



DMD PATHFINDERS

NUTRITION GUIDE



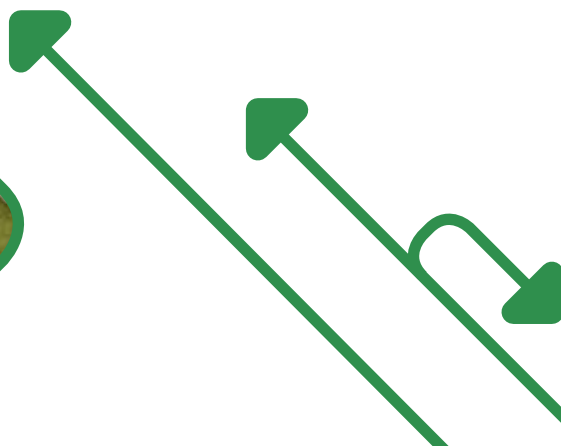


INTRODUCTION

Adults with DMD are living longer as a result of better management of the condition, in particular the use of respiratory interventions (ventilators) and medication to treat heart problems associated with Duchenne. In the UK, average life expectancy has increased to the mid to late 20s, but we also know of many adults living well into their 30s, 40s and 50s.

As clinics see more adults with DMD, a greater understanding is developing of how the condition affects adults, yet this lags behind what is known about children. While more research is being done in clinics, there is still a lack of scientific publications and comprehensive recommendations relating to adults with DMD.

Adults with DMD rely on peer support networks to navigate the complexities of adult life with Duchenne. As a user-led organisation of adults with DMD, DMD Pathfinders has a key role to play in supporting clinicians to develop and share expert recommendations, as well as connecting adults and helping them to share their experiences and learn from each other.





THE NUTRITION GUIDE

This guide provides information on nutrition for adults with DMD. We have worked with health professionals from around the world to pull together existing best practice and experiences of adults with DMD into a useful resource. User experiences were gathered from a survey of 91 adults and families, many of whom have volunteered to share their stories in this guide.

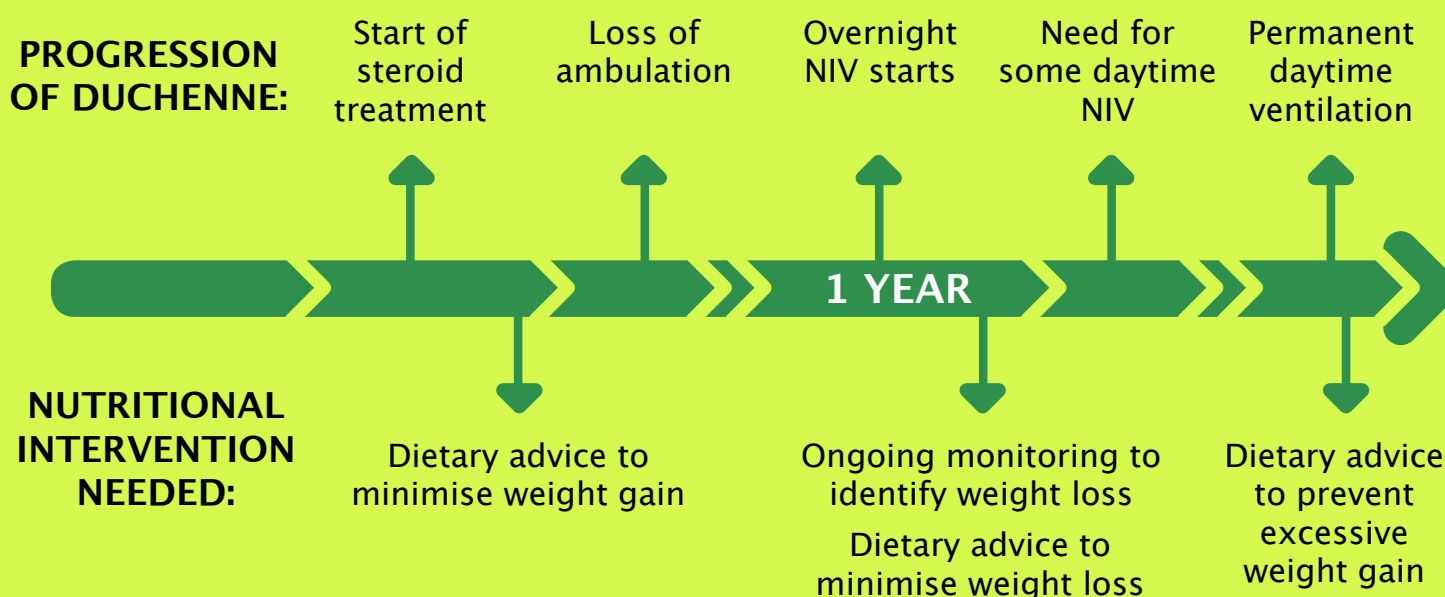
This guide looks at:

- The Nutrition Timeline
- A Healthy Diet
- Nutraceuticals and Dietary Supplements
- Maintaining a Healthy Weight
- A Good Blended Diet (Case Study)
- Eating with DMD
- Chewing & Swallowing Problems
- Ventilation & Nutrition
- Feeding Tubes
- Managing Bowels & Constipation
- Common Gastrointestinal Problems
- Nutrition Top Tips

As with all our guides, the information contained within is no substitute for attending regular appointments with health professionals who specialise in Duchenne. If you have any concerns or questions, we advise that you always consult a specialist health professional in the first instance.



THE NUTRITION TIMELINE



As Duchenne progresses, nutritional issues also change. During childhood, especially when steroids are used and at the time when children start using a wheelchair, there is a particular risk of obesity which can impede breathing and reduce muscle function and mobility. Dietary advice to assist with maintaining a healthy weight is required.

In teenage or adult years, as eating becomes more difficult and respiratory function declines, energy requirements may increase while food intake often declines, putting individuals at significant risk of undernutrition. Interventions to gain or maintain weight may be required, including dietary changes and if necessary the placement of gastrostomy tubes, to insert food directly into the stomach.

Difficulties with swallowing (dysphagia) also present problems for people with DMD and may require compensatory strategies to avoid life-threatening aspiration pneumonia. This may start in childhood but is not usually noticed until later in life.

Where dietary and swallowing interventions are successful, and adults become permanently ventilated, energy requirements decrease and once again individuals become at risk of becoming overweight.



