Feeding difficulties are known to occur with advancing age in Duchenne muscular dystrophy (DMD). We evaluated the role of videofluoroscopy swallow study (VFSS) in a group of 30 DMD patients with feeding difficulties. Indications for feeding assessment were: respiratory infections potentially attributable to aspiration (n = 10) and/or episodes of choking (n = 24) subdivided into isolated choking events (n = 8) and regular choking during swallowing (n = 16). Indications for assessment were analysed in relation to the VFSS results. Median age at assessment was 17.13 years (range 6–31.4). Twenty-four VFSS were performed. Prolonged chewing and effortful bolus transport for solids increased with age. Swallow trigger was normal in the majority of cases. All patients had some post-swallow pharyngeal residue around the laryngeal inlet increasing in volume with age. Although this residue did not result in aspiration, it was worse in patients that were frequently choking. Three patients intermittently had penetration of the supraglottic space that did not reach the vocal folds during the swallow. Our results suggest that reported swallowing problems when assessed are not always associated with difficulties on VFSS. It is the oral phase of swallowing that is most significantly affected in DMD. The pharyngeal phase is well triggered but is weak with incomplete pharyngeal clearance leaving pharyngeal residue. Insufficient or effortful chewing coupled with weak clearance may predispose them to choking episodes either as a one off event or with increasing frequency with age. This study suggests that VFSS may not be of additional benefit to careful feeding history and observation in DMD with feeding difficulties.

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1. Introduction

Duchenne muscular dystrophy (DMD) is the most frequent neuromuscular disorder occurring in childhood and is due to mutations of the Xp21 gene. DMD is clinically characterised by progressive muscle weakness leading to loss of ambulation by 13 years of age. Respiratory and cardiac functions are progressively affected. Feeding and nutritional problems have been reported to be frequent after the first decade, often associated with gastrointestinal dysfunction and reduced weight gain. Feeding difficulties have been reported in approximately 30% of DMD patients younger than 25 years. It has also been reported that in boys with DMD reduced bite force and malocclusion may also have an impact on chewing and create feeding difficulties. Weakness in muscles of the swallow mechanism can result in choking. We have recently published the results of a survey in an Italian cohort of patients with DMD. Our study included patients up to 35 years and we found an overall incidence of 28% of feeding problems. In the great majority of cases, however (20% of the whole cohort), there were difficulties in mastication that appeared to be related to a combination of weakness of the masticatory muscles and malocclusion. These were uncommon in patients younger than 18 and progressively increased with age. Swallowing difficulties occurred in 18%, mainly above the age of 18 and consisted of episodes of choking on solids and less frequently on liquids. In that study, however, swallowing difficulties were only reported subjectively by the patients or their families. Video fluoroscopic swallow studies (VFSS) were not systematically performed and we could not therefore establish whether choking symptoms were related to actual swallowing difficulties or establish specifically which phases of swallowing were affected. While swallowing patterns have been described in other neuromuscular conditions, so far no systematic study has explored swallowing difficulties in DMD. The aim of this study was to evaluate the extent of swallowing problems in a group of DMD patients referred to speech and language therapy (SLT) services because of possible symptoms of swallowing difficulties. We also tried to establish whether these results could help us better understand the extent and severity of the problems and provide general guidelines for our clinical practice.

2. Patients and methods

This was a retrospective case note review study of those DMD patients referred to SLT for feeding assessment. All the neuromuscular patients in our Unit are interviewed about feeding difficulties, specifically asking about chewing or swallowing difficulties or any other feeding problems. As a result of this, all the patients, who report episodes of choking or other swallowing difficulties or who have recurrent respiratory infections potentially attributable to aspiration, are referred for an assessment by a speech and language therapist (SLT). Poor oral intake associated with weight loss as an isolated indication for feeding assessment does not warrant VFSS. The Dubowitz Neuromuscular Centre is a tertiary referral centre for children and adolescents with neuromuscular disorders. Patients have previously been followed up into adulthood. More recently, and with the improved survival, patients are encouraged to transition into adult services soon after the age of 16. From the 349 boys with DMD that are registered on our clinical database at the Dubowitz Neuromuscular Centre, we excluded those lost to follow up in the last 10 years. Of the 267 remaining patients, 123 were ambulant (either independently or in knee-ankle-foot orthoses (KAFOs) and 144 had lost ambulation. Thirty of the 267 (11%) had presented with symptoms warranting referral for a feeding assessment by a speech and language therapist. These 30 patients formed the basis of this study. Five of these 30 patients had died at the time of case note review. Median age at death was 22.5 years. The median age of survivors was 19.4 years.

