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Assessment and management of nutrition and growth in Rett syndrome

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Abstract

Objectives—We developed recommendations for the clinical management of poor growth and weight gain in Rett syndrome through evidence review and the consensus of an expert panel of clinicians.

Methods—Initial draft recommendations were created based upon literature review and 34 open-ended questions where the literature was lacking. Statements and questions were made available to an international, multi-disciplinary panel of clinicians in an online format and a Microsoft Word

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formatted version of the draft via email. Input was sought using a 2-stage modified Delphi process to reach consensus agreement. Items included clinical assessment of growth, anthropometry, feeding difficulties and management to increase caloric intake, decrease feeding difficulties and consideration of gastrostomy.

Results—Agreement was achieved on 101/112 statements. A comprehensive approach to the management of poor growth in Rett syndrome is recommended that takes into account factors such as feeding difficulties and nutritional needs. A BMI of approximately the 25th centile can be considered as a reasonable target in clinical practice. Gastrostomy is indicated for very poor growth, if there is risk of aspiration and if feeding times are prolonged.

Conclusions—These evidence- and consensus-based recommendations have the potential to improve care of nutrition and growth in a rare condition and stimulate research to improve the current limited evidence base.

Keywords

Rett syndrome; gastrointestinal; growth; feeding difficulties

INTRODUCTION

Rett syndrome is a neurological disorder caused by mutations in the *MECP2* gene.[1] Core features include a period of normal development followed by regression with loss of communication and hand function skills, the development of hand stereotypies and impaired gait.[2] In addition, Rett syndrome is associated with epilepsy,[3] scoliosis[4] and poor growth.[5, 6]

Growth retardation is one of the supportive criteria for Rett syndrome. [2, 7] Poor growth has been observed as early as three months of age [8] and may continue into adulthood.[5, 6, 9, 10] Contributing to this may be a number of factors affecting food intake, both separately and in combination, [5, 6, 9, 10] including feeding difficulties, oromotor dysfunction and other digestive tract disorders, factors which themselves are likely to be determined by genotype[11] and, as such, are an intrinsic component of the disorder. There are additional neurological complexities such as apraxia, autonomic dysfunction including hyperventilation, disrupted sleep patterns and the development of scoliosis, each of which is likely to have some influence on feeding and growth. Thus, unlike many other neurological disorders, Rett syndrome can be considered as a pervasive disorder of growth[12] requiring specific strategies for management over and above what is typically required for children with other developmental disabilities.[13] Data from the Australian Rett syndrome population indicated that feeding difficulties manifested as prolonged feed times in approximately three quarters; there were requirements for food to be pureed, mashed or chopped in approximately two thirds; gagging or choking with food or liquids occurred in approximately one quarter; and one fifth required enteral nutritional support.[6] A recent US survey of 983 families with a daughter with Rett syndrome found similarly that 28% had a gastrostomy.[5] Those with a late truncating *MECP2* mutation including C terminal deletions were more likely to have higher BMI and weight Z scores compared to other mutations, although there was less apparent variation in height by mutation.[6] Recommendations that take into account specific issues associated with Rett syndrome could contribute usefully to the nutritional management in this disorder.

There are few studies available on the management of poor growth in Rett syndrome and these are generally restricted to small case series,[10] narrative,[14] or systematic reviews and position statements in the general neurodevelopmental literature.[15–17] As Rett syndrome is rare, most clinicians typically have exposure to very few patients. There is a need for

systematic review of the literature and the pooling of the expertise to indicate current best practice with regard to nutritional assessment and management.

A consensus building process using the Delphi technique was recently employed to develop clinical recommendations for the assessment and management of a range of gastro-intestinal problems in Rett syndrome. This paper describes the development of recommendations relating to the assessment and treatment of issues relating to calorie intake and feeding difficulties including consideration of gastrostomy.

MATERIALS AND METHODS

This project took the format recommended for the development of clinical guidelines by the National Health and Medical Research Council, Australia. The project was directed from the Telethon Institute for Child Health Research, Western Australia from 2009 to 2011 and was approved by the Ethics Committee of The University of Western Australia.

