement Therapy History



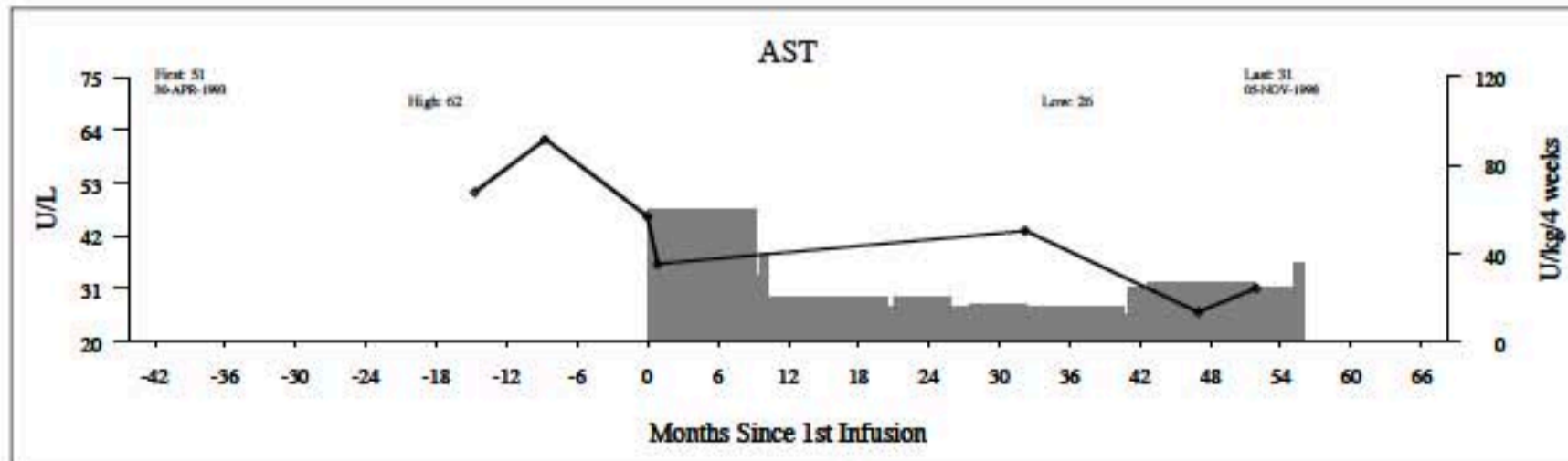
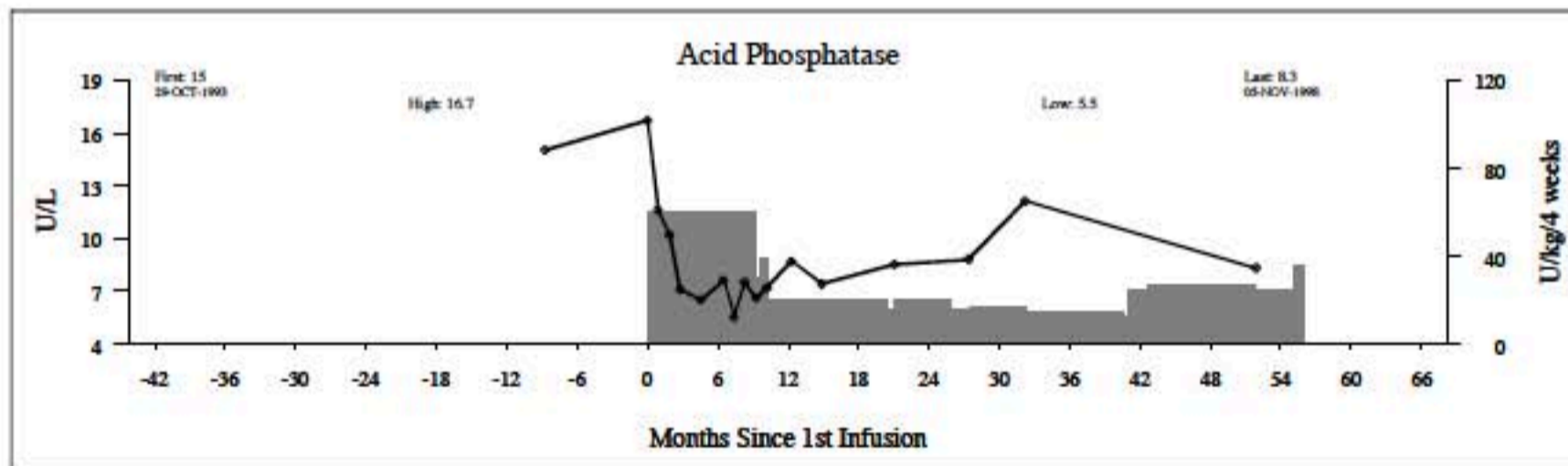
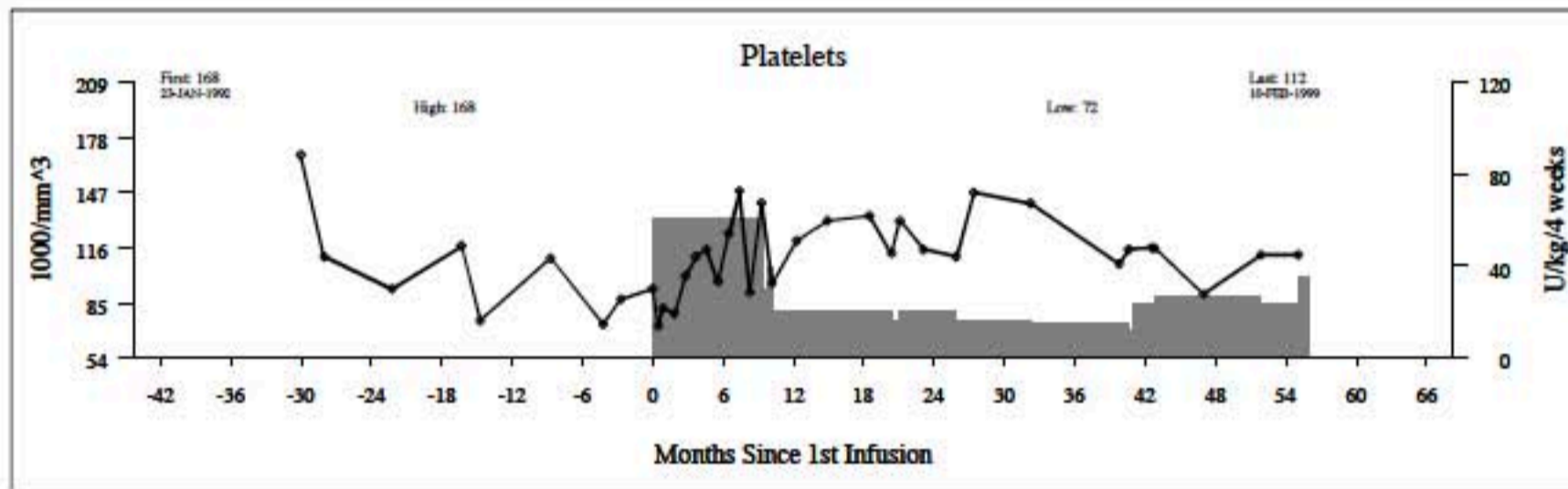
