

Organic Acidemias				
Condition	Method	Tier	Biomarker	Value
3-Hydroxy-3-Methylglutaryl-CoA Lyase Deficiency	MS/MS	1st	C5OH	≥ 1.08 μMol/L
	MS/MS	1st	C6DC	≥ 0.10 μMol/L
3-Methylcrotonyl-CoA Carboxylase Deficiency, 3-Methylglutaconyl-CoA Deficiency, and Multiple Carboxylase Deficiency	MS/MS	1st	C5OH	≥ 2.0 μMol/L
	MS/MS	1st	C5DC	≥ 0.60 μMol/L
Glutaric Acidemia (Type 1)	MS/MS	1st	C5	≥ 0.60 μMol/L
	MS/MS	1st	C5/C2	≥ 0.05
Isovaleric Acidemia	MS/MS	1st	C5/C3	≥ 0.50
	MS/MS	1st	C3	≥ 6.92 μMol/L
Methylmalonyl-CoA Mutase Deficiency or Cbl A/B/C/D Deficiency	MS/MS	1st	C3/C2	≥ 0.20
	LC-MS/MS	2nd	MMA	≥ 3.0 μMol/L
Mitochondrial Acetoacetyl-CoA Thiolase Deficiency and 2-Methyl-3-Hydroxybutyryl-CoA Dehydrogenase Deficiency	MS/MS	1st	C5OH	≥ 1.08 μMol/L
	MS/MS	1st	C5:1	≥ 0.19 μMol/L
Propionyl-CoA Carboxylase Deficiency	MS/MS	1st	C3	≥ 6.92 μMol/L
	MS/MS	1st	C3/C2	≥ 0.20
	LC-MS/MS	2nd	MCA	≥ 1.0 μMol/L