Literature review and parent perspectives

A literature search was performed. Databases included PubMed, Medline, the Cochrane Library, EMBASE, CINAHL, PsychINFO, Proquest Health and Medical Complete and Web of Science; online libraries included those of the World Health Organisation, CMA – Clinical Practice Guidelines, Geneva Medical Research, the National Guideline Clearinghouse, National Electronic Library for Health, Scottish Intercollegiate Guidelines, National Institute for Health and Clinical Excellence, and the Trip Database. Search and key words included combinations of Rett syndrome, cerebral palsy, developmental disability, intellectual disability, co-morbidity, gastrointestinal, growth and feeding. The search was limited to full papers in English from 1986 to 2012. Statements relevant to the clinical assessment and management of poor growth in Rett syndrome were extracted from the full text.

Rettnet, an online email information interchange for parents/persons with an interest in Rett syndrome,[18] was reviewed to collect parent and caregiver perspectives on poor growth and contributing factors such as calorie intake and feeding difficulties. Postings from January 2008 to March 2009 were reviewed.

Initial expert panel recruitment, guideline development and redrafting using the modified Delphi process

Clinicians in the disciplines of developmental paediatrics, child neurology, clinical genetics, paediatric gastroenterology, pediatric surgery, dietetics, speech therapy and nursing with experience with Rett syndrome from different countries were identified through networks of researchers, publications, and the Australian Rett Syndrome[4] and InterRett[19] databases. Network sampling using collegial recommendations was used to expand the sample. Potential participants were contacted by email to request their participation with English as the language used. Fifty seven clinicians were successfully contacted and 38 (66.7%) agreed to participate as the expert panel.

Referenced statements extracted from the literature were categorized into topic areas and accompanied by a 5-point Likert scale for agreement rating (strongly agree, agree, neither agree or disagree, disagree, strongly disagree) with space for comments. If there was no literature or in response to Rettnet postings, an open-ended question on management was constructed. Parents who were participants in the Australian Rett Syndrome Consumer Reference Group reviewed the proposed research plan and scope of the statements. The statements and questions were listed in a Microsoft Word document format and an online version created using HTML form and PHP script. Participants could indicate their level of

agreement with dropdown menus and type in comments in the spaces provided. Responses could be saved, edited and submitted in stages. Data received were stored in a MySQL database (Sun Microsystems, Inc., Cupertino, USA) on a secured server located at the Telethon Institute for Child Health Research.

For the first round of the Delphi process, members of the expert panel provided feedback on the emailed Microsoft Word document or online version which was username and password protected. Panel members were asked to respond to nominated sections relevant to their professional scope. A pre-determined level of consensus was established: consensus was attained where a minimum of 70% of responses were within one response category of the median response. The second guideline set was informed by these responses and returned comments and was then sent for consensus assessment. This process was a modified Delphi process because consensus or lack of consensus was clear at each time point and therefore we did not send panel members their previous responses together with the median responses for the group. A level of evidence using the Scottish Intercollegiate Guidelines Network grading scheme[20] was applied to each item for which there was consensus: level 1 representing evidence from systematic reviews and randomized controlled trials (RCTs), level 2 case control or cohort studies, level 3 case reports or case series and level 4 expert opinion.

RESULTS

Literature review and parent perspectives

Search of the scholarly databases revealed 300 citations as potentially relevant. Of these, 77 articles and 3 sets of guidelines from online libraries were retrieved and reviewed in full text: 31 supported the final guideline draft. The first draft of the recommendations was reviewed to check that it contained the main issues raised by families in the Rettnet postings.

Expert panel participation

Of the 38 clinicians agreeing to participate, 27 (71.1%) provided data. Seven (25.9%) were child neurologists, six (22.2%) were gastroenterologists, four (14.8%) were clinical geneticists, 3 (11.1%) were paediatricians, two (7.4%) were dieticians and two (7.4%) were speech therapists. There was also one (3.7% each) disability specialist, nurse, and dysphagia and feeding specialist.

