Modification of the Paediatric Gastro-oesophageal Reflux Disease Symptom and Quality of Life Questionnaire (PGSQ) for children with cerebral palsy: a preliminary study

Sarah Mills , Catherine Tuffrey, Lee Tbaily, Mark Tighe

ABSTRACT

Objective  Gastro-oesophageal reflux disease (GORD) is a common condition affecting children, characterised by the passage of gastric contents into the oesophagus causing pain, vomiting and regurgitation. Children with neurodisability (such as cerebral palsy; CP) are predisposed to more severe GORD due to coexisting gut dysmotility and exclusive/ supplemental liquid diet; however, there are no existing tools or outcome measures to assess the severity of GORD in this patient group. For children without CP, the ‘Paediatric Gastro-oesophageal Symptom and Quality of Life Questionnaire’ (PGSQ) assesses symptoms and response to treatment, but the questions are not suitable for children with significant cognitive impairment. We aimed to adapt the existing PGSQ assessment tool to enable use in evaluating children with CP and GORD.

Patients/Interventions  Cognitive interviews were conducted by the research team with six parents/carerers of children (aged 3–15) with CP (Gross Motor Function Classification System level V) who have current or past symptoms of reflux. They were asked to interpret the questionnaire using a ‘think-aloud technique’ and offer suggestions on alterations to questions. Reasons for changing questions included confusing/difficult to understand questions, differing interpretations of questions and response choices not applying to the patient group.

Results  The PGSQ was modified iteratively following each interview. Overall, parents/carerers reported that it was acceptable to recall information over the past 7 days. In the final version, it was felt the questions were relevant, useful and related to symptoms that were observed. It was easy to comprehend with no uncomfortable questions. Suggestions for future work included a section specifically focusing on the school day answered by school staff and home life answered by carers who assist them in the home.

Conclusions  We have adapted the PGSQ to improve relevance and acceptability for families/carerers of children with symptoms of GORD and neurodisability. Further work is needed to validate the questionnaire for this patient group.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Gastro-oesophageal reflux disease (GORD) is extremely common in children with cerebral palsy and can be problematic. There are several validated symptom questionnaires for children with GORD without comorbidities.

WHAT THIS STUDY ADDS

⇒ We have adapted the existing Paediatric Gastro-oesophageal Symptom and Quality of Life Questionnaire to improve face validity for families/carers of children with symptoms of GORD and cerebral palsy.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ This modification will aid in assessing efficacy of pharmacological treatments for GORD in children with cerebral palsy and potentially has significant cost-saving implications if treatments can be initiated/discontinued based on accurate symptom assessment.

INTRODUCTION AND BACKGROUND

Gastro-oesophageal reflux (GOR) is a common problem, characterised by the passage of gastric contents into the oesophagus. GOR affects approximately 50% of infants less than 3 months old; however, most children improve with age, with less than 5% of children with vomiting or regurgitation in infancy continuing to have symptoms after the age of 14 months. In some children, GOR is associated with troublesome symptoms or complications, known as GORD disease (GORD). Children with neurodisability, such as cerebral palsy (CP), are more likely to suffer from GORD, due to coexisting gut dysmotility, exclusive/ supplemental liquid diet and other medications (eg, medications for dystonia/epilepsy). Gastrointestinal complications can include oesophagitis...
and stricture formation, and extraintestinal sequelae can include secondary anemia, chronic respiratory disease and faltering growth.\(^4\)\(^5\)

It is estimated that there are currently 9000 children across the UK with CP and GORD. Antacids (proton pump inhibitors (PPIs), histamine H2-receptor antagonists (H2RAs)) and prokinetics are treatments that are often continued long term in many children without clear evidence of ongoing efficacy. Long-term treatment results in increased workload for parents/carers, treatment costs\(^6\) and potential adverse side effects.\(^7\) This area has been highlighted as a National Institute for Health and Care Excellence (NICE) research recommendation (NG1)\(^8\) as there is a lack of evidence in this patient cohort despite their significantly increased risk of morbidity and mortality due to aspiration and respiratory complications.\(^9\) These children are also more likely to be referred for fundoplication due to failure of medical treatments.\(^5\)\(^9\)