A HEALTHY DIET

In general, adults with DMD should strive to have a healthy diet balanced in energy, protein, fluid, calcium, vitamin D, and other nutrients. Adults should aim to include a variety of foods from the five main food groups: fruit; vegetables; meat, poultry and alternatives; dairy and breads and cereals.

MICRO-NUTRIENTS

Adults are at risk of low bone mineral density as a result of not walking or standing, and sometimes due to the use of steroids. Recommended care guidelines are to take a daily multivitamin supplement with vitamin D and minerals if calorie intake is low. Additional vitamin D supplements may be required in the case of vitamin D deficiency (DMD Care Considerations Working Group, 2018). Regular monitoring of vitamin D levels is recommended.

PROTEIN

A dietary allowance of 0.95 g/kg bodyweight per day is recommended for those aged 4–13 years; 0.85 g/kg per day for those aged 14–18 years; and 0.80 g/kg per day for those aged 19 years or older.

FLUIDS

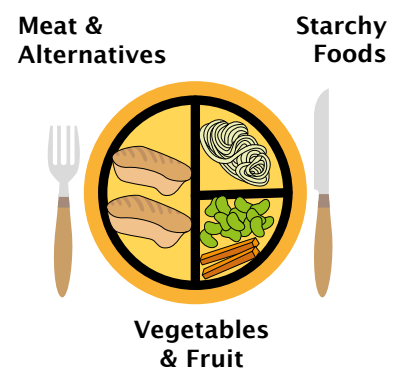
Recommended fluid intake in the care guidelines is based on weight, with 1500 mL + 20 mL for each kg over 20 kg (DMD Care Considerations Working Group, 2018)

OTHER CONSIDERATIONS

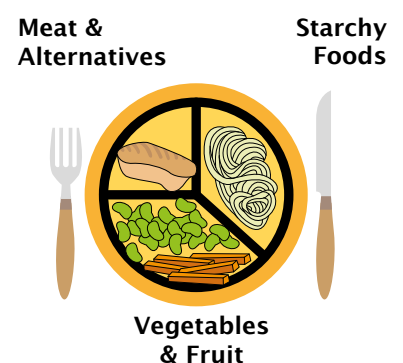
Adults with chewing and swallowing difficulties may start eating softer foods to ensure they are still eating enough. For those not supplementing their eating with a feeding tube, it is important to ensure modified diets remain well balanced. A dietitian can help with this.

Although there is no evidence to suggest specific dietary intolerances of people with DMD (e.g. to dairy or gluten), individuals with DMD may have their own specific dietary intolerances. Any changes in diet should be closely monitored to identify any specific problem issues.

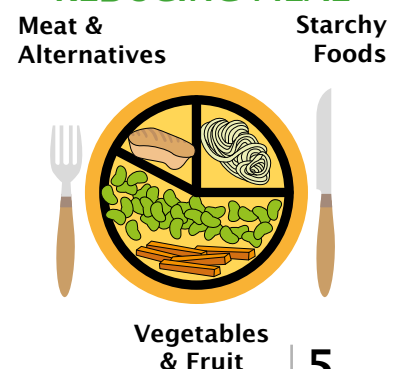
TRADITIONAL MEAL



HEALTHY 'BALANCED' MEAL



WEIGHT REDUCING MEAL





NUTRACEUTICALS AND DIETARY SUPPLEMENTS

TOP TIP

When deciding whether to use additional supplements, it is important to fully research the evidence available and talk to a specialist dietitian (where possible) to ensure you are aware of any risks.

There have been a range of nutritional supplements investigated in boys with DMD. The most promising supplement is creatine monohydrate. A systematic literature review (considered the highest level of evidence) demonstrated that creatine supplementation had some short to medium term benefits with regard to strength and muscle mass in Muscular Dystrophy (Davidson et. al., 2015).

This review included data from five clinical trials conducted in ambulatory boys with DMD. However, there is a lack of long term evidence for creatine in DMD, and no data that describes how it affects adults with DMD.

Some other supplements that have been investigated in clinical trials for boys with DMD include: glutamine, Coenzyme Q10, fish oil, leucine and carnitine (Davoodi et. al. 2012; Davidson et. al., 2015). There has not been strong evidence of an effect with these supplements nor is there any data for these supplements in adult populations.

There have been many supplements investigated in the DMD mouse model. Whilst there have been some interesting results in these studies, these findings do not always translate directly into humans. Caution is recommended when interpreting results from these studies.

DON'T BELIEVE EVERYTHING YOU READ

There are a wide range of unsubstantiated claims regarding specific nutrients and diets to treat many different conditions, including Duchenne. Claims have become increasingly sophisticated, often being promoted by medical professionals and linking to articles in journals. However, often these journals have no quality control or peer review process and those medical professionals may earn money from products they recommend.

Some supplements offer promise but haven't yet been subject to clinical trials to prove if they work. However, some supplements or specific diets have been studied and have not been found to work.

In researching suggested diets and supplements, it is important to identify the evidence base supporting their use (publications, clinical data, human trials, animal studies) and whether these are from a reputable source (e.g. a recognised specialist DMD clinic or peer-reviewed journal). Take care not to believe everything you read.



MAINTAINING A HEALTHY WEIGHT

Energy balance is an important concept for understanding weight management. The body uses energy all the time, even when at rest or asleep. Energy balance is when the energy obtained from diet (measured in calories) meets the energy that the body needs.

As an individual with DMD stops walking, energy needs often decrease making obesity a greater risk. As an individual experiences respiratory decline, energy needs increase making undernutrition a greater concern.

As adults switch to permanent ventilation, breathing difficulties decrease and nutritional requirements also decline. A study by Gonzalez-Bermejo in 2004 identified that adults with DMD who are on permanent ventilation had a lower resting energy expenditure, suggesting they need fewer calories. This explains why some people start to put on weight once they have 24/7 ventilation.

Energy balance can also change in the short term, as energy requirements are increased during illness and infection, after surgery and when breathing becomes more difficult.

OBESEITY

In children and some young adults, care must be taken to avoid obesity, particularly when using steroids which can be associated with increased appetite and weight gain. Weight gain can sometimes be an issue for older adults as well, where they are permanently ventilated and in good health.

Obesity causes problems with breathing and mobility as the body must work harder to use muscles, and it also makes it harder for moving and handling by family and carers. If necessary, weight reducing diets may be needed to restore a healthy weight.

It is important that a proactive approach is taken before a child or adult becomes overweight. It's easier, and healthier to prevent weight gain in the first place than to lose weight afterwards!