2.1. Speech and language therapy assessment

The SLT case notes were reviewed and data collated on indication for feeding assessment, feeding case history and oral examination and need for VFSS.

2.2. VFSS

VFSS was performed according to local protocol for range and volume of consistencies offered. Three teaspoons of smooth semi-solids, a thin liquid and a chewable solid would be offered. The environment was made to match the typical feeding environment based on pre-VFSS assessment and observation. Patients would typically be positioned in sitting typically in their own wheelchair in which they would usually eat. If they could they would feed themselves using adapted cutlery as required or be fed by a familiar carer. Most boys drank using a straw.

VFSS looked at the oral and pharyngeal phases of swallowing. In the oral phase, the bolus is manipulated and formed and held prior to swallowing. Oral transit then measures the ease of bolus transport into the pharynx for swallowing. The pharyngeal phase describes the onset of swallow trigger, and strength and efficiency of bolus clearance with a note of the location and volume of any pharyngeal residue. Assessment is made of airway closure during the swallow and any penetration or aspiration of material into the airway, including any response made to aspiration. In the healthy child the swallow usually triggers at the valleculae. There should be no significant residue noted in the pharynx after the clearing swallow. Deep penetration of the airway and aspiration are abnormal.

The VFSS data were reviewed and rated using the Steele measures (Steele C.M., 2006, Videofluoroscopy scales, personal communication, October 2006) for onset of pharyngeal swallow and range of hyoid movement and competence of airway closure during the swallow. A note of any residue was made to assess bolus clearance from the valleculae and pyriform sinus. Depth of any airway invasion using the aspiration/penetration scale were noted.
Additional clinical information, including growth, gastrostomy tube feeding and gastro-oesophageal reflux (GOR) were noted. Other data on disease progression, survival, faltering growth, frequency of respiratory infections, and non-invasive ventilation (NIV) were also collected. Results on patients were analysed in three age groups in keeping with the natural history of the disease (i.e. up to loss of ambulation, puberty, adulthood).

3. Results

3.1. Speech and language therapy assessment

Of the 30 patients, three had been seen on more than one occasion. Each assessment is included as a separate episode, totalling 35 feeding assessments. Median age at feeding assessment was 17.13 years (range 6–31.4). Table 1 provides details of the groups subdivided according to age.

Indications for feeding assessment were: respiratory infections potentially attributable to aspiration (n = 10) and/or episodes of choking (n = 24). Acute management of respiratory infections was at the patients’ local hospital so exact records of severity and diagnosis of chest infection were not available at case note review. Chest infection was noted as significant if it required antibiotics or hospital admission and occurred greater than three times in a year. Choking episodes were subdivided into those who had experienced an isolated choking event (n = 8) and those who were having an increase in the frequency of regular choking during swallowing (n = 16). Reduced oral intake leading to weight loss was an associated indication for feeding assessment in 12 patients and an isolated reason in five. Feeding difficulties were occasionally associated with changes to their NIV equipment or following scoliosis surgery (n = 4). Fig. 1 shows the indications for feeding assessment divided into age groups. Indications were frequently co-existing.

3.2. Feeding assessment

From the case histories, it was found that modifying food textures for ease of chewing and choking avoidance was common across all age groups (> 80%). Most reported having soft or chopped food, with sauces added and drinks available to help clear food residue. Patients of group 1 were all able to feed themselves with assistance by using adapted cutlery, an elbow rest or by finger feeding. Fifty percent (n = 7) of group 2 continued to manage with assisted feeding but this fell to 33% (n = 5) in group 3.

Observation of facial appearance and oral examination was noted in all cases. Facial weakness indicated by an inability to bury their eyelashes on closure, rounded cheeks, open mouth posture at rest and limited cheek retraction on smiling was noted in almost 50% of the cases. Only one patient in group 1

![Fig. 1 – Indications for feeding assessment.](image-url)

