Concise Report

Oropharyngeal dysphagia in juvenile dermatomyositis (JDM): an evaluation of videofluoroscopy swallow study (VFSS) changes in relation to clinical symptoms and objective muscle scores

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Objective. To determine if objective, validated scores of muscle weakness and function [manual muscle testing (MMT), childhood myositis assessment scale (CMAS)] or scores of general disease activity or function [childhood health assessment questionnaire and physician global assessment of disease activity visual analogue scale (VAS)], can predict children at risk of swallow abnormalities in juvenile dermatomyositis (JDM) measured by videofluoroscopic swallow studies (VFSS).

Methods. Patients were referred for speech and language dysphagia assessment upon diagnosis of JDM or flare of disease. VFSS was used to document a swallow score indicating severity of swallow dysfunction. Clinical symptoms, examination findings and objective scores of disease activity were analysed. Any correlation was looked for using chi-squared Fisher exact test and linear regression models.

Results. Fourteen patients with inflammatory myopathy (age 2–16 years) had clinical assessments and VFSS. VFSS was abnormal in 11 children (79%). Only two children were asymptomatic at assessment, but both had swallow dysfunction, including aspiration, on VFSS. In contrast, three of the symptomatic children had a normal VFSS. No relationship was found between objective disease severity scores and VFSS swallow score.

Conclusions. This study failed to show any correlation between swallow score and objective measures of muscle strength and function (MMT/CMAS) or general disease activity and function [physician VAS/childhood health assessment questionnaire (CHAQ)]. In the absence of a more accurate objective method to determine which children with active JDM are most at risk of swallow dysfunction and aspiration, all children with active dermatomyositis should be referred for speech and language assessment and VFSS.

Introduction

In spite of the prevalence of dysphagia in inflammatory myositis, reported in 29–44% children, it can be overlooked until symptoms are severe [1–3]. Silent aspiration in children with dysphagia, from any cause, has been recognized [4, 5]. Although swallow abnormalities are not universal in juvenile dermatomyositis (JDM), recognition of unsafe swallow is crucial to avert aspiration and lung damage, preventable by nasogastric or parenteral feeding. Swallow dysfunction may occur due to weakness of oropharyngeal, laryngeal and oesophageal musculature. X-ray videofluoroscopic swallowing study (VFSS) is the procedure of choice in children to delineate pharyngeal and oesophageal phases of swallow [4, 6].

Many groups have compared clinical examination of dysphagia, due to any cause, with VFSS findings. Those involving children have recognized (as in adults) that clinical features cannot accurately predict aspiration [5, 6, 7]. A study of 13 adults with myositis and dysphagia showed lack of supporting clinical signs and a substantial number of false negatives for laboratory tests in recognition of swallow dysfunction detected by VFSS [8]. An abstract by Punaro et al. describing 31 children with dermatomyositis demonstrates that clinical indicators do not predict aspiration [9].

VFSSs expose children to radiation, are expensive, time consuming and can be an unpleasant, frightening experience for some children. If there was a way of predicting which children with dermatomyositis were at risk of aspiration, resources could be targeted accordingly. This study aims to establish predictive factors that could determine children at risk of aspiration from JDM, particularly looking at objective scores of muscle strength and function, which have not been considered in previous studies.

Patients and methods

Patient recruitment and analysis

Following ethics approval, children with probable or definite JDM defined by Bowan and Peter criteria [10] were referred prospectively for speech and language assessment upon diagnosis or flare of their condition. Data collection took place following informed written parental consent and patient assent. All children were evaluated and treated between July 2003 and April 2005 at Great Ormond Street Hospital, London, UK. Each patient underwent a clinical speech and language assessment and a videofluoroscopy of swallow, recorded whilst consuming various food and liquid consistencies, in a seated position, as per standardized protocol (available upon request). The dysphagia outcome severity scale (DOSS) was used to determine severity levels for all patients who had undergone videofluoroscopy assessment. This seven-point scale was developed to systematically rate the functional severity of dysphagia based on objective assessment [11].