TOP TIP

If you have more calories than your body needs, you will gain weight, and if you have fewer calories than your body needs, you will lose weight. This means to maintain a healthy weight you need to be aware of both changes in the amount of calories in your diet as well as changes to your energy needs.

TOP TIP

You should consider using nutritional supplements or meal replacement drinks during periods of illness or other times when your energy requirements increase.

TOP TIP

Both obesity and under-nutrition are harmful! Be sure to get weighed regularly and adapt your diet as needed to maintain a healthy weight.

UNDER-NUTRITION

For older teenagers and adults, under-nutrition is a significant risk and individuals can quickly become underweight. This may be due to loss of appetite associated with respiratory problems, difficulties with chewing and swallowing or a result of increased nutritional requirements from increased work of breathing.

Under-nutrition adversely affects respiratory muscles, reducing muscle mass and strength. It additionally affects immune function, wound healing, sensitivity to oxygen (prolonging ventilator weaning) and psychosocial function.

It can often be challenging for adults and their families to recognise when they are becoming underweight, particularly if they are used to watching what they eat and keeping weight down. It may require a shift in mind-set.



From when I was 10 I was concerned about getting too large and about how that may affect my heart, so I basically cut fast food out altogether. When I was 17 my weight dropped quickly, but then for 10 years I was fairly stable. Then in November 2017 I had a bowel blockage and after that was resolved my weight was 22kg; I was pretty shocked! So now I'm determined to gain weight at this stage. I'm drinking resource drinks and porridge and dessert on top of my normal diet!

Scotty, Australia

TOP TIP

Where you are concerned about significant weight gain or weight loss, loss of appetite or difficulties with eating, it is important you get in touch with a dietitian and/or speech and language therapist.

WHAT DO I NEED TO LOOK OUT FOR?

In reviewing nutritional requirements, it is important to consider a number of factors:

Weight – where possible adults with DMD should be weighed regularly and attention should be paid to any unintentional weight changes over time. Weight is the best indicator of achieving energy balance so is important to monitor regularly, every 6 months if stable and more regularly if unwell. It is important to consider other signs of weight loss if it is difficult to get weighed regularly such as the fit of your clothes.

Nutritional intake/appetite – adults with DMD should monitor how much they are eating compared to normal, including snacks in between meals, and in particular how often meals are left unfinished.

Fatigue or difficulty with eating – this can result from increasing problems with chewing and swallowing or respiratory problems. This may be an early sign that weight loss could become a problem.

Changes in energy needs – If fighting an infection or starting to need use of the ventilator more during the day, adults with DMD are likely to need more energy and more calories than usual.

WHAT DOES BMI MEAN FOR ME?

BMI scales have been shown to be inadequate in diagnosing obesity in children with DMD (Pessolano et al. 2003). As DMD progresses in adulthood, the loss of muscle also makes it difficult to reliably assess healthy weight using BMI.

NICE guidelines (2017) define malnutrition as a BMI of less than 20 kg/m² and unintentional weight loss of greater than 5% within the last 3–6 months. Obesity is defined as more than 25 kg/m².

Using this scale, many older adults will be classified as underweight. For adults in this category, it is vital to access dietetic services to ensure appropriate advice can be given to prevent under-nutrition. However, even after these interventions, adults may struggle to reach a “normal” weight on a BMI scale when adequately nourished.

More research is needed to identify a DMD-adjusted BMI scale or similar tool which accounts for muscle loss in determining a healthy weight. For now, BMI guidelines can be a useful guideline but must be interpreted in the context of individual weight history, energy requirements and nutritional intake.

Where possible, adults with DMD should attend an adult clinic specialising in DMD which has access to a specialist dietitian or speech and language therapist to allow tailored advice and multidisciplinary management. If this is not possible, a referral to local dietetic and speech and language therapy services should be made.

HOW TO GET WEIGHED

Some clinics use a weighing scale which weighs the individual in their wheelchair and subtracts the weight of the wheelchair from this. Care must be taken to ensure consistency by removing any additional equipment, bags etc. on the wheelchair each time.

Where an individual gets a new wheelchair or makes modifications to it, it will be necessary to weigh the wheelchair separately the first time it is used on the scale, in order to compare previous weight history.

Some clinics and community teams use a weighing scale that attaches to a hoist/Hoyer lift and weighs the individual directly. These can also be purchased privately.

STRATEGIES TO LOSE WEIGHT

- Reduce any sugary drinks e.g. fizzy drinks, juice, alcohol. Focus on water as the main drink with some dairy (e.g. milk) for bone health.
- Reduce frequency of high calorie, low nutrient foods e.g. takeaways especially fried food, bakery products e.g. croissants, Danish pastries, muffins, crisps, chocolate
- Include low calorie snacks e.g. vegetable sticks (raw or steamed) with salsa, berries
- Include lean proteins at meals and snacks: e.g. tin tuna, boiled eggs, chicken
- Focus on having 2 servings of fruit and 5 servings of vegetables daily.
- Eat 3 regular, balanced meals a day and plan your meals for the weeks.

STRATEGIES TO GAIN WEIGHT

- Focus on increasing the nutrient density of foods rather than the amount of food you eat e.g. adding fats (butter, margarine, oil, cream) and protein (milk powder, full cream dairy, meat/chicken/fish, eggs, tofu)
- Eat more nutrient-dense snacks e.g. full fat dairy (yoghurt, cheese, custard), peanut butter on crackers or bread, sweet or savoury muffins
- Eat and drink more milky drinks/condensed soups e.g. flavoured milk
- You can be prescribed high calorie drinks and liquid supplements in addition to normal meals by your dietitian.

TOP TIP

It is important you are weighed more regularly if you are at risk of undernutrition. Once a year or once every six months may not be sufficient to pick up on significant and rapid weight loss which could put your life at risk. More regular weighing may require referral to a local dietetic service.



CASE STUDY: BATTLE THE BITE

Matt Roberts is 24 and living with DMD. Matt and his family have worked hard to adapt his diet to allow him to enjoy a varied and delicious range of foods, all pureed to enable easy swallowing. DMD Pathfinders caught up with Matt to find out more:

WHEN DID YOU START BLENDING YOUR FOOD?

About two and a half years ago.

WHAT ARE SOME OF YOUR FAVOURITE MEALS?

Chicken Casseroles, Curry, Roast Dinner, Leek and Potato Soup, Steak Diane

HOW WELL IS THE BLENDED DIET WORKING OUT FOR YOU?

