

Supplementary Material**Supplementary Methods****Clinical Scoring**

Patients were analyzed according to a previously established clinical scoring system that assessed the degree of regression of vision, motor function, language ability, and intellect as well as the frequency of Grand Mal seizures (1). Scoring assessments were made retrospectively using patient charts, patient questionnaires, and detailed interviews of parents and care givers, as well as prospectively in 6-month intervals. Clinical scoring was assessed as previously described for vision, intellect, language, and motor function as follows: normal function (3); minor but readily recognized dysfunction (2); severe dysfunction (1); total loss of function (0) (1). Because the frequency of seizures of CLN3 patients can vary due to different anticonvulsive medications, the epilepsy scoring was not used for description and classification of different disease courses of the 25 patients in this study. Scoring assessments for each patient were conducted and counter-checked independently by two child neurologists at the NCL specialty clinic. Scores assigned by both child neurologists for each patient were identical, indicating that the scoring data can be obtained and dealt with reliably.

Calculation of Index of Relative Severity

For each patient studied an Index of Relative Severity of the disease was calculated as previously described (1). In this study, however, instead of 1-year periods of observation, 6-month periods of observation were used for the calculation:

$$\text{Index of Relative Severity} = \frac{\sum \text{deviations of Total Scores from median for each 6-month period of observation}}{\text{total number of 6-month periods of observation}}$$

Lebrun et al.

Biomarkers and Modifiers of CLN3 Disease

This index can have positive or negative values. Thus, an index of less than -0.5 corresponds to a severe disease phenotype, and an index between -0.5 and 0.5 defines a “classical” disease phenotype whereas an index higher than 0.5 characterizes a mild disease phenotype. The index was calculated for each of our patients for up to a maximum of fifty 6-month periods of observation (= 25 years) including patients #3 and #4 (Table 2) whose scoring assessments covered more than 25 years.

Microarray Analysis

All experiments were performed using Affymetrix human genome GeneChip U133 Plus 2.0 (Affymetrix, Santa Clara, USA). RNA derived from lymphocytes of 15 different individuals (8 CLN3-patients homozygous for the 1 kb deletion and 7 healthy controls) was used for hybridization. In brief, 100 ng total RNA were used for first strand cDNA-synthesis (Two-Cycle Target Labeling Kit; Affymetrix) according to the GeneChip Expression Analysis Technical Manual. Synthesis of biotin-labeled cRNA was carried out using the IVT Labeling Kit (Affymetrix) and cleaned up (Sample Cleanup Module, Affymetrix). For hybridization, 15 µg of fragmented cRNA were incubated with the chip in 200 µl of hybridization solution in Hybridization Oven 640 (Affymetrix) at 45°C for 16 hours. GeneChips were then washed and stained using the Affymetrix Fluidics Station 450 according to the GeneChip Expression Analysis Technical Manual. Microarrays were scanned with the Affymetrix GeneChip Scanner 7G, and the signals were processed using the GeneChip expression analysis algorithm (version 2, Affymetrix). To compare samples and experiments, the trimmed mean signal of each array was scaled to a target intensity of 200.

Analysis of the data set groups with high statistical power was hampered by the small number of samples due to the rareness of the disease and the requirement of CLN3 patients without any medication. To compensate for the low number of suitable samples we chose an

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

experiment design with well defined controls. The control sample selection should gain a preanalytical reduction of degrees of freedom, thus reducing biological background variance. Therefore, each case sample was assigned to a defined control sample that matched in age and gender, two factors which are suspected to have major impact on the phenotype. In the subsequent analysis, paired comparisons between case and control samples were performed with the Affymetrix MAS software (version 5.0, Affymetrix) using default parameters. Wilcoxon's signed rank tests were used to compute change p -values from the perfect match/mismatch signals. For an increase or decrease call the p -value had to be less than 0.003. For the slow and rapid disease progression groups which consisted of merely two pairs an additional cross validation between case and control samples were calculated. Thus, for each group (rapid/slow/average) $n = 4$ comparisons were carried out. Genes were regarded as differentially expressed if they exhibited a minimal absolute fold change of 1.7 and were assigned a dysregulation in the same direction (increase / decrease call by the algorithm) in at least three of the four comparisons. Annotations were further analyzed with interactive query analysis at www.affymetrix.com. Pathways and other functional groupings of genes were evaluated for differential regulation using GenMAPP software (UCSF) as described previously (2,3).

Real-time (RT) PCR

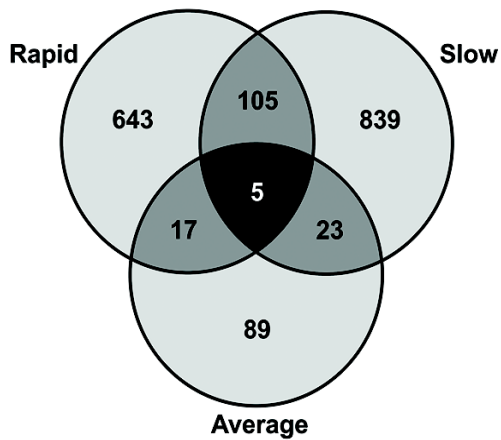
For RT-PCR, TaqMan[®] Gene Expression Assays (Applied Biosystems, Darmstadt, Germany) including pre-designed probes and primer sets for human β -actin (*ACTB*) (Hs99999903_m1), ribosomal protein s18 (*RPS18*) (Hs01375212_g1), *CLN3* (Hs00164002_m1), *DUSP2* (Hs00358879_m1), *CDC42SE2* (Hs00184113_m1), *RAPGEF1* (Hs00178409_m1), *SPIB* (Hs00162150_m1), *MARCKS* (Hs00158993_m1), *BACE2* (Hs00273238_m1), *FLJ10357* (Hs00399913_m1) and mouse β -actin (*Actb*) (Mm00607939_s1), *Cln3* (Mm00487021_m1) and *Dusp2* (Mm00839675_g1) were used. *ACTB* was chosen as the control house-keeping

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease gene in HeLa cells and in SH-SY5Y cells, *RPS18* was chosen as house-keeping gene in lymphocytes. *Actb* was used as house-keeping gene in Cb*Cln3*^{Δex7/8} cerebellar precursor cells. RT-PCR reactions were performed as previously described, using a final volume of 20 μl containing 2 μl template cDNA (4). Quantitation was performed with the Mx3000P™ QPCR System from Stratagene (Amsterdam, Netherlands). The relative expression of mRNA of interest was normalized to the amount of *ACTB* mRNA in the same cDNA using the comparative CT method ($2^{-\Delta\Delta CT}$). For statistical analysis Student's t-test was performed. *p*-values < 0.05 were considered significant.

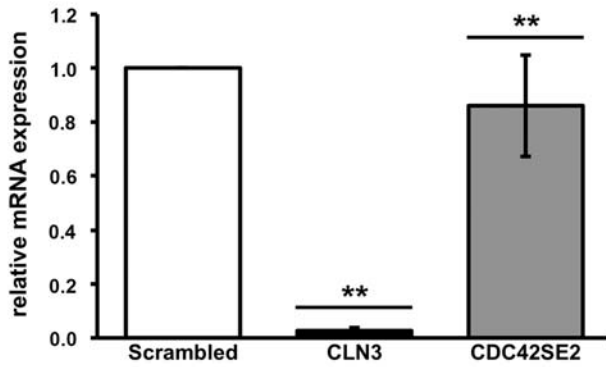
References to Supplementary Methods

1. Kohlschütter A, Laabs R, Albani M (1988) Juvenile neuronal ceroid lipofuscinosis (JNCL): quantitative description of its clinical variability. *Acta Paediatr Scand* 77:867-872.1.
2. Bonner AE, Lemon WJ, You M (2003) Gene expression signatures identify novel regulatory pathways during murine lung development: implications for lung tumorigenesis. *J Med Genet* 40:408-417.
3. Doniger SW, Salomonis N, Dahlquist KD, Vranizan K, Lawlor SC, et al. (2003) MAPPFinder: using Gene Ontology and GenMAPP to create a global gene-expression profile from microarray data. *Genome Biol* 4:R7.
4. Pohl S, Mitchison HM, Kohlschütter A, van Diggelen O, Braulke T, et al. (2007) Increased expression of lysosomal acid phosphatase in CLN3-defective cells and mouse brain tissue. *J Neurochem* 103:2177-2188.

Supplementary Figures



Suppl. Figure 1. Number of dysregulated genes in lymphocytes of CLN3 patients with rapid, average, and slow progression of the disease. RNA microarray analyses were performed in CLN3 patients with rapid ($n = 2$), average ($n = 4$), and slow ($n = 2$) disease progression. The total number of dysregulated genes in the patients in comparison with healthy age and gender matched controls are indicated, and the numbers that were found to be dysregulated in patients with rapid and slow (105), rapid and average (17), or slow and average (23) progression of the disease are given in the overlapping areas. Five genes were commonly up- or down-regulated in all CLN3 patients studied.



Suppl. Figure 2. Relative mRNA expression of *CDC42SE2* in siRNA-mediated acutely *CLN3*-depleted cells. HeLa cells were transfected with *CLN3* siRNA for 72 h in total. The relative mRNA expressions of candidate genes were measured by RT-PCR and normalized to *ACTB* expression. mRNA levels of scrambled siRNA-treated cells (white bar) were set as 1 and used as control. The data are the mean \pm SD of five independent transfection experiments; ** $p < 0.01$ compared with controls.

CDC42SE2 regulates the signaling pathway of the Rho GTPase CDC42 leading to altered local actin dynamics (1). Cdc42 is one of the physiological key players for membrane trafficking, axon growth and neuronal polarization (2,3). In acutely *CLN3*-depleted HeLa cells the *CDC42SE2* expression was slightly reduced by 10 to 40%. The role of *CDC42SE2* in the regulation of actin dynamics needs to be investigated further at the protein level, e.g. both in non-polarized *CLN3*-depleted HeLa cells and in polarized *Cln3*-deficient neuronal mouse cells.

References to Supplementary Figures

1. Pirone DM, Fukuhara S, Gutkind JS, Burbelo PD (2000) SPECs, small binding proteins for Cdc42. *J Biol Chem* 275:22650-22656.
2. Ng J, Luo L (2004) Rho GTPases regulate axon growth through convergent and divergent signaling pathways. *Neuron* 44:779-793.
3. Witte H, Bradke F (2008) The role of the cytoskeleton during neuronal polarization. *Curr Opin Neurobiol* 18:479-487.

Supplementary Tables

Suppl. Table 1. Gene ontology categorization of genes dysregulated in CLN3 patients with average disease progression

Gene Symbol	Gene Name	Fold Change*
NEURON, BRAIN, NERVOUS SYSTEM		
<i>NR4A2</i>	nuclear receptor subfamily 4, group A, member 2	2.9
<i>ROBO1</i>	roundabout, axon guidance receptor, homolog 1 (Drosophila)	2.4
<i>SNF1LK</i>	SNF1-like kinase	2.1
<i>ARHGEF10</i>	Rho guanine nucleotide exchange factor (GEF) 10	2.0
<i>ZEB2</i>	zinc finger E-box binding homeobox 2	2.0
<i>e.s.t.**</i>	full-length cDNA clone CS0DB008YK14 of Neuroblastoma Cot 10-normalized of Homo sapiens (human)	1.9
PROTEOLYSIS, LYSOSOME		
<i>ARTS-1</i>	type 1 tumor necrosis factor receptor shedding aminopeptidase regulator	-2.4
<i>LYZ</i>	lysozyme (renal amyloidosis)	-2.1
<i>ST14</i>	suppression of tumorigenicity 14 (colon carcinoma)	-2.0
<i>XPNPEP3</i>	X-prolyl aminopeptidase (aminopeptidase P) 3, putative	-1.8
<i>ASAH1</i>	N-acylsphingosine amidohydrolase (acid ceramidase)-like	-1.8
<i>DPP4</i>	dipeptidyl-peptidase 4 (CD26, adenosine deaminase complexing protein 2)	-1.8
ENDOCYTOSIS, PROTEIN TRANSPORT, AUTOPHAGY, ENDOSOME		
<i>TMED6</i>	transmembrane emp24 protein transport domain containing 6	1.9
APOPTOSIS		
<i>IL1B</i>	interleukin 1, beta	3.4
<i>LOC728613</i>	programmed cell death protein 6-like	3.4
<i>NLRP2</i>	NLR family, pyrin domain containing 2	2.0
<i>AMFR</i>	autocrine motility factor receptor	1.9
<i>TP53RK</i>	TP53 regulating kinase	-1.8
<i>CYCS</i>	cytochrome c, somatic	1.7
<i>NAIP</i>	NLR family, apoptosis inhibitory protein	1.7

VISUAL PERCEPTION, EYE		
<i>TACSTD2</i>	tumor-associated calcium signal transducer 2	1.4
BLOOD, VASCULAR		
<i>ALAS2</i>	aminolevulinate, delta-, synthase 2 (sideroblastic/hypochromic anemia)	-12.6
<i>HBM</i>	hemoglobin, mu	-3,5
<i>HBB</i>	hemoglobin, beta	-2.4
<i>HBA2</i>	hemoglobin, alpha 2	-1.9
<i>HBD</i>	hemoglobin, delta	-1.8
TRANSCRIPTION, RNA TRANSPORT, DNA INTEGRATION		
<i>SOD2</i>	superoxide dismutase 2, mitochondrial	5.8
<i>ZNF683</i>	zinc finger protein 683	2.2
<i>HBP1</i>	HMG-box transcription factor 1	2.2
<i>NXF3</i>	nuclear RNA export factor 3	2.2
<i>CENPK</i>	centromere protein K	2.0
<i>KIAA1466</i>	KIAA1466 gene	1.9
<i>ZNF124</i>	zinc finger protein 124	1.9
<i>LOC441259</i> /// <i>LOC730323</i> /// <i>POLR2J2</i>	DNA directed RNA polymerase II polypeptide J-related /// similar to postmeiotic segregation increased 2-like 2	-1.7
<i>PARP15</i>	poly (ADP-ribose) polymerase family, member 15	1.7
IMMUNE RESPONSE		
<i>CCL3</i> /// <i>CCL3L1</i> /// <i>CCL3L3</i> /// <i>LOC728830</i> /// <i>LOC730422</i>	chemokine (C-C motif) ligand 3 /// chemokine (C-C motif) ligand 3-like 1 /// chemokine (C-C motif) ligand 3-like 3 /// similar to Small inducible cytokine A3-like 1 precursor (Tonsillar lymphocyte LD78 beta protein) (LD78-beta(1-70)) (G0/G1 switch regulatory protein 19-2) (G0S19-2 protein) (PAT 464.2) /// similar to chemokine (C-C motif) ligand 3-like 3	4.6
<i>e.s.t.**</i>	T-cell receptor beta (TCRB) mRNA (HLA-A3, 29; B7, 44; DR2, 7)	3.0
<i>RGS1</i>	regulator of G-protein signalling 1	2.5
<i>CD83</i>	CD83 molecule	2.5
<i>B2M</i>	beta-2-microglobulin	2.3
<i>PF4V1</i>	platelet factor 4 variant 1	2.2

<i>FCER2</i>	Fc fragment of IgE, low affinity II, receptor for (CD23)	2.1
<i>IGHD</i>	immunoglobulin heavy constant delta	2.0
<i>FCRL5</i>	Fc receptor-like 5	1.9

CYTOSKELETON ORGANIZATION, CELL ADHESION, CELL SURFACE

<i>PTPRK</i>	protein tyrosine phosphatase, receptor type, K	5.6
<i>ANKRD42</i>	Ankyrin repeat domain 42	-2.5
<i>TUBB2A</i>	tubulin, beta 2A	2.0
<i>SDK2</i>	sidekick homolog 2 (chicken)	2.3
<i>FGD2</i>	FYVE, RhoGEF and PH domain containing 2	-1.9
<i>PELO</i>	pelota homolog (Drosophila)	-1.8
<i>MYOM2</i>	myomesin (M-protein) 2, 165kDa	-1.8

CELL CYCLE

<i>RASGEF1B</i>	RasGEF domain family, member 1B	4.3
<i>DUSP2</i>	dual specificity phosphatase 2	2.4
<i>MCC</i>	mutated in colorectal cancers	2.2
<i>CCND3</i>	cyclin D3	1.9
<i>CDC42SE2</i>	CDC42 small effector 2	-1.8

UNKNOWN

<i>PDZK1IP1</i>	PDZK1 interacting protein 1	4.9
<i>KIAA1276</i>	KIAA1276 protein	-4.0
<i>e.s.t.**</i>	transcribed locus	3.1
<i>e.s.t.**</i>	mRNA; cDNA DKFZp762I0915 (from clone DKFZp762I0915)	-2.9
<i>e.s.t.**</i>	full length insert cDNA clone ZD45C02	-2.8
<i>e.s.t.**</i>	transcribed locus, strongly similar to XP_224329.4 similar to p30 DBC protein [Rattus norvegicus]	-2.8
<i>FAM19A1</i>	family with sequence similarity 19 (chemokine (C-C motif)-like), member A1	-2.2
<i>e.s.t.**</i>	transcribed locus	2.1
<i>e.s.t.**</i>	transcribed locus	2.1
<i>LOC283788</i>	hypothetical protein LOC283788	-2.1

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

<i>e.s.t.**</i>	transcribed locus	2.0
<i>e.s.t.**</i>	transcribed locus	2.0
<i>e.s.t.**</i>	hypothetical gene supported by AL713796	2.0
<i>LOC202134</i>	hypothetical protein LOC202134	-1.9
<i>e.s.t.**</i>	mRNA; cDNA DKFZp686D0673 (from clone DKFZp686D0673)	-1.9
<i>e.s.t.**</i>	mRNA; cDNA DKFZp686A22111 (from clone DKFZp686A22111)	-1.9
<i>XAF1</i>	XIAP associated factor-1	-1.9
<i>e.s.t.**</i>	transcribed locus	1.8
<i>e.s.t.**</i>	transcribed locus, strongly similar to XP_001150530.1 hypothetical protein [Pan troglodytes]	1.8
<i>LOC654433</i>	hypothetical LOC654433	-1.8
<i>LOC650392</i>	hypothetical protein LOC650392	-1.8
<i>e.s.t.**</i>	transcribed locus	1.7
<i>e.s.t.**</i>	transcribed locus	1.7
<i>e.s.t.**</i>	transcribed locus	1.7
<i>e.s.t.**</i>	transcribed locus	-1.7
<i>LOC389831</i>	transcribed locus	-1.7
<i>CCDC90B</i>	coiled-coil domain containing 90B	-1.7
LIPID METABOLISM		
<i>SULT1A1</i>	sulfotransferase family, cytosolic, 1A, phenol-preferring, member 1	-1.7
<i>EBPL</i>	emopamil binding protein-like	-1.7
TRANSPORT		
<i>KCNJ2</i>	potassium inwardly-rectifying channel, subfamily J, member 2	-2.2
<i>SLC25A29</i>	solute carrier family 25, member 29	1.7

* dysregulation shown as fold-change compared to healthy controls

** expressed sequence tags

Suppl Table 2A. Gene ontology categorization of genes dysregulated in CLN3 patients with rapid disease progression

Gene Symbol	Gene Name	Fold Change*
NEURON, BRAIN, NERVOUS SYSTEM		
<i>S100B</i>	S100 calcium binding protein B	-10.1
<i>FUT10</i>	fucosyltransferase 10 (alpha (1,3) fucosyltransferase)	3.0
<i>EGR2</i>	early growth response 2 (Krox-20 homolog, Drosophila)	2.4
<i>FOS</i>	v-fos FBJ murine osteosarcoma viral oncogene homolog	2.4
<i>GPR56</i>	G protein-coupled receptor 56	2.3
<i>GPR114</i>	G protein-coupled receptor 114	2.2
<i>SMN1</i> /// <i>SMN2</i>	survival of motor neuron 1, telomeric /// survival of motor neuron 2, centromeric	-2.1
<i>ROBO1</i>	roundabout, axon guidance receptor, homolog 1 (Drosophila)	-2.1
<i>NTN2L</i>	Netrin 2-like (chicken)	-2.0
<i>CNTNAP2</i>	contactin associated protein-like 2	-2.0
<i>SLC18A2</i>	solute carrier family 18 (vesicular monoamine), member 2	-2.0
<i>EGR3</i>	early growth response 3	2.0
<i>CDKN1C</i>	Cyclin-dependent kinase inhibitor 1C (p57, Kip2)	1.8
<i>COLQ</i>	collagen-like tail subunit (single strand of homotrimer) of asymmetric acetylcholinesterase	1.8
<i>NOG</i>	Noggin	-1.8
<i>PTGDS</i>	prostaglandin D2 synthase 21kDa (brain)	1.8
<i>CD97</i>	CD97 molecule	1.8
<i>P2RX5</i>	purinergic receptor P2X, ligand-gated ion channel, 5	1.8
<i>YWHAH</i>	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, eta polypeptide	-1.7
PROTEOLYSIS, LYSOSOME		
<i>ARSA</i>	arylsulfatase A	5.1
<i>CTSW</i>	cathepsin W	3.5
<i>MGC52282</i>	hypothetical locus MGC52282	2.5
<i>GGH</i>	gamma-glutamyl hydrolase (conjugase, foylpolypolyglutamyl hydrolase)	-2.5

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

<i>DPEP3</i>	dipeptidase 3	2.4
<i>ADAM10</i>	ADAM metallopeptidase domain 10	-2.3
<i>e.s.t**</i>	cDNA clone IMAGE:5216666	2.2
<i>LNPEP</i>	leucyl/cystinyl aminopeptidase	-2.1
<i>BACE2</i>	beta-site APP-cleaving enzyme 2	2.1
<i>CAPN7</i>	calpain 7	-1.9
<i>SOLH</i>	small optic lobes homolog (Drosophila)	1.9
<i>PSMA1</i>	proteasome (prosome, macropain) subunit, alpha type, 1	-1.8
<i>PIGK</i>	phosphatidylinositol glycan anchor biosynthesis, class K	-1.8
<i>PLAUR</i>	plasminogen activator, urokinase receptor	1.8
<i>PCSK5</i>	proprotein convertase subtilisin/kexin type 5	-1.7
<i>ADAM8</i>	ADAM metallopeptidase domain 8	1.7
<i>AGBL3</i>	ATP/GTP binding protein-like 3	-1.7
<i>NLN</i>	neurolysin (metallopeptidase M3 family)	-1.7
<i>CTSK</i>	cathepsin K	1.7

ENDOCYTOSIS, PROTEIN TRANSPORT, AUTOPHAGY, ENDOSOME

<i>IGHM</i>	immunoglobulin heavy constant mu	-4.3
<i>IGH /// IGHG1 /// IGHG2 /// IGHG3 /// IGHM /// IGHV4-31</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-3.1
<i>EXOC7 /// IGHA1 /// IGHD /// IGHG1 /// IGHM /// IGHV4-31 /// IL8 /// ZCWPW2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-3.1
<i>IGHA1 /// IGHG1 /// IGHG3 /// IGHM /// IGHV4-31</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-2.8
<i>SYTL4</i>	synaptotagmin-like 4 (granophilin-a)	-2.6
<i>SEC61A1</i>	Sec61 alpha 1 subunit (S. cerevisiae)	-2.5