Children in this patient group are often empirically treated for GORD without investigations to confirm underlying gastrointestinal pathology.\(^4\) Symptoms may be difficult to distinguish from coexisting dystonia, seizures or pain from other pathologies,\(^10\) and assessment is often affected by communication issues where children have cognitive impairment. Investigations to assess severity of GORD include 24-hour oesophageal pH monitoring and upper gastrointestinal endoscopy. One study found that these investigations in otherwise well children have variable correlations with symptoms and may not accurately predict the degree of improvement with treatment.\(^11\)

Frequency and severity of symptoms were shown to vary and were impacted by the types of nutrition consumed, stress, activity levels and intercurrent illnesses. Participants reported that GORD had a major impact on many aspects of the patients' lives, particularly school attendance/performance and participation in extracurricular and social activities. GORD also contributed to general feelings of frustration regarding symptoms, their effect on daily life and the need to take medication.

While assessment tools exist (Paediatric Gastro-oesophageal Symptom and Quality of Life Questionnaire (PGSQ), PEDS-QL GI\(^{12}\)\(^{13}\)), there are no robust data to help clinicians or researchers understand how well these assessment tools correlate with GORD in children with neuromuscular disorders. This is due to both the distributional properties that these tools have in this population and what constitutes a minimal clinically important difference. Establishing a robust outcome measure would allow development of clinical trials, for example, trials assessing efficacy of PPIs versus H2RAs. There are also significant potential cost savings if clinicians could consider initiating or discontinuing anti-reflux medications based on accurate reflux symptom assessment.

Symptom assessments through questionnaires are validated and are currently our most frequently used research tool in assessing improvement in normally developing children. The PGSQ takes on average 7 min to complete in typically developing children and is specific to infants (not assessed in this trial), children or young people (online supplemental appendix 2). The questions are very similar between the age groups, with the phrasing only taking account of the age differences. We aimed to modify the proxy version for parents, as patient and public involvement identified this one as the most likely to be used clinically.

**Aims**

To adapt the pre-existing PGSQ assessment tool to enable use in evaluating children with CP and GORD. This will allow changes in symptoms resulting from treatments to be measured and support clinical trials evaluating treatment efficacy.

**METHODS**

We included children with CP (Gross Motor Function Classification System, GMFCS levels III–V) with symptoms of GORD or on treatment for presumed GORD aged between 2 and 16 years. We only excluded children whose parent(s)/guardian(s) were not able to support their participation in the study in the opinion of the investigator (eg, language/communication issues, health, burden). All parents/carers of children meeting the inclusion criteria were approached about participation either during routine clinic appointments or by the paediatric research team.

Prior permission was sought from Takeda Pharmaceutical International (developers of the original PGSQ) to modify the existing questionnaire. Those who were eligible for recruitment were given the opportunity to participate either by phone, in clinic or by letter. Interviews were carried out by members of the research team trained in cognitive interview methods. Prior to the questionnaire, a standardised script was provided detailing the purpose of the study to ensure that all parents/caregivers received the same information. Interviews were recorded and transcribed using Microsoft Teams or WinScribe. Participants were asked to consider understanding, retrieval of information, judgement, response and construct for each question. A copy of the questions is shared in online supplemental appendix 1.

We focused on development and modification of the questionnaire using techniques described by Willis.\(^14\)\(^15\) This involved the participant talking through their thoughts as they read the questions, to ascertain whether each one reflected important and different dimensions of our patient group. Questions were modified based on parent/carer responses. Reasons for alterations included questions reported as not relevant and confusing or difficult to understand. This allowed relevant adjustments to better fit this subgroup of patients considering their communication issues and associated pathologies. Modifications continued until there were no further issues identified or improvements suggested. We only needed six participants using this method. The COnsolidated
criteria for REporting Qualitative research (COREQ) checklist was completed and is available in the appendices. The study was sponsored by University Hospitals Dorset National Health Service (NHS) Foundation Trust.

**Patient and public involvement**
The public was involved in the design and conduct of this research. Consultation groups were held at two schools for children with profound physical and learning disabilities and complex medical needs (both in Dorset). We outlined our research question to parents of children with CP and they were supportive. One parent was a coapplicant on our funding application to the British Society of Paediatric Gastroenterology, Hepatology and Nutrition. The Children and Clinical Research Group at Southampton NIHR (six children and several parents) reviewed the outcome measures and information sheets and agreed this was an important study. They felt that some of the questions were potentially emotionally challenging so advised that we should administer the questionnaires face to face rather than via the telephone or post. Based on this, participants were given the option to choose to participate in the way which suited them best. On completion of the study, participants will be updated on the results via a study newsletter and dissemination via relevant national charities. In addition to this, consumer members of the NICE Guideline Development Group for GORD in children identified this area as a research priority. Representatives from the NIHR Children Neurosciences Clinical Studies Group provided feedback on the proposed research and felt it addressed an important question.

