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Cystic Fibrosis

Managing oral intake in children with Cystic Fibrosis and Cerebral Palsy

Introduction

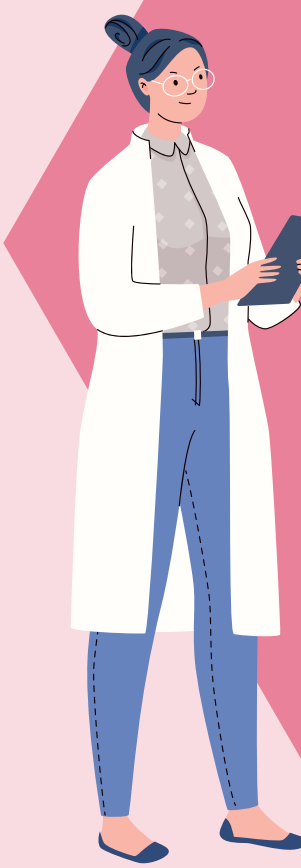
Cystic Fibrosis (CF) and neurological conditions are both life-long, often requiring intense treatment burdens. A concurrent neurological diagnosis can pose unique challenges in the clinical management of CF care.

CF has, historically, been associated with a substantial and advancing multisystem pathology. This comes with a burdensome treatment regimen, complex psychosocial challenges and reduced life-expectancy.[1]

For children with CF (cwCF) who also have a concurrent neurological diagnosis, such as Cerebral Palsy (CP), complex and overlapping challenges will present, especially respiratory health, mobility and nutritional care.

There has also been a transformative change in the management of many, though not all, people with CF (pwCF). Life expectancy is increasing, largely driven by the introduction of newborn screening (NBS), multidisciplinary care and nutritional support, along with rigorous screening and optimisation of comorbidities (e.g. diabetes).[2] More recently, the introduction of cystic fibrosis transmembrane conductance regulator modulators (CFTRm) has brought significant benefits, such as a significantly increased life expectancy, for those who are eligible, and are able to tolerate and access them.[1,3] Consequently, there is now more attention on the complexities of ageing.[4] Managing an ageing CF population along with a concurrent neurological diagnosis will pose new challenges and priorities for pwCF, their families and their healthcare teams.

In cwCF and CP (cwCF/CP) there are several areas with overlapping dietetic concerns, such as growth, nutritional requirements and gastro-intestinal (GI) issues. In this article, we are going to look at patient scenarios for cwCF/CP.



Children with CF and CP

Tables 1 and 2 demonstrate the key aspects of CF and CP.

Table 1: Key aspects of Cerebral Palsy

Causes: Results from abnormal brain development or damage, often during pregnancy, childbirth, or early infancy.
Symptoms: Spasticity, hypotonia, jerky or uncontrolled movements, poor coordination. A spectrum of respiratory, nutritional and digestive complications, including respiratory infections, aspiration, constipation, gastro-oesophageal reflux disease (GORD).
Types: Spastic (most common, stiff muscles), Dyskinetic (uncontrolled movements), Ataxic (balance issues), and mixed types.
Associated conditions: Motor issues, learning disabilities, epilepsy, hearing, vision, or speech difficulties.
Management: Multidisciplinary team (MDT) approach, including physiotherapy, occupational and speech therapy, dietetics. Medication and/or surgery to help manage symptoms and improve function and quality of life (QoL).

