



# C3 Elevated (Isolated)

Elevated C3  
(isolated)

‡Routine labs:

Glucose, electrolytes, blood gas, ammonia, CBC

Assay:  
Urine OA  
Plasma AC  
Plasma Homocysteine

Plasma C3 and C4DC – High  
Urine OA – MMA  
Plasma Homocysteine - Normal

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

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

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

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

Succinyl-CoA synthetase  
(SUCLA2) deficiency

Methylmalonyl-CoA mutase  
(Mut<sup>-</sup>, Mut<sup>o</sup>), CblA or CblB  
deficiency

CblC, CblD, CblF, TC-II, or  
vitamin B12 deficiency

Propionyl CoA carboxylase  
deficiency  
(Propionic acidemia; PA)

False positive  
Consider maternal vitamin B12  
deficiency

Optional Confirmatory  
Testing:  
SUCLA2 sequencing

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

Optional Confirmatory  
Testing:  
Cbl Complement studies  
(fibroblasts)

Optional Confirmatory  
Testing:  
Propionyl-CoA carboxylase  
assay  
(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.