MSUD Dietetic Management Pathway

Presumptive positive screen for MSUD (symptomatic infant)
Refer to Clinical Management Guidelines and Initial Clinical Referral Guidelines and Standards (www.bimdg.org)

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On the same day
admission to local hospital
transfer to specialist centre, if not possible, organise supplies of MSUD Anamix Infant (Nutricia), Isoleucine 50 and Valine 50 sachets (Vitaflo Int) and feeding plan
Liaise with local dietitian and team

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Symptomatic infant
stop breast feeding (continue expressing) and/or standard formula feeds
commence branch chain amino acid free infant formula - MSUD Anamix Infant
commence isoleucine and valine supplements
consider dialysis/haemofiltration dependent on clinical condition and blood leucine level
promote anabolism: provide >120 kcal/kg/day

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Enteral feeding regimen (not fluid restricted 150-180ml/kg):
Branch chain amino acid free (BCAA free) formula - MSUD Anamix Infant
oral or NG to 3g/kg/day protein equivalent (see Appendix 1 - example recipe)

+ glucose polymer to 10% carbohydrate (aim for glucose delivery rate of 10 - 15mg/kg/min)

+ fat to 5% depending on tolerance

+ 200mg isoleucine and 200mg valine supplements: even if already in target treatment range (see Appendix 2 – plasma target treatment range table). Give separately or add to Anamix if full volume will definitely be given

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Enteral feeding regimen (fluid restricted) or not tolerating above:
IV 20% dextrose + added electrolytes (+/- insulin if hyperglycaemic)

+ IV Intralipid 2g/kg/day

+ concentrated branch chain amino acid free (BCAA free) supplement – MSUD Aid III or MSUD Amino5, via NG or NJ to 3g/kg/day protein equivalent (see Appendix 1 - example recipe

+ isoleucine and valine supplements: 200-300mg of each (400mg if being dialysed), even if already within target treatment range (see Appendix 2 - plasma target treatment range table)

Consider BCAA - freeTPN if not absorbing/tolerating feeds and if available

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Monitoring - BCAA levels daily (see Appendix 2 - plasma target treatment range table)
- re-introduce EBM or standard infant formula (as 50mg leucine exchanges - see Appendix 2)
- adjust isoleucine and valine supplements (see Appendix 2 - plasma target treatment range table)
- once stabilised, in classical MSUD 300mg leucine/day is a typical intake

MSUD Dietetic Management Pathway

Presumptive positive screen for MSUD (asymptomatic infant)
Refer to Clinical Management Guidelines and Initial Clinical Referral Guidelines and Standards (www.bimdg.org)

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On the same day
admission to local hospital
transfer to specialist centre, if not possible, organise supplies of MSUD Anamix Infant (Nutricia), Isoleucine 50 and Valine 50 sachets (Vitaflo) and feeding plan
Liaise with local dietitian and team

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Asymptomatic infant
stop breast feeding (continue expressing) and/or standard formula feeds for a maximum of 24hours depending on leucine levels
commence branch chain amino acid free (BCAA free) infant formula
commence isoleucine and valine supplements
promote anabolism: >100 kcal/kg/day

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Enteral feeding regimen (not fluid restricted, 150-180ml/kg):
Branch chain amino acid free (BCAA free) formula - MSUD Anamix Infant
oral or NG to 3g/kg/day protein equivalent (see Appendix 1 - example recipes)

+ 200mg isoleucine and 200mg valine supplements: even if already in target treatment range (see Appendix 2 – plasma target treatment range table). Give separately or add to Anamix if full volume will definitely be given

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Monitoring - BCAA levels daily (see Appendix 2 - plasma target treatment range table)
- re-introduce standard infant formula, breast feeding (see Appendix 2)
- if intermediate MSUD will have much greater tolerance of leucine intake
- adjust isoleucine and valine supplements

Appendix 1 - Example recipes for a 3kg infant

NB: isoleucine and valine 50 sachets contain CHO. If amount provides excess CHO, consider using isoleucine and valine 1000 sachets (will give less CHO). Sachets can be added to feeds as examples below. If feed volumes and doses are changing daily or more often, then sachets are best given separately as a divided dose 3 to 4times/day) . They can be given dissolved in a small volume of sterile water or feed.