Table 2: Key aspects of Cystic Fibrosis

Causes: Genetic Mutation: Caused by mutations in the CF transmembrane conductance regulator (CFTR) gene on chromosome 7. Autosomal Recessive: Must inherit two copies of the defective gene, one from each parent.
Mechanism: The faulty gene causes the CFTR protein to malfunction, disrupting the movement of salt (chloride) and water in and out of cells, leading to thick, viscous mucus.
Symptoms: Most children are diagnosed via NBS, some may be diagnosed in later life. ***Respiratory:*** Persistent cough with thick mucus, wheezing, shortness of breath, frequent lung infections, nasal polyps, chronic sinusitis. ***Digestive/Nutritional:*** Majority are pancreatic insufficient (PI), requiring pancreatic enzyme therapy (PERT) to help treat and prevent steatorrhoea, chronic constipation, abdominal pain, poor weight gain and slow/faltering growth. Fat soluble vitamin supplementation also required.
Associated conditions: ***CF-Related Diabetes (CFRD):*** Damage to the pancreas reduces insulin production. ***Liver Disease:*** Blocked bile ducts leading to cirrhosis. ***Osteoporosis/Osteopenia***. ***Infertility:*** Affects most men (absent vas deferens) and reduces fertility in women.
Treatments: Although no cure, treatments have improved life expectancy. ***Airway Clearance:*** Techniques and devices (e.g. vibration vests, PEP masks) to loosen and cough up mucus. ***Medications:*** CFTRm: Targeting specific genetic mutations to improve protein function. Antibiotics, mucus thinners, bronchodilators. ***Nutritional Support:*** PERT, vitamin supplements (A, D, E, K). ***Lifestyle:*** Regular physical exercise.

Key considerations

Respiratory challenges

Children with CF and CP are at significant risk of severe respiratory infections. The combination of thick mucus in CF and impaired clearance mechanisms (due to motor deficits) in CP can lead to chronic lung infections. Repeated infections can increase energy requirements.

Aspiration risk

Serious respiratory illness in children with CP (cwCP) results from several contributing factors. Oropharyngeal dysphagia, GORD and seizures all increase the risk of aspiration, further complicating lung health. Aspiration can cause respiratory illness due to bacteria being present in the aspirate or chronic inflammatory responses in the lungs. Signs of aspiration include choking, coughing and changes to breathing and voice, but it can also be silent, which is detected using a video fluoroscopy.

Scoliosis is associated with severe gross motor impairment and other comorbidities, and is another risk factor because it reduces lung capacity.[5,13] If aspiration is suspected, it is essential that it is investigated and appropriate treatment initiated.

GI symptoms

Aside from dysphagia, common overlapping considerations in CF and CP are GI issues. In cwCF it's important to also consider potential PI, steatorrhea and abdominal pain. In cwCF/CP, the overarching priorities of supporting nutritional status, growth and QoL need to be carefully navigated. Up to 92% of children with CP have significant digestive issues which can reduce intake and further impact nutritional status, for example GORD, retching, vomiting and constipation.[18]

Figure 1 outlines the assessment and management of children with neurological impairment.

> In cwCF/CP, the overarching priorities of supporting nutritional status, growth and QoL need to be carefully navigated.