**RESULTS**

**Patient demographics**
A total of six participants were enrolled in the study at one secondary care hospital site (University Hospitals Dorset NHS Foundation Trust). Demographic information is detailed in [table 1](#).

The children were either stable on antireflux medications, discontinuing medications for GORD or starting medications for GORD. This was to help demonstrate how the tool was understood by parents in static circumstances, and when treatments were changing.

**Modification pathway**
[Table 2](#) demonstrates how the questionnaire evolved with each cognitive interview. The parent/carer narrative is demonstrated along with the changes that were made to the questionnaire based on this feedback. The original and final versions of the questionnaire can be found in online supplemental appendices 2 and 3.

**DISCUSSION**
This study presents the modification of the pre-existing PGSQ for use in patients with neurodisability (eg, CP and severe learning disability) and GORD.

During the first interview, it was quickly established that questions requiring a response from the child (ie, point to the area where you feel pain) would not be acceptable to parents/carers of children with CP and severe learning disability. Questions regarding physical and social activities were also identified as potentially upsetting to parents/carers, as they highlighted skills that their child may have difficulty with. The most significant modifications, such as the addition or removal of questions and alteration of phrasing, were implemented between version 1 and version 2. Subsequently, parents and carers stated there were no upsetting or distressing questions and that they were mostly representative of their experience of GORD in their child.

One parent felt that they could not answer the questions in the school section because they were not with their child during the school day. They also expressed that their child’s school was used to dealing with problems associated with reflux, reducing the impact on schooling activities and therefore would not be an accurate depiction of the severity of their symptoms. They felt that it would be useful for school staff to complete or contribute to this section of the questionnaire.

Some parents felt that they were able to accurately assess their children’s symptoms and the frequency at which they were experiencing them, however, could not accurately attribute them to reflux rather than another cause.

<table>
<thead>
<tr>
<th>Age</th>
<th>Gender</th>
<th>GMFCS level</th>
<th>No of regular medications</th>
<th>Route of feeding</th>
<th>Previous fundoplication?</th>
</tr>
</thead>
<tbody>
<tr>
<td>9 years 7 months</td>
<td>F</td>
<td>V</td>
<td>4</td>
<td>Gastrostomy</td>
<td>No</td>
</tr>
<tr>
<td>15 years 11 months</td>
<td>F</td>
<td>V</td>
<td>8</td>
<td>Gastrojejunostomy</td>
<td>No</td>
</tr>
<tr>
<td>3 years 3 months</td>
<td>F</td>
<td>V</td>
<td>5</td>
<td>Gastrostomy</td>
<td>No</td>
</tr>
<tr>
<td>9 years 7 months</td>
<td>M</td>
<td>V</td>
<td>10</td>
<td>Oral and gastrojejunostomy</td>
<td>No</td>
</tr>
<tr>
<td>15 years</td>
<td>M</td>
<td>V</td>
<td>7</td>
<td>Gastrostomy</td>
<td>No</td>
</tr>
<tr>
<td>7 years</td>
<td>F</td>
<td>V</td>
<td>5</td>
<td>Gastrostomy</td>
<td>Yes—2016</td>
</tr>
</tbody>
</table>

F, female; GMFCS, Gross Motor Function Classification System; M, male.
Table 2  Table following the parent/carer narrative and how this led to the iterative evolution of the PGSQ