Feeding at 150ml/kg

	Energy (kcal)	Pr Eq (g)	CHO (g)	Fat (g)	Val (mg)	Iso (mg)
68g MSUD Anamix Infant	311	9	33.7	15.6		
4 sachets Valine50	60	0	15.2	0	200	
4 sachets Isoleucine50	60	0	15.2	0		200
Water up to 450ml						
Total (450ml)	431	9	64.1	15.6	200	200
Per 100ml	96	2	14	3		
Per kg	144	3	21	5	67	67

Fluid Restricted: Feeding at 80ml/kg (3.3ml/kg/hr) - 50ml/kg enteral + 30ml/kg IV

	Energy (kcal)	Pr Eq (g)	CHO (g)	Fat (g)	Val (mg)	Iso (mg)
12g MSUD Aid III	39	9	0.5	0		
20ml 8% Valine1000 soln	12	0	2.3	0	200	
20ml 8% Isoleucine1000 soln	12	0	2.3	0		200
11g Glucose polymer	42	0	10.5	0		
Water up to 150ml						
Total (150ml)	105	9	15.6			
Per 100ml	70	6	10.4			
Per kg	35	2	3.5			

+ IV 20% lipids 30ml (10ml/kg): 6g fat (2g/kg), 60kcal (20kcal/kg)
+ IV 20% dextrose 60ml (20ml/kg): 12g CHO, 48kcal (16kcal/kg)

TOTAL: 69kcal/kg, 3g protein equivalent/kg

Fluid Restricted: Feeding at 100ml/kg (4.1ml/kg/hr) - 50ml/kg enteral + 50ml/kg IV

	Energy (kcal)	Pr Eq (g)	CHO (g)	Fat (g)	Val (mg)	Iso (mg)
12g MSUD Aid III	39	9	0.5	0		
20ml 8% Valine1000 soln	12	0	2.3	0	200	
20ml 8% Isoleucine1000 soln	12	0	2.3	0		200
11g Glucose polymer	42	0	10.5	0		
Water up to 150ml						
Total (150ml)	105	9	15.6			
Per 100ml	70	6	10.4			
Per kg	35	2	3.5			

+ IV 20% lipids 30ml (10ml/kg): 6g fat (2g/kg), 60kcal (20kcal/kg)
+ IV 20% dextrose 120ml (40ml/kg): 24g CHO, 96kcal (32kcal/kg)
TOTAL: 87kcal/kg, 3g protein equivalent/kg

Appendix 2 - Biochemical aims of treatment

Plasma Leucine - reduce by >750μmol/l per 24 hours until within target treatment range

Plasma Isoleucine & Valine - maintain in target treatment range
(avoid low levels which are rate limiting for protein synthesis)

Ketones - negative

Plasma Leucine μmol/l	**Guide to re-introducing protein (leucine exchanges)**
> 800	no natural protein/leucine
< 800	re-introduce protein/leucine

	Plasma target treatment range (μmol/l)
Leucine	150-300
Isoleucine	200-400 (400-600 when unwell)
Valine	200-400 (400-600 when unwell)

Volume (ml)	**Leucine exchange**
50ml	Expressed breast milk (EBM)
35ml	Standard infant formula

Appendix 3 MSUD infant formula & Amino acid supplements

MSUD Anamix Infant (Nutricia) – BCAA free infant formula

Standard 15% dilution: *30ml water + 5g (1 level yellow scoop) powder*

	Per 100g	Per 100ml
Energy (kcal)	457	69
Protein equivalent (g)	13.1	2
CHO (g)	49.5	7.5
Fat (g)	23	3.5

Nutricia scoops

1 yellow scoop = 5g powder

1 large blue scoop = 26g powder

MSUD Aid III (Nutricia) – BCAA free amino acid supplement

Suggested dilution 8%: *8.1g (1 level yellow scoop and 1 small blue scoop) + 100ml water*

	Per 100g	Per 100ml
Energy (kcal)	326	26.4
Protein equivalent (g)	77	6.24
CHO (g)	4.5	0.36
Fat (g)	0	0

Nutricia scoops

1 small blue scoop = 1.8g powder

1 yellow scoop = 6.3g powder

1 large blue scoop = 37.2g powder

MSUD amino5 (Vitaflo)- BCAA free amino acid supplement

	Per 100g	Per 6g sachet
Energy (kcal)	332	20
Protein equivalent (g)	83	5
CHO (g)	0	0
Fat (g)	0	0

Valine (Vitaflo)

	Valine50	Valine1000	
	Per 4g sachet	Per 4g sachet	10ml of 8% soln (4g sachet and 50ml water)
Energy (kcal)	15	15	3
L-Valine (mg)	50	1000	200
CHO (g)	3.8	2.9	0.6

Isoleucine (Vitaflo)

	Isoleucine50	Isoleucine1000	
	Per 4g sachet	Per 4g sachet	10ml of 8% soln (4g sachet and 50ml water)
Energy (kcal)	15	15	3
L-Isoleucine (mg)	50	1000	200
CHO (g)	3.8	2.9	0.6

IV Dextrose

% Dextrose	Energy (kcal)/ml
10%	0.34
12.5%	0.43
15%	0.51
20%	0.68

IV Lipids

	kcal/ml	fat g/ml
Intralipid 10%	1.1	0.1
Intralipid 20%	2	0.2
Intralipid 30%	3	0.3
Structolipid 20%	1.96	0.2
Smoflipid 20%	2	0.2

Feed company contact details

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