Clinical/Case Report

American Cleft Palate–Craniofacial Association

The Cleft Palate-Craniofacial Journal
2019, Vol. 56(5) 679-689

DOI: 10.1177/1055665618794070
journals.sagepub.com/home/cpc
SAGE

Tongue Reduction Surgery and Feeding Difficulties in Infants With Beckwith Wiedemann Syndrome: A Case Series

Nicole Prendeville, MRes, MSc[1] and Debbie Sell, PhD[2]

Abstract

Objective: To profile the pre- and post-operative feeding difficulties in infants with macroglossia in Beckwith Wiedemann Syndrome (m-BWS) who have had tongue reduction surgery (TRS) and to pilot a bespoke feeding rating scale.

Design: Retrospective consecutive case series designed with two pre-operative and one 3-month post-operative feeding assessments. A 4-point Likert-type scale was developed and applied retrospectively to describe the feeding behaviors for liquids, purées, and solids. Descriptive and non-parametric statistics were used.

Setting: National service for children with m-BWS at a pediatric hospital.

Patients: Twenty-five infants, age range 4 to 12 months at initial assessment, underwent TRS (median age = 16 months).

Intervention: Tongue reduction surgery.

Outcome Measure: Oral and selected pharyngeal stage feeding behaviors on liquids, purées, and solids.

Results: Pre-operative profile: Most feeding difficulties arose at the oral stage due to the macroglossia impacting important lingual movements. Difficulties were found with lip seal formation, biting, bolus manipulation and tongue lateralization. Aspiration risk was found in >75%. Texture modification was indicated for purées and solids.

Post-operative profile: There were statistically significant differences for each consistency pre- and post-operatively. Eighty-four percent of infants had age-appropriate drinking and eating skills. Mild residual difficulties with biting, tongue lateralization, and bolus manipulation remained for solids in four infants.

Conclusions: Feeding difficulties are common pre-operatively in m-BWS, putting infants at risk of aspiration if left unmanaged. TRS was effective in reducing or eliminating them. This is the first systematic report of infant feeding in m-BWS pre- and post-TRS.

Keywords

Beckwith Wiedemann syndrome, macroglossia, tongue reduction surgery, feeding, speech and language therapist

Introduction

The tongue plays a primary role in the successful orchestration of feeding and swallowing through the transfer of the bolus to the teeth for mastication, bolus formation for stripping against the hard palate and bolus cohesion prior to propulsion towards the esophagus (Robbins et al., 2005; Stierwalt and Youmans, 2007; Wilson and Green, 2009). The age period 6 to 12 months is a critical phase when weaning commences and transition to solids occurs (Delaney and Arvedson, 2008). Tongue enlargement and protrusion can lead to impairment in these microfunctions, resulting in a spectrum of feeding, speech, appearance,

[1] Speech and Language Therapy Department, Great Ormond Street Hospital, London, United Kingdom
[2] Centre for Outcomes and Experience Research in Children's Health, Illness and Disability (ORCHID), Great Ormond Street Hospital, London, United Kingdom

Corresponding Author:
Nicole Prendeville, Speech and Language Therapy Department, Great Ormond Street Hospital, Great Ormond Street, London WC1N 3JH, United Kingdom.
Email: nicolepren@gmail.com

Figure 1. Clinical phenotype.

and dental-occlusal concerns which can have long-term implications for the child and family (Shipster et al., 2006; 2012; Heggie et al., 2013).

Beckwith Wiedemann syndrome (BWS) is a rare congenital overgrowth disorder (incidence 1/13,700; Engstrom et al., 1988). Macroglossia in Beckwith Wiedemann syndrome (m-BWS) is observed in more than 80% of cases (Elliot et al., 1994; Martinez and Martinez, 1996). Figure 1 shows a typical macroglossia presentation before surgery and the change six months after tongue reduction surgery (TRS). TRS is the only means to correct macroglossia (Wolford & Cottrell 1996; Chau et al., 2011; Shipster et al., 2012). Surgery at our center is usually undertaken by the age of 2 years. The technique consists of a midline elliptical excision and an anterior wedge excision, followed by a straight-line suture of the incision borders (Shipster et al., 2012).

Children with m-BWS are commonly cited as having feeding difficulties (Elliot et al., 1994; Weng et al., 1995; Shipster et al., 2006; Tomlinson et al., 2007; Weksberg et al., 2010; Chau et al., 2011; Abeleira et al., 2012; Kadouch et al., 2012; Prada et al., 2012). Shipster and colleagues (2012) detailed six key features associated with the oral preparatory and oral phases of feeding before and after TRS. These were cup or bottle drinking with tongue position beyond the rim/teat, absent lip seal, poor oral retention of a bolus with lateral spillage, adaptive tongue tip use, messy eating, and poor tongue lateralization. However, the study was limited by the wide age range of the children, a small sample size and examination of a small number of feeding behaviors.

No systematic studies exist which examine the possible relationship between m-BWS and deglutition. Unmanaged feeding difficulties have been clinically observed and anecdotally reported by parents leading to longer mealtimes, avoidance of specific food textures, and maladaptive and inefficient feeding techniques. No literature exists on the detailed nature of feeding difficulties in infancy in m-BWS. In part, this is due to the paucity of validated infant feeding assessment tools (Skuse et al., 1995; Arvedson, 2008; Remijn et al., 2014; Thoyre et al., 2014; Benfer et al., 2015), with inconsistent reporting of pre- and post-operative feeding characteristics contributing to a weak evidence base.