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

<i>CENTG2</i>	centaurin, gamma 2	2.3
<i>MYO6</i>	myosin VI	2.3
<i>ATG16L1</i>	ATG16 autophagy related 16-like 1 (<i>S. cerevisiae</i>)	-2.2
<i>EXOC7</i> /// <i>IGH</i> /// <i>IGHA1</i> /// <i>IGHA2</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHG2</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>RAC1</i> /// <i>SIX6</i> /// <i>ZCWPW2</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant alpha 2 (A2m marker) /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// <i>SIX</i> homeobox 6 /// ras-related C3 botulinum toxin substrate 1 (rho family, small GTP binding protein Rac1) /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-2.1
<i>IGH</i> /// <i>IGHG1</i> /// <i>IGHM</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu	2.1
<i>CLEC4F</i>	C-type lectin domain family 4, member F	-2.1
<i>SYTL2</i>	synaptotagmin-like 2	2.0
<i>LOC144571</i>	hypothetical protein LOC144571	1.9
<i>EXOC6</i>	exocyst complex component 6	-1.9
<i>TMED2</i>	transmembrane emp24 domain trafficking protein 2	-1.9
<i>PLDN</i>	pallidin homolog (mouse)	-1.9
<i>PEX13</i>	peroxisome biogenesis factor 13	-1.9
<i>TLK1</i>	tousled-like kinase 1	-1.9
<i>DNM3</i>	dynamamin 3	-1.9
<i>ARL17</i>	ADP-ribosylation factor-like 17	-1.9
<i>SNX11</i>	sorting nexin 11	1.9
<i>SDAD1</i>	SDA1 domain containing 1	-1.8
<i>KPNA3</i>	karyopherin alpha 3 (importin alpha 4)	-1.8
<i>AP3D1</i>	adaptor-related protein complex 3, delta 1 subunit	1.8
<i>ARF4</i>	ADP-ribosylation factor 4	-1.8
<i>AQP3</i>	aquaporin 3 (Gill blood group)	-1.8
<i>ADRB2</i>	adrenergic, beta-2-, receptor, surface	1.8
<i>MAMDC4</i>	MAM domain containing 4	1.8
<i>RAB34</i>	RAB34, member RAS oncogene family	1.7

<i>PTP4A1</i>	protein tyrosine phosphatase type IVA, member 1	-1.7
<i>PLA2G4B</i>	phospholipase A2, group IVB (cytosolic)	1.7
<i>RAB11FIP4</i>	RAB11 family interacting protein 4 (class II)	1.7
<i>SNX19</i>	sorting nexin 19	1.7

APOPTOSIS

<i>IGFBP3</i>	insulin-like growth factor binding protein 3	5.2
<i>NLRP2</i>	NLR family, pyrin domain containing 2	4.2
<i>NAIP</i>	NLR family, apoptosis inhibitory protein	4.0
<i>IL1B</i>	interleukin 1, beta	3.7
<i>TOP2A</i>	topoisomerase (DNA) II alpha 170kDa	-3.4
<i>TXNDC5</i>	thioredoxin domain containing 5	-3.1
<i>COL4A3</i>	collagen, type IV, alpha 3 (Goodpasture antigen)	3.0
<i>CD38</i>	CD38 molecule	-2.3
<i>CDC2</i>	cell division cycle 2, G1 to S and G2 to M	-2.8
<i>BUB1B</i>	BUB1 budding uninhibited by benzimidazoles 1 homolog beta (yeast)	-2.7
<i>IRF5</i>	interferon regulatory factor 5	2.3
<i>PHLDA1</i>	pleckstrin homology-like domain, family A, member 1	2.6
<i>IL6</i>	interleukin 6 (interferon, beta 2)	2.6
<i>GZMH</i>	granzyme H (cathepsin G-like 2, protein h-CCPX)	2.5
<i>STEAP3</i>	STEAP family member 3	2.2
<i>PDCD4</i>	programmed cell death 4 (neoplastic transformation inhibitor)	-2.2
<i>ABCA2</i>	ATP-binding cassette, sub-family A (ABC1), member 2	2.1
<i>TXNDC1</i>	thioredoxin domain containing 1	-2.0
<i>SERPINB9</i>	serpin peptidase inhibitor, clade B (ovalbumin), member 9	2.0
<i>EP300</i>	E1A binding protein p300	-2.0
<i>MRPL41</i>	mitochondrial ribosomal protein L41	1.9
<i>GZMA</i>	granzyme A (granzyme 1, cytotoxic T-lymphocyte-associated serine esterase 3)	1.9
<i>SNCA</i>	synuclein, alpha (non A4 component of amyloid precursor)	-1.9
<i>TRIB3</i>	tribbles homolog 3 (Drosophila)	1.8

<i>CASP2</i>	caspace 2, apoptosis-related cysteine peptidase (neural precursor cell expressed, developmentally down-regulated 2)	-1.8
<i>ALOX12</i>	arachidonate 12-lipoxygenase	-1.8
<i>LOC650083</i> /// <i>YWHAZ</i>	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, zeta polypeptide /// similar to tyrosine 3/tryptophan 5 - monooxygenase activation protein, zeta polypeptide	-1.8
<i>PYHIN1</i>	pyrin and HIN domain family, member 1	1.8
<i>AIFM1</i>	apoptosis-inducing factor, mitochondrion-associated, 1	-1.7
<i>APPBP1</i>	amyloid beta precursor protein binding protein 1	-1.7
<i>NFKBIE</i>	nuclear factor of kappa light polypeptide gene enhancer in B-cells inhibitor, epsilon	1.7
<i>CD14</i>	CD14 molecule	1.7
VISUAL PERCEPTION, EYE		
<i>CRYBB2</i>	crystallin, beta B2	2.9
<i>CYP1B1</i>	cytochrome P450, family 1, subfamily B, polypeptide 1	2.5
<i>ABLIM1</i>	actin binding LIM protein 1	-1.8

* dysregulation shown as fold-change compared to healthy controls

** expressed sequence tags

Suppl. Table 2B. Genes dysregulated in CLN3 patients with rapid disease progression

Probe Set ID	Gene Symbol	Gene Name	Fold Change*
236710_at	<i>C1orf87</i>	chromosome 1 open reading frame 87	26.9
231236_at	<i>ZFP57</i>	zinc finger protein 57 homolog (mouse)	21.5
213482_at	<i>DOCK3</i>	dedicator of cytokinesis 3	12.6
218824_at	<i>FLJ10781</i>	hypothetical protein FLJ10781	7.5
233355_at	<i>PRR17</i>	proline rich 17	7.5
206371_at	<i>FOLR3</i>	folate receptor 3 (gamma)	7.0
220014_at	<i>PRR16</i>	proline rich 16	5.3
210095_s_at	<i>IGFBP3</i>	insulin-like growth factor binding protein 3	5.2
204443_at	<i>ARSA</i>	arylsulfatase A	5.1
214247_s_at	<i>DKK3</i>	dickkopf homolog 3 (<i>Xenopus laevis</i>)	4.4
235937_at	<i>LOC647859</i>	occludin pseudogene	4.4
221690_s_at	<i>NLRP2</i>	NLR family, pyrin domain containing 2	4.2
239944_at	<i>NAIP</i>	NLR family, apoptosis inhibitory protein	4.0
205067_at	<i>IL1B</i>	interleukin 1, beta	3.7
229797_at	<i>MCOLN3</i>	mucolipin 3	3.7
214450_at	<i>CTSW</i>	cathepsin W	3.5
239287_at	---	Transcribed locus	3.5
1569608_x_at	<i>LOC643187</i>	Similar to ankyrin repeat domain 20A	3.4
201681_s_at	<i>DLG5</i>	discs, large homolog 5 (<i>Drosophila</i>)	3.3
1555579_s_at	<i>PTPRM</i>	protein tyrosine phosphatase, receptor type, M	3.3
231776_at	<i>EOMES</i>	eomesodermin homolog (<i>Xenopus laevis</i>)	3.2
206522_at	<i>MGAM</i>	maltase-glucoamylase (alpha-glucosidase)	3.2
230563_at	<i>RASGEF1A</i>	RasGEF domain family, member 1A	3.2
1569954_at	---	Homo sapiens, Similar to AD038, clone IMAGE:3838464, mRNA	3.1
244434_at	---	---	3.1
205987_at	<i>CD1C</i>	CD1c molecule	3.0
235472_at	<i>FUT10</i>	fucosyltransferase 10 (alpha (1,3) fucosyltransferase)	3.0
207067_s_at	<i>HDC</i>	histidine decarboxylase	3.0
230802_at	<i>ARHGAP24</i>	Rho GTPase activating protein 24	3.0
213268_at	<i>CAMTA1</i>	calmodulin binding transcription activator 1	3.0
222073_at	<i>COL4A3</i>	collagen, type IV, alpha 3 (Goodpasture antigen)	3.0
201694_s_at	<i>EGR1</i>	early growth response 1	3.0
205819_at	<i>MARCO</i>	macrophage receptor with collagenous structure	3.0
207840_at	<i>CD160</i>	CD160 molecule	2.9
206777_s_at	<i>CRYBB2</i>	crystallin, beta B2	2.9
243231_at	<i>FLJ39822</i>	hypothetical protein FLJ39822	2.9

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

230110_at	<i>MCOLN2</i>	mucolipin 2	2.9
203535_at	<i>S100A9</i>	S100 calcium binding protein A9	2.9
235221_at	<i>CBLN3</i>	cerebellin 3 precursor	2.8
214085_x_at	<i>GLIPR1</i>	GLI pathogenesis-related 1 (glioma)	2.8
1555745_a_at	<i>LYZ</i>	lysozyme (renal amyloidosis)	2.8
210395_x_at	<i>MYL4</i>	myosin, light chain 4, alkali; atrial, embryonic	2.8
204748_at	<i>PTGS2</i>	prostaglandin-endoperoxide synthase 2 (prostaglandin G/H synthase and cyclooxygenase)	2.8
211597_s_at	<i>HOP</i>	homeodomain-only protein	2.8
227929_at	---	CDNA clone IMAGE:5277945	2.7
209570_s_at	<i>D4S234E</i>	DNA segment on chromosome 4 (unique) 234 expressed sequence	2.7
227819_at	<i>LGR6</i>	leucine-rich repeat-containing G protein-coupled receptor 6	2.7
41644_at	<i>SASH1</i>	SAM and SH3 domain containing 1	2.7
227083_at	<i>B3GALTL</i>	beta 1,3-galactosyltransferase-like	2.7
1560762_at	<i>LOC285972</i>	hypothetical protein LOC285972	2.7
206366_x_at	<i>XCL2</i>	chemokine (C motif) ligand 2	2.6
207072_at	<i>IL18RAP</i>	interleukin 18 receptor accessory protein	2.6
202826_at	<i>SPINT1</i>	serine peptidase inhibitor, Kunitz type 1	2.6
228264_at	<i>PHACS</i>	1-aminocyclopropane-1-carboxylate synthase	2.6
204103_at	<i>CCL4</i>	chemokine (C-C motif) ligand 4	2.6
205207_at	<i>IL6</i>	interleukin 6 (interferon, beta 2)	2.6
217996_at	<i>PHLDA1</i>	pleckstrin homology-like domain, family A, member 1	2.6
203940_s_at	<i>VASH1</i>	vasohibin 1	2.6
220122_at	<i>MCTP1</i>	multiple C2 domains, transmembrane 1	2.5
213849_s_at	<i>PPP2R2B</i>	protein phosphatase 2 (formerly 2A), regulatory subunit B, beta isoform	2.5
204351_at	<i>S100P</i>	S100 calcium binding protein P	2.5
210321_at	<i>GZMH</i>	granzyme H (cathepsin G-like 2, protein h-CCPX)	2.5
231638_at	<i>MGC52282</i>	hypothetical locus MGC52282	2.5
227554_at	---	MRNA; cDNA DKFZp686I18116 (from clone DKFZp686I18116)	2.5
204731_at	<i>TGFBR3</i>	transforming growth factor, beta receptor III	2.5
204141_at	<i>TUBB2A</i>	tubulin, beta 2A	2.5
243189_at	---	---	2.5
214567_s_at	<i>XCLI</i> /// <i>XCL2</i>	chemokine (C motif) ligand 1 /// chemokine (C motif) ligand 2	2.5
205114_s_at	<i>CCL3</i> /// <i>CCL3L1</i> /// <i>CCL3L3</i> /// <i>LOC728830</i> /// <i>LOC730422</i>	chemokine (C-C motif) ligand 3 /// chemokine (C-C motif) ligand 3-like 1 /// chemokine (C-C motif) ligand 3-like 3 /// similar to Small inducible cytokine A3-like 1 precursor (Tonsillar lymphocyte LD78 beta protein) (LD78-beta(1-70)) (G0/G1 switch regulatory protein 19-2) (G0S19-2 protein) (PAT 464.2) /// similar to chemokine (C-C motif) ligand 3-like 3	2.5
242557_at	<i>C6orf12</i>	Chromosome 6 open reading frame 12	2.5
202437_s_at	<i>CYP1B1</i>	cytochrome P450, family 1, subfamily B, polypeptide 1	2.5
201041_s_at	<i>DUSP1</i>	dual specificity phosphatase 1	2.5

219093_at	<i>PIDI</i>	phosphotyrosine interaction domain containing 1	2.5
237035_at	---	Transcribed locus	2.5
230967_s_at	---	Transcribed locus	2.5
205249_at	<i>EGR2</i>	early growth response 2 (Krox-20 homolog, Drosophila)	2.4
243578_at	---	Transcribed locus	2.4
231860_at	<i>BRWD1</i>	bromodomain and WD repeat domain containing 1	2.4
228372_at	<i>C10orf128</i>	chromosome 10 open reading frame 128	2.4
220179_at	<i>DPEP3</i>	dipeptidase 3	2.4
209189_at	<i>FOS</i>	v-fos FBJ murine osteosarcoma viral oncogene homolog	2.4
229497_at	<i>ANKDDIA</i>	ankyrin repeat and death domain containing 1A	2.3
205568_at	<i>AQP9</i>	aquaporin 9	2.3
238376_at	---	CDNA FLJ30967 fis, clone HEART2000309, weakly similar to PTB-ASSOCIATED SPLICING FACTOR	2.3
243882_at	---	---	2.3
204066_s_at	<i>CENTG2</i>	centaurin, gamma 2	2.3
210140_at	<i>CST7</i>	cystatin F (leukocystatin)	2.3
204794_at	<i>DUSP2</i>	dual specificity phosphatase 2	2.3
228256_s_at	<i>EPB41L4A</i>	erythrocyte membrane protein band 4.1 like 4A	2.3
213619_at	<i>HNRPH1</i>	Heterogeneous nuclear ribonucleoprotein H1 (H)	2.3
239412_at	<i>IRF5</i>	interferon regulatory factor 5	2.3
241824_at	---	Transcribed locus	2.3
226625_at	---	---	2.3
237009_at	---	---	2.3
213317_at	<i>CLIC5</i>	chloride intracellular channel 5	2.3
233261_at	<i>EBF1</i>	Early B-cell factor 1	2.3
212070_at	<i>GPR56</i>	G protein-coupled receptor 56	2.3
213343_s_at	<i>GDPD5</i>	glycerophosphodiester phosphodiesterase domain containing 5	2.3
230930_at	<i>LOC338620</i>	hypothetical protein LOC338620	2.3
226675_s_at	<i>MALAT1</i>	metastasis associated lung adenocarcinoma transcript 1 (non-coding RNA)	2.3
203216_s_at	<i>MYO6</i>	myosin VI	2.3
229151_at	<i>SLC14A1</i>	Solute carrier family 14 (urea transporter), member 1 (Kidd blood group)	2.3
240890_at	---	CDNA clone IMAGE:5216666	2.2
229971_at	<i>GPR114</i>	G protein-coupled receptor 114	2.2
227394_at	<i>NCAM1</i>	neural cell adhesion molecule 1	2.2
227001_at	<i>NPAL2</i>	NIPA-like domain containing 2	2.2
218424_s_at	<i>STEAP3</i>	STEAP family member 3	2.2
226722_at	<i>FAM20C</i>	family with sequence similarity 20, member C	2.2
235735_at	---	Full length insert cDNA clone ZC64D04	2.2
226606_s_at	<i>GTPBP5</i>	GTP binding protein 5 (putative)	2.2

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

50221_at	<i>TFEB</i>	transcription factor EB	2.2
229923_at	<i>ZDHHC21</i>	zinc finger, DHHC-type containing 21	2.2
206978_at	<i>CCR2</i> /// <i>LOC729230</i>	chemokine (C-C motif) receptor 2 /// similar to C-C chemokine receptor type 2 (C-C CKR-2) (CC-CKR-2) (CCR-2) (CCR2) (Monocyte chemoattractant protein 1 receptor) (MCP-1-R) (CD192 antigen)	2.1
219529_at	<i>CLIC3</i>	chloride intracellular channel 3	2.1
203979_at	<i>CYP27A1</i>	cytochrome P450, family 27, subfamily A, polypeptide 1	2.1
228518_at	<i>IGH</i> /// <i>IGHG1</i> /// <i>IGHM</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu	2.1
226003_at	<i>KIF21A</i>	kinesin family member 21A	2.1
226047_at	<i>MRV11</i>	murine retrovirus integration site 1 homolog	2.1
213915_at	<i>NKG7</i>	natural killer cell group 7 sequence	2.1
218559_s_at	<i>MAFB</i>	v-maf musculoaponeurotic fibrosarcoma oncogene homolog B (avian)	2.1
216563_at	<i>ANKRD12</i>	Ankyrin repeat domain 12	2.1
212772_s_at	<i>ABCA2</i>	ATP-binding cassette, sub-family A (ABC1), member 2	2.1
217867_x_at	<i>BACE2</i>	beta-site APP-cleaving enzyme 2	2.1
234151_at	---	CDNA: FLJ20976 fis, clone ADSU01764	2.1
228477_at	<i>FLJ10154</i>	hypothetical protein FLJ10154	2.1
1560520_at	<i>LOC401312</i>	LOC401318	2.1
225897_at	<i>MARCKS</i>	myristoylated alanine-rich protein kinase C substrate	2.1
223750_s_at	<i>TLR10</i>	toll-like receptor 10	2.1
227952_at	<i>ZNF718</i>	Zinc finger protein 718	2.1
229607_at	---	---	2.1
219983_at	<i>HRASLS</i>	HRAS-like suppressor	2.1
1569453_a_at	<i>LOC692247</i>	hypothetical locus LOC692247	2.1
213931_at	<i>ID2</i> /// <i>ID2B</i>	inhibitor of DNA binding 2, dominant negative helix-loop-helix protein /// inhibitor of DNA binding 2B, dominant negative helix-loop-helix protein	2.1
1564435_a_at	<i>KRT72</i>	keratin 72	2.1
220646_s_at	<i>KLRF1</i>	killer cell lectin-like receptor subfamily F, member 1	2.1
229261_at	---	Transcribed locus	2.1
205181_at	<i>ZNF193</i>	zinc finger protein 193	2.1
237841_at	---	CDNA FLJ30669 fis, clone FCBBF1000684	2.0
228904_at	<i>HOXB3</i>	homeobox B3	2.0
208961_s_at	<i>KLF6</i>	Kruppel-like factor 6	2.0
218086_at	<i>NPDC1</i>	neural proliferation, differentiation and control, 1	2.0
205863_at	<i>S100A12</i>	S100 calcium binding protein A12	2.0
202833_s_at	<i>SERPINA1</i>	serpin peptidase inhibitor, clade A (alpha-1 antiproteinase, antitrypsin), member 1	2.0
209723_at	<i>SERPINB9</i>	serpin peptidase inhibitor, clade B (ovalbumin), member 9	2.0
239995_at	---	Transcribed locus, strongly similar to XP_530035.1 hypothetical	2.0

		protein XP_530035 [Pan troglodytes]	
216550_x_at	<i>ANKRD12</i>	ankyrin repeat domain 12	2.0
204446_s_at	<i>ALOX5</i>	arachidonate 5-lipoxygenase	2.0
200702_s_at	<i>DDX24</i>	DEAD (Asp-Glu-Ala-Asp) box polypeptide 24	2.0
240633_at	<i>DOK7</i>	docking protein 7	2.0
37996_s_at	<i>DMPK</i>	dystrophia myotonica-protein kinase	2.0
206115_at	<i>EGR3</i>	early growth response 3	2.0
218397_at	<i>FANCL</i>	Fanconi anemia, complementation group L	2.0
238127_at	<i>FLJ41484</i>	hypothetical LOC650669	2.0
213716_s_at	<i>SECTM1</i>	secreted and transmembrane 1	2.0
232914_s_at	<i>SYTL2</i>	synaptotagmin-like 2	2.0
221866_at	<i>TFEB</i>	transcription factor EB	2.0
204083_s_at	<i>TPM2</i>	tropomyosin 2 (beta)	2.0
215101_s_at	<i>CXCL5</i>	chemokine (C-X-C motif) ligand 5	1.9
239146_at	<i>CLDND1</i>	claudin domain containing 1	1.9
1558346_at	<i>COX17</i>	COX17 cytochrome c oxidase assembly homolog (<i>S. cerevisiae</i>)	1.9
224215_s_at	<i>DLL1</i>	delta-like 1 (<i>Drosophila</i>)	1.9
244292_at	---	Full length insert cDNA clone ZD83B06	1.9
218070_s_at	<i>GMPPA</i>	GDP-mannose pyrophosphorylase A	1.9
1564139_at	<i>LOC144571</i>	hypothetical protein LOC144571	1.9
228325_at	<i>KIAA0146</i>	KIAA0146	1.9
225955_at	<i>LOC653506</i> /// <i>METRNL</i>	meteorin, glial cell differentiation regulator-like /// similar to meteorin, glial cell differentiation regulator-like	1.9
227186_s_at	<i>MRPL41</i>	mitochondrial ribosomal protein L41	1.9
225321_s_at	<i>PILRB</i>	paired immunoglobulin-like type 2 receptor beta	1.9
1553589_a_at	<i>PDZK1IP1</i>	PDZK1 interacting protein 1	1.9
1564310_a_at	<i>PARP15</i>	poly (ADP-ribose) polymerase family, member 15	1.9
226299_at	<i>PKN3</i>	protein kinase N3	1.9
232686_at	<i>SIGLECP3</i>	sialic acid binding Ig-like lectin, pseudogene 3	1.9
239135_at	---	Transcribed locus	1.9
213906_at	<i>MYBL1</i>	v-myb myeloblastosis viral oncogene homolog (avian)-like 1	1.9
244869_at	---	---	1.9
242606_at	---	Breast cancer antigen 32004 mRNA, partial sequence	1.9
209473_at	<i>ENTPDI</i>	ectonucleoside triphosphate diphosphohydrolase 1	1.9
206618_at	<i>IL18R1</i>	interleukin 18 receptor 1	1.9
236704_at	---	MRNA; cDNA DKFZp686N0886 (from clone DKFZp686N0886)	1.9
204838_s_at	<i>MLH3</i>	mutL homolog 3 (<i>E. coli</i>)	1.9
202336_s_at	<i>PAM</i>	peptidylglycine alpha-amidating monooxygenase	1.9
215894_at	<i>PTGDR</i>	prostaglandin D2 receptor (DP)	1.9
209197_at	<i>SYT11</i>	synaptotagmin XI	1.9
229560_at	<i>TLR8</i>	toll-like receptor 8	1.9

