C3 Elevated (Isolated)

Assay:
Urine OA
Plasma AC
Plasma Homocysteine

‡Routine labs:
Glucose, electrolytes, blood gas, ammonia, CBC

- Elevated C3 (isolated)

Plasma C3 – Normal
Urine OA – Normal
Plasma Homocysteine - Normal

Plasma C3 – High
Urine OA – MMA
Plasma Homocysteine - Normal

Plasma C3 – High
Urine OA – MMA
Plasma Homocysteine - Normal

Plasma C3 – High
Urine OA – Propionic acid
Plasma Homocysteine - Normal

Plasma C3 – High
Urine OA – Normal
Plasma Homocysteine - Normal

Plasma C3 – High
Urine OA – Propionic acid
Plasma Homocysteine - Normal

False positive
Consider maternal vitamin B12 deficiency
CblC, CblD, CblF, TC-II, or vitamin B12 deficiency
Methylmalonyl-CoA mutase (Mut°, Mut¹), CblA or CblB deficiency
Propionyl-CoA carboxylase deficiency (Propionic acidemia; PA)

Optional Confirmatory Testing:
Propionyl-CoA carboxylase assay (fibroblasts)

Optional Confirmatory Testing:
SUCLA2 sequencing

Optional Confirmatory Testing:
Mut assay/Cbl Complement studies (fibroblasts)

Optional Confirmatory Testing:
Cbl Complement studies (fibroblasts)

Optional Confirmatory Testing:
SUCLA2 sequencing

Optional Confirmatory Testing:
Cbl Complement studies (fibroblasts)

Optional Confirmatory Testing:
Cbl Complement studies (fibroblasts)

Abbreviations/Key:
AC = acylcarnitine
CBC = Complete blood count
Cbl = cobalamin
MMA = methylmalonic acidemia
Mut = mutase
OA = organic acid
TC-II = transcobalamin II

‡ - When the positive predictive value of screening is sufficiently high and the risk to the infant is high, some initiate diagnostic studies that are locally available at the same time as confirmation of the screening result is done.

Actions are shown in shaded boxes; results are in the unshaded boxes

Disclaimer: This guideline is designed primarily as an educational resource for clinicians to help them provide quality medical care. It should not be considered inclusive of all proper procedures and tests or exclusive of other procedures and tests that are reasonably directed to obtaining the same results. Adherence to this guideline does not necessarily ensure a successful medical outcome. In determining the propriety of any specific procedure or test, the clinician should apply his or her own professional judgment to the specific clinical circumstances presented by the individual patient or specimen. Clinicians are encouraged to document the reasons for the use of a particular procedure or test, whether or not it is in conformance with this guideline. Clinicians also are advised to take notice of the date this guideline was adopted, and to consider other medical and scientific information that become available after that date.

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