At our center, all children with a confirmed diagnosis of m-BWS are routinely seen by a speech and language therapist (SLT) who evaluates feeding and age-dependent functional domains (speech, oral motor skill, drooling), in order to determine eligibility for TRS. During feeding, infants with m-BWS can present with the following issues: tongue protrusion, difficulty with bolus manipulation, excessive drooling during feeding, anterior bolus loss, impeded bolus transfer, limited tongue lateralization, a predominance of anterior–posterior tongue movements, bolus expulsion, and inappropriate swallowing of lumps leading to increased aspiration risk. These behaviors can have a negative impact on the appearance of feeding. Compensatory feeding techniques (eg, pacing, positioning, reduced flow rate of teat) and/or a modified diet (ie, texture modification) may be recommended.

There is a range of feeding assessment tools in use including the Schedule for Oral Motor Assessment (SOMA; Skuse et al., 1995), Early Feeding Skills Assessment for Preterm Infants (Thoyre et al., 2005), Observation List Spoon Feeding (van den Engel-Hoek, 2014), the Infant Malnutrition and Feeding Checklist for Congenital Heart Disease (St. Pierre et al., 2010), Eating and Drinking Ability Classification System (Sellers et al., 2013), Mastication Observation and Evaluation Instrument (Remijn et al., 2014), Pediatric Eating Assessment Tool (Thoyre et al., 2014), and the Ability for Basic Feeding and Swallowing Scale for Children (Kamide et al., 2015). However, most of these standardized tools have been developed and validated on children with different feeding/swallowing profiles and etiologies. There are three non-standardized checklists and scales frequently referenced in m-BWS studies: a BWS feeding checklist (Shipster et al., 2012); the Clinical Assessment of Paediatric Neurogenic Dysphagia (Morgan et al., 2008), and the Behavioral Assessment Scale of Oral Functions in Feeding (Stratton, 1981).

All tools were examined to determine whether any of the unique features of feeding in m-BWS were included. Only the SOMA (Skuse et al., 1995) had some relevant items. These included intermittent/incomplete upper lip contact/seal and smooth rhythmic sequence; however, it does not describe tongue protrusion in detail and its impact on key tongue movements during feeding. Furthermore, it does not capture the frequency of occurrence of a feeding behavior which can reflect the severity of impact. The binary rating system (yes/no) was considered inadequate due to the inability to differentiate between a mild and significant impact on feeding. The assessment also does not contain clinically observed compensatory behaviors such as persistent anterior–posterior tongue movements, swallowing whole lumps, or the child's use of their finger to remove (part of) the bolus.

The scale for our study needed to describe discrete feeding behaviors and difficulties and their frequency prior to TRS to inform management, while also examining post-operative feeding skills. A feeding rating scale for infants with m-BWS was developed and piloted (Figure 2). Items for the scale were generated from the clinical observations of feeding difficulties in m-BWS, previous studies in the area (Heggie et al., 2013;

Name: **Hospital No:**	**Date:**
SLT:	**Age:** **Premature: Y N**
Pre-op1 Pre-op 2 Post-op 1	**Data source: AV clinical notes Reports**

1. LIQUIDS **Not Assessed** ☐

Oral stage difficulties: how frequently is difficulty achieving this behaviour observed OR how frequently is this difficulty observed:

1. Lip seal	Never	Occasionally	Most of the time	Constant
2. Tongue protrusion under nipple/teat/spout/ cup rim	Never	Occasionally	Most of the time	Constant
3. Rhythmic nutritive suck	Never	Occasionally	Most of the time	Constant
4. Bolus transfer	Never	Occasionally	Most of the time	Constant
5. Anterior loss	Never	Occasionally	Most of the time	Constant
6. Prolonged feed times	Never	Occasionally	Most of the time	Constant

Pharyngeal stage difficulties:

7. Cough	**Never**	**Occasionally**	**Most of the time**	**Constant**
8. Risk of Aspiration	No		Yes	
9. Modified diet	No		Yes	
10. VFSS* required	No		Yes	

*VFSS – videofluoroscopy swallow study

Notes:

Figure 2. Feeding rating scale.

2. PURÉES **Not Assessed** ☐

Oral stage difficulties: how frequently is difficulty achieving this behaviour observed OR how frequently is this difficulty observed:

1. Bolus removal	**Never**	**Occasionally**	Most of the time	**Constant**
2. Lip seal	Never	Occasionally	Most of the time	Constant
3. Tongue protrusion while eating	Never	Occasionally	Most of the time	Constant
4. Bolus manipulation	Never	Occasionally	Most of the time	Constant
5. Lateral tongue movements	Never	Occasionally	Most of the time	Constant
6. Bolus transfer	Never	Occasionally	Most of the time	Constant
7. Anterior-Posterior tongue movts	No		Yes	
8. Anterior loss	Never	Occasionally	Most of the time	Constant
9. Swallows lumps whole	Never	Occasionally	Most of the time	Constant
10. Drooling while eating	Never	Occasionally	Most of the time	Constant
11. Spits out bolus	Never	Occasionally	Most of the time	Constant
12. Prolonged feed times	Never	Occasionally	Most of the time	Constant

Pharyngeal stage difficulties:

13. Cough	**Never**	**Occasionally**	**Most of the time**	**Constant**
14. Risk of Aspiration	No		Yes	
15. Modified diet	No		Yes	
16. VFSS required	No		Yes	