Clinical notes for each patient and research data collected from the Juvenile Dermatomyositis National JDM Registry and Repository (UK and Ireland) through the Juvenile Dermatomyositis Research Group [1], were reviewed to determine demographic data, clinical symptoms and examination findings at time of VFSS, as well as manual muscle testing (MMT) [12], childhood myositis assessment scale (CMAS) score [13],
childhood health assessment questionnaire (CHAQ) [14] and Physician global assessment of disease activity visual analogue scale (VAS) [15]. A record was taken of time from onset of symptoms to diagnosis and time from diagnosis to VFSS. Medication use at the time of and preceding VFSS, was recorded. Laboratory findings including creatine kinase (CK), lactate dehydrogenase (LDH), alanine aminotransferase (ALT) and aspartate aminotransferase (AST), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), full blood count (FBC) and auto-antibodies (ANA, anti-ENA, anti-DNA) were analysed and correlated to VFSS findings. The presence of interstitial lung disease was evaluated by pulmonary function tests (spirometry, plethysmography and transfer factor), chest X-ray and, wherever appropriate, high-resolution computed tomography (HRCT) scan.

**VFSS technique.** All children were assessed by VFSS performed by a paediatric radiologist in conjunction with a speech and language therapist. A fluoroscopy unit (Siemens Polystar digital unit- Siemens AG, Erlangen, Germany) connected to a medical high definition medical quality videorecorder (Sony FBO 9500 MDP) was used. Videofluoroscopy studies were performed in a lateral position with the patient in a seated position. Each patient was asked to swallow a food and/or liquid bolus of high density (EZPaque 100% wpv) barium sulphate suspension. Images were acquired at 15 pulses/s.

**Statistical analysis.** Data were analysed using SPSS software. Chi-square looked for correlation between an abnormal VF result and clinical findings including systemic features, symptoms of weakness or swallow dysfunction, dyspnoea, myalgia, arthritis, vasculitis, ulcerative disease, oedema and calcinosis. Chi-square was also used to look for correlation between VFSS abnormalities and elevated muscle enzymes (CK, LDH, ALT, AST) or inflammatory markers (ESR, CRP), low haemoglobin, raised white cells or platelet count, evidence of interstitial lung disease (pulmonary function tests and HRCT) and medication use (corticosteroid, methotrexate, hydroxychloroquine, cyclophosphamide). Regression analysis determined any relationship between swallow severity score and CMAS, CHAQ, physician VAS or MMT.

**Results**

**Patient characteristics**

Speech and Language Therapy (SALT) assessment and VFSS was carried out in 14 patients (5 males, 9 females; 10 white, 1 black, 2 Asian, and 1 mixed race). The mean age at onset of the disease was 7.2 years (range 2.4–14.6) with mean age at diagnosis of 8.1 years (range 2.6–15.6). Mean time from diagnosis to video fluoroscopy was 15.2 months (range 0–120). Four children had overlap features with polyarticular arthritis (2), systemic lupus erythematosus (1) and scleroderma (1). Such children were included in the analysis provided that the pre-dominant clinical presentation was myositis. All children had active disease at the time of evaluation. Nine children had a muscle biopsy consistent with JDM in addition to evidence of raised muscle enzymes at time of presentation with proximal muscle weakness and characteristic skin changes. Five children did not have a biopsy, but had evidence of an inflammatory myopathy on T2 weighted STIR Magnetic Resonance Images with clinical and laboratory features of JDM. All children co-operated with SALT assessment and VFSS.

Five patients (36%) were evaluated at disease onset whereas the remaining nine (64%) had VFSS assessments during disease flares. Twelve patients (86%) had symptoms of palatal or oropharyngeal weakness at time of VFSS including dysphonia (10 out of 14, 71%) and dysphagia (9 out of 14, 64%). Some patients complained of mild symptoms at time of VFSS including food ‘sticking in the throat.’ Two children (14%) were asymptomatic at the time of VFSS, including one who had never reported swallow symptoms. Three patients (25%) had symptoms of dyspnea and abnormal pulmonary function tests, including one with evidence of pulmonary fibrosis on HRCT. All patients with abnormal lung function had abnormal VFSS, including aspiration (two out of three) and liquid in the pyriform fossa (associated with risk of aspiration).

