

Title page

Title: Simultaneous quantification of alpha-aminoadipic semialdehyde, piperidine-6-carboxylate, pipercolic acid and alpha-aminoadipic acid in pyridoxine-dependent epilepsy

Running head: Simultaneous quantification of PDE biomarkers

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Table S1. The cone and collision energy for the detection of α -AASA, P6C, α -AAA, d3- α -AAA, PA and d9-PA.

Name	Q1(m/z)	Q3(m/z)	Cone/V	CE/V	RT/min
α-AASA	202.2	81.9	25	25	1.86
P6C	184.1	81.9	35	18	2.37
α-AAA	274.3	98.1	30	22	2.41
d3-α-AAA	277.3	101.1	30	22	2.38
d9-PA	195.2	93.1	30	18	1.87
PA	186.2	84.1	35	28	1.85

Table S2. The limit of detection (LOD) and limit of quantification (LOQ) for all analytes.

Metabolites (μmol/L)	urine (LOD)	Plasma (LOD)	DBS (LOD)	Urine (LOQ)	Plasma (LOQ)	DBS (LOQ)
α-AASA	0.063	0.072	0.020	0.265	0.137	0.090
P6C	0.103	0.148	0.046	0.394	0.481	0.189
PA	0.026	0.037	0.021	0.103	0.128	0.081
α-AAA	0.021	0.032	0.023	0.098	0.116	0.083

