

CASE STUDY: DYSPHAGIA MANAGEMENT AND NUTRITION SUPPORT IN A TEENAGE BOY WITH ATAXIA TELANGIECTASIA



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Paediatric dysphagia can affect children of all ages and it has a significant impact on nutrition if it is not managed effectively.

Poorly managed dysphagia also increases the risks of aspiration, recurrent chest infections and potentially, extended hospital stays (1). Dysphagia can occur amongst a wide range of patients (see Table 1) and symptoms can vary in severity. Problems can occur at each stage in the eating, drinking and swallowing process (see Table 2) resulting in varying symptoms (see Table 3).

In this article, I will discuss the joint dietetic and speech and language therapy (SALT) intervention for a teenage boy with dysphagia and ataxia telangiectasia (AT).

Case study

Emma has been working as a Paediatric Dietitian for five years and her caseload includes HETF, disability, coeliac disease, cystic fibrosis, PKU and childhood obesity. She has been a local Coeliac UK group organiser for 18 months.

At the age of 13, Adam was referred to our joint dietetic/SALT clinic due to weight loss and concerns from his parents regarding a marked reduction in his oral intake of food and fluids. He had begun to retch and cough during mealtimes and there were concerns regarding his oral coordination of fluids. He was diagnosed with AT prior to the family moving to the area. AT is a rare

genetic disorder which affects up to one in 40,000 people (4). Involving a number of systems in the body, it is a degenerative disorder. As the disorder progresses, one of the many health issues that AT patients experience is increased difficulty in coordinating movements. This affects walking, speech and eating/drinking ability. Cognitive development is also limited and slows or stops after the age of 10 to 12 (5). Dysphagia is common in AT patients and the limited evidence available suggests that it occurs when the disorder advances (6, 7) and aspiration (silent or frank) is a significant issue as AT patients reach their mid-teens (7). Dysphagia in AT patients is further exacerbated by increasingly poor posture and poor hand-to-mouth coordination. Texture modified diet and fluids, also enteral nutrition support, are often required as patients move into their teenage years. For more information about ataxia telangiectasia visit: www.atsociety.org.uk/about-at

Adam's parents reported that he had always been a 'good eater'. He would eat most of the family foods he was offered ▶

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and his appetite was good. According to his previous medical notes his weight had been tracking along the 50th centile. However, his weight had been static for almost 12 months when he arrived in clinic and it had drifted to the 25th centile. Over a period of six to eight months, Adam had become fussier with the foods he would accept, either at his new school and at home. He was refusing mixed textured foods, e.g. spaghetti bolognese, baked beans and sandwiches, which he had previously eaten well. He would accept some foods containing soft lumps, which were a more unified texture. He was happy to eat 'bite and melt' foods, such as corn based crisps and ice cream wafers. Soft, smooth foods, e.g. porridge, yoghurt, mashed potato were well tolerated too. Staff at school had noticed that he would retch or cough when eating drier, soft foods, such as the inside of a jacket potato with tuna. This was one of his favourite meals. Parents confirmed that this was also an issue at home. The retching and coughing usually occurred about five minutes into the mealtime. Adam coped well until then and he would fatigue if he was encouraged to eat any longer. The retching and coughing did not occur once he'd had a rest following his main meal. He was then offered his pudding, which he would manage well if it was fairly smooth and soft. He enjoyed his pudding and would often manage a large portion of sponge cake and custard, natural yoghurt and pureed fruit or homemade rice pudding, without any coughing or retching.

There were also concerns that Adam was not coping well when drinking thin/unthickened fluids. Parents and school staff had observed Adam coughing and spluttering when taking more than a few sips. He had a 'wet' sound to his voice after drinking and he would refuse to take more than 100ml liquid at any time.

Adam's nutritional assessment showed that he would manage to meet his requirements on four to five days of the week, but there were days when his intake wasn't so good. I discussed this with Adam's parents who were already fortifying his food with extra butter, cheese, milk and olive oil. Mum was struggling to see where she could add more calories to the food he ate. In light of Adam's dysphagia symptoms, supplement drinks were not recommended at this time and parents agreed to use a powdered supplement (Pro-Cal), which was added to the foods he was tolerating

Table 1: Types of patients affected by dysphagia (2, 3)

Acquired brain injury, e.g. cerebral palsy, stroke
Birth defects
Cleft palate
Degenerative disorders, e.g. hereditary ataxia syndromes gastroesophageal disorders, including reflux
Heart or lung conditions
Genetic conditions, e.g. Down's syndrome, Rett's syndrome
Head and/or neck cancer
Neuromuscular disease, e.g. Duchenne's muscular dystrophy
Oral sensory or oral motor problems
Prematurity

Table 2: Stages of the eating, drinking and swallowing process (2)

Oral preparatory phase - food is chewed and made in to a suitable sized bolus
Oral phase - food/liquid is moved around the mouth using the tongue, towards the back of the throat
Pharyngeal phase - the food/liquid is swallowed, it passes through the pharynx to the oesophagus

well, e.g. his puddings and soft main meal foods. SALT recommended that he was given a texture 'D' pre-mashed dysphagia diet (8).

