Newborn Screening Follow-up for Isolated C5 Acylcarnitine Elevations
(also applies to any plasma or serum C5 acylcarnitine elevations)*

- Isolated elevation of C5 acylcarnitine

**Perform:**
- OAU / Organic Acids Screen, Urine
- ACYLG / Acylglycines, Quantitative, Urine
- AND 1 of the following:
- ACRN / Acylcarnitines, Quantitative, Plasma
- ACRNS / Acylcarnitines, Quantitative, Serum

- Plasma or serum C5–High
- Urine isovaleryl/glycine–High
- Urine 3-methylbutyryl/glycine–Normal

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**Call for consultation to determine the necessity for molecular testing:**
- IVDA / Isovaleryl-CoA Dehydrogenase (IVD) Mutation Analysis (A282V)
- Isovaleryl-CoA Dehydrogenase (IVD) Sequencing

**Did patient receive antibiotic treatment (ie, ampicillin)?**

**NO**

- Isovalerylcarnitine–High
- 3-Methylbutyryl/glycine–Normal

**STOP**

**Diagnostic for isovaleryl-CoA dehydrogenase deficiency**

**YES**

- Isovalerylcarnitine–Normal
- 3-Methylbutyryl/glycine–Normal

- Isovalerylcarnitine–Normal
- 3-Methylbutyryl/glycine–High

**STOP**

**Diagnostic for short/branched chain acyl CoA dehydrogenase (SBCAD) deficiency**

**Consider confirmatory testing for SBCAD gene**

**Routine labs performed locally: Glucose, electrolytes, blood gas, ammonia**

**Perform:**
- FAO / Fatty Acid Oxidation Probe Assay, Fibroblast Culture
- FIBR / Fibroblast Culture

**STOP**

**Likely pivalic acid (antibiotic) artifact**

* Interpretive report provided for all tests in this algorithm