Notes:

Figure 2. (continued)

3. SOLIDS **Not Assessed** ☐

Oral stage difficulties: how frequently is difficulty achieving this behaviour observed OR how frequently is this difficulty observed:

1. Bite	Never	Occasionally	Most of the time	Constant
2. Lip seal while eating	Never	Occasionally	Most of the time	Constant
3. Tongue Protrusion while eating	Never	Occasionally	Most of the time	Constant
4. Bolus manipulation	Never	Occasionally	Most of the time	Constant
5. Lateral tongue movements	Never	Occasionally	Most of the time	Constant
6. Bolus transfer	Never	Occasionally	Most of the time	Constant
7. Anterior-Posterior tongue movts	No		Yes	
8. Rotary Jaw movement	Never	Occasionally	Most of the time	Constant
9. Anterior loss	Never	Occasionally	Most of the time	Constant
10. Swallows lumps whole	Never	Occasionally	Most of the time	Constant
11. Increased drooling while eating:	Never	Occasionally	Most of the time	Constant
12. Finger to remove bolus	Never	Occasionally	Most of the time	Constant
13. Prolonged feed times	Never	Occasionally	Most of the time	Constant

Pharyngeal stage difficulties:

14. Cough	**Never**	**Occasionally**	**Most of the time**	**Constant**
15. Risk of Aspiration	No		Yes	
16. Modified diet	No		Yes	
17. VFSS required	No		Yes	

Notes:

Rating: Frequency of occurrence

Never **0**	Occasionally **1**	Most of the time **2**	Constant **3**

Variables with binary response categories: Yes = 1 No = 0

Figure 2. (continued)

Shipster et al., 2012), items adapted from existing validated feeding tools (Skuse et al., 1995), parental reports of feeding difficulties, and best practice guidelines on clinical feeding dysphagia assessment (Royal College of Speech Language Therapists, 2009; Smith Hammond and Goldstein, 2006). To ensure face and content validity, the relevance, clarity, and interpretation of items were discussed with a group of SLT dysphagia specialists and one expert SLT in m-BWS.

Novel behaviors in the scale included difficulty with bolus manipulation, lateral tongue movements and bolus transfer, lip and tongue structure, function and position during eating and drinking. The scale is sub-divided into 3 sections based on the bolus consistency (liquids, purées, and solids) and contains 43 items. Each subscale is categorized into the oral and pharyngeal stages of deglutition. An item "cough" during or after bolus intake was included as a clinically observed behavior and indicator of pharyngeal stage difficulty and aspiration risk. Other clinical signs of aspiration included eye tearing and stress signals related to increased work of breathing: reddening of the face, noisy or wet upper airway sounds when feeding, grimacing while feeding, and a wet vocal quality during or immediately after feeding (Wolf and Glass, 1992; Arvedson and Brodsky, 2002). Two management items were included for all consistencies: the need for a modified diet and the indication for a videofluoroscopy swallow study (VFSS). The frequency of a feeding difficulty was coded using an ordinal adjectival scale (Streiner and Norman, 2008): constantly (3), most of the time (2), occasionally (1), and never (0) for most behaviors. Behaviors that occurred "most of the time" or "constantly" represented the greatest problem. Dichotomous scores were assigned to anterior–posterior tongue movements, aspiration risk, diet modification, and referral for VFSS. The scale was piloted on three patients not in this study cohort, to ensure comprehensiveness, and was revised with some items removed, indicative of a preliminary contribution to the content validity of the scale (Bowling, 2014).

This study aims:

i. to profile the feeding difficulties of infants with m-BWS prior to TRS,
ii. to examine the changes post-operatively,
iii. to describe typical feeding management recommendations, and
iv. to pilot a novel systematic approach to the analysis of feeding and drinking.

The National Health Service Hospital Trust Research and Development Department and the University School of Health Sciences Ethics Committee approved this project as a service evaluation which followed ethical and governance principles.

Methods

Patients

Clinical data from a consecutive series of 25 infants were analyzed within a single-center national service for children with m-BWS (April 2013 to March 2015). The inclusion and exclusion criteria are described in Table 1.

Table 1. Inclusion and Exclusion Criteria.

Inclusion Criteria	Exclusion Criteria
A confirmed genetic or clinical diagnosis of BWS with associated macroglossia	No confirmed diagnosis of BWS with macroglossia
Aged between 4 and 12 months at initial SLT assessment (pre-op 1[a])	Older than 12 months of age at initial pre-operative assessment
Was being weaned onto solids	Had a coexisting neurodevelopmental diagnosis
Less than 24 months old at the post-operative assessment	Had a mild macroglossia phenotype that did not require surgical management
Was fully orally feeding	Older than 24 months at post-operative assessment
Feeding difficulties on clinical assessment pre-operatively attributed to macroglossia	History of a previous tongue reduction procedure
Had undergone TRS	Exclusively nonorally fed
Complete pre-operative and post-operative feeding assessment data sets	Had not undergone the post-operative assessment

Abbreviations: BWS, Beckwith Wiedemann syndrome; SLT, speech and language therapist; TRS, tongue reduction surgery.
[a]Pre-op 1 indicates first pre-operative assessment.

Table 2. Median Age at Each Assessment.