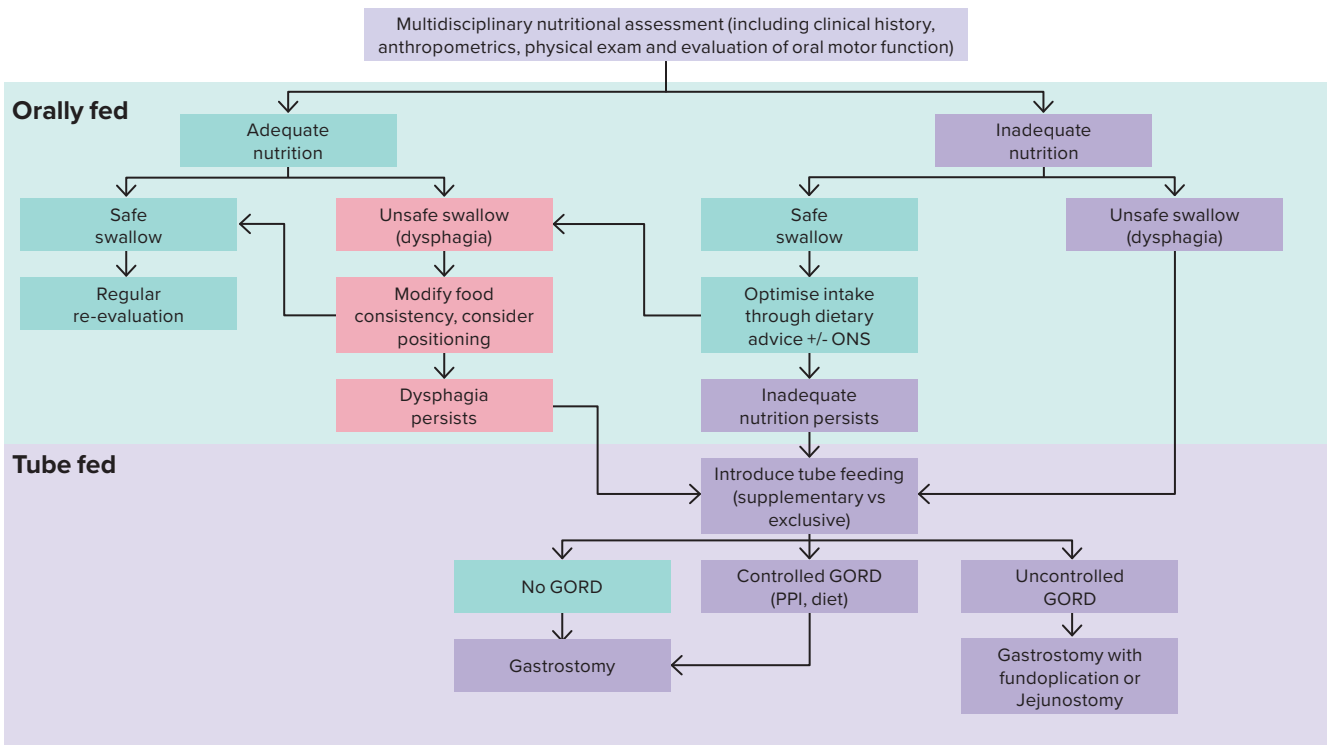


Figure 1 : Nutritional management of children with neurological impairment, adapted from Romano et al, 2019.[6]

GORD: Gastro-oesophageal Reflux Disease, ONS: Oral Nutritional Supplement, PPI: Proton Pump Inhibitor.

Growth assessment

If the child can stand, standard growth charts should be used, whereas measuring growth can be challenging if they are unable to stand. Segmental measurements are reliable and valid to obtain an estimated height, calculated from ulna length, knee height and tibial length.[7]

Assessment of undernutrition/malnutrition in cwCF/CP should not be based on CP-specific growth charts. CwCP often have a low body size (both height and weight) and so a different composition from neurologically developed children. According to ESPGHAN, signs of malnutrition include a weight for age that is 2 standard deviations below the mean and fat mass measurements (triceps skin fold, arm muscle area) below the 10th percentile.[6]

Body mass index (BMI) has long been identified as an independent predictor of mortality in CF and was endorsed for clinical evaluation in cwCF.[8,9] BMI became the defining standard, recommending children and young people (2 to 20 years) maintain a BMI at or above the 50th percentile. However, with improved treatments, CF is moving from a condition associated with undernutrition to one increasingly associated with appropriate nutritional status and overnutrition.[10] BMI does not distinguish between fat mass (FM) and fat free mass (FFM), or lean body mass (LBM), which reflect different health status - the focus is now on body composition.

Bio-electrical impedance (BIA) is recommended to assess body composition in cwCF and is non-invasive.[11] A study of cwCP has also highlighted that standard measures, like BMI, may not adequately reflect nutritional risk, especially in those with greater motor impairment.[12] A comprehensive assessment of body composition is recommended to guide individualised, function-based nutritional strategies.[12]

Assessment should also include measurement of bone mineral density and micronutrients, both routinely recommended in CF, as well as other signs of malnutrition (e.g. skin condition, pressure areas).[11]

Energy requirements

Many cwCP have decreased energy requirements compared to neurotypical children, and these differences increase with impairment severity. For example, wheelchair bound cwCP reportedly have 60-70% of the energy requirements of neurotypical children, but any physical activity should be considered.[13,25]

ESPGHAN recommends using dietary reference standards for typically developing children to estimate the energy requirement for cwCP.[6] As energy expenditure is based on basal metabolic rate, which is strongly influenced by body size and composition (especially FFM), this may overestimate energy needs due to the low weight and height of many cwCP and variation in FFM (muscle tone).