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
242457_at	---	Transcribed locus	1.9
241068_at	---	Transcribed locus	1.9
244696_at	---	---	1.9
206255_at	<i>BLK</i>	B lymphoid tyrosine kinase	1.9
229121_at	---	CDNA FLJ44441 fis, clone UTERU2020242	1.9
210069_at	<i>CHKB</i> /// <i>CPT1B</i>	choline kinase beta /// carnitine palmitoyltransferase 1B (muscle)	1.9
210889_s_at	<i>FCGR2B</i> /// <i>FCGR2C</i>	Fc fragment of IgG, low affinity IIb, receptor (CD32) /// Fc fragment of IgG, low affinity IIc, receptor for (CD32)	1.9
205220_at	<i>GPR109B</i>	G protein-coupled receptor 109B	1.9
211275_s_at	<i>GYGI</i>	glycogenin 1	1.9
205488_at	<i>GZMA</i>	granzyme A (granzyme 1, cytotoxic T-lymphocyte-associated serine esterase 3)	1.9
238949_at	<i>LOC401805</i>	hypothetical gene supported by NM_144726	1.9
230245_s_at	<i>LOC283663</i>	hypothetical protein LOC283663	1.9
211996_s_at	<i>DKFZp547E087</i> /// <i>LOC23117</i> /// <i>LOC440345</i> /// <i>LOC440353</i> /// <i>LOC613037</i> /// <i>LOC728888</i>	KIAA0220-like protein /// hypothetical gene LOC283846 /// hypothetical protein LOC440345 /// nuclear pore complex interacting protein pseudogene /// similar to Protein KIAA0220	1.9
1557987_at	<i>LOC641298</i>	PI-3-kinase-related kinase SMG-1 - like locus	1.9
216834_at	<i>RGS1</i>	regulator of G-protein signaling 1	1.9
200986_at	<i>SERPING1</i>	serpin peptidase inhibitor, clade G (C1 inhibitor), member 1, (angioedema, hereditary)	1.9
219386_s_at	<i>SLAMF8</i>	SLAM family member 8	1.9
204275_at	<i>SOLH</i>	small optic lobes homolog (Drosophila)	1.9
220140_s_at	<i>SNX11</i>	sorting nexin 11	1.9
204099_at	<i>SMARCD3</i>	SWI/SNF related, matrix associated, actin dependent regulator of chromatin, subfamily d, member 3	1.9
235274_at	---	Transcribed locus	1.9
211052_s_at	<i>TBCD</i>	tubulin folding cofactor D	1.9
204929_s_at	<i>VAMP5</i>	vesicle-associated membrane protein 5 (myobrevin)	1.9
242429_at	<i>ZNF567</i>	zinc finger protein 567	1.9
205639_at	<i>AOAH</i>	acyloxyacyl hydrolase (neutrophil)	1.8
208710_s_at	<i>AP3D1</i>	adaptor-related protein complex 3, delta 1 subunit	1.8
205839_s_at	<i>BZRAP1</i>	benzodiazapine receptor (peripheral) associated protein 1	1.8
209933_s_at	<i>CD300A</i>	CD300a molecule	1.8
1570166_a_at	---	CDNA clone IMAGE:4719554	1.8
214974_x_at	<i>CXCL5</i>	chemokine (C-X-C motif) ligand 5	1.8
206073_at	<i>COLQ</i>	collagen-like tail subunit (single strand of homotrimer) of asymmetric acetylcholinesterase	1.8
203104_at	<i>CSF1R</i>	colony stimulating factor 1 receptor, formerly McDonough feline sarcoma viral (v-fms) oncogene homolog	1.8

213183_s_at	<i>CDKN1C</i>	Cyclin-dependent kinase inhibitor 1C (p57, Kip2)	1.8
222279_at	<i>RP3-377H14.5</i>	hypothetical protein FLJ35429	1.8
242996_at	<i>MTRF1</i>	mitochondrial translational release factor 1	1.8
225147_at	<i>PSCD3</i>	pleckstrin homology, Sec7 and coiled-coil domains 3	1.8
225688_s_at	<i>PHLDB2</i>	pleckstrin homology-like domain, family B, member 2	1.8
223152_at	<i>PPP1R12C</i>	protein phosphatase 1, regulatory (inhibitor) subunit 12C	1.8
214041_x_at	<i>RPL37A</i>	Ribosomal protein L37a	1.8
1552449_a_at	<i>hCG_1741344</i> <i>/// SCGB1C1</i>	secretoglobin, family 1C, member 1 /// similar to secretoglobin, family 1C, member 1	1.8
227591_at	<i>SH3BP5</i>	SH3-domain binding protein 5 (BTK-associated)	1.8
205861_at	<i>SPIB</i>	Spi-B transcription factor (Spi-1/PU.1 related)	1.8
203887_s_at	<i>THBD</i>	thrombomodulin	1.8
226489_at	<i>TMCC3</i>	transmembrane and coiled-coil domain family 3	1.8
227393_at	<i>TMEM16J</i>	transmembrane protein 16J	1.8
218145_at	<i>TRIB3</i>	tribbles homolog 3 (Drosophila)	1.8
33132_at	<i>CPSF1</i>	cleavage and polyadenylation specific factor 1, 160kDa	1.8
228333_at	---	Full length insert cDNA clone YT94E02	1.8
210606_x_at	<i>KLRD1</i>	killer cell lectin-like receptor subfamily D, member 1	1.8
228592_at	<i>MS4A1</i>	membrane-spanning 4-domains, subfamily A, member 1	1.8
228658_at	<i>MIAT</i>	myocardial infarction associated transcript (non-protein coding)	1.8
39650_s_at	<i>PCNXL2</i>	pecanex-like 2 (Drosophila)	1.8
212187_x_at	<i>PTGDS</i>	prostaglandin D2 synthase 21kDa (brain)	1.8
240413_at	<i>PYHIN1</i>	pyrin and HIN domain family, member 1	1.8
232231_at	<i>RUNX2</i>	runt-related transcription factor 2	1.8
207351_s_at	<i>SH2D2A</i>	SH2 domain protein 2A	1.8
209679_s_at	<i>LOC57228</i>	small trans-membrane and glycosylated protein	1.8
221769_at	<i>SPSB3</i>	splA/ryanodine receptor domain and SOCS box containing 3	1.8
205315_s_at	<i>SNTB2</i>	syntrophin, beta 2 (dystrophin-associated protein A1, 59kDa, basic component 2)	1.8
212956_at	<i>TBC1D9</i>	TBC1 domain family, member 9 (with GRAM domain)	1.8
243049_at	---	---	1.8
206170_at	<i>ADRB2</i>	adrenergic, beta-2-, receptor, surface	1.8
227113_at	<i>ADHFE1</i>	alcohol dehydrogenase, iron containing, 1	1.8
220416_at	<i>ATP8B4</i>	ATPase, Class I, type 8B, member 4	1.8
213578_at	<i>BMPRIA</i>	bone morphogenetic protein receptor, type IA	1.8
203973_s_at	<i>CEBPD</i>	CCAAT/enhancer binding protein (C/EBP), delta	1.8
202910_s_at	<i>CD97</i>	CD97 molecule	1.8
218148_at	<i>CENPT</i>	centromere protein T	1.8
218938_at	<i>FBXL15</i>	F-box and leucine-rich repeat protein 15	1.8
223836_at	<i>FGFBP2</i>	fibroblast growth factor binding protein 2	1.8
226171_at	<i>FLJ20209</i>	hypothetical protein FLJ20209	1.8

228283_at	<i>MGC61571</i>	hypothetical protein MGC61571	1.8
206478_at	<i>KIAA0125</i>	KIAA0125	1.8
229473_at	<i>MAMDC4</i>	MAM domain containing 4	1.8
201058_s_at	<i>MYL9</i>	myosin, light chain 9, regulatory	1.8
213116_at	<i>NEK3</i>	NIMA (never in mitosis gene a)-related kinase 3	1.8
203845_at	<i>PCAF</i>	p300/CBP-associated factor	1.8
210845_s_at	<i>PLAUR</i>	plasminogen activator, urokinase receptor	1.8
212823_s_at	<i>PLEKHG3</i>	pleckstrin homology domain containing, family G (with RhoGef domain) member 3	1.8
210448_s_at	<i>P2RX5</i>	purinergic receptor P2X, ligand-gated ion channel, 5	1.8
225738_at	<i>RAPGEF1</i>	Rap guanine nucleotide exchange factor (GEF) 1	1.8
236717_at	<i>LOC165186</i>	similar to RIKEN cDNA 4632412N22 gene	1.8
210613_s_at	<i>SYNGR1</i>	synaptogyrin 1	1.8
220384_at	<i>TXNDC3</i>	thioredoxin domain containing 3 (spermatozoa)	1.8
229202_at	---	Transcribed locus	1.8
226700_at	<i>U2AF1L4</i>	U2 small nuclear RNA auxiliary factor 1-like 4	1.8
236190_at	<i>XYLT1</i>	Xylosyltransferase I	1.8
205180_s_at	<i>ADAM8</i>	ADAM metallopeptidase domain 8	1.7
228032_s_at	---	CDNA FLJ36663 fis, clone UTERU2002826	1.7
227152_at	<i>C12orf35</i>	chromosome 12 open reading frame 35	1.7
204494_s_at	<i>C15orf39</i>	chromosome 15 open reading frame 39	1.7
228891_at	<i>C9orf164</i>	chromosome 9 open reading frame 164	1.7
1558185_at	<i>CLLU1</i>	chronic lymphocytic leukemia up-regulated 1	1.7
208094_s_at	<i>CCDC130</i>	coiled-coil domain containing 130	1.7
205081_at	<i>CRIP1</i>	cysteine-rich protein 1 (intestinal)	1.7
1555486_a_at	<i>FLJ14213</i>	hypothetical protein FLJ14213	1.7
212657_s_at	<i>IL1RN</i>	interleukin 1 receptor antagonist	1.7
240613_at	<i>JAK1</i>	Janus kinase 1 (a protein tyrosine kinase)	1.7
206584_at	<i>LY96</i>	lymphocyte antigen 96	1.7
227999_at	<i>PWWP2</i>	PWWP domain containing 2	1.7
224710_at	<i>RAB34</i>	RAB34, member RAS oncogene family	1.7
228030_at	<i>RBM6</i>	RNA binding motif protein 6	1.7
218638_s_at	<i>SPON2</i>	spondin 2, extracellular matrix protein	1.7
209403_at	<i>LOC653380</i> /// <i>LOC653498</i> /// <i>LOC727735</i> /// <i>LOC729837</i> /// <i>LOC729873</i> /// <i>LOC729877</i> /// <i>TBC1D3</i> /// <i>TBC1D3C</i>	TBC1 domain family, member 3 /// TBC1 domain family, member 3C /// similar to USP6 N-terminal like /// similar to TBC1 domain family member 3 (Rab GTPase-activating protein PRC17) (Prostate cancer gene 17 protein) (TRE17 alpha protein) /// similar to TBC1 domain family, member 3	1.7
239988_at	---	Transcribed locus	1.7
244695_at	---	Transcribed locus	1.7

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
225605_at	<i>TP53I13</i>	tumor protein p53 inducible protein 13	1.7
213527_s_at	<i>ZNF688</i>	zinc finger protein 688	1.7
226344_at	<i>ZMAT1</i>	zinc finger, matrin type 1	1.7
201850_at	<i>CAPG</i>	capping protein (actin filament), gelsolin-like	1.7
202450_s_at	<i>CTSK</i>	cathepsin K	1.7
201743_at	<i>CD14</i>	CD14 molecule	1.7
213398_s_at	<i>C14orf124</i>	chromosome 14 open reading frame 124	1.7
227668_at	<i>C17orf56</i>	chromosome 17 open reading frame 56	1.7
218565_at	<i>C9orf114</i>	chromosome 9 open reading frame 114	1.7
226713_at	<i>CCDC50</i>	coiled-coil domain containing 50	1.7
203409_at	<i>DDB2</i>	damage-specific DNA binding protein 2, 48kDa	1.7
218552_at	<i>ECHDC2</i>	enoyl Coenzyme A hydratase domain containing 2	1.7
212087_s_at	<i>ERAL1</i>	Era G-protein-like 1 (E. coli)	1.7
231647_s_at	<i>FCRL5</i>	Fc receptor-like 5	1.7
226184_at	<i>FMNL2</i>	formin-like 2	1.7
1565567_at	---	Full length insert cDNA YN68A11	1.7
1556839_s_at	<i>SPTBN5</i>	Homo sapiens, clone IMAGE:4704591 /// Spectrin, beta, non-erythrocytic 5	1.7
203927_at	<i>NFKBIE</i>	nuclear factor of kappa light polypeptide gene enhancer in B-cells inhibitor, epsilon	1.7
203574_at	<i>NFIL3</i>	nuclear factor, interleukin 3 regulated	1.7
223464_at	<i>OSBPL5</i>	oxysterol binding protein-like 5	1.7
219095_at	<i>PLA2G4B</i>	phospholipase A2, group IVB (cytosolic)	1.7
203317_at	<i>PSD4</i>	pleckstrin and Sec7 domain containing 4	1.7
225739_at	<i>RAB11FIP4</i>	RAB11 family interacting protein 4 (class II)	1.7
202358_s_at	<i>SNX19</i>	sorting nexin 19	1.7
201072_s_at	<i>SMARCC1</i>	SWI/SNF related, matrix associated, actin dependent regulator of chromatin, subfamily c, member 1	1.7
220684_at	<i>TBX21</i>	T-box 21	1.7
236833_at	<i>TTC16</i>	tetratricopeptide repeat domain 16	1.7
242801_at	---	Transcribed locus	1.7
239489_at	---	Transcribed locus	1.7
1555888_at	<i>UBR5</i>	Ubiquitin protein ligase E3 component n-recogin 5	1.7
202780_at	<i>OXCT1</i>	3-oxoacid CoA transferase 1	-1.7
223298_s_at	<i>NT5C3</i>	5'-nucleotidase, cytosolic III	-1.7
202268_s_at	<i>APPBP1</i>	amyloid beta precursor protein binding protein 1	-1.7
232395_x_at	<i>AGBL3</i>	ATP/GTP binding protein-like 3	-1.7
217610_at	---	CDNA FLJ38765 fis, clone KIDNE2014489	-1.7
205544_s_at	<i>CR2</i>	complement component (3d/Epstein Barr virus) receptor 2	-1.7
228828_at	---	Homo sapiens, clone IMAGE:5215917, mRNA	-1.7
215121_x_at	<i>IGL /// IGLV2-14 /// IGLV3-25</i>	immunoglobulin lambda locus /// immunoglobulin lambda variable 4-3 /// immunoglobulin lambda variable 3-25 /// immunoglobulin	-1.7

	/// <i>IGLV4-3</i>	lambda variable 2-14	
212096_s_at	<i>MTUS1</i>	mitochondrial tumor suppressor 1	-1.7
225944_at	<i>NLN</i>	neurolysin (metallopeptidase M3 family)	-1.7
202165_at	<i>PPP1R2</i>	protein phosphatase 1, regulatory (inhibitor) subunit 2	-1.7
200730_s_at	<i>PTP4A1</i>	protein tyrosine phosphatase type IVA, member 1	-1.7
200848_at	<i>AHCYL1</i>	S-adenosylhomocysteine hydrolase-like 1	-1.7
201020_at	<i>YWHAH</i>	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, eta polypeptide	-1.7
212008_at	<i>UBXD2</i>	UBX domain containing 2	-1.7
222430_s_at	<i>YTHDF2</i>	YTH domain family, member 2	-1.7
1557953_at	<i>ZKSCAN1</i>	zinc finger with KRAB and SCAN domains 1	-1.7
201629_s_at	<i>ACPI</i>	acid phosphatase 1, soluble	-1.7
205512_s_at	<i>AIFM1</i>	apoptosis-inducing factor, mitochondrion-associated, 1	-1.7
238587_at	<i>STS-1</i>	Cbl-interacting protein Sts-1	-1.7
235117_at	<i>CHAC2</i>	ChaC, cation transport regulator homolog 2 (E. coli)	-1.7
212515_s_at	<i>DDX3X</i>	DEAD (Asp-Glu-Ala-Asp) box polypeptide 3, X-linked	-1.7
200043_at	<i>ERH</i>	enhancer of rudimentary homolog (Drosophila)	-1.7
226715_at	<i>FOXK1</i>	forkhead box K1	-1.7
1561155_at	---	Full length insert cDNA clone ZD60E09	-1.7
217655_at	<i>FXDY5</i>	FXDY domain containing ion transport regulator 5	-1.7
201626_at	<i>INSIG1</i>	insulin induced gene 1	-1.7
225613_at	<i>MAST4</i>	microtubule associated serine/threonine kinase family member 4	-1.7
208698_s_at	<i>NONO</i>	non-POU domain containing, octamer-binding	-1.7
213652_at	<i>PCSK5</i>	proprotein convertase subtilisin/kexin type 5	-1.7
210371_s_at	<i>RBBP4</i>	retinoblastoma binding protein 4	-1.7
225166_at	<i>ARHGAP18</i>	Rho GTPase activating protein 18	-1.7
230407_at	---	Transcribed locus	-1.7
224593_at	<i>ZNF664</i>	zinc finger protein 664	-1.7
39248_at	<i>AQP3</i>	aquaporin 3 (Gill blood group)	-1.8
223641_at	---	CDNA FLJ36838 fis, clone ASTRO2011426	-1.8
225638_at	<i>C1orf31</i>	chromosome 1 open reading frame 31	-1.8
221823_at	<i>C5orf30</i>	chromosome 5 open reading frame 30	-1.8
230426_at	<i>DLD</i>	dihydrolipoamide dehydrogenase	-1.8
227847_at	<i>EPM2AIP1</i>	EPM2A (laforin) interacting protein 1	-1.8
226432_at	<i>ETNK1</i>	ethanolamine kinase 1	-1.8
226159_at	<i>LOC285636</i>	hypothetical protein LOC285636	-1.8
218733_at	<i>MSL2L1</i>	male-specific lethal 2-like 1 (Drosophila)	-1.8
235067_at	<i>MKLNI</i>	muskelin 1, intracellular mediator containing kelch motifs	-1.8
227639_at	<i>PIGK</i>	phosphatidylinositol glycan anchor biosynthesis, class K	-1.8
1555247_a_at	<i>RAPGEF6</i>	Rap guanine nucleotide exchange factor (GEF) 6	-1.8
201894_s_at	<i>SSRI</i>	signal sequence receptor, alpha (translocon-associated protein)	-1.8

		alpha)	
1560680_at	<i>LOC732014</i>	Similar to Heterogeneous nuclear ribonucleoprotein A1 (Helix-destabilizing protein) (Single-strand RNA-binding protein) (hnRNP core protein A1) (HDP)	-1.8
215424_s_at	<i>SNW1</i>	SNW domain containing 1	-1.8
207983_s_at	<i>STAG2</i>	stromal antigen 2	-1.8
201663_s_at	<i>SMC4</i>	structural maintenance of chromosomes 4	-1.8
222315_at	---	Transcribed locus	-1.8
238468_at	<i>TNRC6B</i>	trinucleotide repeat containing 6B	-1.8
222420_s_at	<i>UBE2H</i>	ubiquitin-conjugating enzyme E2H (UBC8 homolog, yeast)	-1.8
201096_s_at	<i>ARF4</i>	ADP-ribosylation factor 4	-1.8
207206_s_at	<i>ALOX12</i>	arachidonate 12-lipoxygenase	-1.8
209811_at	<i>CASP2</i>	caspase 2, apoptosis-related cysteine peptidase (neural precursor cell expressed, developmentally down-regulated 2)	-1.8
207828_s_at	<i>CENPF</i>	centromere protein F, 350/400ka (mitosin)	-1.8
201059_at	<i>CTTN</i>	cortactin	-1.8
209839_at	<i>DNM3</i>	dynamamin 3	-1.8
233898_s_at	<i>FGFR1OP2</i>	FGFR1 oncogene partner 2	-1.8
222830_at	<i>GRHL1</i>	grainyhead-like 1 (Drosophila)	-1.8
204686_at	<i>IRS1</i>	insulin receptor substrate 1	-1.8
231798_at	<i>NOG</i>	Noggin	-1.8
218039_at	<i>NUSAP1</i>	nucleolar and spindle associated protein 1	-1.8
204019_s_at	<i>SH3YL1</i>	SH3 domain containing, Ysc84-like 1 (<i>S. cerevisiae</i>)	-1.8
201417_at	<i>SOX4</i>	SRY (sex determining region Y)-box 4	-1.8
213109_at	<i>TNIK</i>	TRAF2 and NCK interacting kinase	-1.8
237753_at	---	Transcribed locus	-1.8
212330_at	<i>TFDP1</i>	transcription factor Dp-1	-1.8
223106_at	<i>TMEM14C</i>	transmembrane protein 14C	-1.8
200638_s_at	<i>LOC650083</i> /// <i>YWHAZ</i>	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, zeta polypeptide /// similar to tyrosine 3/tryptophan 5 - monooxygenase activation protein, zeta polypeptide	-1.8
200965_s_at	<i>ABLIM1</i>	actin binding LIM protein 1	-1.8
222740_at	<i>ATAD2</i>	ATPase family, AAA domain containing 2	-1.8
222696_at	<i>AXIN2</i>	axin 2 (conductin, axil)	-1.8
227227_at	---	CDNA FLJ32605 fis, clone STOMA1000175	-1.8
238010_at	<i>C1orf174</i>	chromosome 1 open reading frame 174	-1.8
203482_at	<i>C10orf6</i>	chromosome 10 open reading frame 6	-1.8
208077_at	<i>C9orf38</i>	chromosome 9 open reading frame 38	-1.8
1557238_s_at	---	EST clone 111681 mariner transposon Hsmar1 sequence	-1.8
210387_at	<i>HIST1H2BG</i>	histone cluster 1, H2bg	-1.8
228160_at	<i>LOC400642</i>	hypothetical gene supported by BC041875; BX648984	-1.8
217148_x_at	<i>IGLV2-14</i>	immunoglobulin lambda variable 2-14	-1.8

203820_s_at	<i>IGF2BP3</i>	insulin-like growth factor 2 mRNA binding protein 3	-1.8
221503_s_at	<i>KPNA3</i>	karyopherin alpha 3 (importin alpha 4)	-1.8
202729_s_at	<i>LTBP1</i>	latent transforming growth factor beta binding protein 1	-1.8
1564776_at	<i>LENG10</i>	leukocyte receptor cluster (LRC) member 10	-1.8
218558_s_at	<i>MRPL39</i>	mitochondrial ribosomal protein L39	-1.8
223244_s_at	<i>NDUFA12</i>	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 12	-1.8
221757_at	<i>PIK3IP1</i>	phosphoinositide-3-kinase interacting protein 1	-1.8
210759_s_at	<i>PSMA1</i>	proteasome (prosome, macropain) subunit, alpha type, 1	-1.8
218607_s_at	<i>SDAD1</i>	SDA1 domain containing 1	-1.8
239411_at	---	Transcribed locus	-1.8
236220_at	---	Transcribed locus	-1.8
232991_at	<i>ARL17</i>	ADP-ribosylation factor-like 17	-1.9
209846_s_at	<i>BTN3A2</i>	butyrophilin, subfamily 3, member A2	-1.9
203357_s_at	<i>CAPN7</i>	calpain 7	-1.9
242974_at	<i>CD47</i>	CD47 molecule	-1.9
1566142_at	---	CDNA FLJ37949 fis, clone CTONG2009156	-1.9
242568_s_at	---	CDNA FLJ38922 fis, clone NT2NE2011691	-1.9
226456_at	<i>C16orf75</i>	chromosome 16 open reading frame 75	-1.9
232879_at	<i>CRTC3</i>	CREB regulated transcription coactivator 3	-1.9
1556821_x_at	<i>DLEU2</i>	deleted in lymphocytic leukemia, 2	-1.9
1552621_at	<i>POLR2J2</i> /// <i>POLR2J3</i>	DNA directed RNA polymerase II polypeptide J-related /// RPB11b2 protein	-1.9
222859_s_at	<i>DAPPI</i>	dual adaptor of phosphotyrosine and 3-phosphoinositides	-1.9
1562831_a_at	<i>LOC283089</i>	hypothetical protein LOC283089	-1.9
236887_at	<i>KIN</i>	KIN, antigenic determinant of recA protein homolog (mouse)	-1.9
231975_s_at	<i>MIER3</i>	mesoderm induction early response 1, family member 3	-1.9
224823_at	<i>MYLK</i>	myosin, light chain kinase	-1.9
236930_at	<i>NUMB</i>	Numb homolog (Drosophila)	-1.9
206099_at	<i>PRKCH</i>	protein kinase C, eta	-1.9
236223_s_at	<i>RIT1</i>	Ras-like without CAAX 1	-1.9
201011_at	<i>RPNI</i>	ribophorin I	-1.9
201239_s_at	<i>LOC653566</i> /// <i>SPCS2</i>	signal peptidase complex subunit 2 homolog (S. cerevisiae) /// signal peptidase complex subunit 2 homolog pseudogene	-1.9
222727_s_at	<i>SLC24A6</i>	solute carrier family 24 (sodium/potassium/calcium exchanger), member 6	-1.9
204467_s_at	<i>SNCA</i>	synuclein, alpha (non A4 component of amyloid precursor)	-1.9
209754_s_at	<i>TMPO</i>	thymopoietin	-1.9
210379_s_at	<i>TLK1</i>	tousled-like kinase 1	-1.9
242693_at	---	Transcribed locus	-1.9
231784_s_at	<i>WDSOF1</i>	WD repeats and SOF1 domain containing	-1.9
217378_x_at	---	---	-1.9