Pre-operative 1 = 8.0 months (IQR = 5.5-10.0 months)	Pre-operative 2 = 11.0 months (IQR = 10.0-13.0 months)	Post-operative 1 = 19.0 months (IQR = 17-22 months)

Abbreviation: IQR, interquartile range.

The study cohort was 52% male. All children were British with ethnicities including Caucasian (n = 22), Asian (n = 2), and African (n = 1). Median age at surgery was 16.0 months (interquartile range = 14.0-18.5 months). Median ages at each data point are shown in Table 2. Five (20%) infants were premature, all of whom were born within the late preterm category of 34 to 36 weeks (World Health Organization, 2018). All the children presented with age-appropriate development and were considered typically representative of the population of m-BWS.

Intervention

Keyhole reduction surgery was undertaken by one of 2 surgeons. One surgeon conducted 76% (n = 19) of the surgeries.

Feeding Assessment

Depending on their age and progress with weaning, each infant was given three food consistencies (liquids, purées, and solids) by the parent. All children were fed in their usual seating position. The assessment was audio-video recorded.

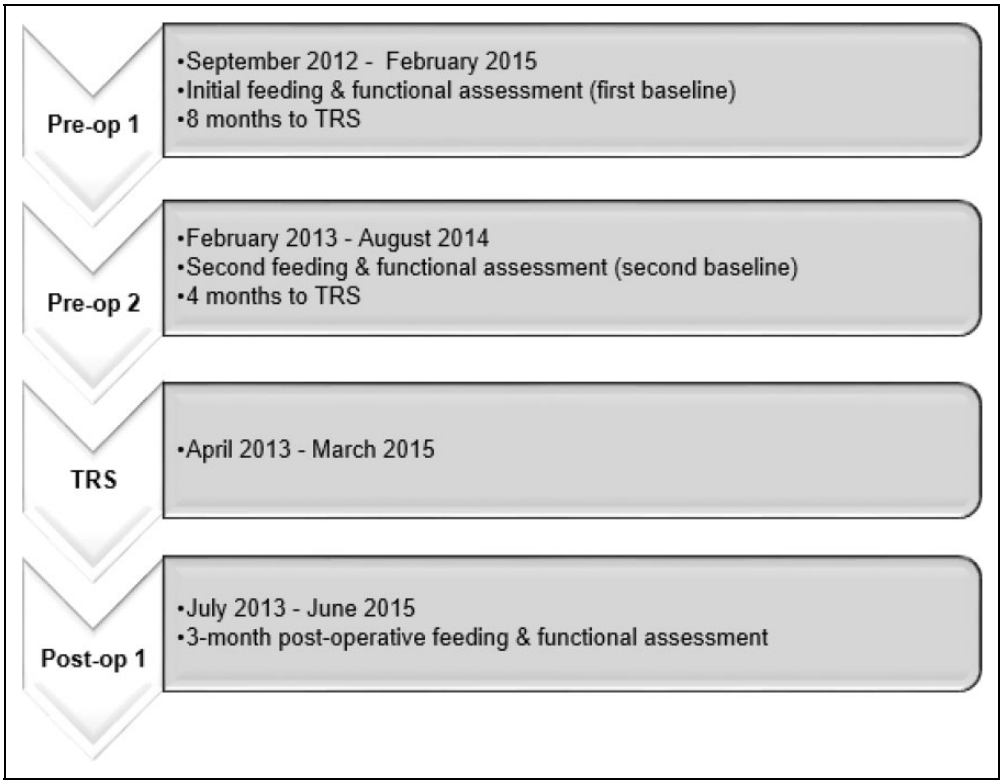

Figure 3. Time line from pre-operative to post-operative SLT feeding assessment. SLT indicates speech and language therapist.

Repeated feeding measures were taken at two pre-operative feeding assessments (pre-operative 1 and pre-operative 2) and one 3-month post-operative assessment (post-operative 1). The infants served as their own controls. Figure 3 depicts the time line of feeding assessments undertaken by the SLT.

Outcome Measure—Rating Scale

Once a complete pre- and post-operative data set was available for each patient, the investigator completed the rating scale for each of the 3 data points based on the descriptive SLT clinical notes/reports and video recordings of feeding assessments. The item "prolonged feed times" was subjectively measured based on parental report and observation of the duration of a bottle-feed and food intake which was qualified by parental report of this being representative of feeding times at home.

Main Outcome Measures

Oral and selected pharyngeal stage feeding behaviors following TRS were the main outcome measures.

Statistical Methods

Quantitative data were generated from the rating scale. The data were coded according to the ordinal scale values for frequency of occurrence (0-3) and the nominal values for dichotomous items (1, 0). The data were entered into SPSS statistics (version 22), anonymized, and analyzed (IBM Corp, 2013). Descriptive statistics were used to profile the group by examining the occurrence of each feeding behavior over time and to characterize those who presented with aspiration risk and required diet modification. Mean groupwise consistency scores over time of the ordinal items were generated. Given the small sample size, the predominantly ordinal data and the lack of variance post-operatively, a number of non-parametric assumptions were conformed to, making the Friedman analysis of variance (ANOVA) test of proportions and pairwise comparisons appropriate to test for differences (mean rank) over the 3 data points (2 tailed). The ANOVA was conducted on the mean scores for the ordinal items within each subscale. Post hoc analyses were conducted using Wilcoxon sign ranked test to ascertain between which data points the differences existed. A Bonferroni correction for the 3 comparison tests was applied; therefore, all effects were reported at a 0.01 ($0.05/3 = 0.0167$) level of significance (Field, 2009).