Thirteen patients (93%) had constitutional symptoms with prominent fatigue (92%), headache (54%), irritability (46%), fever (31%) and weight loss (23%). Gastro-intestinal involvement (diarrhoea and abdominal pain) was seen in two (14%) patients and one patient (7%) had cardiac involvement (pericarditis). All patients (100%) had skin involvement including heliotrope rash, Gottron’s papules and peri-ungual erythema. Eleven (79%) had symptoms of weakness, nine (64%) had myalgia and eleven (79%) had arthritis.

Eight children (57%) were taking oral prednisolone and disease-modifying medication at the time of VF; including methotrexate (6), azathioprine (1), and hydroxychloroquine (1).

**VFSS results**

VFSS was abnormal in 11 out of 14 (79%) children (Table 1). Children with no symptoms of abnormal swallowing at the time of VFSS (n = 2) were found to have VFSS abnormalities as well as aspiration, including one with no symptoms of weakness. Three children had a normal VFSS examination, despite swallow symptoms at the time of examination including dysphonia (n = 3).

### TABLE 1. Videofluoroscopy results

<table>
<thead>
<tr>
<th>Pt</th>
<th>Time (months) from symptom onset to VFSS</th>
<th>Time (months) from diagnosis of JDM to VFSS</th>
<th>Muscle symptoms</th>
<th>Swallow symptoms at time of VFSS</th>
<th>VF result</th>
<th>Swallow severity score</th>
<th>Treatment pre VFSS</th>
<th>Clinical disease state</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12.0</td>
<td>0</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia and dysphonia</td>
<td>Aspiration thin liquids</td>
<td>3</td>
<td>None</td>
<td>CS;MTX</td>
</tr>
<tr>
<td>2</td>
<td>40.8</td>
<td>28.8</td>
<td>Weakness</td>
<td>None</td>
<td>Delay in liquid swallow</td>
<td>4</td>
<td>CS;MTX</td>
<td>Chronic</td>
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<tr>
<td>3</td>
<td>126</td>
<td>120</td>
<td>Weakness</td>
<td>No</td>
<td>Post swallow residue</td>
<td>6</td>
<td>CS;AZA</td>
<td>Chronic</td>
</tr>
<tr>
<td>4</td>
<td>7.2</td>
<td>1.2</td>
<td>Weakness</td>
<td>Yes; dysphagia</td>
<td>Trace residue</td>
<td>6</td>
<td>CS;MTX;HCQ</td>
<td>Chronic</td>
</tr>
<tr>
<td>5</td>
<td>26.4</td>
<td>24</td>
<td>None</td>
<td>No</td>
<td>Aspiration</td>
<td>3</td>
<td>CS;MTX;HCQ</td>
<td>Chronic</td>
</tr>
<tr>
<td>6</td>
<td>1.2</td>
<td>0</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia and dysphonia</td>
<td>Nasal regurgitation</td>
<td>5</td>
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<td>7</td>
<td>10.8</td>
<td>10.8</td>
<td>Weakness</td>
<td>Yes; dysphagia</td>
<td>Liquids in pyriform fossa</td>
<td>4</td>
<td>CS;MTX</td>
<td>Chronic</td>
</tr>
<tr>
<td>8</td>
<td>6.4</td>
<td>4.8</td>
<td>Weakness</td>
<td>Yes; dysphagia</td>
<td>Liquids in pyriform fossa</td>
<td>4</td>
<td>CS;MTX</td>
<td>Chronic</td>
</tr>
<tr>
<td>9</td>
<td>7.2</td>
<td>1.2</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia and dysphonia</td>
<td>Aspiration</td>
<td>3</td>
<td>CS</td>
<td>Frare</td>
</tr>
<tr>
<td>10</td>
<td>14.4</td>
<td>2.4</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia and dysphonia</td>
<td>Aspiration of liquids</td>
<td>3</td>
<td>CS;MTX;HCQ;CYP</td>
<td>Frare</td>
</tr>
<tr>
<td>11</td>
<td>0</td>
<td>0</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia</td>
<td>Pharyngeal wall weakness</td>
<td>6</td>
<td>None</td>
<td>Onset</td>
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<tr>
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<td>0</td>
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<td>Yes; dysphagia and dysphonia</td>
<td>Aspiration of liquids</td>
<td>3</td>
<td>CS;MTX;HCQ;CYP</td>
<td>Frare</td>
</tr>
<tr>
<td>13</td>
<td>6</td>
<td>0</td>
<td>Myalgia</td>
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<td>Normal</td>
<td>Normal</td>
<td>CS;MTX</td>
<td>Frare</td>
</tr>
<tr>
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<td>25.2</td>
<td>19.2</td>
<td>Weakness and myalgia</td>
<td>Yes; dysphagia and dysphonia</td>
<td>Normal</td>
<td>Normal</td>
<td>CS;MTX</td>
<td>Frare</td>
</tr>
</tbody>
</table>