Initial guideline draft and redrafting using the modified Delphi process

The initial guideline draft comprised sections on the clinical assessment of calorie intake, feeding difficulties, anthropometric measures and consideration of gastrostomy; and treatment to increase calorie intake, address feeding difficulties and use of gastrostomy. It included 46 statements, 34 questions and a reference list, and all of the Rettnet topics were represented. The responses to these questions were either supportive of the responses to the statements, or were used to construct items for round 2. Twenty eight items were included in the second round. The final document comprised 112 separate statements listed in Tables 1 to 6 and there was agreement for 101 of these. These are presented together with levels of evidence, the median responses and the percentage of responses within one category of the median.

Assessment of calorie intake, feeding difficulties, anthropometry

The items describing the assessment of calorie intake and feeding difficulties are shown in Table 1. There was consensus that nutritional assessment should be undertaken twice annually in those aged up to 12 years and annually thereafter. Information should be sought on the quantity and variety of food intake as well as texture preference and tolerance. A 24

hour diet recall is one mechanism for assessing food intake. It was considered important also to include enquiries or observation of meal time stress for the child and the carer. Functional capacity for self-feeding, chewing and swallowing can be also assessed from carer report, by direct observation or through video, or a referral to a speech therapist or occupational therapist for assessment can be valuable. The Schedule for Oromotor Assessment (SOMA) is an assessment tool for oromotor function in pre-verbal children [21, 22] and has been used clinically for those with Rett syndrome.[9] Questions to carers should include drooling, constipation, vomiting and regurgitation, appetite and the length of a typical meal time. In the course of the consultation the clinician should assess the need for physical positioning and postural supports, special equipment use, modification of food consistency and prompting during mealtimes. Video fluoroscopy can be used to aid diagnosis of aspiration or other respiratory concerns and to inform texture modification needs (Table 1).

The items describing anthropometric and biochemical assessments are shown in Table 2. The measurement of weight was considered a critical part of clinical assessment. It was acknowledged that measuring height is more problematic especially when some girls and women are unable to stand or have a spinal deformity. For those who can stand a stadiometer should be used and for those under 2 years or unable to stand supine length should be measured. Height can be estimated by measuring lower leg height for those to the age of 12 years who cannot stand.[23] The algorithms for estimating height in centimeters are (3.26 × tibial length) + 30.8, or (2.69 × knee height) + 24.2. [23] (It was agreed that a standard growth chart should be used to plot anthropometric measures over time and that a BMI>25th centile was a reasonable target for clinical practice, with attention not to exceed the normal range. (Table 2)

There was strong consensus that it was important to check the oral health of the patient and where applicable to make the appropriate dental referral. It was also agreed that the clinician should be aware of the possible additional energy requirements associated with the breathing abnormalities in Rett syndrome.[6] However to date this has not been substantiated in a clinical study. [24] In line with guidelines for the gastrointestinal care of children more generally,[25] biochemical tests that should be undertaken as part of a nutritional assessment include haemoglobin, ferritin, white blood cell count, differential count, albumin/protein, urea, creatinine, electrolytes, vitamin D and calcium. For those at risk of specific micronutrient deficiency, eg, those on some anti-convulsant medications, with poor growth, with symptoms suggestive of gastro-intestinal malabsorption or receiving at least 50% of their nutritional needs via enteral support, it was recommended that in addition to the above they should have testing of folate and B12. The frequency of biochemical testing should be determined by growth status. (Table 2)

Management to increase calorie intake and to reduce feeding difficulties

It was agreed that energy requirements should be based on serial growth measures but if underweight, as indicated from serial growth charts, the energy requirements should be above the recommended calorie intake for weight[26]. Energy dense foods are the most appropriate way of increasing caloric intake [26–28] and frequent snacks of high calorie nutritional supplements can be administered. The panel agreed that a daily diet containing the recommended daily allowance for essential nutrients was optimal, administered during frequent small feeds offered throughout the day.[14] There is no evidence that gluten free or lactose free diets are associated with improved growth. (Table 3)