For cwCF, energy requirements were set at 120%-200% of the recommended energy intakes.[14] With the changing landscape in this new era of CFTRm, energy requirements have yet to be defined. Therefore, growth parameters are still the best method for assessment.

As a starting point, choose a method to calculate requirements. If growth is appropriate, this should be considered as a success. Growth should be monitored 3-6 monthly, depending on the age of the child; more frequently if it falters.

Protein requirements

There is no evidence to suggest that protein needs of cwCP differ. Protein requirements for typically developing children are therefore recommended.[6] If undernourished, additional protein (along with energy) may be required to promote 'catch-up' growth, e.g. up to 2.0 g/kg per day of protein and an additional 20% increase in energy intake.[15]

Micronutrients

In cwCF, ESPGHAN recommendations for micronutrients for CF should be followed.[11] CwCF/CP can be at risk of micronutrient deficiencies, particularly calcium, iron, zinc, vitamins C, D and E, and selenium,[18] especially if tube fed.[6] Starting with the requirements for typically developing children is recommended.[6]

Nutritional management

Both conditions can impact weight gain. CF causes digestive issues and nutrient malabsorption, while CP can involve feeding and swallowing difficulties and digestive issues, necessitating specialised diet plans.

There is no evidence to show whether optimising nutritional intake in young people with CP prevents respiratory illness. However, in people without CP, there is evidence that insufficient nutrition leads to weakened immune responses.[16]

If cwCP have poor nutritional status, dietitians should look to optimise nutritional intake whilst managing the risk of aspiration and digestive issues.[17]

Children with CF and neurological conditions will fall into one of several categories of nutritional support, depending on symptoms. Nutritional status, the ability to consume adequate quantities of food and fluids, and pulmonary aspiration risk all affect the type of nutritional support required. Impaired swallowing, poor co-ordination of breathing with swallowing during meals, and GORD all contribute to the requirement for nutritional support and impact the route of feeding. GI symptoms, including constipation, retching and vomiting, can further complicate management. The level of support required is dynamic, depending on how either condition develops. ESPGHAN has published a useful guide to support decision-making.[18]

Options include:

- Oral intake with food texture modifications
- Oral intake with food fortification and oral nutritional supplements (ONS)
- Oral intake plus enteral tube feeding (ETF)
- All nutrition by tube and nil by mouth (NBM).

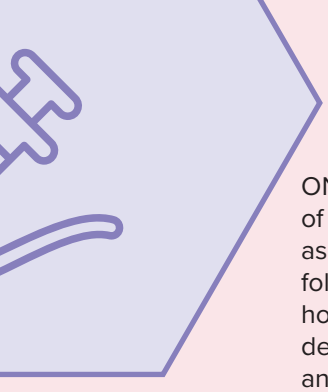
The first option, where possible, should involve oral nutritional support, aiming to increase the energy, protein and micronutrient content of the diet. The trial of oral intake will also depend on the child's age and severity of malnutrition.[6]

The decision to modify the diet and/or use of non-oral feeding methods is individual, depending on a wide range of factors. Firstly safety, with the risk of aspiration and respiratory disease discussed, allowing families to make a fully informed decision. Other factors for consideration are growth, weight gain and QoL.

Food texture modification needs to be assessed in conjunction with a speech and language therapist using the International Dysphagia Diet Standardisation Initiative (IDDSI) classification. As respiratory symptoms are common in cwCP, especially at mealtimes, it is important to consider the risk of respiratory illness associated with pulmonary aspiration regularly.[5]

If faltering growth is an issue, food fortification and ONS have an important role to play. As well as the standard ranges, other options include compact varieties for smaller volumes, higher density requirements and fibre options. Fibre recommendations for cwCP are in line with typically developing children.[18]

> Children with CF and CP face overlapping respiratory, nutritional and feeding challenges that demand a highly individualised MDT approach.