Really well as it allows me to eat much more without getting as tired from lots of chewing. Before blending my food I lost weight and went to about 5 and a half stone, I'm now nearly 10 stone and I'm still really enjoying food.

WHAT KIND OF FOODS BLEND WELL, AND WHICH ONES DON'T BLEND WELL?

Ideally you need foods that are quite liquidy or meals with lots of sauce. Big bits of meat or vegetables will cause trouble with the blender so don't add too much

at once. Rice and noodles do not work very well, but beans will blend well. Soups are very good but things like bacon don't work well. Adding things like cream is good to increase calories.

DO YOU HAVE ANY OTHER TIPS FOR BLENDING FOOD?

Add stock to make foods easier to blend and try not to have too much meat as it can make it a bit too claggy (to use the technical term). Chopping things up a bit smaller as well might help. Putting the food through a sieve after blending normally gets rid of most lumps.

HAVE YOU TRIED NEW MEALS AND DIFFERENT FOODS SINCE YOU STARTED BLENDING?

Yes, lots of different soups but my parents usually make and try different recipes anyway so there are lots of different things.

DO YOU TAKE ANY FOOD SUPPLEMENTS AS WELL AS THE BLENDED FOOD?

I have done before and sometimes when I need to go out and don't have time to eat a meal. I lost loads of weight a few years ago and dietitians prescribed them to me but I no longer need them, they are just convenient calorie boosts now.

DO YOU EVER GET BORED OF BLENDED FOOD?

Sometimes yes. I do occasionally really crave a bacon sandwich,

or burger and chips, or rice-based dishes, but I do get plenty of variation with blended food. Also, I am still able to eat some non-blended food such as bread depending on the consistency. Ultimately, it is necessary to intake the correct amount to be healthy so I just get on with it as much as possible. I think certain textures are the thing I miss most. However with blending you still get great tastes and flavours but it's just in a slightly different form.



Chicken with Vermouth, Taragon and Cream



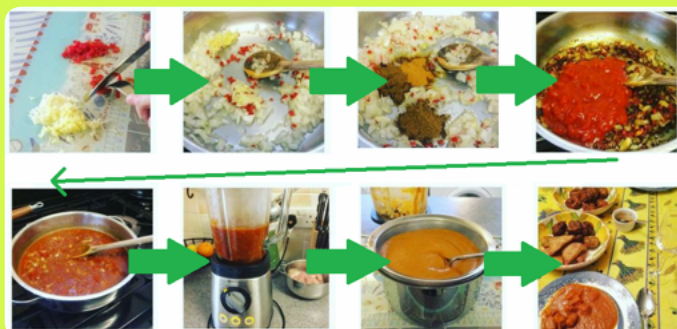
Steak Diane with Asparagus, Potatoes, Peppers and Tomatoes

Recipe

Chicken Tikka Masala

Fridays are for curries right? This chicken tikka masala is a household fave. The best thing about discovering good puréed recipes is that families can eat the same meal, together, regardless.

1. Fry onions, garlic, chilli & ginger
 2. Add chilli powder, tumeric & garam masala
 3. Add brown sugar, salt & pepper, tomato purée, tin of tomatoes & half can of water
 4. Cook on hob with lid on for 1hr
 5. Purée until smooth (for everyone eating)
 6. Add diced chicken, cook
 7. Purée and sieve for those who need it or leave as it is for those who don't.
- Done





EATING WITH DMD

As young people and adults with DMD lose upper body strength, they steadily require more support with eating and drinking. At first, this means needing help to cut up food. As upper body strength continues to deteriorate assistance is required with feeding either by a carer or other feeding aids. As adults become older, and respiratory needs increase, further adaptation may be required to enable eating and drinking whilst using a ventilator.

As well as reduced physical strength, chewing and swallowing becomes significantly more difficult. Adults frequently experience additional problems with their jaw meaning that they can't open their mouths wide enough to eat certain foods unless chopped finely or pureed.

If swallowing is very impaired, some adults may become unable to eat by mouth and require nutrients solely through a feeding tube. Most adults do however manage to eat and drink (even if only in small quantities) well into adulthood, either with or without a feeding tube. This is likely to require adaptations to meals.

According to our survey, 88% of respondents (including feeding tube users) were able to eat. A feeding tube does not prevent an individual from eating and drinking by mouth. It can relieve the pressure on meal times and make food enjoyable again.

There is no one size fits all approach to managing difficulties associated with eating. Sometimes a feeding tube will be necessary, in others it may be optional. A neuromuscular dietitian and speech and language therapist will be able to advise you of the different options available to you.



I use a spork – a cross between spoon and fork – and a knife / fork combo – both with a chunky handle and very light weight. The blade is curved so it's easier to rock to cut food (rather than sawing). They are good if someone is using a neater eater or neater arm support – as you only need one hand. I also have a plate with sides which is great for pushing food to the edges then trapping it against the fork, or spoon. It's quite heavy which makes it stay put (especially if you stand it on anti slip mat) which helps when eating with one hand.

Josh, UK

EQUIPMENT TO MAKE EATING EASIER



The Neater Eater – an electronic machine which can be controlled by the user to pick up food from a plate and move it to their mouth, enabling independent feeding.



Plate Guards – as upper body strength deteriorates, it is often easier to eat with one hand (often supported by the other hand). Eating one-handed can make it difficult to pick up food on a fork without pushing it round your plate. Plate guards provide a surface against which food can be pushed onto a fork/spoon easily.



Specialist Cutlery – the weight of cutlery can often be a problem for adults who are still able to feed themselves. Lightweight cutlery can make a big difference.



Blenders – the essential tool for adults on blended diets. It's possible to blend pretty much everything, but we can't promise you will always love the results! A bit of experimentation is needed to find out what's nice and what isn't.



Straws – these come in all shapes and sizes. Standard straight or bendy straws usually serve the basic purpose, while extra-long straws can provide the extra reach necessary to reach a drink without moving. Wide straws can also be useful for sucking up thicker liquids and chunkier blended foods. If you're watching your environmental impact, silicone straws offer a reusable alternative that works for hot drinks and can still bend.

TOP TIP

Take your time to eat and don't let anyone rush you. Allow yourself plenty of time to eat. Consider eating smaller more frequent meals throughout the day if eating a large dinner exhausts you. Eating well is an essential part of maintaining your health and must be prioritised.

THE IMPORTANCE OF POSITIONING

Positioning can make all the difference to eating, as a good position makes it easier for swallowing solid foods. Where possible, adults should try to sit as upright as possible when eating.