Pt, patient; swallow severity score 1–7: 1 = severe abnormal swallow; 7 = normal swallow.

CS, Corticosteroid; MTX, Methotrexate; AZA, Azathioprine; HCQ, Hydroxychloroquine; CYP, Cyclophosphamide; CIS, Ciclosporin.
and dysphagia \((n = 2)\). Two children with normal VFSS had received treatment with corticosteroid and methotrexate, but one child had not received treatment at the time of assessment. Other children had an abnormal VFSS \((n = 11)\) whether they had received medical intervention \((n = 8)\) or not \((n = 3)\), as shown in Table 1. There was no relationship between swallow severity score and MMT, CMAS, Physician VAS or CHAQ (Fig. 1). There was no statistically significant correlation with abnormal VFSS and symptoms at presentation or at the time of VFSS (data not shown). Medication at time of VFSS did not predict an abnormal or normal result. The presence of interstitial lung disease, raised muscle enzymes or inflammatory markers, abnormal FBC or positive ANA did not correlate with abnormal VFSS (data not shown). There was no significant correlation between an abnormal clinical assessment and an abnormal VFSS examination \((P = 1.0; \text{ sensitivity } 81.1\%; \text{ positive predictive value } 75\%)\). The time from the onset of symptoms to VFSS did not correlate with an abnormal VFSS result.

Children found to have severe abnormalities on VFSS were treated aggressively with corticosteroids and immuno-suppressant medication, in addition to being kept nil by mouth until they had a reliable swallow.

**Discussion**

This study has shown that swallow dysfunction and aspiration is not restricted to children who are severely weak with JDM. It is important to prevent aspiration damage in these immuno-suppressed patients. The study aimed to determine if objective scores of muscle weakness or function could predict those children at risk of swallow abnormalities. The CHAQ is a valid, reliable measure of general function shown to correlate well with measures of disease activity in JDM, with good internal reliability, construct validity and strong responsiveness [14, 16]. The CMAS score is a quantitative, non-invasive method designed to measure proximal muscle strength, function and endurance across a wide age range [13]. It has excellent convergent validity, responsiveness and interrater and intrarater reliability [13, 17]. MMT is a quantitative measure of muscle strength in clinical practice and trial settings, using a 5 or 10 point scale for the neck, trunk, proximal and distal muscle groups. It is sensitive to detecting change in muscle strength when muscles are moderately weak, with good intrarater and interrater reliability [12, 13]. Physician global assessments of disease activity and damage in juvenile inflammatory myopathies have shown high intrarater reliability [15].

![Fig. 1. Linear regression analysis of VFSS scores and CHAQ score, CMAS score, MMT score and physician VAS. Swallow severity score 1–7: 1 = severe abnormal swallow, 7 = normal swallow. Manual muscle testing (MMT) 0–80: 0 = severe weakness, 80 = full muscle power. Childhood myositis assessment scale (CMAS) 0–53: 0 = extreme weakness, 53 = normal muscle strength and stamina. Physician VAS 0–100: 0 = extremely unwell, 100 = extremely well.](http://rheumatology.oxfordjournals.org/)

