

Supplemental Table 6. Differences in the levels of lyso-Gb₃ and analogues in plasma and urine, and Gb₃ in urine at treatment-switching (pairwise analysis). Lyso-Gb₃ and analogue values are expressed as median (Q1-Q3). Interval values are expressed as mean (SD) and represent the time between the date of measurement and date of treatment. Lyso-Gb₃ (+50) data could not be analyzed due to many results below the limit of detection. LOD: limit of detection.

	switch from agalsidase-beta to agalsidase-alfa			switch from agalsidase-alfa to agalsidase-beta			switch from ERT to migalastat		
	before	after	p	before	after	p	before	after	p
Plasma (nM)									
n	18			5			9		
interval (years)*	0.79 (1.20)	0.82 (0.63)		0.40 (0.65)	1.08 (1.20)		0.06 (0.13)	0.92 (0.61)	
Lyso-Gb ₃ (-28)	0.12 (<lod -0.43)	0.14 (0.10-0.36)	0.14	0.11 (<lod -0.34)	<lod (<lod -0.06)	0.25	0.13 (0.04-0.43)	0.30 (0.25-0.38)	0.20
Lyso-Gb ₃ (-2)	2.70 (2.21-3.49)	2.82 (1.64-3.97)	0.13	2.94 (1.96-3.60)	1.80 (1.47-2.93)	0.19	2.80 (1.78-4.06)	3.21 (2.34-3.64)	0.50
Lyso-Gb ₃	10.49 (7.21-14.22)	9.68 (7.04-14.94)	0.20	9.05 (8.98-10.75)	6.96 (5.80-8.90)	0.06	9.31 (6.71-12.28)	9.30 (8.91-12.60)	0.25
Lyso-Gb ₃ (+16)	0.39 (0.22-0.79)	0.55 (<lod -1.06)	0.37	0.59 (0.23-1.05)	<lod (<lod -0.77)	0.25	0.42 (0.14-0.45)	0.48 (0.19-0.57)	0.30
Lyso-Gb ₃ (+18)	0.31 (<lod -0.37)	0.34 (0.16-0.94)	0.07	0.60 (0.26-0.79)	<lod (<lod -0.34)	0.06	0.42 (0.24-0.72)	0.64 (0.56-0.93)	0.04
Lyso-Gb ₃ (+34)	<lod (<lod -0.31)	<lod (<lod -0.47)	0.08	<lod (<lod -0.28)	<lod (<lod -0.21)	1.00	<lod (<lod -0.16)	0.34 (<lod -0.38)	0.13
Lyso-Gb ₃ (+50)	<lod (<lod -<lod)	<lod (<lod -<lod)	-	<lod (<lod -<lod)	<lod (<lod -<lod)	-	<lod (<lod -<lod)	<lod (<lod -<lod)	-
Urine (pM/mM cre)									
n	14			4			9		
interval (years)*	0.75 (0.93)	0.90 (0.63)		0.77 (0.48)	1.08 (1.20)		0.20 (0.42)	0.69 (0.37)	
Lyso-Gb ₃ (-28)	5.73 (1.98-11.32)	7.00 (<lod -17.64)	0.31	5.55 (0.37-13.34)	3.06 (0.40-7.38)	0.75	10.32 (6.01-12.41)	16.62 (7.55-23.23)	0.02
Lyso-Gb ₃ (-12)	48.93 (28.26-67.24)	46.99 (34.62-95.40)	0.19	43.71 (21.81-96.81)	33.12 (24.20-39.71)	0.63	48.41 (30.95-58.26)	48.76 (46.26-75.05)	0.16
Lyso-Gb ₃ (-2)	6.90 (<lod -14.88)	9.94 (<lod -21.37)	0.18	8.34 (1.17-17.69)	3.78 (2.07-12.41)	0.88	16.47 (10.10-19.09)	16.49 (8.88-30.25)	0.65
Lyso-Gb ₃	10.50 (<lod -17.62)	10.04 (1.98-22.97)	0.47	7.67 (4.55-18.30)	8.18 (4.90-23.46)	0.63	10.29 (3.98-21.35)	8.60 (3.11-12.83)	0.36
Lyso-Gb ₃ (+14)	13.53 (10.05-21.36)	13.72 (7.94-31.17)	0.39	11.19 (4.05-30.12)	11.48 (8.07-19.53)	0.88	13.08 (11.08-25.10)	15.06 (13.10-21.00)	0.82
Lyso-Gb ₃ (+16)	156.03 (99.81-220.87)	146.44 (74.18-251.77)	0.81	89.61 (63.52-186.13)	92.25 (80.34-134.64)	1.00	131.15 (81.39-196.69)	167.63 (105.80-168.99)	0.50
Lyso-Gb ₃ (+34)	61.72 (46.39-126.17)	51.08 (42.42-121.16)	0.72	58.97 (48.78-95.76)	43.74 (35.85-77.63)	0.63	52.34 (44.26-89.31)	77.46 (66.86-100.55)	0.04
Lyso-Gb ₃ (+50)	41.34 (32.54-90.63)	49.47 (27.30-82.66)	0.81	53.03 (36.95-60.41)	50.69 (31.05-81.21)	0.88	37.70 (31.96-61.32)	50.41 (37.51-63.02)	0.42
Gb₃ (µg/mmol creatinine)									
	11.03 (7.76-16.41)	13.98 (8.10-25.48)	0.36	8.63 (4.98-15.55)	5.19 (4.91-10.01)	0.63	7.50 (5.46-13.54)	28.14 (13.81-44.79)	0.007