Results

The results are reported according to each of the study's aims.

(i) Pre-operative Presentation

Pre-operatively, all infants presented with a number of difficulties with liquids, purées, and solids with varying frequency and severity. For the purposes of this report, those characteristics which occurred most frequently (ie, most of the time or constantly) are reported. The 5 premature infants displayed

Table 3. Results Overview From the Pre-operative and 3-Month Post-operative Data Points.[a]

	Pre-operative 2			Post-operative 1		
Feeding Difficulty	Liquids	Purées	Solids	Liquids	Purées	Solids
Tongue protrusion under teat of bottle/while eating	100%	91%	92%	0%	0%	0%
Biting	-	-	78%	-	-	12%
Difficulty with lip seal	91%	82.5%	91%	0%	0%	8%
Tongue lateralization	-	100%	92%	-	0%	16%
Bolus manipulation	-	78%	96%	-	0%	12%
Risk of aspiration	74%	78%	87%	4% (n = 1)	8%	0%
Cough	9%	4%	13%	0%	4%	0%
Anterior loss	35%	56%	35%	0%	0%	4%
Modified diet	17%	91%	100%	4%	0%	8%
Prolonged feeding times	9%	48%	63%	0%	0%	8%
Required VFSS	8%	17%	9%	4%	0%	0%

Abbreviation: VFSS, videofluoroscopy swallow study.
[a]Behaviors occurring most of the time or constantly.

feeding patterns similar to the rest of the group. None of the infants had required hypoglycemia management beyond the initial few days of life; therefore, this was not an influencing factor in their feeding profile.

Table 3 provides an overview of those characteristics which caused difficulties from the 43 feeding behaviors in the scale. There were no statistically significant differences between the two pre-operative data points for any consistency on a Wilcoxon signed rank test: liquids $Z = -.307$ $P > .01$; purées $Z = -.525$ $P > .0167$; solids $Z = -.221$ $P > .0167$. For this reason, only data collected at the second pre-operative data point will be discussed, henceforth called the pre-operative data point.

Liquids. Pre-operatively all children presented with constant tongue protrusion under the teat. Ninety-one percent had difficulty achieving a lip seal around the teat and 24% had difficulty achieving a rhythmic sucking pattern. Anterior loss of liquids was noted in 35% of the group. Seventy-four percent presented with aspiration risk requiring compensatory feeding techniques. A modified diet was introduced for 17% to manage the aspiration risk and 8% required VFSS to instrumentally assess their swallow safety (Table 3).

Purées. Thirty-nine percent struggled to remove a bolus from a spoon; 82.5% had difficulty achieving a competent lip seal; 91% had tongue protrusion while eating; 100% had tongue lateralization problems; 78% and 87% had bolus manipulation and bolus transfer difficulties respectively. Fifty-five percent had anterior bolus loss 78% presented with aspiration risk, 91% required diet modification, and 17% required VFSS (Table 3).

Solids. Complete data were available for 14 patients due to the difficulties in progressing to solids. Difficulty occurred most of the time or constantly with the majority of behaviors for solids. Seventy-eight percent had difficulty biting solids; lip incompetence was noted in 91%; 96% had bolus manipulation difficulty. Ninety-two percent had difficulty with tongue lateralization. Coughing on solids was noted in 13%; 87% presented with aspiration risk, and all of the children required texture modification (Table 3).

(ii) Post-operative Presentation

Resolution or reduction of oral preparatory and oral stage feeding difficulties was the pervasive finding 3 months after TRS. Table 3 demonstrates changes over time for a range of feeding behaviors for each consistency showing the significant improvement in feeding skills.

The Friedman ANOVA test was carried out for each consistency which demonstrated a statistically significant difference across the data points: liquids (n = 23), χ^2 (2) = 36.575, $P < .01$; purées (n = 20), χ^2 (2) = 31.692, $P < .001$; solids (n = 14), χ^2 (2) = 21.143, $P < .001$. The Wilcoxon signed rank test (2 tailed) pinpointed a statistically significant difference between the pre-operative and post-operative data points for each consistency: liquids $Z = -4.205$; $P < .01$; purées $Z = -3.933$, $P < .0167$; solids $Z = -3.29$; $P < .0167$. Improvement in individual feeding behaviors associated with each consistency was found post-operatively. Tongue protrusion was eliminated for all 3 consistencies. Apart from one infant who occasionally coughed on purées and a second who presented with an arrhythmic suck and occasional cough due to a history of reflux and isolated oropharyngeal dysphagia, no other issues with liquids or purées were found.

Across the 3 consistency domains, most residual feeding difficulties affected solids (Table 3). Eight percent of the whole group had occasional difficulty with biting. Twelve percent had occasional difficulty with bolus manipulation of solids and 16% had occasional difficulty with lateral tongue movements. Eight percent continued to require minor diet modification to manage harder solids due to reduced lateral tongue movements, bolus manipulation and transfer. Coughing and aspiration risk on solids were both eliminated post-operatively. The majority of the group had efficient post-operative feeding skills, allowing them to eat age-appropriate, typical family meals and drink effectively.

(iii) Feeding Management

Seventy-four percent of the group presented with aspiration risk on liquids pre-operatively. This was managed in most infants using compensatory techniques including pacing, semi-upright positioning, and reduced flow rate of teat. Pre-operatively, about one-fifth required texture modification for liquids, 91% required this for purées, while all required this for solids, thereby reducing the effects of tongue protrusion, poor lip seal, and limited tongue lateralization. These modifications enabled the infants to safely meet their nutritional requirements by having a smoother, manageable consistency.

