Metabolic Crisis Intervention Kit : method of administration

1. Trisodium Citrate 1g/ 5 ml
    a. Dilute with NaCl 0.9% if needed
    b. Not to many data available in the literature: one study about administration in the intensive care unit 0.3 mmol/kg/h up to 0.5 mmol/kg/h = 90 to 150 mg/kg/h (sodium-potassium citrate mainly given orally for cystinuria: 1.5-2 mEq/kg/day in 3 doses)
2. L-Arginine HCl 21% = 4.2 g/20 ml
    a. To be diluted: hypertonic (2000 mosm/l)
    b. Preferably administration through a central vena : cave extravasation!
    c. Dilute with NaCl 0.9%
    d. Not to administer as IV bolus
    e. Intermittent infusion: administration time dependent of urea cycle defect: 100-700 mg/kg/day as continuous IV infusion
3. L-Carnitine 1 g/ 5 ml
    a. Slow IV: over 2-3 minutes: diluted or not with NaCl 0.9%
    b. Continuous infusion – diluted or not with NaCl 0.9%
    c. Intermittent infusion - diluted or not with NaCl 0.9%
4. Sodium Benzoate 500 mg/ 5 ml
    a. Loading dose (250 mg/kg): infused over 90 minutes: may be diluted with glucose 5% of glucose 10% to 20 mg/ml, may also be infused undiluted
    b. Maintained as a continuous IV infusion (250 mg/kg/day)
5. Vitamin B1 100 mg/ 1 ml
    a. Slow IV: over 5 minutes
    b. IV infusion: dilute with NaCl 0.9%, glucose 5%, Ringer Lactate
    c. May be added total parenteral nutrition solution
    d. IM
6. Vitamin B2 10 mg/ 1 ml
    a. Slow IV: over 5 minutes
    b. IV infusion: dilute with NaCl 0.9%, glucose 5%
    c. May be added to total parenteral nutrition solution

Referenties:

- Pediatric Injectable drugs, 10th edition (The Teddy Bear Book), 2013
- http://www.uptodate.com/home/lexicomp-and-uptodate-collaborate-launch-integrated-patient-education-hospitals-emrs-and-other
- Package insert: l- Arginine HCl 21%-Braun
- http://www.medicinesforchildren.org.uk/
- https://www.kinderformularium.nl/