The majority of the panel members supported the promotion of optimal participation in feeding activities. For example, for some girls it may be worth a trial of intensive therapy to optimize abilities as new skills can be learned.[9] Physical positioning and postural support such as supporting jaw and avoiding hyperextension may be applicable.[29–31] There may also

be a need for special equipment use[30] or prompting and socializing during meals.[31, 32] (Table 4)

Consideration of gastrostomy

The benefits of gastrostomy on which the panel agreed were that it would allow shorter mealtimes and reduced chest infections for the affected individual and an improved quality of life for caregivers.[33] A gastrostomy in itself was not likely to result in reduced vomiting, reflux, constipation or pain. The panel agreed that gastrostomy should be considered where there is failure to thrive despite efforts to increase caloric intake, where oromotor dysfunction was causing an unsafe swallow and where the length of the feeding time was causing stress for both carer and child. However they also agreed that complications included infection, leaking, dislodgement or migration of the tube, intestinal perforation and an increased risk of reflux. A summary of the benefits and complications of gastrostomy and gastro-jejunostomy in relation to Rett syndrome is presented in Table 5.

If gastrostomy is performed, clinical monitoring following insertion of a gastrostomy should include a general and nutritional assessment, review of gastro-intestinal function and fluid status, check on feeding tube and stoma site and importantly management of equipment and any support needs for the home.[17] (Table 5) The panel agreed that aggressive nutritional therapy and gastrostomy placement can favourably alter adverse trajectories for growth in Rett syndrome.[34, 35] There was strong consensus on the need for parents to have practical and emotional support before and after the insertion of a gastrostomy tube[34]. Fundoplication is not done routinely but needs to be considered in the presence of severe gastro-oesophageal reflux. There was also agreement that a trial with a nasogastric tube could assist in determining the potential for weight gain that may occur with gastrostomy feeds and correct existing malnutrition while waiting for a gastrostomy. A gastro-jejunostomy tube can be used where gastrostomy feeds are not tolerated[36, 37] but the panel agreed that disadvantages are their tendency to migrate back into the stomach, for the tube to obstruct and the need for slow continuous feeds. In some settings the dependence on physician support for changing gastro-jejunostomy tubes may also be a disadvantage.

DISCUSSION

Poor growth is one of the supportive diagnostic criteria of Rett syndrome,[2] usually observed as decreased velocity of head growth, height and weight for age. The reasons for poor growth in Rett syndrome are not well understood but contributing factors include genotype[6, 38] and the motor impairments that affect chewing and swallowing.[5] This project identified relevant literature relating to Rett syndrome and developmental disability and with parental input and expert clinician consensus, integrated the evidence using a modified Delphi technique. We have worked systematically to develop recommendations that assist clinicians and families to engage in best practice.[39] Our document is practical and comprehensive as a guide to the assessment and management of poor growth and feeding difficulties in Rett syndrome.

There is a clear understanding that comprehensive clinical attention to the medical health issues of Rett syndrome is required, including feeding and nutritional support as we have described. We recruited an international and multi-disciplinary panel to provide expertise to discuss the relevant issues. Very little “drop out” of panel members occurred and using methods similar to our previously developed guidelines for scoliosis in Rett syndrome,[40] the group engaged in timely consultation using email and online surveys. In the absence of high level evidence, this document represents a current best effort to provide clinicians with practical guidance in this important area of care. Our recommendations are comprehensive with a life span approach and will be useful, not as a recipe for clinical practice, but as a

guide for embedding the monitoring and management of poor growth within routine management. A significant limitation of this project is the paucity of peer-reviewed literature and therefore, literature in the general neurodevelopmental field and the consensus of experts played particularly important roles.