(iv) Piloting a Novel Feeding Rating Scale

Investigation of the internal consistency of the items within each consistency scale was conducted using Cronbach α. A Cronbach α level of between .70 and .95 was accepted (De Vellis, 2012). Cronbach α was calculated for item-total correlations and *if item deleted* to ascertain the homogeneity of the scale (Streiner and Norman, 2008). Items are considered to correlate well with the total α score if they are above .30 (Kline, 1999; De Vaus, 2002). At both pre-operative data points, the overall α for all consistencies was strong (liquids pre-op 1 $\alpha = .785$, pre-op 2 $\alpha = .756$; purées pre-op 1 $\alpha = .738$, pre-op 2 $\alpha = .754$; solids pre-op 1 $\alpha = .854$, pre-op 2 $\alpha = .801$). This shows good representation of the underlying construct of feeding difficulty per bolus consistency. Post-operatively, it was not possible to calculate Cronbach α due to low variance in the data.

Discussion

This investigation aimed to describe the feeding skills in infants with m-BWS prior to TRS, to examine the changes that occurred post-operatively, to describe feeding management strategies, and to pilot the novel scale. In doing so, this study has provided a detailed profile of the feeding difficulties that can present in the first year of life, not previously described for this condition.

The results demonstrated that the majority of feeding issues associated with m-BWS in infancy were related to the oral preparatory and oral stages of feeding. These included difficulties with lip seal, achieving a rhythmic suck, removing purées from a spoon, bolus manipulation, tongue lateralization and anterior loss. When the specific feeding skill of tongue lateralization was impacted, chewing skills which should subsequently emerge were affected, limiting the range of solids in the diet. The combination of difficulties meant that the group was unable to progress along the normal feeding hierarchy to firmer textures and had difficulty with controlling liquids, resulting in aspiration risk. This highlights the importance of a detailed clinical assessment in identification of the specific feeding difficulties to enable prompt, appropriate management.

Post-operatively, these issues either resolved or significantly reduced, allowing the group to eat age-appropriate, typical family meals and drink effectively. Residual difficulties were occasionally evident at the post-operative assessment. These related mostly to the biting and chewing of solids, usually occurring infrequently without greatly impacting the infants' ability to progress with this texture.

Strengths of the Study

This study was characterized by the use of a consistent assessment protocol in a consecutive series of infants in a narrow age range. It is a relatively large sample in a low-incidence condition, compared with previous studies. Efforts were made to control for sample size, power, and the evaluation of missing data in order to undertake meaningful analysis. The double baseline enabled examination of the stability of behaviors pre-operatively and confirmed minimal spontaneous change during this period, thereby giving confidence to changes being attributable to the surgery.

Studying infants under the age of 12 months at initial assessment allowed examination of early feeding skills as well as weaning and transition to family mealtimes (Arvedson and Brodsky, 2002). This was critical to addressing the clinical question of feeding skills in infancy because of clinical reports and parents' concern during this period. The upper age limit of 24 months at the post-operative data point introduced an element of homogeneity within the group. This aimed to align with the maturation of rotary jaw motion (Wilson and Green, 2009), one of the defining features of advanced eating skill.

The feeding rating scale was developed to create a consistent method of documenting and reporting feeding skills in this population. This study has evaluated the appropriateness of the items to inform possible development of a feeding assessment tool in the future for this population. Initial steps were taken to ascertain the validity of the subscales by examining internal consistency. A good item–total correlation and high Cronbach α demonstrated good representation of the underlying construct of feeding difficulty per bolus consistency. These features contribute to the face and content validity of the scale. The scale has the potential to be used with other populations including children with oral malformations including hemangiomas or other oral abnormalities, oral–facial–digital syndrome, facial or tongue palsy, as well as children with trisomy 21 who present with tongue thrust, all of whom can exhibit tongue-related feeding difficulties which may/may not occur independently of a pharyngeal stage swallowing dysfunction.

Limitations

The retrospective design of this service evaluation is known to be associated with bias. A clear limitation of this study is the use of a novel rating scale which did not have a comparison group, such as an age-matched group of children who were unoperated or with developmental feeding norms. It is acknowledged that this would be critical in a more detailed study on tool development and long-term outcome analysis. Furthermore, strictly the findings cannot be generalized to the wider population of m-BWS due to the non-experimental nature of a service evaluation

(Hess, 2004; Trochim, 2005). The limitations of the impairment-based-only approach to reporting outcomes are also acknowledged. While the item regarding duration of feeding attempted to capture how feeding difficulties affected the infant and family's quality of life, more rigorous methods to fully describe parental experiences are required to reflect the impact of feeding difficulties upon family mealtimes and interactions (World Health Organization, 2002).

Future Research

Analysis of this group one year post-operatively would help differentiate between maturational change and true persisting oral stage feeding deficits. It is particularly important that the residual, albeit mild, difficulties are profiled over a longer time period, given that self-reported eating difficulties by adults have been described (Tomlinson et al., 2007). Further psychometric testing of the scale is recommended to examine test–retest stability and inter- and intra-rater reliability. Normative data would give criterion validity to the tool allowing for known-group validation. One-to-one interviews or focus groups could be undertaken to examine the impact on quality of life for the child and family.