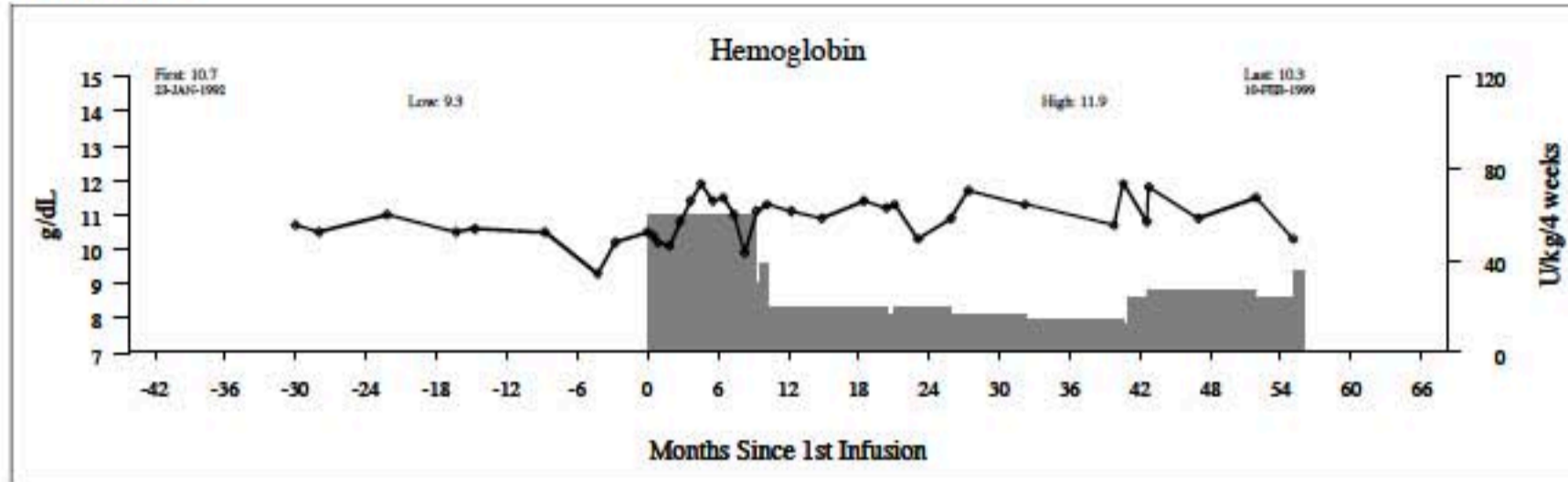
The previously recorded dose value is carried forward when an interim dose value is missing; Last Dose: 0.0 signifies a treatment interruption.