ONS can significantly contribute to the intake of micronutrients (and potentially fibre) as well as additional energy and protein. An initial follow-up in 1-3 months is usually sufficient; however, the length of time to trial an ONS depends on age and nutritional status. Infants and those with a poorer nutritional status will need to be reviewed more frequently.[19] If nutritional status continues to be of concern, ETF should be considered.

Nutritional goals are for optimal growth and nutritional status, but it can be a fine balancing act. For those who can eat but require supplementary tube feeding, allowing the child to eat what they want may provide enjoyment and add to QoL. However, food choices may not always be the most nutritious, leading to nutritional imbalances. For example, if less ETF is given and poor nutrient dense foods consumed, nutritional depletion may become an issue. If the full prescribed ETF is given and the child is allowed to eat high energy but poor nutrient dense foods, then excess weight gain may occur. Feeding should not be stressful for the child or family or take excessive amounts of time to the exclusion of other activities, e.g. a maximum of 30 mins per feed. If families are spending many hours a day trying to feed their child it can significantly impair QoL, and this is often when the option to tube feed is decided.[20]

Whatever route and regimen are used, families need to be supported with options to explore different feeding scenarios.

Considerations for ETF

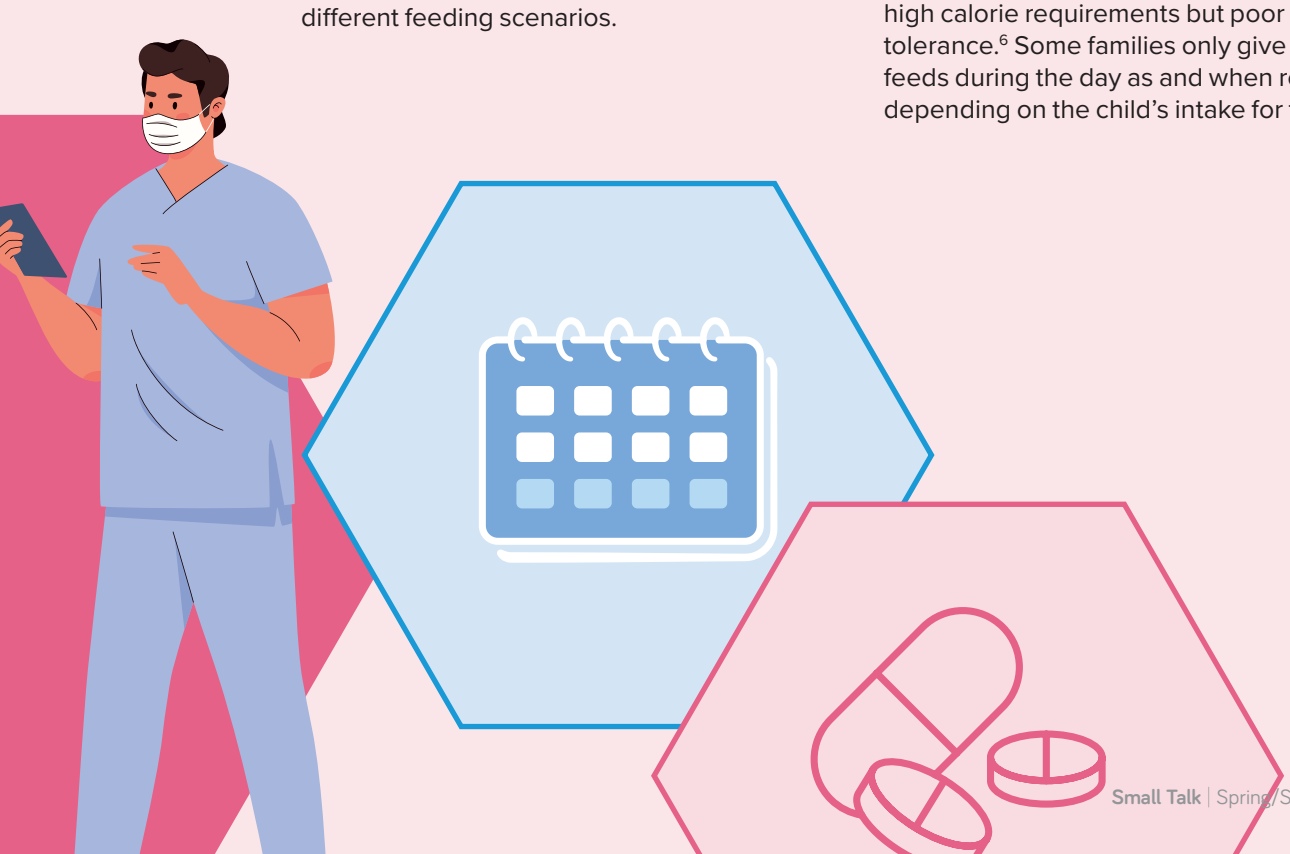
Indications for ETF include:[19,20]