Growth problems are reported frequently, often by families who are puzzled that their daughter has an apparently good appetite. Assessment of ten girls with Rett syndrome using doubly labeled water found that the energy expenditure of involuntary activities including hand stereotypies did not increase and in this small sample appeared not to have contributed to growth failure.[24] The contribution of autonomic abnormalities including hyperventilation to growth failure has not been assessed to date. Some females gain excess weight in association with gastrostomy. Our panel recommended a regimen of feeding that aims for at least >25th centile as a minimum with due attention not to exceed the normal range. New centile curves based on 816 girls and women with Rett syndrome now provide additional information with which to monitor clinical progress and determine the necessity for intervention.[41] Our recommendation would be approximately equivalent to the 50th centile values on the new Rett syndrome specific growth charts which were not available at the time of the Delphi process.

Poor feeding skills can contribute to poor growth including difficulties chewing and swallowing foods and involuntary tongue movements,[5] presumably a result of altered muscle tone and dyspraxia as well as poor motor control. Hypersalivation, hyperventilation and breath holding can further interfere with feeding. These difficulties are compounded by poor communication skills that make it difficult for the girls and women to express what or how much they want to eat. As a result, fewer calories may be consumed than are necessary for growth and development. Cross sectional studies have found that in women with Rett syndrome, hand function can be poorer[42] and the delay to first swallow longer[31] reducing abilities to self-feed. These findings are consistent with observations of increased muscle tone and paucity of movement with advancing age.[43] There is a limited evidence base for the training of feeding skills to improve feeding[32, 44, 45] and benefits have nevertheless been observed for girls as well as women.[45] This justifies assessment and a trial of intensive treatment for some because gains can be made, but more systematic investigation of the effectiveness of these therapy strategies is necessary to clarify this recommendation.

The decision by a clinician to recommend a gastrostomy and for a family to accept this is difficult in the management of Rett syndrome. Health outcomes of gastrostomy have been studied in general[33, 46] and include advantages of improved height and weight gain.[35] However, there are potentially many other benefits of gastrostomy such as improved respiratory health, decreased mealtime stress and improved carer quality of life. There is a need to examine these issues systematically so that clinicians can support the emotive components that accompany this clinical decision making. There was no consensus that gastrostomy should be accompanied by fundoplication, and clinical practice appears to vary. It should be considered with caution in those with significant air swallowing given the case reports of rare stomach perforation.[47, 48] Our panel recommended in some instances a trial of nasogastric feeding may be used to assess the potential for weight gain, although some panel members described the associated discomfort as counter-productive. Clearer assessment of this management method is also required.

Rett syndrome is a complex and overwhelming disorder with many aspects to its medical management best undertaken using the expertise of specialists working in the context of a multi-disciplinary team. We have developed practical baseline recommendations for the monitoring and management of poor growth and feeding difficulties in Rett syndrome using a comprehensive, multidisciplinary approach. The level of evidence for the statements was

low and the expert panel drew heavily on clinical experience when determining their agreement. This document provides important leads for the planning of further research. For current clinical care however, the recommendations can be used as a basic plan to promote discussion between caregivers and their clinicians and as an aid for clinical management.

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Table 1

Assessment of calorie intake and feeding difficulties

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Nutritional assessment should be conducted			
approximately 6 monthly in girls younger than 5 years		Agree	22/25 (88.0)
approximately 6 monthly between the age of 5–12 [27, 49]	3	Agree	16/20 (80.0)
yearly during adolescence and adulthood		Agree	19/20 (95.0)
2. Nutritional assessment should include [10, 13]			
caregiver reports of food intake	3, 2+	Agree	24/26 (92.3)
caregiver reports of variety of foods		Agree	25/26 (96.2)
caregiver reports of texture preferences and tolerance		Agree	25/26 (96.2
food intake assessed with a 24 hour diet recall including type and quantity of food		Agree	20/26 (76.9)
3. Meal time stress			
for the carer can be assessed by the time taken to feed a standard meal.		Agree	17/20 (85.0)
for the girl or woman can be assessed by associated symptoms of choking, gagging and crying		Agree	20/20 (100)
can also be assessed by viewing a video recording of meal time.		Agree	20/20 (100)
can be assessed by direct observation		Agree	20/20 (100)
4. Functional capacities for self feeding, chewing and swallowing can be assessed by			
direct observation		Agree	19/20 (95.0)
video of meal time		Agree	20/20 (100)
video fluoroscopy		Agree	19/20 (95.0)
detailed history from carers		Agree	20/20 (100)
5. Assessment includes caregiver report of[9] feeding difficulties	3	Strongly agree	26/26 (100)
drooling		agree	25/25 (100)
constipation		Agree	24/25 (96.0)
regurgitation		Agree	25/25 (100)
appetite		Agree	26/26 (100)
length of typical meal time		Strongly agree	23/23 (100)
6. Assess need for[29]			
physical positioning and postural supports	2+	Strongly agree	21/23 (91.3)
special equipment and utensil use		Agree	21/23 (91.3)
prompting and socialising during mealtimes		Agree	23/24 (95.8)
modification of food consistency		Strongly agree	26/26 (100)
7. Request assessment of feeding, chewing and swallowing function	3	Strongly agree	25/26 (96.1)