Weight and Height



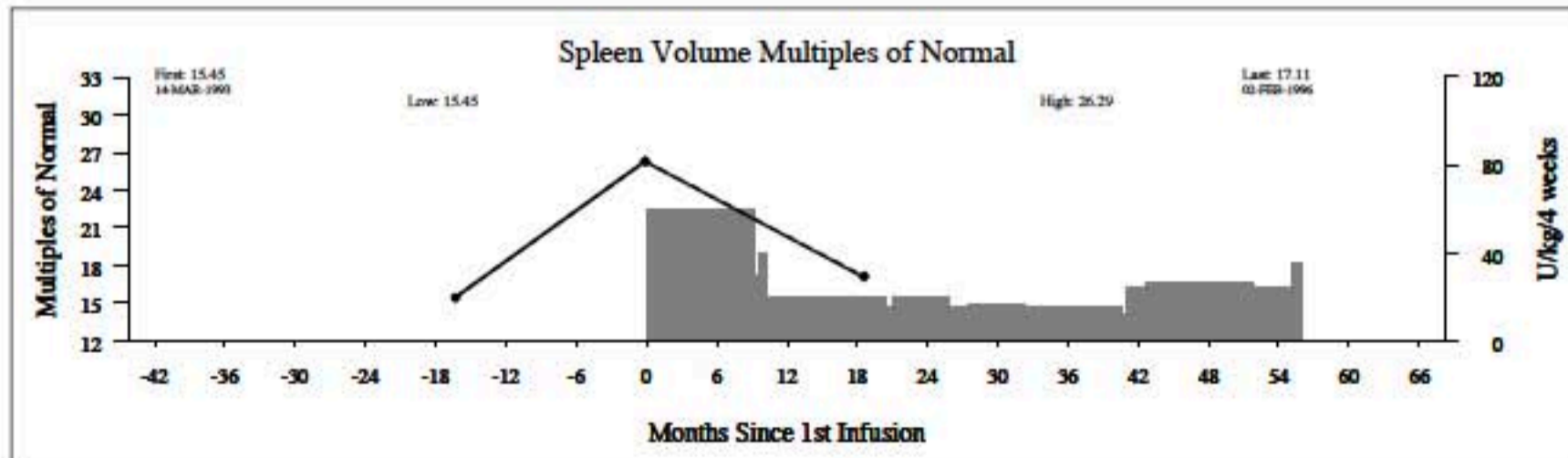
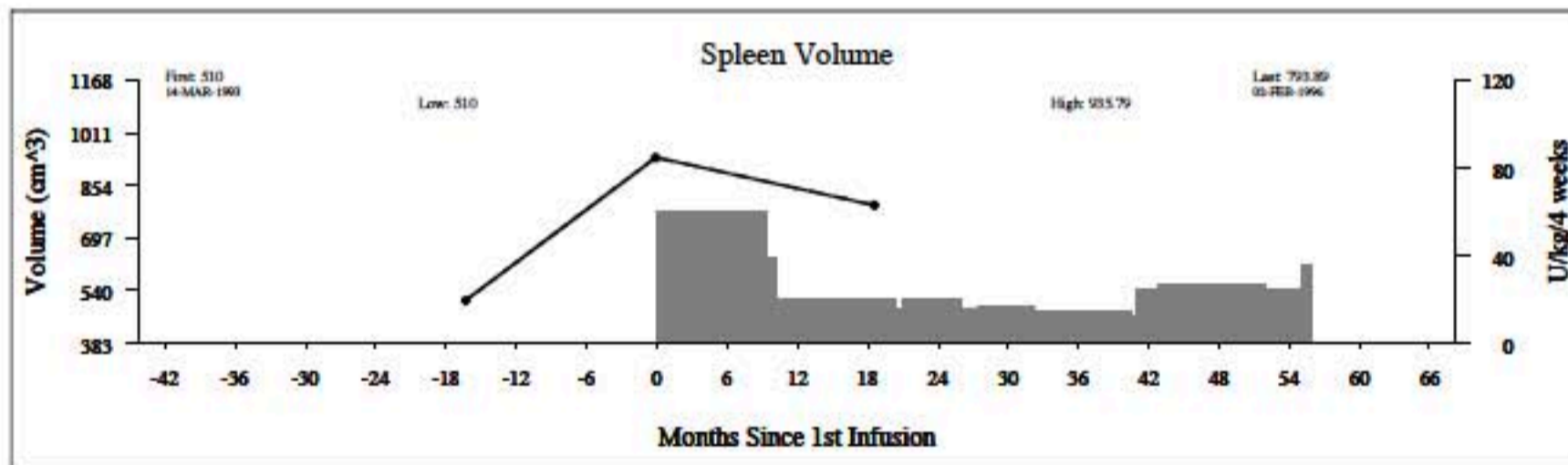
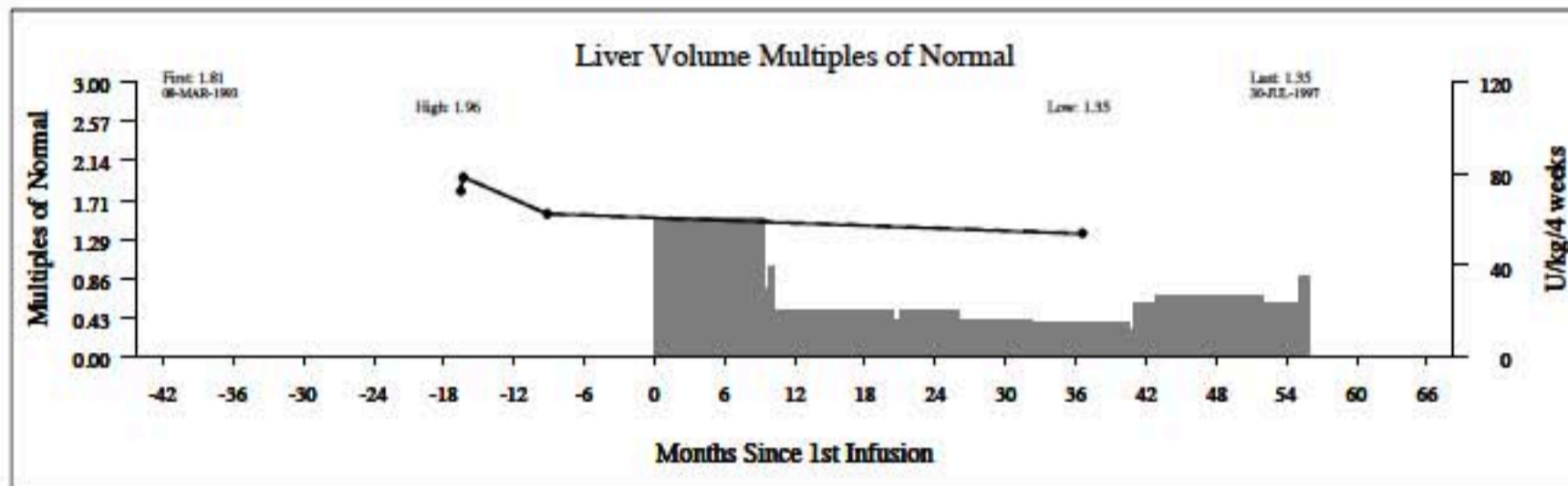
