

Nutrition and hydration

Irène Benigni

Dietician, member of paramedical council of French Rett Syndrome Association

Nutrition

The health and wellbeing of patients largely depends on their diet. Malnutrition and growth retardation are frequently associated with Rett syndrome [1], [2].

They may result in a range of causes:

- High level of energy expenditures and specific needs (especially in peri surgical arthrodesis cases)
- Feeding difficulties linked to:
 - Inability to self-feed and drink
 - Bad oral state
 - Weak chewing due to muscular rigidity of the masseters (mastication muscles)
 - Swallowing difficulties
 - Sensory feeding disorder inducing an adverse reaction to the eating and/or drinking process and impairing food and/or drink intake
 - Excessively slow eating

Digestive problems also interfere with eating, especially GOR (Gastro-Oesophageal Reflux), abdominal bloating and constipation.

Nutrition status must be assessed

- Children

It is essential to record and chart children's weight (monthly) and height (annually). If weight appears to be stagnating or decreasing, malnutrition is beginning. If growth slows, malnutrition is suspected to become chronic. About 50% of Rett individuals are undersized compared to standard and this difference increases with age [3].

The weight/height index (measured weight x 100/standard weight for child's height) should be between 90 and 110%. Below 90%, malnutrition risk occurs.

- Adults

A 5% weight loss in a month or 10% in 6 months or a Body Mass Index inferior to 18.5 kg/m² are alert factors.

Nutritional requirements of patients with Rett syndrome

Nutritional requirements can vary significantly from one person to another.

They seem to be high in Rett syndrome and many girls reputedly have large, healthy appetites yet remain malnourished. [2]

About 80 % of the parents estimate that their daughter is well nourished even if she is malnourished. [1]

No hypothesis has been confirmed concerning this paradox: energy expenditure increased in case of spasticity, stereotypies, other cases ...

Other girls have feeding difficulties and cannot eat enough to reach their nutritional requirement.

The causes of malnutrition must be identified so as to be treated if possible, and enriched food must be given to improve the dietary intakes.

Enriched food

Add into the meals in order to concentrate the calories in a small volume:

We can add proteins with concentrated milk, powdered milk, cheese, growth milk as a drink, eggs, jam, tuna or salmon crumbs ...

We can add fat with butter, margarine, oil, cream, chocolate spread, peanut butter, almond purée ...

We can add carbohydrates with starch, cakes, crumbles, almond powder, nut powder, breadcrumbs ...

We can also give high-energy and high-protein food supplements produced by the pharmaceutical industry, commercialised as desserts, soups, mashed cereals, fruit juices, protein powder, maltodextrin ...

Proposed drinks with nutrient intake (milk, fruit juice)

Specific needs

The prevalence of osteoporosis in Rett syndrome is very high [4]. Nutritional status as well as intakes of calcium and vitamin D are important as prevention measures [5]. A deficiency in vitamin D can lead to the occurrence of osteoporosis. It can be useful to give high-fat fish one or twice a week and to give the patient a supplement of vitamin D.

A deficiency in calcium is multifactorial but can be caused or worsened by insufficient intake [3]. It is important to give dairy product three times a day and to control their intake.

Adapting food texture to chewing abilities

Some Rett individuals can get tired quickly during eating, due to the masseter muscles rigidity, so chewing becomes weak.

A grinding mill or a blender has to be used, or food must be pureed or mashed to avoid suffocation, to decrease digestive pain and, to improve digestion and increase the patient's eating pleasure.

In case of trouble with swallowing, ground foods will be easier to ingest.

Minced foods:

We will give this texture to persons whose chewing is weak, but whose lateral movement of the tongue is adequate and who have no swallowing problems. Meat must be minced, and vegetables crushed. Sometimes, small cereal grains like rice, semolina, some pasta products, lentils, peas are to be avoided because it is difficult to collect and homogenize them in the mouth. All desserts except fruit too hard to chew are allowed.

Ground foods:

This texture is adopted for RS individuals:

- who suffer from swallowing difficulties,
- or who do not chew and are unable to move their tongue laterally.

Foods must be finely ground separately. Cheese spread must be ground with purees or soups; otherwise, when ingested on their own, they stick to the mucous membrane.

Recommended desserts are: ground fresh fruit, apple sauce, milk desserts with or without any starch, or any pastry into which a binder of any kind has been added.

Check as causes of feeding difficulties

- Possible dental pain, orthopaedic pain, osteoporosis pain
- Sensory feeding disorder that can be reduced with appropriate reeducation by a speech therapist
- Digestive problems, especially gastro-oesophageal reflux, abdominal bloating and constipation, prevalence of those troubles is estimated about 74% [6] and must be systematically suspected and treated.

If feeding difficulties remain important, tube feeding can complement or replace oral nutrition. This is a system of nutrition that uses nasogastric or more often gastrostomy tube feeding. It allows to improve growth and weight gain. [7]

Gastrostomy is always the result of a collegial and consensual decision involving the patient, the family and the doctor(s).

Hydration

Hydration balance is often precarious in people with Rett syndrome, either due to inadequate intake (refusal to drink, inability to drink themselves) or due to increased losses (hyperthermia, drooling, vomiting, diarrhoea, high heat).

Practical advice

Offer drinks during and outside meals.

Only water is essential but some Rett girls do not appreciate water because of the lack of sensory information (no taste, no texture, inadequate temperature).

We can propose:

- Flavoured water, infusions, sparkling drink, fruit juice, dairy drink, tea, coffee, chicory, smoothies, yoghurt drinks, or salted drinks such as soup, vegetable stock...
- All these drinks can be offered at different temperatures (warm temperature is often appreciated) and/or thickened with starch or gelified with agar or gelatin, especially in the case of swallowing disorders

The prevention and fight against the risk factors require a multidisciplinary approach involving “cure and care”.

This team should include a speech therapist, a gastroenterologist and a dietician specialized in developmental disabilities.

Nutrition represents one of the most important aspects of treatment in Rett Syndrome but it is often neglected. It is the cornerstone by which all other forms of therapies are made possible.

Such problems create an additional burden for families and they may need to spend huge amounts of time feeding their daughters or sons.

References

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