Item	Level of evidence[a]	Median response	n/N (%)[b]
by the appropriate allied health professional[50]			
8. The SOMA[c][21, 22] can be used to assess feeding in girls and Women with Rett syndrome[9]	3	Agree	13/15 (86.7)
9. Video fluoroscopy can be used to aid diagnosis of aspiration or other respiratory concerns [31, 50]	3	Agree	21/23 (91.3)
10. Video fluoroscopy can inform texture modification needs [31, 50]	3	Agree	21/23 (95.7)

[a] Scottish Intercollegiate Guidelines Network,

[b] numerator is the number of responses with median response or 1 category either side and denominator is the number of clinicians in the panel whose expertise was relevant to this item,

[c] SOMA - Schedule for Oromotor Assessment

Table 2

Anthropometric measures and other clinical assessments

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Weight should be measured as part of clinical assessment [10, 13, 15, 49]	3,3,3,2+	Strongly agree	24/24 (100)
2. Measure height using a stadiometer if the girl or woman is able to stand [10, 15, 49]	3	Strongly agree	16/16 (100)
3. Supine length should be obtained if younger than 2 years or if the girl or woman is unable to stand[15]	3	Agree	24/24 (100)
4. In the presence of skeletal deformities, alternative measures such as lower leg length or upper arm length may be obtained [15, 49]	3	Agree	18/22 (81.8)
5. BMI > 25th centile in the general population can be considered as a reasonable target in clinical practice (approximately equivalent to 50th centile in Rett syndrome[41]), with attention not to exceed the normal range		Agree	13/18 (72.2)
6. Height may be monitored using knee height if the person is unable to stand straight		Agree	14/14 (100)
7. Anthropometric measures over time in girls with Rett syndrome should be assessed with standard and Rett syndrome-specific growth charts[41]		Agree	15/16 (93.8)
8. Carefully observe the teeth and mouth to check oral health as may interfere with oral intake and refer to a dentist when appropriate[15]	3	Strongly agree	22/22 (100)
9. Biochemical testing includes the following tests[25]	4		
Haemoglobin, Ferritin		Agree	23/23 (100)
WBC count, differential count		Agree	22/23 (95.7)
Albumin/Protein		Strongly agree	21/23 (91.3)
Serum urea nitrogen, creatinine, electrolytes		Agree	23/23 (100)
Vitamin D, Calcium, Phosphate and alkaline phosphatase		Agree	23/23 (100)
An expanded panel including B12 and Folate should be considered if at risk of specific micronutrient deficiency, eg, those on some anti-convulsant medications or less than the 25th centile on BMI, or those with symptoms suggestive of gastro-intestinal malabsorption		Agree	23/23 (100)
10. Any girl or woman receiving at least 50% of daily nutritional needs via enteral support should have the following biochemical tests as indicated by growth status[17]	3		
Full blood count and ferritin		Agree	23/23 (100)
Albumin/ Protein		Agree	23/23 (100)
Urea, electrolyte and Creatinine		Agree	22/23 (95.7)
Liver function tests		Agree	20/22 (90.9)
Calcium/Phosphate/Alkaline Phosphatase/Magnesium		Agree	21/22 (95.5)
Vitamin D		Agree	22/23 (95.7)
Vitamin B12/folate, if at risk of specific micronutrient		Agree	21/23 (91.3)

