National Centre for Inherited Metabolic Disorders

Calculation Booklet:

A guide for Nurses and Healthcare Professionals

Foreword

This booklet was developed initially to assist nurses caring for children with Inherited Metabolic disorders. We would like to acknowledge Caroline O'Connor, Catherine McDonnell and the staff of St Brigid's ward responsible for its initial development. The booklet has been further developed over the last number of years, edited by Eilish O'Connell, and reviewed by Dr Ellen Crushell and numerous members of the wider healthcare professional team in the NCIMD, Temple Street Children's University Hospital. Its aim is to provide information regarding the calculations involved in the management of Children with different Inherited Metabolic Disorders.

Contents

1. USEFUL FORMULAE

1.1. Drug Calculations

What you want ÷ what you have x dilution = Volume to be given (Watt, 2003:43)

All units of measurement used must be the same – i.e. all milligrams

e.g. Flucloxicillin 125mg has been prescribed. The stock ampoules are 250mg and you have diluted it to a final volume of 5mls with water for injection. The calculation will be

125mg ÷ 250 x 5mls = 0.5mg x 5mls = 2.5mls

1.2. Calculation of Glucose Content of Intravenous Dextrose

This formula is used when nursing patients with suspected or confirmed hyperinsulinism to assess the amount of glucose required to maintain the blood glucose level within normal limits.

Formula:

Dextrose % of Infusion x Rate divided by weight divided by 6

See section 3.1

1.3. Blood Sugar Levels

Blood glucose levels are measured in two different units: mg / dl or mmol / l

In USA the most common unit is mg / dl while in Canada, Australia and most of Europe mmol / l is more commonly used.

The correlation between the two units is a factor of 18

mmol / l x 18 = mg / dl

1.4. Surface Area

$$\frac{\sqrt{\text{Wgt (Kg) x Hgt (cm)}}}{3600} = \text{BSA } (m^2)$$

i.e. square root of (weight multiplied by height divided by 3600)

(http://www.al-nasir.com/www/PharmCalc/mob_exec_calc.php?ID=bsa)

1.5. Anion Gap

(Na + K) – (Cl + Bicarbonate)

This formula is useful in aiding the diagnosis and treatment of acidosis.

Normal reference range = 7 – 16mmol/L

1.6. Tubular Reabsorption of phosphate - formula

http://baspath.co.uk/calculations/renal_tubular_reabsorption_of_ph.htm

1.7. Calculation of additives per bag of infusing fluid

Amount of additive i.e. Sodium / Potassium / phosphate / Solvito / Peditrace / Vitlipid divided by number of bags which will infuse in 24 hour period.

Watt, S. (2003) Safe administration of medicines to children: part 1. Paediatric Nursing. 15(4): 40-43.

2. CALORIE CONTENT OF INTRAVENOUS SOLUTIONS

Intravenous Solution	Cals / ml	CHO Content gr/ 100mls	FAT Content gr / 100mls	Amino acids Content gr / 100mls
GLUCOSE				
Dex. 2.5 %	0.1 Cal	2.5gr	0	0
Dex. 5 %	0.2 Cal	5.0gr	0	0
Dex. 10 %	0.4 Cal	10gr	0	0
Dex. 12.5%	0.5 Cal	12.5gr	0	0
Dex. 15 %	0.6 Cal	15gr	0	0
Dex. 20 %	0.8 Cal	20gr	0	0
Dex. 25 %	1.0 Cal	25gr	0	0
Dex. 30 %	1.2 Cal	30gr	0	0
NATURAL PROTEIN				
Vaminolact	0.24 Cal	0	0	6.53g/100mls (5.8 g protein/100mls) MSUD 14 mls = 1 exchange PKU 19 mls = 1 exchange HCU 19 mls = 1 exchange Other conditions 15 mls = 1 exchange of amino acids
Aminoven 25 (Central use only)	0.6 Cal	0	0	15g/100mls MSUD 11 mls = 1 exchange PKU 9 mls = 1 exchange HCU 7 mls = 1 exchange Other conditions 7 mls = 1 exchange
AMINO ACIDS (SYNTHETIC PROTEIN)				
MSUD Amino acids With 10 % Dextrose	0.64 kcals	10gr	0	7.5 g/100mls
MSUD Amino acids With 20 % Dextrose	0.98 kcals	20gr	0	7.5 g/100mls
GA TYPE 1 Amino acids With 10 % Dextrose	0.64 kcals	10gr	0	7.5 g/100mls
GA TYPE 1 Amino acids With 20 % Dextrose	0.98 kcals	20gr	0	7.5 g/100mls
PA / MMA Amino acids With 10 % Dextrose	0.64 kcals	10gr	0	7.5 g/100 mls
PA / MMA Amino acids With 20 % Dextrose	0.98 kcals	20gr	0	7.5 g/100 mls
FAT				
Smoflipid 20 %	2 cal	0	20 gr / 100mls	0
Intralipid 20 %	2 cal	0	20 gr / 100mls	0