202760_s_at	<i>AKAP2</i> /// <i>PALM2-AKAP2</i>	A kinase (PRKA) anchor protein 2 /// PALM2-AKAP2 protein	-1.9
213060_s_at	<i>CHI3L2</i>	chitinase 3-like 2	-1.9
220477_s_at	<i>C20orf30</i>	chromosome 20 open reading frame 30	-1.9
219179_at	<i>DACT1</i>	dapper, antagonist of beta-catenin, homolog 1 (<i>Xenopus laevis</i>)	-1.9
215659_at	<i>GSDML</i>	Gasdermin-like	-1.9
223777_at	<i>MGC13005</i>	hypothetical protein MGC13005	-1.9
219657_s_at	<i>KLF3</i>	Kruppel-like factor 3 (basic)	-1.9
222510_s_at	<i>MKRN2</i>	makorin, ring finger protein, 2	-1.9
223311_s_at	<i>MTA3</i>	metastasis associated 1 family, member 3	-1.9
222826_at	<i>PLDN</i>	pallidin homolog (mouse)	-1.9
200968_s_at	<i>PPIB</i>	peptidylprolyl isomerase B (cyclophilin B)	-1.9
1556009_at	<i>PEX13</i>	peroxisome biogenesis factor 13	-1.9
1564494_s_at	<i>P4HB</i>	procollagen-proline, 2-oxoglutarate 4-dioxygenase (proline 4-hydroxylase), beta polypeptide	-1.9
219493_at	<i>SHCBP1</i>	SHC SH2-domain binding protein 1	-1.9
216850_at	<i>SNRPN</i>	small nuclear ribonucleoprotein polypeptide N	-1.9
208761_s_at	<i>SUMO1</i>	SMT3 suppressor of mif two 3 homolog 1 (<i>S. cerevisiae</i>)	-1.9
204361_s_at	<i>SKAP2</i>	src kinase associated phosphoprotein 2	-1.9
217979_at	<i>TSPAN13</i>	Tetraspanin 13	-1.9
240302_at	---	Transcribed locus	-1.9
239016_at	---	Transcribed locus	-1.9
242617_at	<i>TMED8</i>	Transmembrane emp24 protein transport domain containing 8	-1.9
221205_at	---	---	-1.9
1556007_s_at	<i>CSNK1A1</i>	Casein kinase 1, alpha 1	-1.9
238653_at	---	CDNA FLJ43454 fis, clone OCBBF2034906	-1.9
214769_at	<i>CLCN4</i>	chloride channel 4	-1.9
211792_s_at	<i>CDKN2C</i>	cyclin-dependent kinase inhibitor 2C (p18, inhibits CDK4)	-1.9
232599_at	<i>EXOC6</i>	exocyst complex component 6	-1.9
1559964_at	<i>FLJ38717</i>	FLJ38717 protein	-1.9
239432_at	<i>FLJ31306</i>	hypothetical protein FLJ31306	-1.9
211641_x_at	---	Isolate Middle91 immunoglobulin heavy chain variable region (IGVH)	-1.9
1559600_at	---	MRNA; cDNA DKFZp547G2314 (from clone DKFZp547G2314)	-1.9
222582_at	<i>PRKAG2</i>	protein kinase, AMP-activated, gamma 2 non-catalytic subunit	-1.9
204427_s_at	<i>TMED2</i>	transmembrane emp24 domain trafficking protein 2	-1.9
210935_s_at	<i>WDR1</i>	WD repeat domain 1	-1.9
233239_at	---	CDNA: FLJ21229 fis, clone COL00740	-2.0
207389_at	<i>GPIBA</i>	glycoprotein Ib (platelet), alpha polypeptide	-2.0
228568_at	<i>Gcom1</i>	GRINL1A combined protein	-2.0
210948_s_at	<i>LEF1</i>	lymphoid enhancer-binding factor 1	-2.0

200644_at	<i>MARCKSL1</i>	MARCKS-like 1	-2.0
227722_at	<i>RPS23</i>	ribosomal protein S23	-2.0
205857_at	<i>SLC18A2</i>	solute carrier family 18 (vesicular monoamine), member 2	-2.0
202223_at	<i>STT3A</i>	STT3, subunit of the oligosaccharyltransferase complex, homolog A (<i>S. cerevisiae</i>)	-2.0
237544_at	---	Transcribed locus	-2.0
243514_at	---	Transcribed locus	-2.0
222777_s_at	<i>WHSC1</i>	Wolf-Hirschhorn syndrome candidate 1	-2.0
228974_at	---	CDNA FLJ42233 fis, clone THYMU3000420	-2.0
220710_at	<i>C15orf28</i>	chromosome 15 open reading frame 28	-2.0
233995_at	---	Clone HQ0663 PRO0663	-2.0
219301_s_at	<i>CNTNAP2</i>	contactin associated protein-like 2	-2.0
202221_s_at	<i>EP300</i>	E1A binding protein p300	-2.0
81737_at	---	Homo sapiens, clone IMAGE:4271781	-2.0
204108_at	<i>NFYA</i>	nuclear transcription factor Y, alpha	-2.0
218340_s_at	<i>UBE1L2</i>	ubiquitin-activating enzyme E1-like 2	-2.0
1555194_at	---	CDNA clone MGC:21733 IMAGE:4517792	-2.0
207431_s_at	<i>DEGS1</i>	degenerative spermatocyte homolog 1, lipid desaturase (<i>Drosophila</i>)	-2.0
226811_at	<i>FAM46C</i>	family with sequence similarity 46, member C	-2.0
244741_s_at	<i>MGC9913</i>	Hypothetical protein MGC9913	-2.0
216576_x_at	<i>NTN2L</i>	Netrin 2-like (chicken)	-2.0
208097_s_at	<i>TXNDC1</i>	thioredoxin domain containing 1	-2.0
226107_at	---	CDNA FLJ13495 fis, clone PLACE1004425 /// Full-length cDNA clone CS0DI084YF13 of Placenta Cot 25-normalized of Homo sapiens (human)	-2.1
225644_at	<i>CCDC117</i>	coiled-coil domain containing 117	-2.1
1552410_at	<i>CLEC4F</i>	C-type lectin domain family 4, member F	-2.1
211798_x_at	<i>IGLJ3</i>	immunoglobulin lambda joining 3	-2.1
210878_s_at	<i>JMJD1B</i>	jumonji domain containing 1B	-2.1
225567_at	---	MRNA; cDNA DKFZp762E1314 (from clone DKFZp762E1314)	-2.1
226864_at	<i>PKIA</i>	Protein kinase (cAMP-dependent, catalytic) inhibitor alpha	-2.1
227396_at	<i>PTPRJ</i>	protein tyrosine phosphatase, receptor type, J	-2.1
241299_at	---	Transcribed locus	-2.1
205506_at	<i>VILI</i>	villin 1	-2.1
215252_at	---	CDNA: FLJ21350 fis, clone COL02751	-2.1
222848_at	<i>CENPK</i>	centromere protein K	-2.1
229070_at	<i>C6orf105</i>	chromosome 6 open reading frame 105	-2.1
212730_at	<i>DMN</i>	desmuslin	-2.1
1559205_s_at	---	Homo sapiens, clone IMAGE:5745627, mRNA	-2.1
211548_s_at	<i>HPGD</i>	hydroxyprostaglandin dehydrogenase 15-(NAD)	-2.1
228316_at	<i>FLJ31438</i>	hypothetical protein FLJ31438	-2.1

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

217157_x_at	---	Immunoglobulin kappa chain, V-region (SPK.3)	-2.1
1553873_at	<i>KLHL34</i>	kelch-like 34 (Drosophila)	-2.1
231866_at	<i>LNPEP</i>	leucyl/cystinyl aminopeptidase	-2.1
211048_s_at	<i>PDIA4</i>	protein disulfide isomerase family A, member 4	-2.1
227692_at	<i>GNAI1</i>	guanine nucleotide binding protein (G protein), alpha inhibiting activity polypeptide 1	-2.1
233303_at	---	Homo sapiens, clone IMAGE:4295366, mRNA	-2.1
234764_x_at	<i>IGL@ /// IGLV1-44</i>	Immunoglobulin lambda variable 1-44 /// Immunoglobulin anti-HBsAg lambda light chain (LM25) /// Immunoglobulin lambda locus	-2.1
225997_at	<i>MOBK1A</i>	MOB1, Mps One Binder kinase activator-like 1A (yeast)	-2.1
218951_s_at	<i>PLCXD1</i>	phosphatidylinositol-specific phospholipase C, X domain containing 1	-2.1
213194_at	<i>ROBO1</i>	roundabout, axon guidance receptor, homolog 1 (Drosophila)	-2.1
203852_s_at	<i>SMN1 /// SMN2</i>	survival of motor neuron 1, telomeric /// survival of motor neuron 2, centromeric	-2.1
206283_s_at	<i>TAL1</i>	T-cell acute lymphocytic leukemia 1	-2.1
223324_s_at	<i>TRPM7</i>	transient receptor potential cation channel, subfamily M, member 7	-2.1
219503_s_at	<i>TMEM40</i>	transmembrane protein 40	-2.1
208324_at	---	---	-2.2
209789_at	<i>CORO2B</i>	coronin, actin binding protein, 2B	-2.2
1558220_at	<i>MUC20</i>	Mucin 20, cell surface associated	-2.2
204545_at	<i>PEX6</i>	peroxisomal biogenesis factor 6	-2.2
202731_at	<i>PDCD4</i>	programmed cell death 4 (neoplastic transformation inhibitor)	-2.2
1559025_at	39334	septin 9	-2.2
202558_s_at	<i>STCH</i>	stress 70 protein chaperone, microsome-associated, 60kDa	-2.2
219449_s_at	<i>TMEM70</i>	transmembrane protein 70	-2.2
1570135_at	<i>ZNF230</i>	zinc finger protein 230	-2.2
209869_at	<i>ADRA2A</i>	adrenergic, alpha-2A-, receptor	-2.2
220521_s_at	<i>ATG16L1</i>	ATG16 autophagy related 16-like 1 (S. cerevisiae)	-2.2
216166_at	---	CDNA: FLJ21256 fis, clone COL01402	-2.2
224829_at	<i>CPEB4</i>	cytoplasmic polyadenylation element binding protein 4	-2.2
1552622_s_at	<i>LOC441259 /// LOC730323 /// POLR2J2 /// POLR2J3</i>	DNA directed RNA polymerase II polypeptide J-related /// PMS2 postmeiotic segregation increased 2 (S. cerevisiae)-like /// RPB11b2 protein /// similar to postmeiotic segregation increased 2-like 2	-2.2
1558922_at	---	Full length insert cDNA clone YF43G08	-2.2
216765_at	<i>MAP2K5</i>	Mitogen-activated protein kinase kinase 5	-2.2
210543_s_at	<i>PRKDC</i>	protein kinase, DNA-activated, catalytic polypeptide	-2.2
223989_s_at	<i>REXO2</i>	REX2, RNA exonuclease 2 homolog (S. cerevisiae)	-2.2
1558117_s_at	<i>USP31</i>	ubiquitin specific peptidase 31	-2.2
214895_s_at	<i>ADAM10</i>	ADAM metallopeptidase domain 10	-2.3
212097_at	<i>CAVI</i>	caveolin 1, caveolae protein, 22kDa	-2.3

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

214768_x_at	<i>IGKC</i>	Immunoglobulin kappa constant	-2.3
230793_at	<i>LRRRC16</i>	leucine rich repeat containing 16	-2.3
203470_s_at	<i>PLEK</i>	pleckstrin	-2.3
238774_at	---	---	-2.3
1569312_at	---	CDNA clone IMAGE:4067166	-2.3
1562836_at	---	CDNA FLJ11653 fis, clone HEMBA1004538	-2.3
232967_at	<i>XPA</i>	Xeroderma pigmentosum, complementation group A	-2.3
1569408_at	<i>EIF2C4</i>	Eukaryotic translation initiation factor 2C, 4	-2.4
226558_at	<i>LOC653071</i>	similar to CG32820-PA, isoform A	-2.4
228737_at	<i>TOX2</i>	TOX high mobility group box family member 2	-2.4
214710_s_at	<i>CCNB1</i>	cyclin B1	-2.4
1562927_at	---	Homo sapiens, clone IMAGE:5215971, mRNA	-2.4
208927_at	<i>SPOP</i>	speckle-type POZ protein	-2.4
227974_at	---	Transcribed locus	-2.4
218542_at	<i>CEP55</i>	centrosomal protein 55kDa	-2.5
203560_at	<i>GGH</i>	gamma-glutamyl hydrolase (conjugase, foylpolypolyglutamyl hydrolase)	-2.5
211645_x_at	---	Immunoglobulin kappa light chain (IGKV) mRNA variable region, joining region, and constant region	-2.5
235965_at	---	MRNA; cDNA DKFZp779C0742 (from clone DKFZp779C0742)	-2.5
200962_at	<i>RPL31</i>	ribosomal protein L31	-2.5
207426_s_at	<i>TNFSF4</i>	tumor necrosis factor (ligand) superfamily, member 4 (tax-transcriptionally activated glycoprotein 1, 34kDa)	-2.5
219003_s_at	<i>MANEA</i>	mannosidase, endo-alpha	-2.5
223062_s_at	<i>PSAT1</i>	phosphoserine aminotransferase 1	-2.5
222385_x_at	<i>SEC61A1</i>	Sec61 alpha 1 subunit (<i>S. cerevisiae</i>)	-2.5
238407_at	---	Transcribed locus	-2.5
230493_at	<i>TMEM46</i>	transmembrane protein 46	-2.5
1556592_at	---	CDNA FLJ40061 fis, clone TESOP2000083	-2.6
235126_at	<i>LQK1</i>	LQK1 hypothetical protein short isoform	-2.6
211352_s_at	<i>NCOA3</i>	nuclear receptor coactivator 3	-2.6
227703_s_at	<i>SYTL4</i>	synaptotagmin-like 4 (granuphilin-a)	-2.6
207165_at	<i>HMMR</i>	hyaluronan-mediated motility receptor (RHAMM)	-2.6
209218_at	<i>SQLE</i>	squalene epoxidase	-2.6
1557252_at	---	CDNA FLJ36213 fis, clone THYMU2000671	-2.7
203755_at	<i>BUB1B</i>	BUB1 budding uninhibited by benzimidazoles 1 homolog beta (yeast)	-2.7
217281_x_at	<i>IL8</i>	Interleukin 8	-2.7
202589_at	<i>TYMS</i>	thymidylate synthetase	-2.7
203213_at	<i>CDC2</i>	cell division cycle 2, G1 to S and G2 to M	-2.8
215356_at	<i>ECAT8</i>	ES cell associated transcript 8	-2.8
210012_s_at	<i>EWSR1</i>	Ewing sarcoma breakpoint region 1	-2.8

216557_x_at	<i>IGHA1</i> /// <i>IGHG1</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-2.8
243495_s_at	---	MRNA; cDNA DKFZp686E18224 (from clone DKFZp686E18224)	-2.8
235412_at	<i>ARHGEF7</i>	Rho guanine nucleotide exchange factor (GEF) 7	-2.8
205692_s_at	<i>CD38</i>	CD38 molecule	-2.9
233416_at	---	CDNA FLJ30762 fis, clone FEBRA2000575	-2.9
215177_s_at	<i>ITGA6</i>	integrin, alpha 6	-2.9
232952_at	---	CDNA FLJ11942 fis, clone HEMBB1000652	-2.9
244631_at	<i>LOC389834</i> /// <i>LOC642398</i> /// <i>LOC727834</i>	hypothetical gene supported by AK123403 /// hypothetical LOC642398 /// hypothetical protein LOC727834	-2.9
225655_at	<i>UHRF1</i>	ubiquitin-like, containing PHD and RING finger domains, 1	-2.9
217022_s_at	<i>IGHA1</i> /// <i>IGHA2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant alpha 2 (A2m marker)	-3.0
236641_at	<i>KIF14</i>	kinesin family member 14	-3.0
231925_at	---	CDNA: FLJ23006 fis, clone LNG00414	-3.0
1556054_at	---	Full length insert cDNA clone ZD45C02	-3.0
201747_s_at	<i>SAFB</i>	scaffold attachment factor B	-3.0
215262_at	---	Clone 24629 mRNA sequence	-3.1
216510_x_at	<i>EXOC7</i> /// <i>IGHA1</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>ZCWPW2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-3.1
211430_s_at	<i>IGH</i> /// <i>IGHG1</i> /// /// <i>IGHG2</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-3.1
221491_x_at	<i>hCG_1998957</i> /// /// <i>HLA-DRB1</i> /// /// <i>HLA-DRB3</i> /// /// <i>HLA-DRB4</i> /// /// <i>HLA-DRB5</i> /// /// <i>LOC730415</i>	major histocompatibility complex, class II, DR beta 1 /// major histocompatibility complex, class II, DR beta 3 /// major histocompatibility complex, class II, DR beta 4 /// major histocompatibility complex, class II, DR beta 5 /// hypothetical protein LOC730415	-3.1
221253_s_at	<i>TXNDC5</i>	thioredoxin domain containing 5	-3.1
229026_at	<i>CDC42SE2</i>	CDC42 small effector 2	-3.2
231721_at	<i>JAM3</i>	junctional adhesion molecule 3	-3.2
237594_at	---	---	-3.4
1558438_a_at	---	Immunoglobulin epsilon chain	-3.4
201292_at	<i>TOP2A</i>	topoisomerase (DNA) II alpha 170kDa	-3.4
236419_at	---	Transcribed locus	-3.4

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

203764_at	<i>DLG7</i>	discs, large homolog 7 (Drosophila)	-3.5
240608_at	---	---	-3.6
219308_s_at	<i>AK5</i>	adenylate kinase 5	-3.7
206641_at	<i>TNFRSF17</i>	tumor necrosis factor receptor superfamily, member 17	-3.8
216560_x_at	<i>IG@</i>	immunoglobulin lambda locus	-3.9
202503_s_at	<i>KIAA0101</i>	KIAA0101	-4.0
235839_at	---	Transcribed locus, strongly similar to XP_001139021.1 hypothetical protein [Pan troglodytes]	-4.1
1555039_a_at	<i>ABCC4</i>	ATP-binding cassette, sub-family C (CFTR/MRP), member 4	-4.2
201890_at	<i>RRM2</i>	ribonucleotide reductase M2 polypeptide	-4.2
216491_x_at	<i>IGHM</i>	immunoglobulin heavy constant mu	-4.3
221634_at	<i>RPL23AP7</i>	ribosomal protein L23a pseudogene 7	-4.4
205826_at	<i>MYOM2</i>	myomesin (M-protein) 2, 165kDa	-4.6
218948_at	<i>QRSL1</i>	glutaminyl-tRNA synthase (glutamine-hydrolyzing)-like 1	-4.8
206632_s_at	<i>APOBEC3B</i>	apolipoprotein B mRNA editing enzyme, catalytic polypeptide-like 3B	-5.1
212592_at	<i>IGJ</i>	Immunoglobulin J polypeptide, linker protein for immunoglobulin alpha and mu polypeptides	-5.1
202870_s_at	<i>CDC20</i>	cell division cycle 20 homolog (S. cerevisiae)	-5.2
221122_at	<i>HRASLS2</i>	HRAS-like suppressor 2	-5.2
204836_at	<i>GLDC</i>	glycine dehydrogenase (decarboxylating)	-5.6
209773_s_at	<i>RRM2</i>	ribonucleotide reductase M2 polypeptide	-5.6
218663_at	<i>NCAPG</i>	non-SMC condensin I complex, subunit G	-6.1
202338_at	<i>TK1</i>	thymidine kinase 1, soluble	-6.6
219918_s_at	<i>ASPM</i>	asp (abnormal spindle) homolog, microcephaly associated (Drosophila)	-7.0
209642_at	<i>BUB1</i>	BUB1 budding uninhibited by benzimidazoles 1 homolog (yeast)	-7.1
1561276_at	<i>DOCK5</i>	dedicator of cytokinesis 5	-7.2
207245_at	<i>UGT2B17</i>	UDP glucuronosyltransferase 2 family, polypeptide B17	-7.3
207324_s_at	<i>DSC1</i>	desmocollin 1	-8.3
209686_at	<i>S100B</i>	S100 calcium binding protein B	-10.0
240703_s_at	<i>HERC1</i>	hect (homologous to the E6-AP (UBE3A) carboxyl terminus) domain and RCC1 (CHC1)-like domain (RLD) 1	-14.2

* dysregulation shown as fold-change compared to healthy controls

Suppl. Table 3A. Gene ontology categorization of genes dysregulated in CLN3 patients with slow disease progression

Gene Symbol	Gene Name	Fold Change*
NEURON, BRAIN, NERVOUS SYSTEM		
<i>GJB6</i> /// <i>IGKC</i>	immunoglobulin kappa constant /// netrin 2-like (chicken) ///	-13.9

<i>NTN2L</i>	gap junction protein, beta 6	
<i>NTN2L</i>	Netrin 2-like (chicken)	-10.2
<i>EPHB1</i>	EPH receptor B1	-9.1
<i>NRG1</i>	neuregulin 1	4.9
<i>DOCK7</i>	dedicator of cytokinesis 7	3.9
<i>PNOC</i>	prepronociceptin	-3.8
<i>PAX5</i>	paired box 5	-3.6
<i>NR4A2</i>	nuclear receptor subfamily 4, group A, member 2	3.6
<i>NEFL</i>	neurofilament, light polypeptide 68kDa	-3.5
<i>CNTNAP2</i>	contactin associated protein-like 2	-3.1
<i>SYPL1</i>	synaptophysin-like 1	-3.0
<i>C7orf16</i>	chromosome 7 open reading frame 16	-2.6
<i>HHEX</i>	hematopoietically expressed homeobox	-2.5
<i>VEGFA</i>	vascular endothelial growth factor A	2.2
<i>CDKN1C</i>	cyclin-dependent kinase inhibitor 1C (p57, Kip2)	-2.2
<i>EPHA4</i>	EPH receptor A4	2.2
<i>GM2A</i>	GM2 ganglioside activator	-2.1
<i>CLN5</i>	ceroid-lipofuscinosis, neuronal 5	-2.1
<i>GM2A</i>	GM2 ganglioside activator	-2.0
<i>LPHN1</i>	latrophilin 1	2.0
<i>MBP</i>	myelin basic protein	-1.9
<i>COLQ</i>	collagen-like tail subunit (single strand of homotrimer) of asymmetric acetylcholinesterase	1.9
<i>MEF2C</i>	myocyte enhancer factor 2C	-1.9
<i>NET1</i>	neuroepithelial cell transforming gene 1	-1.8
<i>RAB23</i>	RAB23, member RAS oncogene family	1.7
PROTEOLYSIS, LYSOSOME		
<i>IGH</i> /// <i>IGHG1</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma	
<i>IGHG2</i> ///	1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m	
<i>IGHG3</i> ///	marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) ///	
<i>IGHM</i> ///	immunoglobulin heavy constant mu /// immunoglobulin heavy	
<i>IGHV4-31</i>	variable 4-31	-15.2

