



## Very Long Chain Fatty Acids, Pristanic and Phytanic Acids in Serum GC-MS Analysis Kit

Peroxisomes are organelles surrounded by membrane in the form of spheres. Peroxisomes play an important role in beta oxidation of very long chain fatty acids (C22:0, C24:0, C26:0) and alpha oxidation of 3-methyl fatty acids (phytanic acid). The Zellweger syndrome is best known peroxisomal disorders. Zellweger syndrome is a rare disease characterized by decreased or absent peroxisomes in the cells of the liver, kidney and brain. Increased levels of very long chain fatty acids (VLCFA) in blood and decreased plasmalogen levels in erythrocytes are the most important parameters to aid in diagnosis. Very long-chain fatty acids, (VLCFA) pristanic and phytanic acid analysis to aid in diagnosis of peroxisomal biogenesis disorders (Zellweger Syndrome Spectrum), X-linked adrenoleukodystrophy (X-ALD). The diagnosis of X-ALD may be raised by the above clinical signs or symptoms, including isolated adrenal insufficiency and is commonly achieved by laboratory evaluation of increased concentrations of VLCFA in plasma. Testing typically includes eight VLCFA parameters: the level of C22:0, C24:0, C26:0, pristanic and phytanic acid and the ratio of C26:0/ C22:0, C24:0/C22:0, pristanic acid/phytanic acid.

### Highlights of the Analysis Kit



Total run time is 14 min.



All analytes, including pristanic and phytanic acids, are analyzed in a single sample preparation



Consuming small volume of patient's sample



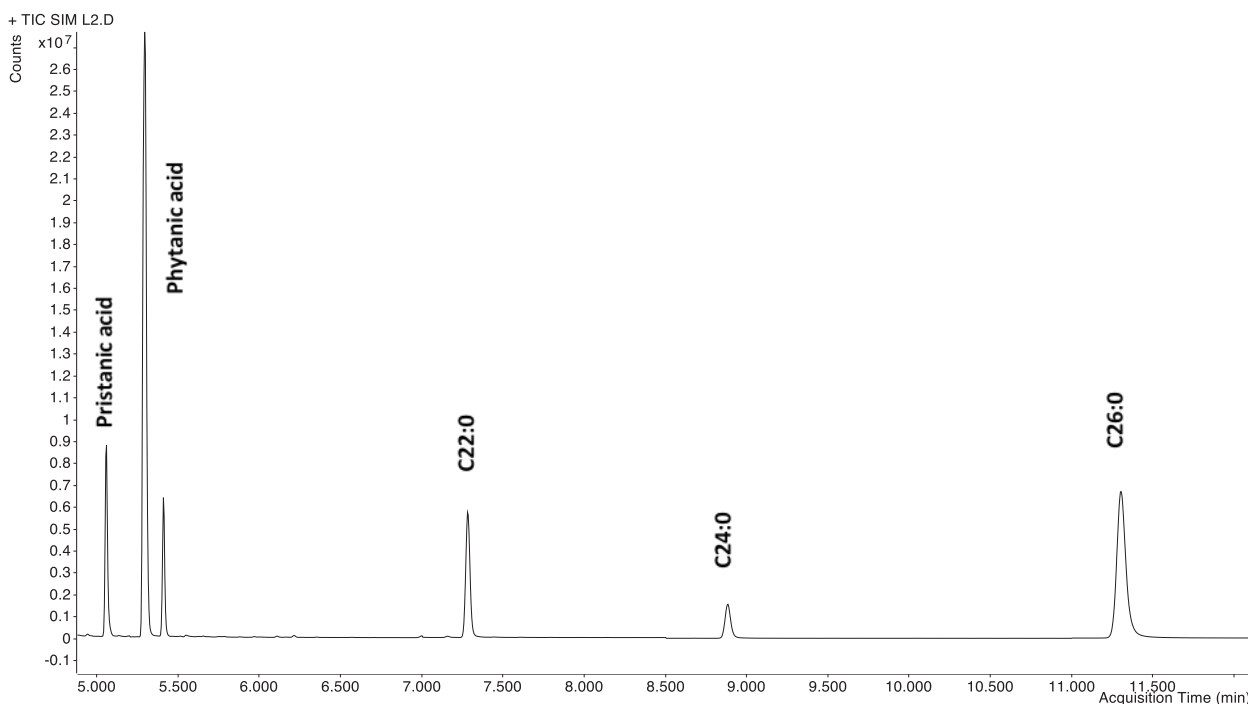
Long life span of GC column

Parameters		
Pristanic acid Phytanic acid	Docosanoic acid (C22:0) Tetracosanoic acid (C24:0)	Hexacosanoic acid (C26:0)
Sample Type		
Serum		

### Sample Preparation

1	Pipette 200 µl serum sample into a glass HPLC vial
2	Reagent mix prepare into the volumetric flask on ice in fume hood. Reagent 3 is added drop-wise while swirling on Reagent 1 and Reagent 2, vortex for 10 sec.
3	600 µl of Reagent-mix, vortex for 15 sec.
4	Locate the vial into the empty glass tube. Incubate in water bath at 90 min 70°C
5	After cooling to room temperature, the methyl derivatives are transferred in eppendorf, extracted 800 µl Reagent 4 and centrifuge at 3500rpm 2 min.
6	Pipette 600 µl extracted serum samples are dried under stream of nitrogen, dissolved 150 µl Reagent 4, and vortex 10 sec.
7	Transfer the final solution into the insert of HPLC vials

### Example Chromatogram



Selected ion chromatogram (SIM) of pristanic, phytanic acid, C22, C24 and C26

## Method Performance

All results were obtained using Agilent 7890B GC 5977B MSD instrument

Analytes	LOQ (nmol/mL)	Linearity (nmol/mL)	Recovery		Repeatability			
					intra-day		inter-day	
			LLQC* (%)	HLQC** (%)	LLQC (%CV)	HLQC (%CV)	LLQC (%CV)	HLQC (%CV)
Pristanic Acid	0.18	0.2 – 6.4	86	100	2.536	1.365	3.62	2.63
Phytanic Acid	1.635	1.0 – 32.0	91	93	4.122	1.919	4.65	2.22
C22:0	3.046	6.0 – 192.0	80	83	0.497	1.087	2.82	2.24
C24:0	2.161	6.0 – 192.0	92	99	2.878	1.406	3.4	3.65
C26:0	0.159	0.25 – 8.0	110	119	4.199	3.031	4.25	4.62

\* LLQC: Low-level quality control

\*\* HLQC: High-level quality control



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