

Organic
Acidaemias
Questionnaire
for SSIEM 2014

We would like to try and find out more about feeding practices in children and adults with organic acidaemias throughout Europe and neighbouring countries. We are specifically looking at children with propionic, methylmalonic and isovaleric acidaemia.

We would like to present these findings at the SSIEM meeting in Innsbruck. If we have enough information, we would like to consider publication of this data. Anyone who contributes data will be included as an author. We would be very grateful if you would take time to complete the following questions by **2nd August, 2014**.

Please do not hesitate to contact us if you do need any further explanation. Thank you and Best wishes

Name and country of metabolic centre

- Name of Dietitian to be included in the author list (this has to be the person who has collated the data for each centre or played a major role in writing the paper). We have to explain the role of each author to the journal
- Name of other dietitians to be acknowledged at the end of the paper

How many patients are on a prescribed protein restriction with organic acidaemias?

Condition	Total Numbers of patients	Aged < 1y	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia	Neonatal diagnosis				
	Late diagnosis				
Methylmalonic acidaemia – Vitamin B12 responsive	Neonatal diagnosis				
	Late diagnosis				
Methylmalonic acidaemia – Vitamin B12 non- responsive	Neonatal diagnosis				
	Late diagnosis				
Isovaleric acidaemia	Neonatal diagnosis				
	Late diagnosis				

Please only include patients who have been prescribed protein restriction (even if compliance is poor).

Please indicate how many patients are diagnosed early (i.e. less than 30 days of age) and how many post 30 days (late diagnosis). If age of diagnosis is unknown please indicate this.

Please calculate the total protein (g/kg/day) you have prescribed for your patients on protein restriction with organic acidaemias for each of the age bands

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia					
Methylmalonic acidaemia – Vitamin B12 responsive					
Methylmalonic acidaemia – Vitamin B12 non-responsive					
Isovaleric acidaemia					

Total protein includes natural protein and protein equivalent from precursor-free L-amino acid supplements e.g. XMTVI Anamix infant

NB: precursor-free L-amino acid supplements = medical foods

How many of your patients have precursor -free amino acid supplements (e.g. XMTVI Anamix infant) in the following age groups

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia					
Methylmalonic acidaemia – Vitamin B12 responsive					
Methylmalonic acidaemia – Vitamin B12 non - responsive					
Isovaleric acidaemia					

NB: precursor-free L-amino acid supplements = medical foods

Please calculate the dose of precursor-free amino acid supplements (e.g XMTVI Anamix Infant) used **for each of the age bands**. Please give this in both g/kg/daily of protein equivalent and as a percentage of total protein intake.

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia					
Methylmalonic acidaemia – Vitamin B12 responsive					
Methylmalonic acidaemia – Vitamin B12 non-responsive					
Isovaleric acidaemia					

NB: precursor-free L-amino acid supplements = medical foods

Prescription of precursor-free amino acids

- Do you give precursor-free L-amino acids in addition to your usual protein allowance or do they replace natural protein?
- How do you usually give the precursor-free L-amino acids in divided doses, with natural protein, mixed with what ?

NB: precursor-free L-amino acid supplements = medical foods

Type of precursor-free amino acids (medical foods)

- Which is your preferred type of amino acid supplement and why?
 - with added carbohydrate/fat/vitamins and minerals
 - with added carbohydrate/vitamins and minerals
 - L-amino acids only

Comments:

- How do you decide how much protein to give your patients e.g. WHO criteria, ammonia, blood gases, lactate etc?
- Do you allocate patients with mild or severe organic acidaemias the same or different protein allowances?
- Who decides within your IMD team how much protein to allocate patients?

- Do you recommend that animal protein is eaten to make up protein allowance?

Yes

No

Don't know

- Do you restrict odd-chain fatty acids in the diet

Yes

No

Don't know

- How many of your patients use special low protein foods?

- How many of your patients use low protein milk replacements e.g. Sno Pro or Pro-zero?

Adult patients only

- Please state the range of protein you would prescribe in g/day for your patients?
- Are there any differences for any disorder?

How many patients are on tube feeds for feeding?

Condition		Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia	Number of patients					
	% of total patients					
Methylmalonic acidaemia – Vitamin B12 responsive	Number of patients					
	% of total patients					
Methylmalonic acidaemia – Vitamin B12 non-responsive	Number of patients					
	% of total patients					
Isovaleric acidaemia	Number of patients					
	% of total patients					

How many patients are on overnight tube feeds only?

Condition		Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia	Number of patients					
	% of total patients					
Methylmalonic acidaemia – Vitamin B12 responsive	Number of patients					
	% of total patients					
Methylmalonic acidaemia – Vitamin B12 non-responsive	Number of patients					
	% of total patients					
Isovaleric acidaemia	Number of patients					
	% of total patients					

How many patients are on tube feeds for drugs only?

