PRACTICAL ASPECTS OF MANAGING LOW PROTEIN DIETS

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Abstract. Many inborn errors of metabolism can be successfully managed by restriction of natural protein intake, with or without special supplements. In some cases, there is a need for clear advice on management during metabolic crisis brought about by illness. Even when medical facilities and ongoing support are on hand, and special products are available and affordable, management of such conditions is challenging to families. Tailoring diet instruction to the needs and capabilities of the family is most likely to achieve success. Many Australian families cope better with simple guidelines aimed at reducing protein intake within the needs for growth and development and this style of management is also more likely to be successful in countries in which support is limited. Simple guidelines can take the form of increasing intake of certain types of foods and limiting others. Assessment of vitamin and mineral intake is however essential. Breastfeeding will provide less protein than standard infant formula, as well as immune protection, and needs to be encouraged and supported. Restricting meat and other high protein foods with increased intake of cereal and vegetable protein will still substantially decrease protein intake. Increasing intake of low protein vegetable sources or special low protein foods will also by default decrease protein intake. Families need specific guidelines for the care of their child when metabolic decompensation is a risk, with the aim of maintaining adequate energy intake, in forms that the child can eat or drink. A specialist pediatric dietitian can provide the expertise in formulating management plans appropriate for the family and facilities, with ongoing local management. The support of other families dealing with similar conditions can also be invaluable.

INTRODUCTION

Inborn errors of metabolism may be diagnosed by newborn screening or clinically. Diet is the major treatment in many of these disorders, usually by restricting intake of the substrate for the defective enzyme. In some cases special supplements may also provide the product of the affected reaction eg tyrosine in PKU, or may manipulate the balance of amino acids eg essential amino supplements in urea cycle defects. In our clinic, there is distinction between pharmacological supplements, which are prescribed by clinicians, and nutritional supplements ie those that supply nutritional needs. Only nutritional supplements will be discussed here.

The degree of restriction of substrate is dependent on many factors including:

- Individual needs based on tolerance of the affected substrate
- Treatment goals – published for PKU (Medical Research Council Working Party on Phenylketonuria, 1992; National Institutes of Health Consensus) but may need modification if other factors conflict
- Age at diagnosis. Diet changes are harder to make when eating patterns are established
- Facilities for ongoing monitoring eg plasma amino acids and availability of health professionals eg dietitians to provide management and education
- Availability of special products. Amino acid supplements allow sufficient intake of non-affected amino acids in many inborn errors of metabolism, allowing greater restriction of natural protein.
- Nutrient intake. High protein foods are usually rich sources of micronutrients so that intake is compromised on a low protein diet
- Practicality of dietary treatment. Can the family cope with and afford the restrictions involved?
- Times of illness and catabolism. In some inborn errors of metabolism there is a high risk of metabolic decompensation during illness. Families need specific guidelines for the care of their child with the aim of maintaining adequate energy intake, in forms the child can eat or drink.

If natural protein is to be restricted, individual needs for growth may be less than recommended requirements. However, with severe restriction of dietary protein comes

the need for careful monitoring. It is also essential to ensure an adequate intake of energy and micronutrients.

The 1985 FAO/WHO methods and estimates for protein requirements and safe levels of intake have been reviewed and revised (Dewey *et al*, 1996). Average requirement is defined as the lowest mean intake that will maintain functional needs in an individual. Safe level of intake is the amount that will meet or exceed the needs of requirements of practically all individuals in a population (mean requirements + 2 SD) (Table 1).

Table 1. Safe level of intake and mean requirement for protein.