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

<i>ADAM28</i>	ADAM metallopeptidase domain 28	-3.1
<i>DPP4</i>	dipeptidyl-peptidase 4 (CD26, adenosine deaminase complexing protein 2)	-3.1
<i>HLA-DRA</i>	major histocompatibility complex, class II, DR alpha	-3.1
<i>BACE2</i>	beta-site APP-cleaving enzyme 2	-2.8
<i>NAPSB</i>	napsin B aspartic peptidase pseudogene	-2.6
<i>UBE2D3</i>	ubiquitin-conjugating enzyme E2D 3 (UBC4/5 homolog, yeast)	-2.3
<i>IDUA</i>	iduronidase, alpha-L-	2.3
<i>IDS</i>	iduronate 2-sulfatase (Hunter syndrome)	2.2
<i>HLA-DOB</i>	major histocompatibility complex, class II, DO beta	-2.2
<i>ZNRF1</i>	zinc and ring finger 1	2.1
<i>LAMP3</i>	lysosomal-associated membrane protein 3	-1.9
<i>NPEPL1</i>	Aminopeptidase-like 1	1.9
<i>CTSK</i>	cathepsin K	1.9
<i>ABHD5</i>	abhydrolase domain containing 5	1.8
<i>ST14</i>	suppression of tumorigenicity 14 (colon carcinoma)	-1.8
<i>PSMB10</i>	proteasome (prosome, macropain) subunit, beta type, 10	-1.8
<i>DYNC1H1</i>	dynein, cytoplasmic 1, heavy chain 1	1.7

ENDOCYTOSIS, PROTEIN TRANSPORT, AUTOPHAGY, ENDOSOME

<i>EXOC7</i> /// <i>IGHA1</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>ZCWPW2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-15.8
<i>EXOC7</i> /// <i>IGH</i> /// <i>IGHA1</i> /// <i>IGHA2</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHG2</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>RAC1</i> /// <i>SIX6</i> /// <i>ZCWPW2</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant alpha 2 (A2m marker) /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// SIX homeobox 6 /// ras-related C3 botulinum toxin substrate 1 (rho family, small GTP binding protein Rac1) /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-6.7

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

<i>ARL17P1</i>	ADP-ribosylation factor-like 17 pseudogene 1	4.9
<i>IGHM</i>	immunoglobulin heavy constant mu	-4.6
<i>STAP1</i>	signal transducing adaptor family member 1	-3.9
<i>IGHA1</i> /// <i>IGHG1</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-3.1
<i>ARL17</i>	ADP-ribosylation factor-like 17	3.1
<i>SYTL3</i>	synaptotagmin-like 3	3.0
<i>GDI2</i>	GDP dissociation inhibitor 2	-2.8
<i>MARCH3</i>	membrane-associated ring finger (C3HC4) 3	-2.6
<i>STAB1</i>	stabilin 1	2.5
<i>LOC144571</i>	hypothetical protein LOC144571	2.5
<i>SYS1</i>	SYS1 Golgi-localized integral membrane protein homolog (S. cerevisiae)	-2.4
<i>SYTL2</i>	synaptotagmin-like 2	2.3
<i>STX2</i>	syntaxin 2	2.3
<i>NUP54</i>	nucleoporin 54kDa	-2.3
<i>RAB1A</i>	RAB1A, member RAS oncogene family	-2.2
<i>COG5</i>	component of oligomeric golgi complex 5	-2.2
<i>SEC61A2</i>	Sec61 alpha 2 subunit (S. cerevisiae)	2.1
<i>TLOC1</i>	translocation protein 1	-2.1
<i>YIPF5</i>	Yip1 domain family, member 5	-2.0
<i>EXOC7</i>	exocyst complex component 7	-2.0
<i>ZFYVE16</i>	zinc finger, FYVE domain containing 16	-2.0
<i>CCDC91</i>	Coiled-coil domain containing 91	-1.9
<i>RIN3</i>	Ras and Rab interactor 3	-1.9
<i>SORL1</i>	sortilin-related receptor, L(DLR class) A repeats-containing	1.9
<i>VPS13B</i>	vacuolar protein sorting 13 homolog B (yeast)	1.9
<i>SEC31B</i>	SEC31 homolog B (S. cerevisiae)	1.9
<i>TSC2</i>	tuberous sclerosis 2	-1.8

<i>GGA3</i>	golgi associated, gamma adaptin ear containing, ARF binding protein 3	1.8
<i>CLTA</i>	clathrin, light chain (Lca)	1.8
<i>VPS52</i>	vacuolar protein sorting 52 homolog (S. cerevisiae)	1.7
<i>SNX2</i>	sorting nexin 2	-1.7
<i>APIG1</i>	adaptor-related protein complex 1, gamma 1 subunit	1.7
<i>GABARAPL1</i> /// <i>GABARAPL3</i>	GABA(A) receptor-associated protein like 1 /// GABA(A) receptors associated protein like 3	1.7
<i>SNX9</i>	sorting nexin 9	-1.7
<i>RPL3</i>	ribosomal protein L3	-1.7
<i>ATG2B</i>	ATG2 autophagy related 2 homolog B (S. cerevisiae)	-1.7

APOPTOSIS

<i>PAWR</i>	PRKC, apoptosis, WT1, regulator	-3.4
<i>CUL4A</i>	Cullin 4A	-2.7
<i>BIRC3</i>	baculoviral IAP repeat-containing 3	-2.5
<i>BIRC4</i>	baculoviral IAP repeat-containing 4	-2.5
<i>CD27</i>	CD27 molecule	-2.4
<i>ARHGDI1</i>	Rho GDP dissociation inhibitor (GDI) alpha	-2.3
<i>CADM1</i>	cell adhesion molecule 1	2.3
<i>SIAH1</i>	seven in absentia homolog 1 (Drosophila)	-2.3
<i>EAF2</i>	ELL associated factor 2	-2.3
<i>IRF4</i>	interferon regulatory factor 4	-2.3
<i>PMAIP1</i>	phorbol-12-myristate-13-acetate-induced protein 1	-2.2
<i>IGFBP3</i>	insulin-like growth factor binding protein 3	2.2
<i>TXNDC5</i>	thioredoxin domain containing 5	-2.1
<i>ZBTB16</i>	zinc finger and BTB domain containing 16	2.1
<i>LY86</i>	lymphocyte antigen 86	-2.0
<i>MAL</i>	mal, T-cell differentiation protein	-2.0
<i>MBD4</i>	methyl-CpG binding domain protein 4	-2.0
<i>HSPA1A</i> /// <i>HSPA1B</i>	heat shock 70kDa protein 1A /// heat shock 70kDa protein 1B	-2.0

<i>C11orf17</i> /// <i>NUAK2</i>	chromosome 11 open reading frame 17 /// NUAk family, SNF1-like kinase, 2	-2.0
<i>FOXO1</i>	forkhead box O1	-1.9
<i>PARP1</i>	poly (ADP-ribose) polymerase family, member 1	-1.9
<i>NOD2</i>	nucleotide-binding oligomerization domain containing 2	1.9
<i>HSPB1</i>	heat shock 27kDa protein 1	-1.9
<i>MDM4</i>	Mdm4, transformed 3T3 cell double minute 4, p53 binding protein (mouse)	-1.8
<i>FADD</i>	Fas (TNFRSF6)-associated via death domain	-1.8
<i>CIDEB</i>	cell death-inducing DFFA-like effector b	1.8
<i>TIAL1</i>	TIA1 cytotoxic granule-associated RNA binding protein-like 1	1.8
<i>STEAP3</i>	STEAP family member 3	1.8
<i>FAIM3</i>	Fas apoptotic inhibitory molecule 3	-1.8
<i>TFG</i>	TRK-fused gene	1.8
<i>TNFAIP3</i>	tumor necrosis factor, alpha-induced protein 3	1.8
<i>CKAP2</i>	cytoskeleton associated protein 2	-1.7
<i>SON</i>	SON DNA binding protein	1.7
<i>BCL3</i>	B-cell CLL/lymphoma 3	1.7

VISUAL PERCEPTION, EYE

<i>MERTK</i>	c-mer proto-oncogene tyrosine kinase	3.8
<i>RGS9</i>	regulator of G-protein signaling 9	2.7
<i>COL18A1</i>	collagen, type XVIII, alpha 1	-2.1
<i>CYP4V2</i>	cytochrome P450, family 4, subfamily V, polypeptide 2	2.0
<i>CYP1B1</i>	cytochrome P450, family 1, subfamily B, polypeptide 1	2.0

* dysregulation shown as fold-change compared to healthy controls

** expressed sequence tags

Suppl. Table 3B. Genes dysregulated in CLN3 patients with slow disease progression

Probe Set ID	Gene Symbol	Gene Name	Fold Change*
209728_at	<i>HLA-DRB4</i>	major histocompatibility complex, class II, DR beta 4	343.7
243882_at	---	---	29.9
1556172_at	---	MRNA; cDNA DKFZp76210915 (from clone DKFZp76210915)	16.3
238234_at	---	Transcribed locus	16.3
234082_at	---	CDNA FLJ11831 fis, clone HEMBA1006562	11.1
1558796_a_at	<i>LOC728052</i>	hypothetical protein LOC728052	10.4
211389_x_at	<i>KIR3DS1</i>	killer cell immunoglobulin-like receptor, three domains, short cytoplasmic tail, 1	7.5
205767_at	<i>EREG</i>	epiregulin	7.1
224490_s_at	<i>KIAA1267</i>	KIAA1267	6.8
238127_at	<i>FLJ41484</i>	hypothetical LOC650669	6.5
213369_at	<i>PCDH21</i>	protocadherin 21	6.2
231978_at	<i>TPCN2</i>	two pore segment channel 2	6.1
231484_at	---	Transcribed locus	5.8
229187_at	<i>ZNF542</i>	Zinc finger protein 542 /// CDNA FLJ45377 fis, clone BRHIP3019956	5.8
229715_at	---	CDNA FLJ41663 fis, clone FEBRA2027297	5.4
206080_at	<i>PLCH2</i>	phospholipase C, eta 2	5.4
210718_s_at	<i>ARL17P1</i>	ADP-ribosylation factor-like 17 pseudogene 1	4.9
206237_s_at	<i>NRG1</i>	neuregulin 1	4.8
227492_at	<i>LOC647859</i>	occludin pseudogene	4.8
225308_s_at	<i>TANC1</i>	tetratricopeptide repeat, ankyrin repeat and coiled-coil containing 1	4.8
227870_at	<i>NOPE</i>	neighbor of Punc E11	4.5
225627_s_at	<i>CACHD1</i>	cache domain containing 1	4.4
1561651_s_at	<i>TALI</i>	T-cell acute lymphocytic leukemia 1	4.4
203476_at	<i>TPBG</i>	trophoblast glycoprotein	4.4
232232_s_at	<i>SLC22A16</i>	solute carrier family 22 (organic cation transporter), member 16	4.2
211397_x_at	<i>KIR2DL1</i> /// <i>KIR2DL2</i> /// <i>KIR2DL3</i> /// <i>KIR2DS1</i> /// <i>KIR2DS2</i> /// <i>KIR2DS3</i> /// <i>KIR2DS4</i> /// <i>KIR2DS5</i> /// <i>KIR3DL1</i> /// <i>KIR3DL2</i> /// <i>KIR3DL3</i> /// <i>KIR3DS1</i>	killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 1 /// killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 2 /// killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 3 /// killer cell immunoglobulin-like receptor, two domains, short cytoplasmic tail, 1 /// killer cell immunoglobulin-like receptor, two domains, short cytoplasmic tail, 2 /// killer cell immunoglobulin-like receptor, two domains, short cytoplasmic tail, 3 /// killer cell immunoglobulin-like receptor, two domains, short cytoplasmic tail, 4 /// killer cell immunoglobulin-like receptor, two domains, short cytoplasmic tail, 5 /// killer cell immunoglobulin-like receptor, three domains, long cytoplasmic tail, 1 /// killer cell immunoglobulin-like	4.1

		receptor, three domains, long cytoplasmic tail, 2 /// killer cell immunoglobulin-like receptor, three domains, short cytoplasmic tail, 1 /// killer cell immunoglobulin-like receptor, three domains, long cytoplasmic tail, 3	
242868_at	---	Transcribed locus	4.1
210837_s_at	<i>PDE4D</i>	phosphodiesterase 4D, cAMP-specific (phosphodiesterase E3 dunce homolog, Drosophila)	4.1
202861_at	<i>PER1</i>	period homolog 1 (Drosophila)	4.0
237470_at	<i>DOCK7</i>	dedicator of cytokinesis 7	3.9
232720_at	<i>LINGO2</i>	Leucine rich repeat and Ig domain containing 2	3.9
	<i>ANKRD20A1</i> /// <i>ANKRD20A2</i> /// <i>ANKRD20A3</i> /// <i>ANKRD20A4</i>	ankyrin repeat domain 20 family, member A1 /// chromosome 21 open reading frame 81 /// ankyrin repeat domain 20 family, member A3 /// ankyrin repeat domain 20 family, member A2 /// similar to ankyrin repeat domain 20A /// similar to ankyrin repeat domain 20 family, member A2 /// ankyrin repeat domain 20 family, member A4	3.8
1569607_s_at	/// <i>C21orf81</i> /// <i>LOC643187</i> /// <i>LOC644339</i> /// <i>LOC647595</i> /// <i>LOC653436</i> /// <i>LOC727770</i> /// <i>LOC727806</i>		
206028_s_at	<i>MERTK</i>	c-mer proto-oncogene tyrosine kinase	3.8
210890_x_at	<i>KIR2DL1</i> /// <i>KIR2DL2</i>	killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 1 /// killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 2	3.7
220528_at	<i>VNN3</i>	vanin 3	3.6
204622_x_at	<i>NR4A2</i>	nuclear receptor subfamily 4, group A, member 2	3.5
232793_at	---	Primary neuroblastoma cDNA, clone:Nbla04171, full insert sequence	3.4
208426_x_at	<i>KIR2DL4</i>	killer cell immunoglobulin-like receptor, two domains, long cytoplasmic tail, 4	3.4
229413_s_at	---	---	3.3
1560999_a_at	---	Partial mRNA; ID YG31-1, YG81-3B, LG43-4B2	3.3
238899_at	---	Transcribed locus	3.3
236364_at	---	Transcribed locus	3.3
206481_s_at	<i>LDB2</i>	LIM domain binding 2	3.2
233876_at	---	---	3.2
229028_s_at	<i>ARL17</i>	ADP-ribosylation factor-like 17	3.1
235557_at	<i>LOC150763</i>	hypothetical protein LOC150763	3.1
37117_at	<i>ARHGAP8</i> /// <i>LOC553158</i>	Rho GTPase activating protein 8 /// PRR5-ARHGAP8 fusion	3.1
234738_s_at	<i>KLHDC4</i>	kelch domain containing 4	3.1
202948_at	<i>IL1R1</i>	interleukin 1 receptor, type I	3.0
223315_at	<i>NTN4</i>	netrin 4	3.0
1556839_s_at	<i>SPTBN5</i>	Homo sapiens, clone IMAGE:4704591 /// Spectrin, beta,	3.0

		non-erythrocytic 5	
1562255_at	<i>SYTL3</i>	synaptotagmin-like 3	3.0
237216_at	---	Transcribed locus	2.9
204083_s_at	<i>TPM2</i>	tropomyosin 2 (beta)	2.8
1568680_s_at	<i>YTHDC2</i>	YTH domain containing 2	2.8
239799_at	---	CDNA FLJ34826 fis, clone NT2NE2008803	2.8
227062_at	<i>TncRNA</i>	trophoblast-derived noncoding RNA	2.8
238568_s_at	---	Transcribed locus	2.7
241762_at	<i>FBXO32</i>	F-box protein 32	2.7
227330_x_at	<i>LOC389833</i> /// <i>LOC727764</i> /// <i>LOC728312</i>	similar to hypothetical protein MGC27019 /// similar to Tektin-3	2.7
206518_s_at	<i>RGS9</i>	regulator of G-protein signaling 9	2.7
230451_at	<i>C1orf136</i>	Chromosome 1 open reading frame 136	2.7
215388_s_at	<i>CFH</i> /// <i>CFHR1</i>	complement factor H /// complement factor H-related 1	2.7
202887_s_at	<i>DDIT4</i>	DNA-damage-inducible transcript 4	2.7
209524_at	<i>HDGFRP3</i>	hepatoma-derived growth factor, related protein 3	2.7
207072_at	<i>IL18RAP</i>	interleukin 18 receptor accessory protein	2.7
227394_at	<i>NCAM1</i>	neural cell adhesion molecule 1	2.7
212980_at	<i>USP34</i>	ubiquitin specific peptidase 34	2.7
1557263_s_at	---	CDNA FLJ35536 fis, clone SPLEN2002451	2.6
227152_at	<i>C12orf35</i>	chromosome 12 open reading frame 35	2.6
220293_at	<i>C14orf161</i>	chromosome 14 open reading frame 161	2.6
228325_at	<i>KIAA0146</i>	KIAA0146	2.6
236846_at	<i>LOC284757</i>	hypothetical protein LOC284757	2.6
235987_at	<i>PRKXP1</i>	protein kinase, X-linked, pseudogene 1	2.6
205896_at	<i>SLC22A4</i>	solute carrier family 22 (organic cation transporter), member 4	2.6
235763_at	<i>SLC44A5</i>	solute carrier family 44, member 5	2.6
1568742_at	---	CDNA clone IMAGE:2984900	2.6
243469_at	---	Transcribed locus	2.6
228083_at	<i>CACNA2D4</i>	calcium channel, voltage-dependent, alpha 2/delta subunit 4	2.6
230552_at	<i>LOC284412</i>	hypothetical protein LOC284412	2.6
231357_at	<i>CLEC12B</i>	C-type lectin domain family 12, member B	2.5
214250_at	<i>NUMA1</i>	nuclear mitotic apparatus protein 1	2.5
204787_at	<i>VSIG4</i>	V-set and immunoglobulin domain containing 4	2.5
241403_at	<i>CLK4</i>	CDC-like kinase 4	2.5
229748_x_at	<i>LOC389833</i>	similar to hypothetical protein MGC27019	2.5
230970_at	---	Transcribed locus	2.5
206120_at	<i>CD33</i>	CD33 molecule	2.5
204142_at	<i>ENOSF1</i>	enolase superfamily member 1	2.5

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

229272_at	<i>FNBP4</i>	formin binding protein 4	2.5
242903_at	<i>IFNGR1</i>	interferon gamma receptor 1	2.5
211687_x_at	<i>KIR3DL1</i>	killer cell immunoglobulin-like receptor, three domains, long cytoplasmic tail, 1	2.5
1564139_at	<i>LOC144571</i>	hypothetical protein LOC144571	2.5
38487_at	<i>STAB1</i>	stabilin 1	2.5
204794_at	<i>DUSP2</i>	dual specificity phosphatase 2	2.4
205100_at	<i>GFPT2</i>	glutamine-fructose-6-phosphate transaminase 2	2.4
225305_at	<i>SLC25A29</i>	solute carrier family 25, member 29	2.4
1555962_at	<i>B3GNT7</i>	UDP-GlcNAc:betaGal beta-1,3-N-acetylglucosaminyltransferase 7	2.4
1569401_at	<i>CLEC12A</i>	C-type lectin domain family 12, member A	2.4
233819_s_at	<i>ZNF294</i>	zinc finger protein 294	2.4
235798_at	---	---	2.3
242494_at	---	Transcribed locus	2.3
228962_at	---	Transcribed locus	2.3
236571_at	---	Transcribed locus	2.3
228372_at	<i>C10orf128</i>	chromosome 10 open reading frame 128	2.3
209031_at	<i>CADM1</i>	cell adhesion molecule 1	2.3
210070_s_at	<i>CHKB</i> /// <i>CPT1B</i>	choline kinase beta /// carnitine palmitoyltransferase 1B (muscle)	2.3
209626_s_at	<i>OSBPL3</i>	oxysterol binding protein-like 3	2.3
232914_s_at	<i>SYTL2</i>	synaptotagmin-like 2	2.3
244677_at	---	Transcribed locus	2.3
240347_at	---	Transcribed locus	2.3
209967_s_at	<i>CREM</i>	cAMP responsive element modulator	2.3
205059_s_at	<i>IDUA</i>	iduronidase, alpha-L-	2.3
210479_s_at	<i>RORA</i>	RAR-related orphan receptor A	2.3
213434_at	<i>STX2</i>	syntaxin 2	2.3
214686_at	<i>ZNF266</i>	zinc finger protein 266	2.3
221973_at	---	CDNA clone IMAGE:5217021, with apparent retained intron	2.3
235376_at	---	Transcribed locus	2.3
209156_s_at	<i>COL6A2</i>	collagen, type VI, alpha 2	2.3
233937_at	<i>GGNBP2</i>	gametogenetin binding protein 2	2.3
1564150_a_at	<i>LOC256021</i>	hypothetical protein LOC256021	2.3
208003_s_at	<i>NFAT5</i>	nuclear factor of activated T-cells 5, tonicity-responsive	2.3
230563_at	<i>RASGEF1A</i>	RasGEF domain family, member 1A	2.3
1558354_s_at	---	CDNA clone IMAGE:5260583	2.2
235008_at	---	CDNA FLJ25241 fis, clone STM02689	2.2
234196_at	---	CDNA: FLJ21377 fis, clone COL03255	2.2
228742_at	---	Full length insert cDNA clone YQ54B06	2.2

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

210425_x_at	<i>GOLGA8B</i>	golgi autoantigen, golgin subfamily a, 8B	2.2
1556026_at	<i>IDS</i>	iduronate 2-sulfatase (Hunter syndrome)	2.2
210095_s_at	<i>IGFBP3</i>	insulin-like growth factor binding protein 3	2.2
235567_at	<i>LOC283666</i>	Hypothetical protein LOC283666	2.2
230664_at	<i>MGC39900</i>	hypothetical protein MGC39900	2.2
236475_at	<i>MICAL2</i>	Microtubule associated monooxygenase, calponin and LIM domain containing 2	2.2
228846_at	<i>MXD1</i>	MAX dimerization protein 1	2.2
217507_at	<i>SLC11A1</i>	solute carrier family 11 (proton-coupled divalent metal ion transporters), member 1	2.2
227108_at	<i>STARD9</i>	StAR-related lipid transfer (START) domain containing 9	2.2
204619_s_at	<i>VCAN</i>	versican	2.2
210512_s_at	<i>VEGFA</i>	vascular endothelial growth factor A	2.2
224261_at	---	PRO3098	2.2
204497_at	<i>ADCY9</i>	adenylate cyclase 9	2.2
227113_at	<i>ADHFE1</i>	alcohol dehydrogenase, iron containing, 1	2.2
203973_s_at	<i>CEBPD</i>	CCAAT/enhancer binding protein (C/EBP), delta	2.2
228318_s_at	<i>CRIPAK</i>	cysteine-rich PAK1 inhibitor	2.2
224215_s_at	<i>DLL1</i>	delta-like 1 (Drosophila)	2.2
229374_at	<i>EPHA4</i>	EPH receptor A4	2.2
58780_s_at	<i>FLJ10357</i>	hypothetical protein FLJ10357	2.2
213510_x_at	<i>LOC220594</i>	TL132 protein	2.2
219033_at	<i>PARP8</i>	poly (ADP-ribose) polymerase family, member 8	2.2
212753_at	<i>PCGF3</i>	polycomb group ring finger 3	2.2
220952_s_at	<i>PLEKHA5</i>	pleckstrin homology domain containing, family A member 5	2.2
236782_at	<i>SAMD3</i>	sterile alpha motif domain containing 3	2.2
241620_at	<i>SMCHD1</i>	structural maintenance of chromosomes flexible hinge domain containing 1	2.2
236833_at	<i>TTC16</i>	tetratricopeptide repeat domain 16	2.2
214131_at	<i>CYorf15B</i>	chromosome Y open reading frame 15B	2.1
213328_at	<i>NEK1</i>	NIMA (never in mitosis gene a)-related kinase 1	2.1
218888_s_at	<i>NETO2</i>	neuropilin (NRP) and tolloid (TLL)-like 2	2.1
219499_at	<i>SEC61A2</i>	Sec61 alpha 2 subunit (S. cerevisiae)	2.1
1561880_a_at	<i>SIGLECP16</i>	sialic acid binding Ig-like lectin, pseudogene 16	2.1
201996_s_at	<i>SPEN</i>	spen homolog, transcriptional regulator (Drosophila)	2.1
51192_at	<i>SSH3</i>	slingshot homolog 3 (Drosophila)	2.1
213396_s_at	---	Full-length cDNA clone CS0DJ012YG05 of T cells (Jurkat cell line) Cot 10-normalized of Homo sapiens (human)	2.1
1562529_s_at	---	Homo sapiens, clone IMAGE:5747561, mRNA	2.1
228529_at	---	Transcribed locus	2.1
240173_at	---	Transcribed locus	2.1