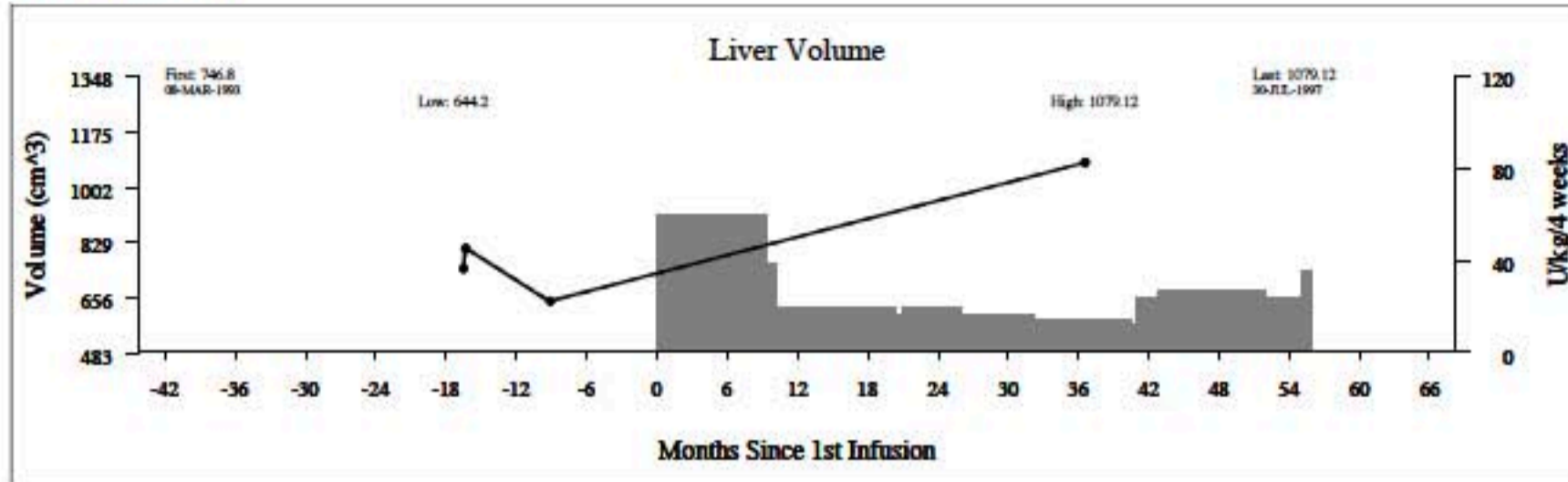
The age-specific percentiles for normal growth are provided (NCHS, 1979).