### Table 1 – Group data

<table>
<thead>
<tr>
<th></th>
<th>Group 1: 6–12 years</th>
<th>Group 2: 12–18 years</th>
<th>Group 3: 18+ year</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of feeding assessments</td>
<td>6</td>
<td>14</td>
<td>15</td>
</tr>
<tr>
<td>Median (year) (range)</td>
<td>9 (6.1–11.8)</td>
<td>16.1 (14.1–17.8)</td>
<td>21.8 (18.2–31.5)</td>
</tr>
<tr>
<td>Mobility</td>
<td>3 Independent walkers</td>
<td>14 wheelchair users</td>
<td>15 wheelchair users</td>
</tr>
<tr>
<td></td>
<td>1 KAFOs</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td></td>
<td>2 Wheelchair users</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non-invasive ventilation</td>
<td>1</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td>Scoliosis surgery</td>
<td>2</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Drugs: prednisolone</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Domperidone/omeprazole</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Lactulose</td>
<td>0</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Mean length of mealtimes (min)</td>
<td>33</td>
<td>41</td>
<td>37</td>
</tr>
<tr>
<td>Gastrostomy recommended</td>
<td>0</td>
<td>9</td>
<td>7</td>
</tr>
<tr>
<td>Inability to self-feed</td>
<td>0</td>
<td>7</td>
<td>10</td>
</tr>
</tbody>
</table>
presented with any drooling. Approximately half of the patients also had macroglossia. A third had dental malocclusion with incidence of anterior open bite increasing with age (Table 2). Poor oral/dental hygiene and incidence of glossopharyngeal breathing also increased with age.

### 3.3. Videofluoroscopic swallow study

Nineteen of the 30 patients proceeded to VFSS. As part of our routine, VFSS is only performed if there are indications of pharyngeal swallowing difficulties noted on SLT assessment, e.g. unexplained or recurrent upper respiratory tract infections (>3 per year), frequent coughing/choking or increased congestion during swallowing. Difficulties in chewing, prolonged mealtimes, inadequate intake or weight loss alone were not considered an indication for VFSS. Isolated choking episodes with no other risk factors for aspiration were usually monitored and only referred for VFSS if more frequent choking or chest infections developed. Decision to perform a VFSS is made by the SLT in liaison with the medical team. VFSS is indicated by a range of often co-occurring factors and is only carried out when information obtained will aid in reaching a diagnosis or is anticipated to alter management recommendations.

VFSS was done once in 19 patients, twice in 1 patient and three times in two patients totalling data on 24 VFSS.

VFSS was performed in all patients in the youngest age group, 6/14 in group 2 and 12/15 in group 3. Of the eight in group 2, who did not have VFSS, six presented with poor oral intake and weight loss that did not warrant specific swallow evaluation and two had isolated choking episodes not associated with acute chest infections or weight loss. Three in group 3 did not proceed to VFSS as the predominant indication for feeding assessment was limited oral intake and poor weight gain. It was thought that VFSS would not further inform and therefore change the likely course of management. One patient in this group also suffered extreme anxiety about choking and it was felt that VFSS may further increase anxiety. He responded well to treatment with paroxetine for his anxiety attacks.

### 3.4. Oral phase

Oral phase difficulties of prolonged chewing and effortful bolus transport for solids increased with advancing age. These difficulties were observed in 3 out of 6 in group 1, 5 out of 6 in group 2 and all 12 in group 3.

### 3.5. Pharyngeal phase

The timing of swallow trigger was assessed by onset of hyoid movement and location of the bolus prior to initiation of swallow. Swallow trigger was mildly delayed (hyoid movement begins 2–3s following arrival of the leading edge of the bolus at the shadow of the ramus of the mandible) in two patients in group 1 and one in group 2, with swallow triggering as the bolus reached the valleculae. A clear view of hyoid movement was obstructed by poor head position and scoliosis that worsened with age, making this measure impossible to assess in four cases in group 3. Where visualised onset was adequate but there was reduced range of hyoid movement during the swallow in all of the patients in group 2 and 3. Presence of residue in all studies after the first swallow made further assessment of onset of swallow trigger more difficult.

All patients had some post-swallow pharyngeal residue, indicative of weak pharyngeal propulsion. Mild residue, described as a thin coating of residual material in the valleculae and/or the pyriform sinus occurred in three patients in group 1, two in group 2 and one in group 3. There was an increase in volume of residue noted with age. Moderate residue described as nearly filling the vallecular and/or pyriform spaces was most common in the older groups, three in group 2 and 10 in group 3. Four cases had a picture of mixed severity of residue with moderate residue in the valleculae and mild in the pyriform sinus (groups 1, n = 2) or moderate residue in valleculae and mild in the pyriform sinus (groups 2 and 3, n = 2). Residue was most substantial for solid rather than liquid consistencies.

The appearance of residue after the first swallow made it appear as if bolus’s were swallowed piecemeal as multiple clearing swallows were triggered to clear residue. In three

### Table 2 – Facial features and oral examination

<table>
<thead>
<tr>
<th>Facial weakness</th>
<th>Macroglossia</th>
<th>Anterior open bite</th>
<th>Malocclusion</th>
<th>Poor oral hygiene</th>
<th>Glosso pharyngeal breathing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1 (n = 6)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group 2 (n = 14)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group 3 (n = 15)</td>
<td></td>
<td></td>
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</tbody>
</table>

●: Affected and ○: not affected.
cases (one in each group) spontaneous swallows were successful in complete clearance of residue. When asked if all swallowed material was gone frequently patients would report it had all gone, when residue remained in the pharynx, indicative of some pharyngeal desensitisation.