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

212599_at	<i>AUTS2</i>	autism susceptibility candidate 2	2.1
207556_s_at	<i>DGKZ</i>	diacylglycerol kinase, zeta 104kDa	2.1
224965_at	<i>GNG2</i>	guanine nucleotide binding protein (G protein), gamma 2	2.1
213931_at	<i>ID2 /// ID2B</i>	inhibitor of DNA binding 2, dominant negative helix-loop-helix protein /// inhibitor of DNA binding 2B, dominant negative helix-loop-helix protein	2.1
206618_at	<i>IL18R1</i>	interleukin 18 receptor 1	2.1
206214_at	<i>PLA2G7</i>	phospholipase A2, group VII (platelet-activating factor acetylhydrolase, plasma)	2.1
226022_at	<i>SASH1</i>	SAM and SH3 domain containing 1	2.1
225962_at	<i>ZNRF1</i>	zinc and ring finger 1	2.1
227755_at	---	CDNA clone IMAGE:4077090	2.1
1559156_at	---	MRNA; cDNA DKFZp686B1142 (from clone DKFZp686B1142)	2.1
219359_at	<i>ATHL1</i>	ATH1, acid trehalase-like 1 (yeast)	2.1
228891_at	<i>C9orf164</i>	chromosome 9 open reading frame 164	2.1
214873_at	<i>LRP5L</i>	low density lipoprotein receptor-related protein 5-like	2.1
205593_s_at	<i>PDE9A</i>	phosphodiesterase 9A	2.1
38671_at	<i>PLXND1</i>	plexin D1	2.1
1558217_at	<i>SLFN13</i>	schlafen family member 13	2.1
214755_at	<i>UAP1L1</i>	UDP-N-acetylglucosamine pyrophosphorylase 1-like 1	2.1
225067_at	<i>ULK3</i>	unc-51-like kinase 3 (C. elegans)	2.1
205883_at	<i>ZBTB16</i>	zinc finger and BTB domain containing 16	2.1
233309_at	---	CDNA FLJ11759 fis, clone HEMBA1005616	2.0
233921_s_at	---	CDNA FLJ12016 fis, clone HEMBB1001707	2.0
228001_at	---	CDNA FLJ26146 fis, clone ADG00290	2.0
228426_at	<i>CLEC2D</i>	C-type lectin domain family 2, member D	2.0
231911_at	<i>KIAA1189</i>	KIAA1189	2.0
225508_at	<i>KIAA1468</i>	KIAA1468	2.0
212307_s_at	<i>OGT</i>	O-linked N-acetylglucosamine (GlcNAc) transferase (UDP-N-acetylglucosamine:polypeptide-N-acetylglucosaminyl transferase)	2.0
204099_at	<i>SMARCD3</i>	SWI/SNF related, matrix associated, actin dependent regulator of chromatin, subfamily d, member 3	2.0
1560259_at	---	Homo sapiens, clone IMAGE:4612205, mRNA	2.0
241824_at	---	Transcribed locus	2.0
228854_at	---	Transcribed locus	2.0
228667_at	<i>AGPAT4</i>	1-acylglycerol-3-phosphate O-acyltransferase 4 (lysophosphatidic acid acyltransferase, delta)	2.0
213715_s_at	<i>ANKRD47</i>	ankyrin repeat domain 47	2.0
228909_at	<i>C21orf86</i>	Chromosome 21 open reading frame 86	2.0
1556323_at	<i>CUGBP2</i>	CUG triplet repeat, RNA binding protein 2	2.0
228391_at	<i>CYP4V2</i>	cytochrome P450, family 4, subfamily V, polypeptide 2	2.0

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

241384_x_at	<i>GK5</i>	glycerol kinase 5 (putative)	2.0
229312_s_at	<i>GKAP1</i>	G kinase anchoring protein 1	2.0
214469_at	<i>HIST1H2AE</i>	histone cluster 1, H2ae	2.0
230753_at	<i>LOC197135</i>	hypothetical LOC197135	2.0
219507_at	<i>RSRC1</i>	arginine/serine-rich coiled-coil 1	2.0
223304_at	<i>SLC37A3</i>	solute carrier family 37 (glycerol-3-phosphate transporter), member 3	2.0
239383_at	<i>ZNF364</i>	zinc finger protein 364	2.0
227810_at	<i>ZNF558</i>	zinc finger protein 558	2.0
226627_at	<i>SEPT8</i>	septin 8	2.0
240188_at	---	---	2.0
232205_at	---	MRNA; cDNA DKFZp434K0722 (from clone DKFZp434K0722)	2.0
240054_at	---	Transcribed locus	2.0
228384_s_at	<i>C10orf33</i>	chromosome 10 open reading frame 33	2.0
202436_s_at	<i>CYP1B1</i>	cytochrome P450, family 1, subfamily B, polypeptide 1	2.0
216260_at	<i>DICER1</i>	Dicer1, Dcr-1 homolog (Drosophila)	2.0
208798_x_at	<i>GOLGA8A</i>	golgi autoantigen, golgin subfamily a, 8A	2.0
226791_at	<i>KIFC2</i>	kinesin family member C2	2.0
47560_at	<i>LPHN1</i>	latrophilin 1	2.0
202336_s_at	<i>PAM</i>	peptidylglycine alpha-amidating monooxygenase	2.0
230494_at	<i>SLC20A1</i>	Solute carrier family 20 (phosphate transporter), member 1	2.0
206073_at	<i>COLQ</i>	collagen-like tail subunit (single strand of homotrimer) of asymmetric acetylcholinesterase	1.9
203591_s_at	<i>CSF3R</i>	colony stimulating factor 3 receptor (granulocyte)	1.9
1554503_a_at	<i>OSCAR</i>	osteoclast associated, immunoglobulin-like receptor	1.9
211106_at	<i>SUPT3H</i>	suppressor of Ty 3 homolog (S. cerevisiae)	1.9
231899_at	<i>ZC3H12C</i>	zinc finger CCCH-type containing 12C	1.9
1558620_at	<i>ZNF621</i>	zinc finger protein 621	1.9
202206_at	<i>ARLAC</i>	ADP-ribosylation factor-like 4C	1.9
213578_at	<i>BMPRIA</i>	bone morphogenetic protein receptor, type IA	1.9
209682_at	<i>CBLB</i>	Cas-Br-M (murine) ecotropic retroviral transforming sequence b	1.9
212851_at	<i>DCUN1D4</i>	DCN1, defective in cullin neddylation 1, domain containing 4 (S. cerevisiae)	1.9
213737_x_at	<i>FLJ32679</i> /// <i>GOLGA8E</i> /// <i>GOLGA8G</i>	golgi autoantigen, golgin subfamily a, 8G /// golgi autoantigen, golgin subfamily a, 8E /// golgin-like hypothetical protein LOC440321	1.9
213343_s_at	<i>GDPD5</i>	glycerophosphodiester phosphodiesterase domain containing 5	1.9
219233_s_at	<i>GSDML</i>	gasdermin-like	1.9
203713_s_at	<i>LLGL2</i>	lethal giant larvae homolog 2 (Drosophila)	1.9
236172_at	<i>LTB4R</i>	Leukotriene B4 receptor	1.9

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
236706_at	<i>LYG1</i>	lysozyme G-like 1	1.9
226225_at	<i>MCC</i>	mutated in colorectal cancers	1.9
220066_at	<i>NOD2</i>	nucleotide-binding oligomerization domain containing 2	1.9
235033_at	<i>NPEPL1</i>	Aminopeptidase-like 1	1.9
218476_at	<i>POMT1</i>	protein-O-mannosyltransferase 1	1.9
213849_s_at	<i>PPP2R2B</i>	protein phosphatase 2 (formerly 2A), regulatory subunit B, beta isoform	1.9
215169_at	<i>SLC35E2</i>	solute carrier family 35, member E2	1.9
1553612_at	<i>ZNF354B</i>	zinc finger protein 354B	1.9
242931_at	---	---	1.9
227929_at	---	CDNA clone IMAGE:5277945	1.9
239476_at	---	CDNA FLJ36491 fis, clone THYMU2018197	1.9
218456_at	<i>CAPRIN2</i>	caprin family member 2	1.9
202450_s_at	<i>CTSK</i>	cathepsin K	1.9
227013_at	<i>LATS2</i>	LATS, large tumor suppressor, homolog 2 (Drosophila)	1.9
223292_s_at	<i>MRPS15</i>	mitochondrial ribosomal protein S15	1.9
219700_at	<i>PLXDC1</i>	plexin domain containing 1	1.9
209889_at	<i>SEC31B</i>	SEC31 homolog B (<i>S. cerevisiae</i>)	1.9
207057_at	<i>SLC16A7</i>	solute carrier family 16, member 7 (monocarboxylic acid transporter 2)	1.9
230707_at	<i>SORL1</i>	sortilin-related receptor, L(DLR class) A repeats-containing	1.9
236254_at	<i>VPS13B</i>	vacuolar protein sorting 13 homolog B (yeast)	1.9
227618_at	---	CDNA FLJ30378 fis, clone BRACE2007953	1.8
235735_at	---	Full length insert cDNA clone ZC64D04	1.8
226885_at	---	Transcribed locus	1.8
243395_at	---	Transcribed locus	1.8
213935_at	<i>ABHD5</i>	abhydrolase domain containing 5	1.8
222023_at	<i>AKAP13</i>	A kinase (PRKA) anchor protein 13	1.8
222761_at	<i>BIVM</i>	basic, immunoglobulin-like variable motif containing	1.8
213956_at	<i>CEP350</i>	centrosomal protein 350kDa	1.8
221188_s_at	<i>CIDEB</i>	cell death-inducing DFFA-like effector b	1.8
226171_at	<i>FLJ20209</i>	hypothetical protein FLJ20209	1.8
208579_x_at	<i>H2BFS</i>	H2B histone family, member S	1.8
227587_at	<i>KRI1</i>	KRI1 homolog (<i>S. cerevisiae</i>)	1.8
230110_at	<i>MCOLN2</i>	mucolipin 2	1.8
233749_at	<i>MSN</i>	Moesin	1.8
219396_s_at	<i>NEIL1</i>	nei endonuclease VIII-like 1 (<i>E. coli</i>)	1.8
212823_s_at	<i>PLEKHG3</i>	pleckstrin homology domain containing, family G (with RhoGef domain) member 3	1.8
222798_at	<i>PTER</i>	phosphotriesterase related	1.8
216834_at	<i>RGS1</i>	regulator of G-protein signaling 1	1.8

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

232231_at	<i>RUNX2</i>	runt-related transcription factor 2	1.8
218494_s_at	<i>SLC2A4RG</i>	SLC2A4 regulator	1.8
218424_s_at	<i>STEAP3</i>	STEAP family member 3	1.8
202405_at	<i>TIAL1</i>	TIA1 cytotoxic granule-associated RNA binding protein-like 1	1.8
243915_at	---	---	1.8
1563629_a_at	---	CDNA FLJ34475 fis, clone HLUNG2003716, moderately similar to RETROVIRUS-RELATED ENV POLYPROTEIN	1.8
209380_s_at	<i>ABCC5</i>	ATP-binding cassette, sub-family C (CFTR/MRP), member 5	1.8
214132_at	<i>ATP5C1</i>	ATP synthase, H ⁺ transporting, mitochondrial F1 complex, gamma polypeptide 1	1.8
213547_at	<i>CAND2</i>	cullin-associated and neddylation-dissociated 2 (putative)	1.8
235721_at	<i>DTX3</i>	deltex 3 homolog (Drosophila)	1.8
209411_s_at	<i>GGA3</i>	golgi associated, gamma adaptin ear containing, ARF binding protein 3	1.8
225341_at	<i>MTERFD3</i>	MTERF domain containing 3	1.8
219304_s_at	<i>PDGFD</i>	platelet derived growth factor D	1.8
218319_at	<i>PEL1I</i>	pellino homolog 1 (Drosophila)	1.8
218956_s_at	<i>PTCD1</i>	pentatricopeptide repeat domain 1	1.8
228806_at	<i>RORC</i>	RAR-related orphan receptor C	1.8
227129_x_at	<i>tcag7.907</i>	hypothetical LOC402483	1.8
226652_at	<i>USP3</i>	ubiquitin specific peptidase 3	1.8
243748_at	---	---	1.8
238545_at	<i>BRD7</i>	Bromodomain containing 7	1.8
229520_s_at	<i>C14orf118</i>	chromosome 14 open reading frame 118	1.8
227668_at	<i>C17orf56</i>	chromosome 17 open reading frame 56	1.8
227188_at	<i>C21orf63</i>	chromosome 21 open reading frame 63	1.8
239203_at	<i>C7orf53</i>	chromosome 7 open reading frame 53	1.8
1560434_x_at	<i>CLTA</i>	clathrin, light chain (Lca)	1.8
228410_at	<i>GAB3</i>	GRB2-associated binding protein 3	1.8
1555913_at	<i>GON4L</i>	gon-4-like (C. elegans)	1.8
214481_at	<i>HIST1H2AM</i>	Histone cluster 1, H2am	1.8
235421_at	<i>MAP3K8</i>	Mitogen-activated protein kinase kinase kinase 8 /// CDNA clone IMAGE:4689481	1.8
236295_s_at	<i>NLRC3</i>	NLR family, CARD domain containing 3	1.8
226843_s_at	<i>PAPD5</i>	PAP associated domain containing 5	1.8
1564310_a_at	<i>PARP15</i>	poly (ADP-ribose) polymerase family, member 15	1.8
1553167_a_at	<i>SEPSECS</i>	Sep (O-phosphoserine) tRNA:Sec (selenocysteine) tRNA synthase	1.8
201070_x_at	<i>SF3B1</i>	splicing factor 3b, subunit 1, 155kDa	1.8
228497_at	<i>SLC22A15</i>	solute carrier family 22 (organic cation transporter),	1.8

		member 15	
219090_at	<i>SLC24A3</i>	solute carrier family 24 (sodium/potassium/calcium exchanger), member 3	1.8
239385_at	<i>TFG</i>	TRK-fused gene	1.8
226604_at	<i>TMTC3</i>	transmembrane and tetratricopeptide repeat containing 3	1.8
202644_s_at	<i>TNFAIP3</i>	tumor necrosis factor, alpha-induced protein 3	1.8
208662_s_at	<i>TTC3</i>	tetratricopeptide repeat domain 3	1.8
230083_at	<i>USP53</i>	ubiquitin specific peptidase 53	1.8
219595_at	<i>ZNF26</i>	zinc finger protein 26	1.8
214760_at	<i>ZNF337</i>	zinc finger protein 337	1.8
243006_at	---	CDNA FLJ30333 fis, clone BRACE2007262	1.7
207275_s_at	<i>ACSL1</i>	acyl-CoA synthetase long-chain family member 1	1.7
203350_at	<i>APIG1</i>	adaptor-related protein complex 1, gamma 1 subunit	1.7
242562_at	<i>DPH4</i>	DPH4, JJJ3 homolog (S. cerevisiae)	1.7
229115_at	<i>DYNC1H1</i>	dynein, cytoplasmic 1, heavy chain 1	1.7
211068_x_at	<i>FAM21C</i> /// <i>FAM21D</i>	family with sequence similarity 21, member C /// family with sequence similarity 21, member D	1.7
211458_s_at	<i>GABARAPL1</i> /// <i>GABARAPL3</i>	GABA(A) receptor-associated protein like 1 /// GABA(A) receptors associated protein like 3	1.7
202364_at	<i>MXII</i>	MAX interactor 1	1.7
207815_at	<i>PF4V1</i>	platelet factor 4 variant 1	1.7
223463_at	<i>RAB23</i>	RAB23, member RAS oncogene family	1.7
214585_s_at	<i>VPS52</i>	vacuolar protein sorting 52 homolog (S. cerevisiae)	1.7
1569052_at	---	CDNA clone IMAGE:3840062	1.7
222048_at	---	CDNA clone MGC:43891 IMAGE:5274560	1.7
243465_at	---	CDNA FLJ32348 fis, clone PROST2007200	1.7
230741_at	---	Full length insert cDNA clone YX74D05	1.7
1558529_s_at	---	Homo sapiens, clone IMAGE:4778855	1.7
40472_at	<i>AGPAT7</i>	1-acylglycerol-3-phosphate O-acyltransferase 7 (lysophosphatidic acid acyltransferase, eta)	1.7
226684_at	<i>ATG2B</i>	ATG2 autophagy related 2 homolog B (S. cerevisiae)	1.7
204908_s_at	<i>BCL3</i>	B-cell CLL/lymphoma 3	1.7
228111_s_at	<i>DNAH1</i>	dynein, axonemal, heavy chain 1	1.7
206589_at	<i>GFI1</i>	growth factor independent 1	1.7
239247_at	<i>LOC401577</i>	Hypothetical protein LOC401577	1.7
205953_at	<i>LRIG2</i>	leucine-rich repeats and immunoglobulin-like domains 2	1.7
212809_at	<i>NFATC2IP</i>	nuclear factor of activated T-cells, cytoplasmic, calcineurin-dependent 2 interacting protein	1.7
243296_at	<i>PBEF1</i>	Pre-B-cell colony enhancing factor 1	1.7
227645_at	<i>PIK3R5</i>	phosphoinositide-3-kinase, regulatory subunit 5, p101	1.7
203056_s_at	<i>PRDM2</i>	PR domain containing 2, with ZNF domain	1.7
201085_s_at	<i>SON</i>	SON DNA binding protein	1.7

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

225246_at	<i>STIM2</i>	stromal interaction molecule 2	1.7
225819_at	<i>TBRG1</i>	transforming growth factor beta regulator 1	1.7
212756_s_at	<i>UBR2</i>	ubiquitin protein ligase E3 component n-recognin 2	1.7
212007_at	<i>UBXD2</i>	UBX domain containing 2	1.7
214659_x_at	<i>YLPM1</i>	YLP motif containing 1	1.7
205437_at	<i>ZNF211</i>	zinc finger protein 211	1.7
235014_at	---	---	-1.7
1557810_at	---	Clone HEA5 Cri-du-chat critical region mRNA	-1.7
227974_at	---	Transcribed locus	-1.7
217588_at	<i>CATSPER2</i> /// <i>CATSPER2P1</i>	cation channel, sperm associated 2 /// cation channel, sperm associated 2 pseudogene 1	-1.7
212042_x_at	<i>hCG_31916</i> /// <i>RPL7</i>	ribosomal protein L7 /// hCG31916	-1.7
1555960_at	<i>HINT1</i>	Histidine triad nucleotide binding protein 1	-1.7
214084_x_at	<i>LOC648998</i> /// <i>LOC652625</i> /// <i>LOC652699</i> /// <i>NCF1</i> /// <i>NCF1B</i> /// <i>NCF1C</i>	similar to Neutrophil cytosol factor 1 (NCF-1) (Neutrophil NADPH oxidase factor 1) (47 kDa neutrophil oxidase factor) (p47-phox) (NCF-47K) (47 kDa autosomal chronic granulomatous disease protein) (NOXO2) /// neutrophil cytosolic factor 1, (chronic granulomatous disease, autosomal 1) /// neutrophil cytosolic factor 1B pseudogene /// neutrophil cytosolic factor 1C pseudogene	-1.7
202732_at	<i>PKIG</i>	protein kinase (cAMP-dependent, catalytic) inhibitor gamma	-1.7
211073_x_at	<i>RPL3</i>	ribosomal protein L3	-1.7
212266_s_at	<i>SFRS5</i>	splicing factor, arginine/serine-rich 5	-1.7
223396_at	<i>TMEM60</i>	transmembrane protein 60	-1.7
238559_at	---	CDNA clone IMAGE:5266242	-1.7
234987_at	---	Full-length cDNA clone CS0DI067YM20 of Placenta Cot 25-normalized of Homo sapiens (human)	-1.7
236198_at	---	Transcribed locus	-1.7
228910_at	---	Transcribed locus	-1.7
213528_at	<i>Clorf156</i>	chromosome 1 open reading frame 156	-1.7
219670_at	<i>Clorf165</i>	chromosome 1 open reading frame 165	-1.7
1559739_at	<i>CHPT1</i>	Choline phosphotransferase 1	-1.7
218252_at	<i>CKAP2</i>	cytoskeleton associated protein 2	-1.7
1553842_at	<i>CXorf20</i>	chromosome X open reading frame 20	-1.7
1554306_at	<i>ITPKB</i>	inositol 1,4,5-trisphosphate 3-kinase B	-1.7
232145_at	<i>LOC388969</i>	hypothetical LOC388969	-1.7
200029_at	<i>LOC392557</i> /// <i>RPL19</i>	ribosomal protein L19 /// similar to ribosomal protein L19	-1.7
200781_s_at	<i>LOC646819</i> /// <i>RPS15A</i>	ribosomal protein S15a /// similar to ribosomal protein S15a	-1.7
214753_at	<i>PFAAP5</i>	Phosphonoformate immuno-associated protein 5	-1.7
202113_s_at	<i>SNX2</i>	sorting nexin 2	-1.7

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
223028_s_at	<i>SNX9</i>	sorting nexin 9	-1.7
1569142_at	<i>TRIM13</i>	tripartite motif-containing 13	-1.7
217823_s_at	<i>UBE2J1</i>	ubiquitin-conjugating enzyme E2, J1 (UBC6 homolog, yeast)	-1.7
239946_at	---	Transcribed locus	-1.8
243798_at	---	Transcribed locus	-1.8
39248_at	<i>AQP3</i>	aquaporin 3 (Gill blood group)	-1.8
202224_at	<i>CRK</i>	v-crk sarcoma virus CT10 oncogene homolog (avian)	-1.8
1570415_at	<i>DDX52</i>	DEAD (Asp-Glu-Ala-Asp) box polypeptide 52	-1.8
227614_at	<i>HKDC1</i>	hexokinase domain containing 1	-1.8
202594_at	<i>LEPROTL1</i>	leptin receptor overlapping transcript-like 1	-1.8
217092_x_at	<i>LOC646912</i>	similar to 60S ribosomal protein L7	-1.8
235061_at	<i>PPMIK</i>	protein phosphatase 1K (PP2C domain containing)	-1.8
239188_at	<i>PPP2R3C</i>	Protein phosphatase 2 (formerly 2A), regulatory subunit B", gamma	-1.8
202659_at	<i>PSMB10</i>	proteasome (prosome, macropain) subunit, beta type, 10	-1.8
218842_at	<i>RPAP3</i>	RNA polymerase II associated protein 3	-1.8
214097_at	<i>RPS21</i>	ribosomal protein S21	-1.8
232338_at	---	CDNA FLJ11553 fis, clone HEMBA1003034	-1.8
235511_at	---	CDNA FLJ36727 fis, clone UTERU2012286	-1.8
244035_at	---	Full length insert cDNA clone YZ11B11	-1.8
236280_at	---	Transcribed locus	-1.8
213892_s_at	<i>APRT</i>	adenine phosphoribosyltransferase	-1.8
	<i>ASNS</i> ///		
	<i>C7orf28A</i> ///	asparagine synthetase ///	
	<i>C7orf28B</i> ///	chromosome 7 open reading frame 28A ///	
	<i>LOC652200</i> ///	chromosome 7 open reading frame 28B ///	
	<i>MGC72080</i>	MGC72080 pseudogene ///	
		similar to CG14980-PB	
1558233_s_at	<i>ATF1</i>	activating transcription factor 1	-1.8
212771_at	<i>C10orf38</i>	chromosome 10 open reading frame 38	-1.8
219006_at	<i>C6orf66</i>	chromosome 6 open reading frame 66	-1.8
205419_at	<i>EBI2</i>	Epstein-Barr virus induced gene 2 (lymphocyte-specific G protein-coupled receptor)	-1.8
203729_at	<i>EMP3</i>	epithelial membrane protein 3	-1.8
232164_s_at	<i>EPPK1</i>	epiplakin 1	-1.8
221601_s_at	<i>FAIM3</i>	Fas apoptotic inhibitory molecule 3	-1.8
206980_s_at	<i>FLT3LG</i>	fms-related tyrosine kinase 3 ligand	-1.8
1558256_at	<i>LOC148189</i>	hypothetical protein LOC148189	-1.8
201755_at	<i>MCM5</i>	minichromosome maintenance complex component 5	-1.8
206036_s_at	<i>REL</i>	v-rel reticuloendotheliosis viral oncogene homolog (avian)	-1.8
226989_at	<i>RGMB</i>	RGM domain family, member B	-1.8
212790_x_at	<i>RPL13A</i>	ribosomal protein L13a	-1.8
213377_x_at	<i>RPS12</i>	ribosomal protein S12	-1.8