Item	Level of evidence[a]	Median response	n/N (%)[b]
deficiency, eg, those on some anti-convulsant medications or less than the 25th centile on BMI, or those with symptoms suggestive of gastro-intestinal malabsorption			
Zinc		*Disagree*	22/23 (95.7)
Copper		*Disagree*	*20/22 (90.9)*
Selenium		*Disagree*	*20/22 (90.9)*
Vitamin A		*Disagree*	15/22 (68.2)
Vitamin E		*Disagree*	17/22 (77.2)
11. Clinicians should be aware of the potential for increased energy requirement in the presence of breath holding and hyperventilation[6]	2+	Agree	21/22 (95.5)

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[b] numerator is the number of responses with median response or 1 category either side and denominator is the number of clinicians in the panel whose expertise was relevant to this item

Table 3

Management to increase calorie intake

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Nutritional intake should be tailored to individual requirements whether the patient is under- or over-weight[26]	3	Strongly agree	25/25 (100)
2. The best indicator of caloric requirement can be obtained by comparing caloric intake to serial growth measures		Agree	19/19 (100)
3. If under-weight, planned energy requirements should be above required calorie intake until satisfactory weight has been attained[49]	3	Agree	21/21 (100)
4. Planned caloric intake should be above energy requirements until satisfactory weight [26–28]	3,3,2+	Agree	17/22 (77.2)
5. To increase calories in the diet for weight gain, give frequent snacks of high calorie nutritional supplements[51]	4	Agree	21/24 (87.5)
6. There is no published evidence that gluten free diets have been associated with clinically improved growth in Rett syndrome[10]	2	Agree	20/22 (90.9)
7. There is no published evidence that lactose free diets have been associated with clinically improved growth in Rett syndrome[10]	2	Agree	21/22 (95.5)

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Table 4

Management to reduce feeding difficulties

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Instructions on feeding techniques should be provided by an experienced nurse, dietician, speech or occupational therapist to all care staff concerned with feeding and should be determined by assessment findings[26]	3	Agree	25/25 (100)
2. Offer frequent small feeds throughout the day [14]	4	Agree	21/25 (84.0)
3. A trial of intensive therapy to optimise abilities to eat is justified in girls who do not eat as new skills can be gained by some girls[9]	3	Agree	18/22 (81.8)
4. Girls and women with Rett syndrome may have a sudden loss of eating skills associated with an acute illness or surgery and supportive treatment may be required until normal feeding returns[9]	3	Agree	23/23 (100)
5. Observation of feeding and barium video-fluoroscopy can inform decisions to modify the consistency of food[26]	3	Agree	23/24 (95.8)
6. If there are needs to provide physical positioning and postural support, support jaw and avoid hyperextension [29, 30, 52, 53]	2+,2+,2+,3	Agree	20/20 (100)
7. If there are needs for special equipment and utensil use, trial different models of utensils, plates, cups and feeding bottles[30]	2+	Agree	21/21 (100)
8. If there is need for prompting and socialising during mealtimes[32, 53]:	3		
Give the food of choice		Agree	18/20 (90.0)
Guided feeding with verbal and physical prompts for at least one meal a day can increase participation in eating		Agree	19/20 (95.0)
The carer needs to be oriented to the breathing pattern and feed when the rhythm of breathing is steady		Agree	20/20 (100)