3. GLUCOSE INFUSION RATE

Calculate the current glucose requirement (glucose required to keep blood glucose within an acceptable range) of a 2.5kg baby who is receiving Dex. 10% at 5 mls per hr

3.1. Long Formula

- Calculate number of mg of dextrose in each ml of IV Fluid

i.e. 10 % solution =

10 grams / 100mls = 10000mg / 100mls = 100mg / 1ml

Therefore a 10% solution contains 100mg = 1 ml

- Multiply by hourly rate to get mg/hr

100 x 5 = 500

- Divide by Baby's Weight

500 / 2.5 =200

- Divide by 60 to get mg / kg / min

200 / 60 = 3.3 mg/kg/min

3.2. Short Formula

Dextrose % x Rate ÷ by weight ÷ by 6

10 x 5 / 2.5 / 6 = 3.3 mg / kg / min

Remember to also include glucose content of oral or nasogastric feeds.

4. ALTERING DEXTROSE CONTENT OF INTRAVENOUS FLUIDS

e.g. Increase a bag of Dextrose (glucose) 10% to Dextrose 15%

Remember % = gram in 100mls (i.e. 1% = 1g)

Example – convert a 500ml bag of Dextrose 10% to 15%.

STEPS:

What do you have?	10 gram dextrose in 100 mls
What do you want?	15 gram dextrose in 100 mls
Deficit	5 grams dextrose in 100 mls

We use Dextrose 50 % when increasing dextrose content of fluids

i.e. 50 grams dextrose in 100 mls

Use drug calculation formula

What you want ÷ what you have x dilution

i.e. 5 ÷ 50 x 100 = 10mls

Therefore, 10mls of Dextrose 50% is needed to increase the dextrose content of 100mls of Dextrose 10% to 15%

NOTE 1: **This is the amount required to alter 100 mls of dextrose 10%. You need to alter a 500ml bag, therefore, 50 mls of Dextrose 50% is required.**

NOTE 2: **When adding Dextrose 50 % to a bag of fluids – ensure the equivalent volume has been removed first from the bag**. For the example above 50mls is removed from a 500ml bag of dextrose 10% and 50mls of Dextrose 50% is added = 15% dextrose.

5. INTRAVENOUS ELECTROLYTES

- Common electrolytes added to IV solutions: sodium, potassium and phosphate.
- Sodium Chloride & dextrose solutions are available (e.g. 0.45% Saline & Dex 5%).
- A 500ml bag of 0.9% NaCl = 77mmols of sodium.

Sodium Chloride: Occasionally a large amount of sodium may to be administered. If this is required 30%Nacl solution may be added to a bag of dextrose to augment the concentration as requested by the metabolic consultant.

Potassium Chloride: Common electrolyte used when administering IV fluids to metabolic patients.

Potassium Phosphate: Less commonly used but when required care must be taken as this solution contains potassium (see example 13)

5.1. Potassium administration – calculation

500ml bags are always used in TSCUH

Example:

- Fluid requirement / day = 2000mls

How many bags will be required in the day? 2000 ÷ 500 = 4 bags

- 4 bags of fluid will be used in a 24hr period (per day).

Patient requires 2 mmols per kg per day of Potassium

⇨ Weight = 30kg

30kg x 2mmol = 60 mmols /kg/ day

You will use 4 bags / day therefore

⇨ 60mmol ÷ 4 bags = 15 mmols of 15% KCL is added to each 500ml bag

15% KCL solution contains 20mmols / 10 mls

⇨ 15mmol ÷ 20mmol x 10mls = 7.5 mls
⇨ 7.5 mls of 15% KCL is added to each 500ml bag

5.2. Sodium administration - Calculation

Example:

- Fluid requirement / day = 1250mls
- How many bags will be required in the day? 1250 ÷ 500 = 2.5 bags

2.5 bags of fluid will be used in a 24hr period (per day).