<table>
<thead>
<tr>
<th>Parent/carer narrative</th>
<th>Changes made: Version 1 → 2</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Interview 1</strong></td>
<td></td>
</tr>
<tr>
<td>► ‘They made sense (but) they weren’t applicable to my daughters’ specific case’ (Q1)</td>
<td>► Alteration of language used</td>
</tr>
<tr>
<td>► ‘It is not possible to do this with a child with complex needs and cerebral palsy; because I can’t show exactly, I don’t know exactly where the pain is at all’ (Q2)</td>
<td>− Q1: What your child has told you → what you have observed</td>
</tr>
<tr>
<td>► ‘I would find it very difficult, and I don’t think it would be accurate at all. I could make something up but that’s not what you want’ (Q2)</td>
<td>− Q3: Unable to eat what he/she wanted → unable to tolerate usual feed</td>
</tr>
<tr>
<td>► ‘It might be quite upsetting because the child often can’t tell you what they do or don’t want to do’ (Q3)</td>
<td>− Q3: Woken up someone in the house → changed sleeping pattern</td>
</tr>
<tr>
<td>► ‘You’d need to ask school, because you’re not there with your child’ (Q4)</td>
<td>− Q3: Felt frustrated/been in a bad mood/worried/upset → changed behaviour</td>
</tr>
<tr>
<td>► Splitting of question r.e. additional medicines/therapies into two separate questions</td>
<td></td>
</tr>
<tr>
<td>► Extra medications</td>
<td></td>
</tr>
<tr>
<td>► Extra treatments for example, massage/alternative therapy</td>
<td></td>
</tr>
<tr>
<td><strong>Interview 2</strong></td>
<td></td>
</tr>
<tr>
<td>► ‘The wording is easy to understand, not confusing’ (Q1)</td>
<td>► ‘(Q1) is really useful because it makes you feel like symptoms of reflux are being recognised’</td>
</tr>
<tr>
<td>► ‘(Q1) is really useful because it makes you feel like symptoms of reflux are being recognised’ (Q3)</td>
<td>► ‘I certainly think these questions describe what can happen within the school day with reflux’ (Q4)</td>
</tr>
<tr>
<td>► ‘It’s good to see it put down like this and to actually get the bigger picture of how it’s affecting everything’ (Overall)</td>
<td>► Addition of a ‘do not know’ column to question 2 and 3.</td>
</tr>
<tr>
<td>► Reported that the questions are not uncomfortable and easy to read</td>
<td>► Addition of extra clarification in the introduction ‘please include each day that the symptoms were persistent/troubling’ to help parents/carers quantify duration</td>
</tr>
<tr>
<td>► ‘(the wording) is quite specific, which is good’ (Q1)</td>
<td>− ‘(I remember) from the last couple of days unless I’ve had a particularly awful day which would stick in (my) mind’</td>
</tr>
<tr>
<td>► ‘If he had an undiagnosed reflux problem (the questionnaire) would make me feel like somebody was listening and wanting to help me’ (Q1)</td>
<td>► Formatting of the questionnaire edited to increase visibility of important points of questions for example, making certain words bold</td>
</tr>
<tr>
<td>► ‘I can’t think of any symptoms that haven’t been covered’ (Q1)</td>
<td>► Changing ‘do not know’ → ‘not relevant/do not know’</td>
</tr>
<tr>
<td>► ‘The questions are comfortable, describe the symptoms of reflux, no suggestions to change’</td>
<td>− ‘maybe ‘unsure’ because sometimes you could be unsure if (their symptoms) are due to reflux symptoms’</td>
</tr>
<tr>
<td>► ‘The first set of questions are definitely relevant because they’re things that are quantifiable’ (Q1)</td>
<td>► ‘The first set of questions are definitely relevant because they’re things that are quantifiable’ (Q1)</td>
</tr>
<tr>
<td>► ‘I’ve never had to fill in a questionnaire when she’s had reflux’ (Q1)</td>
<td>► ‘I’ve never had to fill in a questionnaire when she’s had reflux’</td>
</tr>
<tr>
<td>► ‘(r.e. school) I would be looking at her communications book to see if I could find out the answers’ (Q3)</td>
<td>► ‘(r.e. school) I would be looking at her communications book to see if I could find out the answers’ (Q3)</td>
</tr>
<tr>
<td>► ‘I think the school questions are more directed at children that are in mainstream education rather than special needs schools’ (Q3)</td>
<td>► ‘I think the school questions are more directed at children that are in mainstream education rather than special needs schools’ (Q3)</td>
</tr>
<tr>
<td>► ‘Unless you’re with your child at school for the whole time you’re not going to know the answers’ (Q3)</td>
<td>► ‘Unless you’re with your child at school for the whole time you’re not going to know the answers’ (Q3)</td>
</tr>
</tbody>
</table>

Continued
One parent was surprised that some described symptoms, such as bad breath, were signs of reflux; indicating that the questionnaire can help parents/carers identify lesser-known symptoms. This could ultimately assist with medication dosing and treatment plans as parents would be more likely to recognise and report these issues.