Hematology



- Multiples of Normal Calculations: Liver = Actual Volume(cc) / (Weight(kg) x 25)
Spleen = Actual Volume(cc) / (Weight(kg) x 2)
- Data points in Visceral Organ graphs represent MRI/CT (•) and Ultrasound (◇) only.
- Ultrasound volumetric conversions were calculated using the following formulas: Liver cc = 320.86 + (0.317 x 3 dimensions)
* Elstein, et al., 1997
Spleen cc = 539.50 + (0.344 x 3 dimensions)
- For normal range information, please refer to your institution's Lab Normal Ranges.
- Data under review are not presented in the above graphs.

Visceral Organs



- Multiples of Normal Calculations: Liver = Actual Volume(cc) / (Weight(kg) x 25)
 Spleen = Actual Volume(cc) / (Weight(kg) x 2)
- Data points in Visceral Organ graphs represent MRI/CT (•) and Ultrasound (◇) only.
- Ultrasound volumetric conversions were calculated using the following formulas: Liver cc = 320.86 + (0.317 x 3 dimensions)
 * Elstein, et al., 1997
 Spleen cc = 539.50 + (0.344 x 3 dimensions)
- For normal range information, please refer to your institution's Lab Normal Ranges.
- Data under review are not presented in the above graphs.