TEETH & JAW PROBLEMS

The progression of Duchenne can have a significant impact for the development of the jaw and teeth. This is due to both muscle wastage and enlargement of the tongue, resulting in a number of different problems (Eckardt and Harzer 1996).

MALOCCLUSION: OPEN BITE (ANTERIOR/POSTERIOR), CROSS BITES

This describes the problem whereby teeth do not meet, either at the front or rear of the mouth. This can be common in adults with DMD.

RESTRICTED MOUTH OPENING

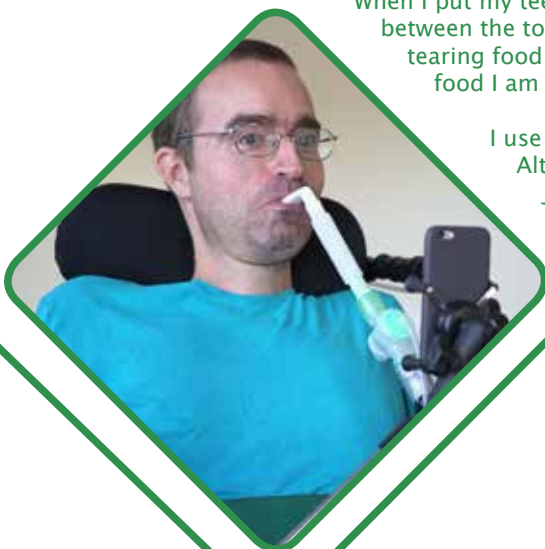
As muscles deteriorate, the jaw, like any other joint, develops contractures which can limit how wide adults with DMD can open their mouth (Symons et. al. 2002). A restricted mouth opening may hamper feeding, oral hygiene and dental care. Some adults report good results from using a jaw stretching system such as Therabite and Orastretch to stretch daily and improve or maintain jaw opening. At the moment there is little research to guide management of restricted jaw opening.

STRESS AND ANXIETY

As eating becomes more difficult, mealtimes can become a source of stress and anxiety. People worry about taking too long to eat, not eating enough or not being able to swallow food properly. It is important that mealtimes are given priority and stress is minimised as much as possible.

When I put my teeth together they don't meet at the front, leaving a big gap between the top and bottom teeth. This can make it difficult for biting or tearing food with my teeth. I always ask my PAs to cut up or break apart any food I am eating to make it easier.

I use a jaw stretcher daily to prevent my jaw from contracting. Although it looks scary, it's really just like stretching any other joint. I've improved my passive jaw opening from 25mm to 40mm which has really helped with eating and cleaning teeth.



Jon, UK



CHEWING AND SWALLOWING PROBLEMS

When we chew, a complex set of muscles operate our jaw to break up our food. In adult DMD, chewing is impaired mainly by an increasing deterioration of the masseter muscle, as well as dental changes such as open bites and cross bites which prevent the upper and lower teeth from meeting.

As chewing gets more difficult, adults with DMD sometimes attempt to swallow larger pieces of food that haven't been fully chewed, making swallowing even more difficult. The greater effort of chewing also causes fatigue, causing people to stop eating before they are full, reducing calorie intake.

Once food is chewed, additional sets of muscles act in order to swallow the food. In DMD, weakness of the tongue and throat (pharyngeal) muscles make it harder to push food through the throat towards the stomach. This means greater effort is required to swallow. If the muscles are very weak, the muscles do not properly close the airway whilst swallowing. This can result in small quantities of food and drink entering the airway. This is known as aspiration.

WHAT HAPPENS WHEN SWALLOWING FAILS

Chewing and swallowing problems can occur with various levels of severity. At the very least, food can be difficult to chew and food gets stuck in the mouth. If throat muscles are also weak, food residues can also get stuck in the throat. These residues can be inhaled into the airway during or after a meal. If food or fluid do enter the airway and are not coughed out, chest infections can occur (known as aspiration pneumonia). There may also be a risk of choking if food blocks the airway.

Chest infections pose a very significant risk to adults with DMD. Frequent episodes of aspiration pneumonia indicate the need to consider gastrostomy to reduce risks of infection from weakened swallowing.

TOP TIP

Try to reflect back up to one year ago (try to think of a significant event where you had a meal) to consider whether you show any new signs of chewing or swallowing difficulty. Ask your family or care team if they have noticed any changes too.

TOP TIP

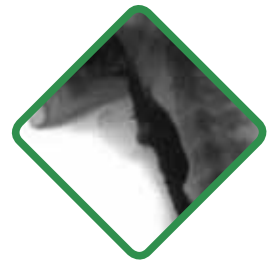
Speech and language therapists don't only help people with speech problems. They are also specialists in chewing and swallowing problems and are essential in the management of Duchenne for all adults.

Not everyone with DMD develops swallowing problems in the same way. Some may experience issues swallowing certain foods or with food getting stuck in the throat, without necessarily experiencing aspiration.

If food residues are still present in the throat after eating and the adult with DMD is not able to detect them, food and drink can enter the lungs without a person being aware (known as silent aspiration). No coughing or choking occurs. Research presented at the UK Swallowing Research Group (UKSRG, 2016) has shown a reduced sensory perception of food residues and aspiration for some adults with DMD. This means adults may be unaware that they are not swallowing effectively and that aspiration is occurring, leading to unexpected or unexplained chest infections.

Some symptoms that suggest a swallowing assessment may be necessary include:

- Increased or extended mealtimes
- Unintentional weight loss
- Prolonged chewing or difficulty in chewing
- Coughing (or attempts to cough) during and/or after meals
- Loss of appetite
- Difficulty moving food around the mouth
- A feeling of food or drink getting stuck in the mouth or throat
- Needing to adapt or avoid certain foods
- Starting to choose softer foods
- Needing to drink with meals
- New or increased number of chest infections



Some of the symptoms listed above, such as weight loss and increased chest infections, might also be caused by respiratory problems. It is therefore important that both swallowing and respiratory assessments are conducted in order to identify the most important issue.

ASSESSING SWALLOWING PROBLEMS

A videofluoroscopic (VFS) study of swallowing (also referred to as a modified barium swallow), requires the adult to swallow a liquid barium solution in front of an x-ray machine. A range of foods and fluids coated in barium are then given to the adult to swallow. The x-ray images help to identify which muscles are changing, assess which (if any) foods and fluids stick in the throat and are at risk of entering the airway. The assessment also helps the Speech and Language Therapist to find the most useful strategies and management advice to keep swallowing as safe as possible.