Complete closure of the laryngeal additus was evident during the swallow in all cases. None of the patients in any group showed distinct aspiration into the airway. One patient in group 2 and two in group 3 intermittently had material that entered the supraglottic space but did not reach the level of the true vocal folds during the swallow. This equates to level two on the penetration aspiration scale where material penetrates the airway but does not reach the true vocal folds and is then ejected.

3.6. Swallowing difficulties and age

Specific choking episodes that are recalled as a single event are most common in the younger age groups. Increased episodes of more frequent choking associated with weight loss and chest infections were seen with advancing age.

Indications for feeding assessment were linked to VFSS results (Table 3). A history of choking was related to the VFSS finding of moderate residue. Isolated episodes of choking are more associated with mild rather than moderate pharyngeal residue.

Chest infections were associated with a delay in the swallow trigger and not with airway penetration. Frequent choking episodes were more associated with airway penetration.

3.7. Management advice for feeding and swallowing difficulties

Safe swallowing advice was frequently given post-feeding assessment including advice to: dry swallow or drink thin liquids after eating to clear pharyngeal residue, to stay upright for 30 min after meals to aid safe clearance of pharyngeal residue and to keep mealtime length to 30 min to avoid swallow fatigue which may then increase risk of aspiration. Optimal head, neck and trunk positioning for safe swallowing was also reinforced. If there was a history of choking episodes, it was ensured that all carers were confident in what to do if their child choked. Texture modification was often in place prior to feeding assessment but was frequently reinforced after feeding assessment to reduce the occurrence of choking and food sticking. In one case, thickened liquids were recommended to reduce the risk of aspiration due to delayed swallow seen on VFSS. Two of the patients in group 3 were advised to use their NIV during mealtimes to reduce the work of glossopharyngeal breathing making it easier to co-ordinate swallowing with respiration and reduce the risk of aspiration of residue during effortful inspiration.

4. Discussion

As documented in other studies there are a subgroup of boys with DMD in our cohort who with age develop feeding difficulties including choking difficulties warranting assessment and intervention. The Dubowitz Neuromuscular Centre is a tertiary referral centre for children and adolescents with neuromuscular disorders. Since 1997, we have used the same criteria for referring patients with swallowing difficulties for SLT assessment. The number of children in our cohort followed at the same Centre who were in need of a SLT assessment was very low (11%) and this may reflect that ours is mainly a paediatric population and the number of patients followed after the age of 18 is relatively limited.

Patients referred for SLT assessment generally present with typical oral facial features characterised by bilateral facial weakness and malocclusion with an increased incidence of anterior open bite, poor oral hygiene and glossopharyngeal breathing with increasing age. One common reason given for feeding assessment was chest infections potentially attributable to aspiration. However, from the VFSS data no patients
were seen to aspirate. For the majority of patients the swallow trigger was within normal limits and in all cases airway closure during the swallow was complete implying the neurological or central control of the laryngeal aspect of swallowing was in tact. The clinical boundary of importance for morbidity and mortality is thought to lie between the penetration aspiration scale 2 and 3, the difference being whether penetrated material is ejected from the airway or not. Children who frequently penetrate the airway may go on to aspirate with fatigue.\(^{15}\) None of the patients seen for VFSS had an aspiration penetration score above 2 and although three had mild penetration of the airway, none were seen to aspirate or penetrate to an extent that would have potentially correlated with an increased risk factor for aspiration. Despite a weak swallow resulting in residue and in three cases penetration of the airway none of the patients were seen to aspirate.

Oral phase difficulties with bolus transport due to malocclusion, reduced tongue strength and coordination may result in inefficient bolus propulsion and subsequent vallecular residue. This may increase with fatigue and be aspirated later in a meal after the VFSS screening period. There is an increased risk of aspiration for some cases due to airway penetration and pharyngeal residue indicative of peripheral pharyngeal muscle weakness. The VFSS only screened swallowing for a short period in time mean length of mealtime is over 30 min. With a prolonged mealtime there may be increased risk of fatigue both in terms of muscle strength and control of respiration which may increase the risk of penetration and aspiration. The results therefore must be interpreted carefully as they do not necessarily reflect the swallow ability with fatigue.