Patient requires 3 mmols per kg per day of sodium

⇨ Weight = 20kg

20kg x 3 mmol = 60 mmols /kg/ day

You will use 2.5 bags / day therefore

⇨ 60mmol ÷ 2.5 bags = 24 mmols of 30%NaCl is added to each 500ml bag

30%NaCl solution contains 50 mmols / 10 mls

⇨ 24mmol ÷ 50mmol x 10mls = 4.8 mls
⇨ 4.8 mls of 30%NaCl is added to each bag

5.3. Phosphate administration – Calculation

Example:

- Fluid requirement / day = 1100mls
- How many bags will be required in the day? 1100 ÷ 500 = 2.2 bags

2.2 bags of fluid will be used in a 24hr period (per day).

Patient requires 0.8 mmols per kg per day of phosphate

⇨ Weight = 12 kg

12 kg x 0.8 mmol = 9.6 mmols /kg/ day of phosphate

You will use 2.2 bags / day therefore

⇨ 9.6 mmol ÷ 2.2 bags = 4.4 mmols of phosphate is added to each 500ml bag

Potassium PHOSPHATE solution contains 0.6mmols/1ml of phosphate

4.4 mmols ÷ 0.6 mmols x 1mls = 7.3 mls

⇨ 7.3 mls of Potassium PHOSPHATE is added to each bag

- ❖ REMEMBER this solution also contains potassium

i.e. there is 1mmol of potassium in 1ml of this solution

As 9.6 mmols of Phosphate/day is required:

9.6 ÷ 0.6 x 1ml = 16mls (will be used /day) = 16mmols of potassium

If this child requires, for example, 2mmols/kg/day of potassium

- ⇨ Weight = 12kg

12kg x 2 mmol = 24 mmols /kg/ day

REMEMBER you will use 16mmols from the potassium phosphate solution

- ⇨ 24 – 16 = 8mmols remain which will be obtained using 15%KCl

You will use 2.2 bags / day therefore

- ⇨ 8mmol ÷ 2.2 bags = 2.7 mmols of 15%KCl is added to each 500ml bag

15% KCl contains 20mmols / 10 mls

- ⇨ 2.7mmol ÷ 20mmol x 10mls = 0.9 mls
- ⇨ 1.4 mls of 15%KCL is added to each bag

Note - If the concentration of Dextrose in the 500ml bag needs to be altered => always alter the dextrose concentration before adding electrolytes to the bag.

6. INFUSIONS

6.1. Insulin Infusion

Insulin administration is very rarely required, however, if a patient is receiving higher percentages of Dextrose (e.g. 20%) hyperglycaemia can occur. The metabolic consultant needs to be informed if the blood glucose is > 11mmols/l. If the patient is receiving > 15% dextrose 4-6 hourly blood sugar level should be checked.

Check doses and method of administration pre-delivery.

Make up solution by adding 50 units of Actrapid to 50 mls of normal saline.

This results in 1 unit per ml. Administer via syringe pump. The infusion can be connected by y-connector to the fluids already in progress (if drugs are compatible). Commencement of an insulin infusion must be discussed with the Metabolic Consultant on call.

6.2. Carnitine

Oral and intravenous form.

- Intravenously: administered as a bolus or continuous infusion diluted in Glucose 5% / 10%, NaCl 0.9%.
- TSCUH: diluted in Dextrose 10% as more calories will be delivered from this diluent rather than from the other solutions mentioned above.
- When prescribed orally, it may be mixed with flavoured drinks or water.

6.2.1. Indications

⇨ Repletion of carnitine stores (carnitine facilitates the passage of long chain fats into the Mitochondria).
⇨ Removal of toxic metabolites in disorders of intermediary metabolism (dose is usually increased during episodes of intercurrent illness).

6.2.2. Side Effects

⇨ It may cause nausea and vomiting, abdominal pain, diarrhoea (in large oral / enteral doses) and fishy body odour especially if oral dose is doubled if unwell.