Lebrun et al.	Biomarkers and Modifiers of CLN3 Disease		
216905_s_at	<i>ST14</i>	suppression of tumorigenicity 14 (colon carcinoma)	-1.8
207130_at	<i>ZMYND8</i>	zinc finger, MYND-type containing 8	-1.8
219008_at	<i>C2orf43</i>	chromosome 2 open reading frame 43	-1.8
202535_at	<i>FADD</i>	Fas (TNFRSF6)-associated via death domain	-1.8
205306_x_at	<i>KMO</i>	kynurenine 3-monooxygenase (kynurenine 3-hydroxylase)	-1.8
216570_x_at	<i>LOC646417</i>	similar to 60S ribosomal protein L29 (P23)	-1.8
219762_s_at	<i>LOC651600 /// RPL36</i>	ribosomal protein L36 /// similar to ribosomal protein L36	-1.8
225742_at	<i>MDM4</i>	Mdm4, transformed 3T3 cell double minute 4, p53 binding protein (mouse)	-1.8
232483_at	<i>MED17</i>	mediator complex subunit 17	-1.8
220007_at	<i>METTL8</i>	methyltransferase like 8	-1.8
201830_s_at	<i>NET1</i>	neuroepithelial cell transforming gene 1	-1.8
206637_at	<i>P2RY14</i>	purinergic receptor P2Y, G-protein coupled, 14	-1.8
219056_at	<i>RNASEH2B</i>	ribonuclease H2, subunit B	-1.8
200909_s_at	<i>RPLP2</i>	ribosomal protein, large, P2	-1.8
215624_at	<i>TSC2</i>	tuberous sclerosis 2	-1.8
1570108_at	---	CDNA clone IMAGE:4815481	-1.9
1554918_a_at	<i>ABCC4</i>	ATP-binding cassette, sub-family C (CFTR/MRP), member 4	-1.9
218981_at	<i>ACN9</i>	ACN9 homolog (<i>S. cerevisiae</i>)	-1.9
219326_s_at	<i>B3GNT2</i>	UDP-GlcNAc:betaGal beta-1,3-N-acetylglucosaminyltransferase 2	-1.9
224990_at	<i>C4orf34</i>	chromosome 4 open reading frame 34	-1.9
210820_x_at	<i>COQ7</i>	coenzyme Q7 homolog, ubiquinone (yeast)	-1.9
238633_at	<i>EPC1</i>	Enhancer of polycomb homolog 1 (<i>Drosophila</i>)	-1.9
224129_s_at	<i>HDPY-30</i>	dpy-30-like protein	-1.9
1554678_s_at	<i>HNRPDL</i>	heterogeneous nuclear ribonucleoprotein D-like	-1.9
201841_s_at	<i>HSPB1</i>	heat shock 27kDa protein 1	-1.9
210045_at	<i>IDH2</i>	isocitrate dehydrogenase 2 (NADP+), mitochondrial	-1.9
226007_at	<i>ISCA2</i>	iron-sulfur cluster assembly 2 homolog (<i>S. cerevisiae</i>)	-1.9
208961_s_at	<i>KLF6</i>	Kruppel-like factor 6	-1.9
230648_at	<i>LOC283663</i>	hypothetical protein LOC283663	-1.9
203362_s_at	<i>MAD2L1</i>	MAD2 mitotic arrest deficient-like 1 (yeast)	-1.9
201670_s_at	<i>MARCKS</i>	myristoylated alanine-rich protein kinase C substrate	-1.9
1555247_a_at	<i>RAPGEF6</i>	Rap guanine nucleotide exchange factor (GEF) 6	-1.9
204951_at	<i>RHOH</i>	ras homolog gene family, member H	-1.9
220439_at	<i>RIN3</i>	Ras and Rab interactor 3	-1.9
1558540_s_at	<i>SLC2A11</i>	solute carrier family 2 (facilitated glucose transporter), member 11	-1.9
208392_x_at	<i>SP110</i>	SP110 nuclear body protein	-1.9
239555_at	---	CDNA clone IMAGE:40114646	-1.9

221234_s_at	<i>BACH2</i>	BTB and CNC homology 1, basic leucine zipper transcription factor 2	-1.9
1570571_at	<i>CCDC91</i>	Coiled-coil domain containing 91	-1.9
240221_at	<i>CSNK1A1</i>	Casein kinase 1, alpha 1	-1.9
207431_s_at	<i>DEGS1</i>	degenerative spermatocyte homolog 1, lipid desaturase (Drosophila)	-1.9
202724_s_at	<i>FOXO1</i>	forkhead box O1	-1.9
205569_at	<i>LAMP3</i>	lysosomal-associated membrane protein 3	-1.9
209200_at	<i>MEF2C</i>	myocyte enhancer factor 2C	-1.9
217408_at	<i>MRPS18B</i>	mitochondrial ribosomal protein S18B	-1.9
204749_at	<i>NAPIL3</i>	nucleosome assembly protein 1-like 3	-1.9
208644_at	<i>PARP1</i>	poly (ADP-ribose) polymerase family, member 1	-1.9
225738_at	<i>RAPGEF1</i>	Rap guanine nucleotide exchange factor (GEF) 1	-1.9
218564_at	<i>RFWD3</i>	ring finger and WD repeat domain 3	-1.9
218974_at	<i>SOBP</i>	sine oculis binding protein homolog (Drosophila)	-1.9
220391_at	<i>ZBTB3</i>	zinc finger and BTB domain containing 3	-1.9
222760_at	<i>ZNF703</i>	zinc finger protein 703	-1.9
228879_at	---	---	-1.9
242714_at	---	---	-1.9
226773_at	---	MRNA (clone ICRFp50711077)	-1.9
228972_at	---	Transcribed locus	-1.9
227189_at	<i>CPNE5</i>	copine V	-1.9
234339_s_at	<i>GLTSCR2</i>	glioma tumor suppressor candidate region gene 2	-1.9
210136_at	<i>MBP</i>	myelin basic protein	-1.9
233951_at	---	CDNA FLJ12204 fis, clone MAMMA1000921	-2.0
1556888_at	---	CDNA FLJ37963 fis, clone CTONG2009689	-2.0
1559437_at	---	CDNA FLJ40815 fis, clone TRACH2010600	-2.0
213484_at	---	Clone 23700 mRNA sequence	-2.0
220987_s_at	<i>C11orf17</i> /// <i>NUAK2</i>	chromosome 11 open reading frame 17 /// NUA family, SNF1-like kinase, 2	-2.0
201390_s_at	<i>CSNK2B</i>	casein kinase 2, beta polypeptide	-2.0
211742_s_at	<i>EVI2B</i>	ecotropic viral integration site 2B	-2.0
204689_at	<i>HHEX</i>	hematopoietically expressed homeobox	-2.0
226879_at	<i>HVCN1</i>	hydrogen voltage-gated channel 1	-2.0
211747_s_at	<i>LSM5</i>	LSM5 homolog, U6 small nuclear RNA associated (S. cerevisiae)	-2.0
207339_s_at	<i>LTB</i>	lymphotoxin beta (TNF superfamily, member 3)	-2.0
215047_at	<i>TRIM58</i>	tripartite motif-containing 58	-2.0
1560112_at	<i>WDFY2</i>	WD repeat and FYVE domain containing 2	-2.0
1565811_at	---	CDNA clone IMAGE:5278245	-2.0
1557527_at	---	CDNA FLJ33848 fis, clone CTONG2005567	-2.0
1556338_at	---	CDNA FLJ39845 fis, clone SPLEN2014452	-2.0

227605_at	---	Transcribed locus	-2.0
204119_s_at	<i>ADK</i>	adenosine kinase	-2.0
223961_s_at	<i>CISH</i>	cytokine inducible SH2-containing protein	-2.0
213846_at	<i>COX7C</i>	cytochrome c oxidase subunit VIIc	-2.0
214802_at	<i>EXOC7</i>	exocyst complex component 7	-2.0
235310_at	<i>GCET2</i>	germinal center expressed transcript 2	-2.0
200800_s_at	<i>HSPA1A</i> /// <i>HSPA1B</i>	heat shock 70kDa protein 1A /// heat shock 70kDa protein 1B	-2.0
220704_at	<i>IKZF1</i>	IKAROS family zinc finger 1 (Ikaros)	-2.0
205859_at	<i>LY86</i>	lymphocyte antigen 86	-2.0
204777_s_at	<i>MAL</i>	mal, T-cell differentiation protein	-2.0
209580_s_at	<i>MBD4</i>	methyl-CpG binding domain protein 4	-2.0
232020_at	<i>SMURF2</i>	SMAD specific E3 ubiquitin protein ligase 2	-2.0
223822_at	<i>SUSD4</i>	sushi domain containing 4	-2.0
1554638_at	<i>ZFYVE16</i>	zinc finger, FYVE domain containing 16	-2.0
212544_at	<i>ZNHIT3</i>	zinc finger, HIT type 3	-2.0
1556588_at	<i>C15orf37</i>	chromosome 15 open reading frame 37	-2.0
1562028_at	<i>CCND3</i>	Cyclin D3	-2.0
1566448_at	<i>CD6</i>	CD6 molecule	-2.0
213060_s_at	<i>CHI3L2</i>	chitinase 3-like 2	-2.0
235060_at	<i>DKFZp547E087</i>	hypothetical gene LOC283846	-2.0
208886_at	<i>H1FO</i>	H1 histone family, member 0	-2.0
215193_x_at	<i>HLA-DRB1</i> /// <i>HLA-DRB3</i> /// <i>LOC730415</i>	major histocompatibility complex, class II, DR beta 1 /// major histocompatibility complex, class II, DR beta 3 /// hypothetical protein LOC730415	-2.0
1555485_s_at	<i>LOC202134</i>	hypothetical protein LOC202134	-2.0
204961_s_at	<i>NCF1</i> /// <i>NCF1B</i> /// <i>NCF1C</i>	neutrophil cytosolic factor 1, (chronic granulomatous disease, autosomal 1) /// neutrophil cytosolic factor 1B pseudogene /// neutrophil cytosolic factor 1C pseudogene	-2.0
208620_at	<i>PCBP1</i>	poly(rC) binding protein 1	-2.0
320_at	<i>PEX6</i>	peroxisomal biogenesis factor 6	-2.0
205933_at	<i>SETBP1</i>	SET binding protein 1	-2.0
224949_at	<i>YIPF5</i>	Yip1 domain family, member 5	-2.0
239606_at	---	CDNA FLJ25345 fis, clone TST01118	-2.1
1559776_at	---	CDNA FLJ36989 fis, clone BRACE2006753	-2.1
226397_s_at	---	Transcribed locus	-2.1
239164_at	---	Transcribed locus	-2.1
235207_at	---	Transcribed locus	-2.1
225331_at	<i>CCDC50</i>	coiled-coil domain containing 50	-2.1
203547_at	<i>CD4</i>	CD4 molecule	-2.1
209082_s_at	<i>COL18A1</i>	collagen, type XVIII, alpha 1	-2.1

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
219518_s_at	<i>ELL3</i>	elongation factor RNA polymerase II-like 3	-2.1
203665_at	<i>HMOX1</i>	heme oxygenase (decycling) 1	-2.1
239835_at	<i>KBTBD8</i>	kelch repeat and BTB (POZ) domain containing 8	-2.1
221194_s_at	<i>LOC51136</i>	PTD016 protein	-2.1
222347_at	<i>LOC644450</i>	hypothetical protein LOC644450	-2.1
204972_at	<i>OAS2</i>	2'-5'-oligoadenylate synthetase 2, 69/71kDa	-2.1
208121_s_at	<i>PTPRO</i>	protein tyrosine phosphatase, receptor type, O	-2.1
218949_s_at	<i>QRSL1</i>	glutaminyl-tRNA synthase (glutamine-hydrolyzing)-like 1	-2.1
212605_s_at	---	AF034176 Human mRNA (Tripodis and Ragoussis) Homo sapiens cDNA clone ntcon5 contig	-2.1
1561004_at	---	Full length insert cDNA clone YY74A01	-2.1
214214_s_at	<i>CIQBP</i>	complement component 1, q subcomponent binding protein	-2.1
204085_s_at	<i>CLN5</i>	ceroid-lipofuscinosis, neuronal 5	-2.1
231152_at	<i>FLJ20309</i>	Hypothetical protein FLJ20309	-2.1
223423_at	<i>GPR160</i>	G protein-coupled receptor 160	-2.1
214918_at	<i>HNRPM</i>	heterogeneous nuclear ribonucleoprotein M	-2.1
207535_s_at	<i>NFKB2</i>	nuclear factor of kappa light polypeptide gene enhancer in B-cells 2 (p49/p100)	-2.1
242201_at	<i>PMS2L5</i>	Postmeiotic segregation increased 2-like 5	-2.1
201079_at	<i>SYNGR2</i>	synaptogyrin 2	-2.1
1552789_at	<i>TLOC1</i>	translocation protein 1	-2.1
221253_s_at	<i>TXNDC5</i>	thioredoxin domain containing 5	-2.1
232286_at	---	CDNA FLJ12187 fis, clone MAMMA1000831	-2.1
235134_at	---	CDNA FLJ30156 fis, clone BRACE2000487	-2.1
209782_s_at	<i>DBP</i>	D site of albumin promoter (albumin D-box) binding protein	-2.1
226099_at	<i>ELL2</i>	elongation factor, RNA polymerase II, 2	-2.1
210889_s_at	<i>FCGR2B</i> /// <i>FCGR2C</i>	Fc fragment of IgG, low affinity IIb, receptor (CD32) /// Fc fragment of IgG, low affinity IIc, receptor for (CD32)	-2.1
35820_at	<i>GM2A</i>	GM2 ganglioside activator	-2.1
206896_s_at	<i>GNG7</i>	guanine nucleotide binding protein (G protein), gamma 7	-2.1
205466_s_at	<i>HS3ST1</i>	heparan sulfate (glucosamine) 3-O-sulfotransferase 1	-2.1
201015_s_at	<i>JUP</i>	junction plakoglobin	-2.1
217266_at	<i>LOC646672</i> /// <i>LOC653232</i> /// <i>LOC728002</i> /// <i>LOC728088</i> /// <i>LOC728576</i> /// <i>LOC730925</i> /// <i>RPL15</i> /// <i>icag7.350</i>	ribosomal protein L15 /// similar to ribosomal protein L15	-2.1
242191_at	<i>NBPF10</i> /// <i>NBPF11</i>	neuroblastoma breakpoint family, member 11 /// neuroblastoma breakpoint family, member 10	-2.1
218320_s_at	<i>NDUFB11</i>	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 11, 17.3kDa	-2.1

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
222450_at	<i>TMEPAI</i>	transmembrane, prostate androgen induced RNA	-2.1
241702_at	---	---	-2.2
1562467_at	---	CDNA FLJ39766 fis, clone SPLEN2000307	-2.2
234260_at	---	MRNA; cDNA DKFZp434E0572 (from clone DKFZp434E0572)	-2.2
1556451_at	---	MRNA; cDNA DKFZp667B1520 (from clone DKFZp667B1520)	-2.2
237412_at	---	Transcribed locus	-2.2
227533_at	---	Transcribed locus	-2.2
213348_at	<i>CDKN1C</i>	cyclin-dependent kinase inhibitor 1C (p57, Kip2)	-2.2
203630_s_at	<i>COG5</i>	component of oligomeric golgi complex 5	-2.2
226757_at	<i>IFIT2</i>	interferon-induced protein with tetratricopeptide repeats 2	-2.2
212314_at	<i>KIAA0746</i>	KIAA0746 protein	-2.2
1552330_at	<i>MGC16385</i>	hypothetical protein MGC16385	-2.2
1554105_at	<i>TMEM185A</i>	transmembrane protein 185A	-2.2
220494_s_at	---	---	-2.2
233406_at	---	CDNA FLJ12038 fis, clone HEMBB1001922	-2.2
50314_i_at	<i>C20orf27</i>	chromosome 20 open reading frame 27	-2.2
223431_at	<i>CNO</i>	cappuccino homolog (mouse)	-2.2
232879_at	<i>CRTC3</i>	CREB regulated transcription coactivator 3	-2.2
206548_at	<i>hCG_1776259</i>	hypothetical protein FLJ23556	-2.2
205671_s_at	<i>HLA-DOB</i>	major histocompatibility complex, class II, DO beta	-2.2
203153_at	<i>IFIT1</i>	interferon-induced protein with tetratricopeptide repeats 1	-2.2
200771_at	<i>LAMC1</i>	laminin, gamma 1 (formerly LAMB2)	-2.2
	<i>LOC133569</i> ///		
	<i>LOC654029</i> ///	ribosomal protein S10 ///	
216505_x_at	<i>LOC728791</i> ///	/// ribosomal protein S10 pseudogene 3 ///	-2.2
	<i>RPS10</i> ///	ribosomal protein S10	
	<i>RPS10P3</i>		
1557293_at	<i>LOC440993</i>	hypothetical gene supported by AK128346	-2.2
	<i>LOC728903</i> ///		
228040_at	<i>RP11-262H14.4</i>	family with sequence similarity 88, member B ///	-2.2
		hypothetical protein LOC728903	
215069_at	<i>NMT2</i>	N-myristoyltransferase 2	-2.2
204286_s_at	<i>PMAIP1</i>	phorbol-12-myristate-13-acetate-induced protein 1	-2.2
207791_s_at	<i>RAB1A</i>	RAB1A, member RAS oncogene family	-2.2
214245_at	<i>RPS14</i>	ribosomal protein S14	-2.2
209865_at	<i>SLC35A3</i>	solute carrier family 35 (UDP-N-acetylglucosamine (UDP-GlcNAc) transporter), member A3	-2.2
208743_s_at	<i>YWHAB</i>	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, beta polypeptide	-2.2
241610_x_at	---	Transcribed locus	-2.3
244177_at	---	Transcribed locus	-2.3
227198_at	<i>AFF3</i>	AF4/FMR2 family, member 3	-2.3

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
238692_at	<i>BTBD11</i>	BTB (POZ) domain containing 11	-2.3
219551_at	<i>EAF2</i>	ELL associated factor 2	-2.3
226475_at	<i>FAM118A</i>	family with sequence similarity 118, member A	-2.3
227410_at	<i>FAM43A</i>	family with sequence similarity 43, member A	-2.3
1559964_at	<i>FLJ38717</i>	FLJ38717 protein	-2.3
206361_at	<i>GPR44</i>	G protein-coupled receptor 44	-2.3
205133_s_at	<i>HSPE1</i>	heat shock 10kDa protein 1 (chaperonin 10)	-2.3
204562_at	<i>IRF4</i>	interferon regulatory factor 4	-2.3
203144_s_at	<i>KIAA0040</i>	KIAA0040	-2.3
205900_at	<i>KRT1</i>	keratin 1 (epidermolytic hyperkeratosis)	-2.3
1560058_at	<i>LOC399900</i>	hypothetical gene supported by AK093779	-2.3
222632_s_at	<i>LZTFL1</i>	leucine zipper transcription factor-like 1	-2.3
212183_at	<i>NUDT4</i>	nudix (nucleoside diphosphate linked moiety X)-type motif 4	-2.3
203243_s_at	<i>PDLIM5</i>	PDZ and LIM domain 5	-2.3
1552621_at	<i>POLR2J2</i> /// <i>POLR2J3</i>	DNA directed RNA polymerase II polypeptide J-related /// RPB11b2 protein	-2.3
232392_at	<i>SFRS3</i>	Splicing factor, arginine/serine-rich 3	-2.3
232365_at	<i>SIAH1</i>	seven in absentia homolog 1 (Drosophila)	-2.3
243780_at	---	CDNA FLJ46553 fis, clone THYMU3038879	-2.3
1558662_s_at	<i>BANK1</i>	B-cell scaffold protein with ankyrin repeats 1	-2.3
206255_at	<i>BLK</i>	B lymphoid tyrosine kinase	-2.3
224867_at	<i>C1orf151</i>	chromosome 1 open reading frame 151	-2.3
215213_at	<i>NUP54</i>	nucleoporin 54kDa	-2.3
1552343_s_at	<i>PDE7A</i>	phosphodiesterase 7A	-2.3
213687_s_at	<i>RPL35A</i>	ribosomal protein L35a	-2.3
212387_at	<i>TCF4</i>	transcription factor 4	-2.3
242110_at	---	Transcribed locus	-2.3
213606_s_at	<i>ARHGDI1A</i>	Rho GDP dissociation inhibitor (GDI) alpha	-2.3
1563674_at	<i>FCRL2</i>	Fc receptor-like 2	-2.3
200869_at	<i>LOC390354</i> /// <i>RPL18A</i>	ribosomal protein L18a /// similar to ribosomal protein L18a; 60S ribosomal protein L18a	-2.3
218866_s_at	<i>POLR3K</i>	polymerase (RNA) III (DNA directed) polypeptide K, 12.3 kDa	-2.3
240383_at	<i>UBE2D3</i>	ubiquitin-conjugating enzyme E2D 3 (UBC4/5 homolog, yeast)	-2.3
1558430_at	---	CDNA FLJ36648 fis, clone UTERU1000138	-2.4
238611_at	---	Transcribed locus	-2.4
232931_at	<i>ASCC3L1</i>	activating signal cointegrator 1 complex subunit 3-like 1	-2.4
219979_s_at	<i>C11orf73</i>	chromosome 11 open reading frame 73	-2.4
206150_at	<i>CD27</i>	CD27 molecule	-2.4
225081_s_at	<i>CDCA7L</i>	cell division cycle associated 7-like	-2.4

