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
- ACRN / Acylcarnitines, Quantitative, Plasma
- ACRNS / Acylcarnitines, Quantitative, Serum

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

Plasma or serum C5–High
- Urine isovalerylglucose–High
- Urine 3-methylbutyrylglycine–Normal

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Did patient receive antibiotic treatment (ie, ampicillin)?

NO

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

STOP

YES

Likely pivalic acid (antibiotic) artifact

Diagnostic for isovaleryl-CoA dehydrogenase deficiency

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

STOP

Diagnostic for short/branched chain acyl CoA dehydrogenase (SBCAD) deficiency
Consider confirmatory testing for SBCAD gene

* Interpretive report provided for all tests in this algorithm