**ELEVATED C5-OH**

**Abbreviations/Key**
- AC = acylcarnitine
- BKT = beta-ketothiolase
- HMG-CoA = 3-hydroxy-3-methylglutaryl-CoA
- IEM = inborn error of metabolism
- MCC = methylcrotonyl-CoA carboxylase
- MCD = multiple carboxylase deficiency
- OA = organic acid
- NBS = Newborn Screening

Action steps are shown in gold (shaded) boxes; results are in plain boxes. Dash lines indicate optional steps.

* = Maternal MCC and holocarboxylase deficiency have been reported as having been identified in newborn screening.
‡ = When the positive predictive value of screening is sufficiently high and the risk to the newborn is high, some initiate diagnostic testing.

**Optional confirmatory testing:**
- DNA analysis and/or enzyme assay

---

**DISCLAIMER:** These algorithms and guidelines were adapted from the American College of Medical Genetics algorithm sheets. They are designed primarily as an educational resource for physicians to help them provide quality medical services. Adherence to these standards and guidelines does not necessarily ensure a successful medical outcome. These standards and guidelines should not be considered inclusive of all proper procedures and tests or exclusive of other procedures and tests that are reasonable directed to obtaining the same results. In determining the propriety of any specific procedure or test, the healthcare provider should apply his or her own professional judgment to the specific clinical circumstances presented by the individual patient or specimen. It may be prudent, however, to document in the patient’s record the rationale for any significant deviation from these standards and guidelines.