A Fibreoptic Endoscopic Evaluation of Swallowing (FEES) involves a small camera being passed through the nose and into the throat on a thin flexible tube. This assessment observes what happens internally during swallowing. Similar to VFS, it shows which muscles are changing and whether food and drink sticks in the throat or enters the airway. The benefit of FEES over VFS is its ability to assess saliva and secretion management. However unlike VFS, it cannot visualise the muscles of the mouth, only the throat.

STRATEGIES TO MANAGE SWALLOWING PROBLEMS

Management of swallowing problems should aim to prevent pneumonia from aspiration of food and drink and reduce weight loss caused by swallowing difficulties. It should be based on clinical assessments and the experiences and preferences of the adult with DMD.

Experiencing problems with swallowing doesn't always mean that the adult with DMD will need a gastrostomy/peg tube. There are a number of strategies that can be tried before this becomes necessary.

Modifications to the texture of the diet may be advised. This may help overcome difficulties with chewing and food sticking in the throat. In some cases adults find sipping drinks during and after meals helps to clear the throat. Other swallowing strategies may include:

- taking time to chew food as much as possible to make swallowing easier
- selecting softer foods such as chicken, minced meat, pureed vegetables, mashed potato etc.
- avoiding the toughest foods such as steak, leafy or stringy salad or crunchy vegetables – consider blending these in a food processor/ blender instead
- if continuing to have problems consider switching to a fully mashed, blended or liquidised diet. This can be supplemented by meal replacement drinks.
- avoid talking whilst eating as this makes it more likely that you will inhale any residues in your throat
- try to sit as upright as possible during and after your meal to ensure food goes down properly
- avoid using cough assist or ventilation immediately after meals where possible (but remember this may become necessary as your breathing declines and should be managed on an individual basis)

While a gastrostomy/peg tube may not be needed straightaway, it is important to think carefully about whether you would consider a feeding tube in the future, and to learn about what is involved.

It is important that strategies to aid nutrition and swallowing are part of a multidisciplinary approach to your care. Strategies such as adapting diets may impact on other areas, for example thickened drinks recommended by a dietitian to boost calories may be harder to swallow and should be used in consultation with a speech and language therapist.



My diet consists of several nutritional shakes, strained soups, cold juices and occasionally sauces on smooth mashed potato, this gives me a good variety of tastes along with my daily nutritional requirements. Most days I will have two scandishakes along with one complan, I will top this up with soup, sometimes meritine which is also a nutritional supplement, or just a normal soup.

Daniel, UK



VENTILATION AND NUTRITION

Ventilation is intrinsically connected to nutrition and swallowing. Inadequate ventilation can have a negative impact on nutrition, while weight loss and undernutrition can make breathing difficulties worse and can increase your vulnerability to infection.

It is recommended that every adult with DMD sees a respiratory specialist at least once every six months to monitor their respiratory needs. As adults with DMD get older, it is typical to increase their use of the ventilator during the day as their respiratory function gets worse. For some people, this may be a sudden change after a chest infection, while for others this can be more gradual. During this transition phase it is often difficult to reliably assess how many hours you should use the ventilator during the daytime.

When your daytime breathing is affected by muscle weakness, your carbon dioxide levels build up, which can cause a loss of energy and appetite. This usually happens before you even notice any difficulty breathing, and over time can lead to unintentional weight loss. This is why being weighed regularly is so important.

Where breathing problems are causing a loss of energy, using a ventilator 30 minutes to an hour before a meal, may help improve your appetite and reduce fatigue during meals. A small research study by Gargulio et al., (2016) has also found improved breathing and swallowing patterns when eating and drinking on a ventilator rather than off the ventilator.



I generally have the mask off and eat a few mouthfuls, then after I've swallowed have the mask placed back on my face and repeat, having someone with good timing holding the mask is essential, if somebody shoves it on as you are swallowing, the food gets blown down your airway and you have a nasty choking session!

Daniel, UK

EATING WITH A VENTILATOR

If necessary, it is possible to eat with a ventilator, as many of our members do, both with non-invasive ventilation and tracheostomy. Sometimes it can take a little while to learn how to coordinate breathing and swallowing effectively, or to find the best ventilator mask.

The use of a ventilator during and after meals needs to be considered carefully. The use of ventilation can improve the swallowing function of an adult with DMD, as they are no longer struggling with breathing. However, poor coordination of breathing and swallowing, often exacerbated by an unexpected ventilator breath, can cause choking episodes. It is important that ventilator settings are properly configured by a respiratory technician to allow the adult to trigger a breath when they are ready. Some adults do struggle to eat and drink on a ventilator, and in the early stages of ventilator set-up it is not advised.



I use a ResMed AirFit N10 nasal mask. I manage quite well when eating, I try to eat soft food and drink to help it go down and I swallow between breaths. It took me a while to get used to doing that.

Joe, UK

Before getting the daytime ventilator I was getting exhausted after only a few bites of food and now I'm not. When I first got it, I was expecting [eating with a ventilator] to be difficult. It was actually pretty simple once I did it. I usually chew and swallow a bite then take 1 or 2 breaths and then repeat. It is important to make sure you swallow everything before taking a breath. I almost aspirated once that way but the cough assist helped get the food out.



Milan, US



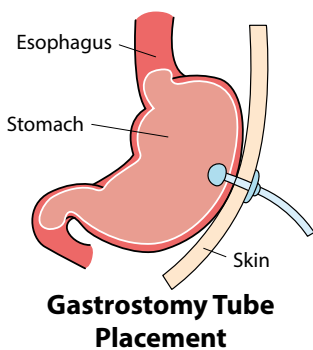
FEEDING TUBES

WHAT IS A FEEDING TUBE AND WHY MIGHT YOU NEED ONE?

A feeding tube delivers a nutrient feed directly into the stomach, allowing an adult with DMD to meet their nutritional needs without having to chew and swallow food.

