On-line Table 1: Number of patients in groups by diagnosis and pathophysiology*

Diagnosis†	Mean Age (year)	Male	Female	Diagnosis Total	Group Total	Lac
Group 1	4				7	
PMLD		2	0	2		0
PMD		3	0	3		0
CMD		1	0	1		0
Hyccin		0	1	1		0
Group 2	6,8				12	
LVWM		3	1	4		3
ME		1	5	6		6
MLC		0	2	2		2
Group 3	6,2				25	
AD		2	2	4		4
ALD		8	0	8		5
MLD		2	5	7		7
GCL		2	4	6		5
CD‡	9,3	1	1	2	3	0
L-2-OH-GA§		0	1	1		0
UL						
HYPO	6,3	8	7		15	0
Rarefaction	4,9	1	2		3	1
DEMY	9,2	3	2		5	1

Note:—HYPO indicates hypomyelination; DEMY, demyelination; PMD, Pelizaeus-Merzbacher disease; PMLD, Pelizaeus-Merzbacher-like disease; CMD, congenital muscular dystrophy; Hyccin, hyccin deficiency; ME, mitochondrial encephalopathy; AD, Alexander disease; ALD, adrenoleukodystrophy; GCL, globoid cell leukodystrophy; MLD, metachromatic leukodystrophy; CD, Canavan disease; L-2-OH-GA, L-2-OH glutaric aciduria; LVMM, leukoencephalopathy with vanishing white matter; MLC, megalencephalopathy with subcortical cysts; UL, undefined leukoencephalopathy.

* Demographics of patients with defined hereditary leukoencephalopathies and UL: mean age, sex, and total by diagnosis and by pathophysiology group. Numbers of cases with detectable lactate (Lac) signal, for each diagnosis and pathophysiology group, are indicated. For UL patients, tentative pathophysiology group was based on clinical examination, MR imaging, and other diagnostic tests and long-term follow-up (bottom 3 rows).

† Diagnosis and pathophysiology groups: 1 HYPO; 2, rarefaction; 3, DEMY.

‡ Two cases of organic aciduria.

§ One case of organic aciduria.

On-line Table 2: Number of patients in groups by ^{1}H-MRSI using linear discriminant analysis

	1, Hypo	2, Rarefaction	3, DEMY
PMLD	1	1	
PMD	2	1	
CMD	1		
Hyccin	1		
LVWM	2	2	
ME		5	1
MLC		2	
AD			4
ALD		2	6
MLD			7
GCL	1	2	3
CD			2
UL			
HYPO	11	4	0
Rarefaction	1	2	
DEMY	2	2	1

Note:—HYPO indicates hypomyelination; DEMY, demyelination; PMD, Pelizaeus-Merzbacher disease; PMLD, Pelizaeus-Merzbacher-like disease; CMD, congenital muscular dystrophy; Hyccin, hyccin deficiency; ME, mitochondrial encephalopathy; AD, Alexander disease; ALD, adrenoleukodystrophy; GCL, globoid cell leukodystrophy; MLD, metachromatic leukodystrophy; CD, Canavan disease; LVMM, leukoencephalopathy with vanishing white matter; MLC, megalencephalopathy with subcortical cysts; UL, undefined leukoencephalopathy.

On-line Table 3: Summary of significant *P* values for post hoc tests in analysis of variance by disease*

	Cho/NAA	NAA/Cr			
Diagnosis	MLD	PMLD	PMD	LVWM	CD
PMD	.007				
PMLD	.039				
LVWM	.005				
ME	.011		.030		.000
MLC			.025		.000
AD		.019	.001	.006	.000
ALD			.002	.024	.000
GCL			.037		.000
MLD		.004	.001	.001	.000
CD	.013	.018	.035	.001	

Note:—PMD indicates Pelizaeus-Merzbacher disease; PMLD, Pelizaeus-Merzbacher-like disease; CMD, congenital muscular dystrophy; Hyccin, hyccin deficiency; ME, mitochondrial encephalopathy; AD, Alexander disease; ALD, adrenoleukodystrophy; GCL, globoid cell leukodystrophy; MLD, metachromatic leukodystrophy; CD, Canavan disease; LVMM, leukoencephalopathy with vanishing white matter; MLC, megalencephalopathy; Cho, choline; NAA, *N*-acetyaspartate; Cr, creatine.

* Cho/NAA appears useful in differentiating MLD from PMD, PMLD, LVWM, ME, and CD. NAA/Cr appears useful primarily in differentiating PMD and CD from ME, MLC, AD, ALD, GCL, and MLD, in addition to differentiating PMLD and LVWM from a few of the other disorders.