Conclusion

This study provides the first systematic profile of feeding examining different textures in a sample of 25 infants with m-BWS. The majority of feeding difficulties in this population arose due to the impact of the enlarged, protruding tongue on important lingual movements which are typically involved in cohesive bolus formation and food propulsion to the back of the oral cavity while maintaining a lip seal to minimize anterior bolus loss. In the group, these functions were negatively affected due to the abnormal structure and position of the tongue during feeding. Over three-quarters of the group presented with aspiration risk, which in all cases was managed with compensatory feeding techniques or texture adjustment. Post-operatively, almost all infants in this study had typical drinking skills, could successfully manage purées, and were eating age-appropriate solids, similar to their peers.

This study provides evidence for feeding-related respiratory problems if left unmanaged. It is recommended that infants born with this condition are referred to a dysphagia specialist SLT as soon as possible following diagnosis, to optimize feeding and to minimize the risk of aspiration. This early intervention could reduce hospital admissions for feeding-related respiratory issues. It is anticipated that this new knowledge will inform pediatricians and neonatologists who typically oversee the care of these infants. Information from this study could be used to inform parent counseling.

The feeding rating scale is of use to specialist and nonspecialist SLTs working with this population, offering a structured approach to assessment, record-keeping, and report-writing. This systematic profiling allows meaningful data sets to be assembled and compared, addressing clinical uncertainties through research in the future.

Authors' Note

This study was presented at the International Cleft Congress, Chennai, India, February 2017; at the UK Swallowing Research Conference, February 2016, London; at the Dysphagia CEN, London, June 2017; and at the Cleft Lip and Palate and Craniofacial Anomalies CEN, Birmingham, November 2017.

Acknowledgments

The authors acknowledge the expertise of Caroleen Shipster, Specialist Speech and Language Therapist, Clinical Lead for the Beckwith Wiedemann Syndrome Macrolgossia Service, Great Ormond Street Hospital, London; Dr Gill Craig, Senior Lecturer in Disabilities & Social Inclusion, School of Health Sciences, City University London for contributing to the original MRes dissertation and Dr Shashi Hirani, Senior Lecturer, School of Health Sciences, City University London for contributing specifically to the statistical analysis of the original MRes dissertation manuscript.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

References

Abeleira MT, Seoane-Romero J, Outumuro M, Caamano F, Suárez D, Carmona I. A multidisciplinary approach to the treatment of oral manifestations associated with Beckwith Wiedemann syndrome: a long-term case report. *J Am Dent Assoc*. 2012;3(2):59-66.

Arvedson JC. Assessment of pediatric dysphagia and feeding disorders: clinical and instrumental approaches. *Dev Disabil Res Rev*. 2008;14(2):118-127.

Arvedson J, Brodsky L. *Paediatric Swallowing and Feeding. Assessment and Management*. Toronto, Ontario, Canada: Singular Publishing Group; 2002.

Benfer K, Weir K, Bell K, Ware K, Davies P, Boyd R. Validity and reproducibility of measures of oropharyngeal dysphagia in preschool children with cerebral palsy. *Dev Med Child Neurol*. 2015;57(4):358-365.

Bowling A. *Research Methods in Health. Investigation Health and Health Services*. 4th ed. Maidenhead, England: Open University Press; 2014.

Chau H, Soma M, Massey S, Hewitt R, Hartley B. Anterior tongue reduction surgery for paediatric macroglossia. *J Laryngol Otol*. 2011;125(12):1247-1250.

De Vaus D. *Surveys in Social Research*. 5th ed. London, England: Routledge; 2002.

De Vellis R. *Scale Development: Theory and Applications*. 3rd ed. London, England: Sage; 2012.

Delaney AL, Arvedson JC. Development of swallowing and feeding: prenatal through first year of life. *Dev Disabil Res Rev*. 2008;14(2):105-117.

Elliot M, Bayly R, Cole T, Temple IK, Maher ER. Clinical features and natural history of Beckwith Wiedemann syndrome: presentation of 74 new cases. *Clin Genet*. 1994;46(2):168-174.

Engstrom W, Lindham S, Schofield P. The Beckwith Wiedemann syndrome. *Eur J Pediatr*. 1988;147(5):450-457.

Field A. *Discovering Statistics Using SPSS*. 3rd ed. London, England: Sage; 2009.

Heggie A, Vujcich J, Portnof J, Morgan A. Tongue reduction for macroglossia in Beckwith Wiedemann syndrome: a review and application of a new technique. *Int J Oral Maxillofac Surg*. 2013;42(2):185-191.

Hess D. Retrospective studies and chart reviews. *Resp Care*. 2004; 49(10):1171-1174.

IBM Corp. *IBM SPSS Statistics for Windows, Version 22.0*. Armonk, NY: IBM Corp; 2013.

Kadouch D, Maas S, Dubois L, van der Horst C. Surgical treatment of macroglossia in patients with Beckwith Wiedemann syndrome: a 20-year experience and review of the literature. *Int J Oral Maxillofac Surg*. 2012;41(3):300-308.

Kamide A, Hashimoto K, Miyamura K, Honda M. Assessment of feeding and swallowing in children: validity and reliability of the Ability for Basic Feeding and Swallowing Scale for Children (ABFS-C). *Brain Dev*. 2015;37(5):508-514.

Kline P. *The Handbook of Psychological Testing*. 2nd ed. London, England: Routledge; 1999.