Future research to determine if there is aspiration at some point after the VFSS screening period would have to either bring the patient back to be re-screened after a period of time post-swallows or use scintigraphy to more accurately assess the quantity and presence of any aspiration and residue over time.

The existence of more residues with solid textures may increase the risk of choking on these consistencies supporting the advice to modify food consistencies for ease of swallowing. VFSS as reported here may not be an essential diagnostic assessment in this population despite their frequent presentation with choking and recurrent chest infections. Chewing difficulties associated with malocclusion and difficult self-feeding may limit oral intake having a consequent effect on growth and maintenance of weight. It is therefore essential that feeding is monitored and assessed to support sufficient oral intake. Assessment using mealtime observation may give sufficient information to inform effective intervention strategies aiming to reduce the length of mealtimes, aid self-feeding or modify the consistency of the diet as required.

Feeding difficulties are most common over the age of 14 years with an increase in choking episodes associated with changes in neck and trunk positioning and worsening respiratory function. Development of malocclusion and anterior open bite results in poor chewing which can cause choking or reduced oral intake resulting in weight loss with a consequent impact on general health. Range of hyoid movement, where able to be visualised, was reduced in all of the patients over 16 years of age indicative of some progressive muscle weakness associated with swallowing. Poor hyoid-laryngeal elevation is associated with reduced airway closure and reduced cricopharyngeal opening, increasing the risk of aspiration.

The amount of residue on VFSS increased with age over the three groups. Residue in the valleculae and pyriform sinus that sits around the laryngeal inlet may increase the risk of choking as residue may penetrate the airway during inspiration at any time after the swallow. Presence of residue may give the sensation of food being stuck in the throat. The amount of pharyngeal residue was worse in patients that reported choking.

Increase in isolated choking episodes with age was often associated with a fear of swallowing and a reluctance to continue eating. Often boys could relate clearly the specific choking episode that had discouraged them from eating. In all the cases, where there was a specific choking episode that triggered, the referral there was not yet an associated weight loss. These episodes appear to be isolated and acute as opposed to the gradual increase in regular choking that is sometimes seen with age in this population.

Choking episodes may also be linked to fast rate of care giver feeding or isolated episodes on particularly difficult consistencies, e.g. swallowing tablets. There may be other specific feeding or texture modification recommendations that can reduce the incidence of such choking events.

Boluses were seen to be swallowed piecemeal, which may also be indicative of lingual and pharyngeal weakness. Frequently, when asked if all the bolus had been cleared from the pharynx the patient would say it had when there was still residue present indicating some desensitisation to residue. Increasing awareness of pharyngeal swallow problems may be of benefit to patient education for strategies such as dry swallowing after a meal, the need to drink fluids to clear solid residue or the importance of staying upright after meals for safe clearance of residue. However, in some cases, it may hypersensitise awareness and thus increases anxiety about potential choking.

Other tests that assess strength of pharyngeal swallow, e.g. myometry or manometry may be useful to further understand the mechanics of the swallow in DMD. Eating while on NIV gave symptomatic relief to older patients who had developed glossopharyngeal breathing. Further VF studies of swallowing on and off NIV would be of interest.

5. Summary

Our results suggest that oropharyngeal swallowing problems are relatively rare in DMD patients and that, even when they are reported by patients, are not always detected on video-fluoroscopy. As previously reported by our groups in another population, it is the oral phase of swallowing that is most significantly affected in DMD with prolonged chewing required potentially due to mild facial weakness, reduced bite force and malocclusion and poor tongue control over bolus preparation and propulsion. This is reflected in prolonged mealtimes and the history of texture modifications to avoid hard and chewy food. In this study, using videofluoroscopy we were able to demonstrate that the pharyngeal phase is well triggered but is weak with incomplete pharyngeal clearance.
leaving pharyngeal residue. Insufficient or effortful chewing coupled with weak clearance may predispose them to choking episodes either as a one off event or with increasing frequency with age.

This study suggests that VFSS may not always be of additional benefit to a careful feeding history and observation, for advising on food texture modification and safe swallowing in patients with DMD presenting with feeding difficulties but is often needed to give an objective assessment of swallowing in patients who report choking episodes and are anxious about mealtimes or in those who have recurrent chest infections and weight loss.

Our results would suggest that when assessed with VFSS, airway protection is complete and there was not one episode of deep penetration or airway aspiration seen during the VFSS. The chest infections, when present are therefore more likely to be due to general respiratory decline rather than specific aspiration episodes.

The population of DMD is changing as survival increases and larger studies in adult DMD patients are essential to better understand the nature and clinical course of these difficulties for optimal future management.

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REFERENCES