There are three main reasons why adults with DMD may require a feeding tube:

1. Recurrent chest infections from aspiration (food getting into the lungs due to swallowing difficulties)
2. Under-nutrition and weight loss which is not responding to swallowing and nutrition strategies
3. Patient preference – where a patient opts to have a feeding tube before severe problems develop (e.g. where they are finding eating more difficult or are at risk of aspiration or malnutrition)



DIFFERENT TYPES OF FEEDING TUBE

Feeding tubes come in three different varieties:

1. A nasogastric tube which goes up the nose and down the throat to the stomach. These tubes are typically only used on a temporary basis, for example to provide necessary nutrients while fighting an infection, or while waiting for a more permanent feeding tube.
2. A gastrostomy tube is a tube that goes through the skin directly into the stomach (via a hole or “stoma” in your skin). These can either be a percutaneous endoscopic gastrostomy (PEG) or a radiologically inserted gastrostomy (RIG), which refers to how they are inserted:
 - A PEG tube involves an endoscope (camera) being inserted via the mouth, down into the stomach, to locate an insertion site for the tube. An incision is made in the skin and the PEG tube is passed into the mouth, down through the stomach and out through the incision, where it is secured in place. A PEG is usually the preferred method of tube insertion when possible.
 - A RIG tube uses X-rays to locate the insertion site for the tube. An incision is made in the skin and the tube is passed through



When I got my feeding tube my doctor explained the procedure to me and I had it done two weeks later. Honestly, the pain was a lot less than I expected. Without it there is no way I could eat enough by mouth and it's really great for taking medicine. In 10 years I've never had any problems with it.

Tim, us

from the outside and secured in place. RIGs may be required where an endoscope/PEG tube cannot safely be passed down the throat.

3. A percutaneous endoscopic jejunostomy (PEJ) or jejunostomy (JEJ tube) goes through the skin directly into the jejunum, the beginning of the small intestine. This may be required if an individual has problems feeding into their stomach.

CONSIDERATIONS FOR GASTROSTOMY PROCEDURE

Gastrostomy insertion should ideally take place in a multidisciplinary specialist centre with a specialist gastroenterologist or radiologist experienced with patients with DMD. The decision to insert a tube is a joint one between the patient and their multidisciplinary team.

Anaesthetics used for the procedure depend on local practice. They may involve local or general anaesthetic or sedation delivered via IV.

An echocardiogram and electrocardiogram should be done before any general anaesthesia. They should also be done before any conscious sedation, regional anaesthesia or local anaesthesia if previous echocardiograms have shown reduced cardiac function or were undertaken over one year previously.

If an individual is on steroids, consideration needs to be given to steroid cover over the period of surgery.

The following drugs should always be avoided during any procedure:

- Inhalational anaesthetic agents, such as halothane and isoflurane. If anaesthetic is required it should be delivered exclusively by intravenous injection due to rhabdomyolysis. Rhabdomyolysis complications are frequently confused with malignant hyperthermia
- Depolarising muscle relaxants, such as suxamethonium chloride, must not be used due to the risk of fatal reactions.
- Postoperative anticoagulation with heparin and/or aspirin is inappropriate and should be stopped pre-procedure.

Non-invasively assisted ventilation and the use of a cough assist is recommended during and after procedure for patients with significant respiratory-muscle weakness. Adults with DMD not currently using NIV or cough assist should be trained in their use prior to any surgery.

(DMD Care Considerations Working Group, 2018)



I definitely want to eat as long as I can by mouth by any means necessary, since I really love and enjoy not just eating, but food in general and love cooking with the help of my PCAs. I'm a foodie. But if my health declines to the point that I can no longer safely eat by mouth, then I would want a feeding tube, because I'm a huge fan of staying alive.

Petr Wronski

TOP TIP

It can be useful to tape the tube to the skin to keep it out of the way, but be sure to use dressings and tape that are not going to irritate your skin.



In my routine, I clean the tube site twice a day, in the morning during my bath/shower with soap and water, and then in the evening with saline water and gauze. To hold the tube in place and protect it, I use two circular cotton wool pads as a small dressing, with an incision to the centre. This is placed around the tube to protect the skin in this area. Gauze is then used as padding underneath the tube and secured with Hypafix tape.

Tyran,
UK

HOW TO USE A FEEDING TUBE

What does it look like?

There are different types of gastrostomy tubes. Some such as low-profile devices sit flush with the stomach. However, gastrostomy tubes are discreet and are not noticeable under the clothes

Eating with a feeding tube

Getting a feeding tube doesn't necessarily mean that adults with DMD won't be able to eat or drink normally. Many adults use a feeding tube to get most of their nutrition and make sure they maintain a healthy weight, but still eat and drink for comfort or when they feel like it. Having a feeding tube removes the pressure on the adult to always make sure they are eating enough. Other adults only use the feeding tube when they are ill or for taking medication.

Using the feeding tube

Specially prepared liquid feeds, which contain all the nutrients the adult needs, are fed into the stomach using the feeding tube. There are different options as to how and when the feed is administered depending on the individual's preferences.

Some people prefer the feed to be delivered by a pump, which can be done overnight or during the day, at different speeds depending on what is best tolerated.

Some people prefer the feed to be delivered at regular mealtimes, using a syringe to insert the food in portions throughout the day. This is known as bolus feeding.

Are there different types of feed?

Standard feeds are designed to meet the complete nutritional requirements of an adult. Many different companies make feeds so there can be small variations in products depending on the company; but essentially feeds are very similar. There are also specialised feeds to meet different nutritional needs e.g. to provide more calories in less fluid, or with increased fibre.

Taking care of your feeding tube

It is important to keep the feeding tube site clean at all times, to avoid any infections and to avoid excess granulation tissue (tissue that forms on wound surfaces during the healing process). It can be useful to rotate the tube a couple of times each day to prevent it sticking to the stomach wall.

The site needs to be kept dry, after being cleaned, to prevent soreness, redness and becoming itchy. Occasional mild dampness on the skin is normal, but if heavily or regularly soiled this should be mentioned to a healthcare professional.

Care must be taken to prevent the feeding tube from being caught or pulled during hoisting and other movements (e.g. moving & handling, physio, sex).

If you have a RIG tube, this is held in place by an inflated balloon of water, so a little more care might be needed. If you have a PEG tube, this is secured by a button inside the stomach, and is rarely dislodged, but pulling the tube could still cause trauma and/or bleeding.



What can go wrong and how to deal with it

It is essential to have a plan in place with your nutrition team if the tube accidentally falls out or migrates. If the tube falls out, the stoma will close within a few hours so it is important to act quickly. If it is a bumper-retained tube, if the tract is kept open, a local hospital can change the tube without needing a procedure. However, if it is a balloon-retained gastrostomy (i.e. held in place with a water filled balloon), a spare balloon tube should be kept at home as these are changed regularly at home.



Before using a silicone g-tube I used flimsy rubber ones and had a lot of problems with clogging. Since I've been using silicone for the last several years it hasn't clogged even once.