6.2.3. Administration

⇨ Please refer to BNF (2011) for information on rate / duration of infusion.

7. MAINTENANCE FLUIDS AND CALORIES

7.1. Normal fluid requirement

For infants under 10kg

Age	Approx. weight	Fluid (ml/kg)
Premature	1 - 2	150 – 200
0 – 6 months	3 - 8	150
7 – 12 months	6 – 10	120

For children over 10kg, normal requirements can be calculated as follows:

11- 20kg	100ml/kg for 10kg +50ml/kg for next 10kg
20 kg and above	100ml/kg for 1st 10kg +50ml/kg for next 10kg +25ml/kg thereafter
	Up to 2500ml per day maximum

7.2. Maintenance Calories

	Boys			Girls		
Age months	Weight	EAR* (Estimated Average Requirements)		Weight	EAR*	
	Kg	Kcal/day	Kcal/kg/day	Kg	Kcal/day	Kcal/kg/day
1-2	5.0		96-120	4.7		96-120
3-4	6.7		96	6.1		96
5-6	7.7		72-96	7.0		72-96
7-12	9.0		72	8.3		72
Years						
1	9.6	770	80	9.0	720	80
2	12.2	1000	82	11.5	930	81
3	14.4	1170	82	73.9	1080	78
4	16.3	1390	85	16.0	1290	81
5	18.6	1480	80	18.2	1360	75
6	21.0	1560	74	21.0	1480	70
7	23.0	1650	71	23.0	1530	67
8	26.0	1750	67	26.0	1630	63
9	29.0	1840	63	29.0	1720	59
10	31.5	2030	64	32.0	1940	61
11	34.5	2130	62	35.9	2020	56
12	38.0	2250	59	40.0	2100	53
13	43.0	2410	56	46.0	2220	48
14	49.0	2630	54	51.0	2340	46
15	55.5	2820	51	53.0	2390	45
16	60.2	2970	49	55.3	2410	44
17	64.0	3080	48	57.0	2460	43
18	66.2	3160	48	57.2	2460	43

Dietetics (2015) Nutritional Requirements for Children in Health and Disease*. 6th Edition, Great Ormond Street Hospital for Children NHS Foundation Trust, London.

Note: Most known patients with Inherited Metabolic Disorders will have specified calorie requirements. Please refer to individual Diet Flow Sheet

8. LIPIDS

8.1. Indications:

Intralipid / Smoflipid is especially valuable in providing a high energy intake to compensate for increased energy expenditure following trauma, infections, severe burns (Ball et al, 1998).

8.2. Administration:

- Slow intravenous infusion (over 24 hour period)
- As part of an all-in-one mixture
- Combination with a glucose/amino acid solution at a Y-connector

8.3. Recommended dosage

The maximum dosage normally prescribed in the Children's University Hospital for patients with Metabolic Disorders is 3grams / kg body weight but this can vary slightly with different metabolic disorders (discuss with Metabolic Consultant).

8.4. Contraindications

⇨ **CONTRA-INDICATED IN DISORDERS OF FAT METABOLISM, LIVER DAMAGE AND ACUTE SHOCK.**

8.5. Calculations

Smoflipid 200mg/ml solution contains 200mg /ml of lipid – i.e. 20g of Fat per 100 mls solution (20g/100ml = 20% solution)

Alternative - Intralipid 20% i.e. 20g in 100mls

Example:

20 kg child requires 2g / kg / day of lipid – (using Smoflipid 200mg/ml solution)

Prescribed volume = 2g / kg = 20 x 2 = 40g (40000mg)

Use drug calculation formula to calculate volume to be infused:

40000 ÷ 200 x 1 = 200mls

- Infusion over 24 hours = 200mls ÷ 24hrs = 8.3 mls/hr
- Remember to calculate calorie content (i.e. Smoflipid / intralipid = 2cals/ml) - 200mls x 2 cals = 400cals obtained from IV Intralipid.
- Observe Liver Function tests / liver size

- As oral feeds are introduced or increased, the combined fat intake must be calculated.