Age	Average requirement g protein/kg	Safe level g protein/kg/d
0-1 month	1.99	2.69
1-2 month	1.54	2.04
2-3 month	1.19	1.53
3-4 month	1.06	1.37
4-5 month	0.98	1.25
5-6 month	0.92	1.19
6-9 month	0.85	1.09
9-12 month	0.78	1.02

For infants, the choice of feeding influences protein intake. Breast milk is lower in protein than standard formula (Table 2) and provides immunological benefits, particularly important for the child at risk of metabolic decompensation. For the bottle-fed baby, use of a formula suitable from birth rather than a follow-on formula after 6 months of age will lower protein intake. Continued use of a formula well into the first year of life and possibly longer, also maintains a lower protein intake compared to cows or goats milk, as well as improving the intake of some vitamins and minerals.

Table 2. Protein content of milks.

	g protein/l
Breast milk	10
Standard formula (Infant formula suitable from birth)	14-18
Follow on formula (Infant formula suitable from 6 months)	21-23
Cows milk	34

If breastfeeding or standard formula provides too much protein then intake can be reduced by supplementing with a protein free formula or disease specific amino acid restricted formula.

Although percentage protein content of foods varies (Table 3), in practical terms protein restriction can be achieved by grouping foods into 3 categories:

- Animal sources of protein eg meat, milk, fish, eggs, chicken
- Cereal protein, potato peas and corn (possibly legumes, nuts)
- Other vegetables and fruit

Within these categories protein content per serve is similar.

Amino acid supplements have a role in the treatment of many inborn errors of metabolism as follows:

- Providing sources of non affected amino acids to allow normal growth and development.
- May provide product eg tyrosine in PKU.
- May improve growth as well as tolerance of affected amino acid (Acosta and Yannicelli, 1994).
- Influential in variation of plasma amino acid levels during the day (MacDonald *et al*, 1996).
- Allow manipulation of amino acid balance, eg essential amino acid supplements in urea cycle disorders.
- Act as a source of energy and other nutrients.

Maintaining energy intake can be a challenge but one that most children appear to regulate well themselves. Dietitians at the Metabolic /PKU clinic at The Children's Hospital at Westmead give advice on energy sources, but rarely calculate dietary energy intake, which is time consuming and often inaccurate, relying instead on growth and weight gain and appetite as guides. Useful sources of energy include

- Milk substitutes – cream, coffee whitener, Duocal
- Fruits and vegetables
- Increasing fat intake with oils and spreads
- Sugar based foods and snacks – but dental care is important
- Lower protein foods available in supermarkets, eg jellies thickened with vegetable gums, tapioca and rice based snack foods and drinks
- Specialised low protein products, eg low protein breads, pastas

Many high protein foods are rich in other nutrients, so that restriction can lead to inadequate intake of vitamins, minerals and essential fatty acids. Such foods may need to be supplemented on low protein diets, and compliance

Table 3. Protein content of foods.

Food	Protein content (%)	Protein content by serve size
Milk	3	400 ml milk = 12 g
Eggs	12	2 eggs = 12 g
Fish	20-25	½ small fillet =12 g
Meat	30	1 small chop or chicken drumstick = 12 g
Nuts	15-25	15 nuts = 3 g
Legumes	5-15	¼ cup baked beans = 3 g
Bread	8	1 slice bread = 3 g
Rice	7 (raw)	½ cup boiled rice = 3 g
Vegetables	1 -5	1 medium potato = 3 g other vegetables except peas and corn < 1.5 g per serve
Fruit	1	< 1.5 g per serve

needs to be monitored. Experience at the Children's Hospital Metabolic Clinic and within the medical literature suggests that compliance, particularly with taking calcium supplements, is not good (Thompson, 2000).

The dietary management of inborn errors of metabolism can be illustrated by PKU. Approximately 10 new cases a year are diagnosed by newborn screening in NSW. The treatment goals are based on the UK guidelines (Medical Research Council Working Party on Phenylketonuria, 1992). The diet is low in phenylalanine, with a protein supplement free of phenylalanine but supplemented with vitamins and minerals. The diet is based on fruits, vegetables and low protein foods with no animal protein or aspartame. The phenylalanine intake is measured in units where 1 unit = 15 mg phenylalanine. This diet is used across Australia with the PKU Handbook (Dietitians Working Party of the Australasian Society for Inborn Errors of Metabolism, 1996) providing information for the families.