Martinez y Martinez R. Clinical features in the Wiedemann Beckwith syndrome. *Clin Genet*. 1996;50(4):272-274.

Morgan A, O Mahoney R, Francis H. The use of pulse oximetry as a screening assessment for paediatric neurogenic dysphagia. *Dev Neurorehabil*. 2008;11(1):25-38.

Prada C, Zarate Y, Hopkin R. Genetic causes of macroglossia: diagnostic approach. *Pediatrics*. 2012;129(2):431-437.

Remijn L, Speyer R, Groen B, van Limbeek J, Nijhuis-van der Sanden M. Validity and reliability of the Mastication Observation and Evaluation (MOE) instrument. *Res Dev Disabil*. 2014;35(7): 1551-1561.

Robbins J, Gangnon RE, Theis SM, Kays SA, Hewitt AL, Hind JA. The effects of lingual exercise on swallowing in older adults. *J Am Geriatrics Society*. 2005;53(9):1483-1489.

Royal College of Speech and Language Therapists. *RCSLT Resource Manual for Commissioning and Planning Services for Speech, Language, Communication Needs—Dysphagia*. 2009. https://www.rcslt.org/speech_and_language_therapy/commissioning/dysphagia_manual_072014. Updated July 14, 2014. Accessed April 10, 2018.

Sellers D, Mandy A, Pennington L, Hankins M, Morris C. Development and reliability of a system to classify eating and drinking ability of people with cerebral palsy. *Dev Med Child Neurol*. 2013;56(3):245-251.

Shipster C, Morgan A, Dunaway D. Psychosocial, feeding, and drooling outcomes in children with Beckwith Wiedemann syndrome following tongue reduction surgery. *Cleft Palate Craniofac J*. 2012;49(4):25-34.

Shipster C, Oliver B, Morgan A. Speech and oral motor skills in children with Beckwith Wiedemann Syndrome: pre-and post-tongue reduction surgery. *Adv Speech Lang Pathol*. 2006;8(1):45-55.

Skuse D, Stevenson J, Reilly S, Mathisen B. Schedule for Oral-Motor Assessment (SOMA): methods of validation. *Dysphagia*. 1995; 10(3):192-202.

Smith Hammond C, Goldstein L. Cough and aspiration of food and liquids due to oral-pharyngeal dysphagia: ACCP evidence-based clinical practice guidelines. *Chest*. 2006;129(1):154-168.

Stierwalt J, Youmans S. Tongue measures in individuals with normal and impaired swallowing. *Am J Speech Lang Pathol*. 2007;16(2): 148-156.

St Pierre A, Khattra P, Johnson M, Cender L, Manzano S, Holsti L. Content validation of the infant malnutrition and feeding checklist for congenital heart disease: a tool to identify risk of malnutrition and feeding difficulties in infants with congenital heart disease. *J Pediatr Nurs*. 2010;25(5):367-374.

Stratton M. Behavioural assessment scale of oral functions in feeding. *Am J Occup Ther*. 1981;35(11):719-721.

Streiner D, Norman G. *Health Measurement Scales. A Practical Guide to Their Development and Use*. 4th ed. Oxford, England: University Press; 2008.

Thoyre S, Pados B, Park J, Estrem H, Hodges E, Mc Comish C, Van Riper M, Murdoch K. Development and content validation of the Pediatric Eating Assessment Tool (Pedi-EAT). *Am J Speech Lang Pathol*. 2014;23(1):46-59.

Thoyre SM, Shaker CS, Prindham KF. The early feeding skills assessment for preterm infants. *Neonatal Netw*. 2005;24(3):7-16.

Tomlinson J, Morse S, Bernard S, Greensmith A, Meara J. Long-term outcomes of surgical tongue reduction in Beckwith Wiedemann syndrome. *Plast Reconstr Surg*. 2007;119(3):992-1002.

Trochim W. *Research Methods: The Concise Knowledge Base*. Mason, OH: Thomson; 2005.

van den Engel-Hoek L, van Hulst KC, van Gerven MH, van Haaften L, de Groot SA. Development of oral motor behavior related to the skill assisted spoon feeding. *Infant Behav Dev*. 2014;37(2): 187-191.

Weksberg R, Shuman C, Beckwith JB. Beckwith Wiedemann syndrome. *Eur J Hum Genet*. 2010;18(1):8-14.

Weng EY, Moeschler JB, Graham JM Jr. Longitudinal observations on 15 children with Wiedemann-Beckwith syndrome. *Am J Med Genet*. 1995;56(4):366-373.

Wilson E, Green J. The development of jaw motion for mastication. *Early Hum Dev*. 2009;85(5):303-311.

Wolf L, Glass R. *Feeding and Swallowing Disorders in Infancy. Assessment and Management*. Austin, TX: Hammill Institute on Disabilities; 1992.

Wolford L, Cottrell D. Diagnosis of macroglossia and indications for reduction glossectomy. *Am J Orthod Dentofacial Orthop*. 1996; 110(2):170-177.

World Health Organization. *Towards a Common Language for Functioning, Disability and Health*. Geneva, Switzerland: ICF; 2002. Available at: www.who.int/classifications/icf/training/icfbeginnersguide.pdf. Accessed June 01, 2018.

World Health Organization. Preterm birth. 2018. Updated February 19, 2018. Available at: http://www.who.int/news-room/fact-sheets/detail/preterm-birth. Accessed June 01, 2018.