Reference: Ball, P.A., Booth, I.W., Holden, C.E. (1998) *Paediatric Parenteral Nutrition*. 3rd edition. Fresenius Kabi Ltd: Milton Keyes

9. VAMINOLACT

9.1. Indications:

- Amino acid (Natural Protein) solution for intravenous nutrition.
- Indicated in children when enteral supply of protein is insufficient, undesirable or impossible (Ball et al, 1998).
- Used for children under 10kgs or less than a year old in CUH (Dunne T. (2008) Parenteral Nutrition: Prescribing Guidelines. The Children's University Hospital, Dublin).
- Note: for the purposes of administration of IV natural protein (rarely used). It is the product of choice for children with an Inherited Metabolic Disorder (specifically disorders of protein metabolism) - as agreed by Consultants in the NCIMD.
- Vaminolact is rarely used in the management of patients with Inherited Metabolic Disorders except as 'exchanges' (each exchange equals 1 gram of protein) as it is a complete protein and contains the precursors of the organic acids or toxic amino acids themselves e.g. Leucine

9.2. Calculations

9.2.1. Example 1

3 grams of Natural Protein (3 exchanges) prescribed for patient.

Vaminolact contains 5.8 grams protein per 100 mls.

Use drug calculation formula to calculate volume to be infused.

⇨ Volume required = 3grams ÷ 5.8 g x 100 = 52mls ÷ 24 hours = 2.1 mls in 1 hour

Vaminolact may be "piggybacked" into Dextrose infusion (via buretrol) or may be infused on its own.

9.2.2. Example 2

Calculate 1 gram (1exchange) / kg protein for 5 kg child = 5 grams / day required.

5 ÷ 5.8 x 100 = 86 mls / 24 hours = 3.6 mls/hr

REMEMBER TO CALCULATE CALORIE CONTENT OF VAMINOLACT (See CALORIE CONTENT OF INTRAVENOUS SOLUTIONS information)

Ball, P.A., Booth, I.W., Holden, C.E. (1998) Paediatric Parenteral Nutrition. 3rd edition. Fresenius Kabi Ltd: Milton Keyes

Thomas, B. (2001) Manual of Dietetic Practice. 3rd edition. Blackwell Science: London

10. VITAMINS AND MINERALS

10.1. SOLVITO

- Yellow mixture of water-soluble vitamins.
- Added to glucose solution (after reconstituted) or Intralipid for Intravenous infusion.
- Intended as a supplement in intravenous nutrition in order to cover the daily requirements of the water-soluble vitamins in both adults and infants.

10.1.1. Dose

Child < 10 kg the recommended daily dose = 1ml of reconstituted mixture per kg body weight.

Adult & child > over 10kg the recommended daily dose = 10 mls (Ball et al, 1998).

10.1.2. Administration

The contents of one vial of Solvito are dissolved by the addition of 10 ml of water for injection and added to one of the following:

- Vitlipid
- Smoflipid 200mg/ml solution / Intralipid 10% or 20% solution
- Glucose Intravenous Solution
- Water for Injections

In Temple Street Children's University Hospital, contents are usually reconstituted with 10 mls of sterile water and the appropriate volume added to a dextrose infusion.

Note:

This solution is light sensitive. Therefore, infusions containing Solvito must be protected.

10.1.3. Calculations

10 mls Solvito to be administered into Dextrose 10% infusion. Rate of Infusion is 20mls per hour

20 mls x 24 hours = 480 mls in 24 hours ….. How many bags? (using 500ml bags)

480 ÷ 500 = 0.96 bags of dextrose 10%

10 mls Solvito ÷ 0.96 bags = 10.4 mls of Solvito must be added to each 500ml bag to ensure that the patient receives 10 mls in 1 day.

Dunne T. (2008) Parenteral Nutrition: Prescribing Guidelines. The Children's University Hospital, Dublin.

10.2. PEDITRACE ® & ADDITRACE ®

Peditrace & Additrace may be prescribed in order to meet the basal requirement for trace elements

Contents:

Peditrace contains traces of Zinc, Manganese, Copper, Fluoride, Selenium and Iodine;

Additrace contains Iron, Zinc, Manganese, Copper, Chromium, Selenium, Molybdenum, Fluoride and Iodine).

It is added to solutions of amino acid or Dextrose.

10.2.1. Dose

Peditrace

Infants & children ≤ 15 kg = 1ml / kg up to a maximum daily dose of 15 mls.