Another theme that emerged during the discussion was that the level of children’s impairment from neurodisability can differ considerably. A questionnaire of this type may not be suitable for all children; however, we aimed for broad applicability, and parents felt this questionnaire helped. One parent commented that the process had made them feel as if they were being listened to and taken seriously regarding their child’s symptoms.

Since there are currently no validated assessment tools in this patient group, this modification could potentially be extremely useful in clinics. A review of our cohort of patients between 2000 and 2015 found that the most common antireflux medication was omeprazole (prescribed in 70% of patients) and that patients remained on this for an average of 35 months. It is widely appreciated that these patients are usually commenced on treatment without investigation to confirm the diagnosis and that it can be difficult to distinguish between GORD and other coexisting pathologies. We hope that the modified questionnaire will assist with assessment of severity of symptoms and treatment response so that management can be optimised, improving patient care, costs and quality of life.

There are several limitations of this preliminary study. Due to the iterative process, the finalised questionnaire was only assessed by one parent/carer, though it is being further tested for face validity. In addition to this, the recruits were all locally identified, therefore, it may not be completely representative of other demographics, communities and socioeconomic variations throughout the rest of the UK. The children were all classified as level V using the GMFCS meaning that they have impairments in all areas of motor function, while this is not a surrogate for their ability to process and communicate, the cohort of children did have associated severe intellectual or learning difficulties. The diagnosis of a neurodisability such as CP covers a wide range of patients with a spectrum of communication abilities; therefore, these proxy questions may not be suitable for children who can self-report. Further work could involve development of self-report versions of the PGSQ suitable for this subgroup of children. We should highlight that the wider validation of the original PGSQ no longer applies to our modified version and we intend to further assess the developed tool including feasibility and test–retest reliability in a larger sample. However, the feedback received by parents/carers was that they felt the questionnaire was relevant to their child and the symptoms they observe them to have which are related to GORD.

**CONCLUSIONS**

We have adapted the existing PGSQ to improve face validity for families/carers of children with symptoms of GORD and neurodisability. More work is needed to ensure that the questionnaire is applicable to as many
children as possible within this patient group. The next phase will involve further assessment of the developed tool including feasibility and test–retest reliability. Future work will be needed to examine construct validity and sensitivity to change.

Acknowledgements We would also like to thank Dr Caroline Storey, Dr Laura Royce and Dr Erika Harterink-Rojas for their contribution to the project.

Contributors SCM performed qualitative data analysis of the interviews and writing and editing of the paper. MT devised the idea for the project and performed the setup of the study including ethical approval and obtaining grant funding. He was also involved in review and editing of the paper and is the guarantor for this study. CT and LT were involved in development of the research idea and reviewing and editing the paper.

Funding The project was funded by a £5000 grant (BiG Award) from the British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN).

Competing interests None declared.

Patient and public involvement Patients and/or the public were involved in the design, or conduct, or reporting, or dissemination plans of this research. Refer to the Methods section for further details.

Patient consent for publication Not applicable.

Ethics approval This study involves human participants and was approved by Health Research Authority IRAS ID: 273604. Participants gave informed consent to participate in the study before taking part.

Provenance and peer review Not commissioned; internally peer reviewed.

Data availability statement Data are available on reasonable request. Study began recruitment in 2018. However data could be provided on request.

Supplemental material This content has been supplied by the author(s). It has not been vetted by BMJ Publishing Group Limited (BMJ) and may not have been peer-reviewed. Any opinions or recommendations discussed are solely those of the author(s) and are not endorsed by BMJ. BMJ disclaims all liability and responsibility arising from any reliance placed on the content. Where the content includes any translated material, BMJ does not warrant the accuracy and reliability of the translations (including but not limited to local regulations, clinical guidelines, terminology, drug names and drug dosages), and is not responsible for any error or/and omission arising from translation and adaptation or otherwise.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited, appropriate credit is given, any changes made indicated, and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/.

ORCID iD
Sarah Mills http://orcid.org/0000-0002-1325-0784

REFERENCES
14 Willis G. Cognitive interviewing; 2011.