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia					
Methylmalonic acidaemia – Vitamin B12 responsive					
Methylmalonic acidaemia – Vitamin B12 responsive non responsive					
Isovaleric acidaemia					

Please indicate the number of patients on nasogastric or gastrostomy feeding tubes in each age group

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric Gastrostomy
Methylmalonic acidaemia – Vitamin B12 responsive	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy
Methylmalonic acidaemia – Vitamin B12 non- responsive	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy
Isovaleric acidaemia	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy	nasogastric gastrostomy

Please tick or list the reasons why your patients are tube fed

Criteria	Yes	No
Prevention of fasting		
Food refusal		
Unsafe swallow		
Inadequate energy intake		
Poor nutritional quality of diet		
To administer precursor-free amino acids		
Mechanical feeding difficulties due to neurological damage		
To administer emergency feeds only		
Other		

Please list all ingredients that you would add to tube feeds for patients with organic acidaemias in the following age categories

Condition	Infants Aged 0-12m	Children Aged 1-10y	Teenagers aged 11-16y	Adults aged >16y
Propionic acidaemia				
Methylmalonic acidaemia				
Isovaleric acidaemia				

How many patients are **currently** taking prescribed energy supplements (oral supplements only)?

Condition	Aged 0-6 m	Aged 6-12 m	Aged 1-10y	Aged 11-16y	Aged >16y
Propionic acidaemia					
Methylmalonic acidaemia – Vitamin B12 responsive					
Methylmalonic acidaemia – Vitamin B12 non- responsive					
Isovaleric acidaemia					

In which situations would you **routinely** prescribe vitamin and mineral supplements?

Situation	Yes	No
To all patients on low protein diets		
To patients only if they have a poor dietary intake on dietary assessment		
To patients only if they have a poor biochemical status		
Other: please state		
What type of supplements do you prescribe?		
Comprehensive vitamin and mineral supplements e.g. Paeditaric seravit, Phlexyvits, Fruitivits		
Partial vitamin and mineral supplements e.g. a multivitamin + iron and zinc purchased from retail shops		
Single vitamin /mineral supplements according to dietary assessment e.g. calcium		
Single vitamin /mineral supplements according to biochemistry e.g. iron or vitamin B12		
Other: please state		

Would you **routinely** prescribe any of the following fatty acid supplements?

Fatty acid supplements	Yes	No
Essential fatty acids (linoleic and α -linolenic acid) e.g. walnut oil		
Linoleic acid only		
α -linolenic acid only		
DHA (\pm EPA) only		
DHA (\pm EPA) + Arachidonic acid		
Other: please state		

Please indicate the concentration of carbohydrate and other type of ingredients you may routinely add to an emergency feed for the following age groups?

Ingredients	Aged 0-6 m	Aged 6-12 m	Aged 1-2y	Aged 3-10y	Aged >10y
10% glucose polymer					
15% glucose polymer					
20% glucose polymer					
25% glucose polymer					
Long chain fat emulsion					
MCT fat emulsion					
Precursor-free amino acid supplements (e.g. XMTVI Anamix infant)					
Electrolytes e.g. sodium, potassium					
Other. Please state					

Which nutritional measurements do you **routinely** undertake and how often do you do these on average?

Parameter	<i>Please circle which of the following measures you assess</i>	<i>Frequency. Please circle the answer that is closest to your practice.</i>
Growth	length/height, head circumference, weight, BMI	Weekly, monthly, six monthly, annually, rarely, never
Nutrient adequacy	3 day diet diary	Weekly, monthly, six monthly, annually, rarely, never
Protein status	total protein, albumin, pre-albumin	Weekly, monthly, six monthly, annually, rarely, never
Plasma amino acids	quantitative amino acid profile	Weekly, monthly, six monthly, annually, rarely, never
Iron status indices	haemoglobin, haematocrit, ferritin, complete blood count	Weekly, monthly, six monthly, annually, rarely, never
Essential fatty acid profile	Plasma essential fatty acids; erythrocyte essential fatty acids	Weekly, monthly, six monthly, annually, rarely, never
Trace element status	Plasma selenium, glutathionine peroxidase, zinc	Weekly, monthly, six monthly, annually, rarely, never
Vitamins	Vitamin B12, vitamin D, vitamin A, vitamin E, folate	Weekly, monthly, six monthly, annually, rarely, never
Metabolic control	Ammonia/blood gases/lactate/plasma or urine MMA/ ketonuria/ plasma carnitine	Weekly, monthly, six monthly, annually, rarely, never
Other		Weekly, monthly, six monthly, annually, rarely, never

Brief/basic information on main drug treatment policy from your unit (this is for background information only for the paper)

Does your unit commonly prescribe the following drugs in organic acidaemais	Propionic acidaemia	Methylmalonic acidaemia – Vitamin B12 responsive	Methylmalonic acidaemia – Vitamin B12 responsive non responsive	Isovaleric acidaemia
Carnitine				
Ammonia scavenger drugs e.g. Sodium benzoate				
Glycine				
Antibiotics e.g. metronidazole				
Laxatives for constipation				