

## EFFECTS OF UCD ON THE BRAIN

# Intellectual, Adaptive and Behavioral Functioning in Children with UCD

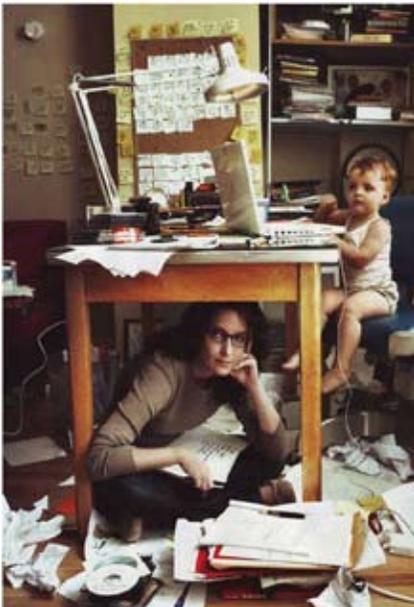
UCD can lead to an accumulation of ammonia in the blood and brain that may result in neurodevelopmental disabilities. This study analyzed data from the Longitudinal Study of UCD to attempt to describe the intellectual, adaptive, and emotional/behavioral functioning of children with UCD. Intellectual functioning refers to the ability to learn, think, solve problems and make sense of the world. Adaptive behavior includes the age-appropriate behaviors necessary for people to live independently and to function safely and appropriately in daily life. Adaptive behaviors include real life skills such as grooming, dressing, safety, safe food handling, school rules, ability to work, money management, clean-

ing, making friends, social skills, and personal responsibility expected based on age. Measures of emotional/behavioral functioning assess one's ability to learn, build and maintain interpersonal relationships, and regulate behavior, feelings, mood, and fears.

These domains were measured through testing and parent questionnaires in 92 children with UCD (33 neonatal onset, 59 late onset). Approximately 50% of children with neonatal onset UCD performed in the range of intellectual disability (ID), including about 30% who were severely impaired. In comparison, only 25% of the late onset group were in the range of ID. There is also evidence that children with UCD

Outcomes for UCD vary widely and are related to many factors including severity of disorder, age of onset, day-to-day metabolic stability, response to treatment, and frequency and severity of hyperammonemic episodes.

have difficulties with some emotional/behavioral and executive function skills (difficulties with behavior regulation, organization, and goal directed behaviors). In conclusion, the study reveals that children with UCD present with a wide spectrum of intellectual and behavioral outcomes. Children with neonatal onset UCDs (presenting in the first 4 weeks of life) have a much higher likelihood of having an intellectual disability, which becomes more evident with increasing age. However, even children with late onset UCDs demonstrate evidence of a wide spectrum of neurocognitive and behavioral impairment, particularly in aspects of attention and executive functioning (intellectual processes). ■



## EXECUTIVE FUNCTION ISSUES

### Metacognitive Problems

- Getting started/initiation
- Organizing Materials
- Working Memory
- Planning approach to tasks
- Difficulty monitoring ones work
- Problems completing tasks

### Behavioral Regulation Problems

- Inflexibility
- Impulsivity
- Emotional Control
- Difficulty monitoring ones own behavior

Children with UCD often experience difficulty with attention and focusing on tasks. Sometimes they do not perform well on neuropsychological tests because of these characteristics. Upcoming research will include studying each disorder separately to determine if there are differences in intellectual, adaptive and behavioral functioning depending on disorder. Adaptive testing methods may be necessary to adequately capture this information.

## References

Krivitzky, LS, Babkian T, Lee HS, Thomas NH, Burke Paull, KL, Batshaw ML. Intellectual, Adaptive and Behavioral Functioning in Children with UCD. *Pediatr Res.* 2009 July; 66(1):96-101