During this joint appointment with SALT, his eating and drinking was observed and it was apparent that Adam fatigued after a short time eating and drinking. He was positioned well in the wheelchair he had. After observing Adam drinking in clinic, his SALT recommended thickening his fluids to stage 1 consistency (9) and this was well tolerated. He was also referred for a videofluoroscopy, which highlighted a number of concerns when Adam swallowed liquids. When taking thin/unthickened fluids from a cup and through a straw, both penetration and aspiration were observed. He had poor palatal elevation and took multiple swallows to clear a single bolus. Poor oro-motor control was noted, which became more evident as Adam fatigued. However, this, along with his swallowing ability, improved when given stage 1 fluids from a cup. Further improvements were seen when the fluid was taken through a straw. Adam's SALT recommended that he always be given stage 1 fluids and to of- ▶

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Food and Fluid Thickener
for patients with dysphagia



Stage 1



Stage 2



Stage 3

Designed to maintain
the original appearance of drinks
which may support compliance
and improve fluid intake

Table 3: Symptoms and signs of dysphagia

Choking
Coughing/retching
Gagging
Colour change during or after feeding (very flushed or becomes very pale)
Difficulty when chewing or taking liquid from a cup
Poor coordination of sucking, swallowing and breathing when bottle fed
Irritability/reluctance during feeds
Lengthy feeding times (greater than 30 minutes)
Refusal of previously accepted food or liquids
Limited intake of food or liquids
History of chest infections
Congestion during or after feeding
Change of breathing rate with feeding
Reporting a sensation of food 'getting stuck' in the throat
'Wet' sounding voice after drinking
Faltering growth
Evidence of food or liquid in any tracheotomy tube in situ

fer them via cup or straw, preferably via the straw when tired.

We provided a written report with our recommendations for staff at Adam's school to follow. We also visited the school to discuss menu options for him with the school cook. She was keen to ensure that she was providing the correct texture of food for Adam. His SALT gave training to Adam's school support worker regarding the thickening of his fluids.

Two months later, we reviewed Adam at school with his parents, the school nurse and his support worker. He had been eating well and was managing his D texture diet. He had gained a small amount of weight, which meant he was tracking along the 25th centile. However, Adam's fluid intake had remained limited. When we discussed this with Adam, he told us that he disliked having his fluids thickened. He didn't like the sensation in his mouth and he

had been refusing fluids because of this. We discussed the benefits of thickening his fluids and Adam understood the risks of having his fluids without a thickener. He agreed to try again using an alternative thickener. This was more acceptable and he was taking more thickened fluids, but still reluctant to drink large amounts. Adam told me that he didn't feel thirsty enough to drink any more.

Subsequent reviews have highlighted further deterioration in Adam's ability to cope with stage 1 fluids. In light of this, his SALT has recommended stage 2 fluids (9). Adam's oral intake of solid food has decreased due to feeding fatigue. He manages to maintain his calorie intake through good food fortification of his texture D diet with added Pro-Cal, as well as high calorie supplement puddings, which are given as snacks twice a day. The deterioration in his condition has led to discussions with Adam and ▶



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his parents about the benefits of a gastrostomy tube, as it may become increasingly difficult for Adam to meet his nutritional and hydration needs. Adam understands that this is likely to be the next step in his feeding and hydration journey. He tells me that, although he worries about how the gastrostomy tube will feel, he

wants to remain well for as long as possible and understands that a gastrostomy will help him to achieve this.

With special thanks to Sioned Darlington (Specialist Paediatric SALT, Wrexham Maelor Hospital, BCUHB)

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Questions relating to: *Dysphagia management and nutrition support in a teenage boy with Ataxia Telangiectasia*
Type your answers below and then **print for your records** or print and complete answers by hand.

Q.1	Define dysphagia, giving examples of signs and symptoms.
A	
Q.2	Describe the three stages of the eating process.
A	
Q.3	What conditions and disorders in childhood can be affected by dysphagia?
A	
Q.4	What are the risks of poorly managed dysphagia in children?
A	
Q.5	Why is dysphagia a problematic condition in patients who are suffering from Ataxia Telangiectasia?
A	
Q.6	In the case study, what were Adam's dysphagia symptoms?
A	
Q.7	Outline the four Dysphagia diet food texture descriptors (DDFTD).
A	
Q.8	What are the risks of not thickening fluids to a dysphagic patient?
A	
Q.9	Describe how a gastrostomy tube would be beneficial to Adam in the case study.
A	

Please type additional notes here . . .