Appendix: Supporting Tables

Table 1: Visceral Organs

Date	Months Since 1st Infusion	Method of Assessment	Weight kg	Spleen				Liver			
				Reported Volume	Calculated Value	Multiples of Normal	Splenomegaly	Reported Volume	Calculated Value	Multiples of Normal	Hepatomegaly
23-MAR-1992	-27.9						YES				YES
06-DEC-1992	-19.4	MRI		15 CM			YES				
08-MAR-1993	-16.4	CT	16.5					8.6 X 12.4 X 12.6 CM	746.8	1.81	
14-MAR-1993	-16.2	MRI	16.5	51 L	510	15.45		.81 L	810	1.96	
18-OCT-1993	-9	MRI	16.5	*SPLENIC INDEX* OF 1020				17 X 10 X 6 CM	644.2	1.56	
15-JUL-1994	-0.1	CT	17.8	16 X 6 X 12 CM	935.79	26.29					
02-FEB-1996	18.5	MRI	23.2	6 X 8.5 X 14.5 CM	793.89	17.11					
26-MAR-1996	20.3	US		5.0 CM				3.0 CM			
24-OCT-1996	27.2	PE		4 CM			YES	8 CM			YES
18-MAR-1997	32	PE		8 CM			YES	10 CM			YES
30-JUL-1997	36.4	MRI	[32.0]	17 CM				11.5 CM X 13 CM X 16 CM	1079.12	[1.35]	
27-NOV-1997	40.4	PE		4 CM				7 CM			
15-MAY-1998	45.9	MRI		MAX LENGTH: 17.5 CM			YES	13 X 11.5 CM MAX LENGTH: 15 CM			YES
11-JUN-1998	46.8	PE		6 CM				11 CM			
05-NOV-1998	51.6	PE		3 CM			NO	2 CM			NO

Multiples of normal are calculated using the nearest weight within 3 months (Child) or 6 months (Adult) of Date of Assessment. When that weight occurs more than 6 months from the Date of Assessment, the value for the weight and the Multiple of Normal are bracketed.

Table 2: Bone - Pain, Crisis and Mobility

Date	Months Since 1st Infusion	Bone Pain	Severity	Bone Crisis	# of Occurrences	Mobility
HISTORY		YES		NO		UNRESTRICTED
23-JAN-1992	-29.9			NO		
23-MAR-1992	-27.9			NO		
08-MAR-1993	-16.4					UNRESTRICTED
14-MAR-1993	-16.2					UNRESTRICTED
30-APR-1993	-14.6	YES	SEVERE	NO		
14-MAR-1994	-4.2					UNRESTRICTED
28-APR-1994	-2.7	YES	SEVERE	NO		
15-JUL-1994	-0.1					UNRESTRICTED
02-AUG-1994	0.5			NO		
30-AUG-1994	1.4	YES	MILD	NO		
13-SEP-1994	1.8			NO		
11-OCT-1994	2.8			NO		
08-NOV-1994	3.7			NO		
22-NOV-1994	4.1	YES	MILD	NO		
06-DEC-1994	4.6			NO		
20-DEC-1994	5.1			NO		
03-JAN-1995	5.5	YES	MODERATE	NO		
31-JAN-1995	6.4	YES	MILD	NO		
28-FEB-1995	7.4			NO		
13-MAR-1995	7.8			NO		
28-MAR-1995	8.3	NO		NO		
25-APR-1995	9.2	YES	MILD	NO		