For children (15 kg or more) – 15 mls Peditrace per day (Ball et al. 1998).

Additrace

Children & adults > 40Kgs = 0.2mls/kg up to a max of 10mls / day (Dunne T., 2008).

10.2.2. Administration

Peditrace or Additrace must be given by slow intravenous infusion over a minimum period of 8 hours. In Temple Street Children's University Hospital, the daily requirement is infused suspended in a dextrose infusion and administered over 24 hours.

10.2.3. Calculations

Example:

Patient weighs 10kg, therefore, requires 1ml/kg/day = 10 x1 = 10mls/day

10 mls of Peditrace needs to be administered in Dextrose 10% infusion.

Rate of Infusion is 30mls per hour

Daily volume of dextrose = 30 x 24 = 720 mls

How many bags will be required in the day (using 500ml bags)?

i.e. 720 mls ÷ 500 ml bags = 1.44 bags

10 mls Peditrace ÷ 1.44 bags = 6.9 mls of Peditrace must be added to each bag to ensure that the patient receives 10 mls in 1 day.

Dunne T. (2008) Parenteral Nutrition: Prescribing Guidelines. The Children's University Hospital, Dublin

10.3. VITLIPID ®N INFANT / VITLIPID ®N ADULT

White oil in water emulsion containing the fat soluble vitamins, A, D, E and K. This vitamin addition is intended as a supplement to Intralipid 10% or 20% in the intravenous nutrition of infants and children.

10.3.1. Dose

Vitlipid infant Pre-term & low birth weight infants up to 2.5 kg bodyweight = 4mls/kg

Infants & children weighing > 2.5 kg = 10 mls/day (Ball et al, 1998).

Vitlipid adult Children > 11yrs & adults = 10mls /day (Ball et al, 1998).

10.3.2. Administration

In the Temple Street Children's University Hospital, Vitlipid is added to smoflipid / intralipid infusion.

Amount to be given divided by number of syringes / bags of Intralipid administered in 24 hours (see calculation example).

10.3.3. Calculations

a) Patient weighs 10kg. Requires 3g/kg/day of lipids.

Total volume of Lipids to be administered in 24 hours = 3 x 10 = 30g/day

(=30000mg/day) (Using Smoflipid 200mg/ml)

30000g ÷ 200 mg x 1ml = 150 mls

This will be administered using 100ml bottles of smoflipid (i.e. 150ml ÷ 100ml = 1.5 bottles)

b) Vitlipid calculation:
10mls Vitlipid (Child weighing 10kg requires 10ml/day) ÷ 1.5 = 6.6 mls

6.6mls of Vitlipid needs to be added to each smoflipid 100ml bottles.

REMEMBER – if added to a syringe where a child requires smaller amount, the dose / day is administered over 24hours.

i.e. if 10mls is required in 24 hours = 10 ÷ 24 = 0.4mls/hr

e.g. 3.5 kg baby requires 2g/kg of Smoflipid = 3.5 x 2 = 7g / day (7000g / day)

7000 ÷ 200mg x 1ml = 35mls smoflipid

As the baby is 3.5kg they will require 10mls of Vitlipid / day

Therefore: 10mls of Vitlipid infant + 35mls of smoflipid = 45mls –» 45 ÷ 24hours = 1.9mls/hour

Where larger volumes are administered, the vitlipid volume does not need to be included in rate / hour.

Dunne T. (2008) Parenteral Nutrition: Prescribing Guidelines. The Children's University Hospital, Dublin

11. INTRAVENOUS SYNTHETIC AMINO ACIDS (SAA)

Synthetic Amino Acid Solutions are used in acute circumstances when the oral or enteral routes are not adequate to provide the essential amount of synthetic protein. The solutions are provided by a company overseas and there it must be noted that there can be a delay of 48 hours before they can be supplied. The percentage of Dextrose in the solutions that can be infused is determined by the presence of peripheral or central line (i.e. 10% for peripheral lines & 20% for central lines). Synthetic Amino Acid solutions may be administered separately or add on to Dextrose/lipids/Vaminolact.

11.1. SAA for Maple Syrup Urine Disease

SAA Solution = without Leucine, Isoleucine and Valine

Standard Solution (as received from Manufacturer)

One litre contains 75 grams of Synthetic Amino Acids (SAA)

Therefore 100 mls contains 7.5 grams of SAA i.e. 7.5% solution

11.1.1. Example

0.25 g / kg SAA prescribed for 15 kg child.