- unable to meet nutritional requirements orally, despite oral nutritional support
- more severe undernutrition
- significant feeding and swallowing dysfunction (with a risk of pulmonary aspiration) or stressful oral feeding sessions/prolonged feeding times.

ETF may be used solely (for those with an unsafe swallow) or to supplement oral intake. The choice of access for ETF will depend on the clinical status of the child but is usually gastrostomy. Nasogastric tube feeding (NGT) should only be used acutely until a gastrostomy can be placed. Post-pyloric feeding, such as a jejunostomy tube, is another alternative if gastrostomy feeding is not tolerated, e.g. with GORD.

Methods of feed delivery

ETF can be administered in several ways: bolus, intermittently, or continuously, using an ETF pump. For example, some children may be able to eat during the day and have a top-up feed overnight or during the evening. Combination feeding is another option, with overnight continuous feeds and boluses during the day to provide sufficient nutrition. This method is suggested by ESPGHAN for children with high calorie requirements but poor volume tolerance.[6] Some families only give bolus feeds during the day as and when required, depending on the child's intake for that day.



Choice of feed

Many commercial enteral feeds are available, including polymeric, semi-elemental and elemental formulas, tailored for different age groups. They vary in energy density, fibre, macronutrient and micronutrient content, osmolarity and packaging.

For increased energy requirements or fluid restriction, a high-energy formula may be useful. Lower-energy formula feeds are available for reduced requirements.

Fibre-containing enteral feeds are recommended, especially if diarrhoea or constipation are experienced.[21] Feeds with dietary fibre have been shown to have potential beneficial effects for the prevention of both diarrhoea and constipation.[22] CwCF would also benefit from a fibre-containing feed, as it is well established that they can have low fibre intakes.[23] In cwCP, insufficient fluid intake may predispose to constipation, especially for those who tend to choke, dribble or vomit. Also, for children with masticatory and swallowing difficulties, a puréed diet may be insufficient to stimulate active peristalsis in the bowel.

> Managing both conditions can be demanding on families, highlighting the need for comprehensive support services.

Special considerations

Where scoliosis is present, especially in moderate to severe cases, this can press on the stomach and other digestive organs, as the unnatural spinal curve reduces torso space, leading to digestive discomfort, GORD, constipation, bloating and nausea. If volume tolerance is an issue, a higher energy formula may be preferred.

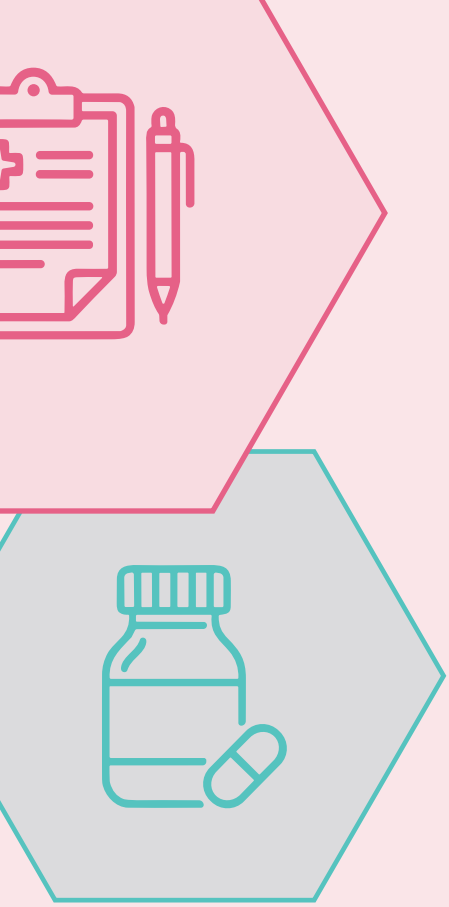
Blenderised food is increasingly being used by parents for their tube-dependent children. To date, there is little published evidence on the potential health benefits or risks of this practice and the best way to use it.[6] The child's needs should be carefully considered. Commercial feeds containing food-derived ingredients are available, providing a potential suitable alternative, so long as the foods used are not contraindicated.