Date	Months Since 1st Infusion	Bone Pain	Severity	Bone Crisis	# of Occurrences	Mobility
09-MAY-1995	9.7			NO		
23-MAY-1995	10.1			NO		
27-JUL-1995	12.3	YES	MILD	NO		
12-OCT-1995	14.8			NO		
30-JAN-1996	18.4			NO		
02-FEB-1996	18.5					UNRESTRICTED
26-MAR-1996	20.3	NO		NO		UNRESTRICTED
18-APR-1996	21			NO		
18-JUN-1996	23			NO		
24-OCT-1996	27.2	NO		NO		
18-MAR-1997	32	NO		NO		
30-JUL-1997	36.4					UNRESTRICTED
04-NOV-1997	39.6			NO		
27-NOV-1997	40.4	NO		NO		UNRESTRICTED
27-JAN-1998	42.4	NOT ASSESSED		NOT ASSESSED		NOT ASSESSED
02-FEB-1998	42.6	NOT ASSESSED		NOT ASSESSED		NOT ASSESSED
11-JUN-1998	46.8	NO		NO		UNRESTRICTED
05-NOV-1998	51.6	YES	VERY MILD	NO		UNRESTRICTED
10-FEB-1999	54.8	NO		NO		UNRESTRICTED

Table 3: Radiologic Skeletal Manifestations

Manifestations	Assessment Method	Visit Date	Months Since 1st Infusion	Finding	Area of Involvement	Side (L/R)	Findings Since Comparison Date		
							Compare Date	Left Side	Right Side
MARROW INFILTRATION	MRI	14-MAR-1994	-4.2	PRESENT	PROXIMAL FEMORAL METAPHYSIS	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	DIAPHYSIS	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	GREATER TROCHANTER	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	DISTAL FEMORAL EPIPHYSIS	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	CAPITAL FEMORAL EPIPHYSIS	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	HIPS TO KNEES	BOTH		NEW	NEW
		14-MAR-1994	-4.2	PRESENT	DISTAL FEMORAL METAPHYSIS	BOTH		NEW	NEW
		02-FEB-1996	18.5	PRESENT	GREATER TROCHANTER	BOTH			
		02-FEB-1996	18.5	PRESENT	DIAPHYSIS	BOTH			
		02-FEB-1996	18.5	PRESENT	PROXIMAL FEMORAL METAPHYSIS	BOTH			
		02-FEB-1996	18.5	PRESENT	CAPITAL FEMORAL EPIPHYSIS	BOTH			
		02-FEB-1996	18.5	PRESENT	DISTAL FEMORAL EPIPHYSIS	BOTH			
		02-FEB-1996	18.5	PRESENT	HIPS TO KNEES	BOTH			
		02-FEB-1996	18.5	PRESENT	DISTAL FEMORAL METAPHYSIS	BOTH			
		30-JUL-1997	36.4	PRESENT	HIPS TO KNEES	BOTH			

Manifestations	Assessment Method	Visit Date	Months Since 1st Infusion	Finding	Area of Involvement	Side (L/R)	Findings Since Comparison Date		
							Compare Date	Left Side	Right Side
MARROW INFILTRATION	MRI	30-JUL-1997	36.4	PRESENT	GREATER TROCHANTER	BOTH	02-FEB-1996	STABLE	STABLE
		30-JUL-1997	36.4	PRESENT	DIAPHYSIS	BOTH	02-FEB-1996	STABLE	STABLE
		30-JUL-1997	36.4	PRESENT	PROXIMAL FEMORAL METAPHYSIS	BOTH	02-FEB-1996	STABLE	STABLE
		15-MAY-1998	45.9	PRESENT	HIPS TO KNEES	BOTH	30-JUL-1997	STABLE	STABLE
		15-MAY-1998	45.9	PRESENT	PROXIMAL FEMORAL METAPHYSIS	BOTH	30-JUL-1997	STABLE	STABLE
		15-MAY-1998	45.9	PRESENT	DISTAL FEMORAL METAPHYSIS	BOTH	30-JUL-1997	STABLE	STABLE
LYTIC LESIONS	MRI	08-MAR-1993	-16.4	PRESENT					
		14-MAR-1993	-16.2	PRESENT					
ERLENMEYER FLASK DEFORMITY	MRI	08-MAR-1993	-16.4	PRESENT	FEMUR	UNKNOWN			
		14-MAR-1993	-16.2	PRESENT	FEMUR	UNKNOWN			

Table 4: Chronic Diseases

Date of Onset	Date Resolved	Prior To ERT	Category	Disease
26-MAR-1996		NO	OTHER DISEASES	ABDOMINAL PAIN

Table 5: Surgical Procedures

Date of Procedure	Prior to ERT	Procedure	Site	Location
No Values Available				