

## 3-Methylcronyl CoA Carboxylase (3-MCC) Deficiency Ages 0-4 years

### Clinic Visits:

- At least every 6 months

### Laboratory studies:

- At diagnosis:
  - Acylcarnitine profile
  - Urine organic acids
  - Maternal Acylcarnitine profile or urine organic acids
  - Carboxylase activity (possible skin biopsy)
  - Carnitine levels
  - Possible biotinidase activity level
- Clinic visits:
  - Carnitine levels

### During illness:

- Call Metabolic Clinic to alert them if your child is sick
  - Poor feeding, vomiting, and lethargy
- Give your child glucose gel or polymer solution if needed (or instructed to do so)
- If your child is not able to keep down glucose containing fluids, you will need to take him or her to the emergency room for IV 10% dextrose (glucose)
- Be sure to bring Emergency Room letter with you to the hospital
- Labs during illness include: comprehensive metabolic panel and ammonia
- Consider medic alert bracelet

### Evaluations:

- Yearly developmental questionnaires to be completed by parents
- Developmental evaluation at 3 years old

### Topics:

- Biochemistry and genetics of 3-Methylcronyl CoA Carboxylase Deficiency
- Diet/management
  - Avoid fasting
  - Management during illness
  - Carnitine supplementation

*Please make sure your child has clinic visits and levels done on time. Call the metabolic clinic at \_\_\_\_\_ to make an appointment, schedule a blood draw, or with any other questions. If you urgently need to reach a Metabolic physician, call \_\_\_\_\_ and ask to page the Metabolic physician on call.*

Thank You,  
Your Metabolic Team