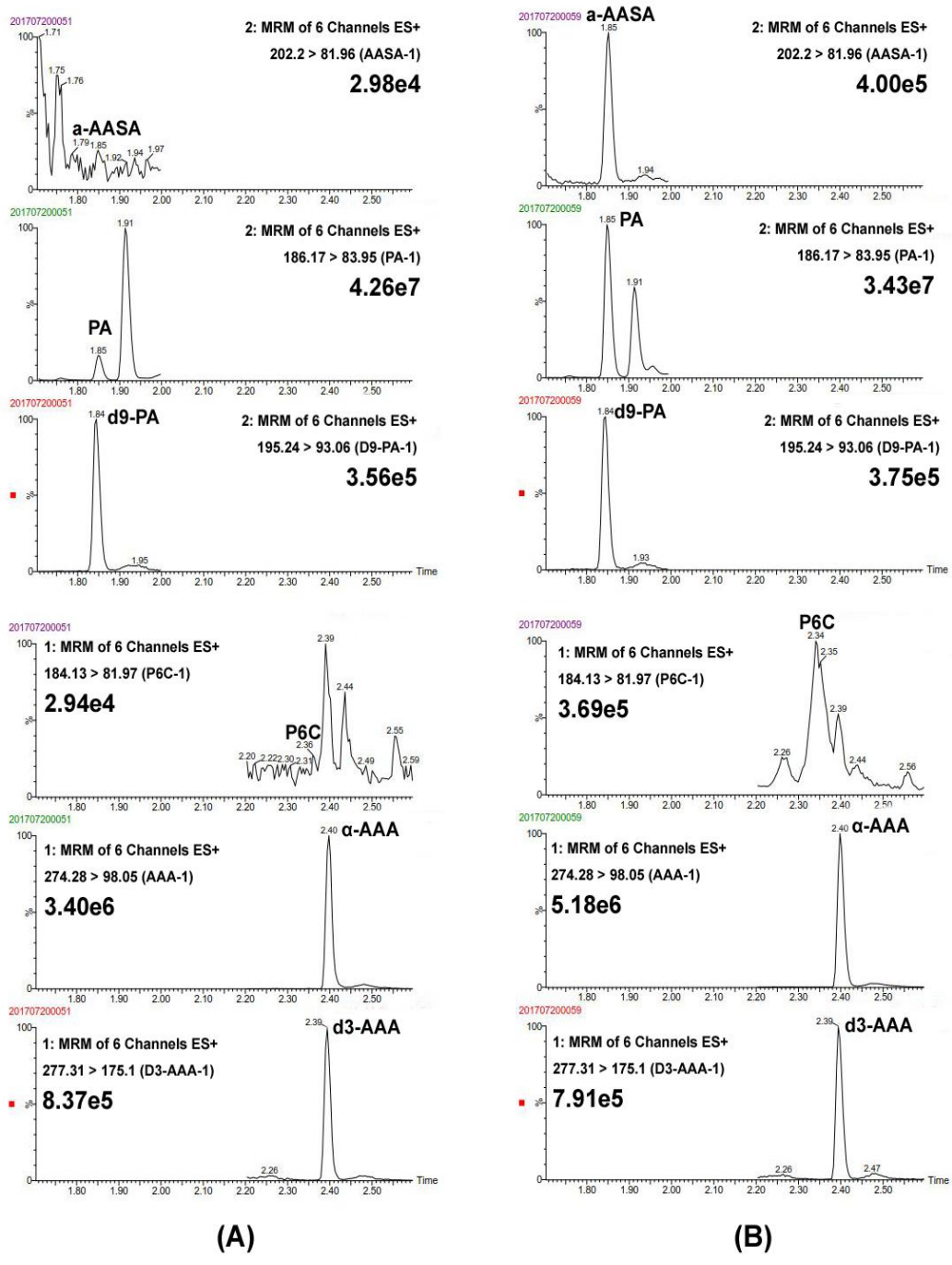


Fig. S1. The classic extract ion chromatogram of a-AASA, P6C, PA, a-AAA, d9-PA and d3-AAA in control and PDE patient in plasma samples.

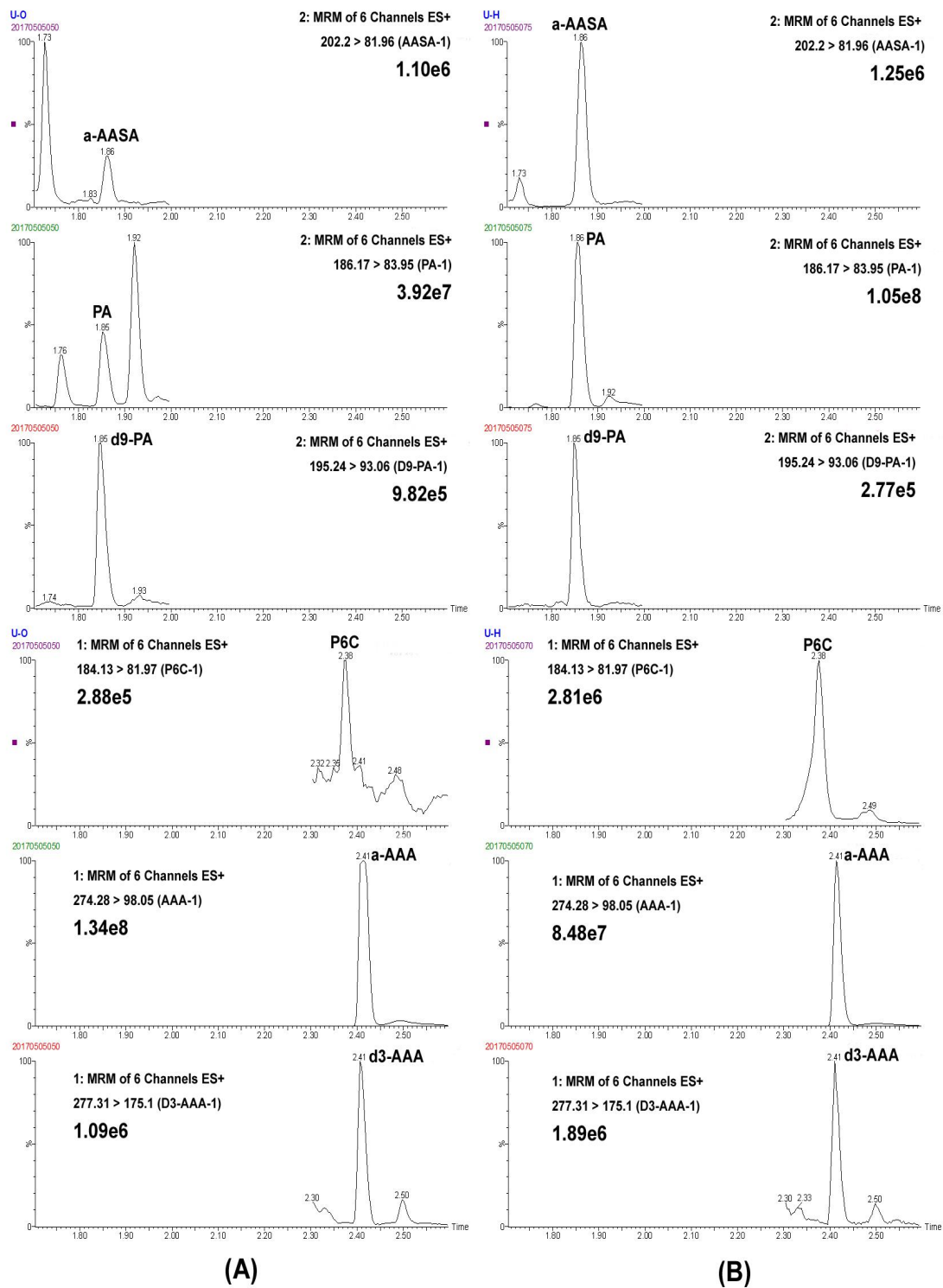


Fig. S2. The classic extract ion chromatogram of a-AASA, P6C, PA, a-AAA, d9-PA and d3-AAA in control and PDE patient in urine samples.