227609_at	<i>EPSTII</i>	epithelial stromal interaction 1 (breast)	-2.4
229390_at	<i>FAM26F</i>	family with sequence similarity 26, member F	-2.4
221004_s_at	<i>ITM2C</i>	integral membrane protein 2C	-2.4
233866_at	<i>KLHL5</i>	kelch-like 5 (Drosophila)	-2.4
205552_s_at	<i>OAS1</i>	2',5'-oligoadenylate synthetase 1, 40/46kDa	-2.4
224670_at	<i>SYS1</i>	SYS1 Golgi-localized integral membrane protein homolog (S. cerevisiae)	-2.4
236226_at	<i>BTLA</i>	B and T lymphocyte associated	-2.4
225334_at	<i>C10orf32</i>	chromosome 10 open reading frame 32	-2.4
202770_s_at	<i>CCNG2</i>	cyclin G2	-2.4
1553856_s_at	<i>P2RY10</i>	purinergic receptor P2Y, G-protein coupled, 10	-2.4
230708_at	<i>PRICKLE1</i>	Prickle homolog 1 (Drosophila)	-2.4
233995_at	---	Clone HQ0663 PRO0663	-2.5
1558418_at	---	Homo sapiens, clone IMAGE:4850148, mRNA	-2.5
229434_at	---	Transcribed locus	-2.5
210538_s_at	<i>BIRC3</i>	baculoviral IAP repeat-containing 3	-2.5
235222_x_at	<i>BIRC4</i>	baculoviral IAP repeat-containing 4	-2.5
229026_at	<i>CDC42SE2</i>	CDC42 small effector 2	-2.5
210279_at	<i>GPR18</i>	G protein-coupled receptor 18	-2.5
229050_s_at	<i>SNHG7</i>	small nucleolar RNA host gene (non-protein coding) 7	-2.5
231972_at	---	CDNA: FLJ21028 fis, clone CAE07155	-2.5
243591_at	---	Transcribed locus	-2.5
219498_s_at	<i>BCL11A</i>	B-cell CLL/lymphoma 11A (zinc finger protein)	-2.5
238071_at	<i>LCN10</i>	lipocalin 10	-2.5
200022_at	<i>RPL18</i>	ribosomal protein L18	-2.5
227891_s_at	<i>TAF15</i>	TAF15 RNA polymerase II, TATA box binding protein (TBP)-associated factor, 68kDa	-2.5
228390_at	---	CDNA clone IMAGE:5259272	-2.5
232319_at	---	CDNA FLJ12360 fis, clone MAMMA1002356	-2.5
232535_at	---	MRNA; cDNA DKFZp434L201 (from clone DKFZp434L201)	-2.5
206398_s_at	<i>CD19</i>	CD19 molecule	-2.5
224735_at	<i>CYBASC3</i>	cytochrome b, ascorbate dependent 3	-2.5
1552622_s_at	<i>LOC441259</i> /// <i>LOC730323</i> /// <i>POLR2J2</i> /// <i>POLR2J3</i>	DNA directed RNA polymerase II polypeptide J-related /// PMS2 postmeiotic segregation increased 2 (S. cerevisiae)- like /// RPB1 lb2 protein /// similar to postmeiotic segregation increased 2-like 2	-2.5
225580_at	<i>MRPL50</i>	mitochondrial ribosomal protein L50	-2.5
200010_at	<i>RPL11</i>	ribosomal protein L11	-2.5
209773_s_at	<i>RRM2</i>	ribonucleotide reductase M2 polypeptide	-2.5
237515_at	<i>TMEM56</i>	transmembrane protein 56	-2.5
210972_x_at	<i>TRA@</i> /// <i>TRAC</i> ///	T cell receptor alpha locus /// T cell receptor delta variable 2 /// T cell receptor alpha variable 20 /// T cell receptor	-2.5

	<i>TRAJ17</i> /// <i>TRAV20</i> /// <i>TRDV2</i>	alpha joining 17 /// T cell receptor alpha constant	
243495_s_at	---	MRNA; cDNA DKFZp686E18224 (from clone DKFZp686E18224)	-2.6
238610_s_at	---	Transcribed locus	-2.6
220231_at	<i>C7orf16</i>	chromosome 7 open reading frame 16	-2.6
206983_at	<i>CCR6</i>	chemokine (C-C motif) receptor 6	-2.6
202503_s_at	<i>KIAA0101</i>	KIAA0101	-2.6
213256_at	<i>MARCH3</i>	membrane-associated ring finger (C3HC4) 3	-2.6
235816_s_at	<i>Rgr</i>	Ral-GDS related protein Rgr	-2.6
217548_at	<i>C15orf38</i>	Chromosome 15 open reading frame 38	-2.6
225795_at	<i>C22orf32</i>	chromosome 22 open reading frame 32	-2.6
232204_at	<i>EBF1</i>	early B-cell factor 1	-2.6
228055_at	<i>NAPSB</i>	napsin B aspartic peptidase pseudogene	-2.6
226117_at	<i>TIFA</i>	TRAF-interacting protein with a forkhead-associated domain	-2.6
201688_s_at	<i>TPD52</i>	tumor protein D52	-2.6
1569128_at	---	Homo sapiens, clone IMAGE:4695648, mRNA	-2.7
236216_at	---	MRNA; cDNA DKFZp313B2312 (from clone DKFZp313B2312)	-2.7
243932_at	---	Transcribed locus	-2.7
1552691_at	<i>ARL11</i>	ADP-ribosylation factor-like 11	-2.7
230896_at	<i>CCDC4</i>	coiled-coil domain containing 4	-2.7
1552552_s_at	<i>CLEC4C</i>	C-type lectin domain family 4, member C	-2.7
232466_at	<i>CUL4A</i>	Cullin 4A	-2.7
214705_at	<i>INADL</i>	InaD-like (Drosophila)	-2.7
210092_at	<i>MAGOH</i>	mago-nashi homolog, proliferation-associated (Drosophila)	-2.7
238931_at	<i>METT10D</i>	methyltransferase 10 domain containing	-2.7
1559413_at	<i>TCP11L2</i>	t-complex 11 (mouse)-like 2	-2.7
241869_at	<i>APOL6</i>	apolipoprotein L, 6	-2.7
207655_s_at	<i>BLNK</i>	B-cell linker	-2.7
228551_at	<i>MGC24039</i>	hypothetical protein MGC24039	-2.7
216850_at	<i>SNRPN</i>	small nuclear ribonucleoprotein polypeptide N	-2.7
232141_at	<i>U2AF1</i>	U2 small nuclear RNA auxiliary factor 1	-2.7
238342_at	---	---	-2.8
235102_x_at	---	Transcribed locus	-2.8
217867_x_at	<i>BACE2</i>	beta-site APP-cleaving enzyme 2	-2.8
217422_s_at	<i>CD22</i> /// <i>MAG</i>	CD22 molecule /// myelin associated glycoprotein	-2.8
230352_at	<i>PRPS2</i>	Phosphoribosyl pyrophosphate synthetase 2	-2.8
220338_at	<i>RALGPS2</i>	Ral GEF with PH domain and SH3 binding motif 2	-2.8
232622_at	---	CDNA FLJ13803 fis, clone THYRO1000187	-2.8

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
235982_at	<i>FCRL1</i>	Fc receptor-like 1	-2.8
1565752_at	<i>FGD2</i>	FYVE, RhoGEF and PH domain containing 2	-2.8
200008_s_at	<i>GDI2</i>	GDP dissociation inhibitor 2	-2.8
228599_at	<i>MS4A1</i>	membrane-spanning 4-domains, subfamily A, member 1	-2.8
205801_s_at	<i>RASGRP3</i>	RAS guanyl releasing protein 3 (calcium and DAG-regulated)	-2.8
208442_s_at	<i>ATM</i> /// <i>LOC651610</i>	ataxia telangiectasia mutated /// similar to Serine-protein kinase ATM (Ataxia telangiectasia mutated) (A-T, mutated)	-2.9
215332_s_at	<i>CD8B</i>	CD8b molecule	-2.9
218640_s_at	<i>PLEKHF2</i>	pleckstrin homology domain containing, family F (with FYVE domain) member 2	-2.9
201258_at	<i>RPS16</i>	ribosomal protein S16	-2.9
215626_at	---	CDNA FLJ11568 fis, clone HEMBA1003278	-2.9
229487_at	---	CDNA FLJ39389 fis, clone PLACE6003621	-2.9
239405_at	---	Transcribed locus	-2.9
202203_s_at	<i>AMFR</i>	autocrine motility factor receptor	-2.9
1558759_s_at	<i>CLASP2</i>	cytoplasmic linker associated protein 2	-2.9
218362_s_at	<i>DIS3</i>	DIS3 mitotic control homolog (S. cerevisiae)	-2.9
1560013_at	<i>PDXDC1</i>	pyridoxal-dependent decarboxylase domain containing 1	-2.9
224503_s_at	<i>ZCCHC2</i>	zinc finger, CCHC domain containing 2	-2.9
204039_at	<i>CEBPA</i>	CCAAT/enhancer binding protein (C/EBP), alpha	-3.0
211991_s_at	<i>HLA-DPA1</i>	major histocompatibility complex, class II, DP alpha 1	-3.0
239975_at	<i>HLA-DPB2</i>	major histocompatibility complex, class II, DP beta 2 (pseudogene)	-3.0
201259_s_at	<i>SYPL1</i>	synaptophysin-like 1	-3.0
224719_s_at	<i>C12orf57</i>	chromosome 12 open reading frame 57	-3.0
229450_at	<i>IFIT3</i>	interferon-induced protein with tetratricopeptide repeats 3	-3.0
	<i>LOC400963</i> ///		
	<i>LOC440589</i> ///		
	<i>LOC441013</i> ///		
217466_x_at	<i>LOC645173</i> ///	ribosomal protein S2 /// hypothetical gene supported by AB082925; BC019021; NM_002952 /// similar to	-3.0
	<i>LOC646294</i> ///	ribosomal protein S2 /// hypothetical LOC645173	
	<i>LOC650901</i> ///		
	<i>LOC729274</i> ///		
	<i>RPS2</i>		
226223_at	---	Transcribed locus	-3.1
204192_at	<i>CD37</i>	CD37 molecule	-3.1
218692_at	<i>GOLSYN</i>	Golgi-localized protein	-3.1
214833_at	<i>TMEM63A</i>	transmembrane protein 63A	-3.1
231979_at	---	CDNA FLJ13266 fis, clone OVARC1000960	-3.1
208269_s_at	<i>ADAM28</i>	ADAM metallopeptidase domain 28	-3.1
	<i>AKAP2</i> ///		
202759_s_at	<i>PALM2-AKAP2</i>	A kinase (PRKA) anchor protein 2 /// PALM2-AKAP2 protein	-3.1
	<i>AKAP2</i>		
242974_at	<i>CD47</i>	CD47 molecule	-3.1

Lebrun et al. Biomarkers and Modifiers of CLN3 Disease

219301_s_at	<i>CNTNAP2</i>	contactin associated protein-like 2	-3.1
211478_s_at	<i>DPP4</i>	dipeptidyl-peptidase 4 (CD26, adenosine deaminase complexing protein 2)	-3.1
210982_s_at	<i>HLA-DRA</i>	major histocompatibility complex, class II, DR alpha	-3.1
216557_x_at	<i>IGHA1</i> /// <i>IGHG1</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-3.1
205267_at	<i>POU2AF1</i>	POU class 2 associating factor 1	-3.1
206759_at	<i>FCER2</i>	Fc fragment of IgE, low affinity II, receptor for (CD23)	-3.2
206206_at	<i>CD180</i>	CD180 molecule	-3.2
235099_at	<i>CMTM8</i>	CKLF-like MARVEL transmembrane domain containing 8	-3.2
228697_at	<i>HINT3</i>	histidine triad nucleotide binding protein 3	-3.2
242234_at	<i>XAF1</i>	XIAP associated factor-1	-3.2
232952_at	---	CDNA FLJ11942 fis, clone HEMBB1000652	-3.3
44790_s_at	<i>C13orf18</i>	chromosome 13 open reading frame 18	-3.3
200962_at	<i>RPL31</i>	ribosomal protein L31	-3.3
244786_at	<i>SNHG10</i>	small nucleolar RNA host gene (non-protein coding) 10	-3.4
217979_at	<i>TSPAN13</i>	Tetraspanin 13	-3.4
227646_at	---	CDNA FLJ39389 fis, clone PLACE6003621	-3.4
208650_s_at	<i>CD24</i>	CD24 molecule	-3.4
226878_at	<i>HLA-DOA</i>	major histocompatibility complex, class II, DO alpha	-3.4
204004_at	<i>PAWR</i>	PRKC, apoptosis, WT1, regulator	-3.4
244413_at	<i>CLECL1</i>	C-type lectin-like 1	-3.5
219179_at	<i>DACT1</i>	dapper, antagonist of beta-catenin, homolog 1 (<i>Xenopus laevis</i>)	-3.5
221916_at	<i>NEFL</i>	neurofilament, light polypeptide 68kDa	-3.5
215694_at	<i>SPATA5L1</i>	spermatogenesis associated 5-like 1	-3.5
220532_s_at	<i>TMEM176B</i>	transmembrane protein 176B	-3.5
209470_s_at	<i>GPM6A</i>	glycoprotein M6A	-3.5
216207_x_at	<i>IGKVID-13</i>	immunoglobulin kappa variable 1D-13	-3.5
221969_at	<i>PAX5</i>	paired box 5	-3.5
228377_at	<i>KLHL14</i>	kelch-like 14 (<i>Drosophila</i>)	-3.6
209583_s_at	<i>CD200</i>	CD200 molecule	-3.7
215859_at	<i>NCLN</i>	nicalin homolog (zebrafish)	-3.7
1552386_at	<i>C5orf29</i>	chromosome 5 open reading frame 29	-3.7
200606_at	<i>DSP</i>	desmoplakin	-3.7
226122_at	<i>PLEKHG1</i>	pleckstrin homology domain containing, family G (with RhoGef domain) member 1	-3.7
209307_at	<i>SWAP70</i>	SWAP-70 protein	-3.7
211654_x_at	<i>HLA-DQB1</i> /// <i>LOC650557</i>	major histocompatibility complex, class II, DQ beta 1 /// similar to HLA class II histocompatibility antigen,	-3.8

		DQ(W1.1) beta chain precursor (DQB1*0501)	
205901_at	<i>PNOC</i>	prepronociceptin	-3.8
235498_at	<i>LRRC44</i>	leucine rich repeat containing 44	-3.9
244172_at	---	---	-3.9
226932_at	---	Transcribed locus	-3.9
244187_at	<i>APOOL</i>	Apolipoprotein O-like	-3.9
203642_s_at	<i>COBL1</i>	COBL-like 1	-3.9
205220_at	<i>GPR109B</i>	G protein-coupled receptor 109B	-3.9
1554343_a_at	<i>STAP1</i>	signal transducing adaptor family member 1	-3.9
1559020_a_at	---	CDNA FLJ14081 fis, clone HEMBB1002280	-4.0
221586_s_at	<i>E2F5</i>	E2F transcription factor 5, p130-binding	-4.0
224559_at	<i>MALAT1</i>	metastasis associated lung adenocarcinoma transcript 1 (non-coding RNA)	-4.0
1569189_at	<i>TTC9C</i>	tetratricopeptide repeat domain 9C	-4.0
234764_x_at	<i>IGL@ /// IGLV1-44</i>	Immunoglobulin lambda variable 1-44 /// Immunoglobulin anti-HBsAg lambda light chain (LM25) /// Immunoglobulin lambda locus	-4.1
1569422_at	<i>FAM129C</i>	family with sequence similarity 129, member C	-4.1
233609_at	<i>PTPRK</i>	Protein tyrosine phosphatase, receptor type, K	-4.3
220606_s_at	<i>C17orf48</i>	chromosome 17 open reading frame 48	-4.4
220068_at	<i>VPREB3</i>	pre-B lymphocyte gene 3	-4.4
209789_at	<i>CORO2B</i>	coronin, actin binding protein, 2B	-4.5
230877_at	<i>IGHD</i>	immunoglobulin heavy constant delta	-4.5
209374_s_at	<i>IGHM</i>	immunoglobulin heavy constant mu	-4.6
215121_x_at	<i>IGL@ /// IGLV2-14 /// IGLV3-25 /// IGLV4-3</i>	immunoglobulin lambda locus /// immunoglobulin lambda variable 4-3 /// immunoglobulin lambda variable 3-25 /// immunoglobulin lambda variable 2-14	-4.8
239292_at	---	Transcribed locus	-4.9
232816_s_at	<i>DDX11 /// DDX12 /// LOC642846</i>	DEAD/H (Asp-Glu-Ala-Asp/His) box polypeptide 11 (CHL1-like helicase homolog, <i>S. cerevisiae</i>) /// DEAD/H (Asp-Glu-Ala-Asp/His) box polypeptide 12 (CHL1-like helicase homolog, <i>S. cerevisiae</i>) /// DEAD/H (Asp-Glu-Ala-Asp/His) box polypeptide 11-like	-4.9
229070_at	<i>C6orf105</i>	chromosome 6 open reading frame 105	-5.0
39318_at	<i>TCL1A</i>	T-cell leukemia/lymphoma 1A	-5.0
205297_s_at	<i>CD79B</i>	CD79b molecule, immunoglobulin-associated beta	-5.1
215379_x_at	<i>IGL@ /// IGLJ3 /// IGLV2-14 /// IGLV3-25</i>	immunoglobulin lambda locus /// immunoglobulin lambda variable 3-25 /// immunoglobulin lambda variable 2-14 /// immunoglobulin lambda joining 3	-5.2
219073_s_at	<i>OSBPL10</i>	oxysterol binding protein-like 10	-5.2
209138_x_at	<i>IGL@</i>	Immunoglobulin lambda locus	-5.3
223565_at	<i>MGC29506</i>	hypothetical protein MGC29506	-5.3
243735_at	---	CDNA FLJ38461 fis, clone FEBRA2020977	-5.4

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
242104_at	---	Transcribed locus	-5.4
227941_at	<i>LOC339803</i>	hypothetical protein LOC339803	-5.4
206641_at	<i>TNFRSF17</i>	tumor necrosis factor receptor superfamily, member 17	-5.7
214669_x_at	<i>IGKC</i>	Immunoglobulin kappa constant	-5.8
214836_x_at	<i>IGKC</i> /// <i>IGKV1-5</i>	immunoglobulin kappa constant /// immunoglobulin kappa variable 1-5	-5.8
215252_at	---	CDNA: FLJ21350 fis, clone COL02751	-6.1
214677_x_at	<i>IGL@</i> /// <i>IGLJ3</i> /// <i>IGLV2-14</i> /// <i>IGLV3-25</i> /// <i>IGLV4-3</i>	immunoglobulin lambda locus /// immunoglobulin lambda variable 4-3 /// immunoglobulin lambda variable 3-25 /// immunoglobulin lambda variable 2-14 /// immunoglobulin lambda joining 3	-6.2
203939_at	<i>NT5E</i>	5'-nucleotidase, ecto (CD73)	-6.2
205861_at	<i>SPIB</i>	Spi-B transcription factor (Spi-1/PU.1 related)	-6.2
1556395_at	---	Full length insert cDNA clone YB44H10	-6.3
206126_at	<i>BLR1</i>	Burkitt lymphoma receptor 1, GTP binding protein (chemokine (C-X-C motif) receptor 5)	-6.3
216401_x_at	---	Immunoglobulin kappa light chain (IGKV gene), cell line JVM-2, clone 1	-6.4
214916_x_at	<i>EXOC7</i> /// <i>IGH@</i> /// <i>IGHA1</i> /// <i>IGHA2</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHG2</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>RAC1</i> /// <i>SIX6</i> /// <i>ZCWPW2</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant alpha 2 (A2m marker) /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// SIX homeobox 6 /// ras-related C3 botulinum toxin substrate 1 (rho family, small GTP binding protein Rac1) /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-6.7
211645_x_at	---	Immunoglobulin kappa light chain (IGKV) mRNA variable region, joining region, and constant region	-6.8
207826_s_at	<i>ID3</i>	inhibitor of DNA binding 3, dominant negative helix-loop-helix protein	-6.8
217148_x_at	<i>IGLV2-14</i>	immunoglobulin lambda variable 2-14	-7.2
208077_at	<i>C9orf38</i>	chromosome 9 open reading frame 38	-7.3
1559139_at	<i>NOC2L</i>	nucleolar complex associated 2 homolog (S. cerevisiae)	-7.5
221651_x_at	<i>IGKC</i> /// <i>IGKV1-5</i> /// <i>IGKV2-24</i>	immunoglobulin kappa constant /// immunoglobulin kappa variable 1-5 /// immunoglobulin kappa variable 2-24	-7.6
210450_at	<i>LOC90925</i>	hypothetical protein LOC90925	-7.7
207245_at	<i>UGT2B17</i>	UDP glucuronosyltransferase 2 family, polypeptide B17	-7.7
235751_s_at	<i>VMO1</i>	vitelline membrane outer layer 1 homolog (chicken)	-7.7
205049_s_at	<i>CD79A</i>	CD79a molecule, immunoglobulin-associated alpha	-7.9
235401_s_at	<i>FCRLA</i>	Fc receptor-like A	-7.9
205379_at	<i>CBR3</i>	carbonyl reductase 3	-8.0

Lebrun et al.		Biomarkers and Modifiers of CLN3 Disease	
230425_at	<i>EPHB1</i>	EPH receptor B1	-9.0
217281_x_at	<i>IL8</i>	Interleukin 8	-9.0
236985_at	---	CDNA FLJ37855 fis, clone BRSSN2014636	-9.4
216576_x_at	<i>NTN2L</i>	Netrin 2-like (chicken)	-10.2
1557910_at	<i>HSP90AB1</i>	heat shock protein 90kDa alpha (cytosolic), class B member 1	-10.7
240405_at	---	Transcribed locus	-10.9
221491_x_at	<i>hCG_1998957</i> /// <i>HLA-DRB1</i> /// <i>HLA-DRB3</i> /// <i>HLA-DRB4</i> /// <i>HLA-DRB5</i> /// <i>LOC730415</i>	major histocompatibility complex, class II, DR beta 1 /// major histocompatibility complex, class II, DR beta 3 /// major histocompatibility complex, class II, DR beta 4 /// major histocompatibility complex, class II, DR beta 5 /// hypothetical protein LOC730415	-11.9
217022_s_at	<i>IGHA1</i> /// <i>IGHA2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant alpha 2 (A2m marker)	-12.3
236203_at	---	---	-12.6
212592_at	<i>IGJ</i>	Immunoglobulin J polypeptide, linker protein for immunoglobulin alpha and mu polypeptides	-13.0
221276_s_at	<i>SYNC1</i>	syncoilin, intermediate filament 1	-13.0
211644_x_at	<i>GJB6</i> /// <i>IGKC</i> /// <i>NTN2L</i>	immunoglobulin kappa constant /// netrin 2-like (chicken) /// gap junction protein, beta 6	-13.9
211430_s_at	<i>IGH@</i> /// <i>IGHG1</i> /// <i>IGHG2</i> /// <i>IGHG3</i> /// <i>IGHM</i> /// <i>IGHV4-31</i>	immunoglobulin heavy locus /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant gamma 2 (G2m marker) /// immunoglobulin heavy constant gamma 3 (G3m marker) /// immunoglobulin heavy constant mu /// immunoglobulin heavy variable 4-31	-15.2
216510_x_at	<i>EXOC7</i> /// <i>IGHA1</i> /// <i>IGHD</i> /// <i>IGHG1</i> /// <i>IGHM</i> /// <i>IGHV4-31</i> /// <i>IL8</i> /// <i>ZCWPW2</i>	immunoglobulin heavy constant alpha 1 /// immunoglobulin heavy constant delta /// immunoglobulin heavy constant gamma 1 (G1m marker) /// immunoglobulin heavy constant mu /// interleukin 8 /// exocyst complex component 7 /// immunoglobulin heavy variable 4-31 /// zinc finger, CW type with PWWP domain 2	-15.7
214777_at	---	Immunoglobulin light chain variable region complementarity determining region (CDR3) mRNA	-18.1
207324_s_at	<i>DSC1</i>	desmocollin 1	-22.6
1558186_s_at	<i>CLLU1</i>	chronic lymphocytic leukemia up-regulated 1	-23.0
227952_at	<i>ZNF718</i>	Zinc finger protein 718	-26.9
209480_at	<i>HLA-DQB1</i>	major histocompatibility complex, class II, DQ beta 1	-87.4
213831_at	<i>HLA-DQA1</i>	major histocompatibility complex, class II, DQ alpha 1	-115.4

* dysregulation shown as fold-change compared to healthy controls