Older children tend to be non-compliant in counting units but they avoid high protein foods. Assessment is made on the basis of biochemical testing and a simpler counting system may be commenced. Concentrating on increasing intake of low protein foods without counting protein and phenylalanine can improve compliance.

In inborn errors of metabolism where there is a high risk of metabolic decompensation during illness (Dixon and Leonard, 1992), families need specific guidelines for the care of their child. Metabolic decompensation may be defined as a loss of metabolic control, often in association with an infection or other acute illness. As a consequence, biochemical balance is lost, and there is a risk of neurological symptoms developing. The unwell protocol is started as soon as the child shows any sign of being unwell (eg upper respiratory infection) and is contraindicated only if the child will not tolerate oral or nasogastric feeding. This protocol was developed to:

- Maintain adequate energy intake to reduce catabolism of body proteins by meeting energy needs with carbohydrate and fat.
- Provide sufficient non-restricted amino acids to enhance protein synthesis, in conditions affecting specific amino acids.
- Reduce intake of natural protein in order to reduce intake of amino acids /nitrogen that cannot be metabolised

The recommendations for individuals during illness are dependent on age, severity of metabolic condition and severity of the acute illness. Table 6 gives guidelines used by the Children's Hospital at Westmead. These are practical guidelines for the child well enough to be managed at home, as it is not expected that the older child will be able to consume full theoretical energy and fluid requirements. The complete protocol is available on request.

A pediatric dietitian specialising in metabolic disorders can enhance the care of children diagnosed with inborn errors of metabolism (Acosta, 1997) by:

- Designing and modifying the diet to suit the individual needs of the child.
- Evaluating nutritional status and nutritional adequacy of diet.
- Educating and supporting the family and child.
- Providing up to date information on product and

Table 4. Guidelines for daily unwell feed.

Age	Final Volume of feed	Protein intake from incomplete amino acid supplement if used	Energy concentration of unwell feed	Natural protein intake or complete protein supplement
0 –12 months breast or bottle fed	Calculate on 150 ml/kg	Minimum of 1g/kg/d (or usual supplement dose whichever is the larger). Do not exceed 3 g protein per kg.	If no supplement Energivit at 1 kcal/ml If supplement addition of Energivit to final concentration of 1 kcal/ml. May need to decrease concentration if vomiting or diarrhoea	Cease breast feeding (mum to express) or standard formula for 12 hours contact doctor. Recommence breast feeding or formula on advice of doctor/ dietitian Low protein solids that child usually eats only
Over 1 year	100 ml/kg to a maximum of 1000 ml per day	0.8g/kg/d (or use the usual supplement dose – whichever is the larger)	Addition of Duocal or Polyjoule to final concentration of 1 kcal/ml of mixture. Older children may tolerate 1.2 kcal/ml (eg >4 years) or 1.5 kcal/ml (eg >12 years) May need to decrease concentration if vomiting or diarrhoea	Aim ½ -2/3 usual protein intake from food / complete protein supplement unless otherwise advised by doctor/dietitian. It may be advisable for no natural protein for 24-48 hours, but this should not exceed 48 hours

food availability.

- Development of practical unwell management.
- Preparing educational material appropriate to country and ethnicity.
- Coordinating care with other health care providers.

Dietitians working in this field also need a support network, since they become local dietary experts despite a very small number of patients. Such support is offered through:

- PNO-METAB-L – a listserv for registered metabolic Dietitians. Contact Rani H Singh, Division of Medical Genetics, Emory University, 2040 Ridgewood Dr, NE, Atlanta GA 300322.
- Australasian Society of Inborn Errors of Metabolism (ASIEM) Dietitian group – contact the author.

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