

**EMERGENCY PROTOCOL
UREA CYCLE DISORDERS
ARGINASE DEFICIENCY**

INTRODUCTION

This protocol is for patients with Arginase deficiency presenting to the emergency department with illness or hyperammonemia. Arginase is one of the five enzymes of the urea cycle associated with a known clinical disorder (collectively known as the Urea Cycle disorders (UCDs)).

Though hyperammonemic crises is less common in arginase deficiency than the other urea cycle defects it does occur in patients ranging from infancy to adulthood and may be secondary to an intercurrent illness, possibly subclinical in presentation. Incipient or frank hyperammonemia may present with anorexia, nausea or vomiting. Lethargy often suggests progressive accumulation of ammonia. Arginase deficiency has also been reported in the newborn and can be devastating despite a less severe degree of hyperammonemia than seen in other neonatal urea cycle defects.