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Table 5

Factors guiding consideration of enteral feeding

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Benefits of gastrostomy include [33, 46]	2+, 3		
Decreased number of feeding times		Agree	19/25 (76.0)
Shorter duration of mealtimes		Agree	24/25 (96.0)
Reduced vomiting and reflux		*Neither agree or disagree*	*22/25 (88.0)*
Reduced chest infection		Agree	24/25 (96.0)
Reduced constipation and pain		*Neither agree or disagree*	*23/24 (95.8)*
2. Gastrostomy should be considered in children with			
Failure to thrive despite efforts to increase the calorie intake		Strongly agree	20/20 (100)
Oromotor dysfunction causing unsafe swallow		Strongly agree	20/20 (100)
Unusually long feeding time with resultant stress to the carer and the girl or woman		Agree	19/20 (95.0)
3. Gastrostomy may also be associated with improved quality of life of caregivers[33]	2+	Agree	24/25 (96.0)
4. *Gastrostomy increases the risk of gastroesophageal reflux disease*[46]	3	*Neither agree or disagree*	*19/23 (82.6)*
5. Complications of gastrostomy include			
Leaking		Agree	17/17 (100)
Dislodgement of the gastrostomy tube		Agree	17/17 (100)
Increased risk of reflux		Agree	15/17 (88.2)
Migration of tube		Agree	15/17 (88.2)
Intestinal perforation		Agree	11/15 (73.3)
Bleeding		Agree	16/16 (100)
6.Gastrojejunostomy or PEG-J tube may be considered in uncontrolled gastroesophageal reflux		Agree	16/17 (94.1)
7. Gastrojejunostomy decreases the risk of aspiration compared with gastrostomy[54]	2+	Agree	14/17 (82.3)
8. The complications of gastrojejunostomy include			
Increased transit time of iquid with poor absorption of some nutrients		Agree	13/17 (76.5)
Dumping Syndrome if the flow is too fast		Agree	17/17 (100)
All the common complications as gastrostomy		Agree	17/17 (100)
9. Clinical monitoring following insertion of a gastrostomy or gastrojejunostomy include review of the following[17]	3		

Item	Level of evidence[a]	Median response	n/N (%)[b]
Nutritional assessment – intake and requirements		Strongly agree	25/25 (100)
General condition, appearance and energy levels		Strongly agree	25/25 (100)
Gastro-intestinal function – vomiting, reflux, bowel function, abdominal distension, pain		Strongly agree	25/25 (100)
Fluid status including feed, water flushes, oral diet and medicines		Strongly agree	25/25 (100)
Check position of feeding tube and health of stoma site with regard to infection, granulation and leakage		Strongly agree	25/25 (100)
Home management of equipment and any support needs		Strongly agree	24/25 (96.0)

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Table 6

The provision of enteral feeding

Item	Level of evidence[a]	Median response	n/N (%)[b]
1. Gastrostomy placement and aggressive nutritional therapy can favourably alter growth failure and under-nutrition in Rett syndrome irrespective of age [34, 35]	3, 3	Agree	23/23 (100)
2. Parents need additional practical and emotional support before and after the insertion of a gastrostomy tube[34]	3	Strongly agree	24/25 (96.0)
3. Gastrostomy alone is justified in children who do not have evidence of significant gastro oesophageal reflux		Agree	15/17 (88.2)
4. Fundoplication should be considered along with gastrostomy in the presence of gastroesophageal reflux disease that is not responsive to medical treatment	3	Strongly agree	17/17 (100)
5. Trial with a nasogastric tube prior to insertion of a gastrostomy tube could help in			
Determining the potential for weight gain and growth improvement		Agree	18/20 (90)
Correcting existing malnutrition if the parents are fearful of surgery/or tube use and unable to commit to a permanent procedure		Agree	20/20 (100)
Short term feeding difficulty whilst waiting for a gastrostomy or if acutely unwell		Agree	19/19 (100)
6. A gastrojejunostomy tube can be used where gastrostomy feeds are not tolerated [36, 37]	3	Agree	18/20 (90.0)
7. A long term gastrojejunostomy tube is indicated in patients who are poor candidates for anti-reflux surgery[15]	3	Agree	14/18 (77.8)
8. Disadvantages of gastro-jejunal feeding include:	3		
Their tendency to migrate back into the stomach needing repositioning under fluoroscopic guidance		Agree	18/18 (100)
Tendency for the tube to obstruct		Agree	17/18 (94.4)
The potential for the tube to act as a lead point for intussusception		*Neither agree or disagree*	*14/16 (87.5)*
The need for slow continuous feeds		Agree	17/18 (94.4)

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