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**FIG. 1.** Linear regression analysis of VFSS scores and CHAQ score, CMAS score, MMT score and physician VAS. Swallow severity score 1–7: 1 = severe abnormal swallow, 7 = normal swallow. Manual muscle testing (MMT) 0–80: 0 = severe weakness, 80 = full muscle power. Childhood myositis assessment scale (CMAS) 0–53: 0 = extreme weakness, 53 = normal muscle strength and stamina. Physician VAS 0–100: 0 = extremely unwell, 100 = extremely well.
Pharyngeal involvement in JDM has been associated with a worse prognosis [18, 19]. More severe JDM associated with a higher CHAQ and physician VAS or a lower CMAS score and MMT score, could suggest a greater chance of an abnormal swallow and identify those children who are at risk of aspiration. In this way, resources could be targeted and radiation doses limited to those children that need VFSS evaluation. However, this study failed to show correlation between swallow score and objective measures of muscle strength and function (MMT/CMAS) or general disease activity and function (physician VAS/CHAQ). In addition, clinical evaluation, patient symptoms and laboratory markers did not accurately predict swallow abnormalities detected by VFSS.

This study is limited by the fact that not all children could be evaluated by VFSS prior to starting medical intervention. Only a small number of children have been analysed and larger prospective studies are needed. There are no control data available on VFSS assessment of healthy children of this age group or patients in remission from JDM. Parameters of swallowing function in older children are usually taken from what is known about normal swallow in the adult literature [20]. The group was heterogeneous, with some children presenting overlap features, although in all, the pre dominant feature at presentation was muscle weakness. Selection bias is possible. Children in this cohort may represent a more severe group of children with JDM due to nature of referrals to Great Ormond Street Hospital. In a study looking at 151 children in a National Registry and Repository for JDM, UK and Ireland [1] 33 out of 114 (29%) had recorded symptoms of dysphagia and 22 out of 126 (17%) had symptoms of dysphonia; compared with 12 out of 14 (86%) in this study with symptoms of dysphagia and/or dysphonia. Thus, findings may not be representative of the disease as a whole. Some children were assessed at presentation and others were assessed during the course of the disease. However, all children had active disease at the time of assessment.

There was no reliability testing between therapists, but all assessments were carried out by dysphagia-trained speech and language therapists, experienced in the use of VFSS. Not all centres will necessarily have this degree of expertise with transferability of diagnostic skills. However, until larger prospective studies are available, this study demonstrates that clinical features and objective scores of disease activity and function are unreliable in predicting swallow abnormality and hence, VFSS should be considered for all children with active inflammatory myopathy.

Further studies are needed to establish how often SALT assessments and VFSS need to be carried out in children with active disease and whether there is benefit in repeating VFSS to assess response to treatment, accepting a significant radiation dose.

Acknowledgements

The authors would like to thank all clinicians and therapists involved in clinical evaluation and collection of objective scores. Particular thanks to S. Maillard, Specialist Physiotherapist at Great Ormond Street Hospital, for calculation of MMT and collection of clinical data. We acknowledge all Speech and Language Therapists involved in the study, especially A. Clarke for her work on analysis of the Videofluoroscopies. The authors also wish to thank all personnel in the Juvenile Dermatomyositis Research Group responsible for data entry, particular Miss V. Brown and Mrs A. Juggins, in addition to recognizing the vital contribution of Dr K. Murray in establishing the Registry and Repository. We thank the patients and their families for agreeing to contribute to the National JDM Registry and Repository (UK and Ireland). The authors wish to thank the European Union for support to the project through a fellowship to Dr S. Garey (contract number AML/B7-311/97/0666/II-0246-F1) as well as the generosity of the Cathal Hayes Research Foundation in its support of the Juvenile Dermatomyositis Registry and Repository (UK and Ireland).

Rheumatology key messages

- Clinical assessment including objective scores of muscle strength and stamina and general quantitative scores of disease activity cannot be used to predict those children with JDM at risk of swallowing dysfunction.
- Silent aspiration is recognized.
- In the absence of better assessment or prediction techniques, all children with active JDM should be referred for SALT assessment and VFSS.

The authors have declared no conflicts of interest.

References