

## The aHUS Eculizumab dosing regimen

The aHUS dosing regimen for adult patients ( $\geq 18$  years of age) consists of a 4 week initial phase followed by a maintenance phase:

- Initial phase: 900 mg of Soliris (Eculizumab) administered via a 25 – 45 minute intravenous infusion every week for the first 4 weeks.
- Maintenance phase: 1,200 mg of Soliris administered via a 25 – 45 minute intravenous infusion for the fifth week, followed by 1,200 mg of Soliris administered via a 25 – 45 minute intravenous infusion every  $14 \pm 2$  days

Pediatric patients in aHUS: Pediatric aHUS patients with body weight  $\geq 40$  kg are treated with the adult dosing recommendations, respectively. In pediatric aHUS patients with body weight below 40 kg, the Soliris dosing regimen consists of:

Patient Body Weight	Initial Phase	Maintenance Phase
30 to < 40 kg	600mg weekly x2	900 mg at week 3; then 900 mg every 2 weeks
20 to <30kg	600mg weekly x2	600 mg at week 3; then 600 mg every 2 weeks
10 to <20kg	600 mg weekly x 1	300 mg at week 2; then 300 mg every 2 weeks
5 to <10kg	300 mg weekly x 1	300 mg at week 2; then 300 mg every 3 weeks

For adult and pediatric aHUS patients supplemental dosing of Soliris is required in the setting of concomitant PE/PI (plasmapheresis or plasma exchange, or fresh frozen plasma infusion):

Type of Plasma Intervention	Most Recent Soliris Dose	Supplemental Soliris Dose With Each PE/PI Intervention	Timing of Supplemental Soliris Dose
Plasmapheresis or plasma exchange	300 mg	300 mg per each plasmapheresis or plasma exchange session	Within 60 minutes after each plasmapheresis or plasma exchange
	$\geq 600$ mg	600 mg per each plasmapheresis or plasma exchange session	
Fresh frozen plasma infusion	$\geq 300$ mg	300 mg per infusion of fresh frozen plasma	60 minutes prior to each infusion of fresh frozen plasma