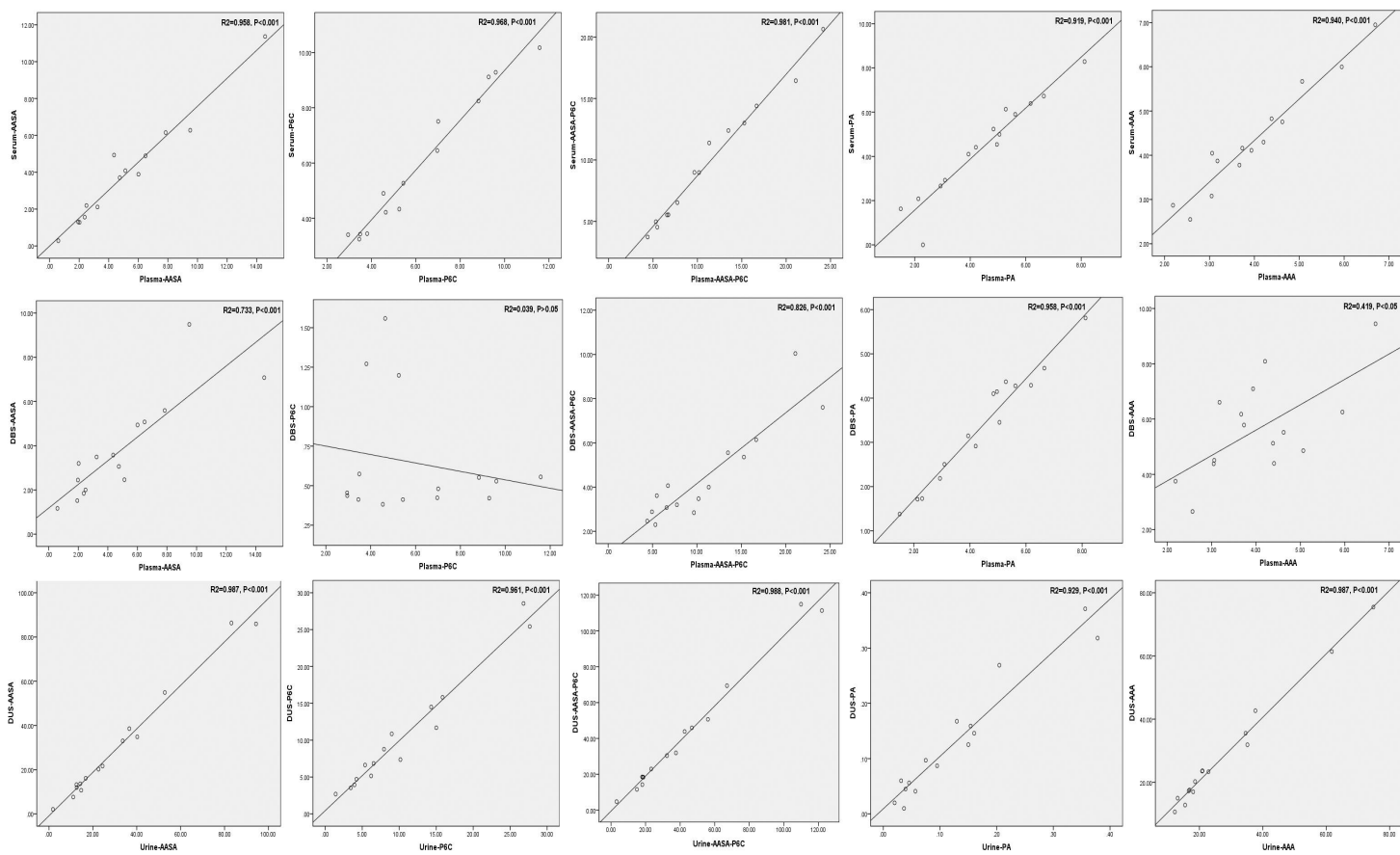


Fig. S3. Correlations of the metabolites concentrations between plasma and serum, plasma and DBS, and urine and DUS.