Whey-based formulas may be beneficial in children with poor feed tolerance due to delayed gastric emptying and GORD.[18,24] Peptide-based formulas are often introduced when standard polymeric whole-protein feeds are not tolerated. These are also available in higher energy options.

Medium-Chain Triglyceride (MCT) feeds are indicated if there are malabsorption issues. As MCTs are absorbed directly into the portal vein, bypassing typical fat digestion, they require less PERT. This can be helpful in tube fed cwCF, especially if NBM, and the PERT must be administered via the tube.

Summary

Dietetic care for cwCP and CF requires an individualised approach to provide adequate macro and micronutrients, fibre and fluids, alongside regular assessment to help ensure optimal growth is achieved. A safe eating experience is essential with any impairment supported. This requires the MDT working with family members to ensure a collaborative approach. Managing both conditions can be demanding on families, highlighting the need for comprehensive support services.



References

1. Cuneo, et al. J Cyst Fibros. 2023;22(4):763-71.
2. George, et al. BMJ. 2011;342:d1008.
3. Yeh, et al. Handb Exp Pharmacol. 2024;283:219-47.
4. Frost FJ, et al. Eur Respir Rev. 2025;34(176):240261.
5. Blackmore, et al. Dev Med Child Neurol. 2016;58(7):780-1.
6. Romano, et al. J Pediatr Gastroenterol Nutr. 2017;65(2):242-64.
7. Stevenson. Arch Pediatr Adolesc Med. 1995;149:658-62.
8. Shepherd, et al. J Pediatr. 1986;109(5):788-94.
9. Borowitz, et al. J Pediatr Gastroenterol Nutr. 2002;35(3):246-59.
10. Szentpetery, et al. J Cyst Fibros. 2022;21(5): 777-83.
11. Wilschanski, et al. Clin Nutr. 2024;43(2):413-45.
12. Atar, et al. Disabil Rehabil. 2026;48(5):1450-62.
13. Stallings, et al. Am J Clin Nutr. 1996;64(4):627-34.
14. Turck, et al. Clin Nutr. 2016;35(3):557-77.
15. Kuperminc, et al. J Clin Nutr. 2013;67(Suppl 2): S21–S23.
16. Dos Santos, et al. Nutr Hosp. 2011;26(6):1322-7.
17. Gibson, et al. Dev Med Child Neurol. 2021;63(2):172-82.
18. ESPGHAN [Online]. 2019. https://espghan.info/files/Dietetic-Management-of-Children-with-Neurological-Impairments-NI.-ESPGHAN-Advice-Guide.-2019.-Ver1..pdf [Accessed March 2026].
19. ASPEN. J Parenter Enteral Nutr. 2002;26(1 Suppl):1SA–138SA (Erratum in J Parenter Enteral Nutr 2002;26:144).
20. Sullivan, et al. Dev Med Child Neurol. 2005;47(2):77-85.
21. Bischoff, et al. Clin Nutr. 2020;39(1):5-22.
22. Lalanne, et al. J Pediatr Gastroenterol Nutr. 2014;59(2):172-6.
23. Poulimeneas, et al. Nutrients. 2020;12(10):3126.
24. Brun, et al. Clin Nutr. 2012;31(1):108-12.

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