

Table 1: Results During Management Periods

| Date | Arginine (66-150 mmol/L) | Ammonia (<35 mcmo/L) | Albumin (39-53 g/L) | Urine Ketone (mmol/L) | Weight (percentile) | Total Protein Intake (50% medical protein/50% natural protein) (g/kg/d) | Calorie intake (kcal/kg/d) |
|----------------|--------------------------------|----------------------------|---------------------------|-----------------------------|---|--|--------------------------------------|
| Visit 1 | | | | | | | |
| Jan. 2016 | 69 | <9 | 43 | 8-16 | 28.4 kg (3 rd percentile) | 0.82 | 50 |
| Oct 2016 | 199 | <9 | 42 | 8-16 | 33.3 (10-25 th percentile) | 0.8 | 50 |
| Visit 2 | | | | | | | |
| Aug 2018 | 31 | <9 | 40 | 8-16 | 44.8 kg (10- 25 th percentile) | 0.75 | 32 |
| Nov 2018 | 17 (Oct 2018) | <9 | 36 | 8-16 | 46.4 kg (10-25 th percentile) | 0.75 | 31 |
| Visit 3 | | | | | | | |
| Sept 2019 | 247 | <9 | 41 | 8-16 | 41.7 (<3 rd percentile) | 0.63 | 35 |
| Feb 2020 | 359 - 468 | 85 Retest 34 | 38 | 8-16 | 44.6 (<3 rd percentile) | 0.83 | 38 |
| Mar 2020 | 336 | 14 | 35 | 8-16 | N/A | | |

Table 2: Results During Procedure:

| Date | SMOF Lipids g of fat/kg/d; (mL/hr) | Dextrose 10% mL/hr | Normal Saline I.V Fluids | Blood glucose (mmol/L) | Urine ketones (mmol/L) |
|-----------|--|--------------------------|--------------------------------|------------------------------|------------------------------|
| Jan. 2016 | 4 (25) | 12 | 33 | 4.1 to 10.6 | 4-16 |
| Aug 2016 | 4 (24) | 12 | 54 | 3.9-7.6 | 0.5-8 |
| Nov. 2019 | 3 (26) | 16 | 39 | 4.2-6.4 | N/A |