0.25 g x 15 kgs = 3.75 g Use drug calculation formula

3.75 ÷ 7.5g x100 mls = 50mls to be infused in 24 hours = 2 mls per hour.

11.2. For Glutaric Aciduria Type 1

SAA solution = without Lysine and Tryptophan

Standard Solution (as received from Manufacturer)

One litre contains 75 grams of SAA

Therefore 100 mls contains 7.5 grams of SAA i.e. 7.5% solution

11.2.1. Example 1

0.5 grams / kg SAA prescribed for 10 kg child

0.5g x10 kg = 5 grams/kg/day Use drug calculation formula

5 divided by 7.5g multiplied by 100 mls = 66.6mls to be administered in 24 hours = 2.7mls /hour

12. INTRAVENOUS ISOLEUCINE AND VALINE

Each solution contains 1g/100mls. They are also provided by a company overseas and therefore may take 24hours to be delivered.

There are no readily available written guidelines on the administration of these medications.

Please consult with a Metabolic Consultant regarding dosage, dilution, method and rate of administration.

13. UREA CYCLE DISORDER MEDICATION

Some of the drugs used in the treatment and management of Urea Cycle disorder are Arginine, Sodium Phenylbutyrate and Sodium Benzoate. The following are examples of drug calculations for each of these drugs. An algorithm for the administration of each drug is available in the Medical Management Guidelines for Children with Inherited metabolic disorders.

Always refer to BNF for administration of above drugs.

Note:

1. Ensure dose is charted in mg. A standard solution is made for administration over a 24 hour period. A Doctor may write the volume to be infused in addition to the dose in mgs on the prescription sheet; however, doses should be checked on each administration to ensure accuracy. Follow the example prescription provided in the algorithms. Remember that change in dose requires a new prescription; however, the solution attached to the patients does not as they are standard solutions.

2. Simultaneous infusion of Sodium Benzoate, Arginine & Sodium Phenyl butyrate is possible – as per TSCUH pharmacy instruction "Arginine, Carnitine, Sodium Benzoate and Sodium Phenylbutyrate may be infused together at a "y" site (there is no data to support this but it has been done in practice)".

3. Administration fluid: 5% / 10% Dex. Dextrose 10% is preferable as it delivers more calories.

4. Calorie content = 0.4 cals / ml (using 10% dextrose)

5. Sodium Benzoate contains Sodium 1.4 mmols/ml
   Sodium Phenylbutyrate contains Sodium 1.1 mmols/ml

13.1. Arginine Calculation

Example of prescription.

Stat dose of 250 mg / kg prescribed for 2.5 kg baby.

2.5 kg x 250mg = 625mgs.

Use Drug calculation formula =

625 mg / 10 000mg x 100 (i.e. 10grams in 100ml solution) = 6.25 mls.

Infused over 90 – 120 mins

See arginine algorithm for drug preparation.

Infuse as prescribed: May be prescribed as a stat / bolus infusion (over 1.5-2 hours) or as a continuous infusion over 24 hours.

13.2. Sodium Benzoate Calculation

Example of prescription

Stat dose of 250 mg / kg prescribed for a child who weighs 10 kg.

250mgs x 10 kg = 2500mgs.

Use Drug calculation formula =

2500 mg / 5 000mg x 100 (i.e. 5grams in 100ml solution) = 50 mls.

Infused over 90 – 120 mins

See sodium benzoate algorithm for drug preparation.

Infuse as prescribed: May be prescribed as a stat / bolus infusion (over 1.5-2 hours) or as a continuous infusion over 24 hours.

13.3. Sodium Phenylbutyrate Calculations

Example of prescription

250 mg / kg prescribed for 15kg child

15 kg x 250 = 3750 mgs = 3.75g

Use Drug calculation formula =

3.75 g / 5g x 100 mls = 75 mls ÷ 24hrs = 3.1 mls per hour

See sodium phenylbutyrate algorithm for drug preparation.

A stat dose of 250mgs / Kg can be administered over 90 mins and thereafter Sodium Phenylbutyrate must be infused as a continuous infusion (BNF, 2011).