Pacien, us

It is not unusual for you to experience some slight seeping around the PEG site, even on a good day. Normally anything spilt on your skin can be easily wiped off without causing much irritation, however when the fluid is slightly corrosive and continually leaking in small amounts it can cause some irritation. In my experience it is a bit of a nightmare to treat this after it begins to get sore, what I find helps is to get a decent barrier cream applied to my skin on a morning, night or whenever it begins to feel itchy.



Robert, uk

I use the Peristeen anal irrigation system that uses water to empty out the bowel, inserted into the rectum using a rectal catheter whilst sitting on the toilet. The water stimulates the bowel and flushes out the stool, leaving the lower half of the bowel empty. My carers received training in the system from a registered nurse and the system is prescribed by my doctor. It took me a while to get used to, but once I got used to it I found it very useful and effective, as it relieves the strain and minimises the strength required when going to the toilet.



Mithun, uk



MANAGING BOWELS AND CONSTIPATION

Constipation is a common symptom for both children and adults with DMD and can be very uncomfortable and dangerous if untreated (Kraus et. al. 2016).

Although comparatively less studied than the muscles involved in chewing and swallowing, muscles involved in defecating (pooping/ pooping) can also be severely affected by Duchenne. It has been shown that food and stool takes longer to pass through the gastrointestinal tract in adults with DMD compared to healthy adults (Cascio and Goetze et. Al. 2016).

Adequate intake of fluids and fibre can often prevent constipation from occurring, but where this is not successful, it is important that constipation is treated promptly and regularly.

Stool softeners, laxatives, and stimulants can be used to deal with constipation and prevent it from occurring:

- Osmotic laxatives work by increasing the water content of your stool to make it easier to pass. These include medications such as Movicol, Macrogol, Lactulose, etc. These are often prescribed for long-term use in adults with DMD and can be effective in treating severe constipation/faecal impaction.
- Hyperosmotic laxatives such as glycerin suppositories and enemas similarly work by attracting water into the bowels to make stools softer. These are inserted up the rectum and have a typically more immediate effect, but are not recommended for long-term use due to the potential for loss of nutrients (e.g. bicarbonate) from the bowel.
- Stimulant laxatives such as dulcolax and senna stimulate the muscles around the gut to move your stool through your intestine. It is unknown whether the benefit of these laxatives is reduced in adults with DMD due to their reduced muscle function.
- Bulk forming laxatives are fibre supplements which increase the fluid content and bulk of stools. However, these can worsen symptoms if fluid intake is not increased and are not very useful.



To manage constipation I have fruit, veg and fruit juice daily, and usually use one sachet of Movicol every 2nd day, and every day if becoming more difficult. It works very well, and I rarely have problems.

Mark, UK



COMMON GASTRO-INTESTINAL PROBLEMS

GASTROESOPHAGEAL REFLUX

Gastroesophageal reflux (commonly known as Acid reflux), is where the tube leading from the mouth into the stomach does not close properly, allowing stomach acid to travel up towards the throat, causing irritation and burning. Gastroesophageal reflux is also known as GORD or GERD when it occurs regularly.

DMD patients are more likely to suffer gastroesophageal reflux and esophagitis due to weaker pharyngeal and hypopharyngeal muscles.

GORD is also a common issue where people take oral steroids (prednisone, deflazacort).

TREATING REFLUX

GORD/GERD can be treated by:

- Changing your diet (avoiding coffee, alcohol, chocolate, and fatty or spicy foods)
- Consider eating smaller, more frequent meals
- Avoid lying flat immediately after meals
- Try using an over-the-counter antacid to neutralise the stomach acid
- Proton-pump inhibitors or H2 receptor antagonists can be prescribed by a doctor to block stomach acid. The benefits of proton-pump inhibitors must be balanced against potential risks, including a higher incidence of community-acquired pneumonia, chronic kidney disease, and bone fracture (Lazarus, Chen & Wilson, 2016; Lambert et. Al. 2015)"

GASTROPARESIS

As skeletal muscle weakness progresses in individuals with DMD, a delay in emptying of the stomach (gastroparesis) can occur. This can cause abdominal pain, nausea, vomiting, a feeling of being full, and loss of appetite. Gastroparesis can be treated by modifying the diet, medication or in some cases a gastrojejunal feeding tube. (DMD Care Considerations Working Group, 2018).

GASTRIC AND INTESTINAL DILATATION

This is where the stomach and/or intestine becomes enlarged and can be common in adults with DMD. It can be both a chronic (long-term) or more severe acute (short-term) condition.

Gastric dilatation presents as a feeling of bloating and an enlarged stomach. Bloating can be caused or exacerbated by a ventilator, in particular when set to a high pressure, causing air to enter the stomach.

Acute gastric dilatation presents itself as a severe localised abdominal pain, bloating and nausea and may cause an elevated heart rate. If untreated, it can cause perforation of the stomach or can cut off blood supply to other parts of the body, which can be potentially fatal. In emergency situations, it can be treated by pumping the stomach to remove a build-up of gas.

TOP TIP

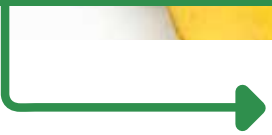
If you have a profiling bed with a tilting function, try tilting it when lying down to reduce reflux

TOP TIP

if you have a PEG or RIG feeding tube, you can relieve bloating by removing air using a syringe attached to the feed port.

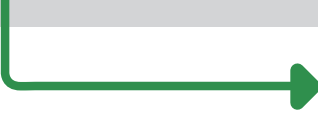


OUR 7 ESSENTIALS FOR DMD NUTRITION

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- 1 Take care of your nutrition in the same way you take care of breathing. It's just as important to get it right! Preventing nutritional problems is often much easier than treating them.
 - 2 Make sure you are regularly seen at a specialist clinic for adults with DMD, with access to a specialist neuromuscular dietitian and speech and language therapist.
 - 3 Find a way to get weighed regularly so you can address issues early and maintain a healthy weight.
 - 4 If you start to experience any problems with eating, chewing, swallowing, drooling, increase in times taken to finish meals, pooing or breathing, get these checked out as soon as possible at a specialist DMD clinic.
 - 5 Don't believe everything you read on the internet about nutrition. Research your sources and ask specialists in a DMD clinic.
 - 6 Try not to worry about feeding tubes. You might not need one, but if you do, many adults with DMD live successfully with a feeding tube and would never go back. If it's necessary for your health, don't delay!
 - 7 If you have a question about nutrition for other adults with DMD, contact DMD Pathfinders or